Usher syndrome: A phenomenological study of adults across the lifespan living in England

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Abstract

Usher syndrome is a rare inherited genetic condition which is one of the main causes of acquired deafblindness in the United Kingdom (UK). Although the condition is not life threatening, it is life altering and will have a significant impact on the lives of not only the person diagnosed with the condition, but also their families, friendship groups and new and existing relationships.

The aim of the study was to develop an understanding of the experiences of diagnosis of and living with Usher syndrome, from the perspective of adults living in England. Specific objectives of the study were to explore the experience of being diagnosed with Usher syndrome; explore the transition from adolescence to adulthood for people who have Usher syndrome; to develop an understanding of the experience of living with Usher syndrome, including support, developmental opportunities and the role of the Deaf community; to disseminate original findings; inform future practice, service development, policy and education and make recommendations for further research relating to the experience of living with Usher syndrome.

To address these aims and objectives, this qualitative, descriptive phenomenological study, conducted interviews with 20 males and females aged between 18-82 years from a variety of demographic locations. To contribute to the trustworthiness of the study, I developed a methodological innovation called 'Multiple Sensory Communication and Interview Methods' (MSCIM) which ensured that as far as possible communication and interview methods were participant led. Three overarching messages from findings were revealed: the importance of ensuring communication is timely, supportive and appropriate; Usher support at the

right time: providing physical and virtual support networks and essentiality of Usher awareness: raising the profile.

This study is unique because it is the first qualitative, descriptive phenomenological study to demonstrate new knowledge to better understand and support people living with Usher in England.

Keywords

Usher syndrome, sensory impairment, D/deafblindness, 'Multiple Sensory Communication and Interview Methods' (MSCIM), social work, qualitative research, descriptive phenomenology

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Chapter 1 Introduction and background

1.1 Introduction

Usher syndrome is a rare inherited genetic condition which is one of the main causes of acquired deafblindness in the United Kingdom (UK) and approximately 3 - 6% of people born *D*eaf [15] will develop it (Genetic Alliance UK, 2012). Persons are regarded as D/deafblind if they have a degree of combined visual and auditory impairment resulting in problems of communication, information and mobility (Deafblind Services Liaison Group, 1993). Although there are general definitions for people who experience D/deafness and D/deafblindness, each individual's experience will be unique to them.

Capital 'D' Deafness refers to a person who considers themselves to be culturally Deaf. The person would most likely be a profoundly Deaf person who would not consider themselves to be disabled, but rather part of a minority group with its own culture, history and linguistics (British Deaf Association, 2007). Deaf people consider being Deaf as part of their heritage and most likely consider themselves proud to be Deaf (Evans and Whittaker, 2010). Deafblind with a capital 'D' refers to people who consider themselves to be culturally Deaf but also experience sight loss.

Small 'd' deafness [16] refers to a person who was likely to have been previously a hearing person or one who was born deaf, growing up in a hearing family with oral communication being the primary means of communication and they would most likely consider deafness to be a disability. People, who are deaf and then experience sight loss, are referred to as deafblind and again will most likely consider their sensory loss to be a disability.

Although Usher syndrome is not life threatening, it is life altering and will have a significant impact on the lives of not only the person diagnosed with the condition but also their families/friendship groups and new and existing relationships. However, the experiences of people who are living with Usher syndrome have rarely been studied to date. This study is a methodological innovation as it is the first phenomenological study to explore what life is like for adults across the lifespan, living with Usher in England.

This first chapter will introduce the doctoral thesis and provide an explanation of Usher syndrome, as well as exploring the rationale and interest behind the study. Disability in the wider context and disability theory pertinent to deafblindness/Usher and D/deafness will also be considered. Guidance and legislative contexts will be discussed and the structure of the doctoral thesis will be provided. The chapter summary will briefly discuss the chapters that will follow. In this thesis the terms Usher syndrome and Usher will be used interchangeably although referring to the same condition.

1.2 Explanation of Usher syndrome

Usher syndrome was first identified by German born ophthalmologist Friedrich Wilhelm Ernst Albrecht von Gräfe also known as von Graefe in 1858 when he discovered the connection between D/deafness caused by an impairment of the auditory nerve which prevents sensory transmission to the brain (sensorineural hearing loss - hearing loss caused by problems with the inner ear) and retinitis pigmentosa (retinal cell degeneration), which can lead to progressive loss of central and peripheral vision (Millan *et al,* 2011). Later, Charles Usher, a British ophthalmologist, discovered the “familial occurrence” (Hope *et al,* 1997 p46) or

familial genetic connection. There are three types of Usher syndrome, type I; II and III (see Table 1.1).

Table 1.1: Usher type and description

Type I	People who experience Usher Syndrome type I are born profoundly d/Deaf and start to experience reduced vision in their young years possibly the first decade. Type I children sometimes experience problems with their balance, have difficulty walking as a child which is often associated with difficulties in the child's vestibular system (Pennings, *et al,* 2003).
Type II	People who experience type II have reduced hearing but are 'not' born d/Deaf. Whilst hearing is affected there does not seem to be obvious balance difficulties as experienced by people with type I. It is not until later that people with Usher syndrome type II start to experience difficulties with their vision. This loss of vision may occur in the second decade or it may be as they get older. It can be very different for different people (Sadeghi *et al,* 2004).
Type III	People who experience type III may not realise they even have it until later in life, the reason being that people with type III do not have any obvious hearing, sight or balance difficulties initially, but rather the losses occur gradually. It may even be that if Usher syndrome is experienced as people get more advanced in years they may expect their hearing and sight to decrease as a natural progression of advanced age, thus not realising that instead of acquired deafblindness they have actually experienced Usher syndrome (Moller *et al,* 2009).

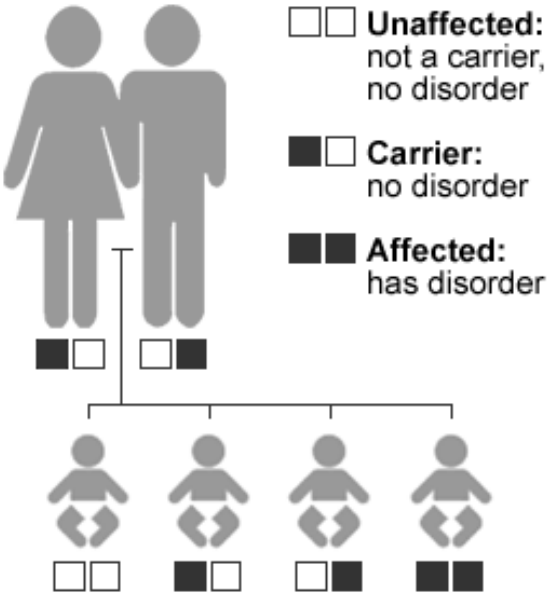
Although, the majority of people who are deafblind are older (Sense, 2002, Kyle and Barnett, 2012) the age range of people with Usher syndrome varies from early years to older age.

One of the characteristics of Usher syndrome is nightblindness or nyctalopia which is a condition where people cannot see well at night or in dark conditions. Often people will experience nightblindness before being diagnosed with Usher syndrome (Sense, 2014b). Usher syndrome is largely an unseen disability as there are no outward visible signs of disability (Sense, 2014b). However, the condition is degenerative and could have a significant effect on not only the life of the person with the condition, but also the lives of any relationships that are formed or children who are born.

Usher syndrome is detected when a person notices deterioration in their vision or, in the case of children and young people, parents/care givers may notice this, and advice is sought to find out the cause. Usher syndrome is diagnosed in various ways; for some people, opticians will diagnose the condition while for others it may be a specialist eye consultant/ophthalmologist [5] or an audiologist [4]. Whilst Usher syndrome is manifested by various hearing loss levels or D/deafness from birth, this hereditary/genetic (passed from parents to their children through genes) condition intensifies to additionally affect sight. At present there is no treatment or cure. The combination of hearing and sight loss is caused by gene mutation (changes in gene structure) and whilst research has been conducted to map mutations which cause Usher, and advanced understanding of genetic causes of Usher have been obtained (Yoshimura *et al,* 2015), this does not provide everyday practical assistance for people who experience Usher or their families (Cohen *et al*, 2007).

Usher syndrome is a recessive disorder which means that a person must inherit a change in the same gene from each of their parents for them to have Usher; if both parents are carriers there is a one in four likelihood (Figure 1.1) that the child will have Usher (National Institute on Deafness and other Communication Disorders, 2016). If a child has one Usher gene then the prediction is that they will not have Usher (however this can vary).

Figure 1.1: Likelihood of child being affected



When a parent is advised that their child has Usher they will often be totally shocked and express that the condition cannot be genetic as “it is not in the family” (Wallber, 2009 p5). However, the parents will often be asymptomatic which means the parents will not have any symptoms and be unaware that they are both carriers of the same gene.

The hearing loss is always sensorineural and the sight loss is always progressive (Wallber, 2009). The sight loss is caused by a condition called retinitis pigmentosa, otherwise known as RP. RP is a degenerative retinal disease with the primary symptoms being nightblindness and loss of peripheral vision, which affects people who are initially sighted, but as a result of RP they gradually lose their vision (Wallber, 2009). Whilst organisations such as RP Fighting Blindness, which is a UK charity that supports people with retinitis pigmentosa and other retinal dystrophies (RP Fighting Blindness, 2016) and innovates and supports medical research to find a treatment or cure for this inherited condition, at present there are no proven treatments to decelerate or stop the condition from deteriorating.

Whilst there is a diagnosis for Usher syndrome, there is no way of predicting the prognosis. Usher is unpredictable because a person could become blind very quickly or it could be that they can still see to read at 50 years. As highlighted earlier there are three clinical types of Usher syndrome (see Table 1.1) with general definitions for each type, however, each individual's experience will be unique to them.

1.3 Rationale and interest

I had professional experience of a variety of sensory roles: technical officer with D/deaf people; Care Manager with D/deafblind adults; social worker with children who are D/deaf and social worker with children with disabilities. The global definition of social work is defined by the International Federation of Social workers (IFSW) as:

> '*A practice based profession and an academic discipline that promotes social change and development, social cohesion and the empowerment and liberation of people. Principles of social justice, human rights, collective responsibility and respect for diversities are central to social work*' (IFSW, 2014).

In the UK, a social worker is required to be registered and is regulated by the Health Care and Professions Council (HCPC, 2012). Whilst working within this field I became aware that provision of services and support for people with sensory needs varied from area to area. In some areas there were specialised services with specially trained sensory workers (visual impairment rehabilitation workers, care managers with deafblind/d/Deaf people) and in others this provision was far more limited. It was a bit of a 'post code lottery' (a situation where a person's access to health services/medical treatment is decided by the area they live in).

When working with people who experience sensory need it became apparent that every person's sensory requirements are unique and individual to them and therefore the services and support they require are unique too. However, without specialist service provision from a service that understands need, it could be argued that specialist; individual, tailor-made services would not be viable. Additionally, where people live in more remote areas, it may be they are quite simply not aware of the services that are available. Furthermore, the shock of being diagnosed with Usher syndrome may result in a person either being in denial and not wanting to access services or not knowing where to begin.

Initially, my interest concerned people who experienced d/Deafness. However after a literature search and review it became apparent that this topic had already received significant and ongoing attention (Young and Hunt, 2009, Young *et al*, 2010 and Young, 2011). As my doctoral objective was to conduct research into an area of sensory need that would make a difference for people who experienced sensory needs/requirements, the chosen research area was reconsidered and other sensory related options explored. I established contacts with organisations for deafblind, sight impaired and D/deaf people to explore what areas of sensory need were currently under-researched or did not receive much attention.

When this thesis was in its infancy, informal discussion with Sense [28] a specialist charity (the terms charity and organisation will be used inter-changeably throughout this thesis) that campaigns to improve services and support for people who experience deafblindness (Sense, 2012) highlighted that whilst all areas of deafblindness were under-researched, Usher syndrome in particular received little attention (Damen *et al,* 2005, Kyle and Barnett, 2012, Ellis and Hodges, 2013), because it is a rare condition with relatively few of the population being affected by it. As the condition was under-researched, it was an area in which organisations

such as Sense and the Royal National institute for Blind People (RNIB) would like wider research participation. Additional discussion with Sense revealed that there was also an interest to support access to research participants; the result was that Sense became primary gatekeepers for the study.

Whilst there had been research conducted from a medical perspective (Pennings *et al,* 2003, Sadeghi *et al,* 2004 Moller *et al,* 2009, Yoshimura *et al,* 2015,) few studies have considered the life experiences of people with Usher (Ellis and Hodges, 2013, Damen *et al,* 2005, Côté *et al,* 2013, Wahlqvist *et al*, 2013, Högner, 2015) and even fewer included participants from the UK (Damen *et al*, 2005, Kyle and Barnett, 2012, Ellis and Hodges, 2013), further highlighting how little research there is, not only with regard to deafblindness (Simcock and Manthorpe, 2014, Simcock, 2016), but especially Usher syndrome (Ellis and Hodges, 2013).

Whilst I had worked with some people who had Usher, their condition had been referred to as deafblindness and I was not aware of the full implications of what experiencing Usher syndrome really meant for individuals. After reading in-depth about the specifics of Usher syndrome, I realised the full extent of the condition and its grave significance. This knowledge about Usher syndrome was essential when conducting the research, especially when realising how unique each participant's experience would be and the essentiality to choose a methodology that reflected this uniqueness.

As Usher syndrome is a rare condition, there is often little public awareness of it, furthermore, "access to this population [people experiencing Usher syndrome] is difficult" (Högner, 2015 p187). As the research for this thesis progressed, I developed techniques to empower people with Usher syndrome as research participants and I published an article entitled 'Empowering people with Usher

syndrome as participants in research' in the British Journal of Social Work (Appendix A).

As developing "a rigorous grounding" in theories and models are essential for social workers to guide their practice (Parker, 2017 p2), theoretical frameworks pertaining to disability were a significant aspect of underpinning this study, to make the most of the usefulness of the research and enable a better understanding of Usher, disability in the wider context and disability research. Therefore, the medical, social and individual models of disability, as well as identity development theory will be considered in the next section. Also to be discussed in the context of disability theory is Deaf culture and where it is positioned. However, to understand where D/deafness and deafblindness/Usher syndrome were positioned in disability theory, firstly terminology used when referring to people who experience disability will be considered.

1.4 Disability theory

1.4.1 Terminology

Terminology used when referring to people who experience disability influences public perception (Aiden and McCarthy, 2014) and therefore needs to be considered when conducting research involving people who experience disability. The Equality Act 2010 definition is that a person is disabled if they have a physical or mental impairment that has a substantial and long-term negative effect on their ability to do normal daily activities.

A report commissioned by Scope (Aiden and McCarthy, 2014), a charity who support people with disabilities to have the same opportunities as people who do not have disabilities, considered current attitudes towards people with disabilities. They suggested that attitudes are a multifaceted collection of beliefs, feelings and values

which affect our perceptions about certain people or situations. They reported that 20 years ago they changed their name from the Spastic Society as they recognised how language had an impact on public perception of people with disabilities. They acknowledged that a lot of uneasiness people feel about disability, may culminate from a lack of understanding and suggested that nearly half of the British public do not know anyone who has a disability. The report further highlighted that people with disabilities are more likely than people who do not have a disability to experience the attitudes of others as a major barrier to education, leisure, transport, access to public services, social contact and accessibility outside the home. However, the report also highlighted that the general public felt more comfortable with someone who had a sensory disability than with someone who had learning difficulties or a mental health condition.

Purdam *et al,* (2008) considered the issue of equality in the lives of people with disabilities and found that inequality exists in terms of people who have a disability being more likely to have less or no qualifications than people without a disability, and be living on lower incomes. Additionally, demeaning or negative language, terminology and labels society give people with disabilities affect not only public attitudes but also the way a person with disabilities sees themselves (Bricher, 2000, Zola, 1993). Bricher posits one option that could be considered is the social model of disability as this model used the "term impairment to describe physical, sensory or cognitive changes" (Bricher, 2000 p784), thus rather than focusing on the person's limitations, it considers the impairment experienced by the person. Bricher (2000) posits that the use of language when using the social model of disability as a theoretical foundation to underpin a study is paramount.

Within research, it must be recognised that the use of language or terminology can affect readers' perceptions of disability and influence the study material, thus

appropriate theoretical underpinning is essential, therefore, a variety of theoretical concepts were explored and are considered next.

1.4.2 Disability in the wider context and disability research

Oliver (1992) reported that developing disability research that contributes to empowering marginalised groups rather than being oppressive or dominatory can be a challenge. Therefore, it is essential that when considering conceptual models of disability, the models do not overshadow the important issues in disability, that of oppression, discrimination and inequality (Oliver, 1990).

There are two possible arguments for the involvement of health and social care professionals in disability research (Bricher, 2000). Firstly, disability discourse argues against traditional research that is based on the medical model in favour of the social model, but it is difficult to argue that all research by health professionals into relevant aspects of disability should cease. However, changes in research production away from the traditional models and towards methods which change the relationship of research production would be a positive response to that criticism.

Secondly, many health and social care professionals struggle to keep up with the burgeoning literature in their own discipline and do not read widely beyond it. Whilst there is a plethora of literature relating to the medical and social models of disability (Bricher, 2000, Oliver, 1990, 1992, French and Swain, 1997) the third model, the individual model, receives less attention (Oliver, 1990, 1992). These three models will be considered next, as well as an argument for positioning the social model of disability as a theoretical foundation to underpin this study (Bricher, 2000), together with the theory of identity development (Erikson, 1982).

1.4.3 Medical model of disability

Bricher posits that the medical model used within health has contributed to, "oppression and marginalisation of disabled people" (p781). Whilst the medical model is criticised for focusing on what a person can and cannot do from a medical perspective, as opposed to barriers presented by society which limit what they can do (Bricher, 2000, Oliver, 1996, Brandon and Pritchard, 2011), it could be argued that there are also positive aspects to the medical model (Evans and Whittaker, 2010). For example within sensory disability where hearing loss has been seen as a problem that needs curing or treating, medical advancement and technology have made it possible for some people who may never have had the opportunity to hear, to attain a level of hearing through cochlea implants[40] .

Within disability discourse the medical model is viewed as being in direct conflict with the social model of disability (Bricher, 2000). However, whilst it could be argued that the social model of disability is essential to further develop societal views and legislation (socio-political issues) with regard to disability, it could also be argued that there is a place for the medical model of disability if it promotes and encourages medical research and advancement. If disability was not seen as an area for which medical intervention was needed, then there would be no medical advancement with regard to disability (Evans and Whittaker, 2010). Also as a person's contact with regard to their disability may have been initially with the health or medical professions, the focus would have been from a medical perspective as opposed to a social one. Moreover, the condition of the person with disabilities may be ongoing and as such require regular or stabilising interventions.

However, whilst the medical model of disability has its place, there is no doubt that the environment and society presents more of a disability than the disability itself,

thus consideration of the social model of disability is essential. Additionally, whilst health professionals may have come from a perspective of good intention, resources have been under their control and thus in some cases has contributed to medical dominance over people with disabilities (Bricher, 2000). Thus whilst the medical model does have a place within sensory research, the next section will explain why the social model of disability was adopted for my study.

1.4.4 Social model of disability

As briefly considered earlier, the initial concept behind the social model of disability was that people were not disabled by their impairments but by the disabling barriers people with disabilities faced in society and as such was a socio-political issue as opposed to an individual one (Oliver, 2013, Bricher, 2000). The social model of disability emerged from the Principles of Disability document first published in the mid-1970s by the Union of the Physically Impaired against Segregation (UPIAS) where they proffered that:

> *"Disability is something imposed on top of our impairments by the way we are unnecessarily isolated and excluded from full participation in society. Disabled people are therefore an oppressed group in society" (UPIAS, 1976 p4)*

Beckett and Campbell asserted that the social model of disability did not emerge by chance but was "born of resistance on the part of disability activists" (Beckett and Campbell, 2015 p271), in particular activist Paul Hunt in the 70s, and then developed by other UPIAS members.

Beckett and Campbell (2015) argue that the social model of disability is a model, not a theory and as such it does not provide an explanation for the disablement process.

However they concur that this model has been productive and enabled creation of unity around disability frameworks and the development of a coherent political strategy. Clearly the model together with avid campaigning has influenced societal and legislative views with regard to disability and strengthened people with disabilities movements (Equality Act 2010, Sense, 2012, Oliver, 2013). People with disabilities challenge the fact that independence is not to be able to do everything themselves but to have control over their lives and make decisions as independent people (Bricher, 2000).

Whilst Oliver concurs that there have been some improvements for a person with disabilities there is still a long way to go in terms of education and employment needs (Oliver, 2013). Oliver further highlights that the limitations of a model such as this one, are that people are seen as a disabled group as opposed to individuals who come from a variety of backgrounds, ages, gender, race, sexuality and so on. Oliver further commented that there have been various opinions and views offered over the years with regards to the model (Swain and French, 2000, Shakespeare and Watson, 2001, Allan, 2010) but regardless of strengths or limitations that have been discussed, he saw this model as a "tool to improve people's lives" (Oliver, 2013 p1024). Oliver further highlights:

> *"that focusing on impairment and difference will only de-politicise the social model and will not lead to the development of any approaches or alternative models that are likely to be useful in developing campaigns to improve or defend the lifestyles of disabled people*" (Oliver, 2013 p1025).

He also adds that when the climate is economically strong there are greater benefits for people with disabilities but when times of austerity occur, benefits and services are decreased with the needs of people with disabilities not being met (Oliver, 2013). Whilst various views have been adopted with regard to the social model of

disability, Oliver (2013) asserted “one thing is for sure; the talking has to stop” (p1026).

Bricher (2000) considers the use of the social model of disability in research and its contribution to equalisation of power relationships, and involvement and empowerment of people with disabilities. However, when exploring wider theoretical concepts of disability, whether it is the medical, social or individual models of disability, it is worth noting Verstraete and Söderfeldt’s comment that “although they disagree on the causes, both the social model and the medical model make suffering a central feature of the disabled experience” (Verstraete and Söderfeldt, 2014 p481). Whilst this comment can appear disempowering as it refers to “suffering being a central feature”, many people with disabilities do experience suffering, inequality and discrimination, in terms of societal perception, environmental barriers and the impact of their condition.

1.4.5 Individual model of disability

The individual model incorporates a whole range of issues in relation to disability which includes the personal experience, psychological and medical aspects of a person’s disability (Oliver, 1990, 1992). The individual model has been critiqued as it has been suggested that this model firmly seats the problem or limitation as being with the person who has the disability, and not with the barriers that are placed on the person with disabilities by the environment or society around them (Oliver, 1983, 1990). However, it could be argued that each person’s experience of their disability will be unique to them and therefore each person’s disability is individual.

Other views are that the individual model has compounded marginalisation and oppression of people with disabilities, thus the individual model makes disability an

individual problem as opposed to a social one (Oliver, 1990, French and Swain, 1997, Moore *et al,* 1998, Bricher, 2000). It is also suggested that people with disabilities have become passive research subjects as opposed to active participants and that people with a disability who are involved in research often do not see it improving their own quality of life (Oliver, 1992). It could be suggested that a successful partnership between researcher and researched is achievable but there needs to be a professional relationship built on mutual respect and understanding, with the participant being able to see a positive reason for their contribution. It could be suggested that without the experiences of people with the disability, there is no research and as such the key contributors are not the researchers but rather the research participants.

Issues also arise around power as a key factor, including the level of involvement that the person with the disability being researched has, this will be discussed further in chapter 3. If it is the researcher who makes all the decisions then the power remains with the researcher, however, where this shifts to being participant led, in terms of the participant choosing the communication and interview methods as opposed to the participant's specifically leading the research; autonomy is increased and as such power heightened (Lloyd *et al,* 1996, Bricher, 2000). Beckett and Campbell (2015) suggested that to be able to state with certainty that what is present is a power relation rather than domination, it is necessary to recognise the "prior existence of the capacity to resist in a given situation" (Beckett and Campbell, 2015 p272). A person having choice or autonomy in a given situation could increase the power relation, but it has been argued that equality in research is contestable as research has been conducted *on* people with disabilities as opposed to *with* them (Bricher, 2000). However, the concept of equalising within disability research is also questionable because as the participants have a disability, there will always be an inequality within the balance of power (Oliver, 1992).

Whilst consideration of disability theory in the wider context has presented an argument for positioning the social model of disability as a theoretical foundation to underpin this study, next the theoretical positioning pertinent to deafblindness/Usher syndrome will be considered.

1.4.6 Identity development theory

People who experience deafblindness are noted to be “seen not only as a vulnerable group” (Simcock, 2016 p1) but as "some of the most vulnerable members of society” (Simcock and Manthorpe, 2014 p2325), therefore it is essential to consider theoretical concepts relating to where deafblindness/Usher is positioned within disability. It could be argued that Identity development is affected when an individual experiences a debilitating degenerative condition such as Usher syndrome as it will inevitably affect identity when transitioning from early to later life.

One of the first theorists to be credited with exploring the concept of identity development (psychosocial development) was Erik Erikson, a developmental psychologist and psychoanalyst who trained under psychoanalyst Sigmund Freud (Kroger, 2007). Erikson considered the concept of ‘ego identity’ as a central disturbance in our psychological lives. This concept of ego identity first appeared when Erikson explored the experiences of veterans who survived World War II (Kroger, 2007). Erikson commented that:

> *“What impressed me the most was the loss in these men of a sense of identity. They knew who they were; they had a personal identity. But it was as if, subjectively, their lives no longer hung together – and never would again….this sense of identity provides the ability to experience one’s self as something that has continuity and sameness and to act accordingly”*

(Erikson, 1963 p42)

Whilst these experiences refer to war veterans they could also pertain to people who experience Usher syndrome. The life that people plan prior to the diagnosis of Usher is often very different to the life that will be their future (see Chapter 4).

Erikson considered that ideal identity development included finding social roles that enable that person to fit into the wider community (Kroger, 2007). However for a person who experiences Usher, the community they may have been part of is affected by the change in their sensory need. For example if a person is part of the Deaf community and uses British Sign Language (BSL) as their first language, when they lose their sight this affects communication as BSL is a visual form of communication. Erikson's life span perspective developed an eight stage cycle (Erikson, 1982) of development which necessitated resolution at different stages of the life span. These stages included:

1. Infant Birth to 18 months
2. Toddler 18 months – 3 years
3. Pre-school 3-5 years
4. School age child 6-11 years
5. Adolescent 12-17 years
6. Young adult 18-39 years
7. Middle age 40 - 64 years
8. Older adult 65+ to death

(Adapted from Erikson, 1998; Walker and Crawford, 2010)

Erikson suggested that as a person journeys through the eight life stage developments, the challenges they face influences the forming of intimate relationships (Walker and Crawford, 2010). Erikson considered the goal is the

ability to form intimate relationships whilst still retaining one's own identity. If these negotiations are successful then our thoughts, emotions and personality will develop and love and commitment will be experienced. However, if unsuccessful, the experiences may be that of isolation and difficulty forming long lasting deep relationships (Walker and Crawford, 2010).

Although, these stages of development are critiqued because they are mainly focused from a male perspective and are Eurocentric and fail to recognise difference and diversity in adults outside of the western world (Walker and Crawford, 2010), it is worth noting that Erikson studied a range of Native American tribes, which included the Sioux and Yurok tribes and considered their difference and diversity (Thornton, 1998). Additionally, the stages are seen as fixed and in reality life does not come in neat packages. However, Erikson theorised these stages in the mid twentieth century and would have been unaware of the future changes that would take place on a global scale.

Another critique of identity development theory is that there are socially constructed expectations and values of our society and if a person does not meet these expectations they may feel they have failed (Walker and Crawford, 2010, Arnett, 2000). However, it could be suggested that there are milestones to reach from the day that a child is born (Crawford and Walker, 2010), with their growth and development being mapped to gauge development and prevent any medical issues being overlooked. If a child is not meeting these milestones, this raises cause for concern (parents, caregivers, healthcare professionals). Whilst there is recognition that people are different, there are also societal expectations that a person will reach a certain point at a certain stage (Crawford and Walker, 2010). As the development of people with Usher will be affected by living with their sensory condition, this theoretical concept while having a place within disability theory also underpins my

research. It is however, also important to remember that whilst theory that is positioned within disability has application in various settings, it is just a theory, and should not be fixed or universally accepted (Walker and Crawford, 2010) and therefore it should be remembered that “wisdom and integrity are active, lifelong, developing processes as are all the strengths included in the life cycle” (Erikson, 1982 p9).

1.4.7 Deaf culture and disability theory

The final area to be contemplated in the context of disability theory is where d/Deafness is positioned. The reference to Capital ‘D’ Deaf [15] and small ‘d’ deaf 31[16] is significant. As explained earlier capital ‘D’ Deafness refers to a person who would most likely be a profoundly Deaf person who would not consider their sensory difference to be a disability or consider themselves to be disabled (Ladd, 2003), but rather part of a minority group with its own culture, history and linguistics (British Deaf Association, 2007). This was highlighted by the British Deaf Association (BDA) when British Sign Language (BSL) attained official language status on 18th March 2003. Deaf people consider being Deaf as part of their heritage and most likely consider themselves proud to be Deaf (Evans and Whittaker, 2010).

However, small ‘d’ deafness refers to a person who most likely would consider deafness to be a disability. A person who has previously experienced the ability to hear music, voices and birds singing and then becomes profoundly deaf, for example as a result of meningococcal infection, can experience this as traumatic loss and be more likely to label themselves with a disability (Evans and Whittaker, 2010, Williams and Evans, 2013). Therefore, a small ‘d’ deaf person would most likely consider themselves disabled by their sensory impairment, as would a person who experiences deafblindness, or Usher syndrome, and thus disability theory that

applies to deaflindness or Usher would also apply to them. For a person who is capital ‘D’ Deaf, their theoretical positioning may be different as they would not consider their sensory difference a disability.

As considered earlier in this section the social model of disability proffers that people are not disabled by their individual limitations but by the disabling barriers they face in society when appropriate adjustments to meet their needs are not applied (Oliver, 1983, 1990, Bricher, 2000, Kitchin, 2000, Bolt, 2005, Oliver, 2013). Harris (1995) discusses her experience as a hearing researcher in a Deaf world in which she explored Deaf culture. Harris reported that:

> “*Deaf people tell me they feel an `affinity’ with disabled people, particularly campaigning groups, yet they have always felt separate and this distinction is important to them*” (Harris, 1995 p296).

Harris identified that the reason for this is that rather than being disabled by their sensory difference they are part of a linguistic and cultural minority group which is central to their identity.

Therefore whilst the social model of disability is crucial for people who are culturally Deaf, to challenge power-loaded relationships within research, theoretical concepts such as Erikson’s identity development would be less likely to be applicable, because Deaf people have a strong cultural identity and would not consider that their development has been affected by their sensory difference i.e. not hearing. Therefore for Deaf people, dialogue or communication in which there is a “collaborative process of knowledge production” (Caretta and Riaño, 2016 p2) is fundamental, as an exchanging of knowledge has the capacity to “create an inclusive space”, which can lead to a “more balanced relationship between power and knowledge” (Caretta and Riaño, 2016 p2).

To summarise, in this section, consideration was given to the way people who experience disability are viewed and how this is influenced by terminology used. Taking this into account necessitates that researchers critically reflect upon theoretical frameworks applied to research to maximise the potential usefulness of research conducted, therefore models of disability and identity developmental theories have been considered to understand where D/deafness and deafblindness/Usher syndrome are positioned not only within disability theory but also within a framework to underpin this study. One example is the social model of disability, which highlighted that people are not disabled by their impairments but rather by the disabling barriers disabled people in society face (Oliver, 2013, Bricher, 2000), therefore, if people with Usher were provided with the appropriate support, challenges that are imposed due to the impact of the condition would be reduced. Exploration identified that whilst the social model of disability provided a framework to underpin the study, identity development theory also influenced it. Next legislation/guidance and its application to deafblindness and Usher syndrome will be considered, together with a brief consideration of application to D/deafness.

1.5 Legislation and guidance

Appropriate legislation and guidance which directly affect people who experience D/deafblindness, Usher syndrome or D/deafness are essential, as without it there would be no provision of support and services to meet the needs of people who experience these sensory conditions. In England the current D/deafblind guidance is ‘Care and Support for Deafblind Children and Adults’ (Department of Health, 2014). Local authorities in England are required to act under this guidance under Section 7 of the Local Authority Social Services Act 1970, which outlines provision with respect to the organisation, management and administration of local authority social services for children, and section 77 and 78 of the Care Act 2014, which

outlines how local authorities should conduct carers needs assessments; should determine who is eligible for support; should charge for both residential care and community care; and places new obligations on local authorities, for adults (Sense, 2014c). This current D/deafblind guidance retains the principles of the previous guidance it replaced, 'Social Care for Deafblind Children and Adults' (Department of Health, 2009), but it also includes updated guidance to reflect the Care Act (2014). Table 1.2 outlines guidance to local authorities to support people who experience D/deafblindness/Usher syndrome.

Table 1.2: Guidance to local authorities (Sense, 2014c)

Ensure that when an assessment of needs for care and support is carried out, this is done by a person or team that has specific training and expertise relating to D/deafblind persons – in particular to assess the need for communication, one-to-one human contact, social interaction and emotional wellbeing, support with mobility assistive technology and habilitation/rehabilitation
Ensure services provided to D/deafblind people are appropriate, recognising that they may not necessarily be able to benefit from mainstream services or those services aimed primarily at blind people or deaf people who are able to rely on their other senses
Ensure that D/deafblind people are able to access specifically trained one-to-one support workers if they are assessed as requiring one; Provide information and advice in ways which are accessible to D/deafblind people
Ensure that a Director-level member of the local authority has overall responsibility for D/deafblind services

There are also assessment regulations (Care Act 2014 s77), which strengthen the policy guidance for adults as they outline the training an assessor must have, it they are to undertake a D/deafblind guidance assessment. This is essential as when assessing the needs and care planning for a person who experiences D/deafblindness/Usher syndrome, specialism is required to ensure person centred communication is used (BSL[6] , Deafblind manual[17] , Hands on signing[7], clear speech); the impact of deafblindness, physically, mentally and emotionally is realised and that mobility issues, safe orientation and social isolation are addressed (Sense, 2014b). Additionally, the legal basis for the provision of communicator guides[41] for the local authorities to provide services falls under the Care Act 2014

(section 77 3a) in which planning the provision by the authority of services to meet needs for care and support is required. The difficulty with guidance is that it is just guidance, and when resources are cut so are specialist support and services. However, guidance is directed by parliament and as such does direct local authorities to adhere to these responsibilities.

In addition to legislation and guidance discussed, another Act of parliament relevant to people who experience D/deafblindness/Usher is the Equality Act 2010, which replaced the Disability Discrimination Act 1995 and protects people who experience disability or sensory impairment against unlawful discrimination which applies to employment settings, including recruitment processes, education settings, provision of goods, facilities and services, exercise of public functions, the disposal and management of premises and associations including private clubs. Whilst this Act incorporates victimisation and harassment it also applies to reasonable adjustments such as allowing a person to write a note to make a request in a shop or take a guide dog onto premises.

Whilst people who experience Deafness would also access provision for assessment and support under the Care Act 2014 and be protected under the Equality Act 2010 a significant legislative milestone which impacted on Deaf people occurred in 2003. The British Deaf Association (BDA) launched a campaign in 1987 to establish official status for BSL as a language in its own right as it is the first preferred language of an estimated 70,000 Deaf people in the UK (BDA, 2007, see 1.4.7). Although the European parliament gave its support from 1988, it was not until March 2003 that the British government established BSL official status as it being a language in its own right.

Whilst organisations such as Sense and the BDA campaign to improve the lives of people with sensory requirements, there is still much to be done in terms of understanding what it is like for people to live with D/deafblindness/Usher syndrome. In times of austerity when services are under threat, there needs to be even stronger legislation and guidance in place to ensure people's support needs can be met.

1.6 Structure of thesis

The doctoral thesis is structured to guide the reader through the thesis in a systematic way. By so doing, the information contained will give an insight to better understand the diagnosis of, living with and experiences of people with Usher syndrome and what can be provided to operate within an equal opportunity framework with regards to independence and aspirations.

Chapter 1 began with an introduction and rationale for the study. An explanation of Usher syndrome was provided (1.2) and theoretical constructs that underpin the study (1.4), legislation and guidance applicable to sensory need (1.5) and structure of the thesis were presented.

Chapter 2 critically reviews literature and considers four key themes that were identified (Table 2.9): impact of diagnosis for people with Usher syndrome; support and intervention for people with sensory impairment; psychological impact for people with sensory impairment and safety and risk for people with visual impairment.

The methods used for the study and the rationale for using a descriptive phenomenological methodology are presented in Chapter 3, together with reasoning

for paradigmatic positioning (Figure 3.2) and discussion relating to trustworthiness (3.7).

In Chapter 4, the findings are presented in four themes with sub-themes (see Table 4.1) and key messages from these findings (see Table 4.4) relating to participants' experiences of diagnosis of, and living with Usher syndrome.

The discussion in Chapter 5 moves away from individual experience and discusses the study findings around the experience of diagnosis of, and living with Usher syndrome, in relation to published literature. Three overarching messages (Table 5.2) were revealed as a result of exploring the four themes from my findings (Table 4.1) and then extracting key messages from these themes (Table 4.4).

Chapter 6 concludes the thesis, outlining: new knowledge, conclusions drawn and key learning; benefits of reflection; strengths and limitations of the study, suggestions for future research and recommendations for practice and service development; recommendations for education; intentions for dissemination of findings and conclusion.

1.7 Chapter summary

In this chapter, the doctoral thesis was introduced and the rationale behind the study provided. An explanation of Usher syndrome was given, as was consideration of disability theory in the wider context and theory pertinent to deafblindness/Usher and D/deafness. Guidance and legislative contexts were discussed and the structure of the doctoral thesis was provided.

Worth noting at this stage is the ancient Chinese proverb as philosophised by Lao-Tzu, a contemporary of Confucius, who reasoned that, a journey of a thousand miles begins with a single step (Seddon, 2006). It could be suggested that this doctoral thesis is the thousand mile journey with chapter 1 being the first step, chapters 2, 3, 4, and 5 being the journey in-between and chapter 6 being the final step. Sometimes for a student starting out on a PhD or other doctorate, the task can appear daunting, however, by considering Lao-Tzu's proverb and viewing each progression as a single step, eventually the journey can be achieved, although as will be considered in chapter 6, there is a question mark as to whether the journey really will end. Whilst the thesis will show how it has contributed to new knowledge, data collected will also be the beginning of future development and research and thus the journey continues.

Chapter 2 Literature review

2.1 Introduction

In chapter 1 an introduction to 'Usher syndrome: A phenomenological study of adults across the lifespan living in England' was provided. In this chapter the research topic will be contextualised with a review of literature that considers the diagnosis of, living with and experiences of Usher syndrome. As already highlighted Usher syndrome is a rare under-researched topic (Kyle and Barnett, 2012, Ellis and Hodges, 2013) thus literature exploration incorporated other areas of sensory need e.g. deafblindness, D/deafness, blindness and visual impairment (especially focusing on retinitis pigmentosa - RP). Considering other areas of sensory loss enabled a wider context to be examined within the literature review. The first section considers the approach chosen to conduct the literature review, using a narrative approach, the search strategy and how the 12 peer reviewed articles and two funded research reports were selected.

2.2 Overview of literature reviewing techniques

This section will consider literature reviewing techniques used in qualitative research, but first the requirements for conducting a literature review will be considered. Key questions when conducting a literature review are: 'what is expected from a literature review? What is the starting point, and what is the end aim?' To clarify this, Table 2.1 outlines the purpose of a literature review.

Table 2.1: Purpose of undertaking a literature review
(Adapted from Wellington *et al*, 2011)

To define the subject area one wishes to explore
To establish research already conducted that relates to the research question
To consider theories, concepts and models used and applied in this subject area
To identify and discuss methods and approaches used in other research by other researchers
To identify gaps and offer evidence of how the present piece of research contributes to filling the gaps

Use of a structure when conducting a literature review enables a clear and organised approach (Green *et al,* 2006). However, whilst a basic structure is useful for undertaking smaller literature reviews, a variety of options are available when literature needs to be reviewed in greater depth, to ensure pertinent literature is considered and enable the researcher to find gaps in literature that are relevant to the study topic. Techniques that were considered included a systematic review, a scoping review, a metasynthesis and a narrative review (Norman and Griffiths, 2014, Boland *et al,* 2014, Wellington *et al,* 2011, Green *et al*, 2006). To decide which was appropriate and relevant to the research study each technique was explored.

A systematic review is rigorous, structured, designed to locate, appraise and synthesise the literature, in order to focus on a specific question, which can be replicated (Wellington *et al,* 2011, Norman and Griffiths, 2014, UyBico *et al,* 2007, Finke *et al,* 2008, Harden *et al,* 2009). Lubben (2013) posits that "systematic reviews intend to answer questions of a particular type" and seek to answer question such as "what strategies work?" (p139). They are particularly suited to large quantitative evidence based studies particularly those with many randomised control trials (Panesar *et al,* 2006). This approach was not useful to this literature review as it was considered the use of keywords and synonyms (Table 2.4) would be useful, which would not be appropriate in the rigorous, structured approach of a systematic literature review.

An alternative option is a scoping review which maps the literature that can be found on a particular topic. This method explores what is currently known about the topic but does not explore deeply the worth or quality of the literature found (Norman and Griffiths, 2014). Whilst, initial scoping identified a limited data set relevant to Usher syndrome, this approach was not considered appropriate for this literature review as greater depth of knowledge was required.

A metasynthesis is, as the name suggests, a synthesis of literature and knowledge found on the topic (Norman and Griffiths, 2014). In addition Walsh and Downe (2005) posit a metasynthesis approach to reviewing literature generates new insights and understandings within qualitative research and is generally conducted where there are large quantities of qualitative data.

An alternative approach to reviewing literature is the narrative method (Green *et al,* 2006, Mulongo *et al,* 2014, Attard *et al,* 2015, Venter *et al,* 2012, Lazenbatt, 2013). Narrative literature reviews are able to cope with small numbers of studies involving limited number of participants and also enable a combination of studies to be explored. While these studies may differ in their methodological approach, they are able to capture an array of perspectives with regard to a social phenomenon (Østergaardan, 2015). Lubben (2013) further considers that the narrative approach to reviewing literature enables the researcher more flexibility in exploring the scope of the review topic, the variety of studies that could be included in the review, the perspective taken for putting the findings together and how the final literature review is presented. However, a limitation of this level of flexibility may be that there is researcher bias with regard to what is presented and discussed (Lubben, 2013, Oakley, 2000).

After consideration of the four literature review options, it became apparent that the narrative approach was compatible with the study as the aim was to draw on a range of literature to develop an understanding of the experiences of diagnosis of, and living with Usher syndrome, from the perspective of adults in England. The aim of the literature review approach for the study was not to explore which strategies work, as is the case with systematic reviews, nor to just scope the literature, which may not enable a deeper analytical understanding, nor to only consider literature and knowledge found on the topic, as with a metasynthesis approach. A narrative approach allows for flexibility in the literature search while considering research material from a variety of methodological approaches (both qualitative and quantitative).

As already outlined Usher syndrome is a rare condition, so the search would have been too narrow if only Usher syndrome was used as a search term. As the purpose of this literature review is to contextualise the research topic within wider literature, other aspects of sensory need were included. A narrative approach allowed search terms to include diagnosis, living with, and experiences of, not only people with Usher syndrome but also deafblindness, d/Deafness and blindness/visual impairment. The narrative approach also evidences that previous research has been reviewed using a structured approach to avoid missing relevant literature and thus, whilst not adopting a systematic review approach, the literature review is structured and organised. To further evidence a comprehensive literature search, tables were constructed to explore search terms (Table 2.3), keywords and synonyms (Table 2.4) and electronic databases (Table 2.5).

Abstracts reviewed and included or excluded are reported upon giving the reasons for decisions made (Table 2.6, Fig 2.1). The selected papers were evaluated using the Critical Appraisal Skills Programme questions sets (CASP-uk.net). CASP

produces assessment tools for use with qualitative and quantitative research and assists in critiquing the rigour, credibility and relevance of a research paper by using a series of questions. As CASP identifies, rigour relates to whether a thorough approach has been used in the research carried out; credibility concerns whether the findings are well presented and meaningful, and relevance explores how useful the findings in the study would be to the proposed study. Whilst this tool has been developed for researchers who are unfamiliar with research it is useful for all researchers in so far as it enables the researcher to conduct a structured evaluation of the literature selected (Dixon-Woods *et al*, 2007, Hannes *et al, 2010,* Attard *et al, 2015).* As the narrative approach is the chosen method for the literature review the next section will consider the approach in more depth.

2.3 Narrative review

Braun and Clarke (2013) outline that a narrative is "an account of events, or more than one event, characterised by having some sort of structure…and other story elements" (Braun and Clarke, 2013 p333) thus the aim is to tell a story. Green *et al* (2006) give structured guidance on the process to contribute to a successful narrative review, identifying that it should be well structured, synthesise the available evidence about the topic, convey a clear message and be as objective as possible.

Mulongo e*t al* (2014) posits that a narrative review is useful to explore what knowledge is known on the topic and suggests that narrative reviews allow the researcher to get a useful overview of the topics being researched. Mulongo *et al* (2014) further proffers that a narrative review is useful for problem identification and can indicate "problems, weaknesses, contradictions, or controversies in a particular area of investigation" (Mulongo *et al,* 2014 p471). Whilst they consider that the expectation is not for the researcher to solve the problems, they suggest that a

narrative review informs and identifies gaps in the literature, thus highlighting areas that may require further research.

Lubben (2013) identified that there can be subjectivity in the search strategy as the researcher chooses what they want to include and what they do not. Similarly, Green *et al* (2006) argue that:

> *"in this rather unsystematic approach, selection of information from primary articles is usually subjective, lacks explicit criteria for inclusion and leads to a biased review*" (Green *et al*, 2006 p104).

Green *et al* (2006) further consider that the author's interpretation and synthesis of information should consider differences between studies, for example if patient samples in one study are different to another or if the research designs are not equitable.

Bias is also an area where researchers need critical awareness for example, in this study, the researcher was previously a sensory social worker. Section 3.7.5 considers the usefulness of reflexivity to address insider bias and subjectivity in greater depth. Green *et al* (2006) identified that researchers need to be aware of potential bias and take preventative action. Hence Green *et al*'s (2006) structure was adopted for conducting the narrative literature review (see Table 2.2). This approach will be referred to as the 5 Ss (Search, Screen, Select, Summarise, Synthesise), 5 stage narrative approach; each of the stages 1-5 will be explained.

Table 2.2: Structure for conducting a narrative literature review -The 5 Ss
(Adapted from Green *et al,* 2006)

Stage 1: Preliminary search of literature – **Search**
Stage 2: Comprehensive narrative screening of previously published information -**Screen**
Stage 3: Selection criteria employed/What was included and why - **Select**
Stage 4: Summarise each study included - **Summarise**
Stage 5: Discussion – Synthesis - Writing/Recording - **Synthesis**

2.3.1 Stage 1 Search

The first stage is to conduct a preliminary search of the literature (Green *et al*, 2006). Search terms were identified and Table 2.3 highlights the order in which searches were conducted. The primary subject was searched first followed by each sub-subject: date parameters were 2005-2015.

Table 2.3: Search Terms in the order in which searches were conducted

Primary Subject	**Sub-subjects**		
1 Diagnosis of Usher syndrome	Diagnosis of deafblindness	Diagnosis of d/Deafness	Diagnosis of blindness/visual impairment (focusing on RP)
2. Living with Usher syndrome	Living with deafblindness	Living with d/Deafness	Living with blindness/visual impairment (focusing on RP)
3.Experiences of Usher syndrome	Experiences of deafblindness	Experiences of d/Deafness	Experiences of blindness/visual impairment (focusing on RP)

Next keywords and synonyms were identified to avoid missing any relevant or significant literature (Table 2.4)

Table 2.4: Keywords and synonyms

Keywords	Synonyms
Usher syndrome	Deafblind
Deafblind	Deaf and blind
	Deaf and vision impaired
	Deaf and sight impaired
	Deaf and partially sighted
	Deaf without sight
	Deaf with sight loss
	Blind and Deaf
	Blind and hearing impaired
	Blind and partially hearing
	Blind without hearing
	Blind and deafened
	Blind with hearing loss
D/deaf	Deaf
	deaf
	Hard of hearing
	Loss of hearing
	Partially hearing
	Hearing impairment
	Deafened
	Hearing loss
Blind	Sightlessness
	Loss of sight
	Partially sighted
	Visual impairment
	Sight impairment
----------AND----------	
Experience	Perception
	Living with
	Knowledge
----------AND----------	
Diagnosis	Analysis
	Finding
	Identification

To further contribute to a systematic and thorough approach to the literature search a range of electronic databases were accessed (Table 2.5).

Table 2.5: Electronic databases used in literature search

Database Name:	Brief Description:
CINAHL (Cumulative Index of Nursing and Allied Health Literature).	Provides journals/texts for nursing/ allied health professionals
PsychINFO	Relates to the field of psychology – Produced by the American Psychological Association
Medline	Incorporates life sciences and biomedical information – Mainly medical/clinical resources
Academic search complete	Index and abstracting service which included full text periodicals/ peer reviewed journals
SocINDEX	Sociological database which covers topics relevant to social work
AMED	AMED (Allied and Complementary Medicine) - produced by Health Care and Information Service at the British Library - relates to complimentary medicine/palliative care.
Scopus	Covers medical, scientific, social sciences, technical related data

The aim was to identify what research relating to Usher syndrome had already been conducted between the year range 2005 - 2015. Table 2.7 summarises the papers that were selected through a process that incorporates choosing the most suitable papers. Prior to 2005, literature was predominately medically focused (Pennings *et al,* 2003, Sadeghi *et al*,2004) whereas in 2005 Damen *et al*'s study explored the lifestyle of people with Usher and acknowledged challenges faced when hearing and sight worsens, which were of interest to my study. 2015 was selected as the end range because no other non-medical literature was found. Searching the literature is different to reviewing it, as the search merely scans the material, considering the abstracts to identify studies that are relevant to the topic, whereas the literature review is the in-depth analysis of the papers identified as relevant (Patton, 2002).

Initially Usher syndrome was searched using a variety of databases (Table 2.5); a preliminary search of Usher syndrome identified 1571 papers of which 974 were linked to Medline and were mainly clinical/medical Usher based papers and only 6 were found in SocINDEX, the sociological electronic database. The search was then

specifically focused to include Usher syndrome and the search terms outlined in Table 2.3 and keywords and synonyms outlined in Table 2.4 applied; this resulted in 1,478 papers being identified for screening.

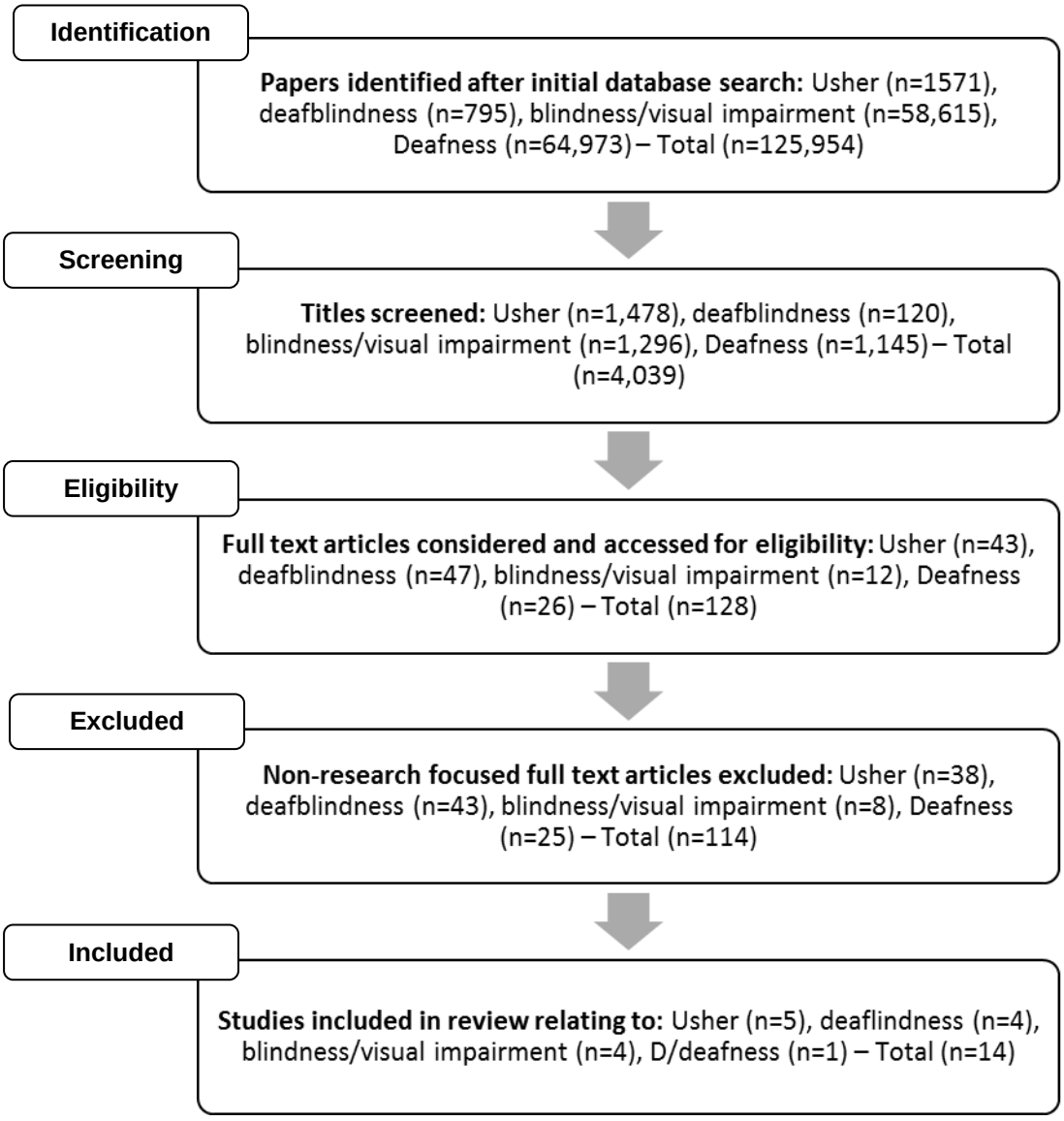
The next search related to deafblindness. The initial search identified 795 papers with many of the papers relating to equipment and communication issues. As with Usher, The original search descriptors were included (Tables 2.3, 2.4) resulting in 120 papers and these titles were selected for screening.

The search relating to blindness/visual impairment specifically focusing on retinitis pigmentosa (RP) resulted in no papers being found so clearly the search criteria was too narrow, therefore, RP was removed. The initial search identified 58,615 papers, 33,046 were medically/clinically related with 1,296 papers being SocINDEX related, and therefore these 1,296 papers were selected for screening.

There were 64,973 papers relating to D/deafness of which 34,459 were medically/clinically related after applying tables 2.3 and 2.4, 1,145 of the 34,459 were selected for screening.

The flow diagram (Figure. 2.1) summarises the literature search process on Usher, deafblindness, blindness/visual impairment and D/deafness.

Figure 2.1: Flow diagram for literature review on Usher, deafblindness, blindness/visual impairment and D/deafness



The screening and selection processes also summarised in Figure 2.1 will next be considered.

2.3.2 Stage 2 Screen

Having conducted a preliminary search to identify published material relating to Usher and other related sensory needs, studies were selected for screening, with the purpose being to include and exclude studies. As the literature was screened to check for relevance to subject material, the inclusion and exclusion details were added to Table 2.6.

Green *et al,* (2006) highlight it is essential to briefly describe the rationale for including or excluding studies from the review. They identify that this helps to keep the papers focused and ensure that papers are included because of "their relevance to the topic rather than how much the author agrees or disagrees with the study" (Green *et al,* p109). They further advise that reasons for exclusion may be "old data (early research), or inappropriate topics" (Green *et al,* p109). To conduct the screening in a structured manner, Table 2.6 was constructed, as it kept the literature review focused and facilitated critical consideration of reasons why material was included or excluded.

Of the 1478 papers screened relating to Usher, after applying including and excluding criteria (Table 2.6), 43 papers were eligible and were considered and accessed. 38 papers were not research focused papers and so these were excluded. With regards to deafblindness, again, after papers being included and excluded (Table 2.6), 47 papers were selected for eligibility however, 43 papers were excluded as they were not research focused. Table 2.6 was again applied to blindness/visual impairment and 12 papers were considered eligible, as 8 were not research focused they were excluded. After screening (Table 2.6) D/deafness 26 papers were eligible, 25 papers were excluded as again, they were not research focused. For summary see Figure 2.1. After screening the next stage was selection.

Table 2.6: Reasons for including and excluding papers

Papers included:	Papers excluded:
• Life, lifestyle and experiences of Usher • Life, lifestyle and experience of deafblindness • Life, lifestyle and experiences of Deafness • Life and experience of blindness • Managing change as a result of Usher • Psychological experiences of Usher • Sociological experiences of Usher • Research with adolescents • Children with sensory need and multiple disabilities • Research focused papers	• Non-research papers • Clinical/medical research • Audiological/audiogram research • Practical based e.g. workshop to raise Usher awareness amongst professionals • Parents • Dementia • Psychiatry • Older people and residential care • Genetics issues • Retinal issues • Cochlear issues • Equipment exploration • Interpreter issues • Molecular science • Opinion/legacy/debate/discussion papers • Deafblindness and learning disability • Blindness and ethnic minority groups • Linguistic issues • Poetry as a social research method

2.3.3 Stage 3 Select

Adherence to stages 1 and 2 led to selection of 14 papers for the literature review (Table 2.7), five related to Usher syndrome, four to deafblindness, four to blindness/visual impairment and one to D/deafness (see Figure 2.1). With regards to Usher syndrome as highlighted previously the majority of the papers identified were from a clinical/medical perspective, which is not surprising as Usher syndrome is a medical condition which requires medical/clinical research in order to understand the nature and treatment approaches of the condition (Yoshimura *et al*, 2015). Four papers selected were peer reviewed research journal articles and one research study selected was a funded report commissioned by Sense, a charitable organisation which supports and campaigns on behalf of people who experience deafblindness (Ellis and Hodges, 2013). Although this was a report, was not peer-reviewed and is yet to be formally published, this was the most recent research study conducted with people who experience Usher syndrome thus it was considered important to include in the review of literature (Table 2.7).

Whilst working on a manuscript relating to 'Empowering people with Usher syndrome as participants in research', which was later published in the British Journal of Social Work, a further funded report by Kyle and Barnett (2012), which briefly referred to Usher syndrome, was identified and included in the literature review (Table 2.7). As the primary participant group were people who referred to themselves as Deafblind as opposed to people who referred to themselves as having Usher, review of Kyle and Barnett's report was included with papers selected pertaining to deafblindness (Table 2.7). Thus five papers were selected for review concerning Usher syndrome (Table 2.7).

With regard to deafblindness much of the literature related to equipment, communication, medical and clinical issues and were not research focused therefore only three papers were selected for critique (Table 2.7). However, Kyle and Barnett's (2012) funded report was also included, totalling four papers selected for review. Four papers relating to blindness/visual impairment were research related and were selected for further critique (Table 2.7). At this stage only one paper relating to D/deafness was selected with other papers discounted for a number of reasons (Table 2.6).

Out of the 14 articles selected, two were funded reports (Ellis and Hodges, 2013, Kyle and Barnett, 2012) and 12 were peer reviewed articles (Table 2.7). Five articles included research with children and young people (Ellis and Hodges, 2013, Högner, 2013, Martens *et al*, 2014, Hadidi and Khateeb, 2014 Argyropoulos and Thymakis, 2015) as these experiences were considered relevant to people with Usher. After conducting stage 1, a preliminary search of the literature, stage 2 screening the literature and stage 3 selecting literature/relevant papers, the next stage is to summarise each study.

2.3.4 Stage 4 Summarise

Summarising encapsulates the studies selected and enables the researcher to determine strengths and limitations of the paper and explore its contribution to the topic, thus highlighting gaps in the literature (Green *et al,* 2006). To enable this stage to be as comprehensive as possible and summarise each article individually, CASP was used to evaluate papers selected and Table 2.7 was constructed.

Table 2.7: Summary of papers selected for narrative literature review

Author(s) and year published	Method	Number of participants, age range and gender	Communication method used	Setting
USHER SYNDROME				
Ellis and Hodges (2013)	Qualitative case study interview based approach exploring lives of people with Usher syndrome	n=42: 14-56 years 16 Male 26 Female	Verbal Spoken English, Written, British Sign Language (BSL) and Braille	United Kingdom (UK) FUNDED REPORT
Damen *et al* (2005)	Quantitative cross sectional survey conducted applying a descriptive questionnaire with closed/open ended questions and multiple-choice to gain data for the Usher lifestyle survey.	n=93: 18 younger than 25 years 41 aged 25-45 years 34 older than 56 years 50 Male 43 Female	Written	France, Germany Ireland, Italy Spain, UK and the Netherlands
Côté *et al* (2013)	Mixed method experimental study initially using a group intervention program and then questionnaires to explore helping person with Usher type II to adapt to deafblindness	n=7: 40-65 years 4 Males 3 Females	Written	Canada
Wahlqvist *et al* (2013)	Quantitative questionnaire by mail based study exploring physical and psychological health in persons with deafblindness that Is due to Usher syndrome type II	n=96: 18-84 years 53% Female 47% Male	Written	Sweden
Högner (2015)	Quantitative, questionnaire based study exploring psychological stress in people with dual sensory impairment through Usher syndrome type II	n=262: 17–79 years 53% Female 47% Male	Written	Germany
DEAFBLINDNESS				
Martens *et al* (2014)	Qualitative observational study exploring effectiveness of	n=4: 15-54 years 2 Females 15 and 26 years	Non-verbal Communication	Holland

	intervention with people who are congenitally deafblind.	2 Males aged 15 and 54 years Plus n=16: Communicators 21-55 years All female	Verbal Spoken English	
Prain *et al* (2010)	Part of a two phase mixed method study. Quantitative study observing the behavior and interactions of adults with congenital deafblindness living in community residences with support staff	n=9: 22-44 years Plus n=9: Paid disability support staff 24-59 years 7 Female 2 Male	Non-verbal Communication Verbal Spoken English	Australia
Prain *et al* (2012)	Second phase of mixed method study. Qualitative study exploring experiences of support workers when interacting with adults with congenital deafblindness	n=8: 24-59 years 6 Female 2 Male	Verbal Spoken English	Australia
Kyle and Barnett (2012)	Mixed method two phase study to explore the Deafblind experience (particularly in comparison with that of Deaf people) and to test a framework for interaction and support by Deaf people for Deafblind people. Study also refers to Usher	n=21 Deafblind people 21-65 years 7 aged 21-35 years 7 aged 36-50 years 6 aged 51-65 years 15 Female 6 Male n=38 Deaf people Age range and gender not specified for Deaf people as intension was to create matched pairs in regard to gender and age. This proved impossible	BSL, manual alphabet (deafblind manual) and hands on signing	UK FUNDED REPORT

BLINDNESS/VISUAL IMPAIRMENT				
Hadidi and Khateeb (2014)	Quantitative study exploring the comparison of social support among adolescents with and without visual Impairments	n=86: Adolescents with visual impairments 12-17 years (7^{th}-12^{th} grade students) 42 visually impaired males, 44 visually impaired females n=73: Sighted adolescents 34 sighted males and 39 sighted females	Verbal Spoken English	Jordan, Arab Region
Kim *et al* (2014)	Quantitative observational study exploring the Influence that ambient sound fluctuations have for people who are visually impaired when crossing the road.	n=14: 18-68 years 9 Males 5 Females	Verbal Spoken English	USA
Brooks *et al* (2014)	Quantitative questionnaire based study exploring substance dependence among people with visual impairments	n=69: 18-78 years 43-Male 26 Female	Verbal Spoken English	USA
Argyropoulos and Thymakis (2015)	Qualitative action research project exploring multiple disabilities and visual impairment	n=1: Female 12 years	Verbal Spoken Greek, Italian, and Albanian	Greece
DEAFNESS				
Sheppard and Badger (2010)	Qualitative Hermeneutic phenomenological study exploring lived experience of depression among culturally Deaf adults	n=9: 21-65 years 2 Males 7 Females	American Sign Language (ASL)	USA

Green *et al,* (2006) advise that prior to the final stage of, synthesising selected literature, which generates deeper understanding and facilitates a combination of

individual studies to form a generic whole (Booth *et al,* 2012, Patton, 2002), the researcher should read through each paper that will be included in the overview and take notes on each one. This process enables familiarisation with the literature selected before embarking on the in-depth process of synthesis. Green *et al*'s (2006) structured approach (Table 2.8) was used for writing up.

Table 2.8: Structure for writing up critique of literature from a narrative perspective
Adapted from Green *et al* (2006 pp110-112)

The purpose of the study reviewed
Synopsis of the content
Research design/ methods used in study
Brief review of findings/themes that arise
Clear and concise summary of major findings of the overview

From reviewing the selected papers and conducting an organised structured review of literature using a narrative approach, four themes were identified (Table 2.9) which linked with the aim of developing an understanding of the experiences of diagnosis of and living with Usher syndrome, from the perspective of adults in England.

Table 2.9: Themes identified from literature review

Impact of diagnosis for people with Usher syndrome (n=1)
Support and Intervention for people with sensory impairment (n=7)
Psychological impact for people with sensory impairment (n=5)
Safety and risk for people with visual impairment (n=2)

Thus the next step was to critique the literature within these four themes. Green *et al* highlight that there are strengths and limitations within each paper as no paper is "perfect" (Green *et al,* 2006 p111), so, tables 2.7 and 2.8 were used to review papers selected. For the purpose of this review, with the exception of Ellis and Hodges (2013), whose study will be explored in two themes: impact of diagnosis for people with Usher syndrome and psychological impact for people with sensory impairment, the primary theme that arose within each research study will be the

main focus of review. Ellis and Hodges's (2013) funded report is an exception because it is the most current UK based study of Usher.

2.3.4.1Impact of diagnosis for people with Usher syndrome

The study commissioned by Sense[28] was the only one that specifically considered the impact of diagnosis for people with Usher syndrome. Ellis and Hodges (2013) explored the lives and experiences of people with Usher syndrome living in the UK. Participants included people who experienced Usher syndrome types I, II and III. The research study aimed to explore the questions; what do people with Usher think about having Usher syndrome? What is the effect of change on the lives of people with Usher? What do people with Usher remember of their diagnosis and what impact did it have on them?

Ellis and Hodges outlined that at the centre of this report was the "voices of people with Usher themselves, telling their own stories" (Ellis and Hodges, 2013 p16). Whilst Ellis and Hodges use the term "voice", within dual sensory loss environment such as deafblindness, having a 'voice' means that people need to be heard, although it may not actually be their voices they use to communicate. For example where a person is culturally Deaf and British Sign Language (BSL) [6] is their first language (see Chapter 1), voice may not be used but rather a visual sign communication method. The report noted that whilst adverts were placed in a variety of settings e.g. Talking Sense [29], Deafblind UK [30], Social services, Teachers of the Deaf, flyers at conferences, professional contacts, the authors' own university website and Ear Foundation [31] letter etc., participants for the study were primarily recruited via two avenues. The first was through social media, which was through personal contacts on Facebook groups about Usher, and the second was through

websites, specifically a website for “RP Fighting Blindness” [32] (Ellis and Hodges 2013 p40). Ellis and Hodges (2013) highlighted:

> “*the fact that this group (RP Fighting Blindness) rather than Sense appeared to be an initial point of contact for some people may indicate they see themselves as visually impaired rather than as having a dual sensory impairment*” (Ellis and Hodges, 2013 p40).

As these participants were primarily visually impaired/blind people, their main method of communication would most likely be speech; therefore the reference to ‘voice’ could be appropriate.

The methodology was a qualitative case study, with data collection through semi structured interviews and focus groups. The participants completed a Self-Image Profile (SIP) tool, which invited individuals to rate on a scale of 0-6 how they felt in certain categories e.g. confident, happy etc. was used. A SIP provides standardised (though non-standardised in this report as delivered in formats which included BSL) measures of self-reported self-image and esteem (Ellis and Hodges, 2013). The use of the SIP will be considered later in the chapter.

Ellis and Hodges postulate that face to face method of interview was most appropriate as the participants’ sight loss had to be taken into account and a written form of interview such as a questionnaire may be “problematic” (Ellis and Hodges, 2013 p36). Two participants used BSL /SSE [33]seven BSL, two Hands on BSL and 31 spoken verbal English. Interviews were recorded and transcribed with nearly all participants who were interviewed in BSL/SSE being recorded on video, along with the interpreters. Where signing participants chose either not to be videoed, or interviewed in a public space for example a café, the interview was recorded using a voice recorder. This aspect of data collection was somewhat confusing as BSL/SSE

is a visual language and as such interviews would need to be video-recorded to ensure transcription was accurate. Recording audibly only may affect the accuracy of the interview content as recording audibly and visually enables cross reference of data collected.

Ellis and Hodges's (2013) rationale for adopting a case study approach was that as they were looking at each person as an individual, "through looking at a range of people, a number of different aspects can be explored" (p35). It appears therefore that they viewed each person as a case. However, it was unclear which case study model was followed; they referred to Stake (1995), which is one of the case study theorists, but they gave no further detail about the design.

Gerring suggests the more case studies there are, the less intensively they are studied, therefore usually "a dozen cases or fewer" (Gerring, 2009 p22) are sufficient. However, Ellis and Hodges (2013) included 42 participants (Table 2.7) all of whom knew they had Usher. Ellis and Hodges (2013) justified a case study approach because they were:

> "*looking through the lives of individuals to gather exploratory information about the way in which they, each individual responded to the challenge of Usher syndrome*" (Ellis and Hodges, 2013 p35),

However, smaller participant numbers, studied in more depth, could have been more suited to this type of study. However, Yin (2009) highlights that a case study approach to research can contribute to providing an insight into causal relationships across a larger population, which appears to have been Ellis and Hodges's (2013) aim.

Ellis and Hodges identified eight themes: 1) The impact of diagnosis on lives; 2) Types of Usher; 3) Mobility; Home, School, public transport, mobility; 4) Communication; Family friends and support networks (including parenthood); 5) Education; 6) Employment; 7) Leisure activities and 8) Self-image profile. Within each theme they discussed change, predictability and diversity. However the specific type of thematic analysis was not explained. Ellis and Hodges did not include anyone under the age of 14 years as "due to the sensitivities of the research it was felt inappropriate" (2013 p39), or people past retirement (reasons for this were not specified). Ellis and Hodges identified that further research should include older people of 70s, 80s and beyond. They also suggested action research to involve people more in research; exploration of gene therapy and retinal implants; finding communities where there are more people with Usher syndrome and collaboration with international colleagues for further research.

Whilst the study included some aspects of the 'impacts of diagnosis for people with Usher syndrome', it is clear that further research needs to be conducted as the report covered a range of themes superficially as opposed to fewer themes in greater depth. There are areas that require further research including greater exploration of early life before Usher, experiences after diagnosis and life with Usher over the life course. The researchers' academic backgrounds were sociology of children (Ellis) and sensory education and learning styles in deafblind children (Hodges). If the research had been conducted by researchers from professional backgrounds such as social work/nursing the methodology and general approach may have been different, thus leading to different findings (Patton, 2002). Next support and intervention will be considered.

2.3.4.2 Support and Intervention for people with sensory impairment

With regard to support and intervention seven research studies will be considered: Damen *et al* (2005), Martens *et al* (2014), Prain *et al* (2010), Prain *et al* (2012), Hadidi and Khateeb (2014), Argyropoulos and Thymakis, (2015) and Kyle and Barnett (2012). However, only Damen *et al* (2005) and Kyle and Barnett (2012) included UK participants. These articles span a variety of sensory losses and a range of settings (Table 2.7). Several aspects relating to support and intervention will be considered in this section, for example, changing needs as condition worsens, especially as people get older; effect support produces; impact of specialist sensory trained staff; social support and support in terms of education and training.

Damen *et al* (2005) investigated three domains: 1) access to information, 2) communication and 3) mobility by following the Nordic definition of maintaining independence in deaf-blindness [34]. Damen *et al* suggested people with Usher syndrome face a "special set of challenges in order to maintain their independence when their sight and hearing worsens" (Damen *et al,* 2005 p309) and therefore conducted a quantitative questionnaire survey relating to maintaining independence for deafblind people, in order to better meet their needs. Whilst the authors were based in the Netherlands, the research was carried out in a number of European countries.

All three types of Usher were considered (types I[35], II[36] and II[37]) in Damen ***et al***'s (2005) study. The results revealed that whilst all participants had Usher, so therefore experienced hearing and sight loss, levels of loss differed with Usher type I patients reporting significantly fewer difficulties as a result of retinitis pigmentosa (RP) than those patients who had Usher type II. Participants (Table 2.7) were from seven

European countries (France, Germany Ireland, Italy Spain, UK and the Netherlands) and each questionnaire was provided in the participants' chosen language. Participants were supported to complete the written questionnaire by family and friends. The language barrier between countries was not discussed. The variables in this study were identified as age and type of Usher, considered hearing loss and the number of retinitis pigmentosa-related sight problems. Usher type was focused on as opposed to age with 60 patients being Usher type I, 25 patients Usher type II, four patients Usher type III and four patients type unknown. The four patients with type unknown were excluded from the study. Participants were accessed via the Charge Association Usher Syndrome Europe (CAUSE) project[37]. Questions were based on 3 main components 1) general personal information 2) clinical information and 3) information about independence. The data were analysed using SPSS[38].

Damen *et al* (2005) reported that three key findings were: 1) Usher type I patients tend to need more help than Usher type II patients; 2) the amount of help needed increases when patients get older/hearing loss worsens, and 3) no patterns in results were seen for the number of retinitis pigmentosa related sight problems. The conclusion was that people with Usher syndrome meet with particular challenges in maintaining their independence as their sight and hearing worsens, so they need equipment and help to remain independent especially as they get older and they fear not knowing when an emergency situation occurs at home.

Damen *et al*'s (2005) study confirmed what might be expected, for example, that people with Usher type I need more help than Usher type II. As people with Usher type I are born profoundly d/Deaf, with little or no residual hearing, losing their sight as well has a disastrous impact, as they are unable to see or hear thus they would need more help. People who have Usher type II however have reduced hearing but

are not born d/Deaf so they would most likely have some residual hearing, therefore, they may have some hearing to support their level of sight.

It was also unsurprising that people need more help as they get older e.g. over 65 years of age will experience age related sight (presbyopia) and hearing (presbycusis) loss (Hearing Link UK, 2015) and so, as their sight and hearing worsens, they will need more assistance. Damen *et al* (2005) identified the need for further research with regard to providing support for people with Usher to enable them to remain safely independent. A positive aspect of this paper was that the abstract was written in four languages, English, German, Spanish and French, thus enabling a wider range of audiences to read the abstract and raising awareness of Usher in a number of languages.

Martens *et al's* (2014) study evaluated the usefulness of the Intervention Model for Affective Involvement (IMAI) to participants who are congenitally[35] deafblind and their communication partners in different settings and different interactional situations.The aim was to explore whether the intervention increased affective involvement between the participants and their communication partners and whether it increased positive emotions and reduced negative emotions in the participants. Martens *et al* (2014) proffer that affective involvement, or the reciprocal sharing of emotions, is vital to control emotions and develop secure attachment relationships.

In Martens *et al's* (2014) study, there were four participants (Table 2.7) who were all congenitally deafblind. The data were collected over 20 weeks of observing interventions of communication partners' interactions with participants. The findings highlighted that affective involvement increased for three participants, while all four participants showed an increase in positive emotions and a decrease in negative emotions. The findings demonstrated that the IMAI can be successfully applied to

people who are congenitally deafblind, and highlighted that support and intervention are essential for people who experience congenital deafblindness, as their disability affects physical, mental and communicative development and affects ability to live independently. The researchers' perspectives included psychology, special educational needs and quantitative social research.

Prain *et al* (2010) conducted research with people who were deafblind, observing the behavior and interactions of adults with congenital deafblindness living in community residences. The purpose of the study was to examine the nature of interactions between adults with congenital deafblindness and the staff who provide their support, and to investigate the dependability of an observation coding system which was originally designed for observing adults with severe intellectual disability. The aim was to investigate the form, frequency, and duration of interactions between adults with congenital deafblindness and their support staff.

Prain *et al* (2010) proposed that people who experience deafblindness as a result of their dual sensory loss require specialist support services in educational and social settings to meet their needs. They outline that there is a distinct difference between a person who experiences congenital deafblindness and one who experiences acquired deafblindness, the reason being that when a person acquires deafblindness they will most likely already have learned age appropriate linguistic and cognitive skills. Thus whilst 80% of deafblind people will have acquired deafblindness, the remaining 20%, require greater specialist intervention and support as their linguistic and cognitive skills will be limited.

The authors advised this study was part one of a two phase mixed method research study (see Prain *et al,* 2012), the first phase being quantitative and the second (which will be considered later in this section) qualitative, however these studies are

reported as two separate studies. Data collection was observations which were filmed and coded (Table 2.7). The coding system used was Cohen k[36] with the entire research team receiving training regarding this system. There were two observers coding the participant's behaviours as they communicated through non-verbal means; one observer was a qualified speech pathologist and the other a psychology honours student. The data were analysed using SPSS.

The results indicated that there were extremely limited interactions between the residents and staff of two group homes for adults with congenital deafblindness. The participants were predominantly observed to be disengaged, with few observations of engagement according to the coding (Cohen's k[36]) schedule's definition of engagement. Interactions between the residents and support staff were rare. The authors concluded that interventions needed to be designed for staff to promote resident engagement in social interaction. The conclusion was that interventions need to be designed to improve and promote resident engagement within their community residences.

Whilst Prain *et al* (2012) refer to their next study as the second phase of their two phase study, as highlighted earlier; they are reported as two separate studies with the second study aiming to gain greater insight into the perspectives of disability support workers and their interactions with adults with congenital deafblindness. Prain *et al* (2012) posit that most previous research studies investigated interactions between children who are congenitally deafblind, their parents or their educators, thus more research needed to be conducted with adults with congenital deafblindness and people they interact with. This phase of the study only focused on support workers whose method of communication with the deafblind clients living in supported housing was primarily non-verbal spoken English. The support workers were asked about their perspective of their interactions with the congenitally

deafblind residents. The methodology was grounded theory with data collected using open ended Interview questions and data analysis conducted using Charmaz's (2006)[39] approach (Table 2.7). Three key themes emerged: 1) the construction of client happiness (the way clients express happiness important to worker); 2) the rationalisation of client disengagement (e.g. clients need time to relax) and 3) imperatives of the staff role (main focus personal care and domestic chores). Prain *et al* (2012) concluded these findings explained the reasons for staff behaviour in their interactions with adults with congenital deafblindness.

As a result of their findings, Prain *et al* (2012) suggested a need for staff policy and procedural documents to be explicit about the importance of social interaction between staff and clients. A final aspect for consideration is that both stage one and stage two of Prain *et al*'s (2010, 2012) research were conducted in Australia; further studies from a UK perspective would be beneficial. Having considered support and intervention in relation to deafblindness the next study relates to visual impairment.

Hadidi and Khateeb's (2014) quantitative study aimed to investigate the availability of social support systems for adolescents with visual impairments (Table 2.7). The data collection instrument used was an Arabic version of the Multidimensional Scale of Perceived Social Support (MSPSS). The MSPSS is used to measure perceptions of support from family, friends and significant others (Zimet *et al,* 1988). Zimet *et al* (1988). The tool's creators assert that it contributes to internal and test-retest reliability, good validity, and is a fairly stable factorial structure which can be translated into different languages. This is highlighted as it was the Arabic language version that was used.

Hadidi and Khateeb found that the level of social support reported by adolescents with visual impairments was higher than that reported by sighted adolescents. In all

three domains of the MSPSS (family, friends, and significant others), students with visual impairments reported receiving more social support than those without impairments and thirdly there was no statistically significant effect for gender. Hadidi and Khateeb suggested that inequalities in experiences of adolescents with visual impairments in developed and developing countries may be attributed to differences in cultural values as well as to the perception of support by those involved.

It could be reasoned that visually impaired adolescents would receive greater support than their sighted counterparts due to their sensory disability. A sensory impairment, whether it is sight, hearing or both, will affect the level of support that a person requires to contribute to safe orientation, sensory appropriate learning needs, employment etc. However despite this anticipated outcome, Hadidi and Khateeb proffer these findings are inconsistent with other Western world focused studies that have been conducted (Bruce *et al,* 2007, Cimarolli and Wang, 2006). Their study populations are the least studied group in the world and only "five percent of studies published were related to visual impairment" (Hadidi and Khateeb, 2014 p417). Clearly further research is needed in undeveloped and developing countries to ensure support, services and intervention are provided to better meet the needs of adolescents who experience visual impairment at a time in their lives when they need to achieve their developmental milestones and reach their full potential.

The aim of Argyropoulos and Thymakis's (2015) study was to examine if a child with multiple disabilities and a visual impairment would achieve better educationally if equipped with support, training and technology to meet their needs. The researchers aimed to investigate children with visual impairment and motor disabilities (impairment to moving and manipulating objects e.g. difficulty clicking on computer mouse). This area of research was considered essential as often these children are

integrated with children who experienced broader multiple disabilities and visual impairment in the educational learning environment. Argyropoulos and Thymakis proposed that children often learn from their environment and where children experience a visual impairment this hinders them from this incidental learning and this, combined with their motor disabilities, creates a unique set of challenges. The study intention was to investigate if providing support by developing keyboarding skills of a student with multiple disabilities and visual impairment via adaptive technology made a difference.

The approach to this study was action research (Table 2.7). As action research is a process of systematic inquiry that assists researchers in finding effective solutions to real problems encountered in daily life (Stringer 2014) this approach was appropriate for this study. The people involved in the study consisted of two groups: the action group included a special education teacher and an expert who specialised in adaptive and assistive technology and was visually impaired themselves; the validation group included special education research methodology specialists.

The findings indicated that students with disabilities who are well equipped and receive up-to-date training in the most appropriate technology to meet their unique needs will have a better chance of achieving their educational and vocational goals. Furthermore, where additional support is instigated for a person who experiences sensory difficulty, generally, that person will developmentally improve as they have sensory tools to support their disability. Although the research study was limited in scope with only one participant taking part, the findings are useful to understand what it is like for an individual to live with multiple disabilities and visual impairment. Additionally, the study was conducted in Greece and similar research in the UK, but with larger participant numbers, could offer a wider perspective.

The next study included was funded by the Big Lottery and prepared by Deaf Studies Trust[42] and Sense[28] (Kyle and Barnett, 2012). The study aimed to examine the experience of being Deafblind, determining if this was similar to Deaf people's experience and discovering if Deaf and Deafblind people could work together (Kyle and Barnett, 2012). The report primarily focused on people who experienced Deafblindness and especially those who had acquired blindness after being Deaf[15]. The participants were referred to as 'Deafblind' with a capital 'D[15]' throughout the report, indicating that the participants were culturally Deaf, and then acquired blindness. The references to Usher were brief, relating to quotes from four participants who experienced Usher syndrome. The first participant referred to their feelings the first time they attended a Deaf Usher weekend, the next explained how their confidence was affected at a party where they were ignored after explaining they had Usher; the third commented how they would like to have normal vision but at the same time they accept they do have Usher so try to be positive, and the fourth person expressed how having Usher can affect friendships. Outside of these four quotes, participants referred to themselves as Deafblind.

The research design was a two phase mixed method study to explore the experiences of people who are Deafblind (particularly in comparison with that of Deaf people) and to test a framework for interaction and support by Deaf people for Deafblind people. A range of communication approaches were used with the Deafblind participants (Table 2.7). Interviews with Deaf people were conducted in BSL. Deafblind researchers for Deafblind participants were provided and Deaf researchers for Deaf people. Kyle and Barnett asserted that the Deafblind researchers helped to form the questions and the methodology (Kyle and Barnett, 2012 p6).

Kyle and Barnett's findings explored the effect support produces when Deafblind people receive informal and formal support. For example they reported that Deafblind people benefited from meeting others who have similar experiences as it increases confidence. With regard to formal professional support, Kyle and Barnett highlighted that although there is a range of support provided, for example, support for Deaf children born into hearing families, counselling for those with have difficulties adjusting to changing sensory needs, and the health of Deaf women, these services are hearing-led, not Deaf led. Thus support is less Deaf aware. They also found that formal care support systems within the Deaf community are not offered to support Deaf people who are at risk, for example, older people. Kyle and Barnett further commented that:

> "*Deafblind people, Deaf people have been considered mostly as recipients of service rather than providers of service. The Deaf community is not empowered to support others*" (p13).

Kyle and Barnett also found that people with Usher needed support with regard to independence, and if they were to seek employment they would need the right services and equipment. They also mentioned benefits of communicator guide provision and services to support a Deafblind person's independence. However, the authors highlighted that for some Deafblind people a guide could be a barrier to independence and suggested further research was required to explore this. Another finding was that although there are Deaf staff members who currently support Deafblind people in services at Sense there has been:

> "*No attempt to date, to try to understand the community relations and the shared experiences which could support a more coherent interaction, particularly within the context of service provision*" (p15).

This finding suggests a need for support in terms of education and training.

The authors identified six themes which were; Deaf people's views on contact (lack of awareness, communication); Deaf views on community (Deaf identity, Deaf community); Dealing with others, (benefits of contact with Deafblind people); Personal circumstances of Deafblind people (independence, environment, being able to make choice); Feelings and hopes (employment, confidence, confidence from others); and other people (Deaf and Deafblind).Whilst these themes are pertinent to culturally Deafblind people, as was highlighted within the report, they are also applicable to people with Usher.

2.3.4.3 Psychological impact for people with sensory impairment

There were five studies that explored psychological impact for people with sensory impairment (Ellis and Hodges, 2013, Côté *et al*, 2013, Wahlqvist, *et al,* 2013, Sheppard and Badger, 2010, Högner, 2013).

As highlighted earlier Ellis and Hodges used a SIP tool and found that people with Usher were "not usually pessimistic" (Ellis and Hodges, 2013 p7) and that they were "doing things they wanted to do, either despite, or occasionally because of, Usher" (Ellis and Hodges, 2013 p7). Whilst Ellis and Hodges acknowledge that at the time of conducting an assessment using SIP, this is merely a snapshot of a person's life, a recent advisory report by Sense entitled 'I fear for my future' (Sense, 2014a) highlighted that deafblind people are becoming marginalised as a result of becoming increasingly isolated and excluded within society. For deafblind people or people with Usher syndrome marginalisation relates to the challenges they face with regard to communication, living independently and safe orientation (Sense, 2014a). Whilst some people with Usher endeavour to have a positive outlook, Ellis and Hodges's (2013) findings highlighted that the reality of living with dual sensory loss is one of

great complexity with regard to living safely and practically on a day to day basis and can affect mental health and well-being.

Côté *et al* 's (2013) aim was to explore how people with Usher type II set and manage personal goals by comparing their results on the meaning of life, serenity, self-determination and goals process (Table 2.7) by using mixed methods. People with Usher type II managed these goals by: 1) goal setting (evaluation of personal desires, negative thoughts); 2) goal planning (to overcome obstacles) and 3) goal pursuits (concrete actions and adjustments). The authors posit that it is well known that as Usher syndrome type II progresses, people experience difficulties in adjusting to change, which can have a psychological impact, they suggest that "one of the factors that may affect the adaptation process is the ability to redirect one's life and set new goals for oneself"(Côté *et al*, 2013 p139).

The researchers initially conducted a pilot project which was spread over two years to show the feasibility of the study and they found that the qualitative data indicated positive effects, as the participants "were able to set, plan and pursue a goal" (Côté *et al*, 2013 p141). Of the fourteen people approached to take part, seven participated. The participants (Table 2.7) received services from Institut de réadaptation en déficience physique de Québec (IRDPQ), had Usher II, lived in Quebec City metropolitan area and were able to communicate in group situations. People with high levels were excluded. To determine high levels of stress Ifelds Psychological Symptom Index (1978) was used and people with high levels were excluded. This tool was adapted by the Quebec Health Survey (which provides information to improve programs and services to meet Quebecers needs), and identifies internal consistency and satisfactory construct validity with scores above 26 being deemed high levels. The researchers recognised that as this was an experimental study, one limitation was there was no control group and this may

have weakened the design. Additionally the small participant number and sampling variability reduces the likelihood of obtaining significant results on the different measurement scales. Data were analysed with SPSS because four independent variables were measured: “meaning of life; serenity; self-determination and goals process” (Côté *et al*, 2013 p143).

Côté *et al* (2013) identified three key results: 1) Intervention (rehabilitation program) partly affected the meaning of life, but not serenity, self-determination or goals; 2) being forced to retire increases stress and a decline in physical well-being, life satisfaction and adaptation and 3) gradual decline in vision had impact on well-being, self-determination and goal realisation process. Côté *et al* (2013) concluded the study was useful because it focused on a better understanding of the impact of managing personal goals in persons with deafblindness, and especially their adaptation process, which was considered an important issue in clinical practice with people who have Usher syndrome. This study was conducted in Canada and similar research within the UK could be useful. The next study reviewed again considers the psychological impact of Usher syndrome, specifically relating to Usher Type II (Wahlqvist *et al,* 2013)

The aim of Wahlqvist *et al’s* (2013) Swedish study was to describe the physical and psychological health of persons with Usher syndrome type II and to explore any differences in terms of gender. The authors suggested that disability in general is difficult to research as each person, whilst being diagnosed with a similar condition, will vary in terms of level of disability and by the person’s individual characteristics. The article considers the effects of comorbidity (the presence of more than one disorder e.g. sight and hearing loss) in relation to the psychological impact it would have on the individual concerned. Wahlqvist *et al* (2013) highlighted Usher syndrome is the leading cause of deafblindness in adults of working age. The

research approach was quantitative with people |(n=96) being selected from the Swedish Usher database (Table 2.7). The participant's health, living conditions, and social relationships were included in the questionnaire. The Usher syndrome II participants were compared with those of a reference group of 5,738 persons who were drawn from a random sample of the Swedish population retrieved from the Swedish Public Health Institute.

The data were analysed using frequencies and logistic regression analysis of significant differences in poor physical and psychological health among the participants with Usher type II and the reference group. Two results emerged, the first being that participants with Usher type II rated their physical and psychological health as significantly poorer than that of the Swedish reference group. They revealed major problems involving headache, fatigue, depression, suicidal thoughts, and suicide attempts. The second result was that male participants experienced greater psychological differences than men in the reference group, for example the men with Usher were six times more likely to attempt suicide (Wahlqvist *et al,* 2013). The variances observed between men and women in the reference group with regard to higher incidence of physical and psychological health were not observed in the Usher group. However when comparing the women in the reference group and the women in the Usher group, the latter reported poorer health. As the study was comparing questionnaire results from 96 people compared to 5,738 this statistically is likely to show differences as one group is so much larger than the other (Wahlqvist *et al,* 2013). There is no indication of controlled comparisons with, for example, age/sex/socio economic group pairings/matches from larger dataset groups.

Wahlqvist *et al* (2013) concluded that the identification of factors associated with physical and psychological health and well-being are important for the design of

future rehabilitation strategies for people with Usher type II and that special focus must be placed on the psychological well-being of men with Usher type II. It is not surprising that the male participants who experienced Usher type II in the study experienced greater psychological difference than their counterparts who did not experience Usher, because, as has already been highlighted, having to cope with either D/deafness or blindness individually is a challenge, but when it is combined the complexity increases.

Wahlqvist *et al* (2013) rationalised the reason for their study was that there had been no research considering the differences in the health of men and women with Usher type II. They cited Sadeghi ***et al'***s (2006) study, which investigated the long-term visual prognosis for persons with Usher type I and Usher type II and found that progression was faster among men than among women. Wahlqvist ***et al's*** (2013) findings that psychological impact was greater for men than for women was therefore interesting and an area that requires further research. Wahliqvist *et al* suggested that although all genders with Usher are at greater risk of suicide, men with Usher are more at risk, which raises concern as suicide is a public health issue. The greater risk of suicide could be because Usher is a condition in which individuals encounter daily challenges. The study was conducted in Sweden but would be pertinent in any country where people experience comorbidity or indeed Usher type II.

The authors are from a rehabilitative background and as such posit that appropriate rehabilitation would improve the psychological wellbeing of both men and women with Usher type II and especially males with Usher type II. It could be argued that rehabilitation is not appropriate for people with progressive conditions because their condition is constantly changing, but the authors were referring to it, in terms of

"access to information, and truly interdisciplinary teamwork" (Wahlqvist *et al,* 2013 p217) which would be applicable.

Högner's (2015) study conducted in Germany aimed to assess stress in people with Usher syndrome type II and the influence of personal variables such as age, gender, and employment on stress. Due to their dual, sensory impairment, Högner commented that people with Usher syndrome are assumed to have a high risk of stress. The research approach was quantitative (Table 2.7). Two questionnaires were used, one being a self-developed questionnaire (SQ) to investigate the frequency and intensity of stress by external stressors within six life domains and the other was the German standardized stress questionnaire Trierer Inventory of Chronic Stress (TICS). These two questionnaires were used to compare frequency of stress between the Usher type II sample group and a reference group. Of the 604 people in the reference group, who were described as being from the Trierer Inventory of Chronic Stress (TICS) population, 314 were female and 290 were male, ranging in age from 16 to 70 years. The analysis method used was factor analysis. The study results were that the greatest stress was seen in the factor orientation and mobility. The second result was that the Usher type II sample showed significantly higher stress on scales that indicate a lack of social-emotional need fulfilment and less stress in those with high expectations.

The results identified that, where a person has difficulty with orientation and mobility, this will increase their isolation whilst decreasing their independence thus limiting autonomy and choice (Högner, 2015). As highlighted earlier in this section, Sense (2014a) too produced a report entitled 'I fear for my future', which highlighted deafblind people are becoming increasingly isolated and excluded within society as a result of reduced limited mobility and safe orientation. Högner (2015) concluded that the results indicate the need for rehabilitation arrangements to reduce stress in

people with Usher type II, especially in the areas of orientation and mobility, chronic worry, and social isolation and in intervention, particular attention should be paid to people who are older, female, and unemployed. The findings relating to women with Usher having higher stress levels than men contrast with those of Wahlqvist *et al* (2013) as their study indicated men were more at risk than women. The authors posited that women already have higher stress levels because they have greater health awareness, more familial expectations in terms of domestic and household duties and have to "prove themselves more than men do in the workplace" (Högner, 2015 p185) and these difficulties, when combined with Usher, increase stress levels. However, Wahlqvist *et al* (2013) and Högner (2015) agree that the results indicate a need for rehabilitation: Wahlquivist in terms of access to information and interdisciplinary teamwork and Högner (2015) in terms of reducing stress in relation to orientation and mobility, chronic worry, and social isolation.

Sheppard and Badger (2010) explored the lived experience of depression among culturally Deaf adults. The aims were to describe depressive symptoms experienced by Deaf adults, describe the American Sign Language (ASL) signs and phrases that express the Deaf adult's perspective of depression, and describe commonalities within the experience of depression among Deaf adults. The research methodology was qualitative and adopted hermeneutic phenomenology (Table 2.7). Sampling was purposive as the researcher sought participants who could express their experiences in American Sign Language (ASL). The interviews were videoed to capture participants' facial expressions and body language. Whilst the researchers do not indicate the exact nature of analysis they advised that analysis was ongoing through the interviews.

Two themes arose, the first being that early emotional chaos, feeling depressed, reaching out and Deaf-belonging as children occurred because there was no

common language within the family. Everyone interviewed was the only Deaf person within the family. The second theme was that nurses must recognize the difficulties experienced by Deaf patients, especially in communication. Sheppard and Badger (2010) concluded that culturally Deaf adults (those who communicate primarily in American Sign Language and view themselves as members of a unique culture) are rarely screened for depression and that few Deaf adults receive the mental health care they need even though symptoms of depression among Deaf adults are no different from symptoms experienced by hearing people. A contributory factor for this may include that communication barriers often make it difficult for people who are Deaf to discuss their symptoms of depression with nurses and other health care providers, therefore, few Deaf people receive appropriate treatment for their depression.

The study highlighted significant psychological impact experienced by the Deaf individuals as when interviewed for this study they described feelings of isolation and that parents expected their children to learn to lip-read, and were often upset when the Deaf child wanted to learn ASL. The study also noted that, where sexual and physical abuse occurred, the Deaf individual did not know how to report it or get help and as a result the Deaf children grew up feeling flawed. The study highlighted that as adults, the Deaf individuals were stared at or laughed at with many feeling hopeless and attempting suicide as a way to escape.

The researchers were clearly Deaf aware and used appropriate terminology and language to indicate that the research was conducted with culturally Deaf persons e.g. using capital ‘D’ Deaf. An interesting finding was in relation to the description of ASL signs and phrases that express the Deaf adult’s perspective of depression. Sometimes within BSL or ASL there may not be a specific sign for a word, for example the word phenomenon, would be translated as ‘strange happening’.

As was highlighted by Argyropoulos and Thymakis (2015), a hearing child is exposed to incidental language and understanding from the day they are born. This would include not just speech and language from their parents, but school, their peers or television. When a person is born profoundly D/deaf, unless they encounter the incident personally, they may not be exposed to the relevant language for an incident. Therefore if a D/deaf person were to be the victim of abuse they may not have the language to initiate requesting assistance with regard to what is happening to them.

Whilst this study was conducted from a nursing profession background, these findings are transferable to other professions, including that of social work as appropriate language and communication are essential for all D/deaf people and those experiencing other sensory needs. To ascertain their experiences and better meet their needs, further research would be needed to explore appropriate communication and professional awareness for not only culturally Deaf adults, but all people with sensory needs when screening for depression. With these factors in mind the next theme to be considered is that of safety and risk.

2.3.4.4 Safety and risk for people with visual impairment

There were two studies within this theme (Kim *et al,* 2014, Brook*s et al,* 2014). Kim *et al* 2014 considered the Influence of ambient sound fluctuations on the crossing decisions of pedestrians who are visually impaired and implications for setting minimum sound levels for quiet vehicles. Following the Pedestrian Safety Enhancement Act 2010 in United States of America (USA), the National Highway Traffic Safety Administration set up a minimum sound level for hybrid and battery electric vehicles. This study investigated the timing and performance of street-

crossing decisions by pedestrians who are visually impaired (that is, those who are blind or have low vision) on selected (intersections) roads.

The methodology was a quantitative observational method with repeated measures t-tests being used for pairwise comparisons (Table 2.7). There was one sighted/hearing experimenter to compare findings. Two key results emerged, the first being that at the residential intersection, the percentage of risky crossing decisions by participants was significantly lower when the decisions were made at lower background noise levels than when they were made at higher background noise levels. The second was that participants were able to make significantly fewer risky crossing decisions during the windows of time when the background noise level was lower at the major and minor street intersection as well.

Kim *et al* (2014) concluded that participants were often able to take advantage of the troughs in ambient sound for making street-crossing decisions, and the decisions made in lower ambient sound level conditions were generally less risky than those made if higher ambient sound level conditions. This was an interesting article that promoted critical reflection. This study was conducted in the USA with people with visual impairment and clearly indicated that sensory loss impacted safety and risk. However, with dual sensory loss of sight and hearing, impacts on safety and risk are even greater. Crossing the roads and safe orientation are just one challenge for people with Usher thus further research is needed to investigate life experiences and ascertain where risk occurs and how best to meet needs to address safety. The next area of safety and risk to be considered is that of substance dependence among people with visual impairments (Brooks *et al,* 2014).

Brooks *et al*'s (2014) research conducted in the USA explored the correlation of substance dependence among people with visual impairments. The purpose of the

research was to examine whether substance dependence among people with visual impairments may be associated with disability related characteristics, physical or sexual abuse history, tobacco use, and sociodemographic characteristics. The methodology was quantitative (Table 2.7) and data collected using questionnaires. Data were analysed using descriptive statistics and chi-square analyses, multivariate, binary logistic regression analysis and SPSS. The participants were recruited from the state's rehabilitation agency for people with visual impairments. There were three key results, the first being that people with visual impairments share some of the same risk factors for substance dependence as the general population. Secondly, the likelihood was greater for males, those who had been physically or sexually abused, and tobacco users and, thirdly, other variables generally predictive of substance use disorders, such as age and education, were not found to be significant. The study results concluded that the rather high probability of substance dependence indicates the need to study substance use disorders among those who are visually impaired, prevent and screen for such problems, provide assistance when such problems are identified, and evaluate prevention and intervention efforts.

To conclude this section Green *et al* (2006) propose suggestions for gaps in knowledge are an important outcome of a thorough review and it is often from these reviews that researchers find areas for future research. This section identified that literature relating to Usher syndrome is scant with significant gaps in literature. Once this stage has been completed the final stage is synthesis. In the next section the gaps in literature will be explored together with the rationale for the research topic of this study.

2.3.5 Stage 5 Synthesis

As discussed earlier, synthesis enables individual evaluations to generate deeper understanding and facilitates a combination of individual studies to form a generic whole (Booth *et al,* 2012, Patton, 2002). The selected papers were evaluated using CASP, as this tool assists in appraising rigour, credibility and relevance of the research studies by using a series of questions (Dixon-Woods *et al,* 2007, Attard *et al,* 2015). The literature reviewed (Table 2.7) concerned the diagnosis of, living with and experiences of Usher syndrome with four themes identified (Table 2.9): 1) Impact of diagnosis for people with Usher syndrome; 2) Support and intervention for people with sensory impairment; 3) Psychological impact for people with sensory impairment and 4) Safety and risk for people with visual impairment.

Of the 14 papers reviewed, only three were UK based; six were quantitative studies, four were qualitative studies and four were mixed methods (Table 2.7). The methodologies used in the qualitative studies included case study (Ellis and Hodges, 2013, Martens *et al,* 2014), action research (Argyropoulos and Thymakis, 2015) and hermeneutic phenomenology (Sheppard and Badger, 2010).

The literature review identified, that although there has been international research conducted (Table 2.7), in the UK, Usher syndrome is clearly an under-researched area of sensory need with only three research studies since 2005 (Damen *et al,* 2005, Kyle and Barnett, 2012, Ellis and Hodges, 2013). Within these studies, various areas for further research were identified. Ellis and Hodges (2013) identified gaps relating to; older people over 70 years; gene therapy and retinal implants; finding communities which have more people with Usher syndrome and collaboration with international colleagues. Although Damen *et al*'s (2005) study was not solely UK focused, as it included six other European countries (Table 2.7) it

identified that further research was needed with regard to providing support for people with Usher syndrome to enable them to remain safely independent and this finding is pertinent to England. Whilst, participants in Kyle and Barnett's study primarily referred to themselves as Deafblind, the findings that identified gaps relating to the relationship between Deafblind people and their communicator guides and also whether guide provision contributes to independence or becomes a barrier, could also apply to people identifying as having Usher. Of the three UK studies conducted, as highlighted earlier, only one was qualitative (Ellis and Hodges, 2013) and this study used a case study methodology.

The review identified limited previous research relating to the impact of diagnosis of Usher syndrome, with only five studies primarily focusing on Usher syndrome (Ellis and Hodges, 2013, Damen *et al,* 2005, Côté *et al,* 2013, Wahlqvist *et al,* 2013 and Högner, 2013) and one study referring to Usher with primary focus of Deafblindness (Kyle and Barnett, 2012). The review also highlighted that four studies adopted a qualitative approach (Ellis and Hodges, 2013, Martens *et al,* 2014, Argyropoulos and Thymakis, 2015, Sheppard and Badger, 2010) of which one adopted a qualitative, hermeneutic phenomenological perspective (Sheppard and Badger, 2010), and was conducted in the USA. Of the other studies, most studies used a quantitative approach (n=7), while three used mixed methods.

With regard to the four themes identified for this review (Table 2.9), whilst some research focuses on support and intervention (Kyle and Barnett, 2012, Damen *et al,* 2005, Martens *et al,* 2014, Prain *et al,* 2010, Prain *et al,* 2012, Hadidi and Khateeb, 2014 and Argyropoulos and Thymakis, 2015), this too is limited as there are only seven studies, two of which are UK based.

There were five studies that explored the psychological impact for people who experience sensory needs (Ellis and Hodges, 2013, Côté *et al,* 2013, Wahlqvist *et al,* 2013, Sheppard and Badger, 2010, Högner, 2013).This theme identified that when a person experiences Usher syndrome there will be ongoing physical and psychological effects.

As regards safety and risk, Kim *et al's (*2014) findings raised concerns relating to advancing technology, for example the impact for people with sensory impairments with regard to electric low sound cars . As technology will continue to advance, this may be an area that requires further research in an ever changing society to ensure as far as possible the safety of people with sensory impairment.

There were variations in the communication methods used within the selected research studies reviewed (Table 2.7), which highlighted the wide variety of communication methods that are used with people who have sensory needs. The methods included written communication (Damen *et al,* 2005, Côté *et al,* 2013, Wahlqvist *et al,* 2013, Högner, 2015), verbal spoken English (Ellis and Hodges, 2013, Martens *et al,* 2014 Prain *et al,* 2012, 2010, Hadidi and Khateeb, 2014, Kim *et al,* 2014 Brooks *et al*, 2014), spoken verbal Greek, Italian, and Albanian (Argyropoulos and Thymakis, 2015), non-verbal communication (Martens *et al,* 2014, Prain *et al,* 2010), American Sign Language (Sheppard and Badger, 2010), British Sign Language (Kyle and Barnett, 2012, Ellis and Hodges, 2013), deafblind manual (Kyle and Barnett, 2012), hands on signing (Kyle and Barnett, 2012) and Braille (Ellis and Hodges, 2013). Having considered the final stage of synthesis, the research questions, study aims and objectives will next be considered.

2.4 Research questions and study aims and objectives

The review of literature led to four themes being identified (Table 2.9) which highlighted gaps that informed my study focus: research questions, aims and objectives.

2.4.1 Research questions

There are two research questions which are:

1) What is the experience of being diagnosed with Usher syndrome?

2) What is life like for people who are diagnosed with, and live with Usher syndrome on a daily basis in England?

The research questions were developed to gain an understanding of participant experiences of what it was like to be diagnosed with Usher and the effects the condition had on day to day living. In the research questions the word experience was used as opposed to experiences. Although the words experience and experiences are similar, the reason the word experience was used, was because I wanted to glean personal, individual experiences from people who experienced Usher syndrome as opposed to collective experiences of people with Usher as a group. Individual experiences were essential for this research study because each person experiencing Usher will have unique and different experiences depending on their levels of sight and hearing loss and these individual experiences were considered valuable to the study.

2.4.2 Study aims and objectives

The aim of the study is to develop an understanding of the experiences of diagnosis of and living with Usher syndrome, from the perspective of adults in England. The objectives are to:

1) Explore the experience of being diagnosed with Usher syndrome

2) Explore the transition from adolescence to adulthood for people who have Usher syndrome

3) Develop an understanding of the experience of living with Usher syndrome, including support, developmental opportunities and the role of the Deaf community

4) Disseminate findings that can inform future practice, service development, policy and education

5) Recommend areas for further research relating to the experience of living with Usher syndrome

2.5 Chapter summary

Conclusions drawn from the literature review were that Usher syndrome is a rare condition with little public awareness and limited understanding of life experiences of people with this condition.

This chapter initially considered requirements for undertaking a literature review (Table 2.1). Various approaches to reviewing literature were explored, however, to support the aims of the study, a narrative structure for conducting literature reviews was selected with the 5 Ss format being adopted (Table 2.2). Whilst the primary source for this structure was based on Green *et al*'s (2006) paper, I developed this further.

Initially literature search terms were identified (Table 2.3) but as there was limited scholarly material with regard to Usher syndrome the literature search was widened to that of deafblindness, blindness/visual impairment and D/deafness. A table identifying synonyms was used to incorporate various terms that may have been used in literature when referring to sensory description (Table 2.4). As a result of utilising various databases (Table 2.5) to search for literature and creating criteria for including and excluding material (Table 2.6), two funded research reports and 12 peer reviewed research articles were selected for critical review (Table 2.7). CASP was used to evaluate papers and an adaptation of Green *et al'*s (2006) structured approach was used for writing up (Table 2.8). This method was adopted to identify four themes (Table 2.9).

So in conclusion, although the review highlighted various methods and methodologies were used to research Usher syndrome, only one study sought to glean experiences of people with sensory requirements (Sheppard and Badger, 2010) and this was with people who were Deaf, not people who experienced Usher syndrome. As the focus of my study is to gain an understanding of life with Usher by exploring life experiences of people with this condition, the next chapter will consider the paradigmatic positioning, rationale behind choosing methods and methodology, the research process, ethical considerations and trustworthiness of the proposed study.

Chapter 3 Methods

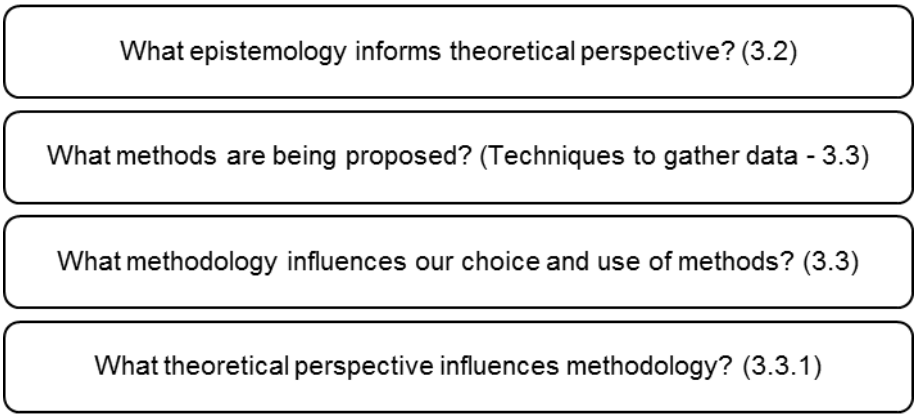
3.1 Introduction

The literature review in Chapter 2 highlighted that, although research has been conducted with people who experience Usher syndrome, internationally and in the UK, there were still gaps (Table 2.7, Chapter 2), relating to understanding the diagnosis of, living with and experiences of people with Usher syndrome. Whilst research has been conducted with people with sensory impairment (Bolt, 2005) few studies have been undertaken involving people with Usher syndrome (Damen *et al,* 2005, Kyle and Barnett, 2012, Ellis and Hodges, 2013, Côté *et al,* 2013, Wahlqvist *et al,* 2013, Högner, 2015) and only a few included participants from the UK (Damen *et al,* 2005, Kyle and Barnett, 2012, Ellis and Hodges, 2013). This chapter considers the paradigmatic positioning, rationale behind choosing methods and methodology, ethical considerations and trustworthiness of the proposed study.

The rationale for choosing a qualitative approach related to the research questions, which were, what is the experience of being diagnosed with Usher syndrome? And what is life like for people who are diagnosed with, and live with Usher syndrome on a daily basis? Also, it was essential to gain participants' narratives of their experiences in their own words. As narrative was core to this study, a quantitative approach was not considered congruent as data could not be analysed from statistical, measurement and/or numerical approaches (Cresswell, 2013, Bernard, 2013, Oakley, 1999).

When conducting research there are two essential elements to consider: The methodologies and methods as well as how these choices can be justified (Crotty, 2015). Crotty (2015) suggested that four questions arise from these two essential elements (See Figure 3.1).

Figure 3.1: Four questions arising from two elements
(Adapted from Crotty, 2015)



Therefore, the paradigmatic choice and epistemological stance taken within the research, the methodology used for conducting the research; ethical issues and how trustworthiness was promoted throughout the study are explored in this chapter.

3.2 Paradigms and epistemology

The term paradigm refers to a way of looking at the world, relates to how inquiry should be conducted and is “a broad term of encompassing elements of epistemology, theory and philosophy along with methods” (Punch, 2014 p14). The paradigm can provide a framework to give structure as it guides how we make decisions and how we conduct the research (Punch, 2014). Lincoln *et al* (2011) suggest that there are five alternative enquiry paradigms: positivism (to attain objectivity and truth); post-positivism (recognise inability to know reality with certainty); critical theory (reality is capable of being apprehended – action orientated); constructivism (reality not already established); and participatory (emphasises participation and action when looking for change). A quantitative approach following a positivist paradigm would not have been appropriate for this study as the aim was to explore the lived experiences of people with Usher

syndrome rather than testing a hypothesis, which is a prediction made about expected relationships between variables (Creswell, 2014).

As the chosen paradigm needs to be compatible with the epistemological stance, methodology, ethical issues and how trustworthiness was promoted (Lincoln *et al*, 2011), this study was conducted within a constructivist paradigm. In constructivism the researcher is involved in the inquiry as they become responsive and reflective to the participants' dialogue (Appleton and King, 1997). Rogers and Pilgrim (2005) highlight that constructivism as a dominant position within sociology holds a fundamental belief that reality is not already established and awaiting discovery, but rather is transitional as a result of our actions as humans and thus reality is constructed. Guba and Lincoln (1994) suggested that reality is socially constructed and what exists depends upon understanding of that reality (or multiple realities) and its socially produced knowledge.

Creswell (2014) suggests that constructivism or social constructivism (terminology is used interchangeably) believes that people pursue understanding of the world they live in. Creswell (2014) further suggests that the main aim of research should be to depend as far as is possible on the participants' "views of the situation being studied" (p8). These principles are fundamental to exploring experiences of people with Usher syndrome.

Constructivism has emerged as "a very powerful model for explaining how knowledge is produced" (Gordon, 2009 p39). Quale (2007) argued that "knowledge must be constructed by the individual knower" (p107), which seems relevant to people with Usher syndrome whose life experiences, are formed depending on their condition and how their experiences affect their life development. Burr (2003)

suggests that knowledge can be defined from construction of phenomena, the phenomenon for this study being living with Usher syndrome.

Epistemology is the study of knowledge and explores what constitutes justifiable knowledge (Steup and Zalta, 2011). Wellington *et al* (2011) suggest that questioning one's own philosophical stance and fundamental assumptions relating to epistemology are essential because they influence selection and usage of methods and methodology. As the stance taken by the researcher informs the methodology and thus provides a context for the process and grounding its logic and criteria (Crotty, 2015) the researcher needs to be clear on their positioning. Whilst each researcher comes from their individual epistemological position, it is natural that a research study will contain assumptions about what is justifiable knowledge, which when combined with their theoretical underpinning and research methodologies and methods produces unique research. Therefore, the researcher's epistemological position explores the way they see the world, how they consider knowledge is constructed and disseminated and how trustworthiness is promoted. The epistemological position taken within this study is that of constructivism as I believe that knowledge is constructed by the life experiences of people with Usher syndrome.

Epistemology also concerns the relationship of the researcher with the participants and questions if they stand alone as separate entities or if they are interlinked (Wang, 2010). For example within this study, the participants will have their personal experiences of Usher syndrome while the nature of my experience was professional, having worked with people who have sensory needs including Usher syndrome. Whittaker (2009) suggests that the researcher brings their own backgrounds and identity, thus it could be natural to bring our own thoughts, values, beliefs, identity, culture, race etc. with us into our research. Therefore although bias could be an

issue, our background also brings positive aspects and insights rather than just bias, therefore bias and the qualitative concept of subjectivity will be discussed further in section 3.7. Also, important is achieving integrity, which not only relates to the researcher's epistemological stance but also their justified belief (Steup and Zalta, 2010), the relationship of the researcher with the participants (Wang, 2010) and philosophical - love of wisdom (Blackburn, 2008), theoretical principles to explain phenomena and underpin assumptions, coherence and methods and methodologies (Wellington, 2011). Interrogating one's philosophical position concerning these aspects is essential to fully plan for the research study (Wellington, 2011).
When questioning one's own epistemological position it is appropriate to consider whether, with regards to knowledge, the aim is to attain objectivity and truth (positivist), to understand how and why things occur (interpretivist) or to consider reality is not waiting to be discovered, but constructed as a product of human activity (constructivism). As previously highlighted, the approach chosen for this study is constructivist.

3.3 Methodology

Methodology is the approach, plan of action or design that informs the choice and use of certain methods and connects these to the preferred outcomes (Crotty, 2015). Wellington *et al* (2011) highlighted that methods and methodologies will be influenced by certain factors, such as personal preferences, inquisitiveness, the researcher's professional background, reasons for embarkation on research, the required end result, the interview topic guide questions, ethical considerations, resources/time: "the nature of the research population and the ability of subjects to give particular responses" (p99). They further highlight that choices will be different just because we are all different people. Whilst there are a number of methodological options that could have been chosen for this study, for example

ethnography, grounded theory or case studies (Cresswell, 2003, Cresswell, 2013), a phenomenological approach was chosen as it was compatible with the research aims.

Use of phenomenology also enables in-depth exploration of complex individual experiences of a phenomenon (in this study, living with Usher syndrome) and then the analysis of the composite experiences of individual experiences to appreciate the essence of the phenomena for all individuals (Cresswell, 2007), which again was an essential element of the study.

Phenomenology has deep historical roots and was mentioned as far back as the 18th century by Johann Heinrich Lambert who was a Swiss polymath (a learned person who draws on knowledge from various different subject areas) and is derived from the Greek “phainein” which translated means, “to appear” (Priest, 2002 p51). Phenomenology has a rich tradition with phenomenologists such as Husserl, Heidegger and Merleau-Ponty and is a “philosophical approach to the study of experience” (Smith *et al,* 2012 p11). A key aspect of phenomenology is that it “provides us with a rich source of ideas about how to examine and comprehend lived experience” (Smith *et al,* 2012 p11). Initially phenomenology was used for the purpose of “solitary philosophical reflection” rather than research (Dowling, 2007 p137). However, as each theorist contributed their ideas, phenomenology as a methodology developed (Zahavi, 2003, Giorgi, 1970, Giorgi, 2012). There are two main approaches to phenomenology: descriptive (Bretano; Husserl; Merleau-Ponty; Giorgi), and interpretative (Heidegger; Gadamer; Smith). Next, both these options will be discussed.

3.3.1 Descriptive phenomenology

Franz Bretano originally coined the phrase descriptive phenomenology in the 19th century (Dowling, 2007), with Edmund Husserl adopting some of Bretano's work, for example, the concept of intentionality which is described as "the internal experience of being conscious of something" (Dowling, 2007 p132). Descriptive phenomenology as a research tradition focuses on participants' experiences from "their viewpoints as fully as possible" and aims to "capture exact and deep layers of life" (Sorsa *et al,* 2015 p9). Heaslip *et al* (2016) posit that:

> "*the purpose of descriptive phenomenological research is to describe a phenomenon and its meanings without interpretation, explanation or construction*" (p1987).

Sorsa *et al* (2015) further highlight that "descriptive researchers will create deep insights through a thorough description of a phenomenon" (p9) and focus on the participants' experiences.

Some theorists when considering the descriptive approach argue that phenomenology is purely descriptive and interpretations fall outside the boundaries of phenomenological research (van Manen 1998). Husserl suggested that people shared pre-reflective experiences and not interpretations of those experiences (Dowling, 2007). Husserl died in 1938 without completing his life-work (Carr, 1970) but other theorists such as Merleau-Ponty and Amedeo Giorgi were influenced by his work and further developed it. Merleau-Ponty focused on phenomenology with regard to arts and politics (Carman, 2008) but Giorgi (2012) explored it in relation to a psychological approach to research. Amedeo Giorgi (1985) explored the phenomenological psychological method for conducting research on human phenomena as lived and experienced and posited that the purpose of research is to

“do justice to the lived aspects of human phenomena” and to do this, “one first has to know someone actually experiences what has been lived” (Giorgi, 1985 pvii). Giorgi further suggests that once the researcher goes beyond the surface and begins to explore participant experiences in more depth, one can work with “description more rigorously and systematically“(Giorgi, 1985 pvii).

Giorgi’s approach comes from describing a situation in daily life that comes prior to any kind of reflexivity with the researcher obtaining description of whatever there is “in front of a person’s eyes and not of that thing’s existence “(Sadala and Adorno, 2002 p283). Paley (2005) suggests that if there is such a concept as objectivity it can only take place in an “interpretative free zone” (Paley, 2005 p110). It is therefore essential for the researcher to adopt a phenomenological stance that will allow them to be open-minded and live that experience as a Gestalt.

The term Gestalt was originally coined by Gestalt psychologist Kurt Koffka in the 20th century and refers to the whole having an independent existence to promote non-judgement and contribute to the openness of the description being given (Koffka, 1935). Adopting this approach places the researcher in a position where they are putting aside their own “natural assumptions about the world“(Sorsa *et al,* 2015 p9), suspending preconceptions and bracketing or placing the “phenomena in Epoche” (Sadala and Adorno, 2002 p283).

Husserl’s concept (Cresswell, 2013) of using the tool of Epoche or bracketing requires a change of attitude to enable the pursuit of philosophical inquiry (Priest, 2002) and includes suspension of bias or personal views and beliefs while analysing data (Seamon, 1979). Giorgi (1997) posits that bracketing of prior knowledge about a phenomenon is essential to be able to see the phenomenon through fresh eyes and describe it as it is being presented without preconceptions.

Seamon suggests “experiencing the action without having to act” (Seamon, 1979 p21); this does not mean that prior experiences are rejected but merely that notions are suspended. For example considering experiences of others collated during interviews and suspending own notions may result in finding that previously held ideas are questioned and fresh ideas embraced (Buttimer and Seamon, 1980). Suspension of bias or personal views, were considered to be core within data analysis to enable a descriptive phenomenological approach of taking participant experiences as a given and using participant comments when coding (Appendix C).

Smith and Osborne (2003) also considers the principle of bracketing and highlight that, the researcher when analysing data needs to suspend preconceptions in order to focus on grasping the experiential world of the research participant. Descriptive phenomenologists attempt to, as far as possible, suspend pre-conceptions when bracketing (Sorsa *et al,* 2015, Koch, 1995). The aim of bracketing is to avoid the researcher influencing the participant’s experiences or understanding of their phenomenon (Sorsa *et al,* 2015).

As the researcher is “the instrument for analysis” across all stages of a qualitative study” (Tufford and Newman, 2010 p81) some authors suggest that bracketing of pre-accumulated knowledge and understanding is a challenge (Chan *et al,* 2013, Levasseur, 2003) and others assert that it is impossible (Heidegger, 1962, Koch, 1995, Johansson and Ekebergh, 2006). However, Drew (2001) suggested that bracketing can contribute to objective description and Chan *et al* (2013) posit that bracketing can contribute to accurately describing “participants’ life experiences” (Chan *et al,* 2013 p2) and as such it could be suggested bracketing is essential for a descriptive phenomenological approach to research which focuses on the rich description of life experiences.

Paley (2005) posits that bracketing is not like “emptying your pockets” or “taking your shoes and socks off “ (p110) and questions how we can identify every conceivable preconception that may affect the findings . Whilst this clearly cannot be achieved it could be suggested that by focusing on description of experiences the researcher focuses on the lived experience of the participant as a given, not interpretations of it and thus takes participant experiences at face value. Additionally “informants are consulted and trusted” (Forsberg *et al,* 2000 p328) as the experiences they share are theirs, personal and unique to them.

Whilst bracketing or suspension of belief (Seamon, 1979) added congruency to this study it is also consistent with the practical role of the social work profession in which a worker is consistently required to suspend their own beliefs in order to practise in an anti-discriminatory manner and provide a person centred service to individuals’ with whom they are working (Parker and Bradley, 2010).

Sorsa *et al* (2015) highlighted that there is the risk of data being affected if researchers do not bracket, however, as bracketing is a conscious task, as researchers’ we need to have a strategy to practice it effectively (Chan *et al,* 2013). Chan *et al* (2013) suggest that as we are human instruments in the research process, in order to bracket effectively, we need to be aware of our own “interests, values, perceptions and thoughts” to be able to put them aside and if we are not aware of our own perceptions and beliefs then it is not possible to bracket and put aside the things that may “influence the research process” (Chan *et al,* 2013 p3). Chan *et al* suggest that reflexivity is the “key thinking activity that helps us to identify the potential influence throughout the research process” (Chan *et al,* 2013 p3). Within my study a reflexive approach was an essential element and is discussed further in section 3.7.

Whichever term is used, Epoche; bracketing or suspension of belief, the action highlights, that “a degree of open mindedness” (Smith *et al,* 2012 p42) needs to be adopted. Having considered descriptive phenomenology, interpretative phenomenology will next be considered.

3.3.2 Interpretative phenomenology

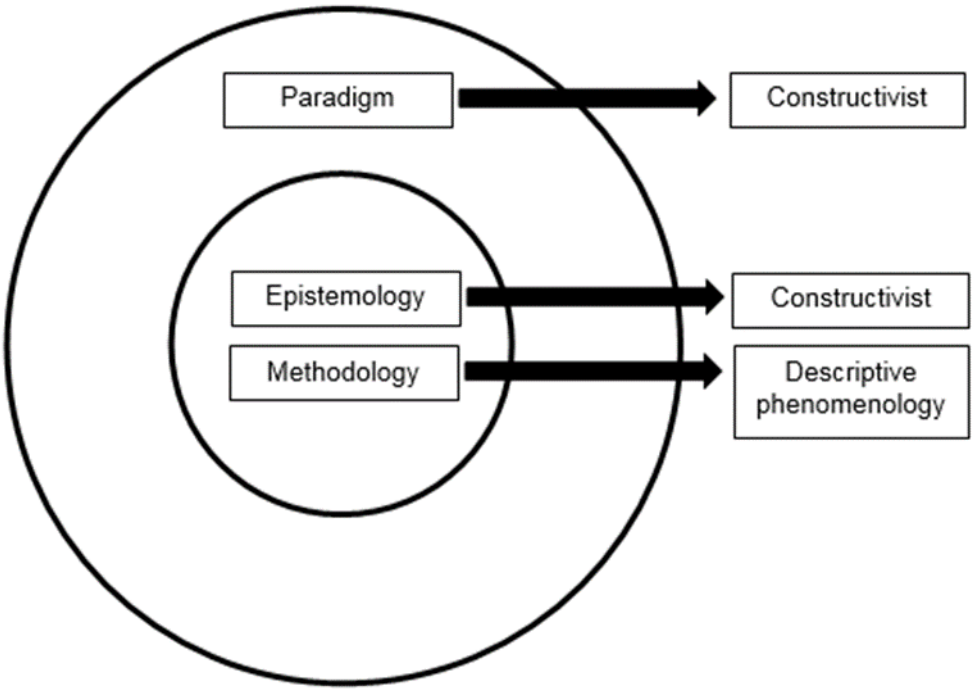
Whilst descriptive phenomenology was adopted for this study, interpretative phenomenology was also considered. The interpretative research tradition was originally based on the philosophical belief of Martin Heidegger (Heidegger, 1962), who was a student of Husserl (Paley, 1997), and built on Husserl’s work but from his own theoretical perspective, from which interpretative phenomenology and the concept of hermeneutics (theory and methodology of interpretation) developed as a research tradition (Lopez and Willis, 2004, Smith *et al,* 2012). Heidegger considered that greater depth of meaning was gleaned by interpretation which could not be achieved if using a descriptive approach (Lopez and Willis, 2004, Silverman, 2011). Interpretative phenomenology considers “research to be the sum of its parts, with both participants and researchers affecting it” (Sorsa *et al,* 2015 p9) and uses previous knowledge to “create new understandings in the research”(p10).

Next Hans-Georg Gadamer studied with Heidegger and was strongly influenced by his philosophical thinking, which inspired him to develop his own concept of philosophical hermeneutics (Gadamer, 1992). A more current theorist is Jonathan Smith who explored interpretative phenomenology further and introduced interpretative phenomenological analysis (IPA) as an approach to “qualitative phenomenological psychological research” (Smith *et al,* 2012 p11) positing it to be a methodology in its own right as opposed to a philosophical strand of phenomenology. IPA was considered as a method for this study as it “connects to

the core ideas unifying the phenomenological philosophers" (Smith *et al,* 2012 p32), however, it was excluded for several reasons. Firstly, IPA is better suited to interviews conducted with smaller participant numbers but in greater depth, often with a series of interviews (Smith *et al,* 2012) and secondly I did not want to collect a "range of possible accounts" which could be interpreted or open to corruption (Paley, 2005 p111). Whilst Paley is not referring to IPA specifically, the principle applies to interpretative phenomenology. Furthermore as the study aimed to describe and report on participant experiences and not interpret them, an interpretative approach would not be appropriate.

So to summarise, whilst other descriptive phenomenological theorists were considered (Bretano, Husserl), Giorgi's (1970, 2012) descriptive phenomenology was adopted for this research, due to its congruency with the paradigmatic position, and the topic being explored. To précis the study's paradigmatic positioning, see Figure 3.2.

Figure 3.2: Paradigmatic positioning



Next methods will be considered.

3.4 Methods

Methods are the techniques or tools that the researcher uses to gather and analyse data related to the research topic or question (Crotty, 2015). The intention for this study was to conduct interviews with 20-25 people with Usher syndrome and analyse the data using thematic analysis while addressing ethical considerations. The data collection method was interviews conducted with 20 people using a range of different communication methods. Firstly, sampling methods will be considered.

3.4.1 Sampling methods

The sampling method adopted was purposive as purposive samples are useful in aiding the researcher to better understand the problem and the research questions

(Cresswell, 2013). Purposive sampling involves the researcher choosing participants', from whom they consider they are likely to "yield useful information" (Whittaker, 2009 p35). Interviews are more successful when participants' have extensive knowledge relating to the research topic and are willing to talk about it (Whittaker, 2009) Furthermore, O'Halloran *et al* (2016) suggests that purposive sampling is compatible with a descriptive phenomenological interview in which description of a participant's lived experience of a phenomenon is required. Therefore, purposive sampling was considered appropriate as in order to address the research topic and the research questions, participants needed to have Usher syndrome to be able to express and describe their experiences of living with it.

The sensory charity Sense was gatekeeper for the study and supported recruitment. There were two Usher groups within Sense, one for Hearing and Sight Impaired (HIS) and another group who were mainly BSL users. Participants were recruited from within these groups and also from the Sense database. Whilst most of the participants were introduced through Sense (Adam, Ben, Carl, Debra, Fred, Gareth, Harry, Iris, Jeff, Kate, Len Monica, Nora, Oliver, Pam, Sara) there were four participants who were recruited via snowballing; their pseudonyms are Eve, Quentin, Susan and Tia.

This phenomenological study recruited a sample of adults with Usher syndrome across a wide demographic area to encapsulate wider participant number and diversity. The lower age limit was 16+; however, there was no upper age limit, gender, nationality or demographic restrictions. In total, 20 people were recruited and they ranged from 18 to 82 years; Table 3.1 summarises the other features of the study participants

Table 3.1: Range of participant gender, nationality and demographic location

Gender	Nationality	Demographic location
Male X 10	Asian (2)	London 6
Female X 10	British Caribbean (1)	Devon (1)
	Irish (1)	Norfolk (1)
	Portuguese (1)	Worcester/Herefordshire (1)
	English (1)	Essex (5)
	British (14)	Berkshire (1)
		Surrey (1)
		Kent (1)
		Cambridgeshire (1)
		Sussex (1)
		Gloucestershire (1)

Participants were given pseudonyms (Table 3.2) because: “anonymity and confidentiality of participants are central to ethical research practice in social research” (Crow and Wiles, 2008 p1); a pseudonym approach gives voice to individuals and also to contribute to anonymity and confidentiality throughout the thesis. Participants were advised that as far as possible their experiences would be anonymised, however, as highlighted by Ellis and Hodges (2013), when involving participants in research from small populations such as Usher there is the potential for other people to recognise them and this could have arisen within Ellis and Hodges’s (2013) study and my own.

Whilst all participants in both my own and Ellis and Hodges (2013) studies were given pseudonyms, one participant in both studies of the same age, nationality and gender used the same analogy of “*Manchester United and Manchester City*” when describing the two Usher groups; the BSL group, and the hearing and sight impaired group. This situation highlighted that in order to minimise, although not eradicate, recognition occurring, a profile of each individual would not be provided. The selected pseudonyms are referred to throughout the thesis.

Table 3.2: Participant number and pseudonyms given

Participant number	Participant pseudonym
1	Adam
2	Ben
3	Carl
4	Debra
5	Eve
6	Fred
7	Gareth
8	Harry
9	Iris
10	Jeff
11	Kate
12	Len
13	Monica
14	Nora
15	Oliver
16	Pam
17	Quentin
18	Ruth
19	Sara
20	Tia

3.4.2 Access and recruitment

Initially, a flyer was created to introduce the study idea and to ascertain if anyone would be interested in contributing. It was hoped that advance advertising and enabling people to consider participation freely would encourage a greater response.

The flyer announced a study to explore what life is like for people with Usher syndrome and was an invitation to people with Usher syndrome to participate in a research study. The flyer contained details of the prospective study, the time commitment required from the prospective participant and researcher contact details. The flyer highlighted that if the interview were face to face the researcher would travel to the prospective participant for the interview, however electronic options such as email were also offered. The flyer identified that communication would be flexible and the option to bring the prospective applicant's own

interpreter/communicator was offered. The flyer was initially circulated by the Head of Research - Sense at a NavNat Event for people with disabilities being held in Hayes. As Sense is a national charitable organisation for people who are deafblind and whose focus are to support and campaign for children and adults who are deafblind this appeared to be a sound start. However, no participants came forward.

Since conducting the research, participants who did engage in this research study conveyed that they are constantly being asked to engage in research because their condition is rare and there are few people able to engage in such research. Some participants expressed a reluctance to be involved in research because they either did not feel it would benefit them or it was enough for them to be coping with living with the practicalities of Usher and they did not have time for research studies. With hindsight, although the flyer was useful in terms of introducing the prospective study and raising awareness of it, it was unrealistic on my part to expect the number of participants required for this study to be recruited via a flyer.

In due course I contacted the Royal National Institute for Blind People (RNIB), Deafblind UK and other sensory organisations to raise awareness using the same flyer. However there were no participants recruited from this approach. Due to the rarity and sensitive nature of the condition, recruitment was very difficult. Sense as gatekeepers, were very supportive and circulated details of the study on a regular basis, but recruitment was still very slow and difficult. The original target age group was 18 – 25 years, as the experiences of young people with Usher were the focus of attention. However, this range was clearly too narrow as recruitment was problematic, therefore the decision was made to make an application to the ethics committee to widen the age range to 16+ with no upper age limit (Appendix L).

Additionally, during this time an opportunity arose to provide communication guide service support[41] to one of the members of the hearing and sight impaired (HSI) group [a peer support group for people experiencing Usher] to a HSI social event. It was following this event that members of the HSI group got to know me better and were more willing to be involved in the research study and agreed to be interviewed. It was useful that as a researcher I had sensory skills required to enable provision of the support required as this presented the opportunity to recruit more participants for the study which would not have been available to me if I did not have these skills.

The combination of the opportunity to provide communication support for a member of the HSI group and widening the age range led to improvement in recruitment resulting in participants being recruited from all over the country (Table 3.1) and aged between 18-82 years. The interviews took place over a period of 14 months. Interviewing began in March 2013 and the next interview was not until July 2013. The final interview took place in May 2014.

3.4.3 Data collection methods

The data collection method chosen was semi structured interviews as this approach whilst being open ended also follows a general script (Bernard, 2013). Kvale (2013) broadens the topic of semi structured interviews to incorporate them from a life world interview perspective in which the aim is to try to "understand themes of the lived daily world from the subjects' own perspectives" (Kvale 2013, p10) and this resonated with the aim for this study. Kvale further suggests that the interview is a "uniquely sensitive and powerful method" (Kvale 2013, p11) for capturing lived experiences of people from their everyday world.

The aim was to interview each participant once using a participant led approach in relation to the interview and communication method. It was expected each interview would last for approximately one hour, with awareness of each participant's needs being taken into account and interview duration being adjusted where needed. In practice, some participants' interviews were shorter than an hour (Quentin, Ruth, both 40 minutes), and some were longer (Kate, 1 hour 40 minutes). As the aim was for interviews to be participant led, whilst I monitored the time, if the person wanted to talk for longer the interview was not cut short. Some interviews were via email (Carl, Harry, Iris, Nora, Oliver and Tia) and therefore the time element was not relevant.

Semi structured interviews are compatible with a phenomenological methodology as they have the ability to "explore the richness of everyday life as it manifests to those who experience it" (Campbell and Scott, 2011 p6). Using a phenomenologically focused topic guide in my study meant the interview questions linked to the study aim and concentrated on gaining participants' experiences of living with Usher syndrome. The five interview questions (Appendix B) were adapted according to communication used, for example where a person communicated using BSL the word order of the question would have been altered to reflect BSL grammatical structure.

With hindsight, question one could have been more phenomenologically attuned by using the word 'experience' instead of 'report'. However, as the participants were immersed in their individual experiences, they proffered their individual unique experiences regardless of the question wording. The questions were topics for use with probes to explore responses. The focus throughout the interviews was on what people expressed as opposed to what they were asked with the overall aim of enabling participants to communicate their experiences in-depth.

A pilot study can be constructive when beginning to collect data, as this testing can establish the content validity and improve questions (Cresswell, 2013). Therefore in this study the first interview was planned as a pilot interview to test the data collection method (interviews), interview topic guide questions and learn from it to improve subsequent interviews. As the data collected were valuable to the study they were included in the analysis. Nothing was changed as a result of the pilot because the research questions worked well and provided rich data.

Flexibility was required as a diverse range of communication methods was needed, depending on when the person had lost their sight, to what degree their sight was impaired and when they had developed their D/deafness/hearing loss e.g. pre/post birth. Whilst in most cases the communication method was established prior to interview, on one occasion I was expecting the participant (Debra) to communicate using visual frame BSL but as the interview progressed, deafblind manual and hands on signing were also used thus highlighting that the communication method was participant led. Table 3.3 outlines communication methods used in interviews with brief descriptions:

Table 3.3: Communication methods used and brief descriptions

Method	Description
Clear speech	Use of clear speech - Voice not too loud or too soft and good articulation. Light and environment used to enhance sound and quality of speech
Written communication	Communication by use of the written word
Visual frame signing - British Sign Language (BSL)	BSL signing within the individual visual field of the person with Usher syndrome (who has restricted sight) to enable use of residual vision (http://www.sense.org.uk/content/sign-systems-and-languages)
Hands on BSL	Based upon BSL - enables the person to feel the signs. With this system, the deafblind person follows the signs by placing his/her hands over those of the signer and feeling the signs produced
Deafblind manual alphabet	A method of spelling out words directly onto the sight impaired/severely sight impaired person's hand. Each letter is denoted by a particular sign or place on the hand (Sense, 2016)

There were some participants who used more than one form of communication for example, visual frame British Sign Language (BSL) and clear speech or visual frame BSL, hands on BSL and deafblind manual. It was interesting to note the link between the choices participants made, for example where the participant used visual/tactile methods of communication e.g. visual frame BSL, hands on signing BSL and deafblind manual, their preferred interview choice was face to face, whereas participants who used clear speech to communicate primarily chose telephone or email interview methods. Written communication was used in two formats; eight participants used email, and one participant wrote brief notes to support signed communication in the face to face interview (Eve). Fourteen interviews were conducted with one contact whilst for six participants there was more than one point of contact as they were conducted via email. In most cases where the interview was conducted face to face, on Skype (video/no video) or telephone, these interviews were conducted in one session. However two participants initially chose email interview and then decided to change their interview method to that of face to face. Table 3.4 shows differences in communication and interview methods chosen by participants and the number of contacts required to complete the interviews.

Table 3.4: Communication and interview methods chosen by the participants and number of meetings required to complete interviews

Participant	Interview method	Communication or combination of communication methods used	Number of contacts
Adam	Face to Face/Email	Clear speech, visual frame BSL, Written communication	3
Ben	Face to Face	Clear speech	1
Carl	Email	Written communication	3
Debra	Face to Face	Visual frame BSL, Hands on BSL, Deafblind Manual	1
Eve	Face to Face	Visual frame BSL, Written communication	1
Fred	Skype no video	Clear speech	1
Gareth	Telephone	Clear speech	1
Harry	Email	Written communication	2
Iris	Email	Written communication	2
Jeff	Skype with video	Clear speech	1
Kate	Telephone	Clear speech	1
Len	Telephone	Clear speech	1
Monica	Face to Face	Clear speech	1
Nora	Email	Written communication	2
Oliver	Email	Written communication	1
Pam	Telephone/Email	Clear speech, Written communication	2
Quentin	Telephone	Clear speech	1
Ruth	Telephone	Clear speech	1
Sara	Telephone	Clear speech	1
Tia	Email	Written communication	1

I utilised a variety of communication and interview methods as I had previous experience of sensory social work where these skills had been learnt in practise. Having sensory knowledge was useful as in interviews participants did not have to interrupt the interview to explain sensory issues, equipment etc. However I was aware that prior knowledge could lead to bias. Thus, as considered earlier, the concept of epoche or bracketing was adopted to limit this occurring. It was also acknowledged that when carrying out flexible communication and interview methods, each could produce different data (Carter and Little, 2007). My paper entitled ‘Empowering people experiencing Usher syndrome as participants in research’ (Appendix A) considers these issues in detail and discusses a methodological innovation which I developed and used during research interviews called MSCIM.

MSCIM is an acronym for ‘Multiple Sensory Communication and Interview Methods’ and was used to “foster research engagement through participant led communication and interview methods” (Evans 2016 p1). Table 3.4 shows variations of communication and interview methods used in the conduct of this research. Although, MSCIM was used to conduct interviews with people with sensory needs, it has significant potential for future use, as highlighted in my article ‘Empowering people experiencing Usher syndrome as participants in research’ because MSCIM can apply across a variety of settings that require “flexibility, adaptability, and the use of multiple methods “(Evans 2016 p1). MSCIM would be useful for people who experience other sensory differences (visual impairment/Deafness), physical disabilities or where English is not the persons first language (Evans 2016). MSCIM would also be useful when conducting research with participants in hard to reach areas such as prisons, remote geographical areas or international research collaboration (Evans 2016). Additionally, using technological tools such as Skype enables participants to maintain a degree of anonymity if they wish to, as they can be involved in research with the video on or off (Evans 2016). MSCIM has a wide range of practical uses, for example in police/job centre interviews and health consultations (Evans 2016). Using MSCIM would enable people to feel more comfortable and empowered in what could be a stressful situation (Evans 2016).

Another consideration when interviewing was that of recognising that participants may become upset while talking about their experiences of Usher, and therefore a contingency plan was needed (Kvale and Brinkmann, 2009, Corbin and Morse, 2003). Participants were offered the option to provide support contact details prior to the interview should they require support during or after interview, however, most declined. Whilst I had experience of service users/clients becoming upset within a social work setting addressing such situations within a research setting can be different due to the differing roles of social worker and researcher (Kvale and

Brinkmann, 2009, Corbin and Morse, 2003). Thus the strategy taken to address this situation was to prepare as far as possible by reviewing appropriate scholarly material and drawing on experiences of others and taking precautions to address this issue should it arise (Kvale and Brinkmann, 2009). I was aware that participants need to feel comfortable in the interview process (Smith *et al,* 2012, Kvale, 2013); this will be considered further in section 3.6 where ethical considerations are discussed.

The data were captured using various methods and included email (written), video camera (visual) and digital voice (audio) recordings. Data from email did not require transcribing as it was in written format and followed the descriptive phenomenological approach of taking participant experiences as a given. Data collected during face to face, Skype and telephone interviews were audio recorded and transcribed (type-written copy) verbatim. Data from participants who communicated using visual frame BSL, hands on BSL and/or Deafblind manual were both visually and audio recorded and included the use of voice over [43]. Voice over was useful as during interviews there was continual translation which was voice recorded. Data from visual and audio recordings were transcribed verbatim. Although, this process was time consuming, it was useful as it enabled cross referencing. All transcribing was conducted by the researcher.

Also considered was the extent the narrative review influenced the data collection. Braun and Clarke (2013) highlighted that the aim of narrative is to tell a story and Green *et al* (2006) posited that the purpose of a narrative review is to provide a structure to synthesise the existing evidence about the topic, deliver a clear message and be as objective as possible. Although the structured approach of the narrative review enabled me to identify gaps in literature, the methodological

approach of descriptive phenomenology enabled collection of unique and individual life experiences of the participants without influence from the data collection.

Another challenge for the researcher when conducting descriptive phenomenological research is, the issue of whether the literature review should be conducted after data collection (Polit and Hungler, 1997, Polit and Beck, 2004, Hamill and Sinclair, 2010) or before (Chan *et al,* 2013). Although there is the view that when conducting descriptive phenomenological research that the literature review should be conducted after the collection of data to avoid pre-conceived assumptions influencing the data (Polit and Hungler, 1997, Polit and Beck, 2004, Hamill and Sinclair, 2010), for my study the literature review was conducted prior to data collection. There were a number of reasons for this choice.

The first was that a research proposal was required by the London South Bank University Research Ethics Committee to grant ethical approval and "meticulous work on the literature review is required to justify the need to carry out the research" (Chan *et al,* 2013 p2). Also it was important that there was a link between the "background knowledge and the research under study" (Chan *et al,* 2013 p3) to prevent questioning "justification for the research need and the overall plan of the study "(Chan *et al,* 2013 p3). Therefore, a literature review prior to data collection was essential. Additionally, I was aware that the topic I was researching, that of Usher syndrome was a rare, under-researched topic and it was unlikely that there would be a plethora of published literature that would influence the data.

Another aspect for consideration during data collection was power dynamics, this will next be discussed.

3.4.3.1 Power dynamics during data collection

Engaging people from marginalised groups such as the deafblind and Usher communities to participate in research has historically proved challenging, mainly due to communication differences between participants and researcher. As people with sensory needs are all different and use different types of communication, multiple communication and interview methods were crucial in the research process. From the conduct of my research an approach called 'Multiple Sensory Communication and Interview Methods' (MSCIM) was developed and used when conducting research with the participants, and then explored in a paper published in the British Journal of Social Work (Appendix A). Though the use of MSCIM is considered throughout this chapter, in this section it will be discussed in relation to power dynamics that can arise when collecting data.

Although in my role as a sensory social worker it was natural for a service user to choose their interview and communication method, within the research this approach led to a measure of unexpected equalising during data collection between the participants and the researcher and this empowering of these individuals as active participants in research contributed to inclusivity and promoted trustworthiness of the research.

After my paper was published a further exploration of the issue of power dynamics between researchers and researched during data collection identified that although there have been studies providing insight into the nature of power between researched and researcher, interviewing, with people classified as global elites (Conti and O'Neill, 2007) foreigner elites (Herod, 1999), and in cross cultural settings (Mullings, 1999), no sensory or Usher specific literature was identified. Fisher (2011) discussed collaborative research relationships between researcher

and organisations, specifically the “municipal network in India[48]” (p456). Although her work is with organisations, the issues identified relating to power of the people being researched is relevant on an individual level as organisations are a group of individuals (albeit with joint aims) and individual power dynamics still arise. For example Fisher writes about the power she perceived she had compared to the limited level of power in reality. This also arose in my current study because although I perceived I had a level of power as a researcher, the reality was that as I was not communicating in my first language, my position of power was reduced due to the power dynamics which occurred due to language differences.

Mullings (1999) considered her experiences of conducting interviews with managers and workers in an information processing company in Jamaica. Again Mullings discusses the ‘insider’/ ‘outsider dilemma as well as positionality during data collection. Mullings discussed that although she felt disempowered in some interviews, when she interviewed the business elites she felt empowered when interpreting the data. This also arose in the current study because at times when I was collecting data and not communicating in my first language, I too felt disempowered.

Herod (1999) discussed interviewing foreign elites who were, “foreign nationals who hold positions of power” (p313). Herod explored the importance of “positionality” (p313), for example whether the researcher is an “outsider” or an “insider” (320), within the research. Herod highlighted that being an “insider” was useful as the researcher was perceived as having knowledge relating to the research being conducted as opposed to being an “outsider” with little or no knowledge. However, he also acknowledged that positioning is not always simple because interviews are fluid and the researcher’s position can change depending on the discussion. In the

current study insider knowledge was useful in terms of understanding equipment issues rather than the participant having to stop their flow and explain.

Conti and O'Neill (2007) interviewed the global elite which included diplomats, private attorneys and member nation trade delegates and shared their experiences of "the strategic content over authority" and their feelings of "despondency that resulted from being talked down to" (p61-82). Conti and O'Neil highlighted that feminist approaches are useful when interviewing in situations with powerful people who mould the interview process. Conti and O'Neill refer to the usefulness of feminist approaches to research when negotiating the power balance between the researched and the researcher. Feminist researchers explore the complex nature of power and critique the relationship between researchers and researched suggesting that this relationship informs and influences the data that is collated in terms of interpretation and representation (Rose, 1997, Conti and O'Neill, 2007). Rose further proffers that for a researcher to examine their position within the research process, reflexivity is essential. The importance of reflexivity was also considered in the current study as a key aspect to the study was researcher reflexivity with regard to the impact of the research on the participants and will be discussed later in the chapter.

Critiquing this literature highlighted that the power dynamics within data collection are often two-way, because in some instances power will shift to the advantage of the researcher but in other instances it will be in the favour of the researched. This concluded that although various approaches can be used to address the issue of power dynamics during data collection between researchers and researched, the literature shows that situations can only be addressed as they arise (Conti and O'Neill, 2007). Having considered the data collection methods and issues that can arise, next data analysis will be considered.

3.5 Data analysis

Three data analysis options were originally considered, that of IPA (Smith *et al,* 2012), Giorgi's "four stage analysis process" (Giorgi, 1985 p10) and Braun and Clarke's "6 phase guide to performing thematic analysis" (Braun and Clarke, 2006 p79). IPA was excluded as reflected upon earlier, leaving two other options for consideration. Whilst Braun and Clarke's approach became the chosen data analysis method for this study as will be discussed later, the reason for excluding Giorgi's process (Table 3.5) will next be considered.

Table 3.5: Giorgi's phenomenological psychological four stage data analysis process
(Adapted from Giorgi, 1985)

Read the entire description in order to get a general sense of the whole statement
Return to the beginning and read through the text once more with the specific aim of discriminating 'meaning units' from within a psychological perspective and with a focus on phenomenon being researched
Examine the meaning units and express the psychological insight contained in them more directly and this is especially true of the "meaning units" most revelatory of the phenomenon under consideration.
Synthesise all the transformed 'meaning units' into a consistent statement regarding the subjects experience and this is usually referred to as the structure of the experience.

Giorgi's approach was not considered compatible to the study because it applied a deep psychological attitude to analysis (Giorgi, 1985, Giorgi, 2009) as a result of his experiences as a psychophysicist, human psychologist and scientific psychologist, specialising in experimental psychology (Giorgi 2002) and the complexity of it did not match with the quality and diversity of the data or the research topic. Giorgi suggests that researcher's needs to analyse the description of accounts given by participants with sensitivity and affinity of their own discipline (Giorgi, 2002) and as my background is that of social work, Braun and Clarke's thematic approach resonated with the practical social work role. Additionally, whereas Giorgi's process

considers more face to face interview data, Braun and Clarke's approach was considered more flexible as my data included visual (BSL, Deafblind manual alphabet), spoken and email data (see Tables 3.3 and 3.4). A more detailed explanation of the reasons for selecting Braun and Clarke's approach to data analysis will next be discussed.

Thematic analysis was originally developed by Gerald Holton, a physicist and historian of sciences in the 1970s (Merton, 1975), and later developed by Braun and Clarke (2006). Braun and Clarke suggested "thematic analysis is a method for identifying, analysing and reporting patterns (themes) within data" (2006 p79). Braun and Clarke's (2006) set of procedures for analysis also contribute to understanding various opinions, ideas, perceptions and experiences which contribute to the drawing of themes from data. Whilst Braun and Clarke's (2006, 2013) guide to performing thematic analysis could also have a psychological perspective as the authors are psychologists, it was considered to be compatible with the quality and diversity of the data, it would be contributory in promoting a transparent and rigorous approach, it is more neutral and has a 'toolbox' approach. There were a number of reasons for adopting this approach, firstly this guide offers various pattern based analytical methods to draw themes from the data (Braun and Clarke, 2013). For example, they consider inductive thematic analysis where analysis is "not shaped by existing theory"; theoretical thematic analysis which is "guided by existing theory"; experiential thematic analysis which "focuses on participants' standpoint", their experiences and how they make sense of these; and constructionist thematic analysis which focus on "how topics are constructed" (Braun and Clarke, 2013 p175).

For this study experiential thematic analysis was selected as this was compatible with descriptive phenomenology in which participant experiences are central. It also

concentrates on where participants see themselves and how they construct and make sense of their experiences (Braun and Clarke, 2013), which were central to this study and compatible to the paradigmatic positioning.

Braun and Clarke's (2006) approach is also compatible with the sample size (twenty participants) and it clearly outlines a set of procedures for analysis (Table 3.6). The thematic approach of capturing "something important about the data in relation to the research question" with a level of representations of "patterned response or meaning within the data set" (Braun and Clarke, 2006 p82) were integral to the study. The researcher took an "active role" (Braun and Clarke, 2013 p225) to developing themes from coded data and creating potential patterns to produce new knowledge, as opposed to a passive role which seeks to "identify something that already exists" (Braun and Clarke, 2013 p225). The approach was also considered compatible to the research topic interview questions and data responses because participant responses shaped themes to enable the data to relate their experiences and tell their story. Table 3.6 outlines Braun and Clarke's guide to performing thematic analysis.

Table 3.6: Braun and Clarke's 6 phase guide to performing thematic analysis
(Summarised from Braun and Clarke, 2006)

Familiarising yourself with the data: Transcribing data, reading and re-reading the data, noting down themes and ideas that initially arise
Generating initial codes: Coding interesting facets of the data in a systematic way across the entire data collection process, collating data that is relevant to each code
Searching for themes: Collating codes into prospective themes, collecting all of the data relevant to each prospective theme
Review themes: Check if the themes that arose initially are compatible to entire data set themes. Generate a thematic map of analysis
Defining and naming themes: Continue to analyse, in so doing refine the specifics of each theme that has arisen. Look at the picture that is emerging from the analysis and name the themes
Producing the report: Final chance for analysis. Relate analysis back to the research question and to the literature review to enable production of a report of your data analysis

Braun and Clarke's thematic analysis approach (Braun and Clarke, 2006, 2013) was used with descriptive phenomenology to enable participant experiences to be taken as a given and themes from the data drawn to ensure participants' experiences were reflected in the findings (see Appendices, C, D, E, F, G, H, I). Four themes were identified, which will be presented in Chapter 4.

Van Manen (1984) highlighted "phenomenological themes are the structures of experience" (p59). However van Manen advises "a thematic phrase only serves to point at, to allude to, or hint at, an aspect of the phenomenon" (1998 p60). So it is essential when using this method of data analysis to also have a broader philosophical perspective to data collated, to enable this, bracketing or epoche (Seamon 1979) was adopted as previously discussed. Ethical considerations will next be discussed.

3.6 Ethical considerations

Kvale (2013) posits that ethical considerations are rooted in the interview inquiry in its entirety. He further highlights that "ethical issues permeate research" and the interviewer needs to be able to "create a stage where the subject is free and safe to talk of private events" (Kvale, 2013 p8). Although participants talked off camera/digital voice recording, only data the participants were willing to share were analysed. For example one participant talked off camera about personal familial Usher experiences but was unwilling for comments to be used within the research, and this was respected. Key ethical considerations which are discussed in this section include: access, confidentiality, informed consent and risk. The study was approved by the London South Bank University Research Ethics Committee (UREC – Reference 1256) on 24th October 2012 (Appendix J) with amendments to age

range (16+ - No upper age range limit) on 1st September 2013 (Appendices K and L) with all participants giving written and verbal informed consent (Appendix M).

3.6.1 Confidentiality

Confidentiality and anonymity were central to the research process. Each participant was allocated an anonymous identifier, any identifying details were removed during transcription and no personal details were included, to prevent recognition. Other people involved in the interviews, for example, an interpreter/communicator [person chosen by the participant to support them with translation and communication during the interview] were asked to sign a confidentiality agreement (Appendix N) stating that the information disclosed during the interview would not be disclosed outside of the interview environment. All data collected were stored securely on an encrypted device**.**

3.6.2 Informed consent

Participation was on a voluntary basis with people who responded to the flyer distributed by Sense contacting me directly to take part in the study or participant details being passed on by Sense for me to contact the individual. Once contact had been made, I sent potential participants a participant information sheet (Appendix O). To gain informed consent, thought was given to individual communication requirements, for example the participant information sheet (Appendix O) may have had to be read to people whose sight loss prevented them from reading it themselves or translated for people who used sign language. Each person was provided with details for them to consider within their individual time frame. Once participants agreed to participate they were asked to give written consent to being part of the study. Consent was voluntary and entirely an individual's choice.

3.6.3 Risks

There were a number of risks identified and these included bodily contact, potential psychological intrusion and misunderstanding of social and cultural boundaries. Each of these will next be considered.

3.6.3.1 Bodily contact

Bodily contact was considered as I anticipated that participants may choose tactile sensory communication methods such as hands on signing[7] and/ or deafblind manual[17] (Table 3.3). In order to minimise risks the form of communication was chosen by the participant. One participant did choose this method (Debra).

3.6.3.2 Psychological intrusion

Psychological intrusion was minimised by fully informing the participants (participant information form), making it clear that they could withdraw from the study at any time up to the point of completion of the thesis, using their own communicators/interpreters, making their own choices fully and without coercion and showing respect and sensitivity at all times. If people became upset they were able to stop or withdraw from the study at any time up to the point of submission of the thesis. Whilst upsets were not anticipated, as discussed in section 3.4.3 participants were offered the option of providing a personal contact prior to the interviewing process to support them, although most declined. Whilst it was not obvious that anyone became upset, there was an incident when Gareth, whilst talking about visual services support, asked to be excused to get a glass of water and was gone for about 5 minutes. When he returned he sounded a little subdued, which prompted me to ask if he was ok. Whilst the participant assured me, all was fine, it did highlight that as this interview was over the telephone, it was not possible to see if the participant had become upset.

3.6.3.3 Misunderstanding of social and cultural boundaries

An issue that may have arisen related to the participants knowing I was a social worker, and they may have misunderstood my role. However, this did not arise as all participants were aware that I was an academic in a university and that the reason for my visit was to collect data that would contribute to a research study as explained in the participant information sheet.

As this study explores disability, misunderstanding of social and cultural boundaries may also have arisen as a person who considers themselves to be culturally Deaf, may not consider themselves to be disabled but rather part of a linguistic minority group, complete with its own culture history and language. However, assumptions were never made and each participant did have their own individual stance. Whilst these issues were considered, no one did withdraw from the study.

Risks relating to conducting interviews in people's homes were also considered. As a social worker, I was trained to lone work and these skills, techniques and precautions would have been utilised had an incident occurred. However, no risk issues arose.

3.7 Trustworthiness

Thompson succinctly defines trustworthiness as "deserving of trust or reliable" (Thompson, 1996 p1114). Trustworthiness in research includes rigour (adhering to specific philosophical perspective), believability and plausibility and can be described as truthfulness and reliability of something or someone (Koch, 1994, Anney, 2014). In this section the meaning of trustworthiness will be considered as it

has an influence on what is meant by Multiple Sensory Communication and Interview Methods or MSCIM.

Smith *et al* (2012) posits that rigour refers to the "thoroughness of the study" and is attained by a commitment from the researcher that the study will be thoughtfully considered including adhering to sensitivity and comfort of the participants (p81). Smith highlights this is achieved by "demonstration of commitment and can be synonymous with a demonstration of sensitivity to context" (Smith *et al*, 2012 p81). In this study commitment and sensitivity were achieved by respectfully arriving on time for interviews and being prepared, willing, and focused to communicate using the individual's preferred communicative method, which was the approach I embraced.

Additionally, to ensure rigour the research data were collected in a systematic and ethical manner. The decisions made were self-conscious and reflexive with decisions having a specific experiential focus e.g. lived experiences of people experiencing Usher syndrome. As identified earlier in this chapter methods and methodologies were compatible to the data analysis method as the aim was to gain lived participant experiences and their sense of the world as they saw it. A significant aspect of sustaining rigour was flexibility surrounding the communicative approach as discussed earlier, for example, being prepared to communicate using each participant's preferred communication method and recognising that as the participants' levels of sight and hearing differed so did their communicative choice.

Ensuring trustworthiness was particularly important in this research study as it may be difficult for people with Usher syndrome to share their experiences with a researcher and so from an ethical perspective, it was essential that the data are trustworthy and conveyed in a credible manner so that the study findings can

benefit others. Finlay (2009) acknowledges that one of the biggest challenges in qualitative research is how to assure the quality and trustworthiness of the research. Shenton suggested that trustworthiness in qualitative research is:

> "*often questioned by positivists, perhaps because their concepts of validity reliability cannot be addressed in the same way in naturalistic work*" (Shenton, 2004 p63).

Giorgi highlights that trustworthiness involves trying to reach real understandings without prejudgments (Giorgi, 2002). One way of achieving this is as discussed previously by using the tool of epoche or bracketing (Sorsa and Astedt-Kurki, 2013).

Guba and Lincoln (1989) suggest that trustworthiness has to be established in research for it to have value and worth. To explore issues surrounding trustworthiness, Guba and Lincoln's (1989) naturalistic criteria for evaluating qualitative research was used. The four criteria include credibility (truth value), transferability (applicability), dependability (consistency) and confirmability (neutrality).

Credibility refers to how the researcher can establish confidence in the truth of the findings of the research. Transferability concerns how the findings from this research study could be applicable in the context of other research studies. Dependability considers consistency and whether the findings of one study would recur if the research study was replicated with the same or similar people in the same or similar context. Confirmability concerns neutrality and whether the findings emerge from participants' own experiences or whether findings are influenced by the researcher's own biases motivations, interests and perspectives. To attempt to address these four criteria in relation to trustworthiness the interview and communication methods used within this research will be critiqued.

3.7.1 Credibility

As considered in the previous section, credibility is linked to trustworthiness and methodological rigour. Patton posited that the credibility of a study hinges on three separate but relating elements, “rigorous methods, the credibility of the researcher and philosophical belief in the value of qualitative inquiry” (Patton, 2002 p584). Guba and Lincoln (1989) refer to the essentiality of the credibility of the research and establishing confidence in the “truth of the findings” (p234).To ensure rigorous methods were achieved, fieldwork incorporated a variety of communication methods to ensure participants were able to express their experiences from a personal perspective and data were analysed with particular focus on credibility.

Credibility was also established by adopting a descriptive phenomenological approach to ensure that as far as is possible, (although this is a contested field) participant responses were taken as a given (Giorgi, 2009) as opposed to interpreted (Heidegger, 1962). After all it was their personal experiences and in order to share these in the research process the participants needed to feel confident their experiences would be taken on face value. Credibility was also established by translating data collected by using different methods. For example where a visual form of communication was chosen by the participant e.g. visual frame BSL/hands on BSL/deafblind manual, the interview was both audio (with voice-over [43]) and video recorded. Voice over was possible as the participant chose to have someone of their choice who was hearing and who was able to communicate using clear speech and a combination of visual forms of communication (Evans, 2017). As I had access to both audio and visual recordings of the interview this enabled me when translating to cross reference and ensure the interview content was credible and truthful.

Patton (2002) discusses that researcher credibility is dependent on “training, experience, track record, status and presentation of self” (p584). It could be suggested that credibility was enhanced as I had over 10 years of professional experience within the sensory field (D/deaf, deafblindness and visual impairment) in which a wealth of sensory contacts, knowledge/skills and sensory communication qualifications (British Sign Language qualifications/level 3; deafblind manual/level 2) were achieved.

3.7.2 Transferability

Transferability refers to the research findings and whether they would be applicable, relevant or appropriate to other studies. Whilst this study focused on Usher syndrome, findings from the data related to areas such as: The need for greater awareness being raised; the essentiality of family support and so on. These findings could be applicable in other areas of sensory or disability research such as experiences of people with visual impairment, people experiencing D/deafness or people with physical disabilities. A further component could be applicability of methods used (MSCIM) which have been described in rich detail. MSCIM (Evans, 2017) could be applicable within other research studies such as with people whose first language is not English (Temple and Young, 2004, Obasi, 2014). It could be suggested that gaining high quality data through using the participants’ preferred methods of communication and interviewing contributed to the richness of data quality (Evans, 2017).

3.7.3 Dependability

Dependability considers whether the findings of one study would recur if the research study was replicated with the same or similar people in the same or similar context. If similar approaches were used and communication and interview methods

participant led, consistency could be achieved. An example of promotion of dependability in this study was with regard to the detailed descriptions provided of how the data collection was conducted thus enabling other researchers to understand methods used. Additionally, the use of MSCIM could also impact on consistency.

3.7.4 Confirmability

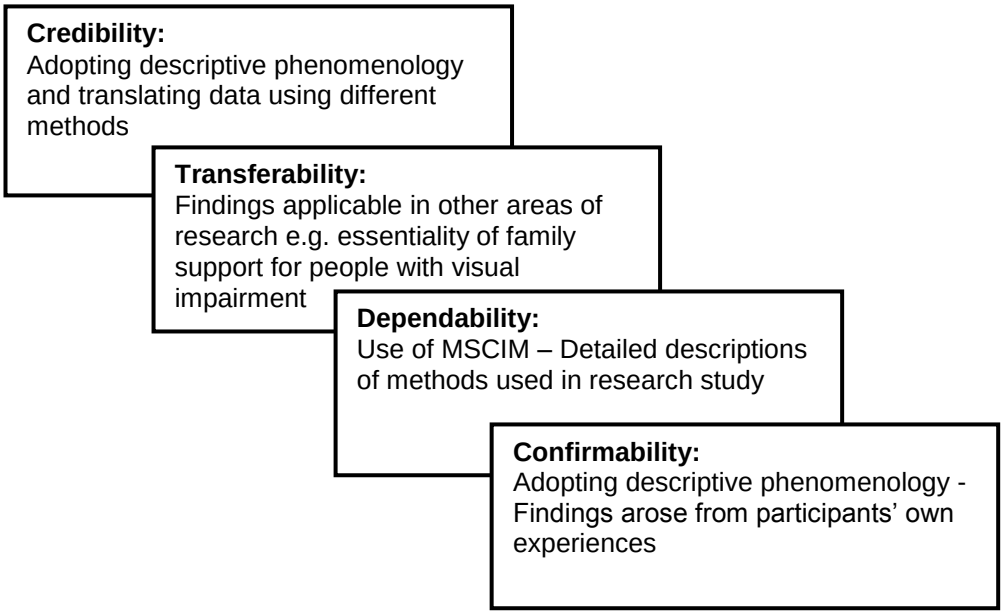
With regard to the final area, confirmability, as has already been discussed my sensory experience from a professional perspective could have influenced the findings. However, using the descriptive phenomenological (Giorgi, 2009) approach, ensured findings arose from participants' own experiences. Additionally my own reflexivity (which will be discussed further later in this section) contributed to the promotion of confirmability. Berger (2015) posits that reflexivity is considered to be a constant internal discussion and critical self-evaluation of the researcher's position within the research.

As a reflexive researcher there was a constant questioning of self to ensure that prior professional experience did not impact on the current study, in order to do this, a reflective diary was kept. However, Berger also highlights some advantages to having prior knowledge. For example where the researcher does have experience, it can encourage participants to be more willing to share their experiences as they may perceive the researcher to be more understanding and sympathetic to their situation. This situation did arise within this study with participants enquiring if I had knowledge of sensory equipment or issues.

Professional sensory knowledge also contributed to the flow of the interview as the participant did not have to stop and explain what they meant when describing

sensory equipment or relaying any other sensory experiences or issues. Additionally, the participant leading on the choice of the interview and communication methods also contributed to trustworthiness as this approach established participant confidence in the credibility of the research findings. Thus to summarise Guba and Lincoln's (1989) four criteria for evaluating qualitative research and how it was applied to this research study see Figure 3.3

Figure 3.3: Guba and Lincoln's four naturalistic criteria for evaluating trustworthiness
(Adapted from Guba and Lincoln, 1989)



Having considered confirmability, insider bias will be reflected upon next.

3.7.5 Insider bias and the impact of reflexivity, subjectivity and neutrality

As identified earlier my role was a sensory social worker and therefore, insider bias requires acknowledgement in order to contribute to promoting trustworthiness. Whilst no credible researcher would advocate for biased distortion of data (Patton, 2002) some theorise that bias is inevitable if the researcher has prior knowledge of the phenomenon being researched and that conducting qualitative research as an insider can be challenging (Asselin, 2003). However, prior experience also brings positive aspects as it can be seen as a useful resource. For example, I think my role as a sensory worker enhanced the study as opposed to biasing it because my knowledge and understanding of sensory issues was useful for participants in interview, in terms of understanding of equipment etc. and being able to be flexible with communication.

To minimise as far as possible insider bias occurring, concepts of reflexivity, neutrality and suspension of preconception in relation to insider bias and the role of the researcher were considered.

Reflexivity is integral to not only the social work role, but also that of the qualitative researcher. As considered in 3.2, in constructivist research the researcher is involved in the inquiry as they become responsive and reflective to the participants' dialogue (Appleton and King, 1997). Shaw (2010) explored the difference between the terminology *reflexivity* and *reflection* as they are often used interchangeably. Shaw highlighted that Woolgar (1988) considers reflection, to be a general set of thoughts concerned with the process and verification whilst reflexivity, is an evaluation of one's self or "turning your gaze to the self" (Shaw, 2010 p234). Tisdall *et al* also highlights that reflexivity is an essential element of qualitative research and defines it as "the thoughtful reflection of a researcher upon the impact of her or his research on the participants….and on the knowledge produced" (Tisdall *et al,* 2009 p229). It is useful to explore the distinctions as often the two terms are considered to mean the same, but clearly they do not.

There are a range of reflexive "research traditions" (Finlay and Gough, 2003 p6) which include: reflexivity as introspection, which involves the researcher's own reflections being primarily used; reflexivity as intersubjective reflection where "researchers explore the mutual meanings emerging with the relationship" (Finlay, 2002 p215); reflexivity as a mutual collaboration which "seeks to enlist participants as co-researchers" (Finlay, 2002 p218); reflexivity as social critique which "acknowledges tensions arising from difference social positions" (Finlay, 2002 p220) and reflexivity as discursive deconstruction where "attention is paid to the ambiguity of meaning in language used and how this impacts on modes of presentation" (Finlay, 2002 p222).

After reflecting on which reflexive model was compatible with my study, reflexivity as introspection was chosen, for two reasons: firstly, because introspection examines "one's own mental and emotional processes" (Thompson, 1996 p522) resulting in "insights emerging from personal introspection" (Finlay, 2002 p214) and secondly because interest begins with discovering "an intense interest, a passionate concern that calls out to the researcher" (Moustakas, 1990 p27).

In this study I had passion for engaging in sensory research following a professional sensory career which spanned more than a decade. Additionally, our role as researchers is to make sense of the life experiences participants tell us with a view of learning more about them, to effect change, whether it is in terms of policy and practice or raising awareness or "enhancing understanding on an individual or institutional level" (Shaw, 2010 p233).

Finlay (2002) highlights the value of using introspection and the usefulness of being reflexive about one's own personal reactions. Within my study, there were occasions when the experiences shared made me feel extremely saddened, by reflecting on my own personal reactions, this enabled me to make sense of the experiences that were being shared and see that the participants were now at a time in their lives where they felt able to share their experiences, which meant they themselves were now in a better place, than when first experiencing Usher.

However, Finlay also warns that there needs to be a balance and researchers need to avoid being too "preoccupied by one's own emotions and experiences"(Finlay 2002, p215) otherwise the participant's voices can get lost. Finlay also highlights that challenges can also arise when using introspection as reflection and researchers need to ensure that their own personal reflections are used as a foundation for knowledge and insights, and that these personal reflection recognises

the links between “knowledge claims, personal experiences of both participant and researcher, and the social context” (Finlay, 2002 p215). In my study, although I was aware of my own reflections, the experiences of the participants were paramount.

Reflexivity requires action in terms of the act of self-reflection, resulting in analysis on the researcher’s part as “one cannot be reflexive about what one takes for granted” (Taylor and White, 2001 p55). Whilst I acknowledged that the participants’ positioning, which could include age, race, gender etc. (Berger, 2015) may affect the research process, the main area of consideration related to my professional sensory knowledge. Therefore, key to the study was my reflexivity with regard to the impact of the research on the participants. Shaw posited that “experiences must be understood within the context in which they happen” (Shaw, 2010 p234) e.g. the participant’s personal experience of life with Usher. Also that “we experience and interpret the world from a particular perspective and we can never fully escape this subjectivity” (Shaw, 2010 p235).

Subjectivity relates to reality and truth, from a subjective view or a variety of sources (Ratner, 2002) and can be used as an “important inroad to understanding and constructing knowledge” (Mruck and Bruer, 2003 p25) and as a tool to improve knowledge (Mruk and Breuer, 2003). Ratner (2002) posits that subjectivity guides everything we do with regards to the research process because it guides our topic choices, selection of methodologies and how we interpret data. Additionally, Bruer *et al* (2002) suggest that “personal, social, and local factors influence the research process” (p246).

Giorgi (2002) asserts that as there is a strong correlation between phenomenology and subjectivity “the very ambition to eliminate subjectivity is an impossible dream” (p10). Giorgi (2002) posits that within phenomenology, “the goal is not to try to

eliminate subjectivity, but rather to try to clarify the role of subjectivity when correct knowledge is attained" (p8). Giorgi (2002) further highlights that the goal is to "study actual situations" (p10), which means that as far as possible the researcher stays true to the descriptive accounts of the participants but still taking into account that subjectivity is present. Ratner (2002) highlights that there is a positive impact on the research project when the researcher reflects on the values and objectives he/she brings to the research. Mruck and Breuer (2003) suggest that the researcher is also a research tool and that there are lots of different skills that researchers bring to the study.

On reflection within my own study there was a positive contribution to subjectivity because I brought sensory skills and knowledge which had a positive impact on the research. For example, the participants were more willing to talk to me and allowed me to collect data because of the sensory knowledge and skills I had. Also, as discussed in chapter 3, when recruitment was difficult, an opportunity arose to provide communication guide service support[41] to one of the members of the hearing and sight impaired (HSI) group to a HSI social event. It was following this event that members of the HSI group got to know me better and were more willing to be involved in the research study and agreed to be interviewed. If I had not had the sensory skills required to enable provision of support, the opportunity to recruit more participants for the study would not have been available to me.

I kept field notes after the interviews, and made notes when conducting interviews via Skype or telephone. The recording of field notes when conducting interviews in visual frame BSL, hands on signing or deafblind manual were not possible during interview, but notes were made after. The keeping of field notes or notes was considered crucial because often as humans we quickly forget information received (Montgomery and Bailey, 2007). Some interviews were emotive, for example Ben

expressed that "*I thought at that time everything hopeless*" and went onto share experiences at college where other students had bullied him. The issue of personal researcher emotion emerged as a result of sensitive experiences shared by the participants. I considered myself to be generally reflexive as this was a skill acquired and practised as a sensory social worker therefore I dealt with my emotions that arose in the conduct of research in a similar way, which was in a practical manner, for example taking physical exercise.

As the interviews progressed, I reflected upon, and became aware of, not only the different ways the condition affected a person practically, for example the inability to drive, but the effect the condition had on the person psychologically, for example depression and the impact of sharing their experiences with a researcher (see Chapter 4). The fact that experiences were sensitive highlighted the inappropriateness of member checking (the process of checking data with participants), as whilst the participants were willing to share their experiences, the sharing of them clearly had an impact on them (Ben) and reliving these experiences again to member check was inappropriate. The participants were prepared to contribute to the interviews but not be contacted back as their experiences were of a complex and delicate nature. Experiences shared were unique and individual. For some people the experiences were very difficult to share (Monica) and for others it was an opportunity to raise awareness and share their narrative (Kate). Reflecting upon the impact of this research on the participants, highlights that whenever an individual experiences an unpredictable, debilitating and deteriorating condition such as Usher syndrome, great sensitivity is needed throughout the whole research process as participant experiences are not just snapshot experiences but their whole life experiences.

Thus to summarise, this study drew on reflexivity as introspection as my reflections were impacted by my personal experiences as a professional sensory worker, which in turn led to fuller understanding of each participant's experiences. Embedding reflexivity into research allows us to come to know our-selves and in turn "use each participant's presentation of their self" (Shaw, 2010 p235) to review and maximise data collected and analysed, and as far as possible reduce insider bias. As reflexivity as introspection was chosen, it could be suggested that reducing insider bias would not be possible, however exploration of neutrality and insider bias considers strategies that could be utilised.

Neutrality arises when considering insider bias, Patton highlights that for research to be considered trustworthy or credible the researcher needs to adopt "a stance of neutrality" (Patton, 2002 p51). In simple terms this means that the researcher does not "set out to prove a particular perspective or manipulate the data to arrive at predisposed truths" (Patton, 2002 p51). Neutrality was an essential element within the study because as highlighted earlier I was aware that experiences of Usher syndrome affected people in different ways and that data shared would be individually unique. Patton suggests that the researcher who adopts neutrality has a commitment "to understand the world as it unfolds" (Patton, 2002 p51), so within the study my position was neutral because I took participant's experiences as a given and enabled participants to feel comfortable in sharing their experiences. Also, as discussed previously whilst subjectivity may influence research because as researchers we make methodological and personal choices which can influence the research process, the researcher can still maintain neutrality by not setting out with pre-conceived ideas or manipulating the data to achieve an end result but rather being committed to gaining and understanding knowledge as it is constructed.

To contribute to a neutral stance being adopted I wished to avoid preconceptions,

impacting, manipulating or affecting data, and so suspension of preconception also known as bracketing/epoche as discussed in section 3.3.1 was adopted. As our thinking is shaped by the literature available (Smith, 2009), professional/personal experiences and different ideas that are presented to us, suspension of preconception was a useful tool to avoid putting particular perspectives on the data collated from people with Usher. Previously considered was the notion that the researcher when analysing data needs to suspend preconceptions in order to focus on grasping the experiential world of the research participant. However, it could be suggested that this is also the case within the interview process to as far as possible, minimise insider bias.

Whilst knowledge accrued from my professional sensory experiences was useful, for example understanding sensory terminology/equipment and having an understanding of what people are going through as a result of their sensory loss, I also considered that suspension of preconception was an essential element within the research as it empowered the participants to share their narrative/experiences and allowed data to tell its own story and unfold freely (Patton, 2002).

Using qualitative evaluative approaches such as Guba and Lincoln's naturalistic criteria (1989) as well as considering the credibility, methodological rigour and insider biases of the research undertaken, contributes to promoting trustworthiness and providing an audit trail to enable other researchers to attempt to replicate the study and strengthen confirmability (Billups, 2014, 2016). Whilst the audit trail does not tell other researchers how to conduct research it does give the researcher's own view on how the process worked for them (Billups, 2016) as outlined in this section.

3.8 Chapter summary

In this chapter the methods were introduced. The researcher's paradigmatic positioning (Figure 3.2) was presented together with the rationale and decision making behind choosing a descriptive phenomenological approach for the study. Additionally, also discussed were the data collection methods adopted; the data analysis method; ethical considerations, trustworthiness in qualitative research and how this was achieved in the study and steps taken to minimise insider bias. In Chapter 4 the findings section of the thesis will be presented.

Chapter 4 Findings

4.1 Introduction

Chapter 3 outlined the rationale for the epistemology and methodology of descriptive phenomenology, and explained the sampling, recruitment, and access, data collection, data analysis, ethical considerations, trustworthiness and reflexivity. In this findings chapter, participants' responses are recorded in their own words, to illustrate the themes and subthemes. No changes to the interview extracts (e.g. grammar/spelling corrections) have been made where the whole interview (Carl, Harry, Iris, Nora Oliver, Tia), or part of the interview (Adam, Pam), was conducted via email; where the translations were from visual frame BSL, hands on BSL or deafblind manual (Adam, Debra, Eve); and where the participant experienced speech difficulties (Ruth). This approach was adopted to enable the person's own description of their experiences to be presented, as well as being congruent with a descriptive phenomenological methodology.

Following presentation of each theme there is a short summary of findings. The chapter concludes with a summary, which outlines key cross cutting messages. This chapter presents each of the four themes in a separate section with their related sub-themes as laid out in Table 4.1.

Table 4.1:Themes 1, 2, 3 and 4

Theme 1 'Diagnosis is the start of the experience' • Early life, Life before Usher • Impact of diagnosis • Experiences after diagnosis
Theme 2 'Familial relationships across the lifespan' • Child diagnosed with Usher • Siblings of a child who has Usher • Being a parent who has Usher • Relationships • Family support
Theme 3 'A sense of belonging' • Interrelationships between communication, culture and community • Friendships and shared experiences
Theme 4 'Experiences of professional support' • Education, employment and benefits • Sensory support: ophthalmology, audiology and social work • Sensory equipment/Guide dog provision • Specialist charity support • Raising Usher awareness

The first theme to be presented is 'diagnosis is the start of the experience'.

4.2 'Diagnosis is the start of the experience'

The age of the participants ranged from 18-82 years and the start of the experience varied for each of them (Table 4.2). Although all participants experienced hearing loss ranging from profound to mild, the addition of visual difficulties led to the diagnosis of Usher syndrome. Receiving a diagnosis had a lot of implications (practical, emotional, physical etc.). Although some participant's knew something was wrong (Gareth, Jeff, Nora, Oliver, Pam, Quentin, Ruth, Sara), when they were diagnosed with the hereditary/genetic condition of Usher, which they knew was progressive, this was the start of the experience.

4.2.1 Early life, life before Usher

There were variations in the age at which participants received a diagnosis, although eight participants (Gareth, Jeff, Nora, Oliver, Pam, Quentin, Ruth and Sara) reported having experienced symptoms of dual sensory loss earlier in life. Table 4.2 presents the age of diagnosis and any prior sight difficulties.

Table 4.2: Age at which participants were diagnosed, time of interview and any prior sight difficulties

Participant	Age diagnosed	Age at time of interview	Expressed sight difficulties prior to diagnosis
Adam	18	49	Yes - noticed clumsiness
Ben	18	29	Yes - noticed clumsiness
Carl	24	49	Yes - noticed clumsiness
Debra	19	82	Yes - noticed clumsiness
Eve	27	54	Yes - noticed narrowing of visual range
Fred	21	54	None - reported main focus was deafness
Gareth	50s	57	Yes - most evidence in teenage years - stopped driving aged 29 years
Harry	26	45	Yes - noticed night vision poor
Iris	30	51	Yes - noticed clumsiness
Jeff	27	55	Yes - noticed aged 18 years. Noticed clumsiness
Kate	15	63	Yes - difficulties recognised at school
Len	36	73	Yes unable to catch ball, when football went out of line of vision
Monica	26/27	39	Yes - noticed clumsiness
Nora	39	59	Yes - symptoms from age 15 years
Oliver	Teens	45	Yes - symptoms from age 5 years
Pam	39	52	Yes - reported sight difficulties prior to diagnosis
Quentin	49	73	Yes - sight difficulties from earliest memories
Ruth	11	69	Yes - reported sight difficulties prior to diagnosis. Noticed clumsiness
Sara	25	32	Yes - reported sight difficulties prior to diagnosis
Tia	16	18	Yes - difficulties recognised at school

Sara, who had experienced sight difficulties prior to a diagnosis of Usher, thought "*there is nothing wrong you know what I mean, it's just one of those things I was just a bit clumsy*". Sara's comment indicates that the experience of having Usher had already begun before she realised it, but had been part of everyday life. Several other participants noticed something was wrong because they experienced clumsiness (Adam, Ben, Iris, Monica and Ruth). Ruth expressed:

> "*They kept saying I was a clumsy child because I kept knocking things like the milk bottle. They used to put it on the table and I kept knocking it*".

Other participants commented that they were clumsy in the dark (Ben, Jeff and Sara) and Adam expressed "*my childhood it was like being a clumsy kid you trip over*". Adam, Carl, Debra, Iris, Jeff, Ruth and Sara expressed that their family noticed something was wrong, for example:

> *"Kept wittering on about the fact I was treading on the cat all the time, walking into things, I was covered in bruises (Sara)".*

Kate and Tia expressed their difficulties were recognised at school but for Carl, Debra and Eve, problems were identified by the optician. All participants, except Len and Tia, commented that they had experienced poor night vision or nyctalopia (nightblindness), which is a condition where people cannot see well at night or in dark conditions such as the cinema or at a club (Sense 2014a). Len reported: "I *was coming home from work one night and I got knocked down by a car*". Len's accident may have occurred because of his vision being affected by the dark, and this was supported by other participants' experiences (Jeff, Kate, Len, Nora, Pam and Sara), as they were also involved in accidents prior to being diagnosed with Usher, for example: "*that particular day I did land on the top of a car bonnet*" (Pam). Jeff also relayed an experience of being involved in an accident due to lack of vision:

> *"I did have a couple of silly little car accidents and that was only purely because I did not see the car in the field of my vision and went into it. Silly but that's erm at the time I didn't really twig I wasn't seeing very well".*

Nightblindness was a clear indicator that the person was experiencing something wrong with their vision but at the time not all realised the severity of the condition. Pam commented:

> "*I know and understand that nightblindness is probably the earliest symptom to manifest itself and didn't realise that I didn't see as well as my peers at*

night or in different lighting situations, that my eyes took longer to adjust, hence more accidents".

In this section, early life before Usher and its individual effect on participants has been reported. Also highlighted was that most participants experienced the effects of Usher prior to diagnosis, although not realising the significance of the seriousness of the condition. In the next section the impact of diagnosis will be presented.

4.2.2 Impact of diagnosis

Prior to diagnosis, some participants were experiencing sight difficulties combined with hearing loss. However, the reality of being diagnosed with a degenerative life altering condition such as Usher syndrome, with its complexities of dual sensory loss, had not yet begun. Sara's comment epitomises the change after diagnosis:

"*When I was younger I went for night walks along a cliff top and I can't see a f***g thing. As long as I was holding onto my mates shoulder right in front of me it should be all alright and we are right on the very edge of the cliff top having a night walk sort of thing. Yeah, erm yeah I think I was far far less cautious pre-diagnosis than after the diagnosis without a doubt".*

However although Sara became more cautious, Ruth did not:

"Make most of life be happy because there are worse things happening than losing sight and hearing. It's not the end of the world it really isn't erm I don't know I've learnt to live with it. I've done abseiling I've done everything".

Whilst participants shared varying experiences post diagnosis, for everyone, there were to be changes ahead that they needed to address. Tia, who was diagnosed with Usher at the age of 16 expressed the reality of these changes:

"Growing up in the past year or so has made me realise some of the restrictions that go with Ushers, although I have lots of coping strategies I will never drive, walk anywhere on my own, read a book again and that makes things different. I am also over reliant on my parents, teachers and support workers and that can be hard".

Several other participants (Adam, Jeff, Len, Monica and Pam) expressed the realities that Usher would present and the support they would need as the condition worsened. Many participants did not realise they were experiencing deafbindness or more specifically Usher and the impact of the diagnosis affected the participants very differently, for example, whilst Pam '*felt relief*', Sara found a diagnosis of Usher to be a shocking experience:

"*and erm he explained what it was and sort of said you'll go blind at some point in the near future and I felt like someone had punched me in the stomach"*.

Such a reaction is wholly to be expected when considering Sara's following comments:

"I was completely like just lost all sort of…I was like…couldn't …really stunned and I was so….(pauses)…it was horrendous really because you, you I mean I don't know if other people have said it to you but you have grown up being deaf and you always think there is a worse thing that could have ever ever happened and that's your sight! Because you rely on your sight so much for communication if you're deaf or partially deaf for lip-reading, sign language all that sort of thing. You think oh God if I lose the sight life won't be worth living".

Other participants also shared the implications of losing sight as well as hearing (Ben, Debra, Eve, Fred, Gareth, Harry, Jeff, Len, Monica, Nora, Pam and Tia), which could lead to a sense of bereavement and grief, as Sara described. However, the impact of diagnosis was not a negative experience for all participants. Where participants had experienced knowing there was something wrong but not actually having a diagnosis they felt a sense of relief. For example Quentin expressed:

> "*I wasn't surprised I just didn't know the name Usher syndrome, it might have been called enema syndrome for all I knew, it didn't make any difference to me what it was called and I was pleased it was recognised, I was pleased that I wasn't alone in this matter. So it didn't make any difference to me. How did I feel about it, it didn't make any difference to me, you hear about people getting really upset I didn't it was just having a name to something which made no problem for me at all. By that time, I had lived most of my time getting used to it, so it wasn't a big shock to me".*

Quentin reported that he had experienced sight difficulties from his earliest memories, so was pleased when he was diagnosed, at age 49 years. Furthermore, he used his expertise about Usher to raise awareness and campaign for equality of access and better conditions for people with Usher:

> "*I campaign a lot for induction loops*[47] *…With success*"

Pam commented about her diagnosis:

> *"Everything fell into place. It didn't freak me out at all. It all made sense. Life made sense to me for the very first time ever when I was diagnosed when I was 39 erm all the clues were there about my sight loss".*

Whilst for some participants, the diagnosis was a shock and for others a sense of relief, Len described his immediate acceptance:

"I was not devastated this might sound weird to you Michelle but it is not strange to me. I just accepted it you know what I mean. Erm I just seem to have accepted it. You know what I mean…I thought oh well, I am going to be blind in a few years' time. Erm [huffs] I was a bit upset but after that I was registered blind by a social worker at [name of hospital] after that say about 3 or 4 weeks, I had a visit from a social worker, all she brought was a letter saying you are now registered blind and she gave me a white [14] stick and that's all she did".

Throughout the interview, Len displayed a relaxed attitude to having Usher:

"*My mum was alive when I was first diagnosed but really, family um yes. None of them seemed to know anything it was just something you accepted my mum said 'you gotta cope' you know what I mean?*"

There are several factors which may affect a person's response to diagnosis, for example age, gender, individual personality, family support and familial attitudes. Additionally, the way the diagnosis of Usher is communicated will affect the individual's experience, for example, Tia expressed feeling "*really angry, following that sadness kicked in".* However, Tia's experience of being communicated the news may have contributed to these feelings:

"*The way I was diagnosed was shocking. My Advisory teacher for the deaf had picked up on my vision loss and advised us to get a referral to a specialist. When I met the first eye doctor she did all the basic tests and told me that we'd got it all wrong and that there was nothing wrong with my eyes aside basic short sightedness. We were over joyed and so relieved, then she said I'll just scan the back of your eyes to be certain, but I'm pretty sure you're fine! Of course the scan said otherwise, she then googled Ushers in*

front of us and said 'Yes I'm afraid you do have Usher Syndrome, it is likely you will go blind'. It was awful".

As Tia commented it was awful to be diagnosed in this abrupt way, but a further factor affecting her response to her diagnosis was her age as Tia was diagnosed aged 16 years, thus at a time when her adult life was beginning to develop, she was told she was "*likely to go blind*". Tia's experience highlights that the way the diagnosis of Usher is communicated will affect the individuals' experience and can have a significant impact on how the person copes with this devastating news. Eve shared her experience of receiving her diagnosis via a third party:

"I had an interpreter person yes and [name of person] with me and interpreter and the doctor told me I would go blind it was very emotional for me".

Eve's experience raises issues around conveying bad news through an interpreter and the impact this news has on an individual (Section 5.2.3 discusses this further). Eve shared that although she had been diagnosed with Usher 27 years previously, she still did not understand it, "*I don't know what that means, I don't know*". However Eve did understand that there would be deterioration in her sight:

"All I know explained sight get worse and worse and field narrower and narrower until can't see. If someone close can see I can't see anything when sun shining can't see it has to be in place without glare, just black shadow".

Eve's experiences highlight the importance of an individual having an understanding of Usher, to be able to be better prepared for the impact of it. Although the diagnosis of Usher impacts on individuals in varied ways, there is no doubt that this condition has a life altering effect. Another aspect to this was with regard to misconceptions of

some participant's (Oliver and Ruth), for example, Oliver believed that diet impacted progression of Usher:

> *"Throughout my life I was always under the impression that my diet had a profound impact on my condition as well"*.

Oliver further commented:

> "*My experience of visiting my doctor was a negative interaction, as he focused on my diet i.e saying that I had the condition because I was not eating enough carrots"*.

Even more extraordinary was Ruth's experience of receiving a diagnosis of Usher when she was 11 years old:

> "*My father nearly fell off his chair they said, they said it was um originally from VD [venereal disease-sexually transmitted disease]"*.

These experiences highlight that it is not only the impact of diagnosis that will be individual and unique but also the perceptions people have of Usher. The next section will explore participants' experiences once they had received their diagnosis.

4.2.3 Experiences after diagnosis

Whilst experiences at the point of diagnosis were not positive for Tia she continued:

> "*I have since been to different hospitals to see different consultants and the experience has been much better*".

Experiences after diagnosis varied for participants; for some the uncertainty about the extent of sight loss was dominant (Ben and Jeff) and others felt apprehension regarding transition from the hearing world into the Usher world. For example, Adam

commented: "*I am not sure with that initial stepping outside of the world and jumping into world around me*". Other participants diagnosed with Usher experienced low feelings, or feelings of depression (Eve and Gareth), and some had suicidal thoughts (Len and Pam).

Eve commented that she felt:

> "S*hocked very depressed shocked and depressed ...stress all hurt and stress. Held in didn't talk to anyone*".

When Eve, was asked if she had been referred to a counselling service she became confused and expressed that she did not know what was meant by a counsellor, she just "*held in*". Gareth shared that he sought professional help:

> "*I was young so um, you become emotionally upset erm think I had to seek professional help I was on Valium for a short time diazepam Erm that kind of thing, very upsetting*".

Whilst some participants expressed feelings of depression (Eve) for others the feeling intensified to feeling suicidal (Pam). Pam expressed:

> "*I was suicidal….. I had to give up dancing any way so...so um you know it did affect me*".

Len shared the following experience:

> "*I went to my GP. He sent me away to a place. Yes a psychiatric place. Yes. (long pause) because I had suicide, yes suicidal thoughts Erm erm. I was there for 8 weeks, yes 8 weeks. I came home I was still depressed and the doctor he prescribed anti-depressants. But um I stayed on as long as I could. I don't know if it did anything but I stayed as long as I could. Years went by and I took anti-depressant. Yes yes*".

These experiences highlight that there is a psychological impact when a person is diagnosed with Usher thus counselling services may be imperative to support some participants. However, these services need to be offered upon diagnosis, not waiting until a person becomes depressed before offering them, and also appropriate to the person's need:

> *"I asked if they had someone I could speak to at [eye hospital] and she was their counsellor, but really a counsellor that gave advice on benefits and things like that so I couldn't even think about things for goodness sake"* (Pam)

The majority of participants (Adam, Ben, Carl, Debra, Fred, Gareth, Harry, Iris, Jeff, Kate, Len, Pam and Quentin) considered that, after the diagnosis of Usher support from sensory specialist charities/organisations, for example Sense and Deafblind UK was essential. Most participants recognised the importance of the knowledge and support these organisations provide (see 4.5.4 for more detail).

After diagnosis Len became confined to his own home, but on one occasion he had to travel by train, as a family member had died. A person who worked for a specialist deafblind organisation was sitting in the same carriage and the participant and the worker chatted. The worker explained how organisations such as the one they worked for could support people with Usher. Len explained the result of this chance meeting:

> *"Yes 3 weeks after [name of town] this lady [person's name], sent me a letter asking if she could come and see me. I said yes of course you can. She did and she made me a member of [charitable organisation]. It was through [charitable organisation] I started to meet people and they had a connection to [charitable organisation]. So I got involved in [charitable organisation] in*

yes 1998, 1998. So that's how my life changed from being a virtual prisoner for 7 years".

It is quite astonishing that this participant experienced being confined to his own home for a prolonged period of time after his diagnosis. However, it is evident that becoming a member of a deafblind support organisation enriched his life. The essentiality of specialist charitable organisational support resonated with almost all of the participants.

Another aspect for attention after diagnosis relates to practical and financial support that a person would require as the condition worsens, for example:

"Just general support information for reading letters, equipment, making appointment, making telephone calls in my circumstance [a bit more with mum] telephone calls for legal stuff" (Adam).

Adam expanded by saying that people who are diagnosed with Usher need support on all levels: "*Realistic attitude as condition worsens e.g. practical, financial and level of support required* ". The final area involving experiences after diagnosis concerns safety.

Experiences highlighted concerns regarding advancing technology and the impact this may have on their safety (Carl and Debra). For example, as technology advances in terms of quietness associated with hybrid electric cars, crossing the road could become a problem. As already highlighted most deafblind people will have some residual hearing or sight even if it is minimal, however, as cars become quieter, any opportunity of hearing their approach will become significantly less. Carl commented:

"*As technology advances potential dangers arise e.g. hearing aids cutting out background noise which is essential for hearing road noise! How can we cross roads when the so called unwanted noises of the traffic is removed to a large degree, so it is so unsafe to use these type of aids! We are more inclined to stay indoors and not go out!*" (Carl)

These safety issues are a cause for concern because a major challenge for deafblind people and people with Usher syndrome is that of safe orientation (Sense, 2014a). If people feel unsafe to venture outdoors this will impact their emotional and physical wellbeing (Sense, 2014a). Carl was concerned that hearing aids (for more information see 4.5.3) were not able to cope with technological advancement:

"*They (the hearing aids) remove a major part of the background noise elements and it is quite seriously bad and dangerous at times to use these new hearing aids while trying to "hear" things like cars on the roads when the blooming aids removes the "noise" of vehicles rumbling along the roads! We have no choice in the matter – just live with it or else!"*

Additionally, Debra worried that if her home should be broken into because of her sight and hearing loss she would be unaware of it:

"*Few years ago decided I can't see its dark, walking into door, can't see CCTV didn't want to live alone any more. Two daughter share, better don't have to worry about people breaking in, so worried. Now daughter's responsibility they can hear"*.

Adam also expressed concerns regarding safety and relayed an experience where he did not see the train track due to his deterioration in sight and hearing:

"*on way to work I was on train line the train going to [name of station] rather than [name of station] and I was just completely off on my own and I realised*

on wrong train had to come back to [name of station] to get on right side and when I got to the flat I didn't really know where I was north or south then I found myself on the platform to the track I had to pick myself out of it. I thought oh my god I was really struggling didn't know where I was the train passed by and I thought this is really serious now".

Adam's experience highlighted that whilst safety is an issue for people who experience deafblindness/Usher syndrome, sometimes the seriousness of the sight and hearing loss will not be realised until an incident occurs. If individual need is considered and support offered at the level required for each person, then the experience after diagnosis may be a little more manageable.

4.2.4 Section summary

This section has presented findings about the 'diagnosis being the start of the experience' and included subthemes relating to: early life, life before Usher; the impact of the diagnosis; and experiences after diagnosis. Table 4.2 provided details of the age range of participants within this study, the age at which they were diagnosed, age at time of interview and whether they had related experiencing symptoms/ difficulties prior to diagnosis. However, whilst this table gives an idea of whether a person had experienced symptoms/difficulties or not, it does not mean that the participant, who did not report having experienced symptoms/difficulties prior to diagnosis, did not have them, it may have been that they did not share these experiences in interview.

As outlined though, some participants had symptoms or experienced difficulties prior to the diagnosis; once diagnosed the reality of the condition was realised and the effects were to become life altering. However for another participant the diagnosis

opened the way to try new adventures. The findings in this theme showed that whilst two people may have a diagnosis of Usher, no two people will have the same experiences as each and every person with Usher is unique and individual.

Three key messages emerged from the findings relating to the theme 'diagnosis is the start of the experience': Usher although not life threatening is life altering; the diagnosis of Usher impacts on individuals in various ways, and the way the diagnosis of Usher is communicated affects individuals' experience, these issues will be discussed in detail in chapter 5. Next the theme 'Familial relationships across the lifespan' will be presented.

4.3 'Familial relationships across the lifespan'

The previous section provided an insight into how 'diagnosis is the start of the experience'. In this section the second theme 'familial relationships across the lifespan' will be presented and will include sub-themes of: child diagnosed with Usher; siblings of a child who has Usher; being a parent who has Usher; relationships, and family support.

A family is defined as, "a group of people from common stock; set of relations living together and descendants of a common ancestor" (Thompson, 1996 p352). A family may also consist of adoptive children, looked after children [9] and kinship care [10.] . A relationship is defined as a "connection or association; an emotional association between…people; a condition or character due to feeling related and kinship" (Thompson 1996 p 854).Thus in this section, when referring to familial relationships, reference is being made to a variety of familial constructs. The first sub-theme to be presented is the child diagnosed with Usher.

4.3.1 Child diagnosed with Usher

Whilst some participants received their diagnosis as adults, others were much younger and so their parents were involved in how, and even whether, they were told. This sub-theme presents the child's experiences when the diagnosis of Usher is given. The findings reveal the differences between not telling the child they have a diagnosis as in the case of Kate and telling a child they have a diagnosis of Usher as experienced by Tia.

Kate highlighted the importance of telling the child they have a diagnosis of Usher and throughout the interview reiterated that "*you must let your children know*" rather than trying to shield or protect them:

> "*I was with one group talking about my situation you know the diagnosis with my parents not telling me and I was saying you must you know let your children know*".

To provide background to this comment the experiences goes back to when Kate was 15 years old. Kate already wore hearing aids and then she "*saw a girl with beautiful red spectacles* "and asked her father to take her to the opticians to get a pair. Kate's father did this, but the optician after carrying out an eye examination, referred Kate to ophthalmology at the hospital. Kate shared the following experience:

> "N*ow I was about 15 at that time erm. My mother took me to the hospital all I remember is being asked to wait outside whilst they spoke to my mother, I thought that was a little bit odd. Then she came out and er yes in her body she was very uptight actually. Held my hand and went, didn't speak didn't say anything at all*".

It later transpired that it was on this occasion that Kate's mother had been told that her daughter had been diagnosed with Usher syndrome and what the prognosis could be for her daughter's future. Kate was taking her O levels and her mother had been told that in addition to deafness her daughter was now going to experience sight deterioration with a possible eventuality of blindness, a shocking parental experience. Kate continues:

> "*Then we went into…we lived in [name of town] at that time and we went into the British Home Stores. There was a hat, it was a pink flamingo, sunglow hat and I just fell in love with it and my mother bought it for me. I thought that was a little bit odd. I have 3 sisters why me*"?

Whilst Kate did not say why her mother bought the hat, one possibility is that her mother wanted her daughter to enjoy colour and beauty whilst she was still able to see it, and this participant at the time of interview was aged 63 years and could still remember the pink flamingo hat even though her sight has significantly deteriorated. Kate at the time questioned why she had been bought the hat and later learned why, but not before experiencing what she described as other strange incidents:

> "*Shortly before doing my A levels 15 going on 16, I found myself being taken on a pilgrimage trip to Lourdes and erm you know actually met up with one or two other children who were seriously ill. One boy had leukaemia and he did actually die later and then a 10 year old boy had cancer. Other people I was with were quite seriously ill and I thought why am I here? You know…when I only have hearing problems*".

The fact that Kate so vividly described this experience 48 years later highlights the impact this experience had on her. However Kate discovered why her parents' behaviour had changed and the reason she was taken to Lourdes:

"Then when I was doing an assignment for one of my essays I asked my younger sister to go into my father's bureau to get some writing paper for me. She brought a pile back and in the middle was a letter. Written to my father from the hospital, the word distinct to me, I didn't read anything else the words it said slowly but surely going blind".

Although, Kate was then aware she would go blind, she did not confront her parents with her discovery and still believed she was dying. Kate then describes a family trip travelling by train:

"W*e had lots of luggage going up an there was a gang plank and I had trouble with my leg, I had shooting pains down my leg and sitting in the 6th form library doing my work and that I couldn't sit or do PE and I thought this is where I am dying*".

After this incident Kate spoke to her parents:

"So anyway [sounds really upset] I waited for my sisters to go to bed and then I approached my parents and said I had been thinking I was going to die. They were quite upset obviously and horrified and I said to them look you have to tell me everything you cannot hide things from me. It so happened that my mother was told at the hospital that I was going to be blind by the time I was 30. So they didn't know anything about retinitis pigmentosa [5] (RP)"

Kate's next comment was profound:

"*I thought it's only my eyes I am not going to die after all*".

Although only one participant relayed this particular experience of her diagnosis being kept from her, the experience highlights that it is difficult for parent(s) to know

when to tell a child they have a condition that will be life altering, especially when the parent is also advised that the condition is genetic/hereditary and they may feel a sense of responsibility themselves (see discussion in chapter 5). However, if the child is not told, then the child will reach their own conclusions, as Kate's experience highlights. Kate's experience has had a lifelong profound effect on her which has resulted in her continued work to raise awareness to educate parents and trainee social workers of the importance of telling a child they have a diagnosis of Usher.

However, another participant's experience demonstrated the result when a child was told of their diagnosis early. Tia's diagnosis was not relayed to her by her parents but rather a health professional, which meant that Tia's parents did not have the daunting task of relaying the news. However, Tia relayed that her "*sight loss has been rapid*", and an early diagnosis helped her to better prepare for her future. Kate's and Tia's stories, demonstrate that whether a child is told they have a diagnosis of Usher or not affects their subsequent experiences.

Another aspect was that a child may find it hard to express difficulties they encounter as a result of Usher, with their parents, as they do not want to upset them, for example:

> *"I've found it hard to express my difficulties as I have not wanted to upset them. Whilst they definitely want what is best for me, it has been hard for us to support each other when we have each been struggling with it individually"* (Tia).

It may be that the child is considering the parents' feelings and emotions as they are aware that the condition is genetic/hereditary. The fact that Usher syndrome is a genetic/hereditary condition arose frequently during the interviews (Adam, Ben, Debra, Harry, Jeff, Kate, Len, Nora, Oliver, Quentin, Sara and Tia). Oliver relayed:

"*My condition is hereditary, but is not treatable. The statistics show there are 354,000 Deafblind people in the UK who has a condition [of Usher]. I understand in terms of relationships the condition manifests itself, with my parents being carriers*".

Harry too was aware his parents were carriers:

"*The Usher gene was passed on to me because both my mum and dad were carriers of the gene but did not have the disease itself*".

The word 'carrier' used by several participants (Ben, Oliver and Harry), means "a person or animal that may transmit a disease or a hereditary characteristic without suffering from or displaying it" (Thompson 1996 p144). Where participants expressed understanding that their parents were carriers of Usher, they imparted this knowledge in a matter of fact way and the participants themselves did not appear to blame their parents for being carriers. It is essential to note that prior to giving birth to their children parents are often completely unaware that together they have a gene that can lead to their child having Usher syndrome. However, diagnosis and realisation that it is their genes that are responsible for the condition contributes to feelings of guilt as expressed by Fred:

"It took a while for her [mum] to accept it [Usher]. She felt guilty I think all parents feel guilty. 9 out of 10 times they feel guilty, it's a natural thing".

Tia echoed this view:

"*If I'm honest I think my parents have found it really hard to cope with what has happened to me. Obviously it is genetic and I think guilt has played a large part in it all"*.

Whilst in most cases participants received support from their parent(s) Sara's experience was different:

> "*Well I got a lot of support from all the family apart from my dad. I met up with him in a pub and told him and he told me it wasn't his fault or his responsibility and he walked out*".

These experiences highlight the differences in the ways that parent(s) cope with their child receiving a diagnosis of Usher.

Although Kate's experiences were many years ago, her comments highlight that parent(s)/caregivers and children may benefit from specialist support (Sense 2014a) as they may struggle to cope with, not only their own feelings, as they see and hear the difficulties that their child encounters on a daily basis, but also the awareness that there is no cure and that the future is likely to get harder for their child. Having considered experiences of the child diagnosed with Usher, the next subtheme presents findings about the siblings of a child who has Usher syndrome.

4.3.2 Siblings of a child who has Usher

The question of how siblings without Usher are affected when their siblings have Usher was also raised. Tia and Carl expressed that their siblings had been helpful with regard to the limitations imposed by Usher:

> "*They are young but have been quick to pick up what has happened to me and have been great guides etc*" (Tia).

> "*My mum and brother were very supportive and did much of the reading of the paperwork for me as by this time I could no longer read the printed form at any size any more*" (Carl).

However, Carl and Tia's experiences were from their perspective as a sibling with Usher, and not from their siblings who did not have the condition, as siblings without Usher were not interviewed as part of this study. Table 4.3 shows that of the 20 participants interviewed five had siblings with Usher, thirteen had siblings without Usher and two had no siblings.

Table 4.3: Participants who have siblings with/without Usher

Participant	Sibling(s)with Usher	Sibling(s)without Usher	No siblings	None mentioned
Adam			X	
Ben		X		
Carl		X		
Debra		X		
Eve		X		
Fred			X	
Gareth		X		
Harry		X		
Iris	X			
Jeff	X			
Kate	X	X		
Len		X		
Monica		X		
Nora	X			
Oliver				X
Pam		X		
Quentin				X
Ruth	X	X		
Sara		X		
Tia		X		

The findings also highlighted that where a person had Usher but there was a sibling in the family who had more demanding needs, the person with Usher's experience was that the attention and support was given to the child with the highest needs:

> *"I terrible speech problem. My sister terrible. Got the same thing as me. She's in a home now. She's got the same as me but she's got more and learning disabilities as well. I had to get on with it. With my little sister my youngest, there are 6 of us, she got all the help under the sun. She had learning disabilities and I was pushed to one side"* (Ruth).

Ruth's experience highlighted that when there is a child in the family needing greater care, attention or regular hospital/medical appointments for an ongoing condition, the child with the lesser needs will receive less support as parental/caregiver time is limited. Therefore it is important to ensure parents are aware of support to which they may be entitled, to support all their children's needs, for example,

charitable/voluntary organisations or local support groups that may be available within their area.

As table 4.3 highlights, within some families there is more than one child in the family with Usher. Although, no two experiences of Usher will be identical, there would be the shared experience of living with Usher and whilst it could be expected they would provide peer support, this was not an aspect they discussed. Having considered siblings and Usher, next the subtheme 'Being a parent who has Usher' will be presented.

4.3.3 Being a parent who has Usher

As identified in chapter 1, a person with Usher will have experienced D/deafness from an early age, and in many cases have been born with it. Therefore the person over time would have built up coping mechanisms and strategies to live life as fully as possible. However, the findings showed that when a person who has Usher becomes a parent additional difficulties are encountered, for example, Debra whose communication methods were visual frame BSL, hands on BSL and Deafblind Manual experienced not being considered suitable to parent her child:

> *"When daughter born mother said doctors approached my mother [participant's mother] and said wouldn't be able to look after baby. When daughter was 6 weeks old wanted to take baby away, social workers. I was no capable of looking after. Grandmother came to live with them to stop baby being taken away".*

Although, it necessitated Debra's mother's support to prevent her first child being looked after, when her second baby was born, Debra's experience was different:

"When second baby born, not talk about taking baby away but health visitor came every day to check. Social worker came every Friday".

Debra's experience was to continue with her family life:

"*Was happy, forgot was disabled, brought family up went with husband in car on holidays*".

Although, the outcome for Debra was positive, initially, her experience was a challenge and necessitated her mother's support. Ruth also experienced a negative perception of her parenting ability and again received her mother's support:

"*No. No. When I did have one of my first child erm they said I wouldn't be able to cope with her and all things like that. But we soon pushed him out the door. No. No social worker no my mum was left with it*".

Whilst both Debra and Ruth's ability to parent were questioned due to their sensory needs, Debra brought up her two children and as adults they remain in regular contact and provide support for her as she has advanced in years. Ruth mothered several children and is also in regular contact with her children. If these children had been removed from their mothers on the grounds that their sensory need impacted their ability to parent, these children would have experienced a very different life to the ones they currently held. However, these experiences must be contextualised within society's attitudes and legislation of that time. These two mothers were 82 years (Debra) and 69 years of age (Ruth). Today, with current legislation such as the Disabled Person Act 1986, National Health Service and Community Care 1990 Act (s47), the Equality Act 2010 and the Care Act 2015, parents with disabilities have legal rights and are entitled to an assessment of the needs and protection against discrimination on the grounds of disability. Having considered the experiences of a parent who has Usher, relationships will be considered next.

4.3.4 Relationships

Several participants (Adam, Gareth, Len, Oliver and Quentin) discussed the forming of new and maintaining existing relationships. For some participants knowing they had a degenerative but unseen condition affected how they felt about building new relationships (Gareth). For others, the inability to socialise in certain environments prevented them from forming relationships (Kate, Len and Oliver), and for others the effects of the lack of socialising had a profound impact on their identify development and ability to form relationships (Len).

Gareth shared how he felt about knowing that the condition was degenerative but unseen:

> "*I had a horrible dark secret, I knew I was diagnosed with RP and I knew what RP meant, it was like a horrible burden on my back that I couldn't quite get off. You know it made me nervous, very nervous*".

Usher is an unseen disability, it is only if a white stick or a red and white stick is used by the person with Usher that they are identified with having a sensory loss. Where sensory equipment is not used a person may not be identified as having a disability. Gareth although referring to Usher as a "*dark secret*", did not want to identify as being different and next expressed:

> "*I played table tennis I was brilliant at table tennis unless I knocked the ball right up in the air. They started laughing I had to try and work my way round that one and pretend I could see it you know I was behaving like a normal sighted person*".

Although, this experience highlighted Gareth's desire to live as a "*normal sighted person*", eventually the effects of Usher will manifest as the condition progresses.

Gareth shared that as a young man he preferred to appear drunk than to have a sensory disability:

> *"The moment I got to discotheque all the lights were dim. I just couldn't see. I was acting like a blind man and I just made out I was drunk".*

Quentin adopted a similar approach to Gareth:

> *"I covered up my inability to not see by pretending I was drunk, because if you are a young undergraduate [name of profession] it's quite alright to be drunk, but it's not OK not being able to see".*

Quentin's comment indicated that he may have felt stigma connected to being visually impaired.

The effects of nightblindness and difficulties seeing in certain environments such as pubs, nightclubs or discos etc. (environments with minimal or limited lighting) had a profound effect on participants with regard to socialising and identity development. Adam relayed the following experience:

> "*When I was going to the cinema with my friends the dim light like the contrast when they turned the light out I was getting glazed out hallucinating sparking out couldn't get my eyes straight out so that sort of scenario".*

Oliver also discussed difficulties with social activities:

> "*Socialising was always difficult, going out was difficult and interacting with people was difficult including going to a nightclub, cinema and restaurant/bar. It was very noticeable to people that something was wrong, that had an affect both professionally and personally*. *I would see people keeping their distance*".

The environment hindered relationship building for Kate as the inability to cope with noise and lack of light prevented her from socialising:

> *"I went with the girls to a house party in [name of town] and you know this day noisy music and darkness I couldn't cope in those situations at all so I couldn't socialise".*

This experienced highlighted how the environment impacted social activities and associated friendships and relationship building. Although, Kate was successful from a professional perspective, she relayed the following experience:

> *"I have not even had a proper boyfriend I might you know gone out once or twice with people but I never had a boyfriend".*

Pam had a similar experience:

> *"Everybody else had a boyfriend I didn't have boyfriend like in the same way and the thing is people know you are different".*

From these experiences it is clear that development in their early years can be influenced by the intrusion of sensory loss. The findings highlighted that Usher not only affects the forming of new relationships but also affects existing relationships. For example Fred shared that:

> "*Erm I only use email …err even then it's difficult to read. You can use certain software that talks to you but when I share a computer with [partner's name] it's not always easy. We don't have space for two separate computers so yeah got partner to think about*" (Fred).

This experience shows that day to day activities, such as computer sharing, are affected when one partner has sight and hearing difficulties and adjustments have to

be made. Fred also acknowledged that sight and hearing loss was a progressive degenerative condition and changes and challenges are ongoing:

> "*It's ongoing all the time, constant changes. The other family and spouses find it difficult to cope with changes"*.

Concern for a partner following diagnosis and progression of the condition was also expressed by Jeff:

> *"Where I could say, don't worry [wife's name] I can do the driving...dah di dah di dah. So [wife's name] is doing all the work (pause) so yes. And erm yes, erm I was happy wife would do all the driving. That meant I could drink at night time when we went out and wife could drive but it did admittedly mean I lost a proportion of independence*".

However, Jeff also shared his concerns:

> *"It worried me that if [wife's name] was out and had gone to pick the kids up from friends house erm erm I am not there to notice there is something not right how it should or shouldn't be and something would happen and I worried erm it was purely for her care nothing else than that"*.

So whilst Jeff was clearly aware that he had to accept the fact that his wife would have more responsibilities due to the diagnosis and subsequent progression of Usher, he was also aware that this raised concerns for him with regards to his wife's wellbeing and safety. To address this situation and alleviate anxieties and concerns Jeff made the following decision:

"*So I decided it would be good to have a Motability car and as you probably know from Motability cars it's you, you use up your benefit to go towards Motability and you have a car for 3 years now and every 3 years you hand that car back and if you want another car you continue the program of having new car every 3 years which means you have a brand new car that is erm road worthy, serviced by the garage you don't have to worry about it".*

Not all partners of the person with Usher were willing to offer support:

"I had no support from my husband. He has never offered any emotional or physical support, only very very recently this is no lie in the last 6 months there has been a change and I don't know why. When I was actually diagnosed he told me that he would do nothing to help me and I would just have to get on with it" (Pam).

Pam was diagnosed when she was 39 years and at the time of interview was aged 52 years, so she experienced 13 years without partner support. Diagnosis can have a devastating impact and to receive little or no support within their family could have a significant effect on the person with Usher. Having considered relationships, the final subtheme is 'Family support'.

4.3.5 Family support

Family support was clearly essential and was discussed on numerous occasions by all participants except Kate, Len and Oliver. Carl's parents encouraged him to have a positive view with regard to his deafness:

"*My parents have always told me to be positive and don't worry about things you do not have control over*".

Carl expressed a positive view of his sensory needs throughout the interview, whilst realistically recognising aspects where sensory need could present challenges. Jeff too showed how positive family support can contribute to well-being:

> *"I do know of a couple of members who in [Usher support group] have admitted they suffered stages of depression. Fortunately I have a very supportive wife, 3 great kids".*

However negative family attitudes with regard to sensory need, contribute to a negative impact on the person with Usher, for example Monica expressed:

> "*My parents taught me nothing. I have been bumbling through life, just bumbling through life*".

Monica expressed that her sensory limitations had significantly affected her life from education and career aspirations to marriage and family. Whilst she expressed that she now felt in a good place in her life, greater family support at an earlier stage could have been beneficial.

Although lack of support can impact the person with Usher, so too can overprotectiveness, which Fred described:

> *"Trouble is my parents became overprotective because they were worried about me"* (Fred)

In contrast to overprotectiveness in parents, other participants' experiences were that their parents did not want to accept that their child had a disability, for example:

> "*Then my mother she, she didn't really want to know I had hearing problems. And she used to say, you can't hear because you always got your head in a book you see. And my father being a big man in the army did not want his*

son to be disabled so for a long time it wasn't accepted in the family that I had problems" (Quentin).

When families do not treat their children any differently because they have a disability this could contribute to a child just being another member within the family and more resilient in the world outside which may not be so understanding:

"They were quite supportive (family). They took the mickey out of me a bit. I was just one of the family" (Gareth).

Gareth expressed in the interview that his parents and his sibling had been supportive with his condition and that the family was a safe space to go to when the condition worsened and he was in a position where he could no longer work. Findings from this section support the idea that each family will deal with their child's disability in a different way.

4.3.6 Section summary

This section has presented findings about 'Familial relationships across the lifespan', which included the four subthemes of: child diagnosed with Usher; siblings of a child who has Usher; being a parent who has Usher; relationships and family support. Participants revealed the differences between telling and not telling a child they have Usher. Whilst it is difficult for a parent to communicate bad news, participants' experiences showed consequences that can occur if a parent does not tell their child about their condition and the child's ability to be prepared if they do. The effects of Usher with regard to siblings showed how siblings without Usher could be supportive. Participants shared the impact of Usher being hereditary/genetic, and the effect it had on parents from the perspective of the child who has Usher. For those participants diagnosed as a child with Usher, they were

unwilling to upset their parent(s)/caregiver around their awareness that Usher is genetic/hereditary as their parents had their own feelings to cope with.

A further family issue highlighted was that parenting capacity may be doubted when a mother has sensory requirements/needs. Both mothers' experiences presented in this section occurred some years ago and such practices may not occur today, however, social workers need to be sensory aware to ensure best practice and best interests of the child and family. The findings highlighted that relationships were affected because, although Usher is unseen, the prognosis is life altering. Some participants with Usher expressed that they were not 'normal' because of their sight and vision difficulties but they did not want to be labelled as being different, especially if they felt there was stigma attached to the sensory impairment. The final subtheme, familial relationships across the lifespan, highlighted that positive family support resulted in a positive attitude to Usher. The findings indicated that most families support the person with Usher, but a lack of support not only affected the participant's attitude to the diagnosis of Usher but also the way they coped with living with Usher on a daily basis.

Two key messages emerged from the findings in this theme: A diagnosis of Usher impacts on new and existing relationships and Usher being an unseen hereditary/genetic condition has consequences for the person with Usher and their families. These messages will be discussed in greater detail in chapter 5. Next, the third theme, 'A sense of belonging' will be presented.

4.4 'A Sense of belonging'

The third theme 'A sense of belonging' included two sub-themes: Interrelationships between communication, culture and community; and friendships and shared experiences.

4.4.1 Interrelationships between communication, culture and community

The differences in communication methods within the Deaf community lead to different expectations from people who consider themselves to be culturally Deafblind as opposed to those who would consider themselves to be deafblind. Debra, when talking about her position within the Deaf community commented:

> *"All Deaf, same culture same, happy together*".

However, Gareth expressed:

> "*I go to [name of support group] member's day who use sign language. Don't have much to do with them they keep to themselves in their own little groups*".

Similarly, Jeff's experience was that:

> "*The Deaf community not at all, they are a world to their own".*

Being part of the Deaf community is a strong bond for people who consider themselves to be culturally Deaf. The findings showed that even after a diagnosis of Usher, capital 'D' Deaf[15] people will see themselves as culturally Deaf with its linguistic difference, history, culture and sense of belonging. Debra expressed:

> "*Few years ago social worker came to help me, what do you want me to put on the form deafblind? No Deaf after that the sight got worse. See myself as part of the Deaf culture with culture, history".*

Debra expressed that shared communication with other people whose first language is BSL[6] made her feel at ease:

> *"Communication is easy I feel good, same intelligence, same side."*

However it was not only shared communication that contributed to being part of the Deaf community but also a shared view of humour and shared understanding of culture, linguistics and history:

> *"Hearing world I can't pick it up comedy and laughter it means nothing to me I don't understand but Deaf laugh at same thing"* (Debra)

However, Fred's experience was different:

> *"I had peripatetic teachers mostly from the deaf school to the hearing teachers just keeping check on you. You feel so different. There was a time when I went back do the deaf school for a week or two because they knew my English and maths not so good. That made me realise there is no way I want to be part of the Deaf community at that time"*.

Several participants (Adam, Ben, Mary, Fred, Gareth, Harry, Jeff, Monica, Nora, Quentin) linked being part of the Deaf community with the ability to sign. Harry felt that the ability to sign was dependent on opportunity to use it:

> "*I did try to learn some BSL a few years ago but with no one to use it with soon forgot it"*.

Ben felt that whilst he had been part of the Deaf community this had changed with the progression of Usher:

> *"Yes I did but now I don't. Deaf people see things in a different way. Now I can't see that well. I haven't seen any of my friends from [name of school] for a very long time".*

When other participants were asked about involvement with the Deaf community they commented:

> *"Deaf community. Hardly at all. The Deaf Community uses sign language and I can't use sign language"* (Jeff).

> *"I find Deaf people are locked in their own little world, specially when signing so I didn't bother going*" (Nora)

> *"When I came to [name of town] yes there was a Deaf group, I went to. But they did all sign language and I am not a signer so I didn't go much"* (Monica).

These comments again highlight that the use of sign language as a communicative method is linked to being part of the Deaf community. However, not all deaf people with Usher see themselves as part of the Deaf community and not all necessarily use sign language.

Quentin was adamant that he did not consider himself to be part of the Deaf community expressing, "*I don't even accept the term Deaf Community*", going on to explain:

> *"Not because I am against the Deaf Community but the only community I live in is the normal UK community".*

Quentin further clarified:

> *"Because it's completely against what I want, because I want integration"*.

Another finding relating to the uniqueness of cultural Deafness pertained to a personal identity number being given to each Deaf child upon school entry. Having

worked in a sensory environment for many years my understanding was that a child being given a number upon entry to school had been a negative experience as it took away the child's identity as rather than having a name, the child was now just a number. However, Debra expressed that using her identity number was positive and she still used it to this day:

> *"Yes still use number now with friends"*.

I was surprised by this and asked Debra why she used it:

> "*Because of speech and lipreading one girls name was [girl's name] I was [participant's name] [names are similar], so used numbers instead it made lip reading easier"*.

It transpired that at school the friends referred to each other by number and still used these numbers allocated as children at the Deaf club:

> *"Yes at Deaf club use them. Said friends name by lips but sign number. Example [participant's name and number]"* (Debra)

When Debra was asked how she felt about using a number to identify herself she responded:

> *"Looking back likes it because it's similar to we recognise the voice of someone we have not seen for long time, but for me it's the number that stays with that person. Yes, can't recognise voice, so number is added thing to that person, add number who they are I think fantastic"*.

Debra continued to further highlight the practical benefits of using a number as a means of identification:

> *"On clothes had numbers, could not see sometimes took wrong clothes. All remember number and all excited"*.

Overall, there were varied experiences and views about belonging to the Deaf community but these were closely linked with communication methods used by individuals. Therefore, the next aspect to be considered is how a change in eyesight can affect relationships with friends and family.

As BSL is a visual form of communication, acquired deterioration of a person's sight affects their ability to communicate with the people they would usually communicate with. Ben expressed:

> *"Then after that I went to the [name of school] School in [name of town] which is a signing school …..I can't anymore it just doesn't make sense I can't see where the hands are supposed to go".*

Although Ben stopped seeing friends, Debra dealt with her situation differently. Debra had been educated in a school for Deaf children and her first language, BSL, had been used since birth. Debra was culturally Deaf and had experienced a life-long sense of belonging from being part of the Deaf community. However when her eyesight started to deteriorate she found that it became harder for her to communicate with her friends as sign language is a visual language and she relied on her sight to receive information. Debra's friends had not encountered a person who used sign language to communicate but then was no longer able to see what they were signing. Debra relayed:

> *"They don't know how to communicate with me"*.

Debra's friends stopped communicating with her and she became isolated and not wanting to attend the Deaf club, even though she had attended it her whole life. Here are extracts from her interview, expressing how her deteriorating sight affected her being part of the Deaf community:

"Prob this year friends stopped talking. April last year worked as treasurer in Deaf club really happy and over years moved when more quiet. Didn't feel part of (Deaf) community, things started to change. Realised not seeing Deaf friends. It's culture. Life is ok but need friends. …. Didn't want to see Deaf friends but daughters said should. When went back to Deaf club quiet again. Coffee morning all Deaf came up and tried to talk, but couldn't. Realised friends attitude was changing". […] *"Friends came and tried to talk but can't see them. So they just said hello will come back and talk later but didn't. Didn't get any news. Never came back. Felt not included"*. […] *"Next time coffee morning daughter went to see what happening and find out what going on with light etc. Daughter watched. Went to person in Deaf club and said have to learn to communicate in different way. Sign language not enough"*.

At this stage Debra's daughter tried to resolve the situation but was not successful, so a charity that supports and campaigns on behalf of Deafblind/deafblind were asked to attend a meeting at the Deaf club to explain what Usher syndrome is and how it affects a person's communicative method:

"Meeting at [town name] came.[name of worker]and[name of worker] explained what Usher syndrome is and genetics, history what it means how sight gets worse" (Debra).

Debra explained how she felt:

"Then I talked and explained how I felt, wanted to talk with them, know it's difficult, need hands on".

Debra then explains, that although her friends realised there were communicative difficulties, they did not know how to resolve them:

"*After that friends realised. They did not realise problem. No one really understood how bad it was"*.

The person who attended the meeting from the specialist D/deafblind charity then demonstrated how little vision she had. Debra relayed the following experience:

> *"Did something very crucial stood at front and showed how little she could see. Asked all to stand up and brought arms in to show how much she could see. All the Deaf stood up. [Name of person from D/deafblind charity] show me with your hands where you can see and they just all copied [name of worker]. They really didn't understand what she was trying to explain. [Name of worker, colleague of person carrying out demonstration] said they are not understanding. Man named [name of Deaf man] he got it he came out to front and told people off. Said just like sheep, stop, think. Did it again half got it. When the Deaf got it [name of friend] a Deaf friend burst into tears, she had no idea there was just no understanding. Remarkable been Deaf all their lives, they come across people with Usher syndrome and don't understand totally remarkable".*

This experience highlights how sensory changes affect relationships and that communication is affected by acquired disability. The loss of sight together with already having loss of hearing significantly impacts communication and as a result did impact these relationships**.** However, where awareness is raised and people are made aware that alternative communication methods can be used such as hands on signing [7] or deafblind manual 31[17], these obstacles can be overcome, as was the case in this situation. However, Debra's experience does highlight the essentiality of family support. This participant was in her 80s and had lost confidence. She had her family to advocate on her behalf and knew which sensory charity to approach for

support, but another person in the same situation may not have had this support network and therefore the outcome may have been different.

Having discussed Deaf culture and its significance in belonging, the next aspect to be presented is whether there is an Usher culture. As previously discussed, there are two groups of people who experience D/deafblindness, those who are culturally Deaf and whose first language is BSL and those who previously had speech and were part of the hearing community. Adam's experiences identified two Usher cultures, one group who sign, and the other group who are hearing and sight impaired and communicate using speech:

> *"Yes, there are two Usher culture. The Usher cultures which are more with sign language Usher if you go down to probably made contact [name of deafblind charity worker] or [name of deafblind charity worker]. With the network 2 group in Usher sign language Usher and the HIS-Hearing and Sight Impairment. It's a little subgroup basically when I became member of [deafblind charity) the Usher was sign language group they had own AGM Xmas parties twice a year and one in [month of year] and one in [month of year] we all get together 2 groups like Manchester united and Manchester city, like East Berlin and West Berlin. I am in both groups".*

The idea of whether there was an Usher culture was not expressed by any other participants in this study. However, being able to sign is linked to being part of the Deaf community, so if the communication method used affects community affiliation, the notion of Usher culture is likely. Next, the impact of shared experiences when living with Usher will be presented.

4.4.2 Friendships and shared experiences

Len's experience highlighted the isolation experienced as a result of Usher. However, this subtheme shows how people living with Usher are united by shared experiences in which support networks are put in place and friends made:

> *"The Usher and the [name of group] do try to bring people together, made trip to [name of Palace] and museum arrange event for people to join in"* (Adam).

Some participants wished to be more actively involved in the charity and events organising, whilst others were happy to just be part of the group:

> *"Now with [deafblind charity], I got in touch with a lovely lady [name of worker] from [deafblind charity]. She said there was this support group. It was actually just a bunch of people with hearing and sight issues and they meet a few times a year. Social activities, you can share your experiences and learn from them, how you cope"* (Jeff).

Jeff's comment highlights the positive role of specialist Deafblind/deafblind charities in the lives of people who experience D/deafblindness. A diagnosis of Usher can lead to isolation (Len) but charities signpost people so they can meet other people with Usher. Meeting up with other people who have Usher can have three-fold benefits; the person feels less alone; they can learn coping mechanisms from shared experiences; and they can learn what support may be available. Jeff expressed that when he was in a situation where he had to decide to accept or decline a particular area of sensory medical intervention, whilst he did his research to decide, he also discussed it with his support group:

> "*Having association with the support groups you learn things you don't learn if you just do research*".

Jeff asserted:

> *"I am a great believer a problem shared is a problem halved. So talking to people who have similar like-minded problems is a great asset"*.

However, Jeff also acknowledged that everyone has different personalities and some people may not want to talk about their situation, but he further explained the possible benefits of sharing experiences with like-minded people:

> *"Now a person may not be comfortable to talk about it at the time. It is about gaining trust and bonding with people. But it opens up the door or the window whatever you want to call it. People may say I don't have problems with my eyes we all have problems with our eyes. But…what is the specific problem with your eyes? Oh I get this sort of this oh yes….yes then you are talking about it. If you don't talk about it people worry. I am not suggesting that they do, but people might think whatever it is problem could be life threatening it can bring on stresses to a person so yes. Like-minded people".*

Kate also expressed:

> *"I am going to [name of town] in 2 weeks' time. This Friday I am going to [name of town] after that I am going up to[name of town]. I am … I mean yes I am getting busy I enjoy it and it gives me a chance to meet other people".*

Some participants travelled some distances to meet up with other people with Usher (Debra, Fred, Gary, Iris, Jeff, Len) with whom they could share their experiences and learn from others. Additionally where a common experience was being shared age was no barrier:

"Club that was nice a lot of people object to being with older people I was in my 30s late 30s 39 it never bothered me how old somebody is if you have common ground" (Pam).

Pam continued:

"I found that very interesting. Um err it it gave me a lot of information about how to communicate effectively because I am very good my central vision I call it my central vision …I had one tunnel that went one way and another tunnel that goes the other way. A lot of people say about central vision but mine has never been straight ahead that is why I had problems socializing but I learned a lot of things about ways to communicate because some peoples type of hearing loss very different to mine and they need different tones to the voice".

Ruth expressed how meeting up with other deafblind people had a positive and inspirational impact:

"It wasn't until I met the deafblind people and they were all doing things and I thought I want to have a go at doing it. If they can do it I'm doing it and I did [sounds proud]. This is where we have got to get them all in to meet other people and to other people you think I ain't as bad as them. Join them".

However, whilst friendships and shared experiences were made by meeting face to face and the support of specialist charity support groups, the findings showed that the use of technology was another portal for sharing experiences. The introduction of technology such as Twitter [20], Facebook[18], FaceTime[19] Skype [21] iPad[23], IPhone[24], Kindles[25], speech to text[26], video conferencing[27] and Email [22] mean a person with Usher can access people with the same condition on a wider scale. The study findings showed extensive engagement with these technologies as a way of

communicating, developing relationships and sharing experiences. Adam shared how Twitter had enabled him to access people living with Usher, all over the world:

> *"Now we have Face book and Twitter now I am making more network across USA and Canada. Made contact with D/deaf organisations around world"*.

Tia shared:

> *"I use Facebook frequently, amongst other things it's a great way to communicate with other people with Usher/ RP who I might not have otherwise come across. I use email also but do not have Skype"*.

The type of technology chosen by the participant was dependent on their level of sight and hearing loss. Tia highlighted that technology does make a positive difference to living with Usher:

> *"Technology important for someone with Usher it keeps us in touch with the sighted/ hearing world. For me it allows me to write (touch type) read (screen reader) and braille (braille note) easily in a mainstream environment"*.

As identified previously, for some people who experience D/deafblindness, being in touch with the sighted/hearing world may not be their environment of choice; however as is evidenced by Tia's comment, it is very important for some people. Adam shared his experiences of social media:

> *"Not all bad now have Facebook and Twitter so good. I mean I am on Facebook every night then I am on the group, given a couple of one liner's and world people are not alone, they have gone through something. One person the other day said had bad day started screaming her head off couldn't do things around house and then it's like I say I have been there I have done that then you can have a little laugh. You are not physically there but Facebook…then you try to erase bad feeling and put icing on top. Don't*

worry it's not end of world. Reassure and encourage be supportive they feel better".

Another finding that emerged was that some people at the time of a diagnosis of Usher, may not want to know further information or meet like-minded people but technological facilities such as Facebook are readily available 24/7 and can be accessed whenever the person with Usher is ready to do so. This was expressed by Sara:

"When I was first diagnosed I like I say I didn't want to know anything, I didn't want to know anyone but there came a point when I did wanna find things out and to be honest the only way I was finding things out was to be honest searching through Facebook for other people with the condition or other people that know about it".

Facebook enabled Sara to access information at a time that was right for her. Technological support mechanisms such as this may be useful for people with Usher to find information, obtain support, and facilitate friendships with like-minded people with the same condition, when they are ready and without formal involvement.

Whilst Len's experience of support from a D/deafblind specialist organisation was positive, for others, such as Sara approaching this conventional method was not for them. Despite recognising their value, they did not want to be affiliated or involved with organised deafblind charities, but rather opted for social media options such as Facebook:

"I have met a lot of people via Facebook and thought we'll stay in touch but thought not with a barge pole and they do sort of meet for drinks that sort of stuff but not via [name of deafblind Charity]. These are also people who do

not feel that [name of deafblind charity] or [name of deafblind charity] is for them. [Name of deafblind Charity] are doing something at the moment where they are trying to bring those people back into the fold. I don't know maybe I have a very old fashioned idea about what [name of deafblind charity] is all about "(Sara).

Debra aged 82 years conveyed experiences of using FaceTime to communicate with her daughter using BSL, "*iPad FaceTime fantastic. Technology important, Facebook Email"*. One example given of using the iPad was where Debra and her daughter communicated using BSL when her daughter was in a pub and Debra was in her bedroom. Debra said it was sometimes easier to talk on FaceTime because it was, *"very well lit, good back light"*. It was interesting to note that it was encouragement from a D/deafblind charity representative who demonstrated the communicative benefits of an iPad for a D/deafblind person that prompted the purchase:

"Bought iPad next day. Very clear. Very clear. Whatever doing will flash up on screen when it rings. Find Skype OK but FaceTime better. Deaf friends have iPad too" (Debra).

However, as previously highlighted technology will vary from person to person depending on their sight and hearing loss, as highlighted by Kate:

"I can't cope with it. It's the first kindles that came out they are not suitable for me the other ones iPad you can have kindle going on there and I was shown can adjust font size. The only problem is with the icons running finger across them. I thought I would like to get one. I miss reading, reading books, also with writing class we have to do exercise of writing and reading it out afterwards I did this but of course with my writing I use black ink but couldn't see to read it. If I had I pad I could see to write on it and also read it back".

The findings highlighted the resilience, open mindedness and adaptability of people with sensory needs/ requirements when learning new skills to adapt to sensory needs. For example exploring tactile options instead of vision reliance and exploring options available with regard to adjustable sound and vision (Debra and Kate). For some participants, as highlighted by Kate, the deterioration of their sight and vision resulted in them losing out/missing out on something they had previously enjoyed, for example reading books. In some cases although not all, technology can be used to address some of these issues. The use of technology was not affected by age. Whilst the youngest person who used technology to access a wider network to contact people with Usher was 18 years, the oldest person in this study was 82 years. These experiences highlight that a person is never too old to learn new things, especially if it enables continued contact and communication with family, friends and society in general. Len further commented that he had not needed technology up till now, but as friends started to use forms of technology for example email, this affected contact and communication:

> *"Up till now I have managed to go along without it. I find I miss out most of my friends are on Email. Sorry I just can't get into it"*.

So to summarise, for some people living with Usher, technology opens up a whole new world and enables them to interact with a wider group of people and access environments that otherwise would have been unattainable. However, technology for some participants was not considered so beneficial and so personal choice is important.

4.4.3 Section summary

This section presented the findings about ‘A sense of belonging’ and explored the subthemes: Interrelationships between communication, culture and community, and friendships and shared experiences. Although as the findings highlighted, some participants did not feel they were part of the Deaf community, others did. For those who did, it is not about segregation or merely a shared communication method, but rather being part of a culturally Deaf community complete with a shared understanding of culture, linguistics, history, storytelling and humour which contributes to a sense of belonging.

Also, presented was the possibility of whether there is an Usher culture. As there are two groups: the hearing and sight impaired and BSL user groups who communicate in different ways and participants related different experiences, it is likely that there would be two Usher cultures. However, further research would be needed to explore this possibility.

Also considered was how friendships were established and experiences shared through the use of technology and its impact on the lived experience of people with Usher. However, although technology can have a positive influence for people with Usher, it is not acceptable for everyone so there should not be assumptions about the use of technology.

Three key messages emerged from the findings within the theme ‘A sense of belonging’: Acquired disability affects communication and in turn, relationships; Deaf/Deafblind culture can contribute to a sense of belonging, and that technology can support a sense of belonging; these will be discussed in chapter 5. The next section presents the final theme ‘Experiences of professional support’.

4.5 'Experiences of professional support'

The fourth and final theme, 'Experiences of professional support' presents findings concerning: education, employment and benefits; sensory support; sensory equipment/guide dog provision, specialist charity support and raising Usher awareness. The first subtheme to be presented is education, employment and benefits.

4.5.1 Education, employment and benefits

4.5.1.1 Education

Most participants (Adam, Ben, Carl, Gareth, Harry, Jeff, Kate, Len, Nora, Pam, Quentin, Ruth and Tia) expressed that they required reasonable educational adjustment to meet their sensory needs. Ben shared positive experiences of support he received in the partially hearing unit (PHU) in his primary school and Tia expressed that her support from school was "*incredible*". However, Nora had less positive educational experiences:

> *"I did have problem hearing and understanding what teachers was saying – Fell behind with my education".*

Oliver found innovative and unique ways to adjust to his situation within the educational environment:

> "*I started to lip read at age 12/13 as I struggled at school and I wanted to find a way to get to grips with what was being said. By this I found an alternative method to grab information by lip reading* [1]".

Although, Oliver utilised this method of communication, lip reading is a skill that not all people who experience D/deafness can achieve. The method enables a D/deaf person to look at the person's lips they wish to read (e.g. teacher, doctor, friend, family etc.) in order to receive the information conveyed. This can be difficult in

some cases, for example where the lips being read have extensive facial hair, do not move very much or are overly expressive. Also in a teaching situation, the teacher would need to be aware not to obscure the view of their lips, by, for example putting their hand over their mouth.

Quentin also utilised a variety of methods to maximise learning in an educational environment, commenting:

> *"I used to use all kinds of substitutes. I always sat at the front of the lecture theatre you know and I couldn't hear when anyone turned their back to me. When the teacher turned his back to the class so I had to follow other people you know. So I just got by"*.

This experience highlights the importance of educational support to enable a young person in a learning environment to achieve their maximum potential as opposed to '*getting by*'. Also important was the provision of sensory equipment and human aids to communication, as discussed by Adam, Ben, Carl, Len, Monica, Pam and Tia. These aids included hearing aids (Len), advisory teacher for the deaf and advisory teacher for the blind (Tia) and note taker and equipment such as neck loop [2] (Adam). Tia's experience from an email transcript, demonstrates aspirations that can be achieved with the appropriate support:

> "*My support from my school/ 6th form has been incredible. I have the most amazing support workers whom I owe my survival in a mainstream to. Despite losing pretty much all my useful vision in 18 months I completed 8 successful GCSEs, am about to sit my A-Levels and have been accepted at a [name of university] to Study History and Politics. I owe a lot of my achievements to my two SWs and my amazing, versatile teachers*".

Whilst some participants were able to adapt and glean the best from their educational experiences, others were not. For example Nora and Len's experiences resulted in less positive educational experiences. Len shared the experience of being bullied when wearing sensory equipment (a body worn hearing aid [3]).

> "*Yes that was what I had. I used to get bullied so I wouldn't wear it at school. I was very self-conscious of it I really was. In the end I had to leave it at school in the head master's office I had to go down to the office and collect it each day. Because if I had two at home I would have left it at home. I got the cane two or three times for not wearing it"*.

Len portrays a very difficult situation for a child to be placed in, bullied by other children for wearing a hearing aid, and disciplined by the school for not wearing it. Where participants did not experience receiving an individualised approach to education to meet their sensory needs this impacted their concerns regarding obtaining employment, as highlighted by Nora who commented "*because my education wasn't good I was worried I wouldn't get a job*". In the next section benefits and unemployment will be considered.

4.5.1.2 Benefits and employment

It is likely that educational opportunities received as children and young people affect employment opportunities in adulthood. The previous section highlighted that where children have a sensory need the support received or not received within education can significantly affect not only confidence with regard to applying and seeking for employment, but also achievement (Nora and Tia). Whilst several participants were initially in employment (Carl, Fred, Gareth, Jeff, Kate, Len and Nora), as Usher progressed and became more degenerative this affected their ability to work (Gareth and Jeff). Nora explained the effect on her work:

"*My last job became more difficult as my eyes were deteriorating and I started working with new staff there were not as understanding and helpful as my previous colleagues, this made things more difficult hence early retirement*".

However, although some participants were medically retired due to their dual sensory loss, Gareth encountered a very negative experience when required to attend the job centre:

"*I was forced to go to the job centre; my brother had to take half a day off. I went there with a red and white [13] stick and a personal listener wires I used headphones, she still said to me at the end there "could you please look at the monitor and check what I have written". I started waving my stick around and said I can't see the monitor let alone the text. She said we still think you should apply for work. I don't need all this sorry I put letters in front of her from [specialist eye hospital] hospital form the [Ear Nose and Throat hospital]l and then she just ignored it all*".

The participants involved in this study have a recognised long-term condition often supported by medical evidence. Fred, Gareth and Jeff expressed that they were very upset that they were no longer able to work in their chosen careers or provide as they had previously for their families.

These findings portray difficulties encountered when living with Usher, and the effects on individuals. Next, specialist sensory support will be presented.

4.5.2 Sensory support: audiology, ophthalmology and social work

The findings highlighted that people who are living with Usher, require support from a variety of sources, including audiology, ophthalmology and sensory social work.

4.5.2.1 Audiology and ophthalmology

Whilst the participants at one time or another attended audiology [4] and ophthalmology [5] it became apparent that experiences differed, for example, Jeff felt that it would be advantageous for audiology and ophthalmology to provide a dual sensory loss clinic for the following reasons:

> *"I think it would be better to be joined up. I get the perception that if I have hearing problem they are not taking into consideration my eye sight problems".*

Sara too initially agreed with this:

> "*I think they would be erm because whenever I go to [name of hospital] for the RP although they obviously know about the condition and they know its Usher syndrome and they know it's a dual sensory loss and they know it does has an impact each sensory loss has an impact on the other one, they don't actually take it into consideration if that makes sense. So they have never asked about how the hearing is and it's kinda like you kind of think to yourself as it's a condition that involves the two it would make sense to have an idea what each one is doing*".

However, Sara then went on to express that she felt it better to keep hearing and sight hospital appointments separate for the following reasons:

"*Would it weaken their expertise in terms of eyes if they had other sensory sort of experts there because that is where the funding has got to go as well*".

Tia concurred with Sara's views and offered the following experience:

"Both departments have not spoken to each other (to my knowledge anyway) but I do not think they've needed to, both have managed their specialism well".

However Monica expressed, *"there should be a department just for Ushers…. Then people can get help and support".* Several participants (Jeff, Monica, Pam, Quentin, Ruth and Tia) experienced separate appointments for hearing and sight; only Debra had attended a clinic which was tailored to meeting the needs of a person with dual sensory loss.

"Only one in country that has dual sensory hospital. [Name of charitable organisation worker] said about clinic, don't know what can do. Glad did. GP referred to clinic. Had to ask for funding as not live in [area where dual sensory loss clinic is located]. Services not in our area".

Debra's experience raises issues regarding equity and fairness of service provision as only a small minority of people with Usher have access to dual sensory loss services. However, with the current economic climate and cuts in health and social care, it may not be realistic to expect a change in the way services are delivered:

"*But you know I think the communication about the whole aspect of Ushers would be good within…probably not gonna happen anytime soon given all the NHS cuts! At the moment*" (Sara)

Harry highlighted the benefits that can be received from the audiology department:

"*I also asked at the Audiology dept at my local hospital and they said they would send me details of lip reading and sign language courses*".

Thus whilst sensory issues need addressing from a medical perspective, there are also social and communication issues that need considering. Overall, participants had varied experiences with regard to audiology and ophthalmology.

4.5.2.2 Sensory social work

Whilst sensory support and sensory social work may at first appear to be separate, the findings highlighted that each of these areas impact and support the other as often multi-agency work is required to support a person with Usher.

A sensory social worker provides a specialist area of social work and works with people who experience sensory impairment, which is the loss of one or more of a person's senses e.g. sight, hearing, touch, taste etc. However the participants who contributed to this study experienced sensory loss in relation to the combination of sight and hearing loss (deafblindness/Usher). Thus whilst a sensory social worker would practise within guidelines required for a registered social worker (HCPC, 2012), the primary focus of their practice is with people who experience sensory needs/requirements. The experience of people with regard to sensory social work support was diverse:

"I didn't get referred to anyone or anything erm… I was referred to the local hospital, to [name of hospital] erm and they kinda told me my diagnosis and stuff and said you'll have to come back (pause) for check-ups once a year and then my ..obviously my GP knew and then um…I have over the years spoken to the GP about like counselling and stuff because I think that sometimes you do need to talk about these sort of things to the professionals

and they've said there is no need for it (laughs, sadly). They said it's a medical condition rather than an emotional need and all that sort of stuff. But then I went and saw someone because I thought oh well ...it was something on a sliding scale then, and they said they didn't think I needed to speak to anyone because I was dealing with it very well. I said well yeah I am, but this is the time to speak to someone else because I have got the awareness to try and seek help before I sink into some kind of like pit, do you know what I mean?"(Sara)

Having already considered the impact of diagnosis of Usher and the impact Usher has on familial relationships across the lifespan it is somewhat disconcerting that for Sara, Usher was considered to be solely a medical condition without considering wider impacts and thus referral to specialist services was not deemed necessary. Part of the role of a sensory social worker is to provide practical support to people with sensory needs/requirements to promote empowerment, equality of opportunity and access to services with provision of these contributing to a person's physical and emotional wellbeing. Sara's experience showed that she had awareness to "*seek help before sinking into a low state*"; however Len's lack of specialist sensory social work affected his mental wellbeing:

"I had a visit from a social worker all she brought was a letter saying you are now registered blind and she gave me a white stick and that's all she did…. I didn't know about [specialist charity supporting deafblind people], nothing. I didn't know anything like that existed. I had packed up work I was on my own".

Len continued:

"*I just feel that once you have been registered, been registered by the local authority blah blah blah, there should be a system where people I don't know get a visit every so often*".

Although Pam did get a follow up visit, it was not a positive experience:

"*I had contact with social services they didn't do much but a cane came through my letter box, my door and then another social worker came to see me from the visually impaired and said... oh she asked me what I could see and what I couldn't I thought I can only see what I have ever seen. I didn't understand about how much vision I had lost. She said it doesn't go black you know, well I had black all around my eyes but I was thinking I had hair in my eyes or whatever, that kind of thing. She said you are fine you don't need any kind of help. That's what she said to me*".

Pam expressed that she had a history of mental health issues and suicidal thoughts and that she thought "*oh I have been abandoned*". However, Pam did go on to contact social services again and insist that she received help and a specialist assessment of her sensory need was carried out:

"*Then I got back in contact with them I said I feel...I am not going to leave my house properly if I don't have some kind of …it was [specialist charity supporting deafblind people] actually it was [specialist charity supporting deafblind people] had actually back then asked me to go to [city name] for a full day assessment*".

However, where specialist social work support was provided, there appeared to be inconsistency depending on geographical location and the level of the social workers' sensory experience and knowledge. Carl experienced better support and services when he moved area:

"[Town name] was a good move for me as the support from the council was miles better".

Eve also relayed an experience of social work intervention:

"*She was hearing. Little bit understand deaf not really. [Family member name] find me new social worker [social worker name], other social workers not very good at all. It was [Family member name] found social worker at time. Communicator my social worker [social worker name] in [town name] they were the best perfect. Before that other social workers not good. [Town name current residence] not so good not good at all"*.

Eve shared her experiences of the specialist sensory support she received in different geographical locations. Eve became quite upset during interview when telling about the change in services and support she was currently receiving after having to move home to a different geographical location due to ongoing health issues. Eve commented on previous support received in the area in which she previously lived:

"*Yes in [town name previous residence] lots of support"*

However Eve then expressed that her support changed after she moved into a new location:

"The social worker does not visit. Before in *[town name previous residence]* they used to visit me every few weeks, [social worker name], [support worker name]. In *[town name current residence]* very different. Very different".

Eve then talked about the impact the change of social worker was having upon her:

"*Remember my move [social worker name] the social worker was seeing me every 2 weeks it's very different for me. [Town name current residence] it*

difficult to get around. Scary and they expect me to visit office its dangerous lots of cars easier to get around [town name participant lived in previously, prior to being moved due to health issues] quieter. I ask them visit my flat they say no our office".

Experiences such as these highlight the lack of awareness amongst professionals who work in the sensory field as safe orientation can be difficult for people who experience deafblindness/Usher (Sense 2014a, 4.2). Eve then shared her experience of the impact of lack of specialist social work support:

"*The social worker doesn't visit at all. Only Guide [communicator guide] Thursday, don't understand very confused. I have said I want social worker to visit me to talk to me they ignore me. It is 6 months since I have been asking for a home visit. It's far. I don't want [town name current residence]. My knees. Their office is far. I have told doctor, I have stress, I can't see. People throw rubbish, there are steps to go down and I have fallen"*.

Eve also explained how lack of communication skills contributed to an already difficult situation:

"*Here [town name previous residence] very good. Could sign. Deaf person could meet me. Come to [town name, previously lived] would explain. [Town name current residence] area is very different to [town name previously lived]. I want to move back to [town name of previous residence]. [Town name of current residence] they have emails. They say email any problems*".

Another participant who was aged 45 years at the time of interview, and had experienced symptoms since the age of five years expressed the limited level of his support:

"I've only been receiving support since October 2012 even though I have had the condition [Usher syndrome] all my life".

Whereas Len's experiences were some years ago, Eve and Oliver's were current and were occurring at the time of the interviews in August and November 2013 respectively. Adam who at the time of the interview was aged 49 years proffered reasons for lack of support:

> *"No support at that time with Usher unknown"*.

However, Adam felt optimistic that this would change as more became known about the condition:

> "*Know more now with technology at my time unknown not very clear*".

Monica felt strongly that anyone working with her, whether they were in the role of social work/support work or carer should have specialist deafblind knowledge, she expressed:

> "*Even my carer didn't know. You know and um so yes. I have left the care agency. Yes I did like the carer but I need a new carer now. She has left the agency too. I was her favourite client. I am looking for a new carer now. Support. Not specially trained for my eyesight and hearing just a generic kind of carer. Agency so just helps with whatever I need help with. But she doesn't have specialist training regarding hearing and eyesight and with my particular needs, so I have to let her go. They don't have training and I have to spend hours and hours training them when that time could be spent on the many many things I need help with and I have spent many hours training them from generic agency I am with. It's very exhausting and very tiring. I feel like an unpaid workshop leader; teacher, mentor. You don't get paid for it"*.

The findings highlighted the stark contrast for people living with Usher, between where specialist sensory support is provided and where it is not. However, where sensory services have been provided, Fred experienced these can be withdrawn:

> "*Change things, what services? Yes can't take the condition away but could make things better Firstly they should keep sensory teams absolutely. That is total stupid, experience we have had some wonderful people but with the changes they have gone now you have lost that valuable expertise. Shared things when it comes to eyes and ears the doctors can do it separate but when it comes to rehab you have to take the combination into consideration. It's a sensory impairment not one or the other".*

This comment highlights the essentiality of D/deafblind social workers/experts understanding the complex needs of people with Usher in order to provide appropriate and individual support. As has been mentioned previously it is not sight loss or hearing loss but a combination of both (sight loss + hearing loss) which adds to greater complexities being experienced by the person who has Usher syndrome.

Carl also shared his experiences of the impact of appropriate sensory support:

> "*The Sensory Support Unit (SSU) in the [name of council] Council is very good too and they came out on several occasions to give me mobility training with the long white cane, put tactile blobs and lines on the domestic equipment. I went back to college to do my Maths A level again and the SSU came out to give me more mobility training, alongside the [association for the blind] own community worker helping me too".*

Monica expressed how being supported to apply for a bus pass made a significant difference:

"*She helped me through social services to get the bus pass. That was a big deal actually"*.

A strong finding that arose within this theme was that sensory expertise is essential. Monica's experience regarding the importance of support being at the right time in the life of the person with Usher is profound:

"*I have been bumbling through life, just bumbling through life. So [children] need to be taught life skills, independence skills, how to adapt, the quicker they start the better. Stops a lot of problems way down the line. Asking for a lot of trouble and then society has to pick up the pieces"*.

Monica concluded the interview with the comment:

"*I think it's a ticking time bomb if people do not get support at the right time*"

There were varied experiences of support amongst other participants, for example:

"*Since I have been diagnosed I have found the varying support out there making an individual more independent. The support is varying on a personal and professional basis to assist with socialising, studying and working. There are also support groups to help people who have Usher*" (Oliver).

Jeff initially did not feel the need for social services input:

"*I would confess I didn't feel I needed social work input because I was a great believer you cope and you cope with the system and my problem was not so severe that I needed social services help".*

However at a later stage, Jeff found social services support was beneficial:

"*Through them social services came on board and provided me with a kit and a personal listener for me to have at work, because at that time I was*

attending management meetings and it was felt that maybe you are not picking up things you need".

This highlights that although a person may initially not feel that they want sensory support, provision of support, services and equipment can have a positive effect. Sensory equipment and guide dog provision will be presented next.

4.5.3 Sensory equipment and guide dog provision

The provision of sensory equipment and/or guide dog provision were a central feature within this theme. All participants had been provided with, and were using specialist sensory equipment in one form or another with five participants having guide dog provision (Ben, Pam, Ruth, Sara and Tia). The type of support depended on the individual's hearing and sight loss. First, experiences relating to equipment will be presented, followed by guide dog provision experiences.

4.5.3.1 Equipment

The term, sensory equipment relates to a variety of equipment specially designed to meet the needs of people who experience sensory need/requirement and is constantly changing as technology evolves. For some people if their sight was worse than their hearing, equipment such as a red and white or white cane was paramount for safe orientation and alerting others to their sight loss (Adam, Carl, Fred, Gareth, Harry, Iris, Jeff, Kate, Len Monica, Nora, Oliver and Quentin). For others with sight loss a guide dog was invaluable and for others who had experienced profound hearing loss from birth flashing doorbells/telephone/fire alarm or pager system were useful (Debra and Eve). Equipment provision can have a significant impact on the life of the person who experiences Usher syndrome, however, lack of provision of equipment can lead to isolation, lack of inclusivity and in some cases depression (Evans and Whittaker, 2010).

The type of equipment that a person uses to meet their needs is individual depending on their sight and hearing loss. The findings bore this out with Carl sharing:

> "*By this time, I was needing some help with my computer work with changing the colours from black on white to the reverse scheme of white on black background My brother made me a video inverter module plugged in between my computer and my display to do this without relying on the Windows environment itself and I also used a high spec graphic card to provide magnification (x2 and x4 only) without (again) the Windows knowing about it or providing the magnify function*" (Carl).

Equipment used by participants varied quite considerably depending on their technological abilities. Whilst Carl used quite individual and specialist equipment to meet his needs, Len expressed:

> "*Can't learn technology whether it's because I can't be bothered or my brain won't…..tune in. You know what I mean? I just can't get interested*".

The people that were interviewed all had varying degrees of sight and hearing loss, therefore their experiences regarding equipment required to meet their needs was individual:

> "*I have a symbol cane yes, a quarter of a metre. I don't go anywhere without it*" (Quentin).

Standard pieces of sensory equipment that will be provided to the person with the sensory need include flashing /extra loud doorbells and telephones as relayed by Jeff:

> "*I need to do something to make it a bit more easier in terms of a good telephone, alert me to doorbell that sort of thing*".

However as Jeff highlighted, although he realised he needed a doorbell, he had not considered equipment relating to safety:

> "*I didn't realise at time, but they said you need to be alerted if your fire alarm goes off. I thought actually yes*".

This experience highlights the need for specialist sensory support as without this provision people with Usher may not receive the individual sensory support they need. Sensory equipment is invaluable as it not only keeps people safe, but enables them to be inclusive in society and the community.

Other useful equipment includes mobile phones which due to technological advancement, have greatly changed over time. Iris relayed how she adapted to using her mobile phone:

> "*I only use the mobile on handset free but the problem is difficult to hear if I am outside in the noisy environment therefore I will give to someone who is with me. I also have the land line phone that has the t loop where I switch my hearing aids to the t set*".

Iris continued:

> *"I have a doro mobile phone which has font to choose to the size text that suite me, and I can adjust the volume to who I am talking"*.

Having a range of technological options can also be useful within the family for example, when a familial member cannot or will not sign but would rather use a mobile phone to text or talk:

> *"The other thing with technology is mobile phone I find myself do lot with mobile phone advancement in technology has improved I text but it try to talk*

to my mum because she won't sign. I need mobile phone I can hear so I used neck loop ones. So I can talk to my mum" (Adam).

Other equipment useful for people with sensory needs is speech to text and video conferencing, which provides a range of communication options. Kate expressed that although video conferencing did not work for her, speech to text did and enabled her to be an active participant within an educational setting delivering sensory awareness workshops to students without the delays that may be associated with translation:

"Same with [name of university] we do video conferencing with [name of town] and you know I can't cope with that so what I have is speech to text. There is a person who comes she is paid by [name of university] and she has two don't know if you are aware she has two laptops they are connected, I look at one of them which is adapted to how I like it black background and large yellow font. When people are talking she is typing away so that I can follow what is said. I get dialogue immediately I am able to communicate. We did it with presentation at [name of university] I could follow and was able to join in with the discussion. I said to the students now if it wasn't for this I would not be able to talk to you now".

However, whilst many participants experience benefits from using equipment, Kate found some of them a little difficult to engage with, despite their efforts:

"Got more sophisticated especially with blue tooth [27] and things like that. It's more complicated it's quite frustrating actually".

Conversely, Carl did not welcome the technological advancement in relation to hearing aids:

"Sorry but I guess it still bugs me! I am using old technology and I love these hearing aids but they don't make them anymore so one day I will have to be forced to the new ones! Eeeekkk!"

Whilst the participant's experiences as outlined above highlight the importance of specialist sensory equipment to support individual needs, Fred relayed less positive experiences with regard to use of sensory equipment as he found that people treated him differently within his workplace because he used a white cane, which identified him as being sight impaired/severely sight impaired:

"*Again that started to change things and it's amazing how people start to change when you start to be different in the work place. Yes they started to change in their attitude. They didn't want to talk to you. No you know you have the white cane now it's different. Its strange people can be very funny sometimes*".

Gareth expressed that using equipment such as a red and white cane which identified him as deafblind made him feel vulnerable:

"*And erm with my eyesight I am awfully sorry it is really severe so it is a white stick. Erm it's just light and shade. ….. I sometimes feel vulnerable in the street with a red and white stick*".

Quentin also expressed similar concerns regarding the use of, a white or a red or white cane:

"*It should be red and white but it's white. I don't want to draw too much attention to myself because when I am out, because when I meet someone face to face in a quiet environment I could pass as human you see so I am not as deaf as some people who have a red and white cane. So in other words if I am in a quiet space and I meet someone and I get lost they will*

converse, but if they see red and white cane, they might think my hearing is worse than it is. I should, I am often nagged that I should get a red and white cane. One day I will, but I don't want to present, I don't want to present my difficulties too much".

However, Fred expressed the following about using sensory equipment:

"*At certain times I do going to [specialist charitable charity] functions, lobbies, going to hospital on my own or for treatment it's a red and white stick. They offer me far higher in the queue to be seen quickly so yes I do use the red and white stick as the need arises*".

These experiences highlight that whilst sensory equipment can be beneficial for people experiencing sensory needs/requirement, it can also contribute to vulnerability through identifying the user's disability. However, as will be presented next, guide dog provision evoked a positive experience for all participants who received one.

4.5.3.2 Guide dogs

Out of twenty people interviewed, five (Ben, Pam, Ruth, Sara and Tia) had a guide dog. The findings highlighted the substantial impact guide dog provision had on the participants' lives with all the participants who had a guide dog sharing positive experiences, for example:

"*I used to come back stressed out and [swear word] off with a lot of people. Sometimes people did not respect the type of people [name of guide dog] has made a big difference. I feel a lot safer with [name of guide dog], a lot calmer*" (Ben).

Ben's comment highlighted that guide dog provision contributed to his psychological well-being, inner peace and feeling safer. Ben continued that he considered having the guide dog, had impacted his social life:

> "*I do love dogs and I do love animals he's always with me. He gets a lot of attention. The other day when I was looking through my phone and er my phone book is more girls more and more since I got the dog. They like [name of guide dog] really. One girl asked me if I would like to go for a walk one day. Not me, it's [name of guide dog]. I do feel great"*.

Ben continued:

> "*To be honest [name of guide dog] knows, dogs are very sensitive they can tell if you are happy, angry, if I get angry, [name of guide dog] jumping up and calms me down. He knows when I get angry sometimes upset. He notice, puts his head up*".

Pam expressed that having her guide dog meant although her levels of sight was deteriorating, every-day functioning was enhanced:

> "*I can ...go a bit further than I do and go to unfamiliar places. I can go to familiar places my guide dog knows where to take me"*.

Pam described that having the guide dog had affected her psychological well-being. This was especially poignant as, as noted earlier, Pam related that she experienced depression and suicidal thoughts:

> "*Wonderful. Wonderful for my psychological well-being as well. I don't feel um I feel more in contact with the human race. People come to speak to me"*.

However, Pam advised that she did not get a guide dog so that people would be more ready and willing to communicate but rather to contribute to her safety:

"*I got it because of all the accidents. Got my first guide for all the accidents I had and because I can't hear traffic in the same way. So it was about safe mobility, but also great great companionship erm absolutely amazing"*.

Similarly to Ben, Pam experienced feeling safer and the impact of having a guide dog was so great that she commented:

"*I can't imagine life without a guide dog*".

Ruth too expressed how her dog enabled her to socialise:

"*Yes. But I still go out. I go to the pub every week with my dog and my friend whose blind as well".*

When Sara was asked if she found her guide dog useful, she replied:

"*Oh indescribable how much he helps me, he just yeah…he's my guide, he's my mate , he's been a life saver in so many ways*".

However, Sara also relayed the responsibility that came with having a guide dog. Sara shared how she had experienced rude comments in the work environment when people saw the guide dog and realised she was severely sight impaired. One person said to Sara "*I think it's lovely they have given you this job*". Sara relayed the following:

"*I do a lot of tongue biting you know what I mean particularly with a guide dog, I am aware I am one of those public faces of guide dog for the blind. I work as well. I can't be going round telling people to F off and all that sort of thing, because that brings the business into disrepute and all that sort of thing"*.

Tia who also has a guide dog, related that not only did guide dogs provide the dog but they also provided other services and support to enable independence. Tia had found this useful because, as identified earlier in the chapter, she was going to university and would be living away from home:

> "*My support from Guide Dogs has been fantastic , not only have they provided me with my lovely dog but one of their mobility and rehabilitation officers trained me with a cane and taught me basic living skills in a non-sighted way eg. Cooking, cleaning, mobility at home".*

Guide dog provision clearly had a positive impact on the five participants who had one. However, where a guide dog is provided strict assessment has to take place prior to provision. Furthermore to train and provide a guide dog incurs significant expense, thus whilst some people with sight loss will benefit from this provision clearly not all people will be able to. However, an aspect of professional support which all people with Usher/deafblindness can take advantage of is specialist charity support, though, unless people with Usher are aware of services available, accessibility is not possible.

4.5.4 Specialist charity support

A specialist charity concentrates on a particular area of practice, activity or specific subject field. The specialist charities are highly skilled within their particular field as is the case of specialist charities for people who are deafblind (Sense, 2014a). They provide support and services, carry out research, campaign and raise awareness. Specialist deafblind charities such as Sense and Deafblind UK provide an invaluable service to people who experience deafblindness. Although, people have different perceptions of charitable support, in the main participants were highly appreciative of their support.

Several participants highlighted the essentiality of specialist charitable input (Adam, Jeff, Kate and Tia) as they provide specialist support and services which concentrate on Usher syndrome and other conditions/syndromes associated with deafblindness. Jeff recounts his initial experience of having contact with a specialist deafblind charity:

> "*I remember a lady from [name of charity] coming down and talking to me and erm erm that felt strange to be truthful. Having someone to talk to me*".

Carl commented:

> "*It is only recently in the last couple of years that perhaps I felt that I need to start concentrating on my joint sensory loss and with the problems of my hearing aids, I contacted [name of charity] to seek help from them and I have joined with them. I am involved with their technology section as well as the Usher area with [name of charity]*".

Carl's experience highlights the mutual benefits for both the specialist sensory charity and the person who experiences Usher. The specialist charities have resources but people who actually have Usher, often have experiences and expertise that can only be gained by living with the Usher. Kate also expressed that she had a longstanding involvement with specialist charities:

> "*Over the years up here I have become more involved with the conferences in [name of charity]*".

However, Sara had a different view of some specialist charities:

> "*I know who they are I know what they do I just for me think it's a little bit..this is gonna sound awful but…this is how I feel. I feel that I've got erm a dual sensory loss and am deafblind and all this sort of stuff but I am not the same*

level as the people they help and some of them…does this make sense what I am saying?"

Sara further expressed that she did not feel they were geared up for people like her:

"*They are not necessarily geared to helping people like me. I am a women in her 30s I have a job. My hobbies and my interests so to speak are going out to theatre and pubs and that sort of stuff and I don't necessarily want to go on a residential erm holiday and do outward bound activities, I don't want to make pom poms erm*"

Sara continued:

"*Their latest thing has been a big pom pom drive! Trying to get everyone to make a pom pom and I just don't…well I would never belittle what they do because I do think the charity as an charity they are obviously fantastic but I just don't think they are necessarily for me*".

Such views highlight that whilst some specialist charities will be of benefit and useful for some people, other people will choose not to access them. People have to be ready and willing to access such charities.

Conversely Ben did not access these charities upon diagnosis because he did not know of their existence:

"*Only found out about [name of charity] last year. Mum said she read it in magazine in [name of café] in [town name]. I went there last year to café got to know lady there. Told her Usher syndrome, she said get someone her name [name of worker] , then she came back and it started off from there Then [name of worker] come. Last year in [name of town]. Last year yes they were very nice people understanding*".

Ben had been diagnosed with Usher syndrome 11 years previously and had not been advised that such specialist charities existed. It was purely by chance that his mother read about such specialist charities in a magazine. During this time period Ben expressed during the interview that he had experienced very low moods which led to depression and bullying. It could be argued that the support and services of a specialist charity may have proved to be invaluable to this participant if he had been made aware of them as is highlighted in Ben's next comment:

"*They were very helpful. They did help me out*".

Len also had a similar experience, only discovering support and services that could be provided by specialist charities, as a result of a chance meeting with a specialist charity worker on a train. Len related *"so that's how my life changed".* Again it could be argued that Len's experience may have been very different had he known of the specialist charity's existence. Pam commented on the benefits of being involved in a specialist deafblind charity:

"*Because I needed to keep adjusting to the loss I was experiencing I just needed "to keep busy all the time and that Is what I have done*".

Thus for Pam being involved with a deafblind charity helped her to adjust to living with Usher. Adam also highlighted the benefits specialist charities can provide for young people:

"*We do have the young Usher group so it's kinda like a role model peer group, with their parent and suffering from Usher trying to guide and support with that becoming more supportive to understand not alone*".

So to summarise, participants revealed the importance of raising awareness of specialist organisations. It was, not only to alert the person living with Usher with regards to support and services they could access, but also provide them with the

opportunity to use their experiences to help others should they wish to take a more active role within these organisations. The final subtheme to be presented is raising Usher awareness.

4.5.5 Raising Usher awareness

The findings showed that raising awareness was essential as some participants had never heard of Usher, Quentin commented:

> "*Until I was diagnosed I had never heard of it (Usher syndrome). I had never met anyone with Usher syndrome before in fact even when at the time I was diagnosed. I did not meet anyone with Usher syndrome until I joined [specialist charity]*".

Pam shared Quentin's experience:

> "*No I never met anyone with Usher till I was in my 40s. I hadn't ever heard of it*".

There were several areas in which participants wanted awareness raised: the general public including bus drivers and crematoriums; in the training of social work students and social services; medical staff including general practitioners (GP) and sensory professionals; hearing/sighted children in schools; and the government. However, whilst the participants highlighted that change at governmental level was needed, they demonstrated that as they were living with Usher, they were experts by experience, so worked hard to raise Usher awareness (Adam, Carl, Fred, Gareth, Harry, Iris, Jeff, Kate, Oliver, Pam, Quentin, Ruth and Sara). The first area to be presented is the general public.

4.5.5.1 General public

Ben expressed "t*ry to advertise so people know*". This was poignant because as highlighted earlier Ben lived with Usher, without realising support was available for many years. Nora too shared:

> "*Be good to make people have better understanding of Ushers*".

Iris too had commented that lack of Usher awareness, had affected colleague understanding and as a result contributed to her early retirement. Debra and Pam highlighted the need to raise awareness so people experiencing Usher have an awareness of other people with the same condition. Pam commented that she "*didn't know there were other people out there*" and Debra similarly expressed that more awareness was needed as "*few Usher people, we all live such long way away*". Iris relayed her experience of lack of awareness when using public transport:

> "*I have experience that the bus drivers are not always aware how important for deafblind people to get off the stop they need to get off I had a driver refuse to stop at my stop, a long story. This had now put me off travelling on buses on my own which is now two years ago*".

Iris's experience could have far wider implications than just travel, as this could result in isolation and possibly even affecting a person's mental, physical and/or emotional well-being, thus highlighting the essentiality of awareness being raised to enable people with Usher to have safer orientation.

Another area highlighted by Quentin, was raising awareness for induction loop installation in crematoriums, giving an example of his success:

> "*For example a case I have just one a crematorium at [name of place] and I have been to many funerals there, also my parents and my son. Every time*

they say there is an induction loop when there weren't. I went to a funeral there recently and I wanted to hear the eulogy. It proved they were lying and I thought you are not going to get away with it this time. So to cut a long story short I have threatened to go to court and then they settled and now they have got 3 crematoriums [names of crematorium] now all have induction loops and that is a tremendous success".

Quentin's success highlights the power of active participation of people living with Usher.

4.5.5.2 Training of social work students and social services

The next area to be presented is raising Usher awareness to students on a social work degree program, Kate shared:

"*In fact last Monday I spoke to the MA students about my experiences and about the person visiting me. And they need to know what they are about when they go visiting their clients. To find out what sort of support and help they did need*".

Students receiving sensory awareness training has the potential to improve their practice and subsequently contribute toward good practice. Raising awareness and increasing understanding early in students' training will enable them to utilise their skills and knowledge while on their social work placements. On placement the student completes a portfolio as evidence of learning and is supervised by a practice educator who is a qualified social worker. Kate highlighted that it is not only service user teaching on degree programs that benefits students, but also their contribution to recruitment and admissions of future social work students and also supporting students to prepare and be ready for their work experience placement:

"*I am giving another talk Wednesday with BA students we are involved with admissions and another before they go out and do direct practice assessment*".

Quentin also relayed how he raises awareness by being involved with postgraduates:

"*I do some work with post graduates what she does with me which is quite interesting she goes walking with me, because her PhD is how visual impaired people perceive the countryside*".

Kate explained that after being home visited by social services to carry out an assessment of her sensory need, the person that social services had sent was a student who had no sensory experience. After complaining Kate was asked to be more involved to enable deeper understanding of Usher:

"*I found myself being asked to be involved with this group it took a number of years. So I am now involved with [name of university] service user and carers involvement*".

Training students is essential as lack of awareness for qualified social workers can have serious consequences as described by Debra, when her ability to care for her baby was questioned.

4.5.5.3 Medical staff and sensory professionals

The findings highlighted that staff working in hospitals, GPs, ophthalmology, audiology and opticians all need Usher awareness. Ben expressed that it would be a, "*good idea if doctor and optician offer support at the time*". Fred also expressed the impact of lack of awareness from his GP:

"*I had a situation trying to prove I needed DLA. Doctor said I didn't need it. I went to tribunal and stuff they said why are you there I said this this and this. They said why do you think you should be here? I said thank you very much, am I going to get it? When you get these damn bureaucrat's and people who don't even listen they do it now. People who don't understand anything*".

Fred's experience highlighted that as a result of lack of GP support he was required to attend a tribunal. As previously highlighted in chapter 1, Usher is a serious debilitating condition and people having to fight for recognition and understanding intensifies their difficulties, this was highlighted by Gareth's experiences of attending an appointment at the job centre.

Pam also experienced lack of awareness:

"*I also remember when I was at school that I must have complained about having blurred vision because erm my mum took me to an optician and they said there is really nothing you know glasses aren't going to help but well give her a very weak prescription so so (laughs) so that was another time it was never picked up. It wasn't unusual it's one of those conditions that unless somebody knows they wouldn't be able to diagnose it*".

Pam's experience highlights the value of Usher awareness, as without it diagnosis is not possible. Oliver's and Ruth's experiences highlight that lack of awareness can lead to misconceptions about Usher such as thinking Usher was linked to '*lack of eating carrots*' and connected to 'VD'. Although Pam, expressed that awareness should be raised, she was active in ensuring this occurred:

"*There were 5 sensory teams one of them had social workers for the deaf ….one of them picked up I had Usher syndrome ….wanted me to be a*

spokes- person end I found that very hard to be thrown in at the deep-end and I didn't know if it was something I would be any good at" .

Pam continued:

"*I remember my first going to [name of hospital] in terms of signage and what would help consultants and things like that*".

So although Pam initially found Usher awareness raising daunting, she did it to help others with Usher, again showing importance of service user input.

4.5.5.4 Schools

Another area in which Iris expressed that awareness should be raised was in schools, not only to raise awareness amongst the teaching staff, as highlighted earlier, but also to include hearing/sighted children in school:

"*To have the normal children to have lessons about eyes problems and hearing problems. To include communication skilled such as hand on sign and learn the deafblind manual. This way they can communicate with Usher people and not feel embarrass to communicate with them*".

Iris explained why this was important:

"*When the new generation socialised with deafblind people this will enable for them to work together in the working environment in the future. I wouldn't want the young people not to have the opportunity to go to work and they feel part of the community. Yes I had opportunity to go to work when I had hearing problem. That was difficult but when I had both disabilities that were tougher to cope*".

Iris asserted that raising awareness with school children would have a positive effect for people with Usher and facilitate people with and without Usher working together.

4.5.5.5 The government

Gareth highlighted that an additional area in which Usher awareness raising is essential is within government:

> "*I think the government need to be aware that they can't continue to taint all disabled people with the same brush*".

Gareth continued:

> "*If you have Usher syndrome and there is no treatment or cure that is it. People with long term conditions should not have to go up against the indignity and stress of having to apply every year or two years to be assessed by the government to go back to work, when I have already been forced out of work to be reassessed yet again for personal independent fund and disability living allowance. When it is pretty obvious my vision is not getting any better and it is rubber stamped I have this condition. Valuable resources should not be wasted in making our stress levels really high*".

Although Gareth was aware of the benefits of raising Usher awareness to government, he described challenges experienced:

> "*Yes well years ago I went to [government office] Hall this person said I can understand what it must be like to lose hearing or a sight loss but to have both. It's very difficult you need all the help you can get. All hot air and that was one of the MP's. It's meaningless. They can't understand at grass roots level and when it comes to policies we are all tainted with the same brush. That's not fair just because point 1000001[indicating a very small*

percentage] abuse the benefit system that is a crazy waste of resources. Crazy".

Gareth's experiences highlight the need to raise governmental awareness, so that legislation can be implemented to support people with Usher and this will be discussed further in chapter 5.

The findings show the impact Usher awareness raising achieves. Also, participants sharing their experiences and contributing to this study enabled me to raise awareness by disseminating findings at conferences using PowerPoint/poster presentations and also by writing for publication (Appendix A).

4.5.6 Section summary

This section presented the participants' experiences of professional support and considered sensory social work and sensory support, equipment and guide dog provision, specialist charitable support and raising Usher awareness. One participant expressed the importance of getting support at the right time which also needs to be appropriate and tailored to need because having Usher has an individual profound effect. Although, as stressed, not all people with Usher will require the same support and services, participants' experiences highlighted that Usher awareness needs to be raised so that not only the general public, but medical and social professionals are better able to understand people with Usher and meet their needs. Although, people living with Usher do not want the condition, they learned to live with it, and most of them are dedicated to raising Usher awareness. Two key messages emerged from the theme 'experiences of professional support': Specialist support is necessary regardless of demographics and, there needs to be increased awareness of Usher amongst professionals and the general public.

4.6 Chapter summary

In summary, this chapter explored four themes (Table 4.1) and presented the findings of these themes. Findings showed the effects Usher has on a person living with the condition and the significant changes that they need to make in their lives to adjust and cope with these changes. Also highlighted was the fact it is not only the person with Usher that the condition affects but also the impact on families and friendship groups. Key messages from the four themes were generated, and are summarised in Table 4.4.

Table 4.4: Key messages from findings

Diagnosis is the start of the experience	**Familial relationships across the lifespan**	**A sense of belonging**	**Experiences of professional support**
Usher although not life threatening is life altering	A diagnosis of Usher, impacts on new relationships and the management of existing ones	Acquired disability affects communication	Specialist Usher support is necessary regardless of demographics
The diagnosis of Usher impacts on individuals in varied ways	Usher being an unseen hereditary/genetic condition has consequences for the person with Usher and their families	Deaf/Deafblind culture can contribute to a sense of belonging	There needs to be Increased awareness of Usher amongst professionals and the general public
The way the diagnosis of Usher is communicated affects individuals' experience		Technology can support a sense of belonging	

Next, chapter 5 will discuss these key messages from participant experiences, critically evaluating them with literature and theory to reveal three overarching messages.

Chapter 5 Discussion

5.1 Introduction

In chapter 4 participants' experiences were presented and key messages relating to their experiences of living with Usher were outlined (Table 4.4). For example participants were able to share 'the importance of telling a child they have a diagnosis of Usher' and the implications of Usher being hereditary/genetic. The literature review (chapter 2, Table 2.7) highlighted, that there are few studies that have explored the impact of diagnosis of Usher syndrome on individuals, with only two exceptions identified (Ellis and Hodges, 2013, Kyle and Barnett, 2012). This is the first English study to explore in depth the lived experiences of people with Usher of all types (I,II,III) and a range of ages (18-82 years), using a phenomenological methodology as well as giving participants the opportunity to choose their preferred method of communication and interview method (MSCIM, see 3.4.3).

In this chapter, the discussion moves away from individuals and discusses the study's findings in the context of previous research about living with Usher and sensory loss, and with critical review of other literature relating to life altering, genetic but not terminal conditions. To begin each of the four themes (Table 4.1) will be discussed individually, outlining key messages from findings and critically discussing them in the context of existing published literature and theory. After discussing all four themes individually, then three overarching messages from the 'key messages' will be identified (Table 5.2). The discussion begins with the first theme from the findings, 'diagnosis is the start of the experience'.

5.2 Discussion: Theme 1 – 'Diagnosis is the start of the experience'

Three key messages arose from the first theme 'diagnosis is the start of the experience', Usher although not life threatening is life altering; the diagnosis of Usher impacts on individuals in varied ways, and the way the diagnosis of Usher is communicated affects an individual's experience. The first key message, Usher although not life threatening is life altering, will next be discussed.

5.2.1 Usher although not life threatening is life altering

The complexity of losing both sight and hearing is described as not loss of sight *and* hearing but sight *times* hearing (Sense, 2014a). If a person has their sight and loses their hearing, they have one sense to rely on and vice versa if a person can hear, but where a person loses both senses this has serious consequences for the person experiencing the loss. Therefore although Usher/D/deafblindness is not life threatening, it is life altering as is highlighted in the literature (Ellis and Hodges, 2013, Kyle and Barnett, 2012, Högner, 2015, Wahlqvist *et al,* 2013, Kim *et al,* 2014, Prynn and Cecil, 2015, Cecil, 2015) and in my study.

Ellis and Hodge's (2013) qualitative case study with participants aged 14-56 years highlighted that a diagnosis of Usher will have life altering effects, for example: they identified that for some participants it would necessitate the use of sensory equipment such as a white [14], or red and white cane 31[13], as well as being an end to driving. As participants in Ellis and Hodges's study had not used sensory equipment previously, and had been able to drive if they chose to, living with Usher altered their lives. These findings were also found in my study as participants lives were altered when they needed to access equipment and/or guide dog provision and choices were reduced in terms of driving.

Kyle and Barnett's (2012) mixed method two phase study referred to the impact of diagnosis for people with Usher and primarily explored the Deafblind experience of 21 people who were Deafblind in comparison with that of 38 people who were Deaf ranging between ages 21-65 years, to test a framework for interaction and support by Deaf people for Deafblind people. The study aims were to examine the experience of being Deafblind, exploring if this was similar to Deaf people's experience and discovering if Deaf and Deafblind people could work together. Kyle and Barnett also highlighted the life altering effects of Deafblindness in terms of employment choices, access to the environment and the option to drive being take away from the person as their sight worsened. Kyle and Barnett highlighted that Deafblindness increased reliance on other people, for example: the need for a communicator guide[44] to access the environment and related frustrations a Deafblind person may experience, if the guide or the timetable is changed. As these incidents would not have been experienced if the person was still sighted, again, they highlight the life altering effects D/deafblindness imposes. Difficulties accessing the environment were also expressed by participants in my study.

Although there were similarities between my study findings and Kyle and Barnett's (2012), in terms of changing sensory needs altering participants' lives with regard to future employment, accessing the environment and no longer being able to drive, their focus was on Deafblindness, not life altering effects of Usher, as with Ellis and Hodges's (2013) study, and my study. The key differences between people who have Usher and people, who are D/deafblind, are that Usher is congenital/genetic/hereditary, whereas D/deafblind people may not have been born with the condition (Sense, 2014a). As D/deafblindness affects people in different ways, the life altering effects for people experiencing Usher and people who experience D/deafblindness with be unique to them as individuals.

Ellis and Hodges's (2013), Kyle and Barnett's (2012), and my findings, that Usher syndrome/D/deafblindness imposes the need for life altering sensory equipment, increased reliance on others and that life choices in terms of driving are affected, links in with the discussion in chapter 1 about the theoretical concept of identity development (Erikson, 1982) because although not life threatening, Usher syndrome is life altering in terms of the life the person may have expected to live, and the one they ultimately experience. Erikson (1982) theorised that ideal development necessitated resolution at different stages of the life span, but when sight and hearing deteriorate and life choices are impacted, a person's opportunity for growth and development are affected. For example: development is hindered when a person's opportunity to drive is affected, which in turn may affect their employment prospects and also, where a person needs specialist sensory equipment or support (communicator guide) to cope with the practicalities of day to day living.

Despite Usher not affecting life expectancy, it is progressive, cannot be treated and can affect a person's safety which means the effects are disabling. These aspects of being diagnosed with a non-life threatening but life altering condition were confirmed by the participant's in my study and also previously discussed by Högner (2015), Wahlqvist *et al* (2013) Kim *et al* (2014) Prynn and Cecil (2015) and Cecil (2015).

Högner's (2015) quantitative, questionnaire based study found that living with Usher contributed to difficulties with safe orientation and mobility, which in turn would lead to increased isolation and reduced independence, resulting in limited autonomy. Similarly, in my study participants experienced Usher related restrictions, such as not being able to walk to places alone and drive, which was life altering, limited their life choices and was different to the life they may have planned or expected.

Wahlqvist *et al*'s (2013) quantitative questionnaire study highlighted that living with Usher was life altering as it affected the participant's physical and psychological health. Participants in their study reported they experienced headaches, fatigue, depression, suicidal thoughts and suicide attempts. The authors further reported that people with Usher need support to cope with living with Usher, in terms of access to information and interdisciplinary teamwork because although Usher is not life threatening it is a complex condition which makes everyday tasks a challenge. Whilst in my research study participants did not report headaches or fatigue, they did express experiences of depression and suicidal thoughts.

Living with Usher also raises life altering safety issues, for example: road safety. Kim *et al* (2014) quantitative observational study explored issues around safety and risk discussing the crossing decisions of pedestrians who are visually impaired and the effect quieter vehicles will have on their safety. In Kim *et al*'s study participants' decisions were compared with the sighted experimenter's decisions to determine the level of their risk. In my study there were clearly concerns for people with sensory needs about safe orientation as participants highlighted their reticence to travelling alone due to vulnerability and the dangers of quieter vehicles. Similarly in Ellis and Hodges's (2013) study electric vehicles were also referred to as being a problem because they were quiet and affected a person with Usher's ability to access the outdoor environment.

In a commentary, Cecil (2015) identified that cities such as London could have up to 400 electric cabs by next year, while Prynn and Cecil (2015), in an editorial called the advancement of car technology the 'London electric car revolution', and outlined that there are plans to invest £100 million in London to turn it into the "green driving capital of the world" (Prynn and Cecil, 2015 p1). As highlighted by Sense (2014a), safe orientation in general is already a significant challenge for people with sensory

needs, thus the introduction of quieter vehicles poses even greater concerns. Crossing the roads and safe orientation are just one challenge for people with Usher; my findings also highlighted that people with Usher have concerns regarding living alone as they would not hear someone breaking into their home which was not raised in any other studies.

The literature supports participants' views that a diagnosis of Usher alters a person's life in the long-term (Kim *et al,* 2014, Cecil, 2015, Prynn and Cecil, 2015). Safety and risk for people living with Usher inside and outside the home is clearly an area that requires deeper understanding to be able to meet needs and address safety, gain life experiences and ascertain where risk occurs. However, participant experiences also identified that people with Usher are courageous in the face of adversity and although they realise their life will alter, and is likely to get harder, they are realistic and make the best of their future.

As the life altering experience will differ for each individual depending on which stage they are at in their lives, this section included variations in method and age ranges of studies. The only qualitative study was Ellis and Hodges's (2013) and included participant's aged between 14-56 years. Although, other studies included a wide age range of participants (Wahlqvist et al, 2013, Högner, 2015) these were mixed method (Kyle and Barnett, 2012) or quantitative (Wahlqvist et al, 2013, Högner, 2015). The age range within my qualitative study was wider (18-82 years) and therefore the life altering effects of Usher and lifespan experiences from different perspectives at different points in time were able to be discovered.
Other life altering issues such as relationships and employment will be discussed later in the chapter. The second key message that of, 'diagnosis of Usher impacts on individuals in various ways' will be discussed next.

5.2.2 The diagnosis of Usher impacts on individuals in varied ways

The findings highlighted that the diagnosis of Usher impacted on individuals in varied ways. Whilst some participants became cautious, others became more adventurous. Although, some participants experienced depression, suicidal thoughts and feelings of bereavement, for others the diagnosis was a relief, as they now had an explanation for earlier life experiences, and others no longer felt alone as they knew other people with the same condition.

Ellis and Hodges (2013) concurred that the experience of being diagnosed affected people in a variety of ways, which was similar to my study, for some participants it was the beginning of depression and struggle, while for others it was the incentive to do more. Ellis and Hodges (2013) reported that some participants upon diagnosis felt shock and others felt relief that it was nothing more serious, similar reactions were identified in my study.

As loss and bereavement are associated with a diagnosis of Usher, Elisabeth Kübler-Ross's model of grieving is useful to understand it. Kübler Ross (1970) studied people's responses after being diagnosed with a terminal diagnosis (Kübler-Ross 1970) and found that they experienced five stages of grief which included: denial (why me?);anger; bargaining (negotiation e.g. extended life for better lifestyle); depression and acceptance. Kübler Ross's work was developed from James Robertson and John Bowlby's (1952) studies of children separated from their mothers and were later applied by Bowlby and Parkes (Bowlby and Parkes, 1970) to the responses of adults experiencing bereavements (Parkes 2013). Kübler Ross's work raised awareness amongst professionals about people's experiences of terminal illness and thus aided their understanding and ability to support a person who was dying (Hart *et al,* 1998).

However, Kübler-Ross's work has been criticised because the concept of acceptance was based upon weak methodological foundations and strong social expectations and her work reflected social values and expectation of 1969 and, as such, does not reflect cross cultural differences (Hart *et al,* 1998). Another critique is that the model is inconsistent in diagnosis of stages as not all people will display acceptance at the end, some people will still be angry (Hart *et al,* 1998). Hart *et al* (1998) concludes that although models of bereavement such as Kübler Ross's may be useful to understand bereavement, people's experiences and lives cannot be shaped into a particular model, as each person is unique and individual and will deal with bereavement differently. My study identified that similarly, receiving a diagnosis of Usher had varied experiences and responses, ranging from shock to acceptance. Allred and Hancock (2012) explored Kübler-Ross's model in relation to parental response to disability and concluded that a stage model such as Kübler-Ross's did not truly reflect the stages that a parent experiences as its meanings were outdated and the real impact on families could not be determined. Use of Kübler-Ross's model focused on grave sorrow and experience has shown that this is not the case with all parents who have a child with a disability (Allred and Hancock, 2012). Again although the model can be used to support people and guide professionals who are dealing with people who experience loss, it needs to be used as a guide only.

Another aspect to consider is that loss can be viewed from two different perspectives, that of finite loss and infinite loss. Finite loss indicates the loss that ends permanently, for example, death, whereas infinite loss is the loss that never ends, for example, disability and sensory loss (Currer, 2007, Bruce and Shultz, 2001, Evans and Whittaker, 2010), as in a diagnosis of Usher.

Sheppard and Badger's (2010) qualitative hermeneutic phenomenological study of depression in culturally Deaf adults posited that the reason depression occurred was

because people may not receive the mental health care they need. Although these studies do not focus specifically on Usher, but rather culturally Deaf adults, it could be suggested that the study findings are relevant to Usher too, as if people are not screened for depression then they may not receive the appropriate support. However, there are differences between being Deaf and experiencing Usher syndrome, because Usher is progressive therefore people are not only experiencing sensory loss, but they also have to come to terms with the diagnosis of a progressive condition. Within my study, depression was experienced by several participants following their diagnosis thus showing that prevalence and techniques used for early screening for depression in adults diagnosed with Usher could be valuable, as early diagnosis could prevent long term effects.

Although, Usher is a debilitating disease for which there is no treatment, some participants in my study, took comfort in attaining a diagnosis, as they could seek support. Although there is literature relating to how people lived their lives prior to a diagnosis of terminal illness, for example cancer (Montazeri *et al*, 1998, Holick, *et al,* 2008, Geyer, 1991), apart from the report by Ellis and Hodges (2013) there appears to be no other literature relating to life before Usher syndrome. There were some similarities of life before Usher between findings in Ellis and Hodges's study and my study, for example, that 'ignorance is bliss' (Ellis and Hodges, 2013) as the person did not have to face the realities of Usher. In both studies, some participants expressed that they did things they may not have done otherwise if they knew they were diagnosed with deafblindness.

However, whilst the older participants in Ellis and Hodges's (2013) study could remember being told they had Usher, "the majority of the young people" (p53) could not remember. In contrast, in my study, all the participants could remember when they were told, regardless of age, and many of them could recall vivid details many

years later, highlighting the impact of this event. This difference may have been due to the phenomenological methodology used which led to participants' in-depth accounts of the diagnosis experience and as such led to new findings about the experience of diagnosis.

In this section Kübler-Ross's (1970) theory of bereavement was discussed with reference to diagnosis of a life altering condition such as Usher. My findings showed that the diagnosis of Usher impacts on individuals in various ways because no two people diagnosed are the same. The final key message within this theme is the way the diagnosis of Usher is communicated affects an individual's experience.

5.2.3 The way the diagnosis of Usher is communicated affects individual's experience

The final key message relating to diagnosis being the start of the experience is that the way the diagnosis of Usher is communicated affects an individual's experience. Participants in my study discussed how they received news about their diagnosis in a variety of ways: some had the news imparted in an abrupt manner; others received news in a matter of fact approach from the professional, others received news of their diagnosis via an interpreter and others, received the news from their parents but not immediately after diagnosis, leading to misunderstandings.

The first aspect to be discussed is where health professionals have a matter of fact approach to breaking bad news. Bad news is defined as "any information which adversely and seriously affects an individual's view of his or her future" (Baile *et al,* 2000 p 302). In my study, one participant shared how the professional matter of factly advised her she would "*go blind at some point in the near future" (*see 4.2.2). Similarly, although Ellis and Hodges (2013) did not ask participants aged 14-20 years for their feelings and/or opinions on diagnosis, they reported that their older

participants had experiences of being told they had Usher in a harsh, matter of fact manner. One of Ellis and Hodges's participants explained how at the time she was a nurse and when she asked if she would still be able to do her job, was told she would go blind in the future. The participant describes that being told in this harsh, matter of fact manner left her shocked, and she was not given further information about Usher or charitable organisations she could contact for support.

Although Prentice *et al* (2014) suggest that qualified doctors are trained to deal with delivering bad news as they "encounter medical illness on a daily basis" (p272), it could be that some professional desensitisation takes place if people are experiencing being told bad news in a matter of fact manner. However, the participant in my study who experienced being told that she would go blind in the near future was diagnosed nearly 10 years ago, so it maybe that people diagnosed more recently may receive a more sensitive approach to delivery of bad news as professionals become aware of the impact of delivery on the individual (Prentice *et al,* 2014, Baile *et al,* 2000). Although there is literature relating to breaking bad news for people with life threatening conditions such as cancer (Baile *et al,* 2000, Gilbey, 2009, Barth and Lannen, 2011) Usher is a life altering condition, not a life threatening one, and therefore the impact for individuals is different. However, the principles and communication training in medical curricula could also be applicable to giving bad news about life altering conditions, such as Usher.

Baile *et al* (2000) outlined six steps for health professionals when breaking bad news and uses the acronym SPIKES as an aid memoir to remember stages. S = setting; P = perception; I = invitation; K = knowledge; Emotions and S = summarise or strategy. See Table 5.1 for fuller explanation.

Table 5.1: Six steps to breaking bad news (SPIKES)
Adapted from Baile *et al* (2000)

1.	**S**etting up the interview: The health professional mentally prepares themselves for breaking the bad news reminding themselves that although the bad news can be upsetting to the person, imparting of news can enable the person to be prepared for the future
2.	Assessing patient's **p**erception: What do they know about their condition or medical situation?
3.	Acquiring patient's **i**nvitation to impart news: Communicate with the person considering how they would like to know about their condition, does the person want to know all the facts?
4.	Give **k**nowledge and information to the patient: Letting the person know in advance that they are about to receive bad news can lessen the shock and prepare the person for what they are about to hear
5.	Consider the patient's **e**motions: Respond empathically
6.	**S**ummarise and discuss a **s**trategy: People who have a plan for the future are less likely to experience anxiety

Baile *et al* (2000) suggest that the six steps are meant to be sequential, however, people are individual and will respond to the receiving of bad news in different ways and therefore a linear approach to breaking bad news may not always be appropriate. Baile *et al* (2000) also acknowledge that not all steps will be used with every individual. However, there is evidence to show that the six steps work in practice. Kaplan's (2010) study reported observations based on breaking news to a 68 year old woman whose cancer had returned. Kaplan observed that although breaking bad news is a difficult communicative undertaking, following the six step process (SPIKES) can work in practice resulting in easing anguish for both the person receiving the bad news and the health care professional imparting it. Another factor to consider is that breaking bad news is a challenge and distressing for the health professional imparting the news, therefore a matter of fact manner is adopted to prevent overly emotional feelings being displayed. However, no literature could be found to support or refute this possibility.

The second area with regard to the way the diagnosis of Usher is communicated related to receiving the news through an interpreter. Findings in my study highlighted that where news is given using interpreters, it is essential to ensure the person really understands what their diagnosis is and what the prognosis will be. Although Ellis and Hodges (2013) referred to an experience of a participant who had an interpreter with her when she was told she had Usher, and another who was told via a BSL interpreter she had RP (retinitis pigmentosa) but not Usher, there were no other examples in the literature that considered what it is like for a person with Usher to be told they have the condition via a third party.

My findings identified that being given a diagnosis of Usher via an interpreter was an emotional experience. Although receiving bad news is always devastating, receiving that news via a third party as opposed to being informed directly in the person's first language adds greater complexity. When the news is conveyed via a third party the news may be conveyed in a way that the person does not understand, for example where a BSL interpreter is imparting the news. The interpreter may need to interpret words that they do not use in day to day signing and need to pause for clarification possibly finger spelling [8] words to achieve comprehensive understanding, which would interrupt the news being conveyed. Whilst some people who use interpreters may indicate that they do not understand what is being said, others will not, therefore they may not grasp the implications of the diagnosis. A participant in this study, for example, shared that although she had been diagnosed with Usher 27 years previously, she still did not understand it. Although this situation can arise within a sensory setting it could also arise wherever an interpreter is used. Although, there was just one participant in this study who experienced difficulties when being told via a third party, this highlights a need to ensure that health professionals imparting devastating news have awareness and sensitivity to ensure people are informed and understand the implications of the condition.

Although translators breaking bad news to people with Usher or other sensory needs/requirements such as BSL are not considered in the literature, an area that is covered in the literature is that of the experiences of translators breaking bad news to people who are diagnosed with cancer (Prentice *et al*, 2014) and again lessons learned can be applied to Usher. The reason Prentice *et al* (2014) conducted this qualitative research study was because they realised that although health professionals were trained to deliver bad news to people, translators were not. They found that as translators were the first to deliver the news to the patient, they were also the first to receive the patient response, which resulted in the translator feeling bad about imparting news that caused the patient pain. Prentice *et al* (2014) suggested that translators could be better supported by clinical supervision; formal debriefing; reflective practice and, if required, providing formal support. Although this research was conducted with translators for patients whose first language was not English, this research identified that support for BSL sign language, hands on BSL and deafblind manual interpreters would be beneficial as if interpreters are supported to better deliver bad news, the patients will also benefit.

Although there is no specific research relating to the impact on BSL translators giving bad news, I reflected on my own lived experience many years ago when I was engaged as BSL communicator and I communicated with an older woman attending a hospital appointment that she had been diagnosed with cancer. As I was the person who delivered the bad news and the woman was looking at me as I explained the situation in BSL, I remember reminding myself that interpreting was part of my job and I was required to be professional, although I also felt extremely saddened. Although the woman dealt with it calmly, telling me, that I had relayed the news kindly and that she had had a good life, I still remember breaking that news all these years later.

Whiting's (2014c) study which explored what it meant to be the parent of a child with a disability or complex health needs found that another aspect of the way that people received bad news related to the person's personality, life experiences and family support. He found that some people were more resilient than others, and coped with disastrous news, whereas others would not be able to cope with a less severe diagnosis. These findings were similar to those in my study because, where participants had in the main had good life experiences, and received positive family support they were better able to cope with being given bad news even if it was communicated in a negative way.

Although as has been discussed there is literature which discusses breaking bad news, there is no literature specific to Usher. However, the principles contained in the literature could be used to support people who are diagnosed with Usher. The final area to be considered is when a child receives the news they have Usher, but not immediately after diagnosis.

Although, in these findings some participants felt passionately that a child should be told immediately of their diagnosis of Usher otherwise they would fear the worst, others reported that not knowing they had Usher as a young person had enabled them to have experiences they may otherwise have missed out on. Rowland and Metcalf (2013) conducted a systematic review to explore the challenges parents face when communicating genetic risks which included: cystic fibrosis (CF); Duchenne Muscular Dystrophy (DMD); Familial Adenomatous Polyposis (FAP); Hereditary NonPolyposis Colorectal Cancer (HNPCC); Huntington's Disease (HD); Neurofibromatosis (NF) and Sickle Cell Anaemia (Hbo) to their children. Although the authors do not state if the parents knew of the genetic condition prior to diagnosis, they did find that disclosure of genetic risk in families was complex and difficult to broach and often resulted in a delay in disclosure of information and

greater familial pressures due to “misunderstanding, blame and secrecy” (Rowland and Metcalf, p870). Petersen (2006) suggested that when a person learns their condition is genetic/inherited and as such other familial members may be affected, it is difficult for them to disclose the information to other family members. However, in my study often the parent(s) did not know they were carriers until the child was diagnosed, which added to the complexity.

Rowland and Metcalfe (2013) from their literature review looked at the effect when the child is told, highlighting that if a child is told early, the child can be better prepared for the future in relation to “care planning and reproductive decision-making” (p870). The issue of when to tell the child arose in the my study findings as where parent(s) had told a child they had Usher, the child had been better able to cope with rapid sight loss and better prepared them for future Usher eventualities. Although in Ellis and Hodges’s (2013) study, one participant asserted that children are more adaptable, this did not arise in my study. Rowland and Metcalfe (2013) in their summary of the literature continued to highlight that parents telling their children early on, enables the child to develop coping strategies, which enhance their emotional well-being and this too was evident in my study as where a participant was told early on by health professionals that they had Usher they were able to prepare for leaving home and an independent life. Additionally, parents themselves benefit as keeping the secret, causes parental distress and anxiety (Rowland and Metcalf, 2013). Rowland and Metcalfe further highlighted that research indicates that children want to know and want their parent(s) to communicate openly and honestly about genetic conditions. However, there was only one experience given in my study of parent(s) keeping Usher a secret from the child, and is not representative.

The literature discussed in this section identified mixed feelings with regard to telling the child they had Usher. As highlighted, in both my study and Ellis and Hodges's (2013) study, some participants felt life prior to diagnosis was 'bliss' as they were unaware of their future, whereas others felt it imperative to tell the child to enable them to be better prepared for what lay ahead. Whether it is the health professional or parent(s) who has to communicate the diagnosis, it is clear that the task of imparting such news is a difficult one. It is hard for health professionals or parent (s) to have to tell a child they have a serious life altering condition but there may be more distress when the condition is genetic/hereditary and was passed down by the parents themselves, as highlighted by Rowland and Metcalf (2013).

5.2.4 Section summary

Three key messages discussed in this section were: Usher although not life threatening is life altering; the diagnosis of Usher impacts on individuals in varied ways, and the way the diagnosis of Usher is communicated affects each individual's experience. The findings were discussed in relation to previous research and Kübler-Ross's theory of bereavement (1970) was considered and critiqued. The study highlighted that a diagnosis of Usher engendered vulnerability with fears about personal safety, such as crossing roads, especially in the light of hybrid and electric cars, and fear of house burglary. Although, Damen *et al*'s (2005) quantitative survey explored the lives of 93 participants with Usher syndrome and highlighted that as people got older and their sight and hearing worsened; they feared not knowing when an emergency situation occurred, these personal fears have been little raised in previous research about an Usher diagnosis.

Literature relating to breaking bad news with regards to life threatening conditions such as cancer was highlighted but there was no previous studies that specifically

focused on breaking news about an Usher diagnosis. Literature was also considered relating to the complexities of telling a child they have a genetic condition and concluded that the task is difficult but necessary. Next key messages that emerged from the theme 'Familial relationships across the lifespan' will be discussed.

5.3 Discussion: Theme 2 – 'Familial relationships across the lifespan'

Two key messages emerged from the theme 'familial relationships across the lifespan': a diagnosis of Usher impacts on new relationships and the management of existing ones, and Usher being an unseen hereditary/genetic condition has consequences for the person with Usher and their families. The first key message will be discussed next.

5.3.1 A diagnosis of Usher, impacts on new relationships and the management of existing ones

The current study findings showed that a diagnosis of Usher impacted relationships across the lifespan including partners, marital relationships, children and parents. Participants described how Usher affects the forming of relationships as there were difficulties building friendships and meeting partners. Although there is literature which explores disability over the life course (Priestly 2003); the effects of having a disabled child on the family (Reichman *et al,* 2008); blogs on whether relationships could survive disability (Senelick, 2011); online support networks such as Enhance the UK (changing society's view on disability) and studies on the effect of existing relationships (Ellis and Hodges, 2013), there does not appear to be literature directly relating to how Usher impacts new relationships.

Erikson's (1982) theoretical concept highlighted that the challenges people face whilst navigating through the eight stages of development can influence the forming of intimate relationships (Erikson, 1982, Walker and Crawford, 2010). This is compatible with the findings in chapter 4 which highlighted that a diagnosis of Usher impacted on new relationships and the management of existing ones, because for some participants knowing they had a degenerative but unseen condition affected building new relationships; for others, the inability to socialise in certain environments prevented forming relationships, and for others scarcity of socialising had a profound effect on their identify development and ability to form relationships. Martens *et al* (2014) suggested that key to the development of secure attachments within relationships is the need for reciprocal sharing of emotions, to establish a bond and build trust within the relationship. Although Martens ***et al's*** (2014) study was only small scale (n=4) and referred to the relationship between participants and their communication partners, the principles could apply within relationships, especially when one partner experiences Usher.

Jacobson *et al* (1997) conducted a longitudinal 10 year study with children and adolescents with Insulin Dependent Type 1 Diabetes Mellitus ($n = 57$) and an age-matched group who were originally recruited after an acute illness, accident, or injury ($n = 54$) to see whether participants with diabetes experienced problems in close peer relationships in terms of friendships, dating and "love experiences" (p73) more or the same as persons without diabetes. The authors found that there was "less trust and sense of intimate friendship" (Jacobson *et al,* 1997 p73) in love relationships for people with diabetes. However, the research did not suggest that diabetes contributed to problems forming relationships, but rather that the reason for less trust related to lower self-worth on the part of the person experiencing diabetes. Whilst Jackson's study highlights how a condition (and one that is long-term and

may be progressive) affects relationship forming, Usher has additional implications, being genetic and one that affects communication through sensory loss.

In the current study, participants identified how Usher affected the dynamics of existing relationships, when a partner acquires a life altering disability. Similarly, Ellis and Hodges's (2013) study reported on the need for changing roles for people with Usher to maintain their relationships. Other authors have explored the topic but in relation to acquired hearing loss (Hetu *et al,* 1993), chronic illness and disability (Lyons *et al,* 1995) and diabetes, types 1 and 2 (Wiebe *et al,* 2016).

Hetu *et al* (1993) highlighted that acquired hearing loss strongly affects relationships as the deteriorating and progressive sensory loss affects not only communication but coping processes required to live harmoniously as the condition develops. In my study, one participant relayed her experience of how her sight loss impacted the visual form of communication she used, which in turn affected her friendships and the techniques she adopted to overcome the situation. Another participant expressed how practical issues such as sharing a computer required thought to live harmoniously, when one partner had sight loss and one did not, each having different software needs. Lyons *et al* (1995) also discussed how relationships with family and friends change with the onset of chronic illness and disability and suggested coping techniques to maintain close relationships.

Maintaining existing and developing new relationships is important as my study highlighted that supportive relationships had a beneficial effect on the people's experience. Weibe *et al* (2016) in their study of the social context of managing diabetes types 1 and 2 identified how effective management of diabetes was improved with positive social relationships. Weibe highlighted that good support from family and friends, but especially partners contributes to effective diabetes

management and positive outcomes. However, poor support and criticism from familial members was consistently associated with poor diabetes outcomes (Weibe *et al,* 2016). In my study one participant, when referring to people he knew who had depression as a result of Usher, expressed that his outlook was more positive because his wife and 3 children were very supportive. Clearly acquired disability impacts relationships, but as the literature suggests, people with acquired disability and their partners are often willing to support each other to maintain their relationships. This too arose in my study, as when participants were supported either by parent(s), siblings or partners, their experiences were more positive, despite the seriousness of the condition.

Also highlighted in my study by participants was how the progression of Usher resulted in greater responsibility for their partner without Usher, and reduced independence for themselves. In Ellis and Hodges's (2013) study one participant expressed how she stayed at home most of the time now she had Usher. However, Ellis and Hodges highlighted those incidences such as communicator guide provision was a factor with regards to lack of independence. The participant who stayed at home most of the time did not have communication guide support at all, whereas another participant who received 53 hours communication guide support a week reported a full and active life. Ellis and Hodges's participants also reported that their independence was limited, with their partner's responsibilities being heightened. For example one participant recounted how her husband had to do the driving and take her to the toilet if she needed it.

However, Ellis and Hodges also highlighted that often partner support was two ways, as the person with Usher also supported their partners who did not have it. Similarly two way support arose in my study, for example although one participant's wife now did all the driving due to her husband's sight and hearing loss but the

husband supported his wife by ensuring as far as possible the car she drove was reliable by utilising benefits to obtain a Motability car. In both Ellis and Hodges's study and my study, in the main partners were willing to offer support. However, Ellis and Hodges (2013) recounted experiences of participants who when diagnosed with Usher and offered their partners a way out of the relationship as they did not want to burden them; this situation did not arise in my study. The next aspect to be considered with regard to the effect on maintaining existing relationships relates to the impact of parental guilt on the children.

5.3.1.1The impact of parental guilt on the children

Another finding that arose in my study was that children may worry about parent(s) when parents experience guilt, and as the child does not want to upset their parents, they may not look to them for support. Thompson (1996) defines guilt as "feeling of culpability" (p440) and culpability as "deserving blame" (p238) thus guilt can occur when a person feels that they have wronged another person. An example of this occurring was a participant in my study who was diagnosed age 16 and as a grown up child considered her parents' feelings, when considering sharing her own about the diagnosis of Usher, as she felt they already felt guilt as the condition was genetic/hereditary. The only other study that referred to childhood concerns or anxieties for parents when a condition is genetic was Ellis and Hodges (2013) as they referred to one participant giving an account that their parent(s) were worried or upset about their child's diagnosis and another participant who mentioned that their mother bore guilt and projected their fears onto their child.

It is understandable that parents may feel a sense of guilt as they are medically advised that they have passed a serious life altering condition onto their child without themselves being affected. Malpas (2006) acknowledged that parents would

prefer there not to be a genetic condition in the family. James *et al* (2006) considered how guilt and blame can affect parent(s) when a child is diagnosed with a genetic condition. To understand the context, Thompson (1996) defines blame as "assign fault or responsibility to; assign responsibility for an error or wrong" (p96), so blame is the assignation of responsibility for the situation occurring.

James *et al* (2006) interviewed 112 members of 51 families and found that not only does a genetic condition have a medical effect on the families, but also a psychosocial one, as parents need support, information and counselling when a child is diagnosed with a condition that is passed on by their parent(s). They concluded that when genetic counselling is being given, particular attention needs to be given to issues around parental guilt and blame because these have a significant effect on parent(s) and subsequently the way in which they support their children. As discussed previously in section 5.2.3, the issue of feelings of guilt on behalf of the parents when their child has Usher was highlighted as part of this study, and is a thought provoking area because the issue of parental guilt arises not only within Usher but other genetic conditions. For example, Chapman's (2002) study explored issues around pre-natal screening for cystic fibrosis (CF) and the decisions they would have made if they had known that their child had a hereditary/genetic condition. Their views were varied. One of Chapman's participants expressed the importance of genetic screening because then his parents would have had choices as to whether to keep their baby or not. However another participant felt that, due to medical advancements, there was a lot that could be done to enable a person diagnosed with CF to have a healthy life, so even though she had the condition herself, she still felt that she would give birth to a baby with CF. Another participant commented "well the trouble is I sometimes think that everybody wants a perfect child. They want their child to be perfect but that's not like that, sometimes there are hiccups" (Chapman 2002 p202). However, often parents who carry genes that

contribute to hereditary/genetic conditions are completely unaware they do until their child, early or later in life is diagnosed. While there have been medical advancements and people with CF can be better treated, treatments for Usher remain limited. Different types of hereditary conditions evoke different responses, for example although CF is still life limiting and Usher is not, Usher is still increasingly debilitating and life altering.

Lock *et al* (2006) found in their study with relatives of people who had Alzheimer's, that when "genetics is brought up during family exchanges it can bring anxiety or anger" (p287). These exchanges indicated that causation of the condition contributes to family stress. One participants said, "a lot of people just think it's bad genes, you know, that it's just bad genetics. She comes from a bad gene family. It made me question the quality of my own family genetics" (p287). Another participant said "If Alzheimer's happens to me, it happens to me. But I would be much more concerned if I had children" (p281). In both studies by Chapman (2002) and Lock *et al* (2006) participants were presented as more worried about their children having a hereditary/genetic condition, than if they had it themselves; if they had it themselves they could cope, but seeing their children have something they had passed on was less bearable. Within my study participants reported that their parents experienced guilt as parents were carriers and passed down the condition to their children. Some participants struggled to talk to their parents about having Usher but they did not appear to blame the parents for being carriers.

Ellis and Hodges (2013) also referred to Usher being a genetic condition and that siblings of people with Usher became more interested in the geneticness of the condition when they [siblings without Usher] were considering having children.
As discussed, previous studies have reported that acquired disability affects relationships (Hetu *et al,* 1993, Lyons *et al,* 1995, Weibe *et al*, 2016) but, with the

exception of Ellis and Hodges's study, none related to Usher specifically. Although as has been identified, the management of existing relationships has been discussed in literature, the impact for people living with Usher developing new relationships has not been researched. The third key message will be considered next.

5.3.2 Usher being an unseen hereditary/genetic condition has consequences for the person with Usher and their families

In this study participants felt that their condition being largely unseen, had implications associated with genetic/hereditary conditions, such as passing the gene onto their children, and was life altering in terms of the progressive, debilitating nature of the condition. Indeed, one participant referred to Usher syndrome as a "*horrible dark secret*" (see 4.3.4). Similarly, participants in Ellis and Hodges's (2013) study also referred to Usher being a "hidden disability" (p177) as they did not appear blind; these views will be explored next.

5.3.2.1The burden of an unseen condition

There have been studies conducted with people who experience genetic/hereditary conditions, for example, Petersen's (2006) study included people with a variety of genetic conditions such as cystic fibrosis, haemochromatosis, haemophilia, and thalassaemia. Petersen posited that different conditions present a variety of challenges and disruptions as people's lives alter. Petersen (2006) further highlights that there are a range of genetic conditions which are largely unseen and do not show symptoms (asymptomatic), but still have a life altering effect on a person's life.

However, most literature that discusses Usher syndrome as a hereditary/genetic condition is clinical (Cohen *et al,* 2007, Moller *et al,* 2009 and Sadeghi *et al,* 2004),

and does not consider psychological or social aspects of its life altering affect. There are other studies that have investigated living with other hereditary/genetic conditions, for example, Huntingdon's disease[48] (Chapman, 2002), cystitis fibrosis[49] (Chapman, 2002) and Alzheimer's[50] (Goldman *et al,* 2008) and the principles from these findings could be similar to those with people living with Usher and are therefore discussed next.

Chapman's (2002) research provided a phenomenological perspective on what it is like to live with a hereditary/genetic condition; the two conditions that were the primary focus were Huntingdon's disease (HD) and cystic fibrosis (CF). One participant in the study with CF felt they still had a life as although they had limitations they were not incapacitated completely. Another felt that they could be worse off and made the best of what they had and another expressed: "All right this illness is part of my life, but I still do have a life" (Chapman, 2002 p199). Overall, Chapman reported positive quality of life experiences. These findings were similar to my study as some participants also expressed they felt that they were happy and made the most of their life.

However, in my study there were other participants who were depressed by having Usher. Lock *et al* (2008) in their study of people with Alzheimer's reported how one participant had already had genetic testing for breast cancer and psychiatric disorders, as well as Alzheimer's, only to be told that she was at risk of all conditions. This participant relayed how she had a "family where everyone was depressed" (p291). This raises the question that whilst genetic testing and diagnosis of such conditions are essential, does it contribute to depression or affect the way people live their lives. As with Usher, Chapman's study also highlighted that there was no definite prognosis; some participants were seriously debilitated at 40 whereas others could still go on cruises in their 70s. Chapman asserted that "the

voices of people living with genetic conditions should form part of wider bioethical debates arising from such advances" (Chapman, 2002 p195).

It could also be suggested that another aspect of Usher being a "*horrible dark secret*" may relate to the stigma of disability. In my study one participant said she did a lot of "*tongue biting" (*see 4.5.3) particularly when she was with her guide dog and referred to an incident where she was at work and a member of the public said "*I think its lovely they have given you this job*" (see 4.5.3). Therefore although the benefits of having a guide dog have been highlighted in the findings a guide dog is also a visual way of displaying disability and affects public attitudes. Aiden and McCarthy (2014) highlighted that although there has been legislation put in place to challenge public attitudes and discrimination against disabled people, negative attitudes still exist. A reason for such attitudes and perceptions was asserted by Arndt and Parker (2016) who conveyed that barriers are often established by people who do not have a disability themselves or have very little contact with people who do have disabilities. Aiden and McCarthy (2014) further added that research conducted by Opinium (A market research consultancy) found that 76% of people felt that disabled people needed to be cared for. However, it could be suggested that there are tensions between displaying and not displaying disability. On the one hand, the condition being unseen is challenging as people do not understand reasons why people behave in a certain way e.g. bumping into other people because they cannot see, but then when the condition is seen, for example by a person having a guide dog or using red and white cane, it can lead to pre-conceived public perceptions and stigma.

One of Chapman's participants said that he sometimes felt like he was "fundamentally flawed or at least society thinks I am" (Chapman, 2002 p201). In Ellis and Hodges's (2013) study one participant said they tried to be as "normal as

possible” (p139-140). These experiences arose in my findings too, where participants felt they had been treated differently when others realised they had a disability. One participant felt that using equipment identified them as different and another felt that without using sensory equipment such as a red and white cane, which identified him as having a disability he could “*pass as human*” (see 4.5.3). The comment about, passing “as human” implies the perception that a human does not have a disability; it could be suggested that this relates to public and societal perceptions of what is normal (Aiden and McCarthy, 2014). In chapter one Aiden and McCarthy posited that there was a lot of unease about disability and nearly half of the British public did not know anything about disability, so it is not surprising that comments such as this one arose. The participant’s comment bring to mind, the social model of disability which proffers that people are not disabled by their impairments but by the disabling barriers people with disabilities face in society (Oliver, 2013, Bricher, 2000, *UPIAS, 1976*). Whilst application of this model combined with passionate campaigning by and on behalf of disabled people, have influenced societal and legislative views with regard to disability and strengthened people with disabilities movements (Equality Act 2010, Sense, 2012, Oliver, 2013), limitations are highlighted that this model sees people with disabilities as a disabled group rather than individuals who are different and diverse (Oliver, 2013), which may contribute to public and societal perception of disability. It could be suggested that it is unacceptable that people who experience disability, feel that because of public and societal perceptions; they are reticent to use equipment or services that may assist them because it makes their disability more visible. Therefore the person with the disability would rather not use such equipment to enable their condition to be unseen, which in itself is a burden.

As has been revealed by participants in chapter 4 and the discussion of literature in this chapter, living with a genetic condition is a challenge not only for people with

Usher syndrome, but all people who are living with a hereditary genetic condition. Goldman *et al* (2008) suggest the benefits of genetic counselling for people who are affected by genetic mutations. Although their study focuses on people with Alzheimer's the principles could be useful for people with Usher, as they too are affected by the fact that Usher is a hereditary/genetic condition. Just as Goldman *et al* (2008) found that people were eager to receive information with regards to their condition, each person's individual experience would differ, depending on family dynamics, individual personalities and their environments. Therefore, beneficial genetic counselling needs to be individualised, as it may not be for everyone.
The literature discussed highlighted that although issues around the impact of genetic conditions have been considered, Usher syndrome is a genetic/hereditary condition that affects people greatly. The principles from studies relating to other genetic/hereditary conditions are useful when considering Usher. However, regardless of which genetic/hereditary condition it is, the fact remains that unseen conditions, which have a life altering effect can be described as "*a horrible dark secret*".

5.3.3 Section summary

In this section, two key messages were discussed from the theme 'Familial relationships across the lifespan': a diagnosis of Usher impacts on new relationships and the management of existing ones; and also that, Usher being an unseen hereditary/genetic condition has consequences for the person with Usher and their families. Literature was discussed to consider the implications of a condition being genetic/hereditary. Whilst cystic fibrosis and Huntingdon's disease were considered, the impact of Usher being passed on by parents was also discussed and the impact on parent(s) of keeping the Usher secret. The section also discussed how relationships and roles needed to be changed to maintain relationships and how

positive familial support contributed to wellbeing and coping with an acquired condition. Also discussed was the impact when a child felt their parent(s) experienced guilt and how this affected the familial relationship and the tensions between displaying and not displaying disability within society. Next a sense of belonging will be discussed.

5.4 Discussion: Theme 3 – 'A sense of belonging'

Three key messages emerged: Acquired disability affects communication; Deaf/Deafblind culture can contribute to a sense of belonging, and technology can support a sense of belonging. The first key message will next be discussed.

5.4.1 Acquired disability affects communication

In my study, participants' experiences relating to how acquired disability affected their communication were powerful; similarly Aitken *et al* (2000) posited that the devastating effects of deafblindness are most apparent in the area of communication. One participant's experience illustrated how communication differences and lack of understanding affected their sense of belonging. The experience also highlighted that forming and maintaining relationships is closely linked with communication; when communication is affected so too are relationships. Kyle and Barnett (2012) reported that participants who were deafblind felt less a part of the Deaf community reporting that "Deaf people said they did not know how to communicate with Deafblind people. It was as if Deafblind people were foreigners" (Kyle and Barnett, 2012 p7).However other participants in the same study expressed that the differences in Deaf people's experiences, from childhood through to adulthood, shows the Deaf community to be a "rich and varied society which tolerates diversity" for example, in terms of communication (Kyle and Barnett, 2012 p14).

Whilst Ellis and Hodges (2013) found that that the “communication method did not change much” (p11) after diagnosis, in the discussion they reported that Deaf people felt less a part of the Deaf community. Ellis and Hodges also reviewed how people with Usher spoke about themselves by examining YouTube videos, blogs, social media, internet articles etc. and reported the effects of not being able to see when already Deaf and how the loss of vision led to changes in relationships (p32). Erikson’s (1982) theoretical concept of identity development was compatible with the key message that acquired disability affects communication. Erikson posited that ideal identity development included finding social roles that enable a person to fit into the wider community (Kruger, 2007). This idea was compatible with the experience of one participant who highlighted the communication difficulties that acquired disability imposed on her and the effect it had on her relationships with her friends, as BSL is their first language.

In addition to Erikson’s theory, the social exchange theory as presented by George Homans was also compatible with this key message because it explores social behaviour from a psychology and sociology perspective and suggests that social change and stability are a process of negotiated exchanges between two or more people (Cook and Whitmeyer, 1992, Weber and Bouwel, 2005). Furthermore, Homans ideas around social exchange focused on “the actual social behaviour of individuals in direct contact with one another” (Homans, 1961 p3), which again applies to the participant’s situation. Weber and Bouwel (2005) considered that “social structures can arise from and be maintained by individual actions” (p90). Cook and Whitmeyer (1992) concurred with this suggesting that social behaviour can influence outcome because it can be rewarding and facilitate positive outcomes for individuals.

My findings highlighted that negotiating exchanges between, one of the participants and members of their Deaf club, when the participant felt passionately enough to address the impact her acquired deafblindness had on her ability to communicate with her friends, resulted in awareness being raised and social interaction being restored. Social exchange theory questions “if the cost of self-effort is worth the reward” that could be achieved (Weber and Bouwel, 2005 p90). In this participant’s experience the effort was worthwhile, because she was “the expert” in her situation (Parker and Bradley, 2010 p15) and her whole life was entwined with the Deaf community but communication barriers prevented her from being a part of it. However, for social exchange to be successful the outcome needs to be achievable and often relationships with other people are needed to contribute to their success. For example, the participant in my study would not have been as successful in her negotiation if she had not been supported by her daughter and a specialist deafblind charity. A critique of the social exchange theory is that it was developed in the 1960s during a time when freedom of expression and openness was encouraged and in today’s contemporary society freedom of expression is arguably reduced (Miller, 2005).

Social exchange theory has shown that for relationships to be maintained there needs to be negotiation and action. It is evident that acquired deafblindness/Usher does affect communication and consequently relationships, but as shown by the person living with Usher addressing these concerns, a positive outcome can be achieved. Next how Deaf/Deafblind culture can contribute to a sense of belonging will be discussed.

5.4.2 Deaf/Deafblind culture can contribute to a sense of belonging

In my study, culturally Deaf participants after a diagnosis of Usher still referred to themselves as Deaf, not capital 'D' [15] deafblind or small 'd' [16] deafblind. This notion of self-perception was also discussed in Kyle and Barnett's (2012) study where participants who had Usher, but were previously Deaf, referred to themselves as Deafblind, as opposed to having Usher.

The idea of self- perception, leads to consideration of labelling theory. The idea of labelling theory was first theorised by Emile Durkheim, a French sociologist who considered that labelling people who committed crimes satisfied society as people were labelled and contributed to society's desire to control a person's behaviour. Durkheim's book 'Le Suicide (The Suicide)' explored the idea of labelling in more detail (Durkheim, 1897, 1952), however, Howard Becker further developed labelling theory (Becker, 1973), suggesting that labelling theory is concerned with the propensity for minority groups to have negative labels or names given to them. Identity theory posits that labelling affects people's self-concept. For example one participant expressed that he "*could pass as human*" (see 4.5.3), a profound statement which supports ideas around identity theory as it infers that public perception is that disability is labelled as not being human. Also, in my study, being labelled or identified as a member of the Deaf community, was perceived as positive and contributed to a strong sense of belonging.

Some participants experienced feelings that culturally Deaf people were '*locked in their own world*' (see 4.4.1), indicating that people outside of that world were not included within the group. However, it could be suggested that this is no different to a person from another country, and whose first language was something other than English, coming to live in the UK. They may choose to mix with a group of people whose first language is their own. Within this group they may feel affinity and a

shared understanding where they can discuss their culture, heritage and history. This can be likened to people whose first language is BSL and are part of the Deaf community.

One participant in my study expressed that he did not accept the term "*Deaf community*" because he wanted "*integration*" (see 4.4.1), this raises questions around the issues of segregation versus integration and being disabled and being normal. However, it could be suggested that in order for people with a disability and those without to integrate better with each other, awareness needs to be raised to improve understanding. Sense commissioned a report which explored the current attitudes towards disabled people (Aiden and McCarthy, 2014). The report highlighted "both the general public and disabled people believe that more everyday interactions and greater public education about disability will increase understanding and acceptance of disabled people" (Aiden and McCarthy, 2014 p3). The authors suggested that for better integration to take place "public education could help tackle the discrimination and stigma associated with disability" (p14). The participants in Aiden and McCarthy's report were pro integration and it was considered essential to avoid segregation. Whilst this report supports the participant's comment, it is also important to note that personal choice is essential and there were perceived benefits of feeling a sense of belonging by being part of a community group. These findings highlight that people living with Usher are unique and individual.

The uniqueness of Deaf culture was highlighted from findings with regard to experiences of positivity associated with the allocation of personal identification number upon school entry. Although, I was surprised by this practice, the practicalities made sense. For example when a person lipreads, some words can become muddled, for example, bat and hat and some letters, for example, V and F can be difficult to differentiate when a person is lipreading. Also if a person is trying

to read the lips of someone who has significant facial hair, lips that do not move very much or lips that move too much can cause difficulties for the lipreader (Evans and Whittaker, 2010). I critically reflected that as a hearing person, I had seen negative connotations to the issue of personal identity numbers but on reflection, the findings highlighted that this practice contributed to relationship building and a sense of belonging for the participant who discussed it. This participant was the only person in my study who gave this experience, and there does not appear to be any research specific to experiences of giving personal identity numbers to Deaf children upon entry to schools for profoundly Deaf children whose first language is BSL.

Another aspect relating to self-perception was in relation to normalisation. In Ellis and Hodges's study, participants had varying views of whether people should normalise their condition or not. One of Hodges and Ellis's participants felt that they should "act like you are normal" (p63); another expressed "don't try to be normal" (p62) whilst another voiced that they were "knackered trying to be normal" (p114). Similarly, there was also a strong message that participants felt they were not normal because of their sight and vision difficulties in my study. As previously highlighted, one participant when referring to not using recognisable sensory equipment e.g. white/red and white cane said *"I could pass as human*" (see 4.5.3). Another felt that being identified as deafblind changed him as a person and another did not want to use hearing aids which he felt identified him as different. Still, another participant referred to normal children being educated to have awareness of children who did have sight and hearing difficulties. These comments highlighted that people do not want to be labelled as being different but perceived that normality entailed having unimpaired sight and hearing.

The idea of labelling also arose when one of the participants gave her experience that her guide dog visually displayed her disability and gave an account of public

attitudes associated with this. It could be suggested there are tensions between displaying and not displaying disability as although people with a disability need adaptations and support with regards to their disability, public perceptions may lead to pre-conceived ideas, stigmatisation and ultimately labelling. Labelling also resonated with another participant who expressed that the government painted "*all disabled people with the same brush*" (see 4.5.4).

Another area that arose in the findings was whether there were two Usher cultures; those whose first language is BSL, and those who are hearing and sight impaired and use speech. Interestingly, this aspect also arose in Ellis and Hodges's study (2013). In both studies, this topic only featured with one participant and as both participants used the same analogy to refer to the two groups e.g. "*Manchester United and Manchester City*" (Ellis and Hodges, 2013 p102), and both participants seem to be the same age, gender and nationality, there is a strong possibility that this is the same participant. The same participant also referred to the two groups as being like "*East Berlin and West Berlin*" (see 4.4.3). The two analogies given by the participant highlight his perception of the distinct differences between the two groups. Just as there are diverse differences between the two football teams (Ballard, 2017) mentioned and the two German occupation zones (Stein, 1997) referred to, so too could there be differences within Usher culture.

In this section, literature was discussed and critiqued together with an exploration of the social exchange and labelling theories. The social exchange theory was discussed, and highlighted that, for this theory to be effective the outcome needs to be achievable and it also relies on relationships with other people. Differences between Capital 'D' Deafness/Deafblindness and small 'd' deafness/deafblindness were also discussed and the importance of being part of a cultural group for some individuals. Examples were provided of how acquired disability affects

communication and the notion that communication affects person's position within the Deaf community was discussed. Also discussed was the message that a person was not 'normal' if they have sight and vision loss. Next, how technology can support a sense of belonging will be discussed.

5.4.3 Technology can support a sense of belonging

The third key message, 'technology can support a sense of belonging' explored the impact of technology and social media to contribute to participants making contact with other people who were living with Usher syndrome. My study findings identified that technology such as Twitter [20], Facebook[18], FaceTime[19] Skype [21] iPad[23], IPhone[24], Kindles[25], speech to text[26], video conferencing[27] and Email [22] enabled people to not only keep in contact with family and friends but access support networks and share experiences all over the world. This in turn contributed to experiences of reduced isolation and increased inclusion. Ellis and Hodges (2013) too, found that social media such as Facebook was a useful tool for people living with Usher to meet other people with the same condition and not feel alone. One participant in their study expressed the value of Facebook as when she was diagnosed she was told Usher syndrome was rare, which led to her thinking she was alone; use of technology/social media dispelled these thoughts. Another participant in their study highlighted the value of social media in sharing experiences and getting answers to Usher related questions.

Findings from my study highlighted that the type of technology chosen by the participant, not only in terms of interview choice (Table 3.4) but also to communicate with other people with Usher was dependent on their level of sight and hearing loss. Where a person has limited sight but a better level of hearing, Skype may be a form of communication they may choose. For example there was one participant who

chose to be interviewed via Skype with the video camera off, so then whilst he could not see me; I could not see him either. However where a person has limited sight and hearing Skype may not be an option of choice as it may not use any residual hearing and sight to its fullest capacity. However, as discussed in 4.4.2, for a person who is profoundly Deaf an iPad facilitates effective communication. As discussed in chapter 3, the choice of interview and communication method used was participant led (see Table 3.4) which contributed to autonomy and dignity. Regardless of the particular method chosen, the use of technology could make a positive difference to the lives of people with Usher.

Maiorana-Basas and Pagliaro (2014) conducted a nation-wide survey for people living in the USA to explore the technological use, needs, and preferences of people who are Deaf and hard of hearing. Maiorana-Basas and Pagliaro (2014) acknowledged that as technology advances and becomes a necessity for communication and participation in society, education and business, people who are Deaf and hard of hearing also need to access technology. Maiorana-Basas and Pagliaro (2014) highlight that although technology has the potential to reduce isolation and increase independence for people who are Deaf and hard of hearing, accessibility needs to be available in a format that meets sensory needs. Arndt and Parker (2016) who conducted a descriptive qualitative study in the USA with ten adults who were deafblind about their social lives supported Maiorana-Basas and Pagliaro (2014) comments, outlining that it is essential for there to be an understanding of the needs of people who experience the complexities of deafblindness to support them to get access to community involvement and meaningful traditional and virtual social connections.

Developing and sustaining social networks may be a challenge for people who are deafblind (Möller and Danermark, 2007). However, Maiorana-Basas and Pagliaro

(2014) posit that the internet has been useful for providing an equitable environment for people who have individual needs and has contributed to empowerment in terms of presenting opportunities for people to access information and socialise (Maiorana-Basas and Pagliaro, 2014, Barak and Sadovsky, 2008). The notion of empowerment is supported by Barak and Sadovsky (2008) who posited that as "cyberspace" (p1802) provided a platform for people to communicate using visuals, texts and images as opposed to just auditory means, this facilitated concealment of any disability a person may have, offering them more confidence and a greater sense of equality. Maiorana-Basas and Pagliaro (2014) concluded that after conducting their research they found that there was a prominent use of smart phones and social network sites such as Facebook amongst people who are Deaf and hard of hearing and that technology contributed to levelling, "the playing field" (p10) for people with sensory needs. Literature discussed highlights the role technology plays in contributing to a sense of belonging, although at the same time acknowledging a culture of technological change.

Communication theory is pertinent to this key message as it proffers that communication is concerned with "sharing of meaningful interactions with other people in the world" and "concerns the passing of, and receiving and acting on information" (Randall and Parker, 2000 p69). Claude Shannon was an American mathematician and electrical engineer who developed communication theory (Shannon, 1949) and was said to have "laid the foundations for all digital communications" (James, 2009 p257). James further highlighted that without Shannon's work "email and the World Wide Web would not have been possible" (p257). For people to feel a part of something, be it a physical (Usher support group) or virtual (Facebook) community, there needs to be common information that can be shared, to benefit the receiver and the recipient.

These ideas link back to communication theory (Randall and Parker, 2000, Shannon, 1949, James, 2009) as the message needs be conveyed from: the message source, which is then enciphered; then deciphered and the message is received (Shannon 1949). Randall and Parker explain that this process included: choosing a way of transmitting the message for example “language, gesture, writing”; the person receiving the message and decoding or deciphering it by “hearing, seeing, reading” and then the recipient is responding depending on how they interpreted the message by way of a “reply” (p69). The participants in this study used technology and social media as a way of belonging, sharing common experiences and learning more about what life is like to live with Usher. Literature discussed in this section supported technology as a means of enabling people with Usher to have a sense of belonging, whether it was physically or virtually.

If a person has been ensconced in the hearing/sighted world, it can be overwhelming to no longer be part of it, not only being isolated from family and friends but the environment in general. Lieberman and Stuart (2002) surveyed 51 adults who were deafblind about their experiences of social interactions and asserted that as dual sensory impairment often led to isolation and loneliness access to a variety of recreational pursuits are essential. Therefore, having access to technology and social media such as Facebook, Twitter etc. can contribute to people living with Usher no longer being alone.

However, access to technology is dependent on sensory adaptations to meet needs (Maiorana-Basas and Pagliaro, 2014). Maiorana-Basas and Pagliaro found that people who are deaf and hard of hearing have historically looked at new technologies with a level of anticipation with good reason, because often, although technological advancements are made, facilities for full access for Deaf and hard of hearing people are not available, for example, video captioning (displaying additional text/subtitles to provide additional interpretative information). As internet

available informational, entertainment videos or audios are provided whether recorded or streamed live, they are seldom captioned, and there do not appear to be any laws to enforce captioning of all video or audio content on the Internet, unless the content was broadcast first on television with captions (3PlayMedia, 2013, Maiorana-Basas and Pagliaro, 2014). Although, this study was with people who were deaf and hard of hearing, the principles apply for people with loss of sight and hearing which adds to the complexity of technological access.

However, even where access is probable, my findings also showed that some people will not embrace technology whilst others will utilise it to the fullest; one participant accessed people all over the world. Clearly, although, the findings identified that technology in its various forms is useful for people with Usher, the participant group was relatively small in number to really ascertain the full extent to which technology impacts people with Usher. Also, although primarily the benefits of technology were conveyed in this study, it is likely that other people with Usher have different or negative experiences, which if explored may provide a basis for developing a way for technology to be promoted to all people with Usher. One participant in my study highlighted that it was a charitable organisation that had encouraged her to use an iPad. However, not all people will have an awareness of such organisations and therefore could miss out on support and demonstrations of the communicative benefits of technology.

5.4.4 Section summary

In this section three key messages were discussed including acquired disability and how it affects communication, the essentiality of understanding Deaf/Deafblind culture and how it contributes to a sense of belonging, and how technology can support a sense of belonging. Literature and the theories relating to social

exchange, labelling and communication were also discussed. The discussion finished with exploring how people living with Usher syndrome can be supported to utilise technology and social media, with a focus on encouraging everyone with Usher to try it. Next, experiences of professional support will be discussed.

5.5 Discussion: Theme 4 – 'Experiences of professional support'

Two key messages emerged from findings relating to experiences of professional support: Specialist Usher support is necessary regardless of demographics, and there needs to be increased awareness of Usher amongst professionals and the general public.

5.5.1 Specialist Usher support is necessary regardless of demographics

The first key message relating to experiences of professional support relates to specialist support regardless of demographics. Côté *et al's* (2013) mixed method study highlighted the importance of specialist support, positing that lack of it can affect well-being, life satisfaction and realization of personal goals. The participants in my study found that the services and support they received varied from area to area and was a postcode lottery. In light of limited funds being available and no worldwide consensus about how healthcare should be delivered, service provision in the UK has led to a postcode lottery approach (Breuning *et al*, 2010, Russel *et al,* 2013, and Robertson, 2017). Prain *et al* (2010, 2012) highlighted that people who experience dual sensory loss require specialist support services to meet their needs. However, the findings highlighted that some professionals who work within the sensory field display a lack of understanding and empathy about what is like to live with Usher and this could result in a service provision for Usher not being a priority, especially as Usher affects a relatively small section of the population. However,

findings also showed that lack of specialist support can impact a person living with Usher's health and wellbeing, thus specialist Usher support being available regardless of demographics is essential.

Although, there was no literature directly pertaining to specialist support regardless of demographics relating to Usher, this is not surprising as a GP may not have any deafblind people registered in their practice, or any specifically with Usher, so may be unaware of needs and requirements associated with deafblindness. However, there are examples of where the post code lottery approach arises in health care (Jones, 2012, Russell *et al,* 2013, Robertson, 2017), for example within GP commissioning, with GPs having to decide where their parliament allocated budget is spent (Jones, 2012, Russell *et al,* 2013). The reason this system was put in place was because the Health and Social Care Act 2012 abolished primary care and the government felt that GPs were best placed to know where their budgets should be spent (Jones, 2012, Russell *et al,* 2013). The Medicines and Prescribing Centre at the National Institute for Health an Clinical Excellence (MPCNIHCE) have been providing support to primary care trusts to improve services and prevent a post code lottery approach to health care occurring (Russell *et al,* 2013).However, although initiatives such as the MPCNIHCE have been implemented, specific services for Usher are unlikely to receive priority as the group is small in number and often greater funds are allocated to larger populations, for example ageing populations (Robertson, 2017).

Robertson (2017) argues that there are some "rare" exceptions where geographies of care or the post code lottery are justified, for example "population ageing" (p39). Robertson acknowledges that "human services vary across space" (p39) in terms of healthcare and social services support, but highlights that an ageing population presents a growing number of concerns and problems associated with older age

and as such geographies of care are needed. Robertson posited that there are challenges in providing integrated care to older people and people with disabilities, suggesting that as there is a greater shift towards community; care location will become ever more important to older people and those who experience disabilities. Robertson further proffers that integrated care needs to be "geographically informed to be effective" (p39). Thus Robertson is not only supporting the existence of a post code lottery with regards to healthcare, but is suggesting that in some cases, it is necessary to effectively support some groups of people.

Whilst there are analysts who predict demographic based healthcare forecasting (Jones, 2012) and the idea of "constant risk fallacy" (Nicholl, 2007 p1010) to understand demographic need, it could be suggested that healthcare demands which are more prevalent, recognised or advertised, such as cancer, ageing populations or dementia, would gain the greatest attention and thus more funding. Constant risk fallacy occurs because observational studies are used to compare who will require which services then case mix adjustment is used to take into account imbalances between group comparisons. If the risk factors used in the adjustment "are related to risk in different ways in the groups or populations being compared", and then this is ignored, this "commits constant risk fallacy" (Nicholl, 2007 p1010). As "large hospitals are built on such projections" (Jones, 2012 p40) and service provision prepared, it is not surprising that services for a rare condition such as Usher receives little attention.

Although literature is limited relating to the existence of a postcode lottery (Jones, 2012, Russell *et al,* 2013, Robertson, 2017), the literature discussed in this section, supports my study findings that a postcode healthcare lottery exists.
Although, people living with Usher should be able to have specialist Usher support regardless of demographics, in reality, unless there are changes at governmental

level, in which the complexity of Usher is acknowledged, services and support will remain unchanged. Next the need for increasing awareness of Usher amongst professionals and the general public will be discussed.

5.5.2 There needs to be an increased awareness of Usher amongst professionals and the general public

The final key message to be discussed is the need to increase awareness of Usher amongst professionals. Prain *et al* (2012) identified that even where workers have some sensory expertise, still services for people with sensory needs can be lacking. This will be discussed in terms of education, employment, health, the general public and governmental level.

Interviewing participants across a wide age range (18-82 years) enabled different experiences, and perspectives of life experiences of professional support to be gleaned. However, participants highlighted that not all their educational experiences were positive. Although less positive experiences could be deemed to be due to different points in time, with greater awareness, in more recent times, it could also be argued there should be a system where all children have equal access to educational sensory support regardless of their sensory situation or where they live. The UK's guidance and legislation (The Special Educational Needs and Disability guidance 2014, Education Act 2011) and special education needs under part 3 of the Children and Families Act 2014 outlines that local authorities and educational provision must take into account a child's sensory or special educational needs (NDCS, 2015). Argyropoulos and Thymakis (2015) proffered students with a disability that are well equipped, are better able to achieve their educational and vocational goals. Although their research was conducted with students with multiple disabilities and visual impairment, these principles would apply to students with Usher.

With regard to employment, where a person is unable to work due to Usher, findings showed that experiences were not always positive as outlined in 4.5.1. These findings were also discussed by Ellis and Hodges (2013), who reported that Usher affected employment opportunities and highlighted a need for greater awareness. Whilst participants in Ellis and Hodges's (2013) study reported ease of being signed off sick upon diagnosis of Usher and subsequent medical retirement, one participant in this study highlighted that once in a position of being unable to work and claiming benefits, further challenges were encountered. This participant shared that he would rather have still been in employment and in his job and it was due to his condition that he could not.

One of the reasons that Usher is not always recognised as a serious debilitating condition may be because it is unseen (Sense, 2014a, Petersen, 2006). If a person is in a wheelchair, it is clearly visible that they have limitations or a disability. However, when one looks at a person who experiences D/deafness and blindness they do not look like they have a disability. For example if a person with a sensory loss was communicating online, no one would know there was a sensory difference. Whiting (2014c) conducted research with families of children with disabilities and wrote a series of three papers relating to the professional support families received. Whiting found that making sense of their disability was important when supporting families (2014a) and that one of the key aspects of support was interacting positively with children and parent(s). However, the reality of the support children and their parent (s) received highlighted inadequacies and poor practice (Whiting, 2014b). In his third paper, Whiting (2014c) discussed the importance of good communication amongst health professionals to ensure that families are supported. Although his study was with parents of children with a disability, and this was not covered in this study, the principles that there needs to be understanding and awareness amongst professionals to support people with disabilities, remains the same. Whiting

concluded that “If healthcare professionals are to provide the help and support that parents seek, it is essential for them to develop an empathetic understanding of how disability might affect children, parents and families” (2014c p29), this idea of the need for empathetic understanding, also arose within my study where experiences were relayed when the child was diagnosed with Usher.

Although there was only one participant in my study who highlighted that Usher awareness needs to be raised at governmental level, this was still an important finding, as without support at governmental level, changes cannot be implemented nationally, to support people living with Usher. The participant raised this issue as a result of negative experience at the job centre, which had a profound effect upon him.

Whilst there are requirements on local authorities under the Local Authority and Social Services Act 1970 s7 to identify and provide an assessment for people who experience D/deafblindness, cuts in social care raise a question mark over the reality of being able to provide a tailor-made service that identifies and meets the individual needs of all D/deafblind people/people with Usher regardless of their geographical location. Furthermore as highlighted earlier in this chapter, whilst specialist charities endeavour to raise awareness in relation to D/deafblindness and Usher, they have limited resources, thus leaving gaps in development of services and support. Prain *et al* (2012) found in their study that staff policies and procedural documents needed to be explicit when providing support and services for deafblind people to ensure that their needs were addressed. Whilst Hadidi and Khateeb (2014) warned that sensory services need to be tailor made to ensure that people with sensory needs are safe, in terms of orientation, learning needs, employment and so on. Although Hadidi and Khateeb’s findings related to their participants with

visual impairment and was based in Jordan, Arab region, the findings could be relevant to other people with sensory needs.

Sense (2012) meets with all political parties in the House of Commons and the House of Lords to actively campaign and raise the profile of what it is like living with deafblindness. Although much success has already been achieved by Sense, the findings show that people living with Usher still need more support from the government, as identified above. Usher awareness raising is essential as well as greater recognition and integration of the social model of disability, which would contribute to governmental changes and improving the lives of people living with Usher.

The social model of disability argues that people are disabled by disabling and socio-political barriers as opposed to their impairments (Oliver, 2013, Bricher, 2000) and highlights that if this approach was adopted complexities would be recognised by the government. In this section legislation was highlighted which could be useful for raising awareness to the government. However, it was identified, that although charitable organisations such as Sense have actively campaigned for years, poor experiences, such as at the job centre still occur. It could be suggested that people already experience severe consequences of living with Usher and require support in times of need to manage day to day practical activities and also manage personal goals (Côté *et al,* 2013). However, this level of required support can only be provided if recognised and implemented by the government. It could be suggested that for any research to be successful there would be a need for co-working with service users/ expert by experience as co-researchers to develop a strategy to raise governmental awareness as to the complexities of living with Usher, with the aim of preventing incidences such as the 'job centre' experience. Participant experiences highlighted that people with Usher already put much time and energy into either

raising awareness of the condition, or supporting other people living with Usher, and have real life understanding, and therefore, their contribution to research would be invaluable. Whilst suitable strategies would need further research, thought and development, one way to raise awareness could be in the form of mandatory education for all professionals.

Although in this section increasing awareness of Usher amongst professionals has been considered, there is also significant support from sensory charities such as Sense and Deafblind UK. However, they are charities and as such would benefit from greater resources, to provide more provision of services.

5.5.3 Section summary

In this section the final theme was discussed outlining key messages, which were discussed in the context of literature and legislation. The fact that services varied from area to area were discussed and the need for equitable services was highlighted. Although literature discussing GP commissioning was discussed in relation to provision of services for people who experience deafblindness, it was also acknowledged that some GP practices may not have anyone who is deafblind registered in their practice, therefore their sensory knowledge may be limited. Various areas requiring the need for increased awareness amongst professionals were discussed. These related to educational support for students who experience Usher to ensure they are equipped to gain meaningful employment upon leaving education and employment and benefit agencies to consider their training practices, policies, procedures and protocols when working in partnership with people who experience Usher syndrome. Having discussed each of the four themes individually outlining key messages the next section will reveal three overarching messages.

5.6 Three overarching messages from findings

In my study three overarching messages were identified from critically evaluating literature in relation to my study findings, see Table 5.2.

Table 5.2: Three overarching messages from findings

The importance of ensuring communication is timely, supportive and appropriate
Usher support at the right time: Providing physical and virtual support networks
Essentiality of Usher awareness: Raising the profile

These overarching messages were revealed as a result of exploring the four themes from my findings (Table 4.1); identifying key messages from these themes (Table 4.4), and then summarising the information gathered to select three overarching messages (Table 5.2).

The first overarching message, The importance of ensuring communication is timely, supportive and appropriate related to ensuring that a diagnosis of Usher is communicated sensitively (see 5.2.2, 5.2.3), because how the news is communicated can have an impact on, not only how the news it taken by the person receiving it, but also the way in which living with Usher is dealt with in the future.

The second overarching message, Usher support at the right time: Providing physical and virtual support networks, highlighted, the need for specialist Usher support regardless of where someone lives (see 5.5.1), whether it is physical or virtual. This also included the benefits of technological advancement and how support and friendships can be maintained and developed (see 4.4.2, 5.4.3).

The third overarching message, Essentiality of Usher awareness: Raising the profile emphasised the need for people in general to be Usher aware to provide appropriate support, and this especially applied to professionals (see 5.5.2), to

ensure that people living with Usher on a day to day basis are able to better cope with the effects of the condition. The findings also highlighted the essentiality of support for not only the person diagnosed and living with Usher, but also their families, siblings, parent(s), partners, friends etc. However, the study found that participant experiences have revealed that there is still much work to be done and social change and development can only be achieved by raising awareness and appropriate support. Having revealed the three overarching messages, next the chapter will be summarised.

5.7 Chapter summary

In this chapter, each of the four themes was discussed individually outlining key messages. The discussion outlined where findings sat within literature, outlining how my findings supported literature, refuted it and compared and contrasted it. After discussion of each theme three overarching messages were identified (Table 5.2). Although lessons can be learned and principles applied from literature relating to other areas relating to disability (Chapman, 2002, Petersen, 2006), overall there is a lack of comparative literature directly relating to Usher syndrome. The discussion has highlighted how my study's findings have enlightened what life is like for people who are diagnosed with and are living with Usher syndrome.

Next, chapter 6 will conclude the thesis by considering: new knowledge and how my findings contribute to knowledge about the experiences of people living with Usher; conclusions drawn and key learning; benefits of reflection; strengths and limitations of the study; suggestions for further research; recommendations for practice and service development; recommendations for policy and education; intentions for dissemination of findings and conclusion.

Chapter 6 Conclusion

6.1 Introduction

Chapter 5 discussed the key messages (Table 4.4) raised from presentation of each of the four themes (Table 4.1), and critically discussed these in relation to the existing literature. From this discussion three overarching messages from findings (Table 5.2) were identified. This final chapter will start by discussing how my study's findings relating to the study topic contributed new knowledge about the experiences of people living with Usher. The chapter will then present the conclusions drawn and key learning; benefits of reflection; strengths and limitations of the study; suggestions for further research on the basis of my study; recommendations for practice and service development; recommendations for education; intentions for dissemination of findings; and conclusion.

6.2 New knowledge - How my findings contribute to knowledge about the experiences of people diagnosed and living with Usher

As highlighted in chapter 1 (1.1) and the literature review in chapter 2, there are few previous studies that have focused on people living with Usher syndrome. My study is the first descriptive phenomenological study to explore what life is like for adults across the lifespan, living with Usher in England. The study contributes new knowledge related to two areas: empirical findings and the research methods.

6.2.1 Originality of study findings

Different recruitment strategies were adopted to access the participants and this resulted in broad life experiences which enhanced the findings (3.4.2). Previous studies of Usher syndrome (Table 2.7) have focused on the practical and functional effects (Ellis and Hodges, 2013, Damen *et al,* 2005, Côté *et al,* 2013, Wahlqvist *et*

al, 2013, Högner, 2015).The areas of originality within the findings relate to five areas: experiences of diagnosis and living with Usher have improved due to changes in policy, practice and legislation (4.3.3, 4.5.4.5); regardless of age technology plays an important role in supporting a sense of belonging (4.3.2, 4.4.2); the importance of a feeling of belonging and being part of a community (4.4.1); that a diagnosis of Usher engendered vulnerability with fears about personal safety (4.2.3) and sensitivity is required when breaking bad news about a life altering condition such as Usher (4.2.2).

6.2.1.1 Experiences of diagnosis and living with Usher have improved due to changes in policy, practice and legislation

An area of new knowledge is that experiences of support for people diagnosed with Usher have improved from how it was many years ago, a contributory factor being the impact of legislation and guidance (1.5, 4.3.3). The wider age range enabled me, as a researcher, to see different perspectives and participant experiences at different points in time, for example in terms of: educational opportunities (4.5.1.1). With regards to educational opportunities interviewing people from 18-82 years highlighted differences with more recent experiences being more positive. The further back the participant experienced education, the less positive the experience, for example one participant who at the time of the interview was 73 years, was bullied at school by his peers for wearing sensory equipment and was given the cane by the teacher if he did not (4.5.1) . However, the participant who was 18 years at the time of the interview highlighted that she had reached her aspiration of going to university because of positive educational support (4.5.1).

6.2.1.2 Regardless of age, technology plays an important role in supporting a sense of belonging

Experiences around technology varied significantly between the different age groups. The findings revealed that although young people with Usher accessed social media to share experiences and meet other people with Usher (4.4.2) technological development also benefited the participant that was 82 years at the time of interview, as she mastered using FaceTime on an iPad to communicate with her family (4.4.2). Therefore, inclusion of a wide age range in this study contributed to new knowledge of how technology is used by people with Usher of different ages (4.4.2) and also that regardless of age technology plays an important role in supporting a sense of belonging (4.4.2).

6.2.1.3 The importance of a feeling of belonging and being a part of a community

Although there was only one participant who experienced difficulties communicating with her friends within the Deaf community after losing her sight, as BSL is a visual form of communication (4.4.1), this is still an important finding in the study because, it highlights the importance of feeling and belonging and being part of a community, whether it is the Deaf community or other community. The findings showed that relationships were affected because there was a lack of understanding from the participants Deaf friends with regards to Usher. As there does not appear to be any published research exploring the Deaf communities' understanding of Usher, or how Usher affects a feeling of belonging for the person experiencing Usher, this is therefore an area of new knowledge. It may be beneficial to conduct research with people from the Deaf community about their perceptions, awareness and experiences of maintaining networks and friendship groups with Deaf people who acquire blindness. Although, this study provides evidence of how the Deaf

community are unaware of Usher, as mentioned previously, this finding was only raised by one participant.

6.2.1.4 A diagnosis of Usher engendered vulnerability with fears about personal safety

Findings highlighted that a diagnosis of Usher engendered vulnerability with fears about personal safety, such as crossing roads, especially in light of hybrid and electric cars (4.2.3), and fear of house burglary (4.2.3). These personal fears and how best to address safety both inside and outside the home whilst ensuring people with Usher gain life experiences have been little raised in previous research about an Usher diagnosis and thus an area of new knowledge.

6.2.1.5 Sensitivity is required when breaking bad news about a life altering condition such as Usher

Another area of new knowledge related to conveying diagnosis or breaking bad news about a life altering condition such as Usher. The findings highlighted participants were told the news of their diagnosis in different ways, for example: through an interpreter (4.2.2) or in a shocking way that left a lasting impression (4.2.2). Literature relating to breaking bad news with regards to life threatening conditions such as cancer was highlighted (5.2.3) but there was no literature specifically focusing on Usher.

6.2.1.6 The meaning of the findings

So to summarise, new knowledge was produced because the findings offered individual experiences which enabled the development of an understanding of the experiences of, diagnosis of, and living with Usher syndrome, from the perspective

of adults living in England. Development of the findings initially identified four themes (Table 4.1): ‘diagnosis is the start of the experience’; ‘familial relationships across the lifespan’; ‘a sense of belonging’ and ‘experiences of professional support’. Consideration of the four themes and presentation of personal life experiences from the participants as highlighted in chapter 4 identified key messages from findings as presented in Table 4.4.

Key messages from the theme, ‘diagnosis is the start of the experience’ highlighted that although Usher is not life threatening, it is life altering because it affects practicalities of day to day living and future aspirations. Next, the key message relating to the diagnosis of Usher impacting on individuals in various ways was presented and highlighted that because every person experiencing Usher is unique and individual, the impact of the diagnosis will be unique and individual too. The final key message relating to the theme ‘diagnosis is the start of the experience’ highlighted that the way Usher is diagnosed affects individual experience.

The second theme, ‘familial relationships across the lifespan’ highlighted two key messages; the first was that a diagnosis of Usher impacts on new relationships and the management of existing ones. One aspect considered in this key message was the complexities sight and hearing impairment presented when building new relationships especially when socialising in low light environments such as nightclubs, cinemas and restaurants (4.3.4). The second key message that Usher is an unseen hereditary/genetic condition highlighted there are consequences not only for the person experiencing Usher, but their families as well, in terms of parental feelings of guilt when a child is diagnosed with Usher and living with Usher on a daily basis.

The third theme, ‘a sense of belonging’, identified three key messages the first being that acquired disability affects communication, the second, that Deaf/Deafblind culture can contribute to a sense of belonging and thirdly that technology can support a sense of belonging. The first key message highlighted that when a person experiences the combination of acquired sight and hearing loss, communication methods they may have previously used were affected, for example a profoundly Deaf person who communicated using the visual language of BSL. The second key message relates to the first because when communication is affected it can also affect the person with Usher’s, sense of belonging, for example as discussed in chapter 4 which related Debra’s experience of being part of the Deaf community (4.4.1).

The final key message relating to technology highlighted the importance of technology in relation to a sense of belonging because devices such as an IPad enables people to communicate within wider demographics resulting in broader sharing of Usher experiences and reduced isolation.

The fourth theme, ‘experiences of professional support’ revealed two key messages, that, specialist Usher support is necessary regardless of demographics and that there needs to be increased awareness of Usher amongst professionals and the general public.

After, key messages relating to participants experiences of living with Usher were outlined the study findings were critically evaluated in the context of previous literature, from this discussion three overarching messages were identified: the importance of ensuring communication is timely, supportive and appropriate; Usher support at the right time: Providing physical and virtual support networks and essentiality of Usher awareness: Raising the profile. The overarching messages were discovered as a consequence of exploring the four themes (Table 4.4) and

then summarising the information collected to select three overarching messages (Table 5.2). As highlighted previously Usher syndrome is a rare condition and as such, although the critical evaluation of literature outlined where findings sat within the literature this research has clearly identified original contributions to knowledge.

Having discussed areas of new knowledge located within the research, which demonstrated how the findings contributed to knowledge about the experiences of people living with Usher, next originality of the study in relation to methods will be discussed.

6.2.2 Originality of study in relation to methods

The methodology used for conducting this research was descriptive phenomenology (3.3.1). As identified from the literature review (2.3.4, Table 2.7), no previous studies have used this methodology for studying the experiences of people who are living with Usher. In a related field, only one qualitative phenomenological research study was identified (Sheppard and Badger, 2010) and this was with culturally Deaf adults, not people living with Usher syndrome. This methodology was chosen because descriptive phenomenology can contribute to our understanding of the experience of living with Usher by describing individual experiences without interpretation, clarification or construction (Heaslip *et al,* 2016, also see 3.3.1). The methodology was found to work well as a way of understanding the experiences of people living with Usher.

Additionally, theoretical underpinning selected for this study combined with the descriptive phenomenological methodology was also new, as there were no studies focusing on Usher, which combined the social model of disability (1.4.4, 5.3.2.1, 5.5.2), Erikson's identity development theory (1.4.6, 5.2.1, 5.3.1, 5.4.1), and

descriptive phenomenology to better understand how Usher affects adults in England diagnosed and living with Usher over their life span.

Originality was also demonstrated with regard to the narrative approach to the literature review which was adopted for this study (2.3). Although there is a peer reviewed article written in 2006 (Green *et al,* 2006) to guide this process, this work was expanded in chapter 2 to enable a structured comprehensive approach to conducting a narrative literature review and the 5s's model was produced to provide guidance (Table 2.2). There are plans for submitting a manuscript entitled 'Writing a narrative literature review: A novice researcher's guide'.

Another aspect of originality related to Multiple Sensory Communication and Interview Methods (MSCIM). Whilst other studies have been conducted with people whose first language were not the researcher's own (Temple and Young, 2004, Obasi, 2014), after conducting an extensive literature search it transpired that there was no published peer reviewed papers where participant led, individualised interview and communication methods had been utilised for conduct of research with people who had Usher syndrome or other sensory requirements/needs. The development of 'Multiple Sensory Communication and Interview Methods' (MSCIM) for use in the study is therefore a further area of originality. A paper entitled 'Empowering people with Usher syndrome as participants in research' (Appendix A) provided a critical review of MSCIM, which was published in the British Journal of Social Work in 2017.

The discussion relating to issues of power dynamics during data collection (3.4.2) highlighted there was no literature exploring issues of power within research which were Usher specific. Therefore my contribution to the discussion and publication of the paper enhances the thesis and contributes to originality of work relating to Usher

syndrome, especially in terms of raising awareness and contributing to understanding the lived experience of people with Usher, which is essential as this condition is rare, life altering but under-researched.

Having discussed the study's original contribution, next conclusions will be drawn and key learning will be considered.

6.3 Conclusions and key learning

When critically reflecting on the conclusions drawn and key learning, in relation to the commencement; progression; development and construction of this thesis, I found myself asking three questions: Has everything gone to plan? What conclusions have I drawn? And what have I learned?

6.3.1 Has everything gone to plan?

This thesis has in the main gone to plan. Although initially, recruitment was difficult as the condition was so rare and I needed to travel far and wide (3.4.1, Table 3.1) to interview and obtain data, the outcome resulted in rich data for which I feel privileged that people living with Usher shared with me. The aim of the study was to develop an understanding of the experiences of diagnosis of, and living with Usher syndrome, from the perspective of adults in England and by analysing experiences shared, and comparing and contrasting these with existing published literature, greater understanding of the condition has been gleaned (Chapter 5) and suggestions for further research identified (6.6).

6.3.2 What conclusions have I drawn?

As previously highlighted, the aim of the study is to develop an understanding of the experiences of diagnosis of and living with Usher syndrome, from the perspective of

adults in England, one of the key conclusions I have drawn is just how courageous people are when faced with adversity. I felt the people I interviewed were realists, they did not put a glossy sheen on having Usher, and they faced the challenges, whilst still looking to the future. Participants' experiences have been moving and thought provoking and have highlighted the extent of the time and energy that people with Usher put into supporting others with the same condition (5.5.2).

Bastow *et al* (2015) discuss how academics and their research make a difference as it adds to knowledge and makes a significant impact to the world of social science. This was of paramount concern for me, as I not only wanted to complete my PhD but I wanted it to make a difference within the sensory world. Although, literature (chapter 2, Table 2.7) evidences that Usher is a rare condition that needs greater attention, the conclusion I have drawn is that it is only by disseminating findings from my research that a real difference can be made. Completion of this thesis is just the beginning as, in order to do the research findings justice, raise awareness, and truly make a difference to people who live with Usher on a daily basis; more work is needed to disseminate this research via conference and publications.

6.3.3 What have I learned?

The thesis journey has resulted in an abundance of learning as a researcher, for example: challenges that can be experienced when trying to recruit which led to my commitment to travelling far and wide once someone did agree to be interviewed; how time consuming transcription can be, especially when translating in different languages; and the challenges of data analysis.

Also, from a personal perspective, I have learned that not only did my thinking as a scholar intensify, but also my writing style improved as writing the thesis progressed

and when writing for publication. Also, presenting my work at the doctoral support group, doctoral summer school, and attending conference skills workshops has taught me to become a more confident public speaker.

I have also learned the significance of using theory to underpin a study and the impact it had on my research study, for example Erikson's (1982) concept of identity development (1.4.6) and the social model of disability (1.4.4). As identified in chapter 4, Identity development is affected when people live with Usher because the debilitating degenerative condition affects transition from early to later life (chapter 4). For example I learned of the effects of a participant having to give up driving (4.3.4) or never being able to reach that aspiration in the first instance (4.2.2). Applying theory to research helped me understand and make sense of the experiences of what life is like for people living with Usher when transitional stages are affected.

With regards to the social model of disability (1.4.4) I learned that it is not about disabled people doing everything for themselves but rather having support to have control over their own lives. The findings supported this, for example the difference support in education (4.5.1.1), and sensory equipment and guide dog provision (4.5.3) made. It was evident that in order for people to be able to have this control, they need timely and appropriate support in terms of communication, physical and virtual support networks and Usher awareness (Table 5.2).

6.4 Benefits of reflection

Whilst as a social worker, I had always considered the practice of critical reflection to be essential to good practice; the experiences shared by participants reinforced this. A good example of the need for a researcher to be reflexive was demonstrated

in chapter 4 which highlighted my previous held understanding about a personal identity number being given to Deaf children upon school entry. Discussing this with the participant showed that even long held notions need to be reflected upon.
With regards to reflection, I learned the various range of reflexive research traditions (Finlay and Gough, 2003, see 3.7.5). Although, I knew reflexivity was important within social work practise I was unaware of the variety of approaches that could be used within reflexivity (3.7.5) and how to apply these within research practice too.

6.5 Strengths and limitations of study

One of the limitations of the study was in relation to the rarity of the condition. As indicated in chapter one, only 3-6% of people born *D/d*eaf will develop Usher syndrome (1.1) therefore when seeking participants to participate in the study the number of people were limited. As highlighted in section 3.4.2, the interviews took place over a period of 14 months because although interviewing began in March 2013, the next interview was not until July 2013 and the final interview took place in May 2014.

Paley (2005) posits that limited samples used in phenomenology "cannot be representative of any population at all" (p112). However, phenomenological studies are not aiming or claiming to be representative of populations but rather to explore the in-depth nature of the data collected which can lead to a depth of insights into individuals' experiences. The fact that this was a small scale study limited the experiences collated and subsequently the findings.

However, a strength of the study is that people who experienced all three Usher types (I, II, III) were recruited, together with both males and females, various nationalities and wide geographic locations (chapter 3, Table 3.1). Whilst this

outcome was welcome it was surprising as there were only 20 participants recruited. Nevertheless, a larger scale study may provide a more comprehensive account of what life is like for people with Usher in terms of their diagnosis, living with the condition and their experiences of Usher.

Whilst the participants were recruited from wide geographical locations (3.4.1, Table 3.1) if different participants had been included, in other locations the findings may have been different. The study did highlight that support services can vary across the country and including other parts of the country too, could contribute to further understanding of services that arise nationally depending on the person's postcode. This could be taken a step further by the researcher engaging in international collaboration to ascertain if life experiences for people with Usher are unique to England or transferable internationally. This again may depend on if the collaboration is within Europe (Damen *et al,* 2005) or worldwide (Hadidi and Kateeb, 2014).

In Ellis and Hodge's study (2.3.6.1), their primary participant recruitment source was RP fighting blindness and similarly in this study the primary recruitment source was Sense. This form of recruitment has limitations as where people belong to the same organisation this may mean that their responses and life experiences come from a particular perspective. However, within Sense there are two Usher groups or communities, the Hearing and Sight impaired group and the Sign Language Sense group (4.4.1), and as my study had participants from both groups, this did contribute to a difference and diversity within the participants recruited.

A variety of communication methods are needed to undertake research with participants experiencing Usher syndrome and so the researcher needs flexible communication skills, as explained in Chapter 3 (Table 3.3). However, on occasion

the sensory communication chosen by the participant was not the researcher's first language. Whilst as already indicated this led to a measure of equalising between the researcher and the participant, it also placed the researcher at a disadvantage as the researcher's first language was spoken English. Whilst the researcher had the skills to communicate in the participant's preferred way "native fluency" may not be attained in the use of sensory communication if it is not the person's first language (Temple and Young, 2004 p162).

A further limitation of the study may have been insider bias, for example the fact the researcher had been a sensory worker for a number of years (3.7.5); this was addressed as far as possible by promoting trustworthiness in research (3.7) and reflexivity (3.7.5). However, being an insider in the research was also a strength because when the participants were sharing their experiences about sensory support, equipment etc. they expressed positivity about the researcher having insider sensory knowledge/experience, especially when it came to equipment discussions.

Further strengths from the study were that a plethora of areas for future research were identified and these will be considered next.

6.6 Suggestions for further research on the basis of my study

Based on the findings within my study there were several areas where further research relating to Usher syndrome is needed. As highlighted in chapter 1, Usher is rare and as such under-researched. When I present the study at conferences, and I ask the audience, 'what is Usher syndrome'? they rarely know the answer. Whilst the general public have an awareness of deafblindness they are unaware of specific conditions or the impact these have on the life experiences of the individuals who

have them. Chapter 4 presented the findings and discussed four themes and their related sub-themes (Table 4.1). From presentation of these four themes, key messages from findings were identified (Table 4.4) and from these key messages, three overarching messages revealed (Table 5.2).

With regards to the first theme ‘Diagnosis is the start of the experience’, findings highlighted new knowledge that a diagnosis of Usher engendered vulnerability with fears about personal safety (4.2.3, 6.2.1.4). For example: as technology advances and the use of quiet electric cars increases, people with Usher syndrome will experience greater vulnerability and issues of personal safety. Thus this topic would benefit from further research to broaden knowledge relating to personal fears people with Usher experience and to ascertain ways to support them.

Also discussed in this theme were findings relating to delivering life altering news to people with Usher and communicating news of Usher via a third party: for example, an interpreter, this finding identified new knowledge that sensitivity is required when breaking bad news about a life altering condition such as Usher (4.2.2, 6.2.1.5) and thus another area requiring further research.

The findings relating to ‘Familial relationships across the lifespan’ found several areas for further research, the first was that, as all the data collated was from experiences of people who experienced Usher not from their families, further research is required with the families of people with Usher but who do not have it themselves.

Other findings showed that although some children struggled to talk to their parent(s) about having Usher (4.3.1), they did not appear to blame their parent(s) for

being carriers. This was not explored in detail in my study and may be an area for further research.

Associated with this struggle that some children had was the impact on parent(s) of Usher being hereditary/genetic. Although Ellis and Hodges (2013) discussed guilt and parental feelings of responsibility, as with my study, this was also from the grown up child's perspective not the parents (5.3.2). Participants in my study shared that it took time for a parent to accept their child had a diagnosis of Usher because as it is genetic/hereditary their parent(s) experienced feelings of guilt (4.3.1). As parent(s) were not interviewed as part of this study, this would be another area for further research to investigate the parents' perspective with a view to ensuring parents are supported when they experience their child being given a diagnosis of a genetic condition such as Usher syndrome.

Another area for further research is the impact for people living with Usher developing new relationships (4.3.4) because although the impact of the diagnosis on managing existing relationships was considered (Ellis and Hodges, 2013), the forming of new ones was not.

The third theme, 'A sense of belonging' presented two related sub-themes (Table 4.1). The key messages from this theme identified that further research was needed with regards to ensuring that appropriate communication is used when communication is affected by acquired disability (4.4.1), and how people living with Usher syndrome can be supported and encouraged to utilise technology and social media (4.4.2). The discussion also highlighted other areas for future research which included: perceptions of Deaf people about what life is like for people with Usher and, appropriate communication methods with people with Usher.

The final theme ‘Experiences of professional support’ presented findings relating to education (4.5.1.1), employment (4.5.1.2), benefits (4.5.1.2) and sensory social work (4.5.2.2), and highlighted that in all these areas greater sensory awareness and development is required to provide appropriate support for people diagnosed and living with Usher. As Usher is degenerative, there will be ever changing needs, therefore, further research is needed to ensure that professionals understand these needs and offer support which is tailormade to better meet individual needs, regardless of demographics.

Finally, the literature review identified that some research relating to visual impairment has been conducted in under-developed and developing countries (Hadidi and Kateeb, 2014), but there needs to be more studies about Usher conducted in developing countries.

This section has revealed there is a need for further research for people who experience Usher syndrome to meet their needs and better support them. However, Petersen (2006) posits that “patients (or service users) are an untapped resource of expertise in that they often know more about their particular condition and its management than their doctors” (p32). Petersen continues that as people increase their knowledge about their condition they “can become empowered” (p32) and ultimately have “greater control over their lives” (p33). “Therefore, further research with people with Usher could include them as co-researchers.

Next recommendations for practice and service development will be considered.

6.7 Recommendations for practice and service development

The findings (Chapter 4) highlighted that there are voids in appropriate and adequate services for people who experience Usher, supporting previous studies that have highlighted deficits (Chapter 5). Participants shared experiences in which they had encountered negative experiences in employment (4.5.1.2), support from services (4.5.2), education (4.5.1.1), and hospitals (4.2.2). These findings highlight the need for multi-agency working within local authorities, hospitals, specialist organisations, job centres, police and so on. Therefore although specialist sensory training is essential for social workers it is also vital for all professionals working with people with Usher and sensory needs to ensure they understand the full implications of Usher and meet their needs.

This thesis informs social work practice and could help people with Usher to get a better service in several ways: It will raise Usher awareness and outlines communication and interview methods needed to provide individual services and work collaboratively with people living with Usher (Appendix A); it will inform Sense of further research that needs to be conducted, for example, with families of people with Usher but who do not have it themselves, including parent(s) of children with Usher, to enable support to be provided. Within social work practice, social workers conduct assessments of need, and this research will inform social workers and enable them to be more Usher aware when conducting assessments. Finally, as sometimes families themselves are confused by the complexity of Usher and what services are available for them to access; these study findings will help social workers to understand the nature of Usher and the experience of living with Usher to better support families.

Also, although there is current disability legislation for people with Usher (1.5) such as the Health and Social Care Act 2014 and Local Authorities and Social Services Act 1970, when resources are cut, so are specialist support and services (1.5).

6.8 Recommendations for education

The findings highlighted a need for educational support for students who experience Usher to ensure they are equipped to gain meaningful employment upon leaving education (4.5.1.1, 4.2.1, 4.5.4), and also for employment (4.5.1.2, 4.5.4.1) and benefit agencies (4.4.5.2) to consider their training practises, policies, procedures and protocols when working in partnership with people who experience Usher syndrome.

The findings also identified that there was a need for sensory training for student social workers, nurses, occupational therapists etc. who may come into contact with people who experience Usher, and that this training should begin, not, when the student has qualified and already in the field, but when they are at university (4.5.4.2). Within universities, sensory training should be a standard part of the curriculum on social work and other healthcare courses, regardless of which area of practice a student is planning to embark upon. For example: social work and other students studying for healthcare related courses, would benefit from training in communication and collaborative practice to understand the importance of multi-agency working, including working with interpreters; understanding key definitions of sensory need about D/deaf, D/Deafblindness and visual impairment; awareness of how culture and diversity affects sensory impairment, for example when engaging a ‘BSL hands on interpreter’ for a female whose culture does not allow close contact with a person of the opposite gender.

Other areas would include appreciating the essentiality of critical reflection to ensure that services meet individual need including reflecting on if services provided were effective and what adjustments could be made to improve it (3.7.5) and learning about legislation that can be utilised to support people with sensory need (1.5), which is essential in a time of health and social care cuts.

6.9 Dissemination of findings

To date the findings have been disseminated at various conferences throughout the country and a peer reviewed journal article has been published by British Journal of Social Work, focusing on 'Empowering people with Usher syndrome as participants in research'. The purpose of the conferences have been twofold; to share the findings but also to raise awareness of Usher as the findings highlighted lack of awareness. Many people, including health and social care professionals do not know what Usher syndrome is and therefore conferences are an ideal opportunity to share the research findings and provide information about Usher syndrome and the specialist organisations (such as Sense) that will have further information.

I wrote a paper 'Empowering people experiencing Usher syndrome as participants in research' (Appendix A) which was written to develop an understanding that when communicating with people with Usher or indeed any sensory needs , professionals should adopt a person led approach that is specific and appropriate to that individual.

Future plans for dissemination of findings include, a training DVD based on the life experiences of people with Usher, with a suggested title of, 'Usher syndrome: seeing me, through my eyes'. The plan is to liaise with participants who contributed to this study, to involve them in the making of the DVD. Ethical considerations will be

taken into account. The purpose of the DVD is to raise awareness and enlighten professional support agencies such as social services, health, education, police, job centres, housing etc. what it is like for people to live with Usher on a day to day basis and enable support to be available from the point of diagnosis.

Further plans also include a journal article entitled, 'Descriptive phenomenology and social work: Considering compatibility between descriptive phenomenology as a methodology and the practical role of social work'. This study has demonstrated that the phenomenological attitude of bracketing, which is central to descriptive phenomenology is consistent with the practical role of the social work profession, where social workers are required to suspend their own belief's to practise in an anti-discriminatory manner and provide a person centred service to individuals' with whom they are working (Parker and Bradley, 2010).

Other future plans include an article entitled, 'Adapting Multiple Sensory Communication and Interview Method (MSCIM) as a tool in higher education'. Although MSCIM was originally a methodological innovation developed during research conducted with participants experiencing Usher syndrome for the current study, findings relating to the practical ways in which participants utilised technology to communicate could be used in a higher educational setting. One example is utilising Facetime or Skype on an initial practice learning meeting when a student is on placement in a remote area, or when supervising a student studying for their dissertation and the student or supervision is unable to meet face to face, perhaps because of demographics.

Other plans include an article entitled 'Does the meaning of participant led research vary? Comparison of using Multiple Sensory Communication and Interview Methods (MSCIM) in research compared to participants specifically leading on research'. Although this research study referred to participant led in the context of the

participant choosing the communication and interview methods, it could have referred to the participant's specifically leading the research thus highlighting the importance of clarification of what is meant by participant led research.

Finally, there are also plans to develop the literature review for an article entitled 'Is Usher syndrome under-researched? A literature review'. The literature review identified limited published scholarly literature directly relating to Usher syndrome because Usher syndrome is rare and under-researched and so, publishing the literature review would be a further way to raise awareness and also to learn from studies of other areas of sensory need such as visual impairment, Deafness etc.

Although some plans for future dissemination have been considered, it is anticipated that as this research has highlighted innovative ground breaking new knowledge, areas for further developments will arise in the future.

6.10 Conclusion

Chapter 1 introduced the thesis and provided an explanation for Usher syndrome and the background for the study. Chapter 2 reviewed literature relating to not only Usher syndrome but also other sensory impairments to provide a wider context of published literature. Chapter 3 explained the methods used to conduct the study. The fourth chapter presented the findings, the fifth discussed thesis findings in relation to existing published literature and identified three overarching messages (Table 5.2). This final chapter concluded the thesis by highlighting how my findings contributed to new knowledge, identifying what I had learned, the importance of reflection, the strengths and limitations of the study, implications for social work practice and recommendations for further research.

The aim of the study was to develop an understanding of the experiences of diagnosis of and living with Usher syndrome, from the perspective of adults in England. Three overarching messages were revealed (Table 5.2). The first overarching message: the importance of ensuring communication is timely, supportive and appropriate, included findings (4.2) and discussion (5.2.3) relating to ensuring that a diagnosis of Usher is communicated sensitively, as it can have an impact on, not only how the news it taken by the person receiving it, but also the way in which living with Usher is dealt with in the future.

The second message: Usher support at the right time: providing physical and virtual support networks, highlighted the need for specialist Usher support regardless of where someone lives (5.5.1), whether it is physical or virtual. This also included the benefits of technological advancement and how support and friendships can be maintained and developed (4.4.2, 5.4.3).

The final message: essentiality of Usher awareness: Raising the profile, emphasised the need for people in general to be Usher aware to provide appropriate support, and this especially applied to professionals (5.5.2), to ensure that people living with Usher on a day to day basis are able to better cope with the effects of the condition. The findings also highlighted the essentiality of support for not only the person diagnosed and living with Usher, but also their families, siblings, parent(s), partners, friends etc. However, the study found that participant experiences have revealed that there is still much work to be done and social change and development can only be achieved by raising awareness and appropriate support.

The IFSW (2014) defined social work as being a profession that promotes social change and development, taking into account difference and diversity. This study explored experiences from 20 people from a variety of backgrounds, nationalities,

ages, genders and locations and although the small sample is a limitation, common experiences amongst all participants demonstrated the enormity of living with Usher and challenges as a result of the condition.

Chapter one outlined two research questions (2.5.1): what is the experience of being diagnosed with Usher syndrome? And, what is life like for people who are diagnosed with, and live with Usher syndrome on a daily basis in England? The key conclusion from this research is that people who are diagnosed with, and experience Usher do not want everything done for them, but they do need increased support to be able to live with the life altering effects of Usher which affects the practicalities of day to day living, communication and identity development from early years to later life. As discussed in chapter 1, there are legislation and guidance (1.5) which can be used to support people with Usher and use of these have improved support for people with Usher (6.2.1.1), however, as highlighted this can be a bit of a 'post code lottery' (1.3, 5.5.1) so there needs to be increased awareness and specialist resources to enable the general public and professionals to provide support needed. Dissemination of the findings from this research has already (Appendix A), and will continue to contribute towards raising Usher awareness encapsulating the three overarching messages.

Conducting the research and writing the thesis has been the journey of a thousand miles that began with a single step (LaoTzu). Whilst as was considered in chapter 1 it could be argued that the thesis is the thousand mile journey with chapter 1 being the first step, chapters 2,3,4 and 5 being the journey in-between and chapter 6 being the final step, this thesis has demonstrated that there is ongoing and further research to be conducted so the journey needs to continue. The findings need to be disseminated and recommendations for future research followed up not only with

people with Usher but also other disabilities, thus the journey is ongoing and extends beyond the thousand miles.

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NOTES/END NOTES

Notes:

A) In the interest of anonymity participants have been given pseudo names

B) Dual sensory loss – the loss of two senses e.g. hearing and sight. This results in complexity of sensory loss and greater difficulties are experienced by the person. For example if a person loses their sight, they may have their hearing to rely on, but where a person loses both there is nothing to rely on.

End Notes:

1. Lip reading: Where a person who is Deaf/deaf reads the lips of another person to receive information conveyed.

2. Sensory equipment includes such items as mobile phone and Loop/ neck loop. The loop is a device which enables people with a hearing loss to obtain maximum involvement in communication in the environment in which the loop is fitted. The loop has a microphone, an amplifier and loop wire which is installed around the area in which the person sits. The receiver would usually be the person's hearing aid. The neck loop works on a the same principle but is a smaller personal device worn by the person around their neck.

3. Body worn hearing aid – Clips to clothes or in a pocket with a wire that leads to earphone or hearing aid located around the ear area. For people of older generations these may have been worn in a bag which was hung around the neck. These could be very visible and may have contributed to increased vulnerability.

4. Audiology – the study of hearing , balance and related hearing disorders. Often refers to a department in a hospital or clinic which provides hearing aids and other hearing related services

5. Ophthalmology – the study of sight related disorders. Often refers to a department in a hospital or clinic which provides sight related services for example macular degeneration (age related sight impairment, glaucoma (involves a group of diseases to the optic nerve plus a build-up of pressure in the eye that affects sight) or retinitis

pigmentosa (genetic eye disease, gradual deterioration of retina which may incorporate nightblindness and/or tunnel vision)

6. British Sign Language (BSL) – A visual means of communication using signs. BSL has its own grammatical structure and syntax and is recognised as an official language in its own right. Contrary to belief, BSL is regional and sign language varies.

7. Hands on (signing) – A communication method based upon British Sign Language enable the person to feel the signs. With this system, the deafblind person follows the signs by placing his/her hand over those of the signer and feeling the signs produces.

8. Finger spelling – the word is spelt out using either the BSL alphabet (individual signs used for letters of the alphabet) or manual alphabet (this is similar to the BSL alphabet but a more tactile approach where the letters are outlined on the palm of the deafblind persons hand. Fingerspelling is often used to spell out names, places etc.

9. Looked after children – Children looked after by the local authority under Children Act 1989. Sometimes it can be voluntary if parents are struggling to cope (S20) or if a child is at risk of significant harm (S47)

10. Kinship care – Care of child/children by a relative or close family friend.

11. National Deaf Children's Society (NDCS) – An organisation that specialises in supporting and campaigning on behalf of children who experience D/deafness. They also provide advice and a lending service for sensory equipment.

12. Grooming – Where an individual builds an emotional connection with a child to gain their trust for the purposes of sexual abuse or exploitation – Often carried out online/through the internet

13. Red and White cane – A red and white cane identifies the user as being deafblind

14. White cane – A white can identifies the user as being blind.

15. Capital ‘D’ Deafness – A person who considers themselves to be Deaf would most likely be a profoundly Deaf person who would not consider themselves to be disabled, but rather part of a minority group with its own culture, history and linguistics (British Deaf Association, 2007). This was highlighted by the British Deaf Association (BDA) when British Sign Language (BSL) attained official language status on 18th March 2003. Deaf people consider being Deaf as part of their heritage and most likely consider themselves proud to be Deaf (Evans & Whittaker 2010)

16. Small ‘d’ deafness – a deaf person is likely to have been previously a hearing person or one who was born deaf, growing up in a hearing family with oral communication being the primary means of communication. A person who has previously experienced the ability to hear music, voices and birds singing and then becomes profoundly deaf, for example as a result of meningococcal infection, can experience this as traumatic loss and be more likely to label themselves with a disability (Evans & Whittaker 2010)

17. Deafblind manual – The alphabet is similar to that of British Sign Language but is communicated directly onto the hand of the visually impaired/deafblind person. Each letter is placed over the top of the last. There is a slight pause in between words to indicate that a new word has started.

18. Facebook – Online social networking system which allows users to post in-depth posts including photographs.

19. FaceTime - A video-telephony product – A video chat application.

20. Twitter -Online social networking system that allows users to send and receive short posts

21. Skype – Enables a person to video chat with a person using the software application Skype

22 Email – Distribution of messages to one or more than one recipient via a computer or other computerised device such as a tablet or phone

23. IPad – A touch screen tablet PC. It is small and light and easily transportable.

24. IPhone – Is a combination of an IPod (music player), a tablet PC and a phone. Function is similar to an IPad but easier to transport as it is smaller and lighter. An internet enabled smart phone.

25. Kindle – A small hand held device/tablet to enable a person to read books electronically. Depending on the model the internet can also be accessed

26. Speech to text – The conversion of the spoken word to text. The speech to text translator will type verbatim on a PC device. The text is relayed to the person with the sensory needs. The person with the sensory need is able to read the text in a format this is suitable for their sight/hearing loss.

27. Bluetooth – a small device that enables short range wireless connection of mobile phones, PC devices and other electronic devices.

27. Video conferencing – Allows two or more locations to communicate simultaneous by way of a two way video and audio transmissions.

28. Sense - The national (UK) voluntary organization working with and supporting people of all ages who are deafblind or have associated disabilities - Includes information about the condition.

29. Talking Sense - Produced by Sense as above - Offers news, comments and features on the issues that matter to deafblind people, their families and the professionals who work

30. Deafbind UK - National charity providing support services to, and campaigning on behalf of, deafblind adults and their carers - Includes information about the condition.

31. Ear Foundation - Cochlear implants support charity, providing activities and support to families, carers and volunteers with deaf or hard of hearing children. Also provides speech therapy services.

32. Fighting Blindness - Fighting Blindness is an Irish patient led organisation that began in 1983 by a group of families affected by blindness to provide support for

each other. Today this organisation is involved in patient empowerment, advocacy, counselling and funding research into treatments and cures for blindness.

33. Sign Supported English (SSE) – Used to support the spoken work – Borrows signs from BSL and follows English grammatical structure.

34. Nordic definition of maintaining independence in deaf-blindness – in 1980 Nordic countries agreed to a common definition of domains of independence that are adversely affected in deafblindness. These relate to ability to give and receive information (communication) and access to information and mobility

35. Congenitally deafblind – Blind from birth. Either genetic/hereditary or due to the environment

36. Cohen's k or Cohens kappa is a statistic which measures inter-rater reliability (level of concurring between raters) for qualitative items. It is thought this way of measuring is more reliable as takes into account aspects that happen by chance.

37. CAUSE – A project designed at promoting Usher awareness and CHARGE (coloboma, heart defect atresia choanae, retarded growth and development, genital abnormality and ear abnormality) association and was supported by the European Usher Syndrome Network and the CHARGE Network, together with project partner organisations CRESAM and Retina France (France), Fighting Blindness (Ireland), ONCE (Spain) Lega del Filo d'Ora (Italy) Sense (UK) and Blinden instituts sitiftung (Germany)

38. SPSS - A software package used to analyse quantitative data.

39. Charmaz (2006) stage approach to data analysis - (a) several readings of the data to obtain an holistic perspective; (b) initial themes identified, clustered and checked against data, research literature and the first author's observations; (c) a working hypothesis induced from the themes, and (d) constant comparison between data, observations, and literature to test and refi ne the hypothesis.

40. Cochlear implants - A cochlear implant is an electronic medical device that replaces the function of the damaged inner ear. Unlike hearing aids, which make

sounds louder, cochlear implants do the work of damaged parts of the inner ear (cochlea) to provide sound signals to the brain.

41. Communicator guide – A trained person who offers support to people who have developed sight and hearing difficulties to support them with travel to the supermarket to do their weekly shop; Reading letters; Helping to set up a medical appointment and accompanying them to the hospital; Helping an individual to attend a social group where they can meet others with acquired deafblindness (Sense 2014a)

42. Deaf studies trust (DST) - A national charity set up for the benefit of the Deaf community in the UK. It aims to apply research-based knowledge to practical issues for Deaf children and adults. It works also with hard of hearing adults.

43. Voice over – a narration technique used during visual forms of communication e.g. visual frame BSL. A hearing person uses clear speech to translate BSL.

44. Induction loops – Transmits sound directly into hearing aid via a magnetic field. Background noise is reduced to enhance clarity of sound.

45. Huntingdon's disease (HD) - A genetic/hereditary degenerative condition with affects the central nervous system in the brain and which affects behaviour, moods, movement, thinking and judgement (HD Disease Association 2016). As with Usher syndrome, a person many not realise they have the condition until it manifests itself later in life.

46. Cystic Fibrosis (CF) - A genetic/hereditary condition in which the internal organs, especially the lungs and digestive system become clogged with a sticky mucus substance which results in infection and inflammation of the lungs and severe difficulties digesting food. As with Usher, parents often don't know they carry the gene; one in 25 people carry the faulty CF gene without even knowing. If two carriers give birth to a baby, there is a one in four chance that the baby will have CF (CF Association 2016).

47. Alzheimer's is genetic/hereditary - Affects memory thinking and behaviour (Alzheimer's Association 2016). If there are family members who already have the condition, other family member may be aware they may have the condition. However, as it manifests itself later in life, often people will not know they have the condition.

48. Municipal network in India – Local government body to ensure provision and development of primary schools, clean water, lighting in public streets and to oversee and develop public hospitals etc. Networks comprise of Mayor and councillors.

APPENDICES

Appendix A: Article published by British Journal of Social Work:

'Empowering people experiencing Usher syndrome as participants in research'

Abstract

Engaging people from marginalised groups such as the deafblind and Usher communities to participate in research has historically proved challenging, mainly due to communication differences between participants and researcher. Therefore an approach called 'Multiple Sensory Communication and Interview Methods' (MSCIM) was developed and used when conducting research with people who are deafblind and have Usher syndrome. This article considers the value of using MSCIM by critiquing the data collection and interview methods used by the author in a qualitative research study with twenty participants aged 18-82 who experience Usher syndrome. Communication and interview methods were participant led with communication methods including: Clear speech, visual frame British Sign Language (BSL), hands on BSL, deafblind manual and written communication. Participants were given the choice to be interviewed face to face, over the telephone, via Skype (video/no video) or email. Whilst this approach was natural in the researcher's role as a sensory social worker, within the study this approach led to a measure of unexpected equalising between the researched and the researcher and explored how empowering individuals from marginalised groups as active participants in research contributes to inclusivity and promotes trustworthiness in research.

Keywords: Usher syndrome, Deafblind, Empowerment, Inclusivity, Sensory Social Work, Qualitative Research, Trustworthiness, Sensory-Needs,' Multiple Sensory Communication and Interview Methods' (MSCIM)

Introduction

The report 'I fear for my future' (Sense 2014a) highlighted that deafblind people are becoming marginalised as a result of becoming increasingly isolated and excluded within society. Marginalisation is defined as "making or treating as insignificant" (Thompson 1996: 609). Within this deafblind group, Usher syndrome is one of the main causes of acquired deafblindness in the United Kingdom (UK). However, surprisingly very little attention has been paid to it (Ellis and Hodges 2013), especially when people who experience deafblindness/Usher syndrome can experience marginalisation due to challenges faced with communication, living independently and safe orientation (Bodworth *et al* 2011, Sense 2014a). As well as encountering extraordinary challenges on a daily basis people with Usher syndrome receive inadequate support, especially in relation to their communication needs (Sense 2014a). More research is therefore essential to provide greater understanding into this group's experiences, and improve support and services to meet their needs (Wylie *et al* 2013). However, people with Usher syndrome remain under-represented in research studies (Sense 2014a, Ellis and Hodges 2013) and so it is important to consider how to increase their participation in research.

Usher syndrome is a rare inherited condition and approximately 3-6% of people born *D/d*eaf will develop it (Genetic Alliance UK 2012). Capital 'D' Deafness refers to people who are culturally Deaf. The person is likely be profoundly Deaf and would not consider their sensory difference to be a disability. They would consider themselves part of a minority group with its own history, culture and linguistics and be proud of this. In 2003 British Sign Language attained official language status.

Whereas a small ‘d’ deaf person is likely to be previously hearing or if profoundly deaf born into family where primary communication is oral/voice. This person is more likely to consider themselves disabled.

Usher syndrome is primarily characterized by deafness caused by an impairment of the auditory nerve which prevents sensory transmission to the brain (sensorineural hearing loss) accompanied by retinitis pigmentosa (RP - retinal cell degeneration), which can lead to progressive loss of central and peripheral vision. There are three types of Usher syndrome, Types I, II and III. People experiencing type 1 are usually born profoundly d/Deaf and start to experience reduced vision in early years possibly the first decade. Children sometimes experience problems with their balance and have difficulty walking (Moller *et al* 2009). People with type II generally experience reduced hearing but are not born d/Deaf. They have no obvious balance difficulties. Visual difficulties start in the second decade or as they get older with sight and hearing loss varying from person to person (Sadeghi *et al* 2004). The person with type III may not realise they have Usher syndrome until later in life as they initially have no obvious hearing, sight or balance difficulties. Losses occur gradually. (Pennings *et al* 2003). The Usher type experienced by a person can influence their language and communication preferences, for example people who experience types II and III would most likely communicate via speech, whereas a person experiencing type I may use British Sign Language (BSL-Table 1). There is no way of predicting the prognosis of Usher syndrome for an individual, as the condition is unpredictable. A person's sight could deteriorate very quickly or they may still see to read when over 50 years (Sense 2014a). Whilst it is a condition in which people experience sensory impairment, it is an invisible condition as there are no visible outward signs (Sense 2014a). Although the term sensory impairment could refer to loss of one or more senses e.g. sight, hearing, touch, taste etc. for the purpose of this article sensory refers to the loss of sight, hearing or both.

Whilst research has been conducted with people with sensory impairment (Percival *et al* 2006, Bolt 2005) few studies have been undertaken with people with Usher syndrome (Ellis and Hodges 2013, Damen *et al* 2005, Côté *et al* 2013, Wahlqvist *et al* 2013, Högner 2015) and only a few, included participants from the UK (Damen *et al* 2005, Kyle and Barnett 2012, Ellis and Hodges 2013).

People who experience deafblindness are noted to be “seen not only as a vulnerable group” (Simcock 2016:1) but as "some of the most vulnerable members of society” (Simcock and Manthorpe 2014:2325). However, this is not the case for all people with sensory needs/requirements some of whom may not consider themselves to be disabled or vulnerable but rather as members of a linguistic/cultural minority group (Ladd 2003, 2008).

However, the case of Beverley Lewis is an example of vulnerability due to inter alia, deafblindness which led to extremely severe consequences as recorded by Simcock and Manthorpe (2014). Beverley Lewis was a young black deafblind woman who died in 1989 aged just 23 years and weighing under 4 stone; she was found wrapped in newspaper in squalid conditions. Her mother was known to have mental health issues and repeatedly refused services and support. On the day that Beverley Lewis died, her mother was admitted to hospital and she was later diagnosed with schizophrenia. This case highlights that it is essential for people, who experience deafblindness to have a voice and be heard as otherwise serious consequences could occur.

Whilst further research is essential in order to enhance understanding of the experiences of people who have Usher syndrome, their communication differences could affect their participation in research. The aim of this article is to critically

review an approach to communication that was used during research interviews in a study that explored the life experiences of people who have Usher syndrome. The approach entitled 'Multiple Sensory Interview and Communication Methods' (MSCIM), aimed to foster research engagement through participant led communication and interview methods. The article will also consider the social model of disability in relation to the research conducted and the epistemological, methodological and ethical issues that may arise in translation and interpretation of interviews that use different communication methods. The article will consider how empowering individuals from marginalised groups to be active participants in research can contribute to inclusivity and promote trustworthiness in research. Finally, the potential for applying MSCIM in a wider context will be considered.

Background to the research

The researcher's interest in the flexible use of communication and interview methods developed during data collection for a phenomenological study of adults with Usher syndrome as part of a PhD research study that was approved by a university research ethics committee. All participants gave written or verbal informed consent for the interviews. The aim of the study is to develop an understanding of the experiences of diagnosis of and living with Usher syndrome, from the perspective of adults in the UK.

The researcher had worked previously as a sensory social worker whose communication and interview methods in practice were service user led and therefore it was natural to approach sensory research in the same way. However, during the conducting of twenty individualised interviews, it became apparent that by utilising the participants' chosen communication methods a measure of equalising between participants and the researcher took place. This was unanticipated as developing disability research that contributes to empowering marginalised groups,

rather than being oppressive, can be a challenge (Oliver 1999). In most studies the researcher is in a position of power because they decide which questions will be asked, how data will be interpreted and how results should be presented (Caretta and Riaño 2016). However, this paper considers how using MSCIM during data collection can contribute to a measure of equalising within the research relationship.

Communication and interview methods used to conduct interviews

The various communication methods used in MSCIM and a brief description of each of these will first be provided, followed by a summary of the combinations of communication/interview methods and the number of sessions chosen by the participants. The social model of disability will offer the theoretical underpinning as this model proffers that people are not disabled by their individual impairments but by the disabling barriers they face in society when appropriate adjustments to meet their needs are not applied (Oliver 1983, 1990, Bricher 2000, Kitchin 2000, Bolt 2005, Oliver 2013).

Application of the social model of disability in research is crucial, but in order to challenge the power-loaded relationships, a dialogue or communication in which there is a "collaborative process of knowledge production" (Caretta and Riaño 2016:2) is fundamental. An exchanging of knowledge has the capacity to "create an inclusive space", which can contribute to empowering the participant and lead to a "more balanced relationship between power and knowledge" (Caretta and Riaño 2016:2).

The consideration of participation in research from a social model perspective highlights that the communicative choice is essential for the reduction of communication barriers for people who experience disability, Usher syndrome, deafblindness, or other sensory needs. Oliver posits that there is a "firm distinction

between the researcher and the researched" (Oliver 1992:102), based upon the "belief that it is the researchers who have specialist knowledge and skills" (Oliver 1992:102). However, whilst the researcher for the current study had specialist knowledge and skills both in sensory communication and as a researcher, the balance of power was altered by the very fact that the communication and interview method was participant led. As a result, the researcher's first language or preferred interview method was not always the chosen option. Thus issues of inequality within the research relationship although not eliminated, were reduced. Within research relationships, the extent to which research methods disable participants may vary but MSCIM was found to be crucial in this study as people with sensory needs are all different and use different types of communication. Table one shows the variety of communication methods chosen by participants and used by the researcher during the conduct of the twenty interviews.

Table 1: Communication methods used in the interviews

Clear speech	Use of clear speech: voice not too loud or too soft and good articulation. Light and environment are utilised to enhance sound and quality of speech.
Written communication	Communication by use of the written word using a range of formats e.g. large or extra-large print.
Visual frame signing - British Sign Language (BSL)	BSL signing within the individual visual field of the person with Usher syndrome (who has restricted sight) to enable utilisation of residual vision. (Sense 2014b)
Hands on signing BSL	Based upon BSL - enables the person to feel the signs. With this system, the deafblind person follows the signs by placing his/her hands over those of the signer and feels the signs produced.
Deafblind manual alphabet	A method of spelling out words directly onto the sight impaired/severely sight impaired person's hand. Each letter is denoted by a particular sign or place on the hand (Sense 2014b).

It was interesting to note the link between choices participants made. For example where the participant chose visual/tactile methods of communication e.g. visual frame BSL, hands on signing BSL and deafblind manual, their preferred interview

choice was face to face, whereas participants who chose a clear speech method of communication primarily chose telephone or email interview methods.

Whilst an individual may be described as being deafblind, levels of sight and hearing loss can vary, therefore communication methods chosen by a person who experiences Usher syndrome/deafblindness will differ too (Sense 2014a). Whilst it could be argued the most appropriate way to interview a person with a sensory requirement may be a visual communication method for example BSL, Table 2 illustrates the variety of preferred communication methods chosen by the participants which were individualised by the researcher to undertake the interviews.

Table 2: Communication and interview methods chosen by the participants and number of sessions required to complete interviews

Participant	Interview method	Communication or combination of communication methods used	Number of sessions
P1	Face to Face/Email	Clear speech, visual frame BSL, written communication	3
P2	Face to Face	Clear speech	1
P3	Email	Written communication	3
P4	Face to Face	Visual frame BSL, Hands on BSL, Deafblind Manual	1
P5	Face to Face	Visual frame BSL, Written communication	1
P6	Skype no video	Clear speech	1
P7	Telephone	Clear speech	1
P8	Email	Written communication	2
P9	Email	Written communication	2
P10	Skype with video	Clear speech	1
P11	Telephone	Clear speech	1
P12	Telephone	Clear speech	1
P13	Face to Face	Clear speech	1
P14	Email	Written communication	2
P15	Email	Written communication	1
P16	Telephone/Email	Clear speech, written communication	2
P17	Telephone	Clear speech	1
P18	Telephone	Clear speech	1
P19	Telephone	Clear speech	1
P20	Email	Written communication	1

There were differences in the number of sessions that were used for each participant's interview; 14 interviews were conducted in one session whilst 6 required more than one. The number of sessions conducted reflected the participant's chosen interview method; where more than one session was conducted, it was due to email being used. In most cases interviews conducted via telephone, face to face, or Skype (video/no video) were completed in one session. One participant chose Skype with the video off, explaining that this was fairer; the participant could not see the researcher due to the sight loss and the researcher could not see the participant as there was no video. This participant's choice led to some equalising between the researched and the researcher, which at the time of the interview was not anticipated. The other participant who selected Skype chose to have the video on, expressing that they did not mind either way. Whilst Deakin and Wakefield (2014:603) posit that "face to face interviews remain prominent" they acknowledge that "innovative communication technologies such as Skype have facilitated new modes of communication". Two participants initially chose email interview and then decided to change their interview method to that of face to face. Having explained the communication and interview methods used to conduct interviews in the current study, these data collection methods used will be critiqued next.

Critique of data collection methods used

Some participants were initially hesitant about being interviewed as they had previous negative experiences, for example within employment, education or acquiring benefits. Participant 7, when sharing an experience of a Job Centre interview, commented:

> *"I was forced to go to the job centre my brother had to take half a day off. I went there with a red and white stick [red and white cane indicates a person is deafblind, as opposed to a white cane which indicates visual impairment]*

and personal listener wires, I used headphones. She still said to me at the end there 'could you please look at the monitor and check what I have written'? I started waving my stick around and said 'I can't see the monitor let alone the text'. She said 'we still think you should apply for work'. I don't need all this sorry, I put letters in front of her from [specialist eye hospital] hospital and from the [Ear Nose and Throat hospital] and then she just ignored it all".

This participants experience highlights that awareness of Usher syndrome needs raising, so that professionals, society and the general public understand the implications of the condition. However, in addition, the experience confirms the importance of using MSCIM when conducting interviews in a variety of contexts, such as health, education, policing and social care as well as when conducting research. Acknowledging and addressing individual communication needs may empower and encourage participants to engage in research and to be more willing to share personal experiences.

Whilst it is essential to take communicative and sensory needs into consideration during research, each method has strengths and limitations. For example, when using telephone interviewing it is not possible to observe body language and incidental gestures, which may be important in the interview process (Kvale 2013). However, telephone interviews could empower participants as the researcher cannot see their reactions and body language and only hears what the interviewee wants them to. The use of Skype may be advantageous for the participant as that the location is flexible; geographically the participant could be anywhere (Deakin and Wakefield 2014).However the research process may be hindered by poor reception or transmission resulting in interruption of the participant's or the researcher's train of thought. Another disadvantage with using Skype could be that sensitive issues may be in the process of being discussed and suddenly the

transmission stops. It is apparent that whichever method is used there are always strengths and limitations.

Empowerment in the research process and its contribution to inclusivity

As previously highlighted, participant empowerment and some equalising occurred when the video facility on Skype was turned off at the participant's request. There were also other situations where participant empowerment and equalising of power occurred, for example, during interviews where the participants chosen communication method was not the researcher's first language.

Whilst the participants may have felt dis-empowered by the nature of their impairment or because they were subjects of research, the researcher was dis-empowered by not always knowing which communication or interview method the participant would choose and being the one to use their second language, not their first language. For example the researcher was expecting one participant (P4) to communicate using visual frame BSL but as the interview progressed, deafblind manual and hands on signing were also used. The researcher was disadvantaged when the participants chose to communicate using visual frame BSL, Hands on BSL or Deafblind Manual. Whilst the researcher has acquired sensory communication skills, as a hearing person whose first language was spoken English, it was not possible to be as fluent or as efficient as a person for whom visual frame BSL, Hands on BSL or Deafblind Manual is their first language (Temple and Young 2004, Obasi 2014). However, whilst not the case for the researcher, for some hearing people, BSL may be their first language, for example, a hearing child who is raised by one or more parents/guardians or carers who are Deaf (Children of Deaf Adults: CODA) may be bilingual and achieve native fluency (Hoffmeister 2008, Orlansky and Bonvillian 1985).

The other empowering factor was related to the participants' choice of geographical dimension of place or location for the interview. Although the "geographical dimension of place can fundamentally affect the nature of knowledge accessible through a range of methodological techniques" (Anderson *et al* 2010:590), the participants could choose a space where they felt comfortable to share their experiences, which were often of a sensitive nature. Anderson *et al* (2010:596) further suggested that the geographical dimension of place should be given an explicit role in the method in order to help "harness the agency of place in the methodological encounter". Therefore whilst the use of MSCIM positively contributed to a measure of equalisation within the research process, conceivably so does the participant's choice of geographical location.

Promoting inclusivity and trustworthiness in research

Within social work, the principle of inclusivity not only contributes to best practice but also supports the collection of unique data from people who may not otherwise have participated (Bellinger and Elliott 2011). It is essential to recognise that the capacity and resilience of people who use services contributes to inclusivity and is core not only to the social work profession but also to research carried out with service users. Cossar and Neil (2015) highlighted that social work as a profession encourages respect for service users and promotion of their rights/values; this core principle should arguably be reflected in social work research too. Furthermore, Bellinger and Elliott (2011) consider the practicalities of managing inclusive research while not compromising reliability by maintaining the connection between "good practice and how the research was conducted" (Bellinger and Elliott 2011:710). The use of MSCIM within the study supported inclusivity but also, promoted trustworthiness of the research. The concept of trustworthiness in qualitative research has attracted much scrutiny and debate (Shenton 2004, Patton 2002, Finlay 2006, Guba and Lincoln 1985).

One of the biggest challenges in qualitative research is how to assure the quality and trustworthiness of the research (Finlay 2006). Trustworthiness includes rigour, believability and plausibility of the research and can be described as a belief in the truth or reliability of something or someone (Koch 2006, Anney 2014). Believability relates to an acceptance of what is presented to be true, while plausibility concerns the reasonableness or probability of the research (Koch 2006, Anney 2014). Promoting trustworthiness is an important aspect of this research study; it may be difficult for people with Usher syndrome to share their experiences with a researcher and so the data needs to be trustworthy so that the research can benefit others. Participant 13 commented:-

> *"It's like this survey. It is not going to help me directly, it's of no benefit to me now and it's no benefit to me to talk about this, it does not help me at all because I really don't like talking about it. I have talked about it many many times and it does not help me. I am only doing this for other people".*

This comment led the researcher to critically reflect on the importance of the research being trustworthy, so that the participant's views were conveyed in a credible manner. In order to share their personal experiences, the participants needed to feel confident that their experiences would be taken at face value, rather than being translated or interpreted (Giorgi 2009). For example, where participants chose a tactile form of communication e.g. visual frame BSL/hands on BSL/deafblind manual, the interview was both audio (with voice-over) and video recorded. Voice over occurs, where a hearing person interprets, using clear speech, the comments made by the person using the visual form of communication. Voice over was possible as the participant chose to have someone of their choice who was hearing and who was able to communicate using clear speech and a combination of visual forms of communication. This approach was essential to the

study because BSL has its own grammatical structure and a hearing researcher, whose first language is English, could be “oblivious” to their own limitations in translation and also “some of the signed nuances may be lost in translation” (Obasi 2014:73). With reference to her own study Obasi (2014) described how one sign could have had a number of representations in English, for example, a sign in which the hand is clawed and “circulated in the lower stomach area” (Obasi 2014:73) could have indicated anxiety, apprehension or fear. The inclusion of voice over during the interview reduced misinterpretation in translation, thus increasing the trustworthiness of the data. Having access to both audio and visual recordings of the interview enabled the researcher to cross reference and ensure the interview content was truthful during the process of translation.

Epistemological, methodological and ethical issues in translation and interpretation

Phenomenological research is considered to have strong epistemological foundations because experiences are the source of knowledge (Dowling 2007, Racher and Robinson 2003). The use of the social model of disability as a framework for this study sought to address epistemological concerns relating to the research relationship, power balance, oppression and autonomy, with communication and interview methods being participant led. Another factor that was considered in the research conduct was the provision of appropriate support to enable participants to share their knowledge and experiences. Whereas, Damen *et al* (2005) described provision of support for people with Usher syndrome to complete questionnaires in their survey, in the current study participants’ choice to utilise voice over was used as a support mechanism.

The methodologically oriented researcher will question how to avoid inaccuracies within their research (Kvale 2013). In this current study, the researcher needed to

consider methodological issues that arose in translation and interpretation of data. However, Kvale (2013) questions whether we can ever get to know what the subjects really mean. The researcher chose to use descriptive phenomenology (Giorgi 2012) as in this methodology experiences expressed by participants are accepted as a given rather than being open to interpretation. Whilst it could be argued all experiences are open to interpretation, taking participant comments at face value will minimize the possibility of interpretation occurring (Giorgi 2012). Also utilised in the interviews was the concept of back translation (Edwards 1998, Temple and Young 2004) to agree with the participant what has been said was correct. Kvale (2013) suggested that ethical considerations go further than the actual interview situation and are rooted in the interview inquiry in its entirety. He further highlighted that "ethical issues permeate research" and the interviewer needs to be able to "create a stage where the subject is free and safe to talk of private events" (Kvale 2013:8). Whilst Kvale's (2013) views could apply to any research using interviews, in sensory research interviews it is particularly important for participants to feel comfortable with translation and interpretation of their experiences. In the current study, voiceover was utilised when the participant's first language was BSL, but voiceover could also be used to support anyone whose first language is not English or to enable a person to maximize residual hearing. Ethical issues can arise where data are collected using different forms of communication methods or different languages and the research involves translation and interpretation (Temple and Young 2004). To avoid misrepresentation in translation and interpretation, communication and interview methods should be individualised with a significant degree of flexibility. Also as Temple and Young (2004) noted, disseminated work should clearly outline that a variety of communication methods/languages have been utilised rather than presenting the data as having been collected in the language of the researcher.

An additional ethical issue that could have arisen in the study related to personal intrusion (Stalker 1998) or intrusion of privacy. To prevent any feelings of intrusion only data that the participants were willing to share was translated, interpreted and transcribed. For example one participant talked off camera about personal familial Usher experiences but was unwilling for these comments to be used within the research, and this request was respected.

Whilst qualitative research is fundamental to understanding participant experience (Kvale 2013), as highlighted briefly above, there are epistemological, methodological and ethical issues that need considering. Researchers can attempt to be as accurate as possible with regard to interpretation and translation but the very fact that alternative languages are being used may affect complete precision. The next aspects to be considered are applicability and consistency.

Applicability and Consistency

Applicability refers to the findings from the research and whether they would be applicable, relevant or appropriate to other studies. Whilst this study specifically focused on people living with Usher syndrome, findings that related to areas such as the need for greater awareness being raised and the essentiality of family support could be applicable in other areas of sensory research or disability, such as experiences of people with visual impairment, D/deafness (Young *et al* 2008) or physical impairment. A further component could be the applicability of the data collection methods used as MSCIM could be applicable to other research studies, such as with people whose first language is not English. Gaining high quality data through using the participants' preferred methods of communication and interviewing could contribute to the richness of the data quality and achieve greater accuracy.

This study's *consistency* was demonstrated through the detailed descriptions of how the data collection was conducted, thus enabling other researchers to understand the methods used. Additionally, the use of MSCIM could positively affect consistency. The researcher's sensory experience from a professional perspective could have influenced the findings and therefore it was important to maintain neutrality. This was achieved through using descriptive phenomenology (Giorgi 2009), which ensured that findings arose from participants' own experiences. Additionally the researcher's reflexivity contributed to the promotion of neutrality. Berger (2015) explains reflexivity as being a constant internal discussion and critical self-evaluation of the researcher's position within the research. As a reflexive researcher there was a constant questioning of self to ensure that prior professional experience did not impact on the current study. However, there are also some advantages to the researcher having prior knowledge and experience which may encourage participants to be more willing to share experiences, as they may perceive the researcher to be more understanding and sympathetic to their situation. In the current study with people who are living with Usher syndrome participants, enquired if the researcher had knowledge of sensory equipment or issues and when details of professional sensory experience were provided participants gave positive responses:

> *"Oh you have a lot of experience then?"(Participant 17)*
>
> "*I use a neck loop. Do you know what a personal listener is? Aah so you know"*
>
> *(Participant 7)*

The researcher's professional sensory knowledge also contributed to consistency and flow of the interview as participants did not have to stop and explain what they meant when describing sensory equipment or relaying any other sensory experiences or issues.

Future potential of using MSCIM

Whilst this article has focused on MSCIM in relation to research with participants experiencing Usher syndrome, consideration should be given for its use in other research areas. MSCIM can apply across a variety of settings that require flexibility, adaptability and the use of multiple methods. As highlighted earlier, MSCIM could be beneficial where research participants experience other areas of sensory need/disability (visual impairment/D/deafness), or, physical disabilities, or when the participants first language is not English. Whilst the researcher undertaking the current study had sensory communication skills, other researchers may speak more than one language, which could be useful. Additionally for some languages, there are different dialects/regional speech/local standards patterns associated with a single language and that may require the researcher to be flexible and adaptable throughout the interview process.

Sadler *et al* (2010) highlighted that research recruitment of people in hard to reach populations "might be harder to reach because of a variety of personal or sociodemographic characteristics" (Sadler *et al* 2010:369). The use of MSCIM within the interview process may enable research to be conducted with people who are hard to reach due to personal or sociodemographic characteristics such as prisons or remote geographical areas. In addition, interviews could take place where the researcher and the researched are in different countries thus opening up international research opportunities without the expense that such research would usually incur, such as travel and accommodation costs etc. The use of Skype with the video off or email could allow a participant to remain unseen and maintain a degree of control and anonymity if that was their choice. Moreover, in some areas, internet access is not available thus alternative interview methods may be necessary, for example conducting the interview by written communication e.g. letter.

There is also the potential for practical application in a range of other interview settings such as policing, job centres or health environments. Within police interviews, where the person with the sensory need could be the victim or the perpetrator, the use of MSCIM could be advantageous as individualised approaches being used could reduce time and lead to more positive outcomes. Where the person with sensory needs is a victim, the interview method may be more person-centered and sensitively carried out if using an MSCIM approach. For example, where a person has been the victim of a physical/sexual assault, the interview being conducted from a person-led perspective with the victim choosing the location, interview method and communication method, could be empowering and influence the quality and quantity of the information that the person is willing to share. As highlighted earlier, with the experience of participant 7, other sectors, such as job centres may benefit from using MSCIM as an approach. Within health environments the use of the MSCIM model could be advantageous, for example, if a person is asked to share their medical history or other background information as part of their assessment. The use of MSCIM may encourage the person to feel a little more relaxed in what could be a very stressful situation, as they are using methods that they feel comfortable with rather than trying to fit into the interview and communication methods of the healthcare professional who is asking the questions. These examples highlight that MSCIM is flexible, adaptable and participant led, and has the potential to be used in any environment that requires interviews to be conducted using a variety of communication methods. Furthermore, MSCIM contributes to participants in research being empowered and thus they may be more willing to engage in the research/interview process.

Limitations of MSCIM

When using MSCIM in research there are undoubtedly limitations to be considered. Firstly the researcher needs to have a range of communication skills at their

fingertips and the ability to switch between communication methods/languages smoothly and flexibly. In the current study, as the participant's choice of communication method was not always identified until the interview began, the ability to switch between methods of communication/language was essential. The second limitation related to the challenges of translation and interpreting the results generated from using a range of communication methods. This issue was discussed in the section, 'promotion of inclusivity and trustworthiness in research' and highlighted that the use of audio/visual recordings and voice over enabled translation to be cross referenced thus ensuring the interview content was correct.

Conclusion

This article has contributed to the limited published literature relating to the lived experiences of people who experience Usher syndrome by discussing how 'Multiple Sensory Communication and Interview Methods' (MSCIM) were used to promote inclusivity and empower people with Usher syndrome during their participation in research. The issues considered were raised due to the experiences shared by people who have Usher syndrome and who contributed to the current research. The article also considered promotion of inclusivity and trustworthiness in research and epistemological, methodological and ethical issues that may arise in translation and interpretation, due to the conducting of research in a language different to the researcher's own. As a result of the communication and interview method being participant led, the researcher found that participant empowerment and some equalisation of power in the research relationship with participants occurred. As highlighted earlier, participants contributing to disability research may experience inequality due to the fact that they experience disability (Oliver 1992). However, in the current study, the interview/communication method and location for the interview being participant led, contributed to the power dynamics shifting more towards the participant than the researcher, a situation that is unusual when conducting research

with vulnerable, marginalised groups. It would be a positive step towards reducing marginalisation and empowering people with Usher syndrome if Usher awareness was raised and people with Usher syndrome, and those with all sensory requirements, were listened to using their preferred communication and interview methods with MSCIM offered not as a privilege but as a right.

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Appendix B: Interview questions

London South Bank University

Project Title: 'Exploration of the Lived Experiences of People with Usher Syndrome'

The questions are topics for use with follow up probes to explore responses

- What do you report about your knowledge and understanding of your diagnosis/prognosis (of Usher Syndrome)?

- How do you report your experiences of support e.g. parent/carer/social work etc. (with regard to Usher Syndrome)?

- What was your experience of your transition from adolescence to adulthood (with regard to Usher Syndrome)?

- What are your experiences of your developmental opportunities in education, work, socially at home and in relation to peers/siblings/ICT (with regard to Usher Syndrome)?

- What is your experience of the Deaf community?

Thank you again for taking the time to be a part of this project.

Michelle

Appendix C: Codes across transcripts

1) ‘Usher syndrome: A phenomenological study of adults across the lifespan living in England’
2) Data analysis method: Braun and Clarke’s (2006,2013) Six phase guide to performing thematic analysis
3) Descriptive phenomenological approach to capture codes which as theorised by Giorgi takes ‘the participant experiences as a given’ (Giorgi 2012)

Appendix C: Codes across transcripts

Codes	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	Occurrences
Impact of Diagnosis																					1
Shock of diagnosis			X		X	X				X				X		X			X		11
Sadness kicked in																				X	1
People institutionalised for having Usher																X					1
Relief that person had not got something much more serious																X					1
Relief as from young age was treated for anxiety and other mental health issues																X					1
So pleased it was recognised and there person was pleased they wasn't alone in this matter																	X				1
Person honestly felt like someone had made a mistake																				X	1
When person first finding out had Usher Syndrome they had a break down														X							1
Realistic attitude upon diagnosis e.g. vision won't change overnight – coping mechanism			X																		2
Realistic attitude upon diagnosis e.g. practical, financial and level of support required			X			X	X	X	X	X	X	X		X							13
Immediate acceptance of condition, just accepted it												X									3
Didn't make any difference sight and hearing problems now had a name																	X				1
Not a shock, had lived with it most of life getting used to it																	X				1
Didn't freak person out. It all made sense. Life made sense for first time																X					1
Impact of losing sight as well		X		X	X	X	X	X		X		X	X	X		X			X	X	29

Impact of diagnosis		X			X										X			X	X	5
Really messed up and not self at all		X				X									X					6
Living and re-living impact of diagnosis		X				X	X											X		12
Awareness of limitations upon diagnosis			X			X	X				X							X		9
Usher compounds things – harder for barriers and obstacles to overcome															X					1
Diagnosis leads to person becoming depressed					X	X	X													9
Impact of diagnosis - do everything in a hurry before it's too late.						X					X									7
Family overprotective as a result of diagnosis						X														1
Life altering effects							X			X									X	4
Affects people in different ways										X			X							2
The way the person was diagnosed was shocking - 1. First told nothing wrong with eyes 2.Then told there was 3.Eye doctor googled condition 4. Told person they are likely to go blind																			X	1
Post Diagnosis																				
Better experience of ophthalmology consultations after diagnosis																			X	1
Role of……																				
Role of optician in diagnosis		X	X					X			X						X	X		14
Role of eye specialist in diagnosis (also in provision of motability car/DLA etc)		X	X	X	X	X	X	X	X	X	X	X	X		X	X	X	X	X	X	28
Role of audiologist																	X			X	2
The role of the interpreter - Being told emotional news via third party					X																2
Role of professional in career choice adapting to sensory need				X																	1
Research – For solutions																					
Discussion with eye specialist around what research is currently being executed to find solutions			X																		1
Hope for future research - RP magazine 'moving' forward talks about gene therapy							X														1
Hope for future research - Stem cell							X														1

– early stages of research																						
Hope for future research- retinal implants –bionic eye make you see							X															1
Person with usher overloaded with questionnaires asking questions; same old, same old													X									1
In-depth person search about Usher Syndrome – Information is power																X						2
Placental Implants																						
Person had placental implants				X																		1
Diagnosis methods																						
Blood test carried out to diagnose Usher							X															1
Noticing something is wrong																						
Clumsy in the dark		X							X											X		4
Clumsy – noticing something was wrong		X											X						X	X		7
Knocking things over																			X			1
Loss of confidence even before diagnosis as sight affects daily living									X	X												2
Family noticing something is wrong			X	X					X	X									X	X		6
Optician noticing something is wrong			X	X	X						X											4
Person noticing something is wrong			X	X					X	X		X							X			10
School noticing something is wrong											X										X	2
Falling over and walking into things initial indication something wrong		X		X					X	X			X						X	X		8
Poor night vision (nightblindness)		X	X	X	X	X	X	X	X	X	X		X	X	X	X	X	X	X	X		31
Dark colours harder to see																			X			1
Not being able to find own way down a dark street																	X					1
Problems seeing															X		X					2
Losing sight of things when they go out of line of vision											X	X					X	X				4
Better vision in the morning, sunlight etc					X			X														3
Person thought everyone the same poor sight and vision				X						X		X				X						12
Not understanding how much vision had been lost																X						1
Not realising sight problems until problems occur e.g. car accident										X	X											4
Psychological aspect to Usher						X	X					X	X									8
Usher is a psychological blindness as																X						2

well as a physical one																					
Psychological impact on child when parent is diagnosed with usher																X					1
Suicidal thoughts as a result of dual sensory loss.							X					X				X					3
Accidents occurring e.g. not seeing kerb; being knocked down by a car etc											X	X		X		X		X			6
Person not knowing they had Usher, assumed they were deafblind												X					X				4
Noticing something wrong even though not diagnosed													X				X				2
Person always knew they had a problem with sight and hearing but didn't have a name for it																	X				1
Guide Dog Provision																					
Increasing blindness does affect every-day functioning but can be counteracted with for example equipment and guide dog		X									X					X		X			7
Sight improvement after cataracts operation guide dog no longer needed											X										1
Impact of guide dog provision		X									X					X		X		X	5
Support from Guide Dogs fantastic																				X	1
Guide dog contributes to feelings of safety and calmness		X														X		X			3
Positive impact on psychological well being																X					1
Guide dog makes person with usher feel more in contact with human race e.g. other people more ready to communicate																X					1
Safe mobility and companionship																X					1
Guide dog provision makes a big difference																		X			1
Early Diagnosis																					
Undiagnosed Usher can lead to person being treated for mental health issues																X					1
Missed aspirations and dreams when usher is undiagnosed and unsupported																X					1

Routine Usher check													X								1
Person may not have welcomed early diagnosis because they did not want to be drawn attention to																	X				1
Person considered Usher present from birth even though not diagnosed till later																	X				1
Speech therapy to maximise speech e.g. elocution lessons																X		X			2
Check all children for Usher before they start school whatever age they come into school system													X								1
Need for support – counselling services not waiting for person to become more depressed		X			X											X			X		4
Early diagnosis may result in preventing a person living a full life.										X							X		X		6
Managing to travel alone, done it all their life so not different as sight deteriorated																	X				1
Check all children in family if one child is diagnosed										X	X										4
Early diagnosis making people more aware of what their limitations are										X									X		2
Early diagnosis essential to prepare for sensory needs		X		X					X		X		X						X		16
Greater caution after diagnosis																			X		2
Prognosis – usher is a degenerative condition but person **may not** go totally blind					X																1
Prognosis – usher is a degenerative condition but person **MAY** totally blind																X					1
Retinitis pigmentosa (the sight loss part of Usher) can lead to little or no vision – blind					X																3
Progressive degenerative sight condition				X	X	X	X	X	X	X	X	X			X	X	X			X	27
Prognosis unpredictable		X				X	X			X	X	X		X							9
Uncertain future with regard to level of sight loss		X								X											2
When child/adult is diagnosed automatic transfer of records to						X							X								3

social services																					
Automatic transfer of records from medical profession to social services enables services to be put in place						X															1
Person broke so many glasses and crockery and was terrified of being shouted at																X					1
Diagnosis often in teens																X					1
Hearing checks carried out on children of parent with usher																X					1
Optician saying there is nothing wrong																		X		X	2
Eye doctor saying there is nothing wrong with eyes																				X	1
Identity Development																					
Interruption of identity development						X	X						X								13
Nightblindness getting worse in developmental years															X						1
Sensory specialist careers advice to consider changing needs						X			X												8
Person not wanting to be labelled as blind										X											1
Person has experienced all sorts of fantastic things despite being deafblind																				X	1
Noisy music and darkness means person with Usher can't cope in these situations at all so couldn't socialise											X										1
Relationship forming impacted by Usher											X	X		X	X	X					7
Staying in mainstream school allowed person to develop socially similarly to able bodied peers																				X	1
Usher affects professional and personal life															X						1
Person kept sight and hearing problem as quiet as possible – disability not encouraged																	X				1
Can't socialise in the evening											X							X			2
Person can't go out at night because they can't see																		X			1
Problems socialising																X					1

Socialising difficult due to usher. Other people noticing something is wrong – Coming across as disabled at night/dark environments															X						1
Usher limits social circle throughout life							X				X	X	X	X				X			7
Deafness and blindness is socially and personally disabling													X								2
Plans, goals and aspirations dependent on condition/health													X								2
People keep their distance															X						1
No friends																X					1
lack of lighting impacts usher															X						1
labelled as clumsy																X					3
Classed as a clumsy child																		X			1
Accused of being drunk or on drugs																X					2
Anxiety wasn't viewed as due to the fact field of vision was deteriorating																X					1
Shouted at so much for being clumsy person became afraid and fearful in many situations																X					1
Person coping techniques to fit in																X	X				3
Person develops coping strategies to cope with usher																				X	1
Coping techniques to cover up deafblindness in adolescent years/Interruption of identity development							X					X					X				4
Humour adopted as coping technique																X	X	X			3
Parenting																					
People with sensory need seen as not being capable of looking after child				X														X			3
Social work experience not positive but not connected to sensory rather parenting.																	X				3
Lack of appropriate support if parent shows lack of interest/support													X								1
Expectations of disabled people & Derogatory Responses																					
Lower education expectations due to disability		X									X								X		2
Lower expectations towards disabled				X		X					X								X		9

people																					
Patronising attitudes experienced							X									X			X		3
Treated badly by other people		X			X	X		X		X	X	X	X			X	X	X			14
Treated badly by family					X												X				6
Treated badly by school teacher																		X			1
Feeling sad at bad treatment received by other people that people																	X				1
Lack of respect shown		X					X						X								9
People with usher seen as a target/vulnerable		X			X			X			X										4
Vulnerable and taken advantage of		X				X		X													3
Physical abuse experienced		X			X																5
Other peoples responses						X	X					X	X	X					X		11
As usher is an unseen disability it impacts others peoples responses													X	X							3
A lot of tongue biting when people make derogatory comments																			X		1
Annoyance when people are surprised that a person with sensory needs has a job		X																	X		1
Victim of discrimination						X															1
Friendships and Shared Experiences																					
Friendships developed reaching out to other people with the same condition						X	X	X	X	X			X			X		X	X		16
Shared experiences – all being in the same boat – sense of belonging		X				X		X	X	X	X					X		X			14
Share experiences how to cope with Usher, learn from each other										X	X					X		X			8
Facebook and social networking sites contribute to networking and meeting like-minded people		X	X	X							X								X		9
Not liking or forming friendships with every person just because they have usher		X																	X		2
Diagnosis of usher does not affect all relationships		X																	X		2
Diagnosis of usher DOES affect relationships				X							X		X								6
Skype, e mail etc contribute to enhancement of communication for people with sensory needs					X																2

Usher affects developing friendships – No friend's																X					1
Communication																					
Changing sensory needs impact communication		X		X	X	X								X	X	X	X	X	X		16
Deteriorating sight affects communication e.g. ability to lipread																		X			1
Inappropriate communication e.g. talking to people about personal issues in hospital corridor																	X				1
Don't assume - some people they hear the word blind and they see a guide dog and hear the word deaf they see sign language																	X				1
Teach children in schools about Usher – including communication skills such as hands on signing and deafblind manual									X		X	X	X								3
Preferred communication essential to establish and maintain relationships		X		X	X									X					X		6
Accent and dialect affect ability to lipread and understanding of spoken word						X															3
Improvement of lighting in pubs etc to improve communication and reduce isolation								X													1
Family members cannot make out what the person is saying, need other family member to explain														X							1
Taking information by looking at face, gestures to support speech										X											1
Speech improves with age														X							1
Individualised communication needed																	X				1
Big 'D' Deafness & Communication																					
Deaf people see things in a different way		X																			1
Deaf people keep themselves in their own groups							X							X							2
Being part of Deaf community seen as segregation not integration																	X				1

Deaf community are in a world to their own										X											1
Deaf culture seen as negative as it stigmatises people – segregation rather than integration																	X				1
No experience of Deaf community													X								1
Not part of Deaf community																				X	1
Most people with hearing problems do not sign																	X				1
Person does not accept the term Deaf community the only community is the normal UK community																	X				3
Personal Identity number given to Deaf child upon school entry – Used instead of name				X																	4
Personal educational identity number still used even after leaving school				X																	4
Positive attitude to using personal educational identity number.				X																	2
Personal education identity number as means of identification				X															X		4
Being part of Deaf community linked with ability to sign		X		X		X	X	X		X			X	X			X				9
Sign language is out because person can't see																		X			1
Don't assume because someone is deaf they use sign language																	X				1
Not all deaf people with usher see themselves in the deaf community and not necessarily all use sign language																	X				1
All Deaf. Same culture same, happy together				X																	3
Deaf world/hearing world communication difference		X	X																X		1
Deaf children can be sent to boarding school, when home at weekends or holidays difficult to form friendships – boarding school friendships are like family		X		X							X										5
Deaf- school -boarding school				X	X																3
Impact of Deaf-school-boarding school				X																	1

Deaf children sent to boarding school – young age				X							X					X					3
School: protected environment. Person unprepared for 'big' world		X																			2
Fun memories of boarding school		X																			1
Change in eyesight changes relationship with Deaf friends		X																			1
Sign language is a learnt skill some people are scared by		X																			1
Sign language not easy for everyone to pick up/learn		X																			1
Hands on signing hard to pick up/learn for some people		X																			1
Desire to use voice as opposed to sign language				X																	1
Limitations on natural use of sign language as a means of communication.				X																	1
Sign language first language learnt at school (Non English)					X																1
Even after diagnosis of Usher culturally Deaf people see themselves as still culturally Deaf with history and culture				X																	1
Support for people whose first language is BSL to know and understand what Usher is and how to get support					X																2
Tried to fit into hearing world						X		X													3
Deaf school – sign language not used – oral method used						X															1
Not wanting to be part of the Deaf community						X															1
Deaf and hearing united by shared experience e.g. Usher						X															1
Hearing aid users (deaf) and Culturally Deaf peoples (BSL first language) communication part needs to be separate e.g. HIS/BDA						X															1
Not knowing Deaf community existed													X								1
Education																					
Parents being heartbroken they have to send child to boarding school																X					1

Parent(s) not happy child may be sent to boarding school for Deaf											X										1
College/school support required to provide sensory equipment to meet educational need			X								X	X					X				8
Reasonable educational adjustment to meet sensory need			X				X	X		X	X	X				X	X	X			19
Educational support not provided to support individual sensory need													X			X		X			4
Positive attitude to going back to college		X																			1
Not losing out on education side							X														1
Support from school incredible																				X	1
Over reliance on educational support by person who has usher																				X	1
Amazing support workers who contributed to survival in mainstream education environment																				X	1
Difference support makes - Despite losing vision in short time period significant educational achievement																				X	1
Essentiality of school support e.g. teachers/TA's/Support workers to support person with usher to achieve educational aspirations																				X	1
Child comes home from school crying because they can't see – not supported by school.																		X			1
Person went to mainstream school, sat in front of class and got by																	X				2
Blind children sent to residential schools only time with family holiday time.											X										2
Vision of local educational provision for children with sensory need											X										1
Children with sensory need being placed with physically disabled children											X										1
Children with sensory need integrated in mainstream education											X					X	X				3
Sensory need impacts academic achievement											X		X	X				X			6

Children with sensory need separated from mainstream peers for Maths and English											X			X							1
Getting the cane for not wearing sensory equipment – Did not wear because being bullied for wearing it by other non-sensory impaired children												X									1
Bullied for wearing sensory equipment e.g. body worn hearing aid																X					1
Not using sensory equipment after being bullied e.g. body worn hearing aid.																X					1
Treated very differently at mainstream school																X					1
Being scared in school situation – impacted by sensory need																X					1
Audiology & Ophthalmology																					
No usher department/hospital to address combined sensory loss													X			X	X	X		X	5
Attendance at the same hospital but under different consultants for hearing and sight										X											1
Person thinks audiology and ophthalmology would be better joined up										X								X			2
Dual sensory clinic best way forward										X								X			2
Department to specifically address Usher as a condition in its own right													X								1
Joined up working to ensure holistic sensory support												X				X		X			3
Need for joined up working as you get blinder you get deafer																		X			1
Combining audiology and ophthalmology would weaken their expertise and funding in terms of eyes									X										X		2
Person does not think dual sensory clinic required as both audiology an																				X	1

ophthalmology manage their own specialism well																					
Communication between departments ophthalmology/audiology		X		X			X	X				X									7
Joined audiology/ophthalmology appointments/ specialist audiology/ophthalmology worker/coordinator		X				X	X	X	X				X								12
deafness and vision treated separately and coped with separately		X				X	X		X		X								X		9
Raise awareness GP's Hospitals, Audiology, Ophthalmology etc		X					X														2
More support and advise from Audiology and Ophthalmology																			X		1
Support from Audiology and Ophthalmology fantastic																				X	1
Provide support when person with usher asks for it																			X		1
Diagnosed at dual sensory loss clinic							X														1
Provide automatic testing of hearing where sight loss diagnosed and vice versa							X														1
Not questioning the professional e.g. had condition through not eating enough carrots															X						1
Audiology provide regular and consistent support to person																				X	1
Validity of Condition																					
Validation of usher by professionals, organisations, family and friends etc. contributes to person with usher considering reality of condition					X		X												X		2
Participant questioning validity of condition							X														1
Professionals paying more attention to one sensory loss as opposed to other e.g. more to sight that hearing loss													X								1
Benefits & Employment																					
Easier access to services and benefits			X								X					X			X		6
Recognition of condition without			X			X	X				X	X							X		21

having to fight for everything																					
Indignity of benefit system						X	X														5
Person with usher not comfortable asking for help but sensory need warrants it,																X					1
Fear stigma of condition will stop person from working (from employers perspective)									X		X										2
Some colleagues not as understanding and helpful as others														X							1
Education not good because of sensory need, person worried they wouldn't get a job														X							1
Equality of employment opportunity – Provide access to work at point of diagnoses			X								X		X						X		4
Employer not seeing usher as a problem person themselves see it as a problem										X											1
What does the impact of usher mean for work?										X											4
Deafblindness impacting efforts to do a thorough job															X						1
Employers very good, supportive										X											4
Some colleagues not very sympathetic										X											1
Type of work becomes dangerous to continue										X											1
Suitable travelling service for deafblind to travel to work									X	X				X							3
Ushers raises difficulties travelling to work and dealing with crowds														X							1
Employment move to different department as means of reasonable adjustment			X																X		2
Physical impact when appropriate sensory equipment not provided in workplace																			X		1
Equality of opportunity – not just talking about it but really making it happen – everyone is different so make sure everyone starts from the same point		X	X								X	X							X		5

Individualised support						X				X		X			X				X		7
Adapting employment to further meet needs of self and others											X										4
Medically retired prior to retirement age						X	X			X	X	X		X							10
Upset to be medically retired but understood why												X									1
Proper protection and support when bullying harassment occurs											X	X				X					6
Concerns over government making criteria too tight leading to lack of support/less services											X										2
Googling Diagnosis																					
Impact of googling diagnosis																			X		1
Reactions to being given hope																					
Reaction to being given hope		X							X				X						X		5
Slowly bringing of self-back to being a bit more with it and enjoying life																			X		1
Bereavement (Kubler-Ross 1970)																					
Bereavement and grieving for sensory loss		X									X								X		7
Bereavement – Loss of independence		X							X	X						X			X		8
Bereavement - Loss - giving up driving						X	X	X		X						X					7
Bereavement – Loss of giving up something the person loves because sensory loss no longer allow it																X					1
Loss leads impact mental health and wellbeing																X					1
Bereavement – Loss – removal of transport choice/autonomy e.g. bicycle							X		X	X											4
Denial at point of diagnosis														X					X	X	5
Depression upon learning diagnosis		X				X				X		X							X		5
It takes time to *accept* the big changes the usher diagnosis presents		X							X					X			X				5
Desire to get out there/*acceptance*		X				X				X											5
As person gets older they slowly learn to adapt to Ushers (acceptance)														X							1
Anger upon learning of diagnosis						X								X							3

Angry with parents														X						X	2
Coping with anger						X															1
Anger at loss of vision						X															1
Bargaining										X											1
Feelings, Attitudes (Positive & Negative) & Emotions																					
Partaking in project in the hope other people will not go through what person went through													X								1
Helping other people with condition despite feeling no benefit would be received by person themselves													X								1
Self-perceived as different, not ordinary and normal													X								1
Feeling hopeless		X																			1
Difficult choices to make e.g. cochlear implant										X											6
Difficult choices e.g. cochlear implant versus better quality hearing aids										X											1
Why has it happened to me?										X											1
Changes brought upon by usher make person feel strange										X											1
Usher – A horrible dark secret							X														1
Keeping sensory need a secret e.g. not telling husband even though deaf since birth																X					1
Desire to reach full potential		X	X								X										5
Sensory loss impacts aspirations E.g. doing jobs for the sake of the money															X						1
Resilience amongst adversity		X	X			X		X	X	X			X			X		X			16
Being in a deep place mentally and physically			X				X														2
Feelings of things being normal until diagnosis										X									X		3
Realisation of complexity of dual sensory loss			X	X		X		X	X												7
Desire to live a 'normal' life			X	X			X			X	X				X						9
Happy memories before Usher			X							X											2
Sensory loss leads to isolation			X		X		X	X	X			X	X			X					14
Active approach on individuals part to reduce isolation			X					X	X	X	X										7
Individual disadvantage as opposed			X							X	X										6

to individual disability																					
Individual personality impacts condition			X							X		X	X			X		X			11
Positive approach to sensory loss			X							X	X				X						4
Getting on with daily living and life			X	X	X		X			X	X	X			X	X	X				12
Carried on with daily life			X	X						X	X	X					X		X		12
Appreciation for the things they do have												X									1
Desire to retain independence		X	X	X			X			X						X				X	10
Feeling frustrated by loss of sight				X						X											6
Feel need to hide frustration from people				X																	1
Experiencing feeling in a tight spot by not knowing exact usher type							X														1
Worrying for families care e.g. not being able to drive or recognise if there is a fault with the car										X											1
Not being able to drive anymore										X											1
Never being able to drive																				X	1
Never being able to drive, walk anywhere on own or read a book again - Makes things different																				X	1
Avoidance of using the word benefit e.g. use of word scheme/program to describe DLA										X											1
Coping with additional needs e.g. dyspraxia and thinking these are part of the condition of usher															X	X					2
Usher and additional needs																		X			1
Never met anyone with usher before diagnosis																	X				1
Misconceptions																					
Misconceptions: Persons Father was told his daughter's usher originally came from VD																		X			1
Family																					
Genetic condition		X		X				X		X		X		X	X	X			X	X	22
Hereditary condition		X		X				X		X	X	X		X	X		X		X		22
More than one child in the family has usher									X		X			X				X			5
Person, only one in family with usher																				X	1
Younger siblings pick up older sibling needs support																				X	1

Grandparent and grandchild has usher																		X			1
Potentially misses a generation															X			X			2
If one parent does not have the gene then children will not have usher															X						1
Parents with usher not wanting their children to become their carers																X					1
Children whose parents have usher not wanting to become the carer																X					1
Children miss out by parent with usher not driving																X					2
Condition hard to cope with if person has no emotional or physical support from family																X					1
Other family member also deafblind – Having a lot in common														X							1
Parents being carriers															X						1
Parental guilt as condition is hereditary						X													X	X	3
Provision of parental support to enable parent to support child		X				X					X					X			X		4
Provision of parental support to enable parent to support child whatever their age						X					X										2
Provision of support to spouses, siblings, partners and children of parents with Usher						X				X						X					5
Extended family support-raise awareness and enable people outside of immediate family to offer support.																			X		1
Essentiality of family support and understanding		X	X	X	X	X		X	X	X			X	X		X	X	X	X	X	38
Positive attitude to deafness encouraged by family			X					X		X											5
Positive family values/culture – resilience			X			X				X				X							
Lack of interest of condition shown within family													X			X					3
Asking for help but not getting it – Family/Professionals																X					1
Family member nagging to get expertise advise as walking into																			X		1

things																					
Not everyone in the family will have usher		X		X			X				X	X	X	X							10
Siblings seen by person with usher as carriers'		X																			1
Order in which family and friends are told about the diagnosis																			X		1
Limited access to support e.g. family used where participant previously mentioned family member as being perpetrator of physical assault					X																1
Power assigned to family member when used as interpreter					X																2
Shock to family					X	X					X										4
Parents worse dilemma to learn their child is going to have a disability						X															1
Parent not wanting to accept child has disability												X					X				2
Parents find it really hard to cope with when their child is diagnosed with usher																				X	1
Stigma attached to disability family not wanting to accept person has problems																	X				1
Family coping techniques to adapt to condition						X				X	X	X				X	X				10
Person creating own methods to make their life as normal as possible															X		X				2
Not being treated as different because of disability, just being one of the family							X				X			X							3
Parents role of choosing right time to tell child he/she has usher											X										3
Parent's not telling child about diagnosis/prognosis of Usher, Childs view "you must let your child know"											X										1
Change in family attitude upon learning diagnosis e.g. people being kinder											X										1
Person treated differently from how there were treated prior to diagnosis											X										2
Child draws own conclusions if not											X										3

advised they have Usher by parents/caregivers																					
Person draws own conclusions if not advised they have Usher																X					1
Parent holding hand of adult offspring with usher and taking them around											X										1
Over reliance on parent(s) by person who has usher																				X	1
Wrapped up in cotton wool, knowing other children are treated differently from you											X										1
How do children feel when they are treated differently?											X										1
Parents not letting children/young people out of the house unaccompanied											X										1
Teach children with sensory needs life skills to meet their need as parents may not teach these													X								1
Parent can't even speak to partner that child has Usher Syndrome.											X										1
Impact of parent(s)/Caregiver hiding condition from child											X					X					2
What happens to people with Usher if anything happens to their carer/family?											X										1
Child becomes upset on hearing parent has usher as they have 'made fun of them' – didn't realise parent had condition																X					1
Children may need counselling when the realize their parent (s) have usher																X					2
Impact on children when parent is diagnosed with usher																X					1
Impact on child with usher when sibling has a more demanding disability																		X			1
Hard for parents to support child and child to support parent when they are struggling with diagnosis individually																				X	1
Child finds it hard to express difficulties encountered with having usher as they do not want to upset																				X	1

their parents																					
Friends, peers, neighbours and colleagues																					
Essentiality of support from friends peers, neighbours and colleagues			X								X			X							3
Friends, peers and colleagues see the person as different						X	X														2
Specialist Organisation Support																			X		2
Individual choice e.g. not all people with Usher will want to be supported by Sense or Deafblind UK etc			X																X		3
Need to keep busy to adjust to loss e.g. being involved in usher groups/organizations																X					1
People have to ready and willing to access specialist organisations/charities		X								X									X		4
Signposting all the orgs./charities etc., so people can approach their individual choice depending on their personality/need		X									X	X					X		X		6
Importance of specialist organisational support (including legal departments)		X	X	X		X	X	X	X	X	X	X					X	X		X	39
**KEY ** Directory of support networks…e.g. Sense, Deafblind UK, Facebook, counselling services who specialist in deafblind/usher, bereavement services and contact details to be issued at point of diagnosis and available thereafter		X				X						X	X			X	X			X	16
Organised charities not necessarily for everyone			X																	X	5
Mutual benefits: Shared experiences; the person with US and the specialist sensory organisation			X							X								X			5
Person expresses having association with the support groups you learn things you don't learn if you just do research										X											1
People with sensory need volunteer and are involved with individuals with sensory need/sensory											X										5

groups/organisations																					
Sensory Social Work/Sensory Support/Equipment																					
Essentiality of sensory social work support to support person with usher to achieve aspirations																				X	1
Over reliance on social work/support workers by person who has usher																				X	1
Ticking bomb if people do not get support at the right time													X								1
Person felt looked after as professional was clear, honest and empathic even when it is bad news																				X	1
The benefit of being registered blind is you can access social care or social services																X					1
Experience of no support even though person has had condition all their life															X	X		X			3
Insufficient support leaves person feeling abandoned																X					1
Sensory expertise combined with safeguarding experience		X	X	X	X	X	X	X	X		X	X	X	X		X	X	X			37
Specialist sensory visit every so often to ensure person supported												X		X							2
People slipping through system due to circumstances outside of sensory need.													X								1
Joined up working to ensure holistic sensory support												X	X							X	2
Need for dual sensory impairment specialist																				X	1
Provision of communicator guide specialist		X									X					X					4
No provision of communicator guide specialist – not in person with ushers day																		X			1
Automatic referral by hospital to specialist sensory social worker			X						X			X									3
Specialist deafblind worker within local council – nationwide not postcode lottery		X	X							X						X			X		11
Being thrown in the deep end -																					2

professionals expecting person with usher to be a spokesperson when they are adjusting to condition themselves																X					
People not having choices about sensory services they need											X										1
Provision of communicator guide											X					X					4
Specialist sensory equipment needed to support individual needs		X	X		X	X	X	X	X	X	X	X	X	X		X	X	X	X		42
Provide information in sensory format e.g. large print/audio													X								1
Specialist sensory training to adapt to sensory loss			X								X										2
Specialist training regarding hearing and eyesight required to provide person's particular needs to be met													X								2
Signposting and supporting					X								X						X		5
Practical help for daily living			X		X		X		X		X		X			X		X	X		20
Recognition of changing needs			X		X	X	X					X				X					7
Equipment identifies person as having sensory need/disability						X	X	X													6
Equipment identifies person as having sensory need/disability - can lead to feeling of vulnerability							X					X					X				4
Equipment identifies person as different leads to bullying												X									2
Equipment identifies person as having sensory need/disability- positive aspects e.g. being seen quicker in a queue							X														1
Support contributes to independence															X						2
Technology																					
As technology advances potential dangers arise e.g. hearing aids cutting out background noise which is essential for hearing road noise			X																		2
As technology advances potential arise e.g. in terms of hybrids electric cars being silent so crossing roads would become a problem			X																		2
Forced to use new technology when old technology loved but no longer available e.g. hearing aids			X								X										3

Technology important e.g. FaceTime, Facebook, e mail for communication		X	X	X	X	X	X		X	X	X		X			X		X	X	X	17
Facebook great way to communication with other people with Usher/RP who person may otherwise not have come across																				X	1
Technology important for someone with usher it keeps person in touch with the sighted/ hearing world																				X	1
Technology allows person with usher to write (touch type) read (screen reader) and braille (braille note) easily in a mainstream environment																				X	1
Skype/FaceTime prevent isolation and enable communication to be maintained																		X		X	3
Sensory aware technology e.g. tactile instead of vision reliance; adjustable sound and vision						X			X		X		X					X			9
Sensory aware technology available for children – Provide funding for this to happen									X												1
Learning new skills to adapt to sensory needs e.g. technology									X		X										4
Technology enables research – information on Usher as a condition and where to go for support e.g. specialist organisations										X											2
Missing out contact with friends if unable to use technology												X									1
Technology not for everyone														X							1
Safety																					1
Technological advances raise potential safety issues e.g. electric cars		X		X																	2
Vision and sight loss lead to worry people breaking into home and not hearing or seeing				X																	1
Poor eyesight affects safety													X								1
Equality																					
Nationwide equality of services for people with Usher																		X			1
Raising awareness																					

People with Usher are the same as everybody else											X							X			2
Person with usher campaigns to raise awareness																	X				4
Awareness being raised so young people, children and their parents are aware of support available from a young age															X						1
Raising of awareness so general public understand condition		X		X		X					X	X		X	X	X	X				12
Prior to having usher person had not come into contact with anyone with a visible disability																	X				1
Until diagnosis person had never heard of usher – Raise awareness																	X				1
Raising awareness of disability in general – to encourage better understanding																	X				1
People with usher raise awareness about the condition																X					1
Person with usher raising awareness but not sure if they will be any good at it.																X					1
Raise awareness GP's Hospitals, Audiology, Ophthalmology															X	X	X				2
Raise awareness in Deaf community (video, I am Usher)				X																	1
Sensory awareness from people who know about sensory issues, those with sensory need e.g. Service user/Expert by Experience											X										4
Raise awareness – Not end of life - people with Usher relay experiences to people diagnosed/their parents etc				X																	1
Raising awareness of how condition may change communication method e.g. BSL to hands on signing				X																	3
Raise awareness to improve communication for people with changing sensory needs				X								X									2
Raise awareness by demonstrating how little can see – supported by specialist deafblind organisations				X																	1

Lack of awareness and understanding of condition – (inc services such as bus drivers)		X		X					X												5
Lack of awareness and understanding of condition even amongst people who have been Deaf all their lives		X		X																	2
Raise awareness about the condition		X		X		X	X			X	X	X			X	X	X				13
Raise awareness to avoid isolation and segregation											X					X					11
Raise awareness – Lessons for normal children about sight and hearing problems									X												1
Lack of awareness can have serious consequences for the person with sensory needs				X												X					5
Lack of awareness can have serious consequences for the family of the person with sensory needs				X																	1
Older persons day centres, residential and nursing homes to be made Usher aware				X																	1
Lack of awareness and understanding amongst professionals who work in the sensory field					X	X						X				X	X				9
Lack of awareness and understanding amongst professionals						X							X								2
Raise policy/governmental awareness – social care reform bill							X														1
Raise awareness of what people with Usher are entitled to and how to access – Ensure they understand in their chosen method of communication											X										1
Raise awareness to students											X										2
People with sensory need campaign to raise awareness											X										1
It's about recognizing Usher																X					1

Appendix D: Data analysis: Initial codes for stage one analysis

1. Impact of diagnosis
2. Post diagnosis
3. Role of specialist services in diagnosis
4. Research – For solutions Placental implants
5. Diagnosis methods
6. Noticing something is wrong
7. Guide dog provision
8. Equipment, support and raising awareness
9. Early diagnosis
10. Identity development Parenting
11. Expectations of disabled people and derogatory responses
12. Culture difference & diversity
13. Friendships and shared experiences
14. Communication
15. Big ‘D’ Deafness & Communication
16. Education
17. Audiology
18. Ophthalmology
19. Validity of condition
20. Benefits and employment
21. Googling diagnosis
22. Reactions to being given hope
23. Bereavement
24. Feelings, attitude (positive & negative) & emotions
25. Misconceptions
26. Family -Friends, peers, neighbours & colleagues

27. Specialist organisational support

28. Sensory social work/Sensory support/Equipment

29. Safety

30. Technology

31. Raising awareness

Appendix E: Data analysis: Expansion of initial codes for stage one analysis

Child diagnosed with Usher

- Child worries parent life is now restricted
- Parent not telling child about diagnosis/prognosis of Usher. Child's view "you must let your child know"
- Child draws own conclusions if not advised they have usher by parent(s)/caregiver
- Impact of parent(s)/caregiver hiding condition rom child
- Child finding it hard to express difficulties encountered with having usher as they don't want to upset their parents

Siblings

- Effect on siblings who do not have Usher
- Effect on child with Usher when sibling have additional needs/more demanding disability

Parent's whose child is diagnosed with Usher

- Guilt as condition is genetic/hereditary and gene is passed down – parents being carriers
- What support is put in place for parents? And for parents to support child? Hard for parents to support child when they are struggling with diagnosis themselves
- Parents worse nightmare
- Parent not wanting to accept child has a disability – positive/negative impacts. Child not treated any differently V lack of support
- Extra work for parents e.g. hospital appointment's, contact with sensory support etc.
- Parent's role of choosing right time to tell child they have Usher
- Parent's being overprotective – wrapped up in cotton wool

- Misconceptions

Parent who has Usher

- Not wanting their children to become carers
- Impact on child when parent is diagnosed E.g. child(ren) may need counselling
- Parenting – Not being able to look after child

Relationships

- Risks posed when person with usher has to care for aged parent
- Parent can't speak to partner about child having Usher syndrome

Family support

- Essentiality of it
- Positive attitude to deafness
- Having a lot in common when other family members deafblind
- Lack of it
- Inappropriate use of family as interpreter e.g. perpetrator of physical abuse

How change impacts – A theoretical discussion

- Identity development Erikson (Kroger 2007, Arnett 2000) – Whole of section on identity
- Stages of grief (Kubler-Ross 1970) – Whole of section on bereavement

Culture Diversity & Difference

- Big 'D' Deafness & Communication
- Two Usher culture, sign language group and hearing and sight impaired
- All Deaf same culture happy together

- After diagnosis of usher deafblindness people still see themselves as culturally Deaf with history and culture
- Personal identity number given to Deaf child upon school entry – used instead of name; personal identity education number used after leaving school e.g. in e mail; Positive attitude to using personal identity number
- Being part of the Deaf community linked to ability to sign
- Not all deaf people with Usher see themselves in the deaf community and not all necessarily all use sign language
- Change in eyesight changes relationships with friends e.g. not being able to see sign language which is a visual language
- Deaf and hearing united by shared experience - Usher

Communication

- Not taught sign language at home as parents oral
- Learnt sign language later in life, made friends talk more
- Deteriorating sight affects ability to sign
- Sign language can be out when person cant see

Equipment, support and raising awareness

- Guide dog provision
- Early diagnosis
- Friendships and shared experiences
- Education
- Audiology & Ophthalmology
- Validity of condition
- Benefits and employment
- Specialist organisational support
- Sensory social work/Sensory support/Equipment
- Equality

- Raising awareness

Technology

- Technology important e.g Facebook, FaceTime e mail
- Facebook and twitter mean a person with Usher can access people with the same condition worldwide
- Facebook means people with Usher are no longer alone
- If person in family won't sign can use mobile to phone text to talk
- Skype/FaceTime prevent isolation and enable communication to be maintained
- Technology not for everyone

Safety

- As technology advances in terms of hybrid electric cars being silent crossing the road could become a problem
- Vision and sight loss leads to worry people breaking into home and not hearing and seeing
- Poor eyesight affects safety
- Impact of dual sensory loss – safety implications e.g. not seeing platform to the train track

Appendix F: Sample of data analysis chart - Theme 1: 'Diagnosis is the start of the experience'

Early life, life before Usher	Impact of diagnosis	Experiences after diagnosis
Erm when I was at XXXXXX [School] at XXXX [Place] and err then as I say I couldn't see the blackboard and they kept saying I was a clumsy child because kept knocking things like the milk bottle. They used to put it on the table and I kept knocking it. Silly things there were a lot of black cars in them days and black bikes and I knocked a ladies bike over and when she was doing deliveries, because they used to have a bike basket on the front. They used to do home deliveries and I know knocked it flying because I didn't see her and my father took me back to the doctors said there is something wrong with her eyes (Ruth).	You know what I mean. When I was younger I went for night walks along a cliff top and I can't see a f***g thing. As long as I was holding onto my mates shoulder right in front of me it should be all alright and we are right on the very edge of the cliff top having a night walk sort of thing. Yeah, erm yeah I think I was far far less cautious pre-diagnosis than after the diagnosis without a doubt.	Doctor explained shocked very depressed shocked and depressed. Didn't know would be blind first time heard. Why me? Why me? Doctor said I sorry. My step mother said sorry to me for many many years ago. Go on holiday 1992 partially sighted then. XXX [family member] and my husband all went to XXX [place] , saw family I said I blind they said I sure. They shocked, I said can't see in dark. I had stick at home they guide me. People saw stick said blind. I explained to them through day can see but at night not. Shocked. (Eve)
I thought it was very much I was just clumsy you know. I had been travelling the world, lived outside of XXXX [Place] Just come back to XXXX [Place]for work all that sort of stuff it was a way of shutting my mum up because she kept wittering on about the fact I was treading on the cat all the time, walking into things, I was covered in bruises! On my legs like all the time. (Sara)	Make most of life be happy because there are worse things happening than losing sight and hearing. It's not the end of the world it really isn't, erm I don't know I've learnt to live with it. I've done abseiling I've done everything. No, you're normal just because you got deafblindnes you are not abnormal. You can do the things that people are sighted do. You can do. Just take the T off don't say can't say can take the T off. (Ruth)	I was young so um, you become emotionally upset erm think I had to seek profession help I was on Valium for a short time diazepam. Erm that kind of thing, very upsetting. Oh now my lifestyle has completely changed. Talking about years ago when I was first you know…….(long pause) diagnosed my car was taken away then of course my erm you know my bicycle. (Gareth)
Admittedly I did have a couple of silly little car accidents and that was only purely because I did	Growing up in the past year or so has made me realise some of the restrictions that go with	"I went to my GP. He sent me away to a place. Yes a psychiatric place. Yes. (long pause) because I had

not see the car in the field of my vision and went into it. Silly but that's erm at the time I didn't really twig I wasn't seeing very well. (Jeff)	Ushers, although I have lots of coping strategies I will never drive, walk anywhere on my own, read a book again and that makes things different. I am also over reliant on my parents, teachers and support workers and that can be hard. (Ruth)	suicide, yes suicidal thoughts Erm erm. I was there for 8 weeks, yes 8 weeks. I came home I was still depressed and the doctor he prescribed anti-depressants. But um I stayed on as long as I could. I don't know if it did anything but I stayed as long as I could. Years went by and I took anti-depressant. Yes yes". (Len)

Appendix G: Sample of data analysis chart - Theme 2: 'Familial relationships across the lifespan'

Child diagnosed with Usher	Siblings of a child who has Usher	Being a parent who has Usher	Relationships	Family support
I was with one group talking about my situation you know the diagnosis with my parents not telling me and I was saying you must you know let your children know. (Kate)	"They are young but have been quick to pick up what has happened to me and have been great guides etc" (Tia).	"When daughter born mother said doctors approached my mother [participant's mother] and said wouldn't be able to look after baby. When daughter was 6 weeks old wanted to take baby away, social workers. I was no capable of looking after. Grandmother came to live with them to stop baby being taken away". "When second baby born, not talk about taking baby away but health visitor came every day to check. Social worker came every Friday"…. "Was happy, forgot was disabled, brought family up went with husband in car on holidays". (Debra)	"I had a horrible dark secret, I knew I was diagnosed with RP and I knew what RP meant it was like a horrible burden on my back that I couldn't quite get off. You know it made me nervous very nervous". (Gareth)	"My parents have always told me to be positive and don't worry about things you do not have control over". (Carl)
"Now I was about 15 at that time erm. My mother took me	"My mum and brother were very supportive	"No. No. When I did have one of my first child erm they said I	"Socialising was always difficult, going out was difficult and	"I do know of a couple of members who in [Usher

to the hospital all I remember is being asked to wait outside whilst they spoke to my mother, I thought that was a little bit odd. Then she came out and er yes in her body she was very uptight actually. Held my hand and went, didn't speak didn't say anything at all". "Then we went into…we lived in [name of town] at that time and we went into the British Home Stores. There was a hat, it was a pink flamingo, sunglow hat and I just fell in love with it and my mother bought it for me. I thought that was a little bit odd. I have 3 sisters why me"? ….."Shortly before doing my A levels 15 going on 16, I found myself being taken on a pilgrimage trip to Lourdes and erm you know actually met up with one or two other children who were seriously ill.	and did much of the reading of the paperwork for me as by this time I could no longer read the printed form at any size any more" (Carl)	wouldn't be able to cope with her and all things like that. But we soon pushed him out the door. No. No social worker no my mum was left with it". (Ruth)	interacting with people was difficult including going to a nightclub, cinema and restaurant/bar. It was very noticeable to people that something was wrong, that had an affect both professionally and personally. I would see people keeping their distance". (Oliver)	support group] have admitted they suffered stages of depression. Fortunately I have a very supportive wife, 3 great kids".(Jeff)

One boy had leukaemia and he did actually die later and then a 10 year old boy had cancer. Other people I was with were quite seriously ill and I thought why am I here? You know…when I only have hearing problems". (Kate)				
I've found it hard to express my difficulties as I have not wanted to upset them. Whilst they definitely want what is best for me, it has been hard for us to support each other when we have each been struggling with it individually" (Tia).	"I terrible speech problem. My sister terrible. Got the same thing as me. She's in a home now. She's got the same as me but she's got more and learning disabilities as well. I had to get on with it. With my little sister my youngest, there are 6 of us, she got all the help under the sun. She had learning disabilities and I was pushed to one side" (Ruth).		"I covered up my inability to not see by pretending I was drunk, because if you are a young undergraduate [name of profession] it's quite alright to be drunk but it's not OK not being able to see". (Quentin)	"My parents taught me nothing. I have been bumbling through life, just bumbling through life". (Monica)

Appendix H: Sample of data analysis chart - Theme 3: 'A sense of belonging'

Interrelationships between communication, culture and community	Friendships and shared experiences
"All Deaf, same culture same, happy together". …Few years ago social worker came to help me, what do you want me to put on the form deafblind? No Deaf after that the sight got worse. See myself as part of the Deaf culture with culture, history….Communication is easy I feel good, same intelligence, same side…..Hearing world I can't pick it up comedy and laughter it means nothing to me I don't understand but Deaf laugh at same thing". (Debra)	"Now with [deafblind charity], I got in touch with a lovely lady [name of worker] from [deafblind charity]. She said there was this support group. It was actually just a bunch of people with hearing and sight issues and they meet a few times a year. Social activities, you can share your experiences and learn from them, how you cope"…. "Having association with the support groups you learn things you don't learn if you just do research….Now a person may not be comfortable to talk about it at the time. It is about gaining trust and bonding with people. But it opens up the door or the window whatever you want to call it. People may say I don't have problems with my eyes we all have problems with our eyes. But…what is the specific problem with your eyes? Oh I get this sort of this oh yes….yes then you are talking about it. If you don't talk about it people worry. I am not suggesting that they do, but people might think whatever it is problem could be life threatening it can bring on stresses to a person so yes. Like-minded people" (Jeff).
"I go to [name of support group] member's day who use sign language. Don't have much to do with them they keep to themselves in their own little groups". (Gareth)	"I am going to [name of town] in 2 weeks' time. This Friday I am going to [name of town] after that I am going up to[name of town]. I am … I mean yes I am getting busy I enjoy it and it gives me a chance to meet other people".(Kate)
"The Deaf community not at all, they are a world to their own".(Jeff)	"Club that was nice a lot of people object to being with older people I was in my 30s late 30s 39 it never bothered me how old somebody is if you have common ground….I found that very interesting. Um err it it gave me a lot of information about how to communicate effectively because I am very good my central vision I call it my central vision …I

	had one tunnel that went one way and another tunnel that goes the other way. A lot of people say about central vision but mine has never been straight ahead that is why I had problems socializing but I learned a lot of things about ways to communicate because some peoples type of hearing loss very different to mine and they need different tones to the voice”.

Appendix I: Sample of data analysis chart -Theme 4: 'Experiences of professional support'

Education employment and benefits	Sensory support ophthalmology, audiology and social work	Sensory equipment and guide dog provision	Specialist charity support	Raising Usher awareness
"I did have problem hearing and understanding what teachers was saying – Fell behind with my education". (Nora)	"I had a visit from a social worker all she brought was a letter saying you are now registered blind and she gave me a white stick and that's all she did…. I didn't know about [specialist charity supporting deafblind people], nothing. I didn't know anything like that existed. I had packed up work I was on my own….I just feel that once you have been registered, been registered by the local authority blah blah blah, there should be a system where people I don't know get a visit every so often". (Len)	"By this time, I was needing some help with my computer work with changing the colours from black on white to the reverse scheme of white on black background My brother made me a video inverter module plugged in between my computer and my display to do this without relying on the Windows environment itself and I also used a high spec graphic card to provide magnification (x2 and x4 only) without (again) the Windows knowing about it or providing the magnify function" (Carl).	"I remember a lady from [name of charity] coming down and talking to me and erm erm that felt strange to be truthful. Having someone to talk to me". (Jeff)	"Until I was diagnosed I had never heard of it (Usher syndrome). I had never met anyone with Usher syndrome before in fact even when at the time I was diagnosed. I did not meet anyone with Usher syndrome until I joined [specialist charity]". (Quentin)
"I started to lip read at age 12/13 as I struggled at	"I had contact with social services they didn't do much	"I have a symbol cane yes, a quarter of a	It is only recently in the last couple of years that perhaps I felt that I need to	"No I never met anyone with Usher till I was in my 40s. I hadn't

school and I wanted to find a way to get to grips with what was being said. By this I found an alternative method to grab information by lip reading" (Oliver)	but a cane came through my letter box, my door and then another social worker came to see me from the visually impaired and said... oh she asked me what I could see and what I couldn't I thought I can only see what I have ever seen. I didn't understand about how much vision I had lost. She said it doesn't go black you know, well I had black all around my eyes but I was thinking I had hair in my eyes or whatever, that kind of thing. She said you are fine you don't need any kind of help. That's what she said to me… Then I got back in contact with them I said I feel..I am not going to leave my house properly if I don't have some kind of …it was [specialist charity supporting deafblind people] actually it was [specialist charity supporting deafblind people] had actually back then asked me to go to [city name] for a full day	metre. I don't go anywhere without it" (Quentin).	start concentrating on my joint sensory loss and with the problems of my hearing aids, I contacted [name of charity] to seek help from them and I have joined with them. I am involved with their technology section as well as the Usher area with [name of charity]".(Carl)	ever heard of it". (Pam)

	assessment". (Pam)			
"I was forced to go to the job centre; my brother had to take half a day off. I went there with a red and white [13] stick and a personal listener wires I used headphones, she still said to me at the end there "could you please look at the monitor and check what I have written". I started waving my stick around and said I can't see the monitor let alone the text. She said we still think you should apply for work. I don't need all this sorry I put letters in front of her from [specialist eye hospital] hospital form the [Ear Nose and Throat hospital]l and then she just ignored it all". (Gareth)	"[Town name] was a good move for me as the support from the council was miles better". (Carl)	"I used to come back stressed out and [swear word] off with a lot of people. Sometimes people did not respect the type of people [name of guide dog] has made a big difference. I feel a lot safer with [name of guide dog], a lot calmer" (Ben).	"I know who they are I know what they do I just for me think it's a little bit, this is gonna sound awful but…this is how I feel. I feel that I've got erm a dual sensory loss and am deafblind and all this sort of stuff but I am not the same level as the people they help and some of them…does this make sense what I am saying?" (Sara)	"I have experience that the bus drivers are not always aware how important for deafblind people to get off the stop they need to get off I had a driver refuse to stop at my stop, a long story. This had now put me off travelling on buses on my own which is now two years ago". (Iris)

Appendix J: Ethics approval letter

London South Bank University

Direct line: 020-7815 6024

E-mail: dippenas@lsbu.ac.uk

Ref: UREC 1256

Michelle Evans

XXX [Home address]

Dear Michelle,

Re: Exploration of the lived experiences of young people with Usher Syndrome' (UREC 1256)

Thank you for submitting this proposal and for your response to the reviewers' comments.

I am pleased to inform you that your application to the University Research Ethics Committee for the above study has been reviewed. The Chair is able to confirm that the study was completed in keeping with the London South Bank University Code of Practice for Research with Human Participants.

I wish you every success with your research.

Yours sincerely,

Sharon Dippenaar

Secretary, LSBU Research Ethics Committee

cc:

Prof Joan Curzio, Chair, LSBU Research Ethics Committee

24th October 2012

Appendix K: Ethics approval letter age range adjustment

London South Bank University

Direct line: 020-7815 6025

E-mail: mitchen5@lsbu.ac.uk

Ref: UREC 1256

Michelle Evans

XXX [Home address]

Dear Michelle,

Re: Amendments- Exploration of the lived experiences of young people with Usher Syndrome' (UREC 1256)

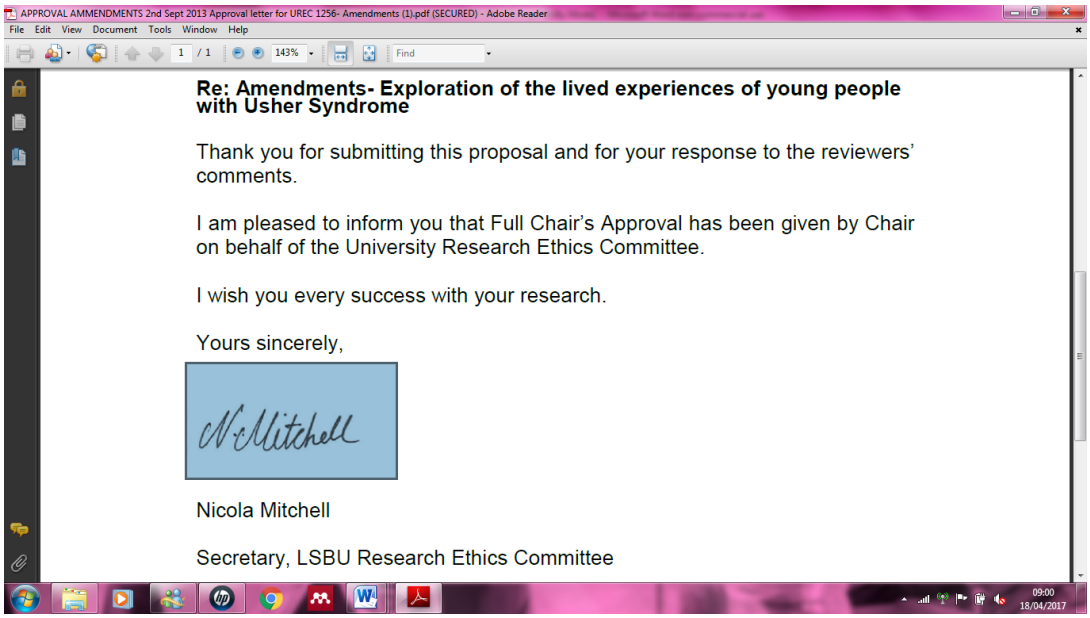
Thank you for submitting this proposal and for your response to the reviewers' comments.

I am pleased to inform you that Full Chair's approval has been given by the Chair on behalf of the University Research Ethics Committee.

I wish you every success with your research.

Yours sincerely,

Nicola Mitchell



Secretary, LSBU Research Ethics Committee

cc:

Prof Shushma Patel, Chair, LSBU Research Ethics Committee

Appendix L: Ethics approval, age range adjustments - Supporting emails

From: Patel, Shushma

Sent: 12 April 2016 10:01

To: Evans, Michelle 9

Cc: Governance : Administration Of Research Ethics Committee

Subject: RE: Ethics ammendment to include 16+ - no upper age limit response to Shushma

Dear Michelle,

thank you for the email.

I am sorry that a formal letter of approval for the amendments was not sent by the then Secretary for the UREC.

The email states that the amendments were approved, so the email is acceptable, and can be submitted as approval.

If you have any problems, please get back to me.

Regards

Shushma

Professor Shushma Patel

Professor of Information Systems

Director of Education & Student Experience

School of Engineering

London South Bank University

103 Borough Road

London SE1 0AA

+44 (0)20 7815 7412

From: Patel, Shushma

Sent: 01 September 2013 15:57

To: Evans, Michelle Deborah 2

Cc: Governance : Administration Of Research Ethics Committee; Curzio, Joan; Popple, Keith John; Goodyer, Annabel May

Subject: RE: Ethics ammendment to include 16+ - no upper age limit response to Shushma

Dear Michelle,

I am really sorry that I have not been able to respond to this earlier.

I am happy with the amendments submitted. Can you please ensure that the contact details for the UREC are changed so that they are for the attention of the Chair of the Committee at ethics@lsbu.ac.uk.

Also, please ensure that the title and UREC number for your study appears as a header on all pages of the participant information sheet.

Please take this email as confirmation and a formal letter of approval will also be sent for the amendment to your application.

Please accept my apologies again for the delay and I wish you well in a very interesting study.

Regards

Shushma

Professor Shushma Patel

Professor of Information Systems

Faculty of Business

London South Bank University

103 Borough Road

London SE1 0AA

+44 (0)20 7815 7412

From: Michelle Evans [mevans223@yahoo.co.uk]

From: "Patel, Shushma" <shushma@lsbu.ac.uk>

From: Evans, Michelle Deborah 2

Sent: 03 July 2013 14:37

To: Patel, Shushma

Cc: Popple, Keith John; Curzio, Joan; Goodyer, Annabel May

Subject: Ethics ammendment to include 16+ - no upper age limit

Dear Shushma

Re: 'Exploration of the lived experiences of people with Usher Syndrome'

The current age range on the study is 18-49. The project is with people who experience Usher Syndrome which is a very rare form of deafblindness. However I have had difficulty recruiting due to age limitations. I now have people responding of varying ages the youngest being 16 the oldest 83 who I would like to include in the

interviews. So could I extend the age range of the participants to 16+ with no upper age limit please?

Also the option for implied consent if the response is via e mail.

I have attached parental participant information and consent forms and young person/child's participant and assent forms.

I await to hear your response

Regards

Michelle

Appendix M: Consent form

London South Bank University

Consent Form (UREC 1256)

People with Usher Syndrome

After having read the Participant Information Sheet (of which I have retained a copy). I consent to take part in the research study;

'Exploration of the lived experiences of people with Usher Syndrome'

The researcher has explained the purpose and proposals of the research study and I confirm I have had the opportunity to read, understand and ask questions about the research study**.**

I have been advised that confidentiality will be adhered to at all times and that the data collected from the study will be safely stored and coded to ensure that information is anonymous.

I have been fully informed about how the data collected for this research study will be used, to whom it may be disclosed, and how long it will be retained. I understand that I am free to withdraw from the study at any time, without giving a reason up until the point of dissertation submission. After which withdrawal is not possible. I agree to the interview being video recorded

I confirm that I voluntarily and without coercion consent to participate in this study.

Participant's Name: (Block Capitals) …….

Participant's Signature: ……

Date:

……..

I as the researcher responsible for this study confirm that I have explained the purpose of the research, what it will be used for and how it will be disseminated. I also confirm that where a communicator/interpreter is used they will be asked to sign a confidentiality agreement to maintain confidentiality at all times.

Researcher's Name: Michelle Evans

E Mail: evansm2@lsbu.ac.uk

Researcher's Signature …….

Date ……..…….

Appendix N: Confidentiality agreement (interpreters/communicators)

London South Bank University

Confidentiality Agreement Form (UREC 1256)

This is to certify that I have read the attached participant information sheet with regard to research being conducted with regard to:-

'Exploration of the lived experiences of people with Usher Syndrome'

The researcher has explained the purpose of the research being conducted. As an interpreter/communicator I am aware that I am not required to contribute to the interviewing process in a personal capacity, but rather simply interpret/communicate for the participant involved in the interviewing process. I have discussed the matter with the participant themselves and the researcher.

I understand my involvement will be confidential. I am also aware that by signing this confidentiality agreement I am agreeing to keep all information and data that arises from the interview confidential. I agree to maintain confidentiality at all times.

I agree for the interview discussion to be video recorded as I am aware this is necessary for translation and data analysis.

I fully agree and consent voluntarily and without coercion to being involved in this research study as an interpreter/communicator.

I also fully agree for the information I convey to be used by the researcher in accordance with the details outlined in the Participant Information Sheet.

Interpreter/Communicator's Name: (Block Capitals) …….

Interpreter/Communicator's signature: …….

Date: …….

As the researcher responsible for this study I confirm that I have explained to the interpreter/communicator named above the purpose of the research.

Researcher's Name: Michelle Evans: evansm2@lsbu.ac.uk

Researcher's Signature: …….

Date: …….

Appendix O: Participant information sheet

London South Bank University

Participant Information Sheet (UREC 1256)

Research Title:

'Exploration of the lived experiences of people with Usher Syndrome'

(E Mail:evansm2@lsbu.ac.uk)

What is the aim of this study?

To explore people's own interpretations of their changing experiences since being diagnosed with Usher Syndrome with a view to developing knowledge and raising awareness.

What is the purpose of the study?

The purpose of this study is to conduct research into finding out about your life experiences as a person with Usher Syndrome. This study is part of Mphil/PhD research being undertaken at London South Bank University.

Why have you been chosen?

You have been chosen to help us and others to better understand what life with Usher Syndrome is like.

Do I have to take part?

It is your choice as to whether you decide to take part or not. If you do decide to take part you will be given this information sheet to keep. You will also be asked to sign a consent form which is a separate document. If you decide to take part you are free to withdraw at any time up to point the dissertation is submitted and without giving a reason. However, after this point withdrawal will not be possible.

What will happen if I choose to take part?

If you agree to take part, you will be contacted to arrange a suitable time and venue for the interview to take place. You will also be asked how you would like the interview to be conducted. You can meet the researcher face to face, meet over SKYPE or communicate via email only. The researcher is skilled in British Sign Language (level 3) and deafblind manual (level 2) but appreciates you may feel more comfortable using your own interpreter.

With your permission interviews will be video recorded where possible. Please discuss with the researcher if you have any concerns with regard to this. You will be also asked to sign a consent form to confirm your participation in this research study is voluntary and that you have been fully informed.

What are the possible disadvantages and risks of taking part?

There is a risk in any interview that you may become upset. To minimise this risk, I will be speaking to you in a professional rather than a personal capacity. At the beginning of the interview, the issue of confidentiality will be discussed and guidelines will be agreed. Whilst upsets are not anticipated, you will be asked to provide a personal contact should a serious upset occur.

What are the possible benefits of taking part?

You may find talking about the experiences of living with Usher Syndrome helpful. Additionally, your contribution may help as a basis for raising awareness of issues

relating to Usher Syndrome and thus contribute to helping people with Usher Syndrome in the future.

Once complete I will consider submitting findings as journal articles, publications, training support package or other avenues to raise awareness with regard to Usher Syndrome and sensory awareness.

What if something goes wrong?

If you are unhappy with any aspect of the research, you can contact me directly or contact my supervisors, whose contact details are below:

Professor Keith Popple popplekj@lsbu.ac.uk

Professor Joan Curzio curziojl@lsabu.ac.uk

Dr Annabel Goodyer goodyeam@lsbu.ac.uk

If the matter remains unresolved and you wish to complain formally to someone outside the research team the University Research Ethics Committee – Chair of the Ethics Committee can be contacted email: ethics@lsbu.ac.uk

If further details are required they are obtainable from the university website: https://my.lsbu.ac.uk/page/research-degrees-ethics

Will my taking part in the study be kept confidential?

All information which is collected about you during the course of the research will be kept strictly confidential. Any information about you which is shared with others (e.g. report and publications or is shared with a supervisor) will have your name and address removed so that you cannot be recognized from it. The information you provide will be treated confidentially and stored in a locked cabinet. Electronic information will be stored on a password protected computer in an environment which is locked when not occupied. An encrypted data stick will be used to store

data. References to you will be coded. The information will be held for a period of five years following submission of the thesis and then confidentially destroyed.

What will happen to the results of the research study?

The results of the research study will be written up as part of my Doctoral Thesis. My research supervisor, marker/moderator and external examiner will have access to the research. Findings may also be written as journal articles, publications etc. Confidentiality will be maintained.

Who has reviewed this research?

It has been reviewed and ethically approved by the London South Bank University Research Ethics Committee.

For further information contact

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If you agree to be part of the research study please contact me and we will make mutually agreeable time.