LIVING WITH CYSTIC FIBROSIS: PATIENTS' EXPERIENCES OF DIAGNOSIS IN ADULTHOOD

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A thesis submitted in partial fulfilment of the requirements of the University of the West of England, Bristol for the degree of Professional Doctorate in Health Psychology

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May 2020

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ACKNOWLEDGEMENTS

This thesis is dedicated to my mum, dad and sister Shashi. Words cannot express my gratitude for your unconditional love and support throughout my life. You have always instilled the value of education and encouraged me to work hard to achieve my dreams.

A huge thank you to the participants that I interviewed for without whom this research would not have been possible. Thank you for your time and inviting me into your world to share your experiences with me.

My supervisors Professor Diana Harcourt, Dr Elizabeth Jenkinson and Dr Alison Pearce thank you for your invaluable feedback, guidance, patience and expertise throughout. You continually challenged me to achieve my potential. Alison, thank you for giving me that opportunity many years ago to come and work for you.

My study buddy Lisa, my heartfelt thank you! Without your encouragement, I would not have been on this programme and would have struggled to get through this doctorate alone. The endless hours we spent working together at weekends was made much more enjoyable with our food feasts, life chats and discussions about our future plans. To my fellow UWE trainees of 2013, it has been a pleasure to be on this journey with you and I hope we continue to cross paths in the future.

Thank you to Eastleigh Goju Ryu Karate Dojo for keeping me sane throughout my doctorate. Karate training provided me with much-needed stress relief and break

from my studies. In the process, I have had the opportunity to make new friends and a chance to continually grow and develop my skills.

Thank you to my friends and family for your positive encouragement throughout. Parsons, where do I start, thank you just does not seem enough. I am eternally grateful for the many motivational pep talks, for your time talking through the interviews and for reading through my drafts. Thank you for being there! Lisa, Shashi, Abhilasha, Ekta, Shiva and Anisha thank you for your continual support and comments on my draft. Hetty thank you for proof reading and your feedback on the final edit. Pawan and Michaela, thank you for your support in the final push in getting it done. Ranveer thank you for always bringing a smile to my face, making me laugh with your mischievous antics and being a much-welcomed distraction from studying.

To my fiancé Deepak, there are not enough words to say thank you. I am truly grateful for the countless times that you kept me company whilst I studied and your emotional support throughout. You always believed in me, even when I didn’t. Thank you for your continual motivation and patience. I look forward to our new adventures together.

And lastly, to Bigi, it is with great sadness that you are not here today to see me completing my studies, but I know you will always be proud.

PREFACE

The Professional Doctorate in Health Psychology requires the successful completion of five competencies: 1) professional skills, 2) behaviour change intervention, 3) consultancy, 4) teaching and training and 5) research. The research competency is split into two parts, comprising the systematic review and empirical study. This thesis has been written to fulfil the requirements of the research competency.

Before undertaking this doctorate, I worked part-time for four years as a volunteer psychology assistant for a regional cystic fibrosis (CF) service. As part of my role, I was tasked with undertaking literature reviews around topics in CF, one such area was adults diagnosed with CF. From the literature, I identified there was a paucity of research within this area and possible scope for a research project. I put together a research proposal and was keen to undertake the project with the support of the psychologist I worked with. However, with limited academic direction and support, the project was not fruitful. When it came to starting my doctorate, I saw this as the perfect opportunity to reignite my previous idea for a research project. The focus of this empirical research is to explore the experiences of adults diagnosed and living with CF in adulthood.

As per the competency requirements, I completed a systematic review within the first year of the doctorate. The title of the review was “Psychosocial interventions that improve coping for people with cystic fibrosis: A systematic review of randomised and non-randomised trials”. As the systematic review did not inform this research it has been included in the appendix (see Appendix A).

ABSTRACT

There is a paucity of research investigating what it is like to be diagnosed and to live with cystic fibrosis (CF) in adulthood. Understanding the experiences of these adults and the impact of the condition can provide information to help healthcare professionals deliver appropriate support for their patients. This research aimed to address this gap in the literature. In-depth semi-structured interviews were carried out with sixteen participants (ten females and six males) diagnosed with CF in adulthood. Using thematic analysis, four themes were identified “No, you can’t possibly have CF”, Emotions around diagnosis, “It did kind of take over my life” and “I no longer wish to argue with it”. These themes described participants’ frustrations with their contact with healthcare providers before diagnosis, their ambivalence around their diagnosis, the various impact and challenges faced with day-to-day living, work-life, finances, relationships, fertility, life plans, the future, treatment burden and their acceptance and adjustment to their CF. A key finding was the mismatch between patient need and healthcare provision. The main recommendations made included: 1) raising a greater awareness amongst non-CF specialist healthcare professionals and the general public of the possibility of receiving a CF diagnosis in adulthood, 2) ensuring CF healthcare professionals delivering the news of an adult CF diagnosis have the appropriate training and support and 3) ensuring CF healthcare professionals adequately assess individual’s information needs and provide appropriate and relevant information. The implications for health psychology practice include support for patients to help make sense of the diagnosis, develop adaptive coping strategies and adjust to living with a chronic condition. The role of the psychologist would involve working with specialist CF

healthcare professionals through providing educative training, reflective practice and supervision.

1.0 INTRODUCTION

This study focusses on exploring the experiences of individuals diagnosed and living with cystic fibrosis (CF) in adulthood. This introductory chapter reviews the current literature around CF in adulthood and childhood and subsequently considers the research in chronic diseases. The chapter follows on to build the case for this study and concludes by stating the study aim and research questions.

1.1 Overview of cystic fibrosis

CF is a progressive autosomal genetic disease affecting over 10,500 people in the United Kingdom (UK) (CF Trust, 2019). Approximately 1 in 25 people are carriers of the recessive gene making it one of the most common life-threatening conditions among the Caucasian population in comparison to other ethnic groups (CF Trust, 2015). It is a multi-system disease, mostly affecting the lungs and pancreas but can be found to impact other organs or parts of the body such as the liver, intestines, reproductive organs and sinuses. CF was first described clinically in children in 1938 (Anderson, 1938) and in 1989, 51 years later, the CF transmembrane conductance regulator (CFTR) gene responsible for CF was discovered (Kreindler, 2010). Mutations of the CFTR gene affect the transportation of salt and water across cell membranes, resulting in thickened mucus production. This leads to frequent respiratory infections, episodic intestinal obstruction, male infertility and pancreatic insufficiency which may lead to malabsorption, malnutrition and diabetes (Spoonhower and Davis, 2016). Reduced lung function is common in CF and is typically measured by the individual's forced expiratory volume in 1 second (FEV_1[1])

[1] FEV_1 is a measurement of the volume of air that a person can exhale during a forced breath in one second.

(Taylor-Robinson *et al.*, 2012). FEV_1 is the most important predictor of survival in CF (Liou *et al.*, 2010) and optimising lung function is the key aim for healthcare professionals (National Institute for Health and Care Excellence [NICE], 2017a). With over 1,400 different mutations of the CFTR gene (CF Trust, 2019), the characteristics, severity and trajectory of CF vary amongst the individuals affected by it, with respiratory failure being the most common cause of death (Berge *et al.*, 2007).

1.2 Diagnosis and management of cystic fibrosis

CF is primarily diagnosed via a sweat test and/or genetic testing (Simmonds and Bush, 2012). Although there have been advances in understanding the genetics and pathophysiology of CF, a cure is currently unavailable. Death in early childhood was common for those with CF (Estrada-Veras and Groninger, 2013). However, with early diagnosis, improved medications and treatment regimes, life expectancy has increased into adulthood. Since 2007, newborns between five and seven days old, have been routinely screened in the UK via a heel prick blood spot test, to identify the most common CF gene mutations. Following the introduction of this programme, infants have received an earlier diagnosis of CF in comparison to those born before 2007. In 2018, the median age of diagnosis for patients aged under 16 was 23 days (CF Trust, 2019). Children aged 5 years in 2016, who were born after the test was introduced, were commonly diagnosed within the first 3 months of life (93%) in comparison to children aged 10 years of age (66%), who were born before the screening programme was in place (CF Trust, 2017). The overall median predicted survival for people diagnosed with CF born between 2014 and 2018 is 47.3 years (CF Trust, 2019). This indicates that CF is no longer a childhood disease but instead

a chronic condition, requiring life-long and complex medical management (Badlan, 2006). Therefore, the long-term management of the condition has become increasingly important.

People with CF often undergo rigorous treatment regimes, usually daily, to maintain good health and control symptoms including chest physiotherapy, airway clearance, exercise, medication and nutritional monitoring (Bishay and Sawicki, 2016). This can often be time-consuming and complex (Sawicki, Sellers and Robinson, 2009). As per the nationally recognised standards of care, people with CF should have their care delivered by a multidisciplinary team of specialist doctors, nurses and allied health professionals at a recognised specialist CF centre[2] (CF Trust, 2011). Children should either receive full care from a specialist paediatric CF centre or shared care within an agreed designated network (CF Trust, 2011). With the increasing complexity of CF in adulthood, full care is recommended to be delivered by a specialist adult CF centre unless exceptional circumstances dictating that adults could receive care through a network CF clinic (CF Trust, 2011). New approaches for the treatment of CF, for example gene therapies, are showing encouraging findings (Alton *et al.*, 2015). However, there is still refinement and experimentation needed before this could be offered as routine treatment. At present, there are promising new medications available for people with specific gene mutations, which are improving people's lung function and quality of life (Condren and Bradshaw, 2013; Vertex Pharmaceuticals Incorporated, 2012). However, in spite of these developments, survival is still

[2] Several requirements need to be met to be referred to as a specialist CF centre such as a sufficient number of patients (a minimum of 100 adults or children), a core multidisciplinary team of trained and experienced CF specialist health professionals and recommended staffing levels, which must be of appropriate number for the size of the patient population. Further information can be found at CF Trust (2011).

dependent on frequent hospitalisations and a lifetime regimen of medication and therapies.

1.3 Cystic fibrosis in adulthood

Although most cases of CF are diagnosed in early infancy or childhood, a small percentage are diagnosed in adulthood. According to the 2018 CF Trust registry data, 834 adults (8.5% of all people with CF in the UK CF Trust registry) were diagnosed with CF at aged 16 or over, with 15 people aged 16 or over newly diagnosed that year (CF Trust, 2019). The first reported case of an adult CF diagnosis was in 1946 (Hellerstein, 1946). Since then an increasing number of case studies involving people diagnosed with CF in adulthood have been documented (De Moraes Júnior *et al.*, 2017; Naderi *et al.*, 2014; Schram, 2012).

There are multiple reasons why a person may not be diagnosed until adulthood. These include: 1) mild or absent symptoms, which may delay a patient accessing medical support, 2) unusual presentation, meaning a medical professional may not suspect CF, 3) the common misconception that CF is a childhood disease and 4) tests (such as the sweat test) presenting normal or borderline results (De Moraes Júnior *et al.*, 2017). It was often previously thought that the adult diagnosis was as a result of a ‘missed’ diagnosis from childhood. However, there is a growing consensus that it results from CFTR mutations with residual function leading to a delayed onset and reduced disease severity (Nick and Nichols, 2016). Individuals diagnosed in adulthood often have a milder form of the disease due to having a less common genetic mutation (Keating, Liu and Dimango, 2010). Improved diagnosis has been made possible by clearer diagnostic criteria, CFTR mutation testing and

awareness by doctors that a diagnosis of CF in adulthood is possible (Santos *et al.*, 2017).

Individuals who receive an adult CF diagnosis differ medically and psychosocially from patients diagnosed as children. Nick *et al.* (2010) found that people diagnosed in childhood, who had survived to age 40 years, had more severe CFTR genotypes and phenotypes in comparison to individuals diagnosed in adulthood. Although, after the age of 40 years, the rate of FEV_1 and death from respiratory complications did not differ between those diagnosed in childhood or adulthood (Nick *et al.*, 2010). Gender differences in CF expression of those diagnosed in adulthood have been reported (Widerman *et al.*, 2000). Patients receiving an adult diagnosis were more likely to be female, married, college graduates and employed full time in comparison to those diagnosed as a child (Nick and Nichols, 2016; Widerman *et al.*, 2000). Those diagnosed in adulthood were more likely to display fewer complications, fewer hospitalisations, less oxygen use, fewer courses of home IV (intravenous), less enzyme use, less pancreatic insufficiency, better lung function and a longer life expectancy compared to those diagnosed earlier in life (Widerman *et al.*, 2000). They were most frequently diagnosed following a respiratory condition (Widerman *et al.*, 2000) and likely to present with other CF complications such as male infertility, intestinal obstruction, pancreatitis and chronic sinusitis (Gilljam *et al.*, 2004).

Despite the milder form of the disease, anecdotally, healthcare professionals have noted that adults diagnosed with CF tend to have difficulty coming to terms with their diagnosis and are slow to begin treatment (Widerman, 2004). It was reported that attempts to engage adult patients, to provide education or support were met with

perceived indifference or withdrawal. This could be because the information and support needs of adults diagnosed with CF have not yet been adequately understood or met (Widerman, 2003). With the prevalence of the neonatal screening programme, it is expected that over time the previously unidentified CF patients who reach adulthood will decrease (Hadjiliadis, 2011). However, with various mutations and residual function of the CFTR gene, adult diagnosis of CF will continue to occur in the future.

Living with a chronic condition that involves complex lifelong care and treatment, places emotional demands on an individual. In a quantitative study, results from 154 CF centres in nine countries across Europe and the United States of America (USA), screening 6,088 patients with CF ages 12 years through adulthood and 4,102 parents, found high rates of depression and anxiety in adolescents and adults with CF and in parent caregivers (Quittner *et al.*, 2014). Raised symptoms of depression were found in 130 adolescents (10%), 913 adults (19%), 1,165 mothers (37%) and 305 fathers (31%). Anxiety was reported by 281 adolescents (22%), 1,503 adults (32%), 1,496 mothers (48%) and 343 fathers (36%). Overall, the elevations of depression and anxiety were two to three times more than community samples (Quittner *et al.*, 2014). Assessing psychological distress in individuals with CF is vital as data suggests that it affects both the disease management and health outcomes. It has been found to be associated with impaired health-related quality of life (Havermans, Colpaert and Dupont, 2008; Riekert *et al.*, 2007), poor lung function (Ploessl, Pettit and Donaldson, 2014), poor treatment adherence (Hilliard *et al.*, 2015), rehospitalisation and increased healthcare costs, lower body mass index (Snell *et al.*, 2014) and relationship difficulties (Yohannes *et al.*, 2012). These studies

emphasise the importance of screening and providing suitable interventions for mental health issues in patients and families with CF. In doing so, it provides the opportunity to improve the quality of life and health of individuals with CF and their parents. It is worth noting that these studies have not specifically stated whether they included individuals diagnosed with CF in adulthood as part of their studies, therefore the understanding of psychosocial issues within the population group is unclear.

1.4 Experiences of being diagnosed and living with cystic fibrosis in adulthood

Diagnosis of CF in adulthood is a unique experience (Widerman, 2005), where individuals can experience short and long-term psychological effects. To date, there have been few published qualitative studies exploring the experiences of adults diagnosed with CF (Blunt *et al.*, 2008; Widerman, 2005; 2004; 2002) and no known quantitative studies reported in this area.

In the most recent qualitative study within this field, Blunt *et al.* (2008) explored the psychosocial impact of an adult CF diagnosis within the UK population. This is the only known research carried out within the UK and was available as a published abstract from a conference presentation, reporting limited details regarding the study. Attempts to contact the research team to access the study findings were unsuccessful. From the available abstract, semi-structured interviews were undertaken with 10 adults and the results yielded various topics including diagnosis process, information needs, psychosocial support, social adaption, comparison to others and past self, loss, identity, emotional impact and coping. Patients reported frustration with past medical contact and time to diagnosis as a key theme. However,

once diagnosed they felt relieved and reported that the consultation was well delivered. At diagnosis, the participants felt overwhelmed by the information given to them. It was felt that it often focused on the negative aspects of CF rather than empowering the individual. Most of the participants felt that they needed psychosocial support including finance, employment information and counselling. Participants reported feelings of isolation as CF was publically perceived as a 'child's disease'. However, they referred to themselves as lucky in comparison to those with child onset CF. Social support was sometimes limited, with some family members distancing themselves due to feelings of guilt or the genetic implications of CF. Blunt *et al.* concluded that an adult CF diagnosis required close follow up and psychosocial support to enable adjustment and acceptance of the diagnosis. Areas such as fertility, employment and financial support were recognised as areas that might be important for discussion with the patient. The findings reported by Blunt *et al.* are interesting and begin to provide a story of the experiences of those diagnosed with CF in adulthood. However, without access to the full research, it is difficult to provide further comments or critique.

With no published papers around the experience of adult diagnosis of CF within the UK, this study draws upon understandings of the adult CF diagnosis experience beyond. In the early 2000's in the USA, Widerman undertook research as part of a larger study exploring the lived experiences of adults who received a diagnosis of CF after age 20. The published research papers (Widerman, 2005; 2004; 2002) refer to the same participants but discussed different aspects of the research findings. In a phenomenological qualitative study by Widerman (2002), 36 participants were asked to describe their adult diagnosis experiences and preferences of having their

diagnosis communicated with them. Individuals reported coming to the attention of specialists when they had ongoing health problems such as coughing, diarrhoea or infertility, which did not respond to treatment advised by their family doctors or for treatment of complications common to CF such as bowel obstruction, haemoptysis and pancreatitis. Individuals reported that despite the specialists suspecting CF and recommending testing for it, they often anticipated that the test would come back negative. The specialists would reassure the individual not to worry about the outcome and in many cases explained that they were too old to have CF. After receiving a positive result, individuals were often left confused and questioned whether they had CF, as the specialists would want to reconfirm this diagnosis. Receiving a diagnosis was a significant life event for those diagnosed in adulthood. Terms such as ‘devastated’ and ‘traumatised’ were used to describe their immediate reaction (Widerman, 2002). Some individuals were angry at their doctors for failing to diagnose their condition, whilst some were angry with their parents or themselves for not taking their symptoms seriously and pursuing a diagnosis sooner. Others were in denial of the seriousness of the condition due to the lack of symptoms or because they had been told that they were ‘too old’ or ‘too young’ to have CF. Some were confused by the diagnosis in that they could not understand why they had CF as an adult, whilst others were not surprised or not upset at receiving the diagnosis. There was no explanation given as to the differences experienced.

In the 2004 qualitative paper, Widerman explored the pre-diagnosis experiences of the same 36 participants diagnosed with CF in adulthood. This study identified four pathways to diagnosis, which were determined by the extent that the individuals experienced pre-diagnosis symptoms and whether they had considered the

possibility of a serious disease. Adults diagnosed with CF had arrived at their diagnosis with very different experiences of both their physical health and the healthcare system. This pre-diagnosis experience affected their post-diagnosis experiences, as well as their readiness for education, treatment and support (Widerman, 2004). For example, in pathway one, participants were characterised with little illness and no awareness of symptoms. As these participants did not have any reason to believe they were unwell at diagnosis they described their experience as a shock. Since they felt well, many initially denied that they had CF, or they minimised the disease. In comparison, those on pathway four, who suspected that they had CF, felt relieved to be diagnosed. Therefore, they were more ready for patient education than those in pathway one, who had a sudden and unexpected diagnosis and needed some time to talk through experiences and to know that their healthcare professional understood what had happened to them.

The focus of the analysis for Widerman's (2005) paper were the themes arising from the adult diagnosis experience and understanding their educational and support needs and preferences following a CF diagnosis. Individuals described their lived experiences of receiving an adult diagnosis through 10 inter-related themes (Widerman, 2005). These included: 1) an increased awareness of death and shortened lifespan, 2) change in lifestyle and self-view to accommodate CF, 3) difference in comparison to "normals" and others with CF, 4) CF competing with typical life stage demands for attention, 5) lack of support and acceptance from families regarding the diagnosis, 6) intrusive impact of CF on daily life and future plans, 7) isolation and loneliness from reduced socialisation, 8) desire to maintain pre-diagnosis lifestyle, 9) altered sense of time with a shrinking future and 10)

uncertainty around the future. Gender-specific themes were identified including mothers reporting the difficulty of parenthood, men being devastated by sterility and homosexual men fearing rejection if their CF symptoms would be mistaken for the symptoms and treatment of AIDS. Themes around illness severity were also identified including the probability of early death, wanting sympathy or feeling self-pity and the importance of exercise for those who described themselves as mildly ill.

Whilst Widerman's (2005; 2004; 2002) research provides useful insight into adults' CF diagnosis experiences, methodological details of the study were unclear. It was difficult to decipher what format the participant data was presented and analysed from. Although the author stated that they read and reread initial interviews when describing the data analysis undertaken, there was no explicit mention of interview transcripts, that interviews were transcribed, or any indication of an audio recording device used. The researcher stated that they completed detailed case notes (recordings of reflections, insights and tentative interpretations) and field notes (recordings of questions, notions and reactions across cases) after each interview. Whilst case and field notes were compiled, it is unclear from the papers if the researcher critically examined their role, potential bias or influence during the research question formation, data collection, recruitment and through the analysis process. These are important considerations when thinking about quality and rigour in qualitative research (see section 3.6 Quality and rigour in qualitative research for further discussion).

1.5 Experiences of being diagnosed and living with cystic fibrosis in childhood, adolescence and adulthood

With limited research in the adult CF literature, the experiences of those diagnosed in childhood and living with CF were explored. Studies investigating the experiences of being diagnosed with CF in childhood have often been reported from the perspective of the parent (Havermans *et al.*, 2015; Carpenter and Narsavage, 2004). Diagnosis of CF was described by parents as a surreal, devastating and life-shattering experience (Jessup and Parkinson, 2010; Carpenter and Narsavage, 2004). Parents reported a range of emotions when they received the initial news of the diagnosis. Shock, fear and disbelief were commonly reported emotions (Jessup *et al.*, 2016; Jessup and Parkinson, 2010). Other feelings included guilt, powerlessness, isolation (Carpenter and Narsavage, 2004) and relief to finally have an explanation of their child's symptoms (Gjengedal *et al.*, 2003).

In a qualitative study, involving focus groups of 14 adults who were diagnosed in childhood and 8 parents of small children diagnosed with CF, it was reported that those who were diagnosed within the first two years after birth were too young to remember their diagnosis experience (Gjengedal *et al.*, 2003). Many of these individuals had heard their "diagnosis story" from their parents, particularly in cases where getting the diagnosis was a challenge (Gjengedal *et al.*, 2003). These participants reported that their parents had worried about their health when they were an infant, taking them to see a health professional who did not take them seriously (Gjengedal *et al.*, 2003). Parents felt that symptoms were brushed off as being benign and reported being told that everything was normal (Gjengedal *et al.*, 2003). In another qualitative study of 20 adolescents, many participants described

vague memories of CF related behaviours such as coughing, doing chest physical therapy and taking medications during early childhood (two to five years) (Christian and D'Auria, 1997). They reported their first awareness of CF occurring between the ages of six to eight years of which many recalled being surprised to find out they had a diagnosis and that they were "sick". For most, their awareness of having a CF diagnosis was triggered due to hospitalisation for a CF related illness (Christian and D'Auria, 1997).

In a qualitative study, involving nine family members in the USA, Carpenter and Narsavage (2004) described three stages in which families moved back and forth through following their child's CF diagnosis: falling apart, pulling together and moving beyond. After the diagnosis (the falling apart stage), families went on to the next phase. This was where families made the required lifestyle adjustments needed for their child with CF and attempted to return to a sense of routine and normalcy. This required families to keep daily routines, to find ways to do things better or more efficiently, to remain vigilant and to constantly adjust to new stressors. This sense of returning to normalcy or reducing differences was consistently reported across other studies (Jamieson *et al.*, 2014; Gjengedal *et al.*, 2003). Participants repeatedly stressed the importance of living a normal life, despite the cost they may have to pay being high in effort (Gjengedal *et al.*, 2003). For instance, parents were aware of the importance of physical activity and tried to motivate and encourage their children to participate in normal play with other healthy peers (Gjengedal *et al.*, 2003). Motivating small children who did not understand the importance of daily treatment was seen as difficult by parents (Gjengedal *et al.*, 2003). The persistent need to take medications, undergo physiotherapy and being hospitalised served as persistent

reminders for some of their incurable illness (Jamieson *et al.*, 2014) or was seen as inconvenient when trying to live a normal life (Gjengedal *et al.*, 2003). For some, treatments were seen as another indication of being different or weaker than their well peers (Jamieson *et al.*, 2014). To protect themselves from exacerbations, individuals had to take more socially related precautions, such as staying away from people with infections and smoke-filled rooms (Gjengedal *et al.*, 2003).

A systematic review exploring children's and adolescents' experiences of CF reported that relationships with others were impacted, particularly as frequent absences from school made it difficult to establish friendships (Jamieson *et al.*, 2014). Participants struggled with how to disclose their diagnosis to others. Many choosing not to tell their peers and teachers or hiding information about their CF to appear normal (Jamieson *et al.*, 2014). Individuals reported feeling excluded when peers avoided them due to their coughing or differences in their physical appearance (Jamieson *et al.*, 2014). Adolescents discussed how their CF cough was one of the most revealing behaviours drawing attention to their differences in school and how being unable to control their cough was embarrassing (Christian and D'Auria, 1997). Many talked of developing a private or social cough or avoided taking medications in public to appear normal and minimise negative social consequences (Jamieson *et al.*, 2014). Participants shared stories of being bullied or abandoned by others who thought CF was contagious (Jamieson *et al.*, 2014). Many participants spoke of embarrassment around their small physical stature (Jamieson *et al.*, 2014). Feelings of frustration and difference were exacerbated by symptoms of their CF such as weakness, fatigue and susceptibility to infections which made it difficult to engage in social and physical activities (Jamieson *et al.*, 2014). Adolescents used pacing as a

strategy to mask their CF symptoms allowing them to participate in physical activities and be seen as equally competent as other peers (Christian and D'Auria, 1997).

In Carpenter and Narsavage's final stage, 'moving beyond', families moved beyond the fear, the guilt and sense of powerlessness and had established normalcy, a sense of control and identified new ways of coping. The stage the families were in was directly influenced by the child's health and wellbeing, meaning that if the child had a setback they might revert to the second stage to readjust and achieve a state of normalcy. Parents and adults with CF talked about having good and bad days, this was particularly the case during ill health or the death of a CF friend, bringing the illness to the forefront (Gjengedal *et al.*, 2003). Parents and older children with CF reportedly adopted a philosophy of "that's life" which helped them make sense of their experiences (Jessup and Parkinson, 2010). Many parents tended to use downward comparisons reporting that there is always someone worse off as a coping strategy (Jessup and Parkinson, 2010). Participants were very conscious of their shortened life expectancy. Whilst not afraid of dying, participants were scared of the risk of serious complications before death (Gjengedal *et al.*, 2003). Rather than waste time worrying about the future, some chose to make the most out of their life (Jamieson *et al.*, 2014). Whilst some worried that they had wasted their time and felt frustrated at how their physical therapy robbed them of a limited resource (Jamieson *et al.*, 2014).

In a qualitative study, Cordeiro *et al.* (2018) investigated the experiences of 12 adults, with an average age of 26, living with CF in São Paulo, Brazil. This is the only known study to have researched the experiences of adults living with CF. The

participants had been living with the disease for an average of 20 years. They reported difficulties living with CF in adulthood, referring to fatigue, the challenges of performing daily living tasks, the need for multiple medications and the intensive time demand to take the treatment. This made it difficult for them to work, to complete studies and to engage in leisure activities, which often isolated them socially. Similar to the childhood and adolescent experience, participants talked about shame and embarrassment in relation to their cough and prejudice experienced from others. For some of the adults, CF was not the centre of their lives. They discussed their coping strategies and shared their positivity, optimism and resilience. Participants also talked about how the fear of death was present in their life and how worsening symptoms made them more fearful. They shared how they lived with the idea of having to have a transplant from childhood or adolescence, the distress experienced waiting for a transplant, the fear of undergoing surgery and the uncertainty of the outcome. The participants shared that after transplant they expected that they would have autonomy, independence, they will be able to get married, have a family, resume their studies, find a good job and undertake activities that gave them pleasure. The authors note that this study was undertaken with a specific group of adults living with CF in the same space, time and with similar cultural characteristics meaning that the transferability of the results is limited.

Overall, the findings from the children's literature have been valuable in raising awareness of what it is like to be diagnosed and to live with CF. However, as initially highlighted, the diagnosis experience is usually reported from the parents' perspective as opposed to the person diagnosed with CF. In the studies reviewed there were some methodological concerns. For example, in Gjengedal *et al.*'s (2003)

study, no information was shared about the recruitment method, making it difficult to ascertain the appropriateness of the sample and subsequently the data gathered to answer the research question. In Carpenter and Narsavage's (2004) research, focus groups were the initial stated method for data capture to describe the lived experiences of families caring for a child with CF. With a voluntary purposeful sampling method, nine family members participated in this study, of which only three could attend the focus group and the remaining six contributed to the study via written narratives. The researchers stated that detailed written responses were provided to the same open-ended questions asked providing comparable data to that collected to a focus group. However, one can argue that it is difficult to clarify meaning from written text in comparison to a focus group setting. It was also unclear whether the relationship between the researcher and participants was adequately considered throughout the research process as this had not been discussed in any of the studies (Jessup and Parkinson, 2010; Carpenter and Narsavage, 2004; Gjengedal *et al.*, 2003; Christian and D'Auria, 1997). In summary, although a lot is known about the experiences of children with CF, as a child, an adolescent and an adult, there is very little published research into the experiences of those diagnosed with CF in adulthood.

1.6 Cystic fibrosis patient information and support needs in adulthood and childhood

There has been a lack of research looking at the information and support needs for CF patients diagnosed in adulthood within the UK healthcare system. Specifically, researchers have not examined what support is currently provided, if this meets patients' needs, if this is acceptable to patients and healthcare professionals, and if

there are any barriers to access and delivery of this support. Access to the right information and support is a key element of effectively managing CF to attain optimal health and quality of life (NICE, 2017b). This can help to reduce anxiety, increase empowerment and confidence with managing symptoms, and care for someone with CF effectively (NICE, 2017b). Not having the right information or support can hinder people in making informed decisions regarding their treatment and care (NICE, 2017b).

Evidence from the studies conducted in the USA can be used to help build a picture as to what support might be beneficial or important to patients diagnosed with CF in adulthood. For example, participants wanted to feel known and cared for by their medical caregivers (Widerman, 2005) and preferred their CF physician being their primary source of information (Widerman, 2003). In a qualitative study involving 36 participants, more than half of the adults were unfamiliar with CF at diagnosis (Widerman, 2002) and after receiving their results, individuals were keen to find out more about the condition. However, their educational needs at the time of their diagnosis were often unmet (Widerman, 2005; 2003; 2002; Widerman *et al.*, 2000) either because the doctor was unfamiliar with individuals being diagnosed with CF in adulthood or because the answers were unknown (Widerman, 2002). Individuals were often left disappointed, annoyed, confused and frightened by outdated and irrelevant information, which reported a life expectancy shorter than their current age and with literature directed towards the parents of young children with CF (Widerman, 2002). Participants were less interested in the biomedical descriptions of CF but instead wanted to know information regarding life expectancy, its possible impact on their life in terms of changes, what to expect in the future, treatment

options, actions they could take, how they could maintain normalcy, research directions and information that would help them to build hope (Widerman, 2005). Those less positive about their diagnosis experience reported that their emotional needs were not addressed, that they were treated impersonally and that they did not receive sufficient information (Widerman, 2002).

In a mixed method study, Widerman (2003) aimed to address the gap regarding the actual and self-perceived knowledge of those diagnosed as adults. This intended to inform the development of educational materials for this population group and to guide healthcare professionals. A questionnaire containing predominately fixed response questions with some open-ended questions was completed by 130 participants from the USA, Europe and other nations with a response rate of 74.3%. Consistent with the findings of Widerman (2002), over two-thirds (67.4%) of respondents reported that they knew little (45.7%) or nothing (21.7%) about CF at diagnosis. Individuals were asked to assess their CF knowledge within the first six months and over one-third stated that they still knew little (30%) or nothing (3.9%), with 7.9% reporting that they were uninterested in CF information at or post-diagnosis. Of the respondents, 93 participants (71.5%) indicated that they received information about CF at diagnosis, from which, 25 individuals (26.9%) responded that they received ‘too little’, 50 individuals (53.8%) stated they received an ‘adequate amount’ and 18 individuals (19.4%) received ‘a lot’. In total, 98 respondents (75.4%) stated they searched for information about CF following their diagnosis but only 30 respondents (30.6%) reported that they were ‘satisfied’ and 9 respondents (9.2%) were ‘very satisfied’. Whilst the data produced from this study was insightful, the method used was not the most appropriate. Instead, a qualitative

semi-structured interview would have been better suited to answer the research question. Also, to note, as the recruitment of the study was predominantly via the internet, individuals that participated were from a volunteer convenience sample, who were information seekers and therefore might be over-represented in the sample. Lastly, the mean time since diagnosis was almost nine years and as the data was self-reported, the time since diagnosis may have affected participants' recall. However, the researcher argued that the open-ended comments from the participants did not reflect difficulty recalling the diagnosis or their information needs at the time.

Widerman (2004) advised for CF healthcare professionals to be aware of patients' pre-diagnosis experience as this would impact on their engagement with the service, their intake of information and their adherence to treatment. Therefore, CF staff were encouraged not to adopt a one-size-fits-all approach. Widerman stated that patients "should be asked how they came to be diagnosed and how they are reacting to their diagnosis" (2004, p.76) and their responses would indicate their needs and could guide the support that healthcare professionals provide. For example, patients whose stories expressed emotions such as anger, confusion, depression, shock or fear should have these emotions addressed by the CF team either before or alongside their clinical care (Widerman, 2004). Whereas individuals who expressed relief or vindication were more inclined to begin their self-care, treatment and be ready to learn more about CF (Widerman, 2004).

With limited research into the information and support needs of patients diagnosed in adulthood, the childhood CF literature was also explored. At the initial diagnosis

phase, parents' narratives referred to the need to fight for information (Jessup and Parkinson, 2010). The majority of parents had lacked pre-diagnosis knowledge of CF and subsequently had to acquire a substantive amount of information (Jedlicka-Köhler, Götz and Eichler, 1996). Parents described various degrees of coping with information. Some parents recounted wanting information instantly and to know as much as possible, whereas others reported receiving too much information, particularly at the time of their child's CF diagnosis, finding this overwhelming (Jessup *et al.*, 2016). This was consistent with findings from a quantitative study undertaken in Austria on parents' emotional and cognitive reactions to their child's CF diagnosis (Jedlicka-Köhler, Götz and Eichler, 1996). The authors stated that parents received a perceived lecture when being told about the CF diagnosis, containing more information than they could assimilate (Jedlicka-Köhler, Götz and Eichler, 1996). Whilst information on how to care for their child was important, the emotional aspects were more of a concern at the time of diagnosis, in comparison to the physical aspects of care (Carpenter and Narsavage, 2004). In all but one case (45 parents), the child's diagnosis was communicated orally without any written educational or illustrated material and 43% stated the explanations given by doctors included medical terms that they were unable to understand at the time (Jedlicka-Köhler, Götz and Eichler, 1996). Parents who reported initial feelings of shock during conversations with healthcare professionals were found to have understood less and retain less information than parents who had not had that kind of stress (Jedlicka-Köhler, Götz and Eichler, 1996). This was because of the incompatibility of the emotional distress and optimum learning leading to difficulties with comprehension. Parents found that the most threatening information was regarding reduced life expectancy and incurability (Jedlicka-Köhler, Götz and Eichler, 1996). Subsequently,

Jedlicka-Köhler, Götz and Eichler made the recommendation for recurrent contact with both parents and the provision of written and audiovisual materials as mandatory.

A qualitative study in a paediatric hospital in Australia was undertaken to understand 10 parents' experiences of initial education following their infant's CF diagnosis from newborn screening (Jessup *et al.*, 2016). Jessup *et al.* reported that parents preferred a short period, for example 48 hours, between being informed of the diagnosis and the education period. A prolonged timeframe led to parents searching for information, often via the internet. For some, this led to regret and fearfulness of the information found, whereas for others this was a neutral or positive experience, allowing them to be more informed and prepared. The majority of parents wanted practical information regarding aspects of the disease or treatment that they could engage, modify and influence. Conceivably, above all else, parents sought information that offered some reassurance and hope. Jessup *et al.* concluded that information needed to be individual in relation to content and pace, practical, timely, culturally relevant, current, conversational and considered through parents' eyes. However, a review of this study highlighted that the research was conducted by multiple researchers and the level of consistency was not reported. Furthermore, the researchers' role and potential influences in the analytical process were not critically reviewed.

Another qualitative study explored the information needs of parents of children with CF prior to their first hospital admission within the UK (Fixter *et al.*, 2017). Data was reported from eight mothers and two parent couples whose children had been

admitted for routine IV antibiotics. Parents unanimously reported receiving limited information, which was mostly medical and practical in nature, with minimal emphasis on the psychological implications of the admission (Fixter *et al.*, 2017). Parents stated that they would have appreciated receiving comprehensive information as they felt this would have enabled them to be better equipped to cope with their child's hospitalisation (Fixter *et al.*, 2017). All parents shared their need for involvement in their child's care and for most parents, feeling in control was important. Relinquishing control to the medical team was perceived by some parents as frustrating (Fixter *et al.*, 2017). The majority of parents interviewed for this research were White British mothers. It is possible that different issues would have presented within the findings if more fathers had participated.

1.7 Experiences of being diagnosed and living with a chronic disease

With limited good quality published research on CF to date and a paucity of research into the experiences of diagnosis in adulthood, the chronic illness literature was reviewed. Chronic diseases are defined as non-communicable illnesses, which endure over time, are usually slow to progress and can have the potential to significantly impact an individual's everyday life (World Health Organisation [WHO], 2018; Moss-Morris, 2013). Chronic diseases have been categorised into four main types: cardiovascular diseases (such as stroke and heart attacks), cancers, chronic respiratory diseases (such as asthma, chronic obstructed pulmonary disease [COPD] and CF) and diabetes (WHO, 2018). Whilst these illnesses may not be equivalent to CF because of clinical features, there are many comparable aspects. For example, many of these conditions can be diagnosed in adulthood, are life-threatening, require an individual to make significant lifestyle changes including

undertaking complex medication routines and can erode the quality of life. The key difference between CF and other chronic conditions is that CF is a genetic condition and although the life expectancy for CF is increasing, individuals still have to face the possibility of an early death (Gjengedal *et al.*, 2003).

For some, the process of arriving at a diagnosis of a chronic condition was a challenge. Cancers, such as non-Hodgkinson's lymphoma (NHL), were often diagnosed when the cancer was at an advanced stage (Wall, Glenn and Poole, 2011). Similarly, COPD was repeatedly underdiagnosed or misdiagnosed and often not identified until the disease had significantly advanced (Price, Yawn and Jones, 2010). In a systematic review of the experiences of patients living with COPD, individuals tended to access treatment for an acute illness episode rather than chronic symptoms of their health (Giacomini *et al.*, 2012).

Participants described a range of emotional reactions when diagnosed with their chronic disease (Chircop and Scerri, 2017; LeBlanc *et al.*, 2016). Shock was a commonly used term to describe individuals' diagnosis experience with various chronic illnesses including cancers such as acute myeloid leukemia (AML) (LeBlanc *et al.*, 2016), NHL (Chircop and Scerri, 2017) and breast cancer (Liamputtong and Suwankhong, 2015), COPD (Ansari *et al.*, 2014) and diabetes (Silva *et al.*, 2018; Due-Christensen *et al.*, 2018). Some participants experienced denial (Silva *et al.*, 2018; Ansari *et al.*, 2014). In Chircop and Scerri's NHL study, some participants expressed disbelief when they were given their diagnosis, particularly as these individuals had experienced some generic symptoms which they did not associate with cancer. Whilst the diagnosis offered participants a label and explained the

symptoms they had experienced, the diagnosis was unexpected. In contrast, one participant felt relieved that she was diagnosed with cancer as the pain she had been experiencing for several months was acknowledged and she was able to begin the appropriate treatment. This was similar to findings from a meta-synthesis of adaption to life following a new type I diabetes diagnosis in adulthood (Due-Christensen *et al.*, 2018). The findings reported that some individuals described their diabetes diagnosis as positive, particularly initially, as taking insulin reduced their symptoms. Others described the onset of their diabetes symptoms as insidious and confusing, leading to feelings of uncertainty of what was happening to them (Due-Christensen *et al.*, 2018). The psychological response to a diagnosis was often expressed as feelings of loss and grief for the person's previous pre-diagnosis life (Due-Christensen *et al.*, 2018). Individuals' response to their diagnosis was influenced by how it was communicated and the type of support that was provided (Due-Christensen *et al.*, 2018). The diagnosis was perceived as a major disruption in regards to the physical, psychological and social aspects of life, particularly concerning their aspirations for the future and life trajectories (Due-Christensen *et al.*, 2018). These findings were consistent with research from the heart failure literature, in which participants reported that the diagnosis and living with their chronic condition led to changes in various aspects of their lives including work, lifestyle and sex life (Paturzo *et al.*, 2016). Studies have reported the negative impact of symptoms on present employment and future work opportunities (Due-Christensen *et al.*, 2018).

Similar to Due-Christensen *et al.* (2018), Bury (1982) proposed that peoples' experiences of chronic illness were conceptualised as a biographical disruption with

a key element being its insidious onset. This theory helps to understand the individual's initial diagnosis experience. Bury stated that "non-communicable diseases do not 'break-out' they 'creep-up'" (1982, p.170). He explained that there was a contrast between their experiences and the common cultural paradigm of the disease, often resulting in reactions of shock, disbelief and anxiety leading onto relief. The next stage was centralised around the emerging disability of the condition and the problem of uncertainty around the impact and the course of the condition, with participants searching for the meaning of events. Bury also discussed how access to medical information about the illness allowed individuals to make sense of the disease separate from their individual self. Lastly, the mobilisation of resources and the availability of social support networks was a key factor. However, the model has been criticised for two reasons by Larsson and Grassman (2012), based on interviews with people who have experienced chronic illness since childhood. Bury stated that a chronic illness constitutes a critical situation for a person, partially due to its unexpected nature. Whereas Larsson and Grassman argued that illness changes do not need to be unexpected to be experienced as disruptive. Larsson and Grassman also challenged the model's implied suggestion that a biographical disruption is a single event that occurs in the early stage of a chronic illness. They reported that biographical disruptions could occur repeatedly over the lifespan of chronically ill and disabled people.

Individuals diagnosed with AML described a mismatch between their informational needs and the type of information healthcare professionals provided (LeBlanc *et al.*, 2016). Some expressed a need for more information whilst others preferred less. In the cases of mismatch, participants expressed dissatisfaction. They described the

information provided by healthcare professionals on their chronic condition as insufficient (Due-Christensen *et al.*, 2018; Marx *et al.*, 2016) and expressed a need for relevant information from those who had personal experiences (Due-Christensen *et al.*, 2018). Many participants struggled with processing complex information about their diagnosis and treatment options due to the overwhelming shock and sadness (LeBlanc *et al.*, 2016). This subsequently had a negative impact on participants' understanding of available treatments and in some cases prompted them to defer decisions about their treatment to their doctor. Participants also tended to minimise the severity of their disease, as a result of denial, due to their interpretations from information given to them by their healthcare professionals (Marx *et al.*, 2016).

People's perception of the seriousness of their diabetes and the impact it had on their health varied: some minimised its significance whilst others perceived the condition to be more serious (Due-Christensen *et al.*, 2018). For some, their diabetes took over their sense of self and lead to a preoccupation with it, whereas others saw their disease as external to them (Due-Christensen *et al.*, 2018). Participants considered their life differently, some reported feeling more responsible whereas others lived by the carpe diem philosophy (Saylor, Hanna and Calamaro, 2019). Individuals reported a continual struggle of finding the right balance between spending all of their time on managing their treatment or spending too little time on their self-management (Due-Christensen *et al.*, 2018). Some perceived that the structure of life and work was a positive factor for adapting to a life with diabetes (Due-Christensen *et al.*, 2018).

Symptoms of people's conditions varied and could be unpredictable. People diagnosed with COPD reported having good days and bad days, which impacted their ability to undertake basic activities in their day-to-day living (Giacomini *et al.*, 2012). Managing their chronic condition involved constant planning and balancing to include the demands it put on their life (Giacomini *et al.*, 2012). Individuals would try to maintain their daily life for as long as possible, which enabled them to cope with feelings of helplessness when they felt at the mercy of their disease (Marx *et al.*, 2016). By continuing with daily life, it made it easier for patients to deny the threat to life and the progression of their disease (Marx *et al.*, 2016). Individuals reported that their diagnosis motivated them to make lifestyle changes, including becoming more active, improving their diet and stopping smoking (Ansari *et al.*, 2014).

Individuals frequently described uncertainty around their prognosis and recognised that death was inevitable (LeBlanc *et al.*, 2016; Paturzo *et al.*, 2016). In some studies, religion or belief in God gave many people inner strength and emotional support (Paturzo *et al.*, 2016; Liamputtong and Suwankhong, 2015).

In several studies, participants discussed social isolation and the impact on their lives (Due-Christensen *et al.*, 2018; Marx *et al.*, 2016; Paturzo *et al.*, 2016; Ansari *et al.*, 2014; Giacomini *et al.*, 2012). With conditions such as COPD and heart failure, social isolation was experienced due to physical limitations of the condition such as breathlessness, which affected individual's mobility (Marx *et al.*, 2016; Paturzo *et al.*, 2016; Ansari *et al.*, 2014; Giacomini *et al.*, 2012).

Studies reported social disruptions in terms of relationships with others and the context of work following diagnosis (Due-Christensen *et al.*, 2018). Some family

members distanced themselves from the person with the illness (Giacomini *et al.*, 2012). Participants experienced poor relationships with healthcare professionals, including reports of lack of compassion, poor listening, hastiness and feeling their distress was ignored by those who focused on objective health measures (Giacomini *et al.*, 2012).

The experiences of those diagnosed with a chronic condition can be explained by two models from the chronic illness literature: the shifting perspectives model of chronic illness (Paterson, 2001) and a working model of adjustment to chronic illness (Moss-Morris, 2013). The application of these models is discussed later (see 5.4 Models and theories in chronic illness literature). Based on a meta-synthesis of 292 qualitative research studies, the shifting perspectives model of chronic illness (Paterson, 2001) reports on the experiences of adults with chronic illness. The model attempts to describe the experience of chronic illness and explain why individuals with chronic illness may engage in behaviours which seem illogical, ill-advised or possibly harmful. Paterson (2001) offered an alternative explanation to that of previous researchers, who described chronic illness as a phased process with a predictable trajectory from diagnosis to mastery or acceptance. Instead, the shifting perspectives model proposed that living with chronic illness is an ongoing and constantly shifting perspective, a complex dynamic between the individual and their environment. Individuals' perspective of the illness defined how they responded to the disease, themselves, health professions, others with whom they had a relationship with and situations that are affected by the illness. Each perspective contained elements of illness and wellness that shifted according to whether illness or wellness were in the experiential "foreground" (see Figure 1). The illness in the

foreground perspective focused on the symptoms, loss, burden and the potential destruction of the disease. Paterson (2001) explained that individuals adopted this perspective to maintain their identity as a sick person. The wellness in the foreground perspective involved appraisal of the illness as an opportunity for meaningful change, with the attention focused on the self, not the diseased body, as the source of identity. Paterson stated that there are several ways that participants may adopt this perspective, particularly through learning about the disease, sharing their knowledge of the disease with others, creating supportive environments and recognising their body's response to the disease. Other reasons that would shift individuals from wellness to illness in the foreground included disease progression, limited skills to manage the disease, stigma related to the disease and interactions with others that emphasised dependence and hopelessness.

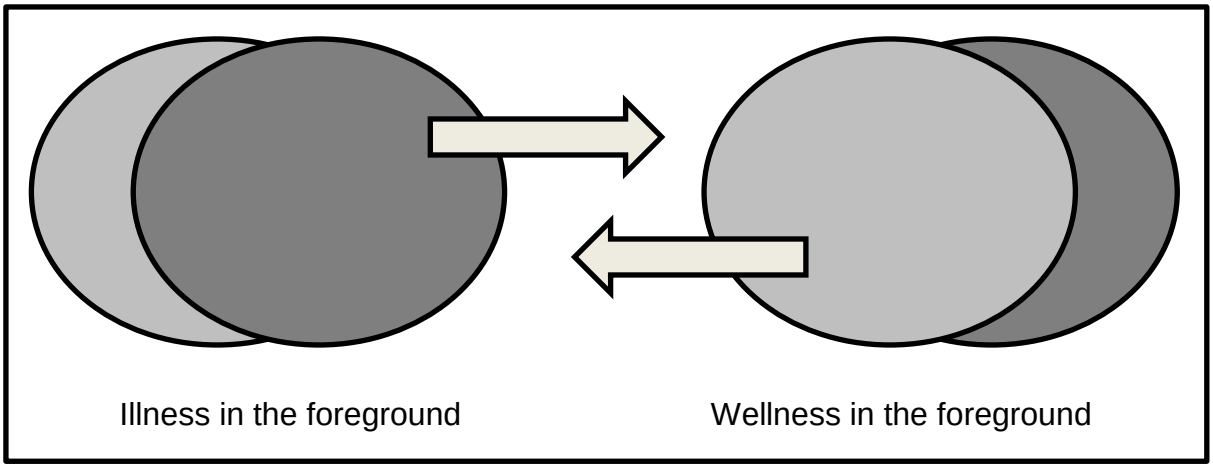

Figure 1: The shifting perspectives model of chronic illness (Paterson, 2001)

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The working model of adjustment to chronic illness (Moss-Morris, 2013) suggests that personal background factors and social and environmental factors influence how people respond to and adapt to illness stressors. Illness-specific factors determine

the extent to which illness stressors or critical events potentially disrupt emotional equilibrium and current quality of life. The adjustment process for critical events requires returning to equilibrium or in the case of chronic stressors requires maintaining equilibrium. Whilst the model recognises that adjustment to new critical illness-related incidents might be an ongoing process, how adjustment changes and whether adjustment outcomes might change, was not explicated (Bogosian *et al.*, 2017).

1.8 Conclusion

All of the known published research investigating the experiences of people diagnosed with CF in adulthood has been carried out in the USA. The previous research was undertaken by one researcher in the early 2000s to inform social work practice. Whilst this sheds light on the patient experience, there is a pressing need to understand the experiences of those receiving their care within the UK, particularly since the healthcare service provision differs. For example, within the UK, integrated psychologists are working within UK CF centre multidisciplinary teams. In addition, the UK CF Trust (2011) standards recommend that psychological support is provided around diagnosis for adults. However, in the USA, social workers are required as part of the core team with psychologists considered a recommended team member and with no specific recommendation for psychological support around adult diagnosis. According to the CF Foundation[3], the patient may see their CF social worker for emotional, social and financial support around living with CF, coping with stress and impact on relationships with friends and family, and support around health insurance cover (CF Foundation, 2018). The CF social worker may refer a patient to

[3] Non-profit organisation in the USA.

a psychologist (CF Foundation, 2018), thus suggesting that the access to psychological support at diagnosis is different in both countries and therefore, the experience might be different.

Since the early 2000s, there has been a growing awareness of CF diagnosis in adults within the healthcare system and there have been developments in the treatments and care offered. Subsequently, there may have been a change in process for making a diagnosis, protocols for care or treatment, which may impact on the patient experience. Adult diagnosis is a growing area for CF departments within the UK. It was identified as an area requiring further research by the UK CF psychosocial group and the regional adult CF centre for which this research was undertaken. Adult CF services were keen to have guidance on how to best support this patient group, especially with the drive within the National Health Service (NHS) to listen to the patient voice and make improvements (Patient Voice South, 2018). Most of the focus of previous research has been on the diagnosis experience with little attention given to the understanding of the ongoing experiences of those living with an adult diagnosis of CF and the implications of having a chronic illness. This research intends to address this gap in the literature.

This research aims to explore and understand the experiences of people diagnosed and living with CF in adulthood within the UK. These findings are important for healthcare professionals providing treatment for those diagnosed in adulthood to ensure that the appropriate care is provided, which is sensitive and comprehensive. Specifically as healthcare professionals providing care for adults rarely encounter undiagnosed CF and, unlike those working in paediatric services, they will have little

experience in communicating the diagnosis (Widerman, 2002). This work aims to inform healthcare professionals about the psychosocial impact of CF, thereby informing the delivery of psychosocial support, as recommended by Blunt *et al.* (2008). The findings will also be beneficial for individuals who are living with and who have been newly diagnosed with CF in adulthood and their families who have little information to validate their experiences or answer their questions. This research will explore the issues and psychological needs faced by patients within this population group and gain wider insight into how people diagnosed with CF as adults adjust to living with their disease. The research aims to provide recommendations to clinically support patients diagnosed with CF in adulthood.

1.9 Research aim and research questions

To understand the lived experience of receiving an adult CF diagnosis and what it means to live with the condition.

Research questions:

1. What is it like to be diagnosed with CF in adulthood?

2. What impact does CF have on an individual's day-to-day life?

3. How do individuals adjust to living with CF in adulthood?

2.0 METHODOLOGY

The following chapter describes the methodology of this study. This is achieved through the discussion of the research design, the researcher's theoretical position, data collection methods, method of analysis, researcher reflections and the interview design. The next chapter refers to the method of the research, including the recruitment process, data analysis, quality and rigour within the research and ethical considerations.

2.1 Research design

A qualitative design was used to explore the experiences of adults diagnosed and living with CF. Using a qualitative approach allowed for the exploration of areas that may never have been anticipated, a priori in an under-studied field (O'Cathain *et al.*, 2014). Qualitative research can be categorised into two areas: experiential and critical (Braun and Clarke, 2013). Experiential qualitative research looks at the views, perspectives, experiences and practices expressed by participants. Adopting this approach, participants are asked to share their reality allowing the researcher to access and make sense of their world. This principle assumes that participants can clearly articulate their experiences and that it makes sense to the researcher. This research project assumed an experiential approach to explore and understand the experiences of those diagnosed with CF in adulthood.

2.2 Theoretical position

Defining the researcher's ontological and epistemological position is crucial in qualitative research as it shapes the approach taken to explore the phenomenon under investigation. It demonstrates quality and rigour within the research through

consistency between the research aims and the underlying paradigmatic assumptions (Haverkamp and Young, 2007). Ontology relates to the relationship between the world and human practices (Kivunja and Kuyini, 2017), specifically whether or not reality is considered to exist independent from human interaction or whether it is constructed through human interpretation. Within qualitative research, there are a range of ontological positions that can be adopted by the researcher, from naive realist to radical relativist (Madill, Jordan and Shirley, 2000). On one end of the continuum, a realist approach assumes a single reality, which can be revealed through research by using the 'correct' methodology (Braun and Clarke, 2013; Madill, Jordan and Shirley, 2000). On the other end of the continuum, a relativist position posits that reality is constructed by interpretation. This position adopts the perspective that there is not just one correct version but instead multiple versions of reality. For this study, a critical realist position was adopted, which sits in between the realist and relativist positions. This was chosen because, from a critical realist perspective, reality holds some 'truth' to produce knowledge to make a difference. This was particularly important to be able to feedback the study findings to inform future practice. However, this reality can only be partially accessed due to individuals' perceptions of reality being subjective and socially influenced, which can vary over time (Willig, 2013).

Epistemology is a branch of philosophy; it is our perceived relationship with the knowledge being uncovered, in particular, what can be known and how meaningful knowledge can be created (Willig, 2013). Similar to ontology, epistemology approaches fall on a continuum from positivism to constructionist. Positivism assumes that there is a direct relationship between what we observe and our

perception of it. It posits that through the application of scientific methods, objective and unbiased data collection, valid knowledge can be obtained. This knowledge will provide the only version of 'truth'. On the other end of the continuum, constructionism sees that knowledge is contextually created, meaning that it accepts an inherent subjectivity of the individual and researcher bringing one's personal and cultural perspectives into the research project (Madill, Jordan and Shirley, 2000). In this study, a contextualist position was chosen. Similar to critical realism, contextualism sits in between positivism and constructionism. In summary, a critical realist, contextualist approach was adopted to explore individuals' experiences of being diagnosed and living with CF in adulthood.

2.3 Data collection

Interviews and focus groups were considered as possible methods of data collection. Interviews are characterised by the researcher asking questions and probes to encourage the participant to talk about a particular topic (Howitt, 2010). Focus groups are an interactive discussion between multiple participants guided by a moderator around the topic of interest (Hennink, 2014). Focus groups were not deemed a suitable method for several reasons. Firstly, focus groups are useful when collecting information about people's attitudes towards a concept, product or idea (Leung and Savithiri, 2009), rather than individual experiences as per the aim of this research (Hennink, 2014). Secondly, an issue with focus groups is that the participants are likely to compete to tell their story. The stories are unlikely to unfold sequentially making the presented picture confusing and difficult to follow (Barbour, 2007). In addition, within a group setting, some individuals may feel reticent or uncomfortable with sharing personal experiences (Hennink, 2014). Thirdly, an

important consideration for this particular study is the fact that contact with other individuals with CF could lead to cross infection, which could be harmful to the individual's health. Lastly, as the participants were geographically dispersed across a region, the logistics of arranging a focus group made it impractical and it may have been potentially inconvenient for participants to travel. For these reasons, face-to-face interviews were chosen as the data collection method. Participants were offered the opportunity to arrange their interview before or after their next clinic visit to avoid travel inconveniences. Alternative forms of interviewing that were presented to potential participants are discussed further (see 2.3.2 Telephone and virtual interviews).

2.3.1 Interviews

Interviews are the most suitable method when little is known about the research topic or when detailed insight is required from individual participants (Gill *et al.*, 2008). They are seen as an appropriate way of gaining insight and collecting rich and detailed data about the participants' experiences and perspectives (Oltmann, 2016) or how participants attribute meaning to those experiences (Forrester, 2010). Interviews are a form of social interaction in which the participant and researcher co-construct the data that is produced.

Face-to-face contact between the researcher and participant is typically seen as the ideal way to collect interview data (Braun and Clarke, 2013). It is argued that a face-to-face encounter is essential for an interview which is flexible, interactive, generative and where meaning and language are explored in-depth (Legard, Keegan and Ward, 2003). A skilled interviewer can build rapport with their participants to help

them feel comfortable, enabling them to share personal information, which may not be captured by other data collection methods such as questionnaires. Interviews can provide a therapeutic and rewarding experience for the participant in comparison to questionnaires, observations and experiments, allowing the participant to talk about their experiences whilst the researcher listens without being critical (Denscombe, 2007). Interviews allow for the discussion of sensitive and personal issues, particularly where participants may not want to talk about issues in a group. It allows for the checking of accuracy and clarifying of meaning with the participant as the data is being collected. Non-verbal expressions conveyed by the participant can be observed. This would not be possible using other methods such as telephone interviews or written text (Opdenakker, 2006). The potential for telephone and virtual forms of interviewing is considered below (see 2.3.2 Telephone and virtual interviews).

The challenges of using interviews as a data collection method are mainly concerning time. For the participant, an interview often takes at least an hour of their time to complete (Braun and Clarke, 2013) and may involve additional travel time. Interviews are also only beneficial if the participant has sufficient insight into the topic area and/or their actions in order to reflect and articulate them. For the researcher, they can be time-consuming to organise, travel to, conduct, transcribe and analyse (Braun and Clarke, 2013). The researcher also needs to find a suitable location to undertake the interview allowing for privacy. Another potential hindrance for face-to-face interviews is the need to audio record the interview to collect the data. This may interfere with the interaction between the participant and researcher, as they may not be entirely relaxed or comfortable with the audio recording device (Willig, 2013). To

overcome this issue in the current study, a relationship with the participant was established to help them feel at ease, through informal conversations when arranging the interview and before the interview began. The placing of the audio device was considered and care was taken to avoid placing it in the participant's eye line.

2.3.2 Telephone and virtual interviews

Telephone interviews are increasingly being used in qualitative research (Oltmann, 2016; Ryan, Coughlan and Cronin, 2009). They are a cost-effective data collection method in terms of time and money, whilst enabling interviews across a wide geographical area (Howitt, 2010). Telephone interviews may allow the opportunity for participants to be interviewed who are reluctant to participate in face-to-face interviews or are from marginalised groups (Oltmann, 2016). They may also encourage participants to share sensitive information, which they might have been reluctant to discuss face-to-face (Opdenakker, 2006).

It is often assumed that telephone interviews are inferior to face-to-face interviews (Novick, 2008). This is because of the lack of non-verbal cues (Howitt, 2010), meaning that it may be difficult to assess the participant's emotional reactions (Sturges and Hanrahan, 2004). Other disadvantages include technology-related problems such as poor sound quality or dropped calls (Oltmann, 2016), as well as the potential for distractions for participants by activities in their own environments (Opdenakker, 2006). Although, these distractions were also reported during face-to-face interviews (Sturges and Hanrahan, 2004). Contrary to common assumptions of telephone interviews being seen as a second-best option in comparison to face-to-

face interviews, research suggests that this is not the case (Egan *et al.*, 2011). Egan *et al.*'s research (2011, p.747) showed that telephone interviews can "produce data equal to that obtained from face-to-face interviews". They found that although non-verbal cues were not visible, other forms of communication such as intonation, pauses and sighs helped with interpretation of the data. Their research also highlighted that "a mixture of methods of data collection can be an effective way to increase sample size without necessarily compromising on the quality of data" (Egan *et al.*, 2011, p.747).

Virtual video interviews involve the participant and researcher conducting an interview using video chat software such as Skype. Similar to telephone interviews they are inexpensive and can be convenient for participants who can speak from their own home or a location of their choice. This offers more accessibility for patients with physical disabilities, mobility issues or for those who are geographically dispersed (Braun and Clarke, 2013). Video interviews, much like face-to-face interviews, enable the researcher to see visual cues. However, they rely on participants to have access to an electronic device such as a computer, laptop or smartphone and be technologically savvy to use the software. For the reasons discussed above, both telephone and video interviews were also presented to participants as alternatives to a face-to-face interview. However, all of the participants opted for face-to-face interviews.

2.4 Method of analysis

To answer the research questions, interpretative phenomenological analysis (IPA) and thematic analysis (TA) were considered as methods of analysis. Both were

deemed as appropriate methods of analysis for exploratory studies (Braun and Clarke, 2013). IPA is concerned with understanding how individuals construe and make sense of their lived experience rather than produce objective knowledge (Smith, Flowers and Larkin, 2009). Usually involving a small number of participants, IPA focuses on identifying patterns of meaning from the detailed exploration of these individuals' subjective experiences and the group. TA is a method for researchers that seek to identify, analyse and report patterns within data (Braun and Clarke, 2006).

Although both IPA and TA could provide valuable insights into the experiences of adults with CF and despite the researcher's previous experience of using IPA as a method, TA was chosen as the most appropriate approach for this research study. As the research question looked to explore and describe the experiences of adults rather than understand their perception of the experience, this guided the decision to choose TA for this study. Unlike IPA which is wed to a phenomenological epistemology and is theoretically bounded, TA is not wed to any pre-existing theoretical framework allowing freedom and flexibility (Braun and Clarke, 2006). Another advantage of using TA as a method is that it is suited to larger sample sizes (Clarke, Braun and Hayfield, 2015; Braun and Clarke, 2006). Using TA also allowed the opportunity to broaden my skills as a researcher.

Due to its theoretical flexibility, several decisions need to be explicitly made, which often are not reported in academic papers (Braun and Clarke, 2006). This includes: 1) the type of analysis undertaken and the claims made related to the data set, 2) whether the themes identified from the data are inductive or deductive and 3)

whether the themes are identified at a semantic or latent level. In relation to the type of analysis undertaken, the reported themes are either a rich description of the entire data set, allowing the reader to get a sense of the key themes or a detailed account of one particular theme or group of themes within the data. Given that so little is known about patients' experiences of being diagnosed and living with CF in adulthood, a descriptive analysis of the whole data set was deemed appropriate to guide understanding of the range of participant experiences and future research. Themes or patterns from the data can be identified as inductive (bottom-up) meaning the themes generated were data-driven or theoretical/deductive (top-down) which is an approach driven by pre-existing theory. An inductive process was used within this research, again to allow new concepts to be identified in an under-researched area and guide future research and practice. The level at which the themes are identified can be semantic (explicit level), meaning the focus is on the surface of the data or latent level going beyond the semantic content and focusing on the underlying ideas, assumptions and conceptualisations regarding the patterns of data. The themes here were identified at a semantic level allowing the analysis to stay close to the participant voice but also showing the progression from the descriptive to demonstrate the significance of the patterns and broader meanings and implications from the data.

2.5 Researcher reflections

Braun and Clarke (2013) discuss the importance of developing qualitative sensibility, meaning a combination of skills or position which fit within a qualitative paradigm. This includes having an interest in process and meaning rather than cause and effect, a critical questioning approach to life and knowledge, not taking things at face

value, being able to reflect on experiences but also to be aware of assumptions and put them aside, the ability to listen intently whilst simultaneously reflecting on what is being said, to have good interactional skills to build trust and rapport and to be able to critically reflect on one's own role as a researcher within the research process. A reflective account in the form of a research diary was kept (see Appendix B Research diary extracts and Appendix C Reflective chapter for general reflections).

Before working within the CF service, my knowledge of CF was basic and limited to what I had learnt from science lessons in school. Sitting in on multidisciplinary team meetings and case discussions, my perception of CF was based on a medical outlook. The patients that I heard about or worked with, in the CF department, were often those who were very unwell and CF played an active role in their day-to-day lives. Any therapeutic patient work that I undertook was predominately with those who had been diagnosed with CF in their early years rather than in adulthood. My presumption of receiving an adult diagnosis of CF was that it would be a shock, the individual would struggle with it and experience difficulties with adjustment post-diagnosis, similar to receiving a diagnosis of any chronic condition. Although I had no direct personal experience with CF, as a young child I was diagnosed with asthma and was known to be a 'sickly child' with constant coughs and colds, regularly requiring antibiotics. Although I have some personal experience of living with a chronic respiratory condition, the severity of it is not the same as living with CF. As a qualitative researcher, it was important for me to acknowledge and reflect on my own experiences, understanding and attitudes of CF as this impacted how I viewed it through the participants' eyes.

2.6 Interview design

A semi-structured interview schedule was developed to guide the participant interviews (see Appendix D). As the intent of the research was exploratory, the questions were open-ended and broad to encourage participants to discuss particular aspects of their life or experience using a non-directive style (Willig, 2013). The interview structure allowed participants to raise issues that may have not been anticipated or included as part of the interview guide. This was particularly important when investigating an area with limited understanding. The interview agenda helped to maintain focus, ensure that the questions asked were non-directive, related to the research questions and functioned as triggers to encourage the participant to talk about their experiences (Willig, 2013). It allowed flexibility within the interview in terms of the order of the questions asked, the wording used to ask them and following up or probing on points of interest.

The interview questions were informed by reviewing the current literature in the area and the purpose of the study. It involved an iterative process in which the questions continued to evolve as interviews were undertaken and the research progressed. In the service where the research was conducted, the CF psychologist (AP) was consulted to review the interview schedule, particularly the order and style of the questions asked. To gain feedback on the interview schedule, several attempts were made to contact the CF patient advisor[4] for the service, however, no response was received. Due to the small pool of available potential participants (see 3.1 Recruitment) and the uncertainty around recruitment success, individuals eligible for

[4] A patient with CF who can be approached to provide insight and help improve the patient experience.

the research were not approached to comment on the questions. Following the first few interviews, no comments were made by the participants nor any uncomfortableness observed when questions were asked, therefore it was assumed that the questions were appropriate.

The schedule covered four main areas including the diagnosis experience, the impact of CF, adjustment to living with CF and the support needs of patients. The first section of the interview involved asking participants to tell their story about how they came to be diagnosed with CF. This allowed participants to become familiar with answering questions and helped to make those who might have been anxious or nervous to feel at ease by developing a rapport. The next part of the interview was related to the impact of CF. This started with participants being asked about the positive and negative impact CF had had in their everyday lives which then moved into specific aspects such as relationships, work and social life. The next section involved questions relating to how participants adapted to living with their CF. The final section asked questions about participants' support needs including who provided support, if this was helpful and how this could be improved. Lastly, a summary of the key points of the interview was fed back to the participant and they were given an opportunity to raise any topics or share information which had not been previously discussed.

3.0 METHOD

This chapter reviews the method undertaken by the researcher of this study. It includes a description of the recruitment, the research process, the participants and the method of analysis. The chapter concludes with a discussion of how quality and rigour were achieved within this research and the ethical considerations.

3.1 Recruitment

Patients diagnosed with CF in adulthood who received their medical care from a regional adult CF service[5] in the South of England were identified. This was done through a review of clinic lists and patient medical records by the CF team and researcher. Participants were recruited from only one CF centre. This was decided upon due to pragmatic reasons related to finances. Furthermore, there were assurances from the services CF psychologist of being able to successfully recruit a sufficient number of participants for the study. Acquiring multi-centred ethics was considered a possibility if the recruitment did not go as intended. A criterion sampling method was used to identify participants for the research study, whereby those who met the first three inclusion criteria were invited to participate (see Table 1 for the specific inclusion and exclusion criteria). It was deemed important to allow at least a minimum of a year since diagnosis, firstly for sensitivity to the patient and secondly to allow time for the individual to have a lived experience of CF. There were no resources to accommodate individuals who did not speak English as part of this research study and therefore those individuals were excluded. Individuals with

[5] This regional centre has been providing care for adult CF patients for over 20 years. At this centre, inpatient and outpatient care is provided for patients.

significant learning difficulties were excluded from the research due to difficulties with communication and their ability to converse about their lived experiences of CF.

Table 1: Inclusion and exclusion criteria for adult CF research participants

Inclusion criteria	Exclusion criteria
Patients who had been given a clinical diagnosis of CF in adulthood.	Patients who had significant learning difficulties.
Patients who were diagnosed with CF at least one year previously.	
Patients who were over the age of 18 years of age.	
Patients who are native or fluent speakers of English.	
Patients who were physically well at the time of the interview.	

A total of 29 patients out of 266 patients registered with the CF centre were identified as having a clinical diagnosis of CF in adulthood. Of the 29 patients, it is unclear how many patients, if any, were excluded based on the study's criteria. The CF psychologist (AP) sent an invitation letter (see Appendix E) and the study information sheet (see Appendix F) inviting the 29 patients to participate in the research. Due to the low response rate following the initial mailing (n=2), a follow-up letter (see Appendix G) was sent to potential participants a month after the initial letter had been sent out. In the initial invitation letter, patients were made aware that they may be contacted by a follow-up letter from the research team. If patients chose not to be contacted or to take part, they were asked in their initial letter to inform the research team, this was recorded and the patient was not asked about the research project again. Patients interested in participating were asked to contact the researcher by

telephoning or e-mailing the CF administration team. Recruitment posters were also displayed around the adult CF service and the service's social media platforms (e.g. Facebook and Twitter) were used to advertise the project (see Appendix H). See Figure 2 for a flow chart of the recruitment procedure and Figure 3 for details of how the participants were recruited.

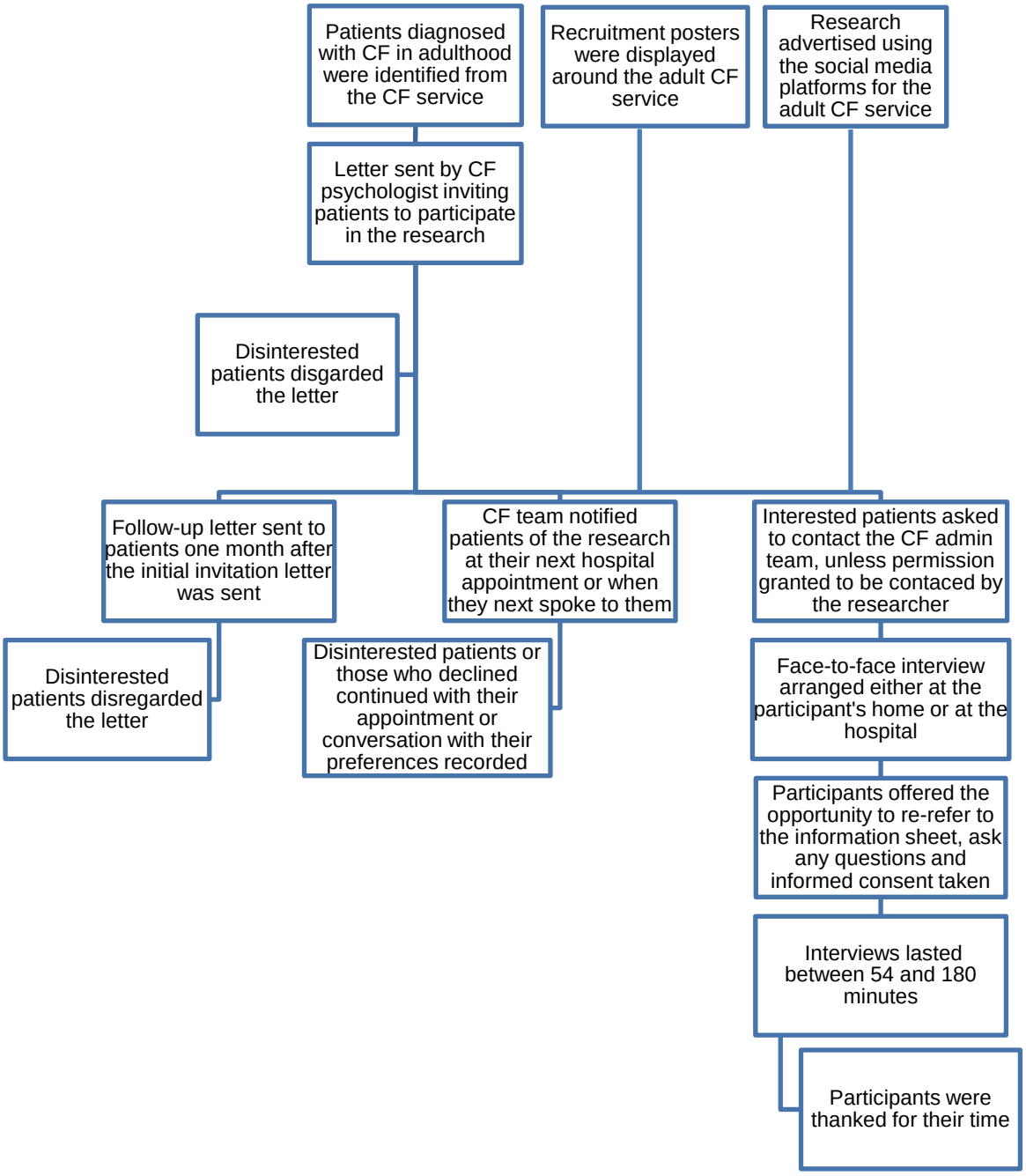

Figure 2: Flow chart of the adult CF patient recruitment procedure

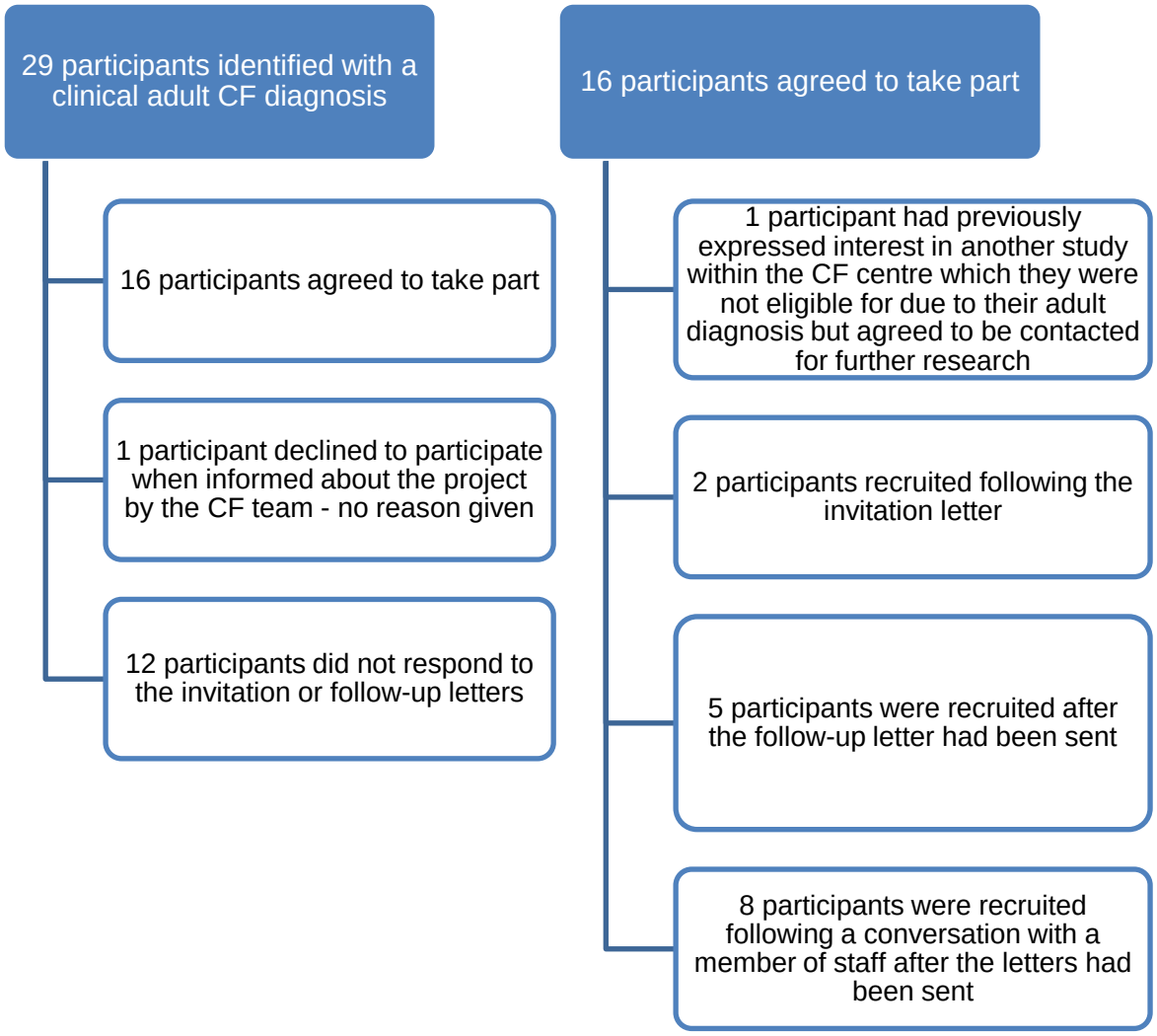

Figure 3: Participant recruitment diagram

3.2 Procedure

Once an expression of interest was made, arrangements for a convenient date and time to carry out the interview were made with the individual over the telephone (n=13), e-mail (n=2) or face-to-face (n=1). All of the participants chose their interviews to be conducted face-to-face. They were held either in a private room in a hospital in the South of England (n=5) or the individual's home (n=11). The interview location was chosen by the participant. For the interviews carried out in the hospital, every effort was made to undertake the interview on the same day as the patient's

hospital appointment to minimise travel. Participants were on their own for the interview except in two cases; one whereby the patient's family member was present for part of the interview and another in which the patient's partner joined at the end. This has been discussed further in the reflection section (see Appendix C – Reflective chapter – Interview experiences).

Interviews were guided following the guidelines of Magnusson and Marecek (2015) and Legard, Keegan and Ward (2003). This involved initially establishing a good rapport with the participant to ensure they were at ease. At the beginning of each interview, a brief introduction to the research project was provided. All participants were given the opportunity to re-refer to the information sheet (see Appendix F) and written informed consent was taken (see Appendix I). A semi-structured interview schedule was used during the interviews (see Appendix D). Participants were encouraged to freely describe their experiences and were asked a series of follow-up questions throughout the interview. Interviews lasted between 54 and 180 minutes with most between 60 and 90 minutes. They were audio recorded, with the participant's consent and later transcribed verbatim mostly by the researcher. A transcription service was also used for time efficiency. Transcription is an important first step in data analysis (Bailey, 2008) therefore, a potential disadvantage was that the researcher was removed from this process. However, to counteract this, the researcher verified the transcripts completed by the transcription service. This also allowed close observation of the data. At the end of the interview, individuals were given the opportunity to reflect on the conversation and were thanked for their time. As a token of appreciation, participants were sent a thank you card and offered the opportunity to be entered into a prize draw to win a £25 gift voucher, which was

stated within the study information sheet and on the consent form. Interviews were carried out between January 2016 and November 2016.

3.3 Participants

In total, 16 participants took part in the study, 6 males and 10 females. The participants were aged between 24 and 69 years at the time of the interview (mean age: 48 years and mean age at diagnosis: 38 years). All participants identified themselves as White British. To situate the sample (Elliott, Fischer and Rennie, 1999), a full description of the demographic details of the participants can be found in Table 2. Pseudonyms have been used to protect participants' anonymity.

Table 2: Participant demographic information

Participant pseudonym	Gender	Age range	Time since diagnosis	Relationship status at the time of the interview	Have children (includes biological and partner's children)	Level of education	Employment status at the time of the interview
Jordan	Female	51 – 60 years	4 years	Married	Yes	NVQ/Diploma	Full-time
Chris	Male	61 – 70 years	17 years	Married	Yes	Not reported	Retired
Robyn	Female	41 – 50 years	7 years	Married	Yes	O Levels/NVQ Level 2	Not working due to health
Alex	Female	41 – 50 years	10 years	Single	No	No formal qualification	Not working
Sam	Female	51 – 60 years	37 years	Divorced	Yes	O Level	Not working
Jamie	Male	61 – 70 years	10 years	Married	Yes	HNC	Full-time
Morgan	Female	31 – 40 years	7 years	In a relationship	No	A Levels	Full-time
Charlie	Male	41 – 50 years	12 years	Single	Yes	No formal qualification	Part-time
Frankie	Male	31 – 40 years	9 years	Married	Yes	National diploma	Full-time
Riley	Male	41 – 50 years	4 years	Married	Yes	Undergraduate	Full-time
Taylor	Female	51 – 60 years	15 years	Married	Yes	Honours degree	Retired
Frances	Male	61 – 70 years	5 years	Married	Yes	City and Guilds	Full-time
Jo	Female	41 – 50 years	11 years	Single	Yes	O Levels	Full-time
Leslie	Female	21 – 30 years	5 years	Single	No	Not reported	Part-time
Max	Female	21 – 30 years	5 years	In a relationship	Yes	NVQ	Not working
Danny	Female	51 – 60 years	23 years	Married	No	Undergraduate	Not working due to health

3.4 Sample size

There is some debate over methods of calculating sample size in qualitative research. The sample size can be influenced by several factors such as what the researcher wants to know, the purpose of the research, what will be useful, what will have credibility and what can be done with available time and resources (Patton, 2015). Data saturation is a widely accepted method in qualitative research, commonly used to indicate the point at which it is deemed unnecessary for any further data to be collected as no new knowledge has been observed (Saunders *et al.*, 2018). However there is uncertainty around how saturation should be conceptualised, inconsistencies in its use and researchers often do not record the methods used to determine how saturation was achieved (Saunders *et al.*, 2018). Unlike quantitative research, which focuses on generalisability and power to determine sample size, factors such as the scope of a qualitative study, the quality and depth of data collected, the nature of the topic, the epistemological and theoretical underpinnings, the amount of data collected per person, the type of analysis, the number of interviews per participant, the use of shadowed data and practical considerations all have an impact on how much data is needed to reach saturation (Baker and Edwards, 2012). As study designs are all unique, “there is no one-size-fits-all method” to achieve data saturation (Fusch and Ness, 2015, p.1409) and “little practical guidance for estimating sample sizes, prior to data collection” (Guest, Bunce and Johnson, 2006, p.59). It was found that the most common sample sizes were 20 and 30, in a sample of 560 PhD studies using qualitative interviews as the method of data collection (Mason, 2010). However, the author noted that most of the sample sizes of the studies were multiples of 10, showing no logical or theory-driven reasoning. Thus, highlighting that the guiding principles of

saturation as a method for sample size were not being followed. Braun and Clarke (2013) recommend that a sample size of between 10 and 20 interviews are suitable for a medium-size project. Whilst it has not been explicitly stated how the authors arrived at this number, it is assumed this value is based upon their extensive experiences as qualitative researchers.

The project aimed to recruit between 15 and 20 participants to include a broad spread of ages and gender from a small sample group of 29 patients. A range of ages allowed the capture of different experiences over time and varied time since diagnosis. In total, data from 16 interviewees was collected. Other studies using semi-structured interviews and thematic analysis have used a similar sample size (Stanyon *et al.*, 2016; Thompson and Abel, 2016; Brunger *et al.*, 2014). Data saturation and practicality were the main factors that influenced the final number of participants recruited for this study. Most of the interviews with the adults with CF were lengthy, generating rich and detailed data from the small pool of potential participants available. At the point of interview 12, it was felt that no new information was coming from the interviews and what had been heard was repetition from previous interviews. After the twelfth interview, a further two participants had expressed an interest in participating in the research. Another two participants who were previously unable to attend their scheduled interviews were still keen to share their experiences. Since the research aimed to have a breadth of experiences and as two of the four participants were from the 21 to 30 years age category, one which had not been represented, the four interviews were carried out. No further participants came forward to take part in the research. With limited time in which to

complete the project and ample data from participants, it was decided to end the recruitment at this point.

3.5 Data analysis

There are several methods of TA (Joffe and Yardley, 2004; Boyatzis, 1998; Aronson, 1994). Data collected from participants was analysed using the TA approach outlined by Braun and Clarke (2013; 2006). They define TA as "a method for identifying, analysing and reporting patterns within data" (Braun and Clarke, 2006, p.79). This version of TA was chosen as it fits with the needs of the research question and offered a systematic and rigorous approach to coding and theme development (Smith 2015; Howitt, 2010) (see Figure 4 for a diagrammatic depiction of the process followed).

Figure 4: Phases of thematic analysis (adapted from Braun and Clarke, 2013; 2006)

As shown in the diagram, the data analysis was not conducted linearly but was an iterative process that involved going back and forth through the phases. The initial stage of analysis involved 'immersing' oneself within the data, becoming familiar with the content and making initial notes of potential items of interest. This process started as the interviews were transcribed verbatim, searching for meaning and patterns within the data as each transcript was read and reread several times. Transcription is considered a key way of familiarisation of data to occur (Bird, 2005). Initial thoughts and key points were noted. The next phase involved complete coding across the entire dataset. Within this stage, interesting elements of data concerning the research question were identified and coded using a word or short phrase. Notes and comments were made alongside two transcripts (see Appendix J). These were compared with the notes made by two people; a fellow doctoral student and a person with a computing background. The transcripts were discussed with individuals from a varied background to get different perspectives to enhance the credibility of the research (Tracy, 2010). These alongside the ideas noted during the transcription stage were used to form tentative codes, which were then systematically applied to code the entire data set. Additional codes were added if the data did not fit a pre-existing code. NVivo[6] was used to aid with the coding process. In the third phase, once all of the transcripts were initially coded, all of the codes were sorted and arranged into potential overarching themes (see Appendix K). Within this stage, the relationship between codes, themes and the different levels of themes were considered. The fourth phase involved two stages of reviewing and refinement. The initial stage involved checking that the extracts under each theme fit. The second stage involved checking if the themes accurately reflected the whole data set. By the

[6] A data analysis software program.

end of this phase, the researcher was clear on what the themes were, how they fitted together and the story it told about the data. Phase five started with the researcher having a thematic map that they were satisfied with. Within this phase, the researcher refined and defined the themes, wrote a clear definition for each theme and began to consider theme names for the final analysis. The sixth phase involved the final analysis and write up of the report. This involved presenting a coherent narrative of the data to the supervisory team, colleagues and participants for feedback, which included extracts to illustrate the participant's story.

There are a number of potential pitfalls that can result in poor analysis. For example, the failure to analyse the data and not go beyond the content, using the interview questions as the reported themes, a weak or unconvincing analysis due to too much overlap between the themes, internally incoherent and inconsistent themes or if the themes do not appear to work (Braun and Clarke, 2006). Therefore, to avoid this, Braun and Clarke's 15-point checklist for good thematic analysis was consulted and followed throughout the phases (see Appendix L).

3.6 Quality and rigour in qualitative research

To ensure rigour, transparency and good qualitative research, guidelines by Tracy (2010) and Meyrick (2006) were followed. Tracy's (2010) eight 'big-tent' criteria aims to present a universal model to demonstrate best practice within qualitative research. Meyrick's (2006) framework aims to provide markers of good quality research across each stage within a qualitative research process. There are a number of alternative criteria that could have been used to inform quality in qualitative research (e.g. Yardley, 2000; Henwood and Pidgeon, 1992; Lincoln and Guba, 1985). However,

Tracy's criteria and Meyrick's framework were chosen because they are simple, practical and comprehensive models with a good fit with the researcher's epistemological and ontological standpoint. Tracy's suggested key markers of quality are: a) worthy topic, b) rich rigour, c) sincerity, d) credibility, e) resonance, f) significant contribution, g) ethics and h) meaningful coherence (see Appendix M for the application of these markers within the current research).

Through each stage of Meyrick's framework, it was ensured that the study followed the principles relating to the relevant stages of the research process. The framework focuses on two principles: transparency (shaded light grey) and being systematic (shaded dark grey) throughout the research process. Appendix N demonstrates how the framework was applied to the study.

3.7 Ethical considerations

This section discusses the ethical issues concerning this study.

3.7.1 Ethics approvals

Full NHS ethical approval was granted for this study on 10th October 2015 by a research ethics committee based in the South of England (Ref: 15/SC/0567) and the University of the West of England (see Appendix O). Research sponsorship and insurance was granted by the University of the West of England (see Appendix P).

3.7.2 Informed consent

Alongside an invitation letter (see Appendix E), potential participants were sent an information sheet (see Appendix F) advising them of the purpose of the study, the

procedure and possible risks involved. It also included information regarding consent, confidentiality and anonymity. Participants had the opportunity to refer to the information sheet before the interview and have any questions answered. If participants were happy to proceed with the interview then they were asked to sign a consent form, which was duplicated, one copy was kept by the participant, one for the site file and one for the patient's medical records.

3.7.3 Patient safety and wellbeing

As a researcher, it was important to be aware that the interviews may have involved discussion of sensitive topics, which may have evoked emotional distress. Participants were advised that they could refuse to answer any questions that they found difficult and could decide to stop the interview at any point. Participants were also advised that they were able to withdraw at any time during the research, even after the interview had taken place, for up to 28 days afterwards. Patients were reassured that their medical treatment was not influenced in any way, even if they decided to withdraw from the study. If it was identified that the patient may benefit from further psychological support, they were provided with the contact details of the CF Clinical Psychologists (AP, RA or LD) working within the service. No participants were referred to the Clinical Psychologists or signposted onto any organisations for additional support following the interviews.

If a patient discussed a topic, shared information or asked specific questions about their CF, which the researcher felt was appropriate to discuss with the CF team, permission was requested to take the most appropriate course of action. For

example, presenting the information to the CF team and if necessary, feeding back the response to the patient.

CF patients can experience a number of symptoms related to their condition, which may include feelings of fatigue or tiredness for example. Care was taken when interviewing participants to be mindful and make adjustments if needed. For example, participants were not interviewed if they were physically unwell, they were advised of the likely length of the interview and that they could pause or stop the interview at any point.

3.7.4 Participant anonymity

Adult diagnosis of CF is rare. With only a small pool of eligible participants for this study who were recruited from the same regional service, there was potential for participants to be identified. To reduce the likelihood of patient identification, participants were given pseudonyms and their age was not explicitly stated. Interview transcripts were anonymised, removing any mentions of people and places.

3.7.5 Researcher safety

For the researcher's safety, a protocol was written and implemented for interviews that took place at the participants' homes. Before the interview, a named contact within the CF team was given the participant's name and contact telephone number, the interview location and the expected time of arrival and departure. On arrival at the participant's home, a text message was sent or a telephone call was made to the named contact and a brief risk assessment was carried out before entering the

participant's home. Contact was made with the named person on completion of the interview. It was agreed that if the named contact had not heard from the researcher 2 to 2.5 hours after arrival, then they would contact the researcher. A code word was agreed to indicate if further assistance was required without alerting the participant. The researcher had access to supervision provided by the CF psychologist if any debrief was necessary. For example, following interviews that involved the discussion of sensitive or distressing accounts.

4.0 RESULTS

The chapter that follows describes and discusses the findings of this study. Four themes were identified during the analysis of the data: “No, you can’t possibly have CF”, Emotions around diagnosis, “It did kind of take over my life” and “I no longer wish to argue with it”. The first theme explores participants’ experiences prior to being diagnosed. It looks at the disparity between individuals’ reported health symptoms with their anticipated and actual outcomes following interactions with healthcare professionals. The second theme focuses on participants’ diagnosis experience and the emotions felt. The third theme explores participants’ experiences of living with CF and the impact it has on daily life, work, relationships and the future. The final theme reviews participants’ relationship with their CF and their acceptance of emotional and physical changes that they may have implemented.

Figure 5 shows each theme and sub-theme. Within the findings, whilst some commonalities have been identified in other research areas such as long term conditions and the cancer literature, the focus of this chapter is to highlight novel findings for being diagnosed and living with CF in adulthood.

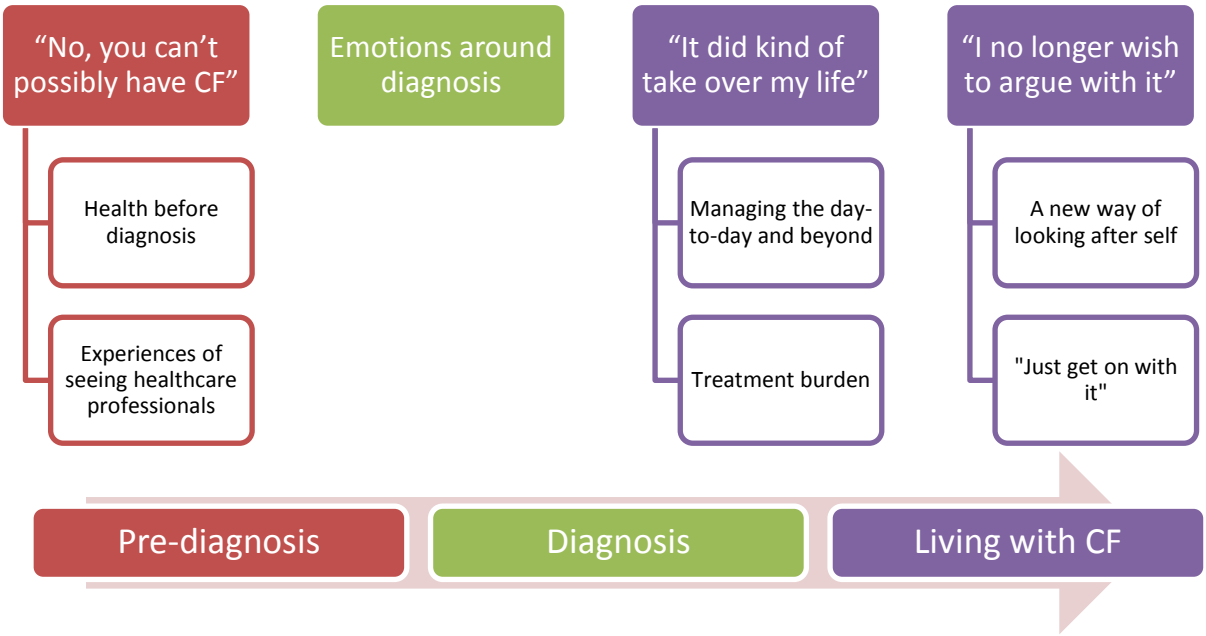

Figure 5: A conceptual map of themes and sub-themes

4.1 "No, you can't possibly have CF"

This theme focuses on participants' pre-diagnosis experience.

4.1.1 Health before diagnosis

From childhood, most of the participants reported being unwell, having "a bad chest" and recurrent chest infections, which often required antibiotics to clear and "a lot of colds" that tended to linger on. One participant, Robyn, also reported suffering from unexplained bowel problems, constipation as well as back and stomach pain. Participants' poor health from childhood continued through into adulthood. Medical advice was sought from individuals' general practitioners (GP), usually by their parents, to identify the problem and to help manage the symptoms. Most of the participants were either diagnosed with another condition, reported being dismissed, or not taken seriously by their healthcare professional. Participants recalled going

back to their doctors repeatedly feeling frustrated that little was done. Six of the participants reported being told that their ill health was due to asthma.

> *"As a child, I'd always had a poorly chest, and I knew that my lungs didn't work properly and it was put down to having asthma"*
>
> Jo, F, 41 – 50 years

> *"I was always bad at school, with, well they always said it was asthma and hay fever"*
>
> Frances, M, 61 – 70 years

Some parents were told that their children would grow out of their symptoms.

> *"[S]ince I was a child, I've had a cough. Nothing serious, but just this kind of cough my mum used to take me to the doctors when I was a child, saying you know she's coughing, and they just say 'oh take her away, she'll grow out of it'"*
>
> Danny, F, 51 – 60 years

Danny and Taylor talked about how their parents accepted what the doctor told them about their health and did not challenge what had been said or pursue things further.

"[W]hen I was younger my mum used to take me to the doctor and then when I was sixteen obviously something else initiated a trip to a consultant, and I can't remember what that was, but to be honest I think I wasn't bad enough that they really pushed it. And my parents aren't, I suppose my dad probably more than my mum, isn't one for arguing with authority, for want of a better word"

Danny, F, 51 – 60 years

"I don't think my parents, you know, they were both professionals, they never thought, you trust what the doctor's telling you"

Taylor, F, 51 – 60 years

Alex discussed how throughout her childhood she was unable to physically keep up when playing with friends. She was regularly unwell and was in need of antibiotics.

"[Y]ou couldn't just do the things what your mates were doing running around and things like that. I would get out of breath, I would have to stop, a couple of times I've been known to pass out, straight out on the floor. Its mad things you know, and never knowing why or what it was caused by, didn't know [pause]. All my life sore throats, colds, flu's, this, that, and everything all the time, was just going on my chest, oh here's another two weeks of antibiotics, you'll be alright, go on, you go home now, you'll be alright, that was it"

Alex, F, 41 – 50 years

In contrast, only a few participants reported feeling well as a child. Despite knowing that something was 'not quite right', participants tended to accept their ill health and made adjustments to their life accordingly, "*as a child you don't think about it too much, you just, accept you know, you have this cough*" (Sam, F, 51 – 60 years). In Charlie's case, he found that staying physically active helped to keep his chest clear and manage his symptoms, therefore his health was not problematic for him at the time.

> *"I was alright growing up, 'cause I was always running about and on my bike and I was pretty fit when I was young, so never really hit me until later on in life"*
>
> Charlie, M, 41 – 50 years

4.1.2 Experiences of seeing healthcare professionals

As an adult, Charlie's health declined following having pneumonia. Whilst Charlie's doctors knew there was something wrong, they could not identify the cause of the symptoms. They provided different treatments to see what worked best. Charlie struggled to be taken seriously by his doctor despite informing them that his sister had CF and had died as a result of it. He reported not being offered testing and instead was sent away with antibiotics. Similar to Charlie's experience, Taylor wanted to get tested for CF following the death of her sister, whom she previously

did not know had CF. However, Taylor was told by her GP and nurses at her surgery that she was “too old” to have it.

> *“[W]hen [sister’s name] died, we said, ‘I need to be tested for CF’ and he [doctor] said ‘no you’re too old, we tested you for emphysema when you were in the [hospital’s name] but you were too young for that’ so I said ‘well if you test me for that, why not test me for CF and knock it out of the equation’, no no no not necessary. So when [sister’s name] died and I had her death certificate, I went in and I said ‘I’m obviously not too old, she was three years older than me, so do you think we should be tested now’, so still never got done, still nothing was done about it”*
>
> Taylor, F, 51 – 60 years

As well as her GP surgery, Taylor encountered other healthcare professionals who stated that she looked “too well to have CF” and a top lung specialist who dismissed the idea of CF, stating that she was “too old” to have it.

Most participants were diagnosed as a result of their recurrent poor health or from receiving care for an acute illness episode whereby they were hospitalised and subsequent further tests identified the individual to have CF. Some participants were diagnosed following the discovery of a sibling or close family member diagnosed with CF and one participant was diagnosed as a result of being unsuccessful in having further children with his wife. By the time participants received their diagnosis, many had spent years going back and forth to their GPs, had been referred to several

specialists for further investigations and had various tests. For some, this led to a negative relationship with healthcare professionals because of a loss in faith and trust as their CF had not been picked up.

> *"I was always a chesty child, I always had a cough, and my parents sent me off to various consultants in [street name] and none of them picked up on CF. So I am quite sceptical about the medical profession"*
>
> Sam, F, 51 – 60 years

> *"I went through a stage of not believing anything anybody said, doctors, nurses, consultants, anybody"*
>
> Max, F, 21 – 30 years

In summary, this theme captures participants' pre-diagnosis experiences and the disparity between individuals' reported health symptoms and their anticipated and actual outcome following interactions with their healthcare professional. Most of the participants had poor health when they were younger, reporting a bad chest and recurrent coughs and colds. Some participants reported very few symptoms as a child but as they got older their health declined. When medical advice was sought from the participants' GP, they were either diagnosed with another condition, usually, asthma or they perceived that they were not taken seriously or felt they were dismissed. Participants recalled recurrent visits to their GP or interactions with

medical professionals where often little was done. For some participants, this led to mistrust and loss of faith in healthcare professionals.

4.2 Emotions around diagnosis

This theme describes participants' diagnosis experience. Upon receiving their diagnosis, many participants felt ambivalent and described their experiences as "*a double-edged sword*" (Jo, F, 41 – 50 years).

> *"[O]n one hand it felt like I had an answer for all this stuff that had been going on in the past, so it was positive. On the other hand, it was quite negative, because it was quite scary. At the time not many people were diagnosed in their, well as an adult. They didn't really know what the prognosis was. They said you're a rare form, and they said chances are you'll be as you are now for the next five years, after that we really don't know"*
>
> Danny, F, 51 – 60 years

Several of the participants were relieved to receive their diagnosis, "*it was like a ton of weight lifted off my shoulders*" (Taylor, F, 51 – 60 years). Getting the diagnosis meant that it opened access to the right support and treatment allowing participants the opportunity for an improved quality of life. For Max, this extended to help from family for tasks that she could not carry out because of her CF, as well as accessing support from her council and GP quicker than she may usually have done if she did not have CF. Participants also reported that their diagnosis was like a "*puzzle*

coming together" (Robyn, F, 41 – 50 years). The diagnosis explained illnesses or what appeared to be random symptoms throughout their life, for example, recurrent coughs, stomach problems, inability to gain weight and infertility issues. It enabled individuals to look back and make sense of their ill health. Previously where they may have felt shame or embarrassment because of their symptoms, they were able to understand or justify why it had been happening to them.

> *"[B]efore I used to hide an awful lot of things, like the tummy problems you get, and being on the toilet, sitting on the toilet for ages, or whatever. I used to hide an awful lot of that, but when I got diagnosed with the CF it was like, huh, I've got a reason for this now, it's not just that I've got really bad eating habits or I've got a rotten tummy or anything like this"*
>
> Taylor, F, 51 – 60 years

When participants were finally diagnosed with CF, they reported a range of emotions and feelings including shock, relief, anger, denial, 'why me?' and 'why did I get missed?'. The most commonly reported was the shock of the diagnosis and the relief to be diagnosed. The shock of their diagnosis was mostly because CF was never mentioned or considered a possibility. Taylor was treated by her GP for recurrent colds and had therefore not anticipated it to be anything more. Following her sister's death, Taylor found out that her sister had CF and later she also tested positive for it.

"I think the initial kind of shock wave, that's exactly what it was, it was a shock wave, and then of course you've got my sister's death, that was the start of that shock wave, 'cause nothing had ever been said about it [CF] before"

Taylor, F, 51 – 60 years

"It was all a massive shock and it was too much to take in and I just immediately thought I was just going to die"

Morgan, F, 31 – 40 years

The shock of the diagnosis was further exacerbated for some as they had not expected to receive a diagnosis when it was given and instead anticipated that they were going for an appointment for further tests. For a few participants, receiving their diagnosis was a destructive life-changing moment.

"[O]nce I got diagnosed, it was like a sledgehammer, smashing me, my whole life was just turned upside down"

Robyn, F, 41- 50 years

In contrast, five participants, four of whom were male, appeared unaffected by the news of their diagnosis. Some of them had anticipated or expected their diagnosis, so when it was finally revealed to them it was not a surprise.

"[T]o be honest it was no surprise when it, the letter came. I was in a hurry read the letter put it on the side and thought ah I knew that, and carried on and it never really affected, I found it the other day actually, on the side, it didn't really affect me"

Frankie, M, 31 – 40 years

"It was a bit weird, 'cause like everybody came in the room and told me, but I was sort of, I knew anyway but, that confirmed it really, so I didn't feel no different"

Charlie, M, 41 – 50 years

Riley shared how his CF diagnosis was not significant for him because he did not understand the impact of it. However, he was more concerned and shocked at the fact that he could not have children naturally. For Riley, it appeared that his infertility was seen separately from his CF.

"I suppose it, came as quite a surprise, I wouldn't say it was a shock 'cause I didn't know, the impact really of what it would be. It wasn't the, CF that was the real surprise and the real shock, it was the fact that I couldn't naturally have children that was the real shock, I think, that was the, so, because I, apart from that I felt I've always been fairly healthy, so I felt healthy, I felt well"

Riley, M, 41 – 50 years

With CF being a life-limiting condition, another common concern and question raised by participants was around their prognosis, bringing up feelings of uncertainties around their future which, for some, always remained at the back of their mind.

> *"[I]t's just a scary illness, you know, you don't know how long you've got, how far you're going to go"*

Alex, F, 41 – 50 years

For some, before their diagnosis, they expected to live to an old age and thought they were 'invincible'. However, having received the diagnosis the reality of their death became real. It was difficult to be told that *"there's no cure, and will very likely reduce your life expectancy as well*" (Jordan, F, 51 – 60 years). Several of the participants commented on wishing they knew about their diagnosis sooner, because they would have made different life choices, particularly choosing not to take up smoking.

> *"[I]t would have obviously been nicer to, know I had it years ago, so I could have actually done, different stuff, if you know what I mean, or I had better treatment, or something"*

Charlie, M, 41 – 50 years

"I would have done my life differently, probably, if I'd have known I had CF when I was, or I had symptoms of CF, when I was in my early twenties and thirties. My life would have been done differently"

Chris, M, 61 – 70 years

One participant voiced regret for taking their health for granted. She questioned if her health would have been better had she known about her CF when she was younger. Six participants spoke about being in a state of denial about their diagnosis and did not want to accept that they had a chronic illness, in Sam's case choosing to bury her head in the sand.

"I was probably my own worst enemy because I did not talk about it, swept it under the carpet, and just carried on as though I didn't have it"

Sam, F, 51 – 60 years

"One part of me says hey, I haven't got CF, I'm convinced I haven't got CF, or a variant of CF. I feel, you know, that I'm convinced I haven't. They [CF team] all say I have, but I am convinced I haven't, you know. But hey, you've got to get a treatment for it, that's fine by me"

Chris, M, 61 – 70 years

A few of the participants were angry, questioning “why me?” and wondered why their CF got missed despite having health problems commonly linked to CF and/or several hospital admissions.

> *“I just thought why me?, again, why me?, not realising that all these previous illnesses, that I’d been growing up with, was actually a result, of the CF. I didn't know that at the time, you know, I just thought ah. And then when it was explained, when I explained to them, ‘cause I’ve been in a few doctors, like meeting things, and they’re like, we can’t understand, with all these illnesses, that not one doctor, picked up on cystic fibrosis. No one can understand, no one can explain it”*
>
> Alex, F, 41 – 50 years

Alex shared how she felt cheated and that all her life she could have had support and treatment for her CF but instead was left on her own to struggle. Alex felt that she had been ignored by healthcare professionals as no one had ever pointed out her CF. Jo equally felt let down by her GP and wondered why they did not ask more searching questions. Prior to being diagnosed, some participants were given a diagnosis for another condition. When they were diagnosed with CF, they struggled to come to terms with it because they believed they had been misdiagnosed. For some, this led to feelings of mistrust with their healthcare providers.

> *“[O]kay, so I’ve got CF, fine, you know or I’ve got a variant of CF, I’ve got to accept it, but I still find that hard to believe, because after getting*

to fifty and told you have emphysema, then someone wakes you up one day and says hey by the way you haven't got emphysema now, you've got CF"

Chris, M, 61 – 70 years

"I didn't have it [CF]; it was a misdiagnosis, and that I had asthma and, a bit of sinuses and a bit of this bronchiectasis thing from chest infections, and that they [hospital healthcare professionals] would realise that they got it wrong and I could just crack on with my life and never see them again"

Morgan, F, 31 – 40 years

Morgan found that it took her a long time to come to terms with the fact that she did not just have asthma but also had CF. The lack of acceptance of her CF led to not engaging in any treatments or taking of medication for many years. This was also similar to Jo's case where she reflected on how she did not allow for CF in her life at the time and therefore her compliance with her medication was low. She talked about how she did not accept CF and how it would change her life. She had to think about coming into hospital every three to six months to have tests and then consider taking time out to do the treatment. It took her a number of years to allow for CF to be part of her life.

At the time of diagnosis, there was a mismatch between the information needs of patients, what was provided and how it was delivered by healthcare professionals. Half of the participants talked about the limited and generalised information available to them at the point of diagnosis. They were often frustrated that the information given to them, or that they had researched, did not correspond to their experiences. Some participants were diagnosed at a time prior to internet access and therefore it was difficult to source information that was current and up-to-date. Other participants shared how overwhelmed they were with the information presented to them.

> *"It was alright until I had my first appointment at the CF clinic, and I was like bloody hell. It was like they gave you so much information and I was like oh I don't understand what any of that means"*
>
> Leslie, F, 21 – 30 years

Jo shared how most of her initial information about CF was provided verbally. She struggled to process the information as she was still trying to process her CF diagnosis. She stated how she would have preferred to have had written information to take away with her.

In summary, this theme identifies how ambivalent participants felt about their diagnosis. There was a sense of relief to get a label for the condition, however, it also brought on concerns about what implications this had for their prognosis. Participants reported feeling many emotions around the time of diagnosis, most commonly a sense of shock as well as a relief to be diagnosed. For many of the

participants, CF was something that had not been mentioned before or even considered a possibility. Some had found that hearing that they had CF was so overwhelming that they were unable to process anything else said to them during their diagnosis appointment. Others were in a state of denial, not wanting to accept that they had a chronic illness, some were angry and questioned why their CF had been missed despite having a lot of common symptoms associated with the disease. For many, receiving the diagnosis was the missing piece of the puzzle, which explained their symptoms. Getting a diagnosis for CF opened access to support and treatment, which allowed participants to have the opportunity of an improved quality of life.

4.3 "It did kind of take over my life"

This theme discusses the impact CF has had on participants' lives.

4.3.1 Managing the day-to-day and beyond

Over half of the participants shared how CF "*impacts on every aspect*" (Jordan, F, 51 – 60 years), it affected all aspects of their day-to-day life including their quality of life, work, finances, relationships, travel and their future. For six of the participants, CF was seen as *"just a bit of an inconvenience*" (Leslie, F, 21 – 30 years).

> *"[N]o-one would even begin to imagine, the day-to-day, devastation on your quality of life, when, you know, you feel like, you can't go out the house, because your stomach's griping, and you're going to need the loo"*

Jordan, F, 51 – 60 years

“I don’t let CF win, until I’m having a really bad day, and then I’m like well do you know what, I hate CF, it’s the worst thing that’s ever, ruined my life, when in theory it had, it’s changed my life, but it’s not ruined it”

Max, F, 21 – 30 years

In contrast, seven participants shared that CF did not affect their day-to-day life, with the biggest impact being that they had to travel to their CF centre several times in the year. Participants commented that they were still able to do everything that they had done before their diagnosis. However, many of these participants appeared to contradict themselves as they also shared instances of where their CF had impacted their lives.

“[I]t hasn’t stopped me doing anything. Touch wood, I’m one of the, I would say lighter sufferers, you know, I don’t have a lot of the bad bits of the CF”

Taylor, F, 51 – 60 years

“[*Y]ou have good days and bad days, you know, if you feel like you’re getting a bit of a cold it can really render you kind of, oh gosh, I can’t do anything, and I like to be out and about doing things, so when you get those bad days, and I just think this is a waste of a day*”

Taylor, F, 51 – 60 years

Seven of the participants shared the challenges experienced as a result of the unpredictability of their CF and the impact that this had on their life. They talked about how they had good days and bad days, meaning that *"you can go to bed feeling fine one night and wake up feeling like a complete train wreck"* (Max, F, 21-30 years). The unpredictability experienced by participants was frustrating, particularly as it would come on all of a sudden.

> *"[M]ost normal people go oh I think I'm coming down with something, think I'm coming down with something. You don't get that normally with CF, it's all or nothing. You feel fine or you feel like utter crap"*

Max, F, 21 – 30 years

This made it a struggle for participants to manage their day-to-day lives as their CF would take over and in some cases, affected their work or ability to work.

> *"[I]t can be a struggle in the way every day is a different, you don't know what you're going to wake up, like tomorrow, you know, some days I wake up I'm leaping about, I'm all good, then the next day I could get up and feel like a ton weight, me head, I can't lift me head, it's just urgh"*

Alex, F, 41 – 50 years

"I could wake up any day and I'd think oh my God, this is really bad [laughs], you know either with a cough or pancreas or whatever, and other days I'd wake up and feel okay, and because it was so unpredictable, it just in some ways it did kind of take over my life in a way I didn't want it to"

Jo, F, 41 – 50 years

The unpredictability of CF made planning a challenge. This was specifically around making commitments to attend social plans as individuals would not know if they would be well enough to attend or not.

"It's difficult like in the future, or like last year I couldn't agree to any social things because I didn't know if I'd feel well enough, so I missed out on a lot of stuff last year"

Morgan, F, 31 – 40 years

Others spoke of similar experiences where they found that they no longer wanted to go out or do anything, noticing changes in their energy levels and their ability to do tasks that were previously considered simple.

"[Y]ou don't want to go out, you know, you just become like, like a recluse, you just don't want to do nothing"

Chris, M, 61 – 70 years

“I’ve got no enthusiasm, no umph, no up and go in me like I used to have”

Alex, F, 41 – 50 years

Ambivalence around work

The impact CF had on work was the most talked about. At the time of the interview, some participants worked full-time, some part-time and some were not working, either due to their CF or retirement. After their diagnosis, some participants had to reassess the appropriateness of their job roles and the environments in which they worked because it was harmful to their health. Some were concerned about how they would cope physically if they worked. Those who were working shared the challenges that CF presented in terms of having to take time out of work for appointments and then having to catch-up with work. Some missed opportunities of being involved with things at work because of their CF. Morgan shared how she would not be included on projects at work by her manager because of a fear that she would be off sick.

“Well it’s like I had my appraisal yesterday at work and my boss was like yeah, I was thinking of giving you this project but we don’t know when you’re going to be sick again and I don’t know, you know what I would do if you were off sick and you hadn’t finished it, or I’d suddenly needed the stuff and you’d done and you weren’t here. And I was a bit

like so I can miss out on a project because I might get ill, but I might not"

Morgan, F, 31 – 40 years

A few of the participants also shared the embarrassment that they faced at work with coughing up phlegm.

"[W]hen I was still working, the ways it would affect is me, is like the coughing, as I said earlier, about coughing up phlegm, now I could be in an interview with a customer for an hour to two hours, if I started a coughing fit and phlegm's flying off, you know, it could be really embarrassing at stages of having to run out of a room"

Taylor, F, 51 – 60 years

Those who were not working often wanted to work. However, they were either unable to find suitable positions that were flexible to their needs enabling them to fit their treatment around, or they had previously worked and struggled to cope. They often felt guilty and frustrated for not working.

"I tried to get a job, and no one would take me on, because, it's like, "oh well you need to be here, all the time, we don't want you having days off", and "oh, you know, we can't keep, expecting you not to turn up,

because, you gotta have some treatment and stuff", there's no jobs out there that are flexible, to fit around what I need"

Robyn, F, 41 – 50 years

However, in some cases not working was beneficial for the participant. For Danny, it gave her the opportunity to look after her health.

"[S]ince I've stopped work, I've been able to do more things, and I have to, I have noticed that I do fatigue more easily … so I'm just having to pace myself a bit"

Danny, F, 51 – 60 years

A few of the participants shared how being diagnosed with CF affected their finances. Robyn was unable to find a suitable work position and if she did, she would lose out on benefits. As a result of this, she felt stuck, feeling that she was unable to improve her financial situation, leading her to blame her CF for her circumstances.

"I can't go and get a job, to make things better because, they will take it away from the benefits, so I'm stuck. I feel so trapped, and then I blame my illness. Yeah, all of that side of it, I then start reverting back to my illness, like, if I didn't have this CF, I'd be able to do anything, you know, and not be told you can't do this, you can't do that, or if you do this we'll take that away"

Robyn, F, 41 – 50 years

At one point, Jo was only receiving statutory sick pay because of the time off she needed from work. This had resulted in her getting into debt in order to pay her bills. Jo also shared the financial pressures caused by the cost of prescriptions, travel to and from the hospital and parking.

> *"I got into a huge amount of debt with all the bills, because you know if you're only getting like sixty, seventy-pound a week to live on, and that all took a long time to sort out and to pay everybody back, so you know, I'm never going to be rich, unfortunately, but yes, so it does have the capacity I think, to really affect your whole, your financial situation, which is one thing that does upset me."*

Jo, F, 41 – 50 years

Prior to her diagnosis, Jordon and her family were looking to move. They were investigating life insurances and financial investments as she and her husband were self-employed. However, as soon as she received her diagnosis, she was unable to get a mortgage or life insurance. Frankie also talked about the challenges he faced when trying to get a mortgage.

"[T]he only time it did affect me, was when I was looking for a mortgage, and it said on there do you know of any, things, on there, of course I just put mild CF, and that was it, stopped dead"

Frankie, M, 31 – 40 years

Danny shared how she and her husband had differing views when it came to money. Her husband was interested in investing their finances, whereas she wanted to spend the money, as she was uncertain about her future.

"[F]or a long time when I was working and we were reasonably financially well off, we'd talk about planning for the future. My husband's always been one for piling money into pensions and things like that and I kept saying well look I don't want to pile all our money into pensions, I don't know what the future holds, I don't want to make money tight for us now for a future that I might not have and I don't think he always got that. I can see where he was coming from, and maybe he got a bit where I was coming from, but somehow it didn't always sink in"

Danny, F, 51 – 60 years

Changes to relationships after diagnosis

Participants shared how their CF not only affected them but also their relationship with others, specifically with their partners and their family. On hearing of the

diagnosis, some close or extended family members tended to distance themselves from the person with CF.

> *"[S]ome people just don't like to hear other people's illnesses, do they? They are just wrapped up in their own world and they don't want to hear, all that negative stuff, so. I've got my brother and my parents, my nephew and my son obviously and [husband's name], although what he's done, he still, he still wants to know what's happening and his mum, and that's it. The rest of the family, just, quietly backed away. The less they know, the better they are, and I'm like, so be it. I'm not gunna try, I'm not gunna force it on 'em"*
>
> Robyn, F, 41 – 50 years

Two of the participants believed that their CF had an impact on their spouse wanting to separate from them. Sam thought her CF inhibited her relationship with men as she was embarrassed about her cough. Participants reported that their partners struggled to know how to deal with their CF and how to support them. Morgan discussed disagreements with her partner about him helping around the house and getting frustrated with him having to stay away from her when he was unwell. Leslie talked about not being in a relationship for a while. Whilst she had not put this solely down to her CF, she believed that it might have had an impact, as bringing up phlegm is unpleasant.

"[O]bviously my parents, and my husband, couldn't cope with it, and I truly believe that it's one of the reasons why he left me [tearful] [pause], he just couldn't cope with it, 'cause he had just lost his dad [pause] to cancer and he couldn't deal with it, and I think, I just don't think he wanted to see me, deteriorate, you know, and go through these bad times, which so annoyed me, 'cause I thought I need you more"

Robyn, F, 41 – 50 years

"Yeah well it definitely probably impacted on my relationship with my son's, mother, yeah for sure, she had it in my mind she wanted try to get away from me I expect, I don't know, it was a bit of a shock to me at the time, well not a shock, but it was, how all of a sudden I really deteriorated"

Charlie, M, 41 – 50 years

Most men with CF are infertile, however, the effect of CF on the fertility of women is less clear. At the time that the participants may have been trying for children, most were unaware that they had CF. Later, when they were diagnosed, they realised their CF was the reason for being unable to naturally conceive children.

"[W]hen we were young in our twenties, mid-twenties to sort of thirty, we tried having children for about, I dunno, yeah three-four years something like that five years, and it didn't happen … we both decided

not to get tested we just, if it was going to happen it was going to happen, and if not, it didn't, and it's only now, having been diagnosed, that you realise your sperm counts really low. So that's probably the reason why, I never had children"

Jamie, M, 61 – 70 years

"[W]e went to the gynaecologists and they checked, and they said 'oh your fallopian tubes are clogged up', didn't know back then, why, so I had surgery, had them cleared out, got to wait for your body to settle down, and then, get them checked, and by the time I went back to get it checked they said 'oh they are clogged up again', 'oh', so I didn't want to keep going through surgery, so we just kinda boshed the whole idea of having more kids, and obviously when I was diagnosed, with CF, it explains, 'cause obviously it was all the build-up of mucus"

Robyn, F, 41 – 50 years

Riley was diagnosed with CF as a result of struggling to conceive children naturally with his wife. For him, the diagnosis was less of a shock than was being unable to naturally have children. He had always felt healthy and being unable to father children naturally challenged his masculinity *"[M]ade me feel less of a man really, like challenged my manlihood"* (Riley, M, 41 – 50 years). He talked about it being *"probably the hardest thing to deal with alongside not being able to have children"*

(Riley, M, 41 – 50 years). For the participants wanting to have a family, fertility issues were an ongoing worry.

> "[*W]e were on about trying for another baby, and I said 'yeah that's fine', I've not been on anything for the last couple of years, and nothing's happened, and I'm like, some parts of me think oh my God is it me, because I know it is harder to conceive with CF"*
>
> Max, F, 21 – 30 years

Of the twelve participants that had children, three referred to their partner's children as their own and nine had their own biological children. Four of these nine participants expressed concern about their children's ill health. They worried that their child's symptoms were similar to their own before being diagnosed with CF. Parents did not want their children to have a similar fate to themselves.

> *"[W]hen he [son] gets a cold, you can hear it on his chest and it really sounds familiar to me, and I worry about him, that he's got it [CF], even though I don't think he has, but then nobody thought I had it"*
>
> Charlie, M, 41 – 50 years

The future – feeling hopeful or hopeless

Some of the participants talked about making plans for their future. This included planning holidays, considering downsizing their homes or thinking about starting a

family. Jamie talked about looking forward to the future, doing activities that he enjoys and overall spoke positively about his future.

> *"[L]ooking forward to the future, looking forward to, the kids getting married, looking forward to retiring, and, doing, things that I like to do, play squash, play racquetball, play tennis, gardening, stuff like that. Anything that's, anything in the future, that is new enjoy doing really. I don't think about what might happen in the future, there's no point, there's no point doing that"*
>
> Jamie, M, 61 – 70 years

Two participants talked about how once they had found out about their diagnosis it had completely changed their life plans. Robyn had wanted to immigrate with her husband and son to another country where she and her husband had grown up when they were younger. When Robyn was diagnosed these plans were put on hold as she realised she was better off staying in the UK. Worries such as the cost and standard of medical care in comparison to the UK impacted Robyn's decision to stay, which left her angry with her CF. She blamed herself for not being able to give her son the life she wanted for him.

> *"I don't want to put myself in, an environment that's gunna, probably make it all go wrong, but then I was angry then because it's like, this damn CF … I just wanted to see my son grow up in a different environment, and we've got a family there as well and I blame myself*

now, although he's doing alright, he's okay, I wished, it wouldn't, it didn't happen to me [pause], because I ruined all that"

Robyn, F, 41 – 50 years

Despite having always had CF, Robyn was resentful of what had become of her life after receiving her diagnosis. She was conflicted by her circumstances, whilst wanting to live life, she chose to be passive and exist, waiting possibly for her own death whilst watching others enjoy their life. She felt that she has aged significantly from not doing anything.

"I'm just sort of like, waiting, sitting around and waiting, not living life, just watching others, live life, and obviously I'm encouraging my son to, go live his life, whether it's in the forces or [country name] or [country name], I want him to, you know, experience life and have fun, have an adventure, just like what I did, when I was younger, and I'm just sitting here watching it, waiting, you know I get bored. I want to do something. I feel, I'm only forty-seven and yet I feel like sixty-seven, do you know what I mean, I feel so old, because I'm not doing anything. I'm sitting around"

Robyn, F, 41 – 50 years

Similar to Robyn, Alex talks about not having anything to look forward to in life and expecting that her health will deteriorate.

"… [I]t's more the death side of it, than what is there to look forward to, there ain't nothing, in my eyes, there's nothing to look forward to, because all I can see is it gets worse, it don't get better, you know I'm not going to kid myself"

Alex, F, 41 – 50 years

Others talked about their worries when they were unwell or in hospital and wondered if they would get better or if their death was imminent.

"I kept thinking how many chest infections you can have before you've damaged all of your lungs and then I was like well you can't, I know in my head you can't put a number on, right, you can have twenty infections in your life, and that's it?"

Morgan, F, 31 – 40 years

Participants realised that death was an inevitable part of their illness and had come to terms with the fact that their life expectancy was likely to be shorter. Some shared how they no longer feared their death.

"I don't intend to go soon, however, I am kind of realistic in the sense that, you know, I'm probably not going to live until, I mean my Nan is in her eighties and still going, just about. So, I think you know that's unlikely"

Jo, F, 41 – 50 years

"I've got no fears about dying any more. Don't want to; I want to live for another twenty years, but you know, if it happens it happens, so I'm not worried about it"

Chris, M, 61 – 70 years

Max shared that even though she was not concerned about her death, she was worried about dying unexpectedly and not having made the necessary preparations.

"[I]t seems to be a lot of my [pause] issues so to speak, come from [pause] death, but I'm not scared of dying, 'cause I know that it's going to happen, and that doesn't scare me. I think it's more a case of if it happens suddenly and nothing is set in place and I've not got control over it, not that you control when you die anyway"

Max, F, 21- 30 years

Two of the participants, Morgan and Max, talked about preparing for the inevitable so it is not on their mind. *"… [T]hinking about the future and what do I need to do, and whatever I need to do I need to do it now before I'm either too sick or I'm dead"* (Morgan, F, 31 – 40 years). Max talked about having to make a will for her son before she died, whilst Morgan also talked about wanting to organise her own funeral.

"… [I]t seems really weird now 'cause I feel fine again, but like even going as far as planning a funeral … Because I'm such a control freak that it has to be done how I want it, and that would be the last thing that I can control, so I want it done how I want it, and I want the music that I want, I want the flowers that I want, and I want, yeah I want to say something"

Morgan, F, 31 – 40 years

"So it's like who at twenty-one, twenty-two, wants to go and make a will for what's going on with their son. Things like that, but it is what it is, and they're the things I have to do to make sure, if anything did happen to me or when something happens to me, everything is in place, so"

Max, F, 21 – 30 years

Some of the participants shared how they wanted to make the most of the time that they had left, therefore trying to do things whilst they had good health.

"We're going to [country name] in April, 'cause we just said right, let's do things whilst I'm well enough, because we really don't know what's around the corner"

Jordan, F, 51 – 60 years

4.3.2 Treatment burden

Participants shared how the number of times they had to come into the hospital for check-ups, annual reviews and inpatient treatment, usually lasting between ten and fourteen days was an inconvenience was an inconvenience. Whilst they realised that the review appointments were for their benefit, some found them to be a burden.

> *"[I]t's almost a bit of a burden … Because I have to come here. I know that sounds awful 'cause I know everything they do is with good intentions. But yeah to come here for check-up's and all the annual reviews and so on and so forth"*
>
> Riley, M, 41 – 50 years

Ten participants shared how taking their medications and doing treatment can be a burden. Some spoke of the vast number of tablets or treatments they had on a daily basis. Others found that taking medication was a tiring and often intrusive chore. They talked about treatment being time-consuming and requiring planning.

> *"I do a nebuliser each morning, you know, I do my puffer first and then my nebuliser, and then it takes a bit of a chunk of the morning, by the time, 'cause I'm very old fashioned I have a bath, so you know, maybe I get up, wake up at seven, and it's probably two hours of, yeah okay, bath and breakfast, and you know, my treatments and physio, so it's quite a chunk actually. And I'm sure people do it much quicker than I*

do, it is quite intrusive, so you know, I'm you have to sort of think, plan ahead a bit"

Sam, F, 51 – 60 years

"[I]t's terrible, because you do them at six in the morning, and you do them again at midday, and at two o'clock, and then again at ten o'clock at night. You have to mix them up, then you have to put them in yourself, and, the drug itself makes you tired, and then like, the fact that you've got to do it three times a day, at unsociable times, is even worse"

Chris, M, 61 – 70 years

At times, participants reported that they were complacent around taking their medication. For a few, the amount of medication, hospital appointments and time involved all became too much and they chose not to comply with their treatment. Max reported that after taking her medication, she felt as though she was going to be sick and therefore sometimes did not take them to avoid feeling that way. Taylor talked about being complacent about using her nebuliser, particularly when she felt well.

"[S]ometimes you can be a bit negligent and think, oh can I be bothered doing the nebulising just now, can I do it, well I'll do one today instead of two"

Taylor, F, 51 – 60 years

This feeling was also shared by Riley. Whilst he had good intentions of taking his medications, he was unsure of its benefits and did not notice the effects of not taking them. Therefore, it became easy to forget to take his medication.

"I have good intentions of taking it [medication]. I take it for three or four days and then not, but I don't feel any worse for not"

Riley, M, 41 – 50 years

Some participants discussed the importance of understanding the purpose of their treatment and what it was trying to achieve. At diagnosis, Leslie found it difficult to comprehend that she needed to have regular medication where before diagnosis she did not need to do so. Leslie struggled with taking her medication as coincidently the more compliant she became, the less well she appeared to be. Over time, she found that her health stabilised and she understood why she needed to take her medications.

"[I]t was just adjusting to not having to do anything to having all of these tablets and all of these inhalers and all these nebulisers that I had to do. I was like, well I've sort of coped this amount a long time without them, why do I suddenly have to do them now and why is it all so important"

Leslie, F, 21 – 30 years

Participants often tended to let their health deteriorate, until it was far worse than it should be before they took any action. They reported that this was because they were either in denial about their health status or that they had not realised how unwell they were. However, once participants identified signs that their health was declining, they took action. This involved either taking medication or getting in touch with the hospital. Six participants described their first hospital admission as frightening because of not knowing what to expect. For Sam, it was terrifying as she had not gone through it before. Feeling breathless was a scary experience and she had not realised quite how unwell she was. Similar to Sam, for Frances and Morgan their hospital admission was a reminder of their CF and that they had something wrong with them. Danny worried about how going into a hospital was a sign of things to come in the future and questioned if it was the start of her health declining.

> *"[I]f you're in hospital you must be sick, you must be sick enough to be in hospital because they don't keep you in for the fun of it, but I don't want to be sick, and in my head I'm not that sick, and so I would struggle with being in there, and I don't like the fact that um, I just don't, I like to be independent, I want to do it all myself, so yeah I struggle"*
>
> Morgan, F, 31 – 40 years

Although Morgan appreciated that in hospital there are set mealtimes and the medication is administered to you, she found this difficult to deal with. For Morgan, it felt as though she was a child as she would lose her independence and control when she got admitted into the hospital. Leslie found hospital admissions difficult as there

were restricted visiting times and her family and friends lived far away. However, with each admission, she found it easier to cope as she became familiar with the ward staff and the processes. As much as some of the participants reported that they did not like going to the hospital, they found that having a course of IV antibiotics was beneficial and made a significant difference to their health. Participants often reported that they felt younger and full of energy after coming in for IV's, not realising how unwell they had become.

> *"[O]nce all the drugs started working, I never felt so well in years, when I came out of hospital that time"*
>
> Charlie, M, 41 – 50 years

> *"I would physically say you bounce as you walk out the door because you feel so, you know, so full of vim and vigour … I've always got the hugest smile on my face as I walk out that door, because I think hi everybody, it's been lovely seeing you, look how good I am, you know. You just feel a million dollars"*
>
> Taylor, F, 51 – 60 years

One of the problems reported by participants of having IV's for two weeks is that it could become an inconvenience and get in the way of plans. Jamie could not continue his usual exercise routine playing racquet sports such as tennis and squash for worry of dislodging his IV line. Leslie shared how she had plans to attend a

festival but had to go to hospital for IV's. As she did not want to miss the festival, she decided to attend it with her IV's attached to her, even though this was an inconvenience.

In summary, this theme explores the impact and the challenges that participants reported facing on a daily basis in all aspects of their life. They experienced good days and bad days in relation to their illness, finding the unpredictable nature of their CF frustrating. Managing day-to-day life and planning for social events was a struggle, as participants would not know if they would be well enough to attend. They had reassessed the appropriateness of their job roles and the environments that they worked in. Taking time off from work for appointments and when they were unwell was an inconvenience, particularly if they had to catch up on work afterwards. A few faced challenges of finding a role that was flexible and could work around their health and treatment regime. For some, their CF affected their finances and life plans including taking out mortgages and life insurance. It also impacted on relationships with others, particularly with partners and family. For some, it affected their fertility and ability to conceive children naturally. Some participants spoke positively about their future, whereas for others, the future brought worries around health and life expectancy. Some had come to terms with their shortened life expectancy and no longer feared death, making preparations including wills and funeral arrangements. Participants wanted to make the most of their time left. They talked about the treatment burden and annoyances of having to attend hospital for appointments and reviews. Hospital admissions were described as a frightening experience as participants were unaware of what to expect.

4.4 "I no longer wish to argue with it"

This theme talks about participants' physical and mental acceptance and adjustment around their CF. Acceptance of CF was not limited to being diagnosed with a chronic condition. It involved the acceptance of the different decisions one might have to make as a result of their diagnosis, the changes in lifestyle that individuals might have to adopt and the acceptance of the limitations of their condition. For some, acceptance and adjustment happened soon after their diagnosis, whereas for others this process took longer.

4.4.1 A new way of looking after self

Jo was one of a few participants for whom it took several years to come to terms with her diagnosis and to accommodate CF in her life. When Jo was initially diagnosed, she understood that she had CF, however, the reality of how this would impact her life and what changes she might have to make were underestimated. Jo tended to choose to socialise with friends and have fun, prioritising other aspects of her life, rather than looking after herself, which was often at the cost of her health.

> *"It was that I was overdoing it. I wasn't accepting that there was sort of certain limitations … I knew that I had it [CF] but my compliance, my drug compliance was pretty erratic … I accepted it physically, it was kind of mentally, that I suppose … and there's things that you need to do, like physio and drugs and inhalers and nebulisers and stuff, that's all going to take time and effort, and I think my problem was that I didn't allow for that in my life, so that was why my compliance was so low"*

Jo, F, 41 – 50 years

Jo struggled with her CF, describing it as *"a small, obnoxious child*" (Jo, F, 41 – 50 years). She did not want her CF to take over and battled with finding the right balance between living her life whilst managing her CF.

> *"I had my battle with CF, because it was about you're not going to define me, and almost CF saying to me well yes you are, you are going to listen, you are going to have to learn to make adjustments"*

Jo, F, 41 – 50 years

Over time, Jo learnt to accept her CF, accommodate it and make adjustments to her life.

> *"I no longer wish to argue with it or to struggle with CF"*

Jo, F, 41 – 50 years

Similarly, other participants recognised that CF was an aspect of their life, but they did not want it to take over or become a part of their identity.

> *"I am not CF, it is a condition that I happen to have, but I am still me"*

Jordan, F, 51 – 60 years

“So now I’m trying to get a balance between yes I’ve got CF, that’s fine, this is what I’ve got to do to stay well, this is what I’ll do if I don’t feel well, and see if I can just crack on with normal life the rest of the time”

Morgan, F, 31 – 40 years

Some of the participants talked about developing an awareness of not looking after themselves, which impacted on their health. A few participants shared that they were always busy doing things which led to burnout. Where previously individuals may have tried to overload themselves when they felt well, they had to learn a new way of balancing and pacing themselves. Others talked about not eating properly, stress or not doing their exercises. Participants talked about the practical steps they took to take care of their physical health and the lifestyle changes that they made. This included quitting smoking, eating meals at the right times, avoiding processed and junk foods and having good quality sleep.

“I think initially we were really just, when I’m well I just need to work, work, work, work, work, ‘cause I don’t know when I’m not going to be well. But now I think I’ve got a better understanding of what to look for”

Jordan, F, 51 – 60 years

Robyn reported that making changes to her lifestyle lead to positive outcomes that allowed her to gain control of her emotional wellbeing.

"It was really really tough to accept it, and it took me I think a good three years, to get over it emotionally, and to adjust to it, physically and emotionally … obviously over the years of having this, and realising that, my whole lifestyles got to change, the diet, activity, work, and everything, to make it more bearable, and then, obviously if that works out and goes, smoothly then the emotional side will, I'll start getting in control of it again"

Robyn, F, 41 – 50 years

Most of the participants talked about the importance of being physically active and how increasing their physical activity had been beneficial to their health. They noticed improvements to their lung function, generally felt well in themselves and were able to fight off illnesses better. Doing regular or increased physical activity was a practical step, which helped them to take or maintain control of their health.

"You know I, I really want to have a decent quality of life. I will work for it. I've got a running machine in the office up there, and a recumbent bike that [husband's name] bought, so we've done exercise, we've done, you know I take vitamin D. We just, yeah we do what we can, because we want to"

Jordan, F, 51 – 60 years

However, one of the challenges reported by two of the male participants was that despite exercise and training they were unable to gain weight or to build muscle, due to their CF.

> *"[T]he trouble with CF is, you don't build up muscle very well, that's the trouble, so, you know I can go out and cycle every day of the week and my leg muscles just don't get any better, I still struggle"*
>
> Francis, M, 61 – 70 years

Having information and understanding about their CF was considered important to help with adopting a lifestyle and routine in which to accommodate their CF and another way of maintaining control.

> *"[B]y being better informed we can actually have a better quality of life, 'cause we know how far to push it, we know what we need to look for"*
>
> Jordan, F, 51 – 60 years

> *"It's got better, I think, yeah, it's definitely better, because I understand it, it's like anything the more you understand something, you learn about something, the more you, you can deal with it"*
>
> Robyn, F, 41 – 50 years

Individuals who appeared well adjusted seemed to either accommodate for CF in their life or the impact of the condition was minimal. CF was not ignored but it remained in the back of their mind. They did what they could to keep as healthy as possible.

> *"I wouldn't say I forget about it [CF], it is always there at the back of my mind, but because I've been so fortunate and because I'm determined to try and keep well, that I've tried to live as well as I can"*
>
> Danny, F, 51 – 60 years

As part of taking care of themselves, participants talked about being aware of and trying to avoid people with colds or places where they might pick up illnesses, with the aim of trying to protect themselves from becoming unwell. Participants talked about being diligent or in some cases paranoid when they went out, particularly when shopping and were conscious of hand hygiene to prevent the spread of infections.

> *"I get paranoid going out shopping because I pick up everybody's germs, when they're coughing and sneezing, you know, it's like ah please get away from me, and guaranteed the next day I'm waking up sore throat"*
>
> Alex, F, 41 – 50 years

4.4.2 "Just get on with it"

Most participants held the attitude of 'getting on with things'. This approach was about moving on with their life, trying to make the best of the situation and maintaining normality rather than complaining or focusing on the negatives. Participants did not necessarily need to have a positive attitude to 'get on with things'.

> *"I'm just like just get on with it, like shit happens and then you die, so there's obviously a reason this stuffs happened, so you just get on with it. You can't change it [CF], so make the best of it basically"*
>
> Leslie, F, 21 – 30 years

> *"[Y]ou either sit around and mope, don't you, and say 'oh dear, oh dear, I've got all these problems' or you say 'balls to it' and get on with it, don't you, and I've you know, and I got on with it"*
>
> Chris, M, 61 – 70 years

For two of the participants, Sam and Frankie, 'getting on with it' meant continuing with life as if they did not have CF, *"I just carried on like a normal person*" (Sam, F, 51 – 60 years).

> *"I don't dwell on stuff like that, and I don't, dwell, I suppose at the time, I sort of carried on as if I didn't have it [CF] anyway"*

Frankie, M, 31 – 40 years

For the others, CF was recognised as an aspect of their lives that could not be changed. Chris talked about feeling down with aspects of his life. However, he expressed that talking about his problems to others did not help. Instead 'getting on with things' by tackling the situation head-on rather than burying his head in the sand was his approach.

> *"I do get depressed, you know, go down the pub, have a couple of pints and pick myself up, dust myself off, let's start all over again. I mean, you know, it's very difficult, because if you don't deal with it, you're going to be worse, aren't you? It's no good crying is it, saying oh I feel so ill. No good coming and talking with you people [psychologists] is it, oh my God, I don't know what I'm going to do, how are you going to help me, you can't, you can be very sympathetic and listen to me, then go home and have your fish and chips and forget me. I've still got my problems, haven't I?"*

Chris, M, 61 – 70 years

Some participants talked of taking a positive and optimistic approach, *"the cup's always half full, it's not half empty ain't it*" (Chris, M, 61 – 70 years), rather than dwelling on the negatives.

> *"[M]y nature is optimistic. I have pessimistic times, but on the whole my outlook is optimistic, so I've got through life by being optimistic and you*

know looking on the bright side and not dwelling too much on the rubbish"

Jo, F, 41 – 50 years

"I'm quite a positive person anyway, for me, when the final diagnosis came, I remember sitting here in the quiet room, and there was a lot of people in the room, and I thought what's all this, and [husband's name] sat there, and he was a bit in shock and I just said look, I said 'there's nothing to worry about', I said 'life's terminal, at least now I know what's wrong with me'"

Taylor, F, 51 – 60 years

Taylor believed that her positive attitude was the reason why she was still alive in comparison to her sister who had died from her CF as she seemed to give up.

"I think her attitude to things and my attitude, are polar opposites. And I think that's why I'm still here at the age of fifty-three and going strong, she just really did seem to just give up"

Taylor, F, 51 – 60 years

Jamie also shared Taylor's belief of how having a positive attitude keeps you well.

"I don't think it matters, what, disease you've got, you know whether you've got, cancer or you've got CF, or you know, whatever, I think having a positive outlook, is a major part of, getting well"

Jamie, M, 61 – 70 years

Participants also talked about focussing on doing practical things, which were within their control rather than focussing on the negatives of CF or its treatment.

"Looking forward not backwards, looking forward, not thinking about all the negative side of CF, treatment you've got to go through, the drugs that you've got to take, that it maybe, maybe life-limiting, maybe, but if it is, and you know, there's nothing you can do about it, except, keep taking medicine, and being, as healthy as you can, in which, okay I keep going back to it I know, but doing as much exercise as you can, that's really important"

Jamie, M, 61 – 70 years

Two of the participants, Sam and Taylor, made references to the attitude of 'getting on with things' being linked to their upbringing.

"And even though I'm feeling exhausted, pretty grotty, you know, I'm afraid it's my upbringing, you know, stiff upper lip and you just carry on"

Sam, F, 51 – 60 years

"I think that's a typically [country name] attitude as well, we always say in [country name], oh there's nothing wrong with you, kick up the backside and get on with it and that's what happens, so it's that kind of attitude"

Taylor, F, 51 – 60 years

Connected to the 'getting on with it' attitude, participants shared how CF is not often spoken about and the different reasons as to why they did not talk about it. Participants tended not to tell people that they had it. Most people's immediate family tended to know that they had CF, but in one case, the family was told that the individual has a bad lung. Some participants shared that they had CF with some friends and work colleagues, but others chose to keep information on a need to know basis. Jordan shared about how she and her family made the decision to be more positive and therefore almost stopped talking about CF. Robyn shared a similar attitude and did not want to talk about her CF, as she did not want people to feel sorry for her. But she also recognised that "what had happened had happened" and having CF cannot be changed. She had become fed up with talking about CF and wanted to move forward in life.

"I don't want to express to people all the time, oh I've got this illness, you know, get the violins out, feel sorry for me, I hate that"

Robyn, F, 41 – 50 years

Others felt similar about their CF. Max found talking about it was negative and therefore tended to avoid bringing it up within conversations with friends and family. Max and Charlie only spoke about their CF if others brought it up or if they were unwell. Frankie and Frances avoided talking about their CF because they did not want their family to worry unnecessarily. Whereas, Sam and Morgan did not want to speak to others about their CF because they felt it was boring and that people would be tired of hearing about it.

> *"I do talk about it, and [pause], like I am quite open; if somebody asks me about it, I will quite openly talk about it, but it is such a negative thing [pause] that it's like, do you know what I mean though, if you're sat with your friends having a chat and it comes up, yeah you can talk about it … But it's not something, it's not a topic of conversation that you'd bring up, because it is all doom and gloom"*

Max, F, 21 – 30 years

Max, Morgan and Frankie tended to share limited details of their CF with their friends and family, such as upcoming or recent hospital appointments.

> *"I never talk about it with the kids. I hate talking about it with anybody. I'll tell people if I get, I'll openly say I've got clinic, next week or something, and all the blokes know, but it was never like clients will say*

'oh, oh do you mind me asking why?' and I'll say 'I've got cystic fibrosis' and they're like 'oh wow, you're really well for it' and that's as far as I go with it really, I don't offer it, because I know how lucky I am, it feels wrong to play that card when I'm not ill with it"

Frankie, M, 31 – 40 years

Others chose to either keep their CF hidden or not tell others because they would not understand or it was difficult to explain it to them.

"[T]here's not many people that know I have it, but that's not because I'm embarrassed about it, 'cause I'm not, it's more because, I don't want to have to then go through explaining [pause], it just takes time doesn't it, to explain what it is that you've got"

Riley, M, 41 – 50 years

Almost all the participants expressed how fortunate they were, particularly in reference to their health. Participants considered themselves "lucky" to be as healthy as they were, to have a milder form of CF and to have lived to their current age, despite CF being a life-limiting condition. Chris shared how some of his friends had died of heart attacks when they were 50 years old. He reported how "lucky" he was to be part of the hospital system because he got regular blood tests, scans, heart, liver and kidney checks, which would identify any issues early on. Other participants referred to themselves as "lucky" for being able to successfully have children

naturally or via IVF and having “a good support network of family and friends” (Leslie, F, 21 – 30 years). Participants often used downward comparison when comparing themselves to those who had CF diagnosed as a child, or others whose CF was worse than theirs and those who had cancer. This seemed to help participants to put things into perspective and adjust better to their circumstances.

> *“[I]n comparison to cancer and heart disease … I just think it is a, you know it’s a cruel disease really, and I’m fortunate that I, you know I’ve lived as long as I have with it, and I think if I’d had a, more aggressive form or I’d had some very serious chest infections, back in my day, people who were born when I was born, in the sixties, you know I wouldn’t have made it to adulthood, so I do thank myself lucky, think myself lucky in that respect”*
>
> Jordan, F, 41 – 50 years

This theme explored participants’ acceptance and adjustment both physically and mentally to their CF. Participants made lifestyle changes to look after their physical health and employed strategies to help them manage their CF. Most of the participants highlighted the importance of carrying out a form of physical activity and noticed a beneficial impact on their health, particularly an improved lung function, the ability to fight off infections and an improved sense of wellbeing. Other lifestyle changes included quitting smoking, eating meals at the right time, avoiding or limiting processed foods and having good quality sleep. Participants also talked about having to be mindful of or avoid people with coughs and colds to prevent picking up

bugs or becoming unwell. Generally, participants talked about having a positive outlook. This attitude was often linked to a ‘getting on with things’ approach. This was about trying to make the best of the situation and maintain normality rather than focusing on the negatives. Participants tried to focus on practical things that they could control rather than dwelling on things outside of their control. They tended not to tell people that they had CF or shared limited details to avoid unnecessarily worrying them, boring them or to avoid it being a negative topic of conversation. Most of the participants talked about how lucky they were to have a milder form of CF and to have reached the age that they had.

5.0 DISCUSSION

This research aimed to understand the lived experience of receiving an adult CF diagnosis and what it means to live with the condition. Sixteen participants, ten women and six men receiving their care from a regional CF centre in the UK were interviewed about their experiences. Their accounts were analysed using thematic analysis. Four themes were identified: “No, you can’t possibly have CF”, Emotions around diagnosis, “It did kind of take over my life” and “I no longer wish to argue with it”. For some adults, the diagnosis of CF was a lengthy process involving multiple interactions with healthcare professionals, from childhood through to adulthood. Often within this, people felt that they were not taken seriously or were dismissed. Being given a diagnosis was seen as a significant life event for some with its impact felt in all aspects of their lives, but for others, it had a limited impact. The findings presented in the previous chapter will be discussed in relation to the research questions and the implications for health psychology practice. The study’s strengths and limitations are reviewed and followed by suggestions which could direct future research and highlight recommendations for good practice.

5.1 Research question 1: What is it like to be diagnosed with cystic fibrosis in adulthood?

Predominantly the participants in this study had poor health before diagnosis, commonly reporting recurrent coughs and colds from childhood. When medical advice was sought from their GPs, individuals reported a misdiagnosis or felt they were dismissed, in some occasions leading to mistrust and loss of faith in healthcare professionals.

Diagnosis was often a lengthy process, which was consistent with the findings of Blunt *et al.* (2008). Not being taken seriously was a common narrative also found amongst parents of children and adults with CF (Giengedal *et al.*, 2003). Participants were told that they were “too old” or “too well” to have CF, which supported previous findings of Widerman (2002). Participants were usually diagnosed with CF following recurrent poor health, an acute illness episode, the diagnosis of a sibling or close family member or through infertility. Many of the participants spoke of their pre-diagnosis experiences at length and how they made sense of these experiences at or post-diagnosis. These findings support previous research that the pre-diagnosis experience is an important time period with implications for future relationships with healthcare professionals and individuals’ readiness for education and support (Widerman, 2004).

For most of the participants, the diagnosis presented a dichotomy. It brought a sense of relief, but also shock and distress. For some, this was seen as a life-changing experience, which was also documented in previous research of families caring for a child with CF, where the diagnosis of CF was described as a “life-shattering” experience (Carpenter and Narsavage, 2004). Individuals experienced a mixture of feelings from shock, relief, denial or anger and questioned ‘why me?’ or ‘why did they get missed?’. These findings were consistent to the diagnosis experiences reported by adults with CF (Widerman, 2002), parents of children with CF (Jessup *et al.*, 2016; Jessup and Parkinson, 2010; Carpenter and Narsavage, 2004; Giengedal *et al.*, 2003; Jedlicka-Köhler, Götz and Eichler, 1996), of other chronic illnesses (Silva *et al.*, 2018; Chircop and Scerri, 2017; LeBlanc *et al.*, 2016; Liamputtong and Suwankhong, 2015; Ansari *et al.*, 2014) and with the five stages of grief (Kubler-

Ross, 1969). An unknown prognosis was a worry for participants, consistent with Widerman's (2002) findings in which participants wanted to know how long they expected to live. Usually, this was a question that healthcare professionals were unable to answer.

A few participants reported that receiving their diagnosis was insignificant. Whilst the understanding of the diagnosis being inconsequential is limited, several speculations could be considered, such as the impact of symptoms at the time of diagnosis on the appraisal of the illness. For example, if an individual felt well and was minimally impacted by the symptoms associated with CF at the time of diagnosis, one may expect that the likely impact of the diagnosis would be irrelevant. Equally, if before the diagnosis an individual experienced numerous symptoms indicating that there was something wrong with their health, it may lead them to anticipate a diagnosis and therefore, it could have a minimal impact.

Interestingly, it was observed that the participants who commented on the insignificance of their diagnosis were mostly male. It is worth noting that there were only a small number of males within this study and the aim of qualitative research is not to make generalisations. Living with CF has been evidenced in previous research to have a greater emotional impact on adolescent girls in comparison to boys (Patterson *et al.*, 2008), therefore, there could be a greater emotional impact on adult women. Another possible reason is that the male participants may have downplayed their symptoms and reactions to the diagnosis to maintain their masculine identity, particularly whilst being interviewed by a female researcher. The reason why these participants reported that their diagnosis experience had a minimal impact is an

interesting area and may be useful to explore further with a greater number of male participants to see if there is a difference between genders and their response to the diagnosis.

The need for information was prevalent throughout participants' narratives. At the time of diagnosis, there was a mix of knowledge amongst participants; some did not know about CF, whereas others had a basic understanding. Participants shared overall that there was a lack of information about CF available to them. Those who looked for information on CF on the internet reported that what they found was irrelevant, as it did not correspond with their symptoms or experiences. Some had been given information verbally which was difficult to process and overwhelming, particularly soon after receiving a diagnosis. These findings were consistent with the CF research in childhood (Jessup *et al.*, 2016; Jessup and Parkinson, 2010; Jedlicka-Köhler, Götz and Eichler, 1996). It also mirrored those found in previous adult CF literature (Widerman, 2005; 2003; 2002) where over two-thirds of those interviewed reported that they knew little or nothing about CF at diagnosis, three-quarters were active information seekers but over half were less than satisfied with the information that they found (Widerman, 2003). Previous participants had expressed frustration at either being given no information or being given cartoons directed towards a paediatric audience (Widerman, 2005). Individuals were interested to know practical information such as what life would be like, longevity, treatment options, research directions and information that would help to construct hope (Jessup *et al.*, 2016; Widerman, 2005).

The findings from this study echoed 9 of the 10 cross-case themes described by Widerman (2005): awareness of death, change, difference, family indifference, intrusion, isolation, normalisation, time and uncertainty. There were also some similarities with the additional themes reported by Widerman (2005) related to gender, illness severity and medical caregivers. Overall, much of the findings from this study were consistent with Widerman's research despite her work being carried out in the USA over 15 years ago with a different medical system to that of the UK. This suggests that the experiences of those diagnosed with CF in adulthood could be considered a universal experience within Western countries.

There were many similarities between the themes of this study and Widerman's research. For instance, in both studies, participants spoke about the adopted changes to their lives following a diagnosis, including their careers and lifestyle choices. There were also several differences noted between the two studies. For example, in Widerman's research, participants spoke of the positive changes CF brought, such as valuing time and relationships, whereas in this study, participants did not reflect on the positives of their CF in the same way. The participants in this research talked about CF being a positive in relation to finally getting a diagnosis and it being the missing piece of the puzzle in terms of explaining illnesses or random symptoms. They also shared that the diagnosis enabled them to access support and treatment. Widerman reported on the theme distraction and found that most of her participants were diagnosed in their late twenties, at a time where CF competed with typical life stage demands for attention, something that was not raised within the current study. Widerman's study also reported on gender-related themes: the difficulty of parenting and the fear of rejection, and illness severity themes: self-

pity/wanting sympathy, none of which were expressed in accounts from the participants in the current study. The differences found between the two studies could be due to the age of participants at the time of diagnosis. In this study, most of the participants were between their late twenties to mid-fifties at diagnosis (mean age at diagnosis: 38 years), in comparison to late twenties (mean age at diagnosis: 28 years) in Widerman's study. In addition, whilst Widerman's research focussed solely on the adult diagnosis experience, the present study provides valuable insight and knowledge on the lived experience of those diagnosed in adulthood. It has specifically focused on the impact of CF on an individual's day-to-day life and their acceptance and adjustment around it. This is important as there is no known published research on the lived experience of those diagnosed with CF in adulthood. The findings are vital for healthcare professionals providing treatment for those diagnosed in adulthood to ensure that the appropriate care is provided.

5.2 Research question 2: What impact does cystic fibrosis have on an individual's day-to-day life?

The findings demonstrate that for many of the participants, CF had a significant impact on various aspects of their lives including work, travel, relationships, finances, and health and wellbeing. This was similar to previous CF research in adulthood (Widerman, 2005), children's research (Jamieson *et al.*, 2014; Gjengedal *et al.*, 2003; Christian and D'Auria, 1997), adults living with CF with a childhood diagnosis (Cordeiro *et al.*, 2018) and chronic illness research (Due-Christensen *et al.*, 2018; Marx *et al.*, 2016; Paturzo *et al.*, 2016; Ansari *et al.*, 2014; Giacomini *et al.*, 2012). The unpredictable nature of their condition was a frustration for participants and made planning a challenge. These results were consistent with the findings of

Giacomini *et al.* (2012). The impact CF had on an individual's work was most commonly discussed by participants, who often had to reassess the appropriateness of their job roles and re-evaluate career plans. CF brought with it the challenges of having to take time out due to appointments or poor health. The embarrassment at work around their cough was reported by participants in the current study. This was consistent with previous research of those diagnosed with CF in childhood (Jamieson *et al.*, 2014; Christian and D'Auria, 1997) and living with CF in adulthood, (Cordeiro *et al.*, 2018) who felt embarrassed with their cough.

Infertility was reported to be a particular concern for one of the participants, even more so than his CF diagnosis. When others received their CF diagnosis, they were able to make sense of their inability to have children, once they found out that infertility is a common characteristic of CF. Devastation by sterility was also found in previous research (Widerman, 2005).

CF was found to have a significant impact on travel insurance, mortgage applications and finances. Travelling required a lot of planning from participants such as making sure that they were well enough to travel and that they had sufficient medication. Financial pressures were a concern when participants were unable to work due to poor health. In Widerman's (2005) study, a theme was identified around insurance worries for medical cover, which was not reported in this study as care for CF patients was provided under the NHS in the UK. Treatment was reported as being a burden due to the vast number of tablets or regimes that they had to complete daily, such as airway clearance or chest physiotherapy. For some, taking medication was tiring, a chore, intrusive, time consuming and often required planning to fit it within

the day. The burden of treatment related to CF is also documented within the children's CF research (Gjengedal *et al.*, 2003). A noticeable difference in the experiences of adults living with CF, who were diagnosed in childhood, was the distress of waiting for a transplant and the uncertainty of the outcome following surgery (Cordeiro *et al.*, 2018). This was not a shared experience with the adults interviewed within this study. This is likely to be explained by the higher lung function of those diagnosed in adulthood and therefore, the topic of transplant becoming a less likely conversation.

Almost half of the participants in the current study described their first hospital admission as a frightening experience as they did not know what to expect. For some, it was a reminder of their CF and of what might come as their health deteriorates. Some lost their sense of control whilst in hospital. Whilst these findings are not known to be documented in the adult literature, they are similar to those reported by the parents of children with CF, prior to the child's first hospital admission (Fixter *et al.* 2017). In Fixter *et al.*'s study, parents reported receiving minimal preparatory information and any that was provided was mostly medical and practical with the psychological aspects of the admission being overlooked. This was raised in the current study by Sam, who felt there was a gap in the provision of psychological support, particularly as she had been admitted into hospital on a Friday. She felt whilst the nurses and doctors had a job to do, the emotional support was lacking. When asked what this support would look like, Sam wanted the opportunity to be able to speak to someone, to be periodically asked how she was doing and if there was anything that she wanted to discuss. In Fixter *et al.*'s (2017) study, parents spoke of their need to be involved in their child's care and that feeling

in control was important. Some participants reported frustration in giving control to the medical team. That was also reported within this study where Morgan described how hospital admissions left her feeling out of control, as she was unable to administer her medication whilst in hospital. One of the parents in Fixter's study talked about learning how to administer IV antibiotics, which allowed her to take back some responsibility. Morgan spoke about being able to administer her medication in the hospital as a way that would help her feel more in control. The research documented that participants took practical steps to look after their physical health and made lifestyle changes following their diagnosis. For most of the participants, this involved a form of physical activity, which they noticed had a beneficial impact on their health, supporting previous research (Widerman, 2005). Participants tended to avoid others with coughs and colds to protect themselves. These findings are similar to that of the children's CF literature (Gjengedal *et al.*, 2003).

5.3 Research question 3: How do individuals adjust to living with cystic fibrosis in adulthood?

Participants tried to maintain a positive outlook and have the attitude of getting on with things to help them adjust to living life with CF. This involved making the best of the situation and maintaining normality rather than focussing on the negatives. This perspective was consistent with the childhood CF research (Jamieson *et al.*, 2014; Jessup and Parkinson, 2010) and for those living with CF in adulthood with a childhood diagnosis (Cordeiro *et al.*, 2018). Research found that parents and older children with CF had a statement or cliché that they referred to in order to make sense of their experiences, this included "That's life" or "It was meant to be. Just do it" (Jessup and Parkinson, 2010, p.360). A possible explanation for adopting the

getting on with things attitude is participants desire to maintain a ‘normal’ life, a phenomenon referred to as normalisation. The importance of the normalisation process was discussed by Gjengedal *et al.* (2003), who state that stigmatised people feel as normal as anyone else. They go onto explain that the feelings of difference come from the attitudes of the ‘normal’, which have been constructed in social relations. Normalisation also serves the purpose of promoting hope for the future (Gjengedal *et al.*, 2003). Robinson (1993, cited by Gjengedal *et al.*, 2003) stated that healthcare professionals were more inclined to focus on the problems rather than getting on with life. This would contribute to disrupting individuals’ practices that supported normalisation. Robinson goes on to explain that healthcare professionals mainly do this because they interpret the normalisation process as denial. Normalisation was also a theme reported within Widerman’s (2005) research.

Despite many reporting that CF had impacted various aspects of their life, it was of particular interest from the findings that half of the participants reported that CF has had little impact on their day-to-day life. Participants referred to themselves as having “mild CF” or as “light sufferers”. Some participants discussed how CF has little or no impact on their life as they were still able to do what they did before their diagnosis. Most people would assume that having a chronic illness would have a substantial impact on one’s life. One of the reasons that may explain these findings was that the symptoms of their condition were milder in comparison to others interviewed, so treatment is minimal and therefore, little impact is felt in their day-to-day lives. Whilst recognising the benefit of having regular appointments, this group of participants shared that they found having to come into hospital for them an inconvenience and burden, usually having to take time off from work or use annual

leave. Participants reported how lucky they were to have reached the age that they had got to and to have a milder form of CF. This was consistent with the findings reported by (Blunt *et al.*, 2008).

5.4 Models and theories in chronic illness literature

A combination of three existing models within the chronic illness literature can help to explain the findings of this research: chronic illness as biographical disruption (Bury, 1982), the shifting perspectives model of chronic illness (Paterson, 2001) and a working model of adjustment to chronic illness (Moss-Morris, 2013). The findings from this study showed similarities to that found by Bury (1982). Participants in this study talked about pre-existing symptoms such as persistent cough and regular colds from childhood. This supported Bury's statement about how a non-communicable disease does not just suddenly happen, but that the impact of the condition is steadily felt. Within this study, participants spoke about the feelings of shock, disbelief and anxiety leading onto relief, which was also reported by Bury. The uncertainty around their prognosis following their CF diagnosis was commonly reported by the participants. Some were found to be searching for meaning to questions such as 'why me?' and 'why did I get missed?'. This mapped the next stage of the model, which was centralised around the emerging disability of the condition and the problem of uncertainty around the impact and the course of the condition, with participants searching for the meaning of events. Bury reported instances in which individuals were 'taken over' by their disease, a perspective shared by many of the CF participants in this study. Bury also discussed how access to medical information about the illness allowed individuals to conceptualise the disease separate from their individual self. Within this research, participants wanted

to know information about their disease but were limited by the information that is known, often with medical professionals learning with the patient as time goes on. Lastly, the mobilisation of resources and the availability of social support networks was a key factor. This summarised the experiences of several of those diagnosed with CF in adulthood. They discussed the disruption that the illness caused in their everyday life with normal activities, due to the unpredictability of the illness, with making plans, excessive tiredness or embarrassment faced due to their symptoms, such as coughing or having to frequently go to the toilet due to bowel problems. Participants spoke about the impact CF had with their work and how they have had to make adjustments such as finding an alternative job or adjusting their hours. In summary, the chronic illness as a biographical disruption model explains the initial aspect of receiving the adult CF diagnosis and some elements of living with the condition. However, the shifting perspectives model of chronic illness was able to explain participants' experiences of living with a chronic illness.

The shifting perspectives model fits well with the experiences of adults with CF, particularly when newly diagnosed, such that the illness takes precedence. Adopting this perspective is beneficial for participants as it forces the person to attend to their CF, learn about what it means to have the illness and to come to terms with it. Paterson (2001) explains that some people will adopt this perspective to maintain their identity as a sick person. The wellness in the foreground perspective involves appraisal of the illness as an opportunity for meaningful change, with the attention focused on the self, not the diseased body, as the source of identity. Paterson states that there are numerous ways that participants may adopt this perspective, particularly through learning about the disease, sharing their knowledge of the

disease with others, creating supportive environments and recognising their body's response to the disease. Within this research, participants felt it was important that their CF did not define them or their life and to 'get on with it'. This connected with the wellness in the foreground perspective in which participants wanted to distance themselves from their sickness and focus on other aspects of their life. The model was able to show the complex relationship individuals had with their CF in explaining the on-going shift between wellness to illness in the foreground and vice versa. An individual's perception of a threat to control was the main reason for when individuals would shift from wellness to illness in the foreground. This explains the experiences of Morgan who would struggle when being admitted into hospital as, firstly, her health had significantly declined and, secondly, she would lose her control around when her meals and medication were administered, putting her into the 'sick' role. Other reasons that would shift individuals from wellness to illness in the foreground included disease progression, limited skills to manage the disease, stigma related to the disease and interactions with others that emphasised dependence and hopelessness. Treatment burden was commonly raised by participants and can be explained by the model. As participants maintained a wellness in the foreground perspective, it was easier to become complacent taking medication as the individual had distanced from their CF. Riley did not know the benefits of taking a particular medication and therefore found it was easier not to take it. As Paterson reports, the major paradox of maintaining the wellness in the foreground perspective is that the management of the disease is vital, requiring attention to not have to pay attention to it. Participants spoke of often allowing their illness to become worse than it should before taking action as the shift between the wellness to illness in the foreground

perspective. CF was spoken about as being in the background which fits this perspective.

Another model that may help to understand the findings of this research is a working model of adjustment to chronic illness (Moss-Morris, 2013). Figure 6 shows an example of how one participant's story has been applied to the model, demonstrating how its application can explain participants' behaviours linked to their CF.

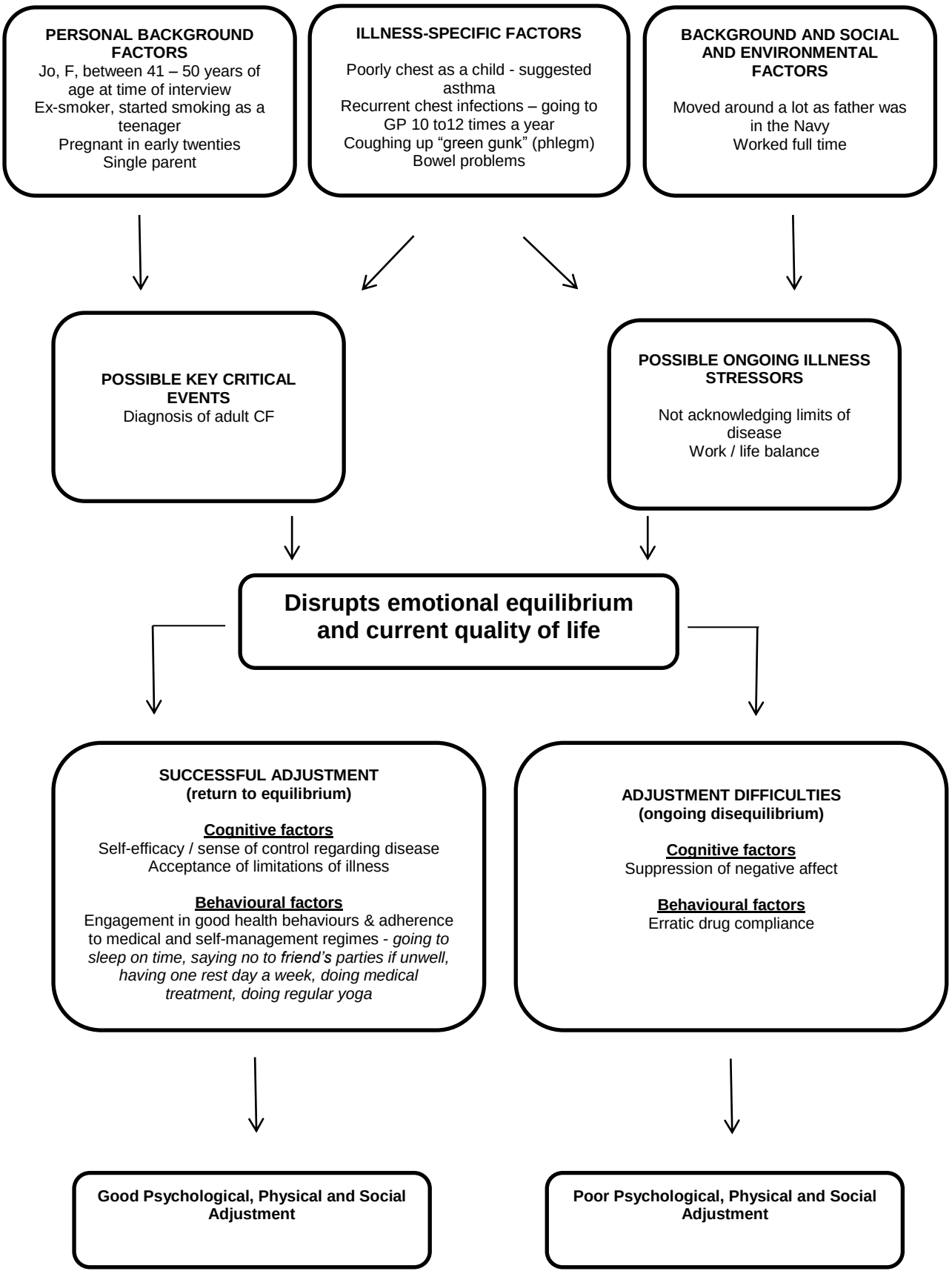

Figure 6: A working model of adjustment to chronic illness (Moss-Morris, 2013)

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5.5 Key discussion points from findings

Across the themes, mismatch was a consistent element throughout the patient journey between their need and healthcare provision. It was evident in the first theme between the participant and their GP or healthcare professional before diagnosis. Participants presented with symptoms relating to their CF, however, these were either dismissed or considered as another condition. At the point of diagnosis, for a few participants, there was a mismatch between the expectation of their hospital appointment and the actual outcome. One participant believed that the purpose of their hospital visit was for further tests, when it had been for them to receive the diagnosis. In another participant's case, there had been a miscommunication regarding the appointment going ahead or not and when she attended the appointment, she was shocked to be given her diagnosis. Further discrepancies were found around the information needs of the participants and what was provided by the CF team or what they found. Some had wanted to find out more information about CF but there was not enough relevant information out there for them, others talked about being overwhelmed with the information that was presented to them. These findings were consistent with those from the chronic illness literature (Due-Christensen *et al.*, 2018; LeBlanc *et al.*, 2016; Marx *et al.*, 2016; Giacomini *et al.*, 2012). Participants also spoke of a mismatch between their identify as having a rare form or milder condition in comparison to someone with 'normal' CF. Paterson (2001) states that an individual's perception of reality is how people with chronic illness interpret and respond to their chronic illness. This is a significant and key point made by Paterson, which explains why there is a mismatch between healthcare professionals' perception of adults who are clinically milder suffers of CF

with patient's perception of adult diagnosis and chronic illness. Individual's relationship to their chronic illness is based on their perception of their reality.

Another element that was consistent across the themes was that of duality. This was expressed by the participants sharing opposite perspectives. For instance, within the theme emotions around diagnosis, individuals shared their ambivalence around their diagnosis. In the theme "It did kind of take over my life", some participants shared that CF impacts on everything whereas others reported that it did not impact on their life. When referring to the future, there were mixed feelings of hopefulness and hopelessness. Lastly, the duality presented itself with participants wanting to find the balance between accommodating for CF in their life and looking after themselves with leading a normal life. This duality element could be explained by the shifting perspectives model of chronic illness (Paterson, 2001). As previously described, the model fits well with the experiences of adults diagnosed with CF. At the time of the interview, some of the participants may have adopted the wellness in the foreground perspective, whilst others adopted an illness in the foreground perspective which may explain the contrast in experiences. The on-going shift between these two perspectives is important for healthcare professionals to be mindful of. For example, an individual may present to a healthcare professional having adopted the wellness in the foreground perspective but might be struggling to manage their CF. Healthcare professionals need to undertake regular follow-up's with individuals, recognise which perspective they may have adopted and subsequently provide the most appropriate support.

5.6 Practical implications and recommendations

As a result of this study's findings, several recommendations have been made below for healthcare professionals working with and supporting people diagnosed with CF in adulthood (see Figure 7). The recommendations may not necessarily apply to all individuals diagnosed in adulthood and do not address all the concerns raised by participants. However, bringing these issues to awareness may have an impact on others' experiences of diagnosis and living with CF in adulthood. The recommendations made are salient throughout the person's journey and should include follow-up as needs may change over time. Figure 8 shows significant time-points in a CF adult patient's journey with suggested interventional points.

Pre-diagnosis

1. Raising greater awareness with non-CF specialist healthcare professionals (e.g. GPs and nurses) and the general public of the possibility for individuals to be diagnosed with CF in adulthood and that CF is no longer considered a children's disease.

Diagnosis

2. Ensuring all healthcare professionals have appropriate training and support when delivering news of an adult CF diagnosis to patients. Breaking bad news can lead to negative consequences for patients, families and healthcare professionals when healthcare professionals lack proper training (Monden, Gentry and Cox, 2016). The following points are recommendations regarding the diagnosis appointment:
    a. Healthcare professionals to set up the expectation and mentally prepare the patient that they may be receiving news of a diagnosis. Some of the ways that this could be achieved include emphasising in the communication with patients that the appointment they have been invited to is important, stating that it is to discuss the outcome of their test results and by advising patients that they may wish to bring someone with them to the appointment.
    b. Healthcare professionals need to allow patients time to process the news of the diagnosis and provide support emotionally before going into the clinically related aspects of its management. Individuals who are feeling anxious at the time of diagnosis may struggle to retain information. It is recommended for information to be written down or for individuals to be sent a follow-up letter.
    c. Individuals may hear comments in different ways. Healthcare professionals are encouraged to check back with patients as to what was heard. Well-meaning comments with the intention to provide support and reassurance can be seen as unhelpful and belittle an individual's experience of their diagnosis. This could have a negative impact on the relationship between the healthcare profession and the patient.

Living with CF

3. Healthcare professionals should provide patients with sufficient information and prepare them prior to an in-patient admission. This could be provided in multiple formats such as a leaflet, website or video link with the opportunity to raise and discuss any questions that they may have. Healthcare professionals working in CF services should be mindful of whether it is the patient's first inpatient visit. It would be beneficial if there was a specialist role for a member of the CF team to check-in with the patient, ask if they had any questions, providing any support or reassurance if necessary, particularly if the admission is over a weekend and access to the team psychologist is unavailable.

4. Healthcare professionals are encouraged to ask patients if they would like more feedback on any results, to provide this information and an opportunity to discuss it with them. Mobile health apps are increasingly popular (Bol, Helberger and Weert, 2018) and could be considered as a way of facilitating this process. A study in China investigating if mobile health apps improve patient experience in Chinese public hospitals concluded that mobile health apps can improve patient experience in relation to accessing health information and making healthcare professional and patient communication more convenient (Lu *et al.*, 2018).

Figure 7: Recommendations for healthcare professionals working with and supporting people diagnosed with CF in adulthood

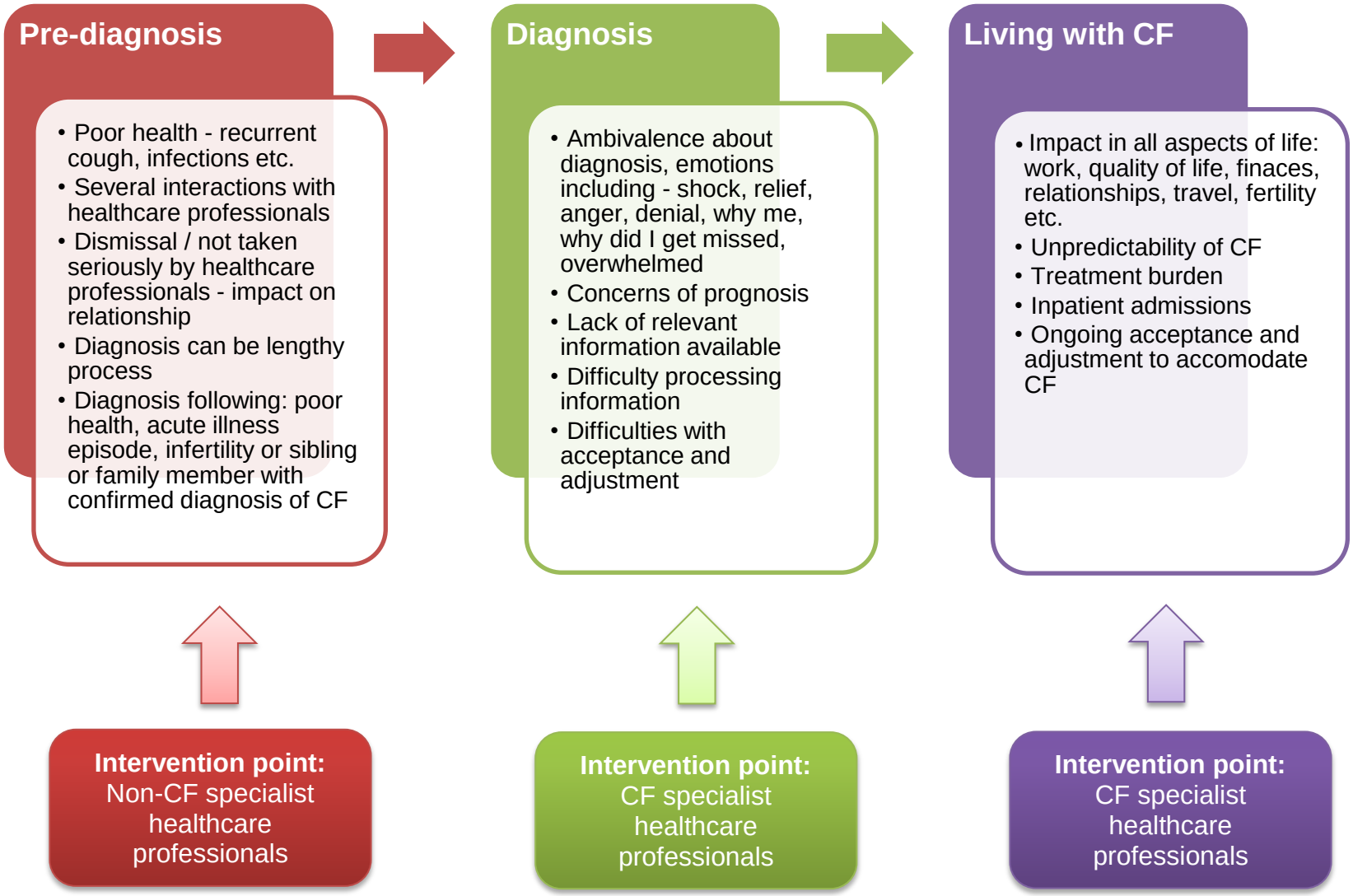

Figure 8: Significant time-points of a CF adult patient's journey with recommended interventional points

5.7 Implications for health psychology practice

As part of the UK CF standards of care, the CF Trust advises for appropriate psychologist staffing levels based on the clinic size (CF Trust, 2011)[7]. For instance, a clinic size of 150 patients should have 1 whole time equivalent psychologist working within the multidisciplinary team. Unlike some long term health conditions, where access to psychological support can be challenging, CF services tend to meet the advised staffing level for psychologists. The CF Trust also recommends that access to psychological support at the time of late diagnosis is essential (CF Trust, 2011).

A key role for the CF psychologist would therefore involve supporting the patient to help them to make sense of their diagnosis. Whilst access to a psychologist is usually available within CF teams, anecdotally, some patients decline this support. A plausible reason is that psychological stigma is associated with a reluctance to seek help (Henderson, Evans-Lacko and Thornicroft, 2013). Therefore, the role of the psychologist would involve working with specialist CF healthcare professionals in three capacities: 1) teaching, 2) reflective practice and 3) supervision. Within the teaching role, the psychologist can inform the CF multidisciplinary team of the reported findings from the research and models related to chronic illness to explain adult experiences of diagnosis and living with CF. The reflective practice role would involve working with CF healthcare professionals, to reflect on their interactions with this patient group to address the reported mismatch experienced and working through the suggested recommendations. The role of providing supervision would

[7] In adult CF centres, the CF Trust recommends to have a 0.5 whole time equivalent (wte) psychologist for 75 patients, 1 wte psychologist for 150 patients and 2 wte psychologists for 250 patients.

involve providing a coaching style support around working with patients who chose not to engage with a psychologist but are struggling with acceptance or adjustment related to their CF. Due to the fast paced environment within the NHS, the ongoing challenge for the CF psychologist would involve working around staff capacity, staff engagement and the time commitment required from healthcare professionals to attend regular training, reflective practice or supervision sessions.

5.8 Strengths and limitations

This research has demonstrated numerous strengths. Whilst most of the current literature has focused on the experience of children, adolescents, or parents of children diagnosed with CF, limited attention has focussed on the experience of those diagnosed and living with CF in adulthood. This study contributes to an area of literature that has been under-researched. The research was descriptive, exploratory and has been able to give voice and insight into the lived experiences of adults diagnosed with CF. Specifically, those living within the UK where previously all the known published research has been within the USA. TA was considered as an appropriate method to answer the research question due to its theoretical flexibility and systematic approach. Although initial recruitment to the study was slow, it quickly gained momentum. Over half of the participants with an adult diagnosis registered with the regional service agreed to be interviewed about their experiences. Thus, highlighting how keen this group of participants was to share their story. It was interesting to note that although the recruitment strategy included advertising the research project via social media and using posters, no participants were recruited via this method. This group of sixteen participants were diverse in terms of experience, a broad range of age groups from early twenties through to late

sixties and a spread of the number of years since diagnosis, which was reflected in the interview data. All the interviews were carried out face-to-face allowing a strong rapport to develop between the researcher and interviewee and the ability for the researcher to notice non-verbal body language.

There were a few limitations to note. The primary limitations of the study were related to the sample, therefore care must be taken for the wider application of the findings. All the participants identified themselves as White British. Whilst CF is a common disease found in the Caucasian population, it is also present in other ethnicities. Individuals who participated in the study were self-selected. People who experience symptoms related to the health condition being investigated are more likely to participate in studies than those who do not have those symptoms (Galea and Tracy, 2007). It would be of interest to speak to those who did not come forward for the study. However, this would be a difficult task and may be received by individuals that they were being coerced into taking part in the study. Within this research, interviews were carried out with some participants who appeared to be reasonably well adjusted with their diagnosis and living with CF had a limited impact on their everyday life. It could be seen that the interviews represented a balance of views. The sample interviewed came from the same geographical area and were recruited from a single service. Future research may benefit from including participants over a wider geographical spread to see if their experiences reflected similar themes found. Further research would also benefit from patient and public involvement (see Appendix C – Reflective chapter – Patient and public involvement for further discussion). Lastly, the interviews relied heavily on participants recalling their experiences of diagnosis, which for a few participants was over thirty years ago.

Since this time, there have been changes in the way services are delivered, how patients would be managed with an adult diagnosis and the available information.

Another limitation to note is that one interview with a participant during a particular timeframe may not be reflective of their overall experience with a chronic illness. Contextual factors unrelated to the illness may colour the responses given by the participant. The data collected is only a snapshot for that given time period. Therefore, it would be beneficial to carry out more than one interview with a participant, whilst being aware of any potential burden on them. This could allow the researcher and participant to reflect on what had been discussed in the first interview and add additional comments or explore any aspects further. It also allows for a greater rapport to develop between the researcher and participant and to discuss aspects that the participant may have been initially reluctant to share.

5.9 Future research directions

From the study's findings, a mismatch between participants' views and their perceptions of healthcare professional's views was identified. As this research only focussed on the views of patients, it would be of interest to explore healthcare professional's attitudes of working with patients diagnosed with CF in adulthood to understand if there is a discrepancy. Anecdotally, healthcare professionals have found it challenging working with this patient group. In particular, going from seeing a very unwell patient diagnosed in childhood to an adult diagnosed patient who is clinically well but psychologically struggling to come to terms with their diagnosis and to live with CF. The themes and items highlighted from participants in this study could be used to develop questions. An online survey tool such as Survey Monkey

could be used as a way to capture data from healthcare professionals. This would allow for data collection over a large geographical area and be a convenient and easily accessible method for healthcare professionals who often are overwhelmed and busy with clinical work. Interviews or focus groups could supplement the questionnaires for further in-depth data. Recruitment could be carried out through existing groups and forums for health professionals, for example through the CF psychosocial group, word of mouth through staff, e-mail and the CF Trust.

As previously stated, within this study, there were no resources to accommodate individuals who did not speak English as part of the research study. Whilst CF is predominantly found within the Caucasian population, it also affects people from other ethnic backgrounds. Further research to explore the experiences of others from different ethnicities would be beneficial.

The present study suggests that living with CF in adulthood impacts relationships with families and partners. Many of the people interviewed were married or in a relationship and had family around them. Further research could consider the experiences of family members of those who have been diagnosed with CF in adulthood to develop understanding and support for this group.

5.10 Conclusion

This research has found that the experiences of people diagnosed and living with CF in adulthood can be broad and diverse. Diagnosis of and living with a chronic condition can be a life-changing experience resulting in difficulties with psychological and social adjustment. However, this may not apply to all individuals and therefore a

‘one-size-fits-all’ approach cannot be used. It was important for participants to have access to relevant information and find a balance between maintaining their life and living with a chronic illness, as Paterson (2001) describes the complex shift between wellness in the foreground and illness in the foreground.

To the researcher’s knowledge, this is the first qualitative study that explored patients’ experiences of an adulthood CF diagnosis with a UK sample. The exception to this is Blunt *et al.*’s (2008) research which is solely available as an abstract published over a decade ago. This study provides valuable insight and knowledge of the lived experience of those diagnosed in adulthood. Its unique contribution is through its focus on the impact of CF on an individual’s day-to-day life and their acceptance and adjustment around the disease. Individuals should be regularly reviewed by their healthcare professionals to identify potential difficulties and be provided with the most appropriate support. Important interventional points are related to the pre-diagnosis phase, diagnosis period and living with CF. Recommendations have been made for greater awareness of CF amongst non-CF specialists and the general public of receiving an adult diagnosis of CF, for healthcare professionals delivering the news of a diagnosis to have the appropriate training and support, ensure they adequately assess individuals needs and provide appropriate and relevant information.

Health psychologists could offer support to both patients and healthcare professionals. This could be through psychological intervention helping the patient make sense of their diagnosis, develop adaptive coping strategies and adjust to living with a chronic condition. Health psychologists can also support healthcare

professionals through providing education on research and models related to chronic illness, reflective practice on the interactions between healthcare professionals and patients and supervision for them to support patients who are struggling with acceptance or adjustment in relation to their CF. Future work could involve investigating healthcare professionals' perspective of supporting adults diagnosed with CF in adulthood to explore the implications for service provision.

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Appendix A – Systematic review

Psychosocial interventions that improve coping for people with cystic fibrosis: A systematic review of randomised and non-randomised trials.

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ABSTRACT

Structured summary

Background: The effectiveness of psychosocial interventions focusing on coping with cystic fibrosis (CF) has not been well established.

Objective: To investigate the effectiveness of psychosocial interventions and whether they provide significant benefits for the psychological and physical wellbeing of individuals on coping with CF in addition to standard care.

Data Sources: Studies were identified from AMED, CINAHL Plus, MEDLINE, PsycARTICLES and PsycINFO. Additional studies were identified through hand searching references and the use of a search engine. Searches were carried out in June 2014.

Study Selection: Any psychosocial intervention (excluding nutritional, physical health based and adherence to treatment interventions) with a comparator of standard care with children, adolescents, and adults with CF were included which aimed to improve coping outcomes (for example quality of life, anxiety, depression, social support, psychosocial adjustment).

Data Extraction: Independent extraction of articles and quality assessment was conducted by two authors.

Data Synthesis: This review included 4 studies, representing data from 366 participants. The studies were diverse in their design and methodology. Two interventions were educational based using a cognitive behavioural approach, one was a written self-disclosure (cognitive based intervention) and one was a hypnosis based intervention. Evidence was found that teaching life skills to children aged eight to twelve is effective in decreasing impact of illness on quality of life.

Conclusions: At present there are insufficient studies on psychosocial interventions on coping with CF. Therefore, no substantive conclusions can be made around the efficacy of psychosocial interventions in coping with CF. Due to the heterogeneity between studies, more high quality research needs to be conducted to assess the efficacy of psychosocial interventions to support coping with CF.

INTRODUCTION

Rationale

Cystic fibrosis (CF), also referred to as mucoviscidosis is an autosomal recessive condition, commonly affecting those of European decent with a low prevalence in Asian and African people (World Health Organisation, 2002). According to the CF Trust (2014), 100,000 people are affected by CF worldwide, including 9,000 people in the United Kingdom (UK). It is a life limiting disease, caused by mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene (Bobadilla et al., 2002). CF mostly affects the lungs and pancreas; but also other organs or parts of the body such as the liver, intestines, reproductive organs, and sinuses, through excessive mucus secretion leading to chronic infections and inflammations (Cystic Fibrosis [CF] Trust, 2014).

With early diagnosis, improved medications and treatment regimes, life expectancy for people with CF has increased. The median predicted survival for someone born in the UK with CF is reported to be greater than 50 years (Dodge *et al.*, 2007). This means that the long term management of the condition has become increasingly important. Currently with no cure, people with CF often have to undergo rigorous treatment regimes, which can be time consuming and complex in order to maintain good health (CF Trust, 2014). Living with CF can have a psychological impact on individuals and families. Therefore, coping is an important area to investigate. It can be assumed that if a person reports good quality of life or psychosocial functioning then they are likely to be using appropriate ways of coping. Interventions that look at increasing coping and lowering stress may find additional benefits in that using active coping strategies to follow treatment regime, the individual is more likely

to stay well (Bartholomew, 1991). Interventions focusing on coping could also be beneficial because there could be a possibility to change maladaptive coping (Abbott, 2003).

The Cochrane review investigating psychological interventions for people with cystic fibrosis and their families state that the updated review will investigate four main target areas for psychological interventions (Glasscoe & Quittner, 2009). One of the areas mentioned is coping. Glasscoe and Quittner (2009) also state "any of these target areas could constitute a separate satellite review that stands alone and explores the area in more depth" (p.79). It was from these recommendations that the review question was derived.

This is a new systematic review with the aim to add to the knowledge of coping with CF. It is reported that in comparison with other conditions, the literature on coping with CF is scarce (Abbott, 2003). Also, the review aims to establish whether there are specific interventions that are effective in helping people to cope with cystic fibrosis.

The outcomes of this review would be relevant to multidisciplinary teams who provide care and work with CF patients worldwide as well as individuals and families with CF.

Objectives

The objectives of this review are:

1.) To investigate the quality of current research of psychosocial interventions in

improving coping in people with cystic fibrosis.

2.) To identify whether in addition to standard care, the effects of the psychosocial interventions provide significant benefits for the psychological and physical wellbeing of individuals with CF.

3.) To compare the effectiveness of the psychosocial interventions in improving coping in people with cystic fibrosis.

METHODS

Protocol and Registration

The review question, participant, intervention, comparison, outcome, study design and inclusion criteria were pre specified and documented in a protocol (appendix 1).

Eligibility Criteria

Study eligibility criteria:

Types of studies: All study designs with a comparison group were included, for example randomised controlled trials (RCTs) and quasi-experimental trials.

Types of participants: Children, adolescents, and adults diagnosed with CF of any age, from any ethnic group were considered. Studies looking at inpatients were excluded from the review whereas studies in an outpatient setting, community setting or at home were included. Patients due for transplant, transplant recipients or CF patients with other co-morbidities or complications as a result of their CF for example CF related diabetes were excluded. This was to reduce the effect of confounding variables on coping.

Types of intervention: Any psychosocial intervention was considered. Exclusions were applied to interventions that looked at nutrition, improving only physical health or adherence to treatment. Interventions without a comparison group were discarded.

Types of outcome measures: Any coping measure related to the individual diagnosed with CF was of interest for this review (for example quality of life, anxiety, depression, social support, psychosocial adjustment). Coping measures related to siblings, parents or caregivers were excluded.

Report eligibility criteria:

Articles only written in English were identified for this review. No restriction was imposed on publication status or publication date.

Information sources

Studies were identified by searching electronic databases, scanning reference lists of articles and journals and contacting specialists within the CF field.

Electronic searches

EBSCO AMED, CINAHL Plus, MEDLINE, PsycARTICLES and PsycINFO were reviewed using a search strategy (appendix 2.) developed by the author. The search terms were developed based on the review question. The search strategy used by Glasscoe and Quittner (2009) was examined to avoid missing key terms. The last search was run on 1 June 2014. The Cochrane Central Register of Controlled Trials was also looked at. Relevant studies were identified using the terms cystic fibrosis and psychology. A brief search using a search engine (Google) was undertaken. Keywords from the search strategy were used in combination for example cystic fibrosis, interventions, coping, and quality of life. In addition, the contents page of The Journal of Cystic Fibrosis was hand searched. References of two similar Cochrane paper were reviewed (Goldbeck *et al.*, 2014; Glasscoe & Quittner, 2009) as well as the references of relevant articles investigating CF and coping.

Searching other sources

Attempts were made to locate unpublished findings through consulting with the managing editor of the Cochrane cystic fibrosis and gene disorders group and the use of list serves.

Search

The following search terms to search all databases were used: cystic fibrosis; mucoviscidos*; psycho*; intervention; therap*; counsel*; transactional; person cent*; client cent*; cope*; coping measure; scale; wellbeing; "well being"; life satisfaction; mood; acceptance; stress; depress*; psychological adjustment.

Limiters were applied in the database to select only journal articles and dissertations, for studies to have been conducted with humans and written in English.

Study selection

The database search identified 836 articles in total (Figure 1.). Two records were obtained through screening the reference list of similar Cochrane reviews and one study was identified through using Google (search engine). All study titles that were considered inappropriate or irrelevant to the research question were discarded. Studies were rejected if they did not fit with the research protocol. This included titles of articles which were medically specific (for example, drug trials), other health conditions which were not CF (for example, cancer, sickle cell disease, thalassemia), health issues related to CF (for example, infertility), studies around transplant, as well as physiotherapy and nutritional based interventions.

Following the initial screening process, the author and secondary reviewer LH reviewed the titles and abstracts of the remaining articles that were to be included in the review using a study eligibility criteria form (appendix 3.). This was carried out independently, in a blinded standardised manner to avoid the possibility of rejection of relevant articles. Although the authors generally agreed on study inclusion or exclusion, any discrepancies were discussed between the two parties and a mutual decision was made. Full text reports for seven studies were retrieved and were assessed for eligibility. Of these, three were excluded for not meeting the inclusion criteria; two articles were conducted in an inpatient setting and the other article did not report the outcomes for CF independently of the other chronic conditions studied in the paper.

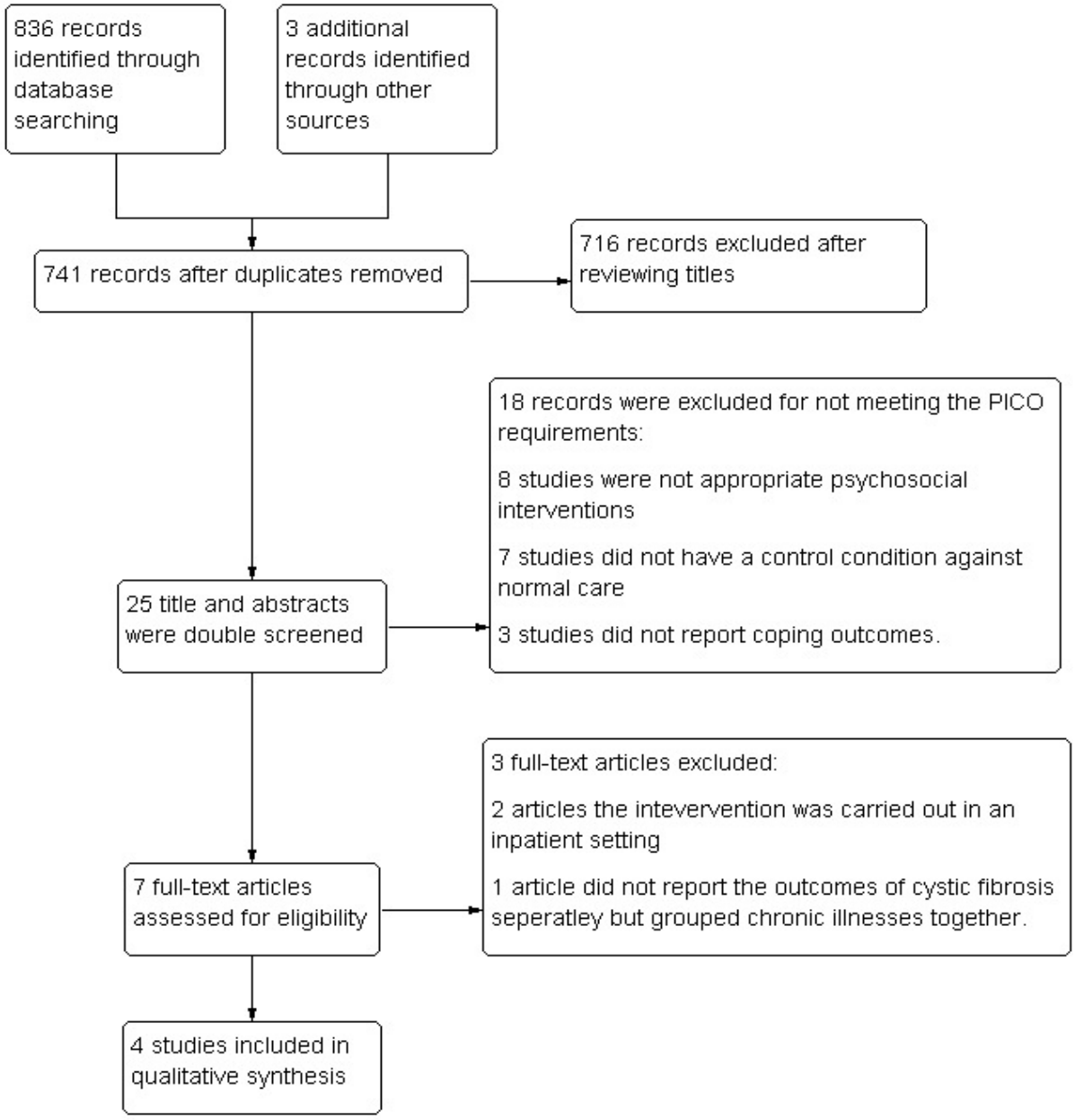

Figure 1. Flow diagram of the study selection process

Data collection process

The author extracted data from the studies using an electronic data collection form. This was based on the Cochrane Collaboration: data collection form template for interventions, version 3 (Cochrane Collaboration, 2014). It was not possible for NS to contact the study authors to request further information. This was because for most of the studies only an address was listed for the contact details and with the age of

the research being published from eight to twenty four years ago, it is likely the study authors had moved on from these addresses. Although advantageous, it was not feasible to request data via the post within the given time of this review.

Data items

Data was extracted from each included research study on the following:

(1) characteristics of the method (including aim of the study, design, start and end date, duration of participation, and if ethical approval was required and informed consent gained)
(2) characteristics of the participants (including the setting, recruitment method, age, gender, ethnicity, relevant sociodemographics, baseline imbalances, withdrawals and missing data)
(3) descriptions of the intervention and control (including the theoretical basis, timing, delivery, duration period, economic information, resource requirements, integrity of delivery)
(4) outcome measures
(5) other information (including author conclusions and references to other relevant studies)

Risk of bias in individual and across studies

Risk of bias at study level was assessed as a part of the data extraction process using the data collection form template for interventions (Cochrane Collaboration, 2014). The author determined the adequacy of the randomisation process and

concealment of allocation prior to assignment, the blinding of participants, data collectors, health care providers, and outcome assessors, and any biases due to incomplete outcome data reported, selective outcome reporting or other potential biases.

Quality assessment was carried out on the four included studies using the Effective Public Health Practice Project (EPHPP) quality assessment tool for quantitative studies (1998). Both the author (NS) and second reviewer (LH) independently carried out the quality assessment of the studies. NS and LH compared their ratings and any discrepancies, mainly due to oversight were resolved through discussion.

Synthesis of results

A narrative synthesis of the studies was undertaken. This was because of the diversity between the participants, interventions, methodology and reported outcomes of the studies to be considered for a meta-analysis.

Additional analysis

It was not feasible to conduct a sensitivity analysis because of the small number of studies currently included, the different types, interventions and outcome measures.

RESULTS

Study selection

Four trials were identified for inclusion in this review. The search of five databases (AMED, CINAHL Plus, MEDLINE, PsycARTICLES and PsychINFO) found 836 citations (Figure 1.). Three additional studies were sourced via alternative methods; two through screening the reference list of a similar Cochrane review (Goldbeck *et al.*, 2014) and the other was identified through using Google. After removing duplicate records, the author screened 741 articles. Of these, 716 studies were discarded because they were considered inappropriate or irrelevant to the research question. After screening the titles, 25 articles remained, of which the abstracts were reviewed by the author and second reviewer using the study eligibility criteria form (appendix 3.). Seven articles were found to meet the eligibility criteria and the full texts were reviewed. Three studies were excluded at this point. Two were identified as being delivered in an inpatient setting and the other study which investigating four chronic conditions did not report the outcomes of the condition separately.

Study characteristics

A summary of the study characteristics are reported in table 1 with further information on the study characteristics provided in appendix 5.

Methods

All of the studies were published in English between 1990 to 2006. Of the included four studies; two were randomised controlled trials (Christian, 2006; Taylor, 2003), one was a controlled clinical trial (Belsky, 1994) and the other a quasi-experimental

trial (Bartholomew, 1997).

Participants

The four studies involved 366 participants who were recruited from CF clinics or centres from the United States of America (USA). Participants were children in one study (Christian, 2006), children and adolescents in one study (Belsky, 1994), adolescents and young adults in one study (Taylor, 2003) and early childhood, middle childhood and adolescents in the final study (Bartholomew, 1997). The ages of participants ranged from under 1 to 29 years across the studies.

Intervention

The studies are diverse in their methodology and design. Two of the interventions were educational and problem solving based using a cognitive behavioural approach for the intervention (Christian, 2006; Bartholomew, 1997). One study assessed the efficacy of a self-management intervention between 12 and 18 months on improving participant's knowledge, self-efficacy, self-management behaviour, health and quality of life (Bartholomew, 1997). The other assessed the effectiveness of the intervention during a home visit and a structured group session to teach problem solving and social skills to improve psychosocial adjustment, functional health and physiologic health (Christian, 2006). One study was a written self-disclosure intervention (Taylor, 2003), using a cognitive approach aimed to improve adherence or health care utilization through asking participants to write about a distressing experience in their life for 20 minutes on three occasions. The last study (Belsky, 1994), children with CF aged 7 to 18 were taught a self-hypnosis technique over a two week period to assess the effects of self-hypnosis on psychological and physiological functioning.

Outcomes

There were a wide range of outcomes reported for the included studies (appendix 5). Of interest for this review were any outcomes related to coping. The outcomes that have been reported include quality of well-being, self-efficacy, self-management (Bartholomew, 1997), locus of control and children's health locus of control, Piers Harris children's self-concept scale, state trait anxiety (Belsky, 1994), psychosocial adjustment, functional health status (Christian, 2006) and subjective health status (Taylor, 2003). All of the studies have looked at pulmonary function; three studies measure this through forced expiratory volume at one second (FEV1) (Christian, 2006; Taylor, 2003; Bartholomew, 1997) and the other (Belsky, 1994) uses peak expiratory flow rate (PEFR). In Bartholomew (1997) the FEV1 was expressed as a percentage of the predicted mean for age, sex, and height using Knudson reference standards.

Table 1: Summary of study characteristics

Study ID	Bartholomew 1997	Belsky 1994	Christian 2006	Taylor 2003
Study design	Quasi experimental (pre/post nonequivalent comparison group)	CCT	RCT	RCT
Number of participants	N=199	N=12	N=116	N=39
Age range	Under 1 to 18 years (mean = 8.6 years)	7 to 18 years	8 to 12 years	15 to 29 years
Study location	2 CF centres, USA	Local CF clinic, USA	1 of 4 CF centres, North Carolina, USA	2 CF centres, southern eastern USA
Intervention	Self-management education program (educational and problem solving)	Self-hypnosis	Life skills educational program (problem solving and social skills)	Written self-disclosure intervention
Psychological basis of intervention	Cognitive behavioural	Other	Cognitive behavioural	Cognitive
Intervention vs Control	n=104 (intervention) n=95 (control)	n=7 (control) intervention not reported	n=58 (intervention) n=58 (control)	n=18 (intervention) n=21 (control)
Duration of intervention	12-18 months	3 appointments in a 2 week period	One home visit and structured group session (2 weeks after home visit)	3 sessions (20 minutes)

Primary outcomes:	Social development and adaptive functions - quality of well-being scale	Locus of control and children's health locus of control	Psychosocial adjustment: 1) Perceived illness experience 2) Children's loneliness 3) Social support (peers & classmates) 4) Self-perception profile (Scholastic competence, social acceptance, athletic competence, physical appearance, behavioural conduct, global self-worth)	Subjective health status: 1) PHQ: a) depression b) anxiety c) somatic complaints d) psychological distress 2) SF-12: a) physical health b) mental health
Primary outcomes:	Self-efficacy	Piers Harris children's self-concept scale	Functional health status: 1) Functional disability	
Primary outcomes:	Self-management questionnaire	State trait anxiety		
Secondary outcomes:	Health Status: 1) NIH score 2) Pulmonary function (FEV1) 3) Weight 4) Height	Lung function (PEFRs)	Physiologic Health Status: 1) Pulmonary function (FEV1) 2) Height 3) Weight	Physiological disease severity: 1) FEV1 2) BMI

Risk of bias within studies

Figures 2 and 3 demonstrate the risk of bias that was judged by the author using the Cochrane risk of bias assessment (2014). Justifications for the authors' risk of bias judgement are described in appendix 4. None of the included studies were judged as

meeting all of the criteria. No studies were excluded from the review as a result of the risk of bias assessment.

Allocation (selection bias)

Sequence generation and allocation concealment was considered low risk for one study (Christian, 2006), unclear risk for one study (Taylor, 2003) and judged as high risk for two studies (Bartholomew, 1997; Belsky, 1994). Bartholomew (1997) and Belsky (1994) did not have a method to randomly generate the allocation of participants. In Bartholomew (1997), the intervention was implemented as part of the medical care and therefore, randomisation was not possible. Belsky (1994) reports that the "two groups were formed on the basis of comparability of age and clinical ratings" (p.284) and so the two groups were not randomised. In both studies, as the groups were not randomised, it is possible that participants could foresee assignment. Both selection biases may impact the outcome of the interventions in a favourable way.

Blinding (performance bias and detection bias)

It can be difficult to completely blind participants, data collectors, health care providers, and outcome assessors. In all four studies, there was an unclear risk of blinding of participants and personnel. There was insufficient evidence from the research study to make judgement in regards to whether there was a bias in the knowledge of being allocated into the interventions. Blinding of outcome assessors was rated as low risk in two studies (Christian, 2006; Taylor, 2003), unclear risk in one study (Belsky, 1994) and a high risk in on study (Bartholomew, 1997). High risk of detection bias is judged in Bartholomew (1997). With the intervention being part of the medical care there is likely to be bias from outcome assessors due to the knowledge of the allocated interventions.

Incomplete outcome data (attrition bias)

Biases due to the amount, nature or handling of incomplete data were assessed as low risk in two studies (Christian, 2006; Taylor, 2003) with an unclear risk in two studies (Bartholomew, 1997; Belsky, 1994).

Selective reporting bias (reporting bias)

Selective outcome reporting bias were judged as low risk for three of the four studies (Christian, 2006; Taylor, 2003; Bartholomew, 1997) and high risk for one study (Belsky, 1994). In Belsky (1994), some outcomes were incompletely reported such as the sample size, means and standard deviations.

Other bias

There was possible type I error bias reported in one study (Bartholomew, 1997). This suggests that some of the effects may have incorrectly been reported.

There are more occurrences of risk of biases in two of the studies (Bartholomew, 1997; Belsky, 1994) in comparison with the other two studies (Christian, 2006; Taylor, 2003). The studies by Christian (2006) and Taylor (2003) use a randomised controlled trial methodology whereas Belsky (1994) is a controlled clinical trial and Bartholomew (1997) uses a quasi-experimental design. The biases reported are likely to reflect the choice of study methodology by the authors.

Overall, across the studies (Figure 3), there is significant selection bias (random sequence generation and allocation concealment). There is an unclear risk of performance bias of all of the studies. Half of the studies present a low risk for detection and attrition bias, a high risk of detection bias in one study and the others

an unclear risk. There is a low risk of bias reported in three quarters of the reported studies with one reported a high risk. Lastly, there is a high risk of other bias reported in one study.

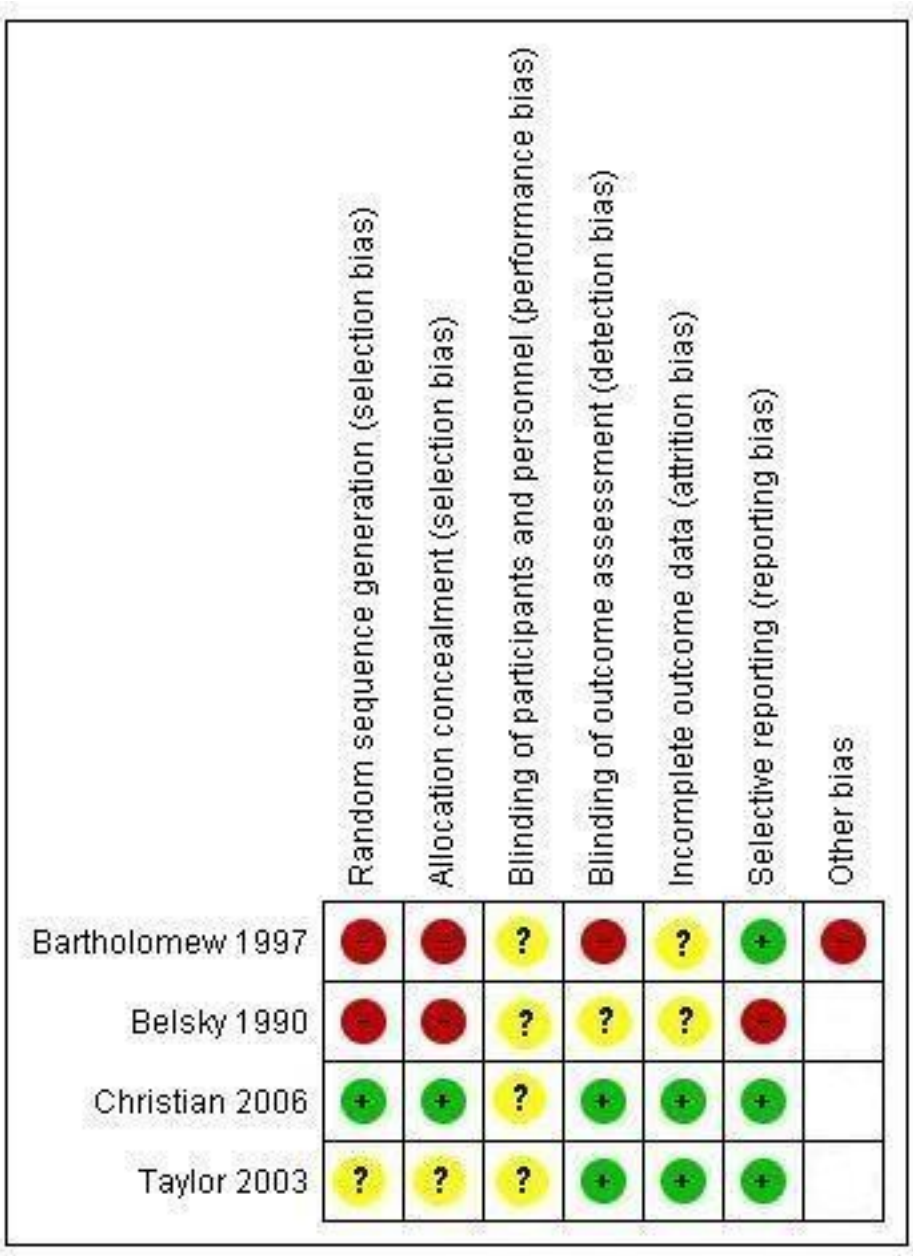

Figure 2: Risk of bias summary: review authors' judgements about each risk of bias item for each included study.

Risk of bias was also assessed using the quality assessment tool (EPHPP, 1998). Table 2 presents a summary of the quality assessment of each included study.

Selection bias

Three studies report a moderate selection bias (Christian, 2006; Taylor, 2003; Bartholomew, 1997) and one study reports a weak selection bias (Belsky, 1994). In both the EPHPP (1998) and the Cochrane risk of bias (2014) assessment tools, a high risk of selection bias is highlighted in Belsky (1994).

Study design

One study reports a moderate bias risk (Bartholomew, 1997) for study design (quasi-experimental) whereas three studies are strong therefore, presenting a low risk of bias (Christian, 2006 [RCT]; Taylor, 2003 [RCT]; Belsky, 1994 [CCT]).

Confounders

Three studies are reported as strong (low risk of bias) for confounding variables (Christian, 2006; Taylor, 2003; Belsky, 1994). One study is reported as weak (high risk of bias) for confounders (Bartholomew, 1997). In the Bartholomew (1997) study a nonequivalent comparison group was used which may present a bias in the reported outcomes.

Blinding

All four studies (Christian, 2006; Taylor, 2003; Bartholomew, 1997; Belsky, 1994) have found to have a moderate risk of bias for blinding from assessors (detection bias) and for study participants.

Data collection methods

All studies (Christian, 2006; Taylor, 2003; Bartholomew, 1997; Belsky, 1994) have a low risk of bias for data collection methods, in that the tools used to detect data are

described as reliable and valid.

Withdrawals and dropouts

Three studies are rated as strong (low risk of bias) for withdrawals and dropouts (Christian, 2006; Bartholomew, 1997; Belsky, 1994). One study was rated as weak (high risk of bias) (Taylor, 2003). In the Taylor (2003) study, less than 60% of the participants (56%) completed the study. From the original 70 participants that were recruited, 14 participants dropped out of the study and 17 patient's data was excluded because of unusual health care utilisation patterns.

Overall the quality assessment of three of the studies is regarded as a moderate risk of bias (Taylor, 2003; Bartholomew, 1997; Belsky, 1994) and one study is rated a strong (a low risk of bias) (Christian, 2006).

Table 2: Quality assessment summary: authors' judgement about study quality using the EPHPP quality assessment tool

Study ID	Bartholomew 1997	Belsky 1994	Christian 2006	Taylor 2003
Selection bias	Moderate	Weak	Moderate	Moderate
Study design	Moderate	Strong	Strong	Strong
Confounders	Weak	Strong	Strong	Strong
Blinding	Moderate	Moderate	Moderate	Moderate
Data collection methods	Strong	Strong	Strong	Strong
Withdrawals and dropouts	Strong	Strong	Strong	Weak
Global rating	**Moderate**	**Moderate**	**Strong**	**Moderate**

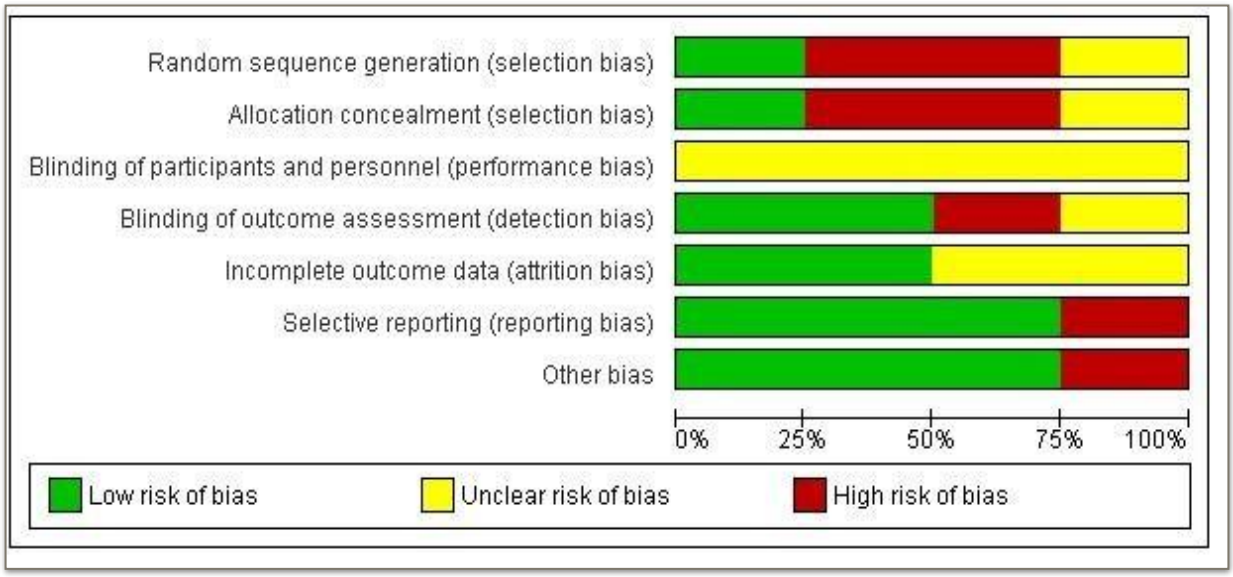

Figure 3: Risk of bias graph: review authors' judgements about each risk of bias item presented as percentages across all included studies

18. Synthesis of results

Preliminary synthesis

Bartholomew (1997) using a quasi-experimental nonequivalent comparison group, assessed the efficacy of a family health education program for CF self-management by parents and patients to increase knowledge, self-efficacy, and self-management behaviours which would improve health and quality of life. Patients were aged from under one to eighteen years (mean age = 8.6 years). The intervention was based on social cognitive theory constructs of self-efficacy, outcome expectations, social reinforcement, and behavioural capability. Families in the intervention group were taught a general strategy of problem solving that involved appraising a situation for specific problems and then planning solutions. It also used a self-paced print format with independent learning strategies for parents and children. The program included instructional modules for respiratory care, nutrition and malabsorption,

communication and coping. The results are reported pre intervention and 1.5 to 2.5 years post enrolment. A significant difference was reported between the intervention and control group for child self-efficacy, child self-management, NIH and pulmonary factor scores. However, no difference was reported for quality of wellbeing and adolescent self-efficacy. Table 3. presents the results of the primary and secondary outcomes for each study.

In a pilot study, using a CCT design, Belsky (1994) aimed to assess the effect of self-hypnosis as a coping strategy on psychological and physiological functioning in children aged seven to eighteen years with CF. In this intervention, children had three appointments in a two week period whereby they were taught a hypnotic technique which lasted 10 minutes. The hypnotic technique instructions were recorded for the children and they were told to listen to the tape once a day. The study reported results at baseline and at a four-month follow-up. A significant difference was reported between the control and intervention groups for the following four outcomes: locus of control, health locus of control, Piers Harris self-concept, trait anxiety. No significant difference was revealed for state anxiety. The study also reports significant findings between the control and intervention groups for lung function (PEFRs).

Christian (2006) carried out an RCT of the effectiveness of an individually tailored intervention session during a home visit and a small group structure intervention session carried out approximately two weeks after the home visit in comparison to normal care. To aim was to assess if the intervention improved psychosocial adjustment, functional health and physiological health in children aged eight to twelve years. Individuals were taught problem solving, social skills, and provided support around the following areas: making sense of their CF diagnosis, explaining

CF related differences, dealing with teasing about CF, and keeping up with peers during physical activity. Outcomes were reported at baseline, three months, six months and at nine months. Significant differences were found between the control group in psychosocial adjustment specifically in the variables perceived impact of illness and loneliness. There was no significant difference between the intervention and control for social support (peers and classmates). No significant differences were found for self-competence, athletic competence and physical appearance. Significant differences were reported for the sub-scale global self-worth, scholastic competence, social competence, and behavioural conduct. There were no significant differences found in functional disability, pulmonary function, weight or height between the intervention and control.

Taylor (2003) carried out a RCT of a three session written self-disclosure intervention compared to standard care with the intention to improve healthcare utilisation, disease severity and subjective health status as well as access the effectiveness, feasibility and acceptability of the intervention applied in chronic disease health care. Participants aged fifteen to twenty nine years were asked to write for a twenty minute period about their most distressing experience of their life focussing on their deepest thoughts and feelings. This was carried out once in a private room within the CF clinic and twice at home. Patients were asked to identify two specific periods over the course of the following five days that they could designate to write alone in a quiet room at home. Individuals were told not to worry about syntax or spelling and were encouraged to connect the topic they wrote about to relationships with others (for example parents, caregivers, friends and so on) and to their past, present or future. The study reported results at baseline and at three months. No significant main effects were found on subjective health status or physiological disease severity.

Table 3. Significant and non-significant findings in primary and secondary outcomes

	Barthlomew (1997)		**Belsky (1994)**		**Christian (2006)**		**Taylor (2003)**	
Intervention	Self-management education program (educational and problem solving)		Self-hypnosis		Life skills educational program (problem solving and social skills)		Written self-disclosure intervention	
Primary outcome / Significant (sig.) or not significant (not sig.)	Social development and adaptive functions - quality of well-being scale	**Not sig.**	Locus of control and children's health locus of control	**Sig.**	Psychosocial adjustment: 1) Perceived illness experience 2) Children's loneliness 3) Social support (peers & classmates) 4) Self-perception profile a) Scholastic competence b) social acceptance c) athletic competence d) physical appearance e) behavioural conduct f) global self-worth)	1) **Sig.** 2) **Sig.** 3) **Not sig.** 4) **Not sig.** a) **Sig.** b) **Sig.** c) **Not sig.** d) **Not sig.** e) **Sig.** f) **Sig.**	Subjective health status: 1) PHQ: a) Depression b) Anxiety c) somatic complaints d) psychological distress 2) SF-12: a) physical health b) mental health	**Not sig.**
Primary outcome / Significant or not significant	Self-efficacy	Child - **Sig** Adolescent **- Not sig**	Piers Harris children's self-concept scale	**Sig.**	Functional health status: 1) Functional disability	**Not sig.**		
Primary outcome / Significant or not significant	Self-management questionnaire	**Sig.**	State trait anxiety	Trait - **Sig.** State **- Not sig.**				

Secondary outcome / Significant or not significant	Health Status: **Sig.** 1) NIH score 2) Pulmonary function (FEV1) 3) Weight 4) Height	Lung function (PEFRs)	**Sig.**	Physiologic Health Status: **Not sig.** 1) Pulmonary function (FEV1) 2) Height 3) Weight	Physiological disease severity: **Not sig.** 1) FEV1 2) BMI		

Exploring relationships within and between studies

The four interventions are carried out in USA and are aimed at different age groups. Three of the interventions are over a short period of time (Christian, 2006; Taylor, 2003; Belsky, 1994) whereas Bartholomew (1997) is between twelve and eighteen months. The studies have been grouped together based on their risk of bias assessment.

All four studies have used theoretical underpinning around their intervention. They recognise that as life expectancy in CF has increased, more needs to be done to support individuals to manage their chronic condition physiologically as well as their psychosocial needs. Taking an educational approach, Bartholomew (1997) aimed to increase knowledge, self-efficacy and self-management and by doing so hypothesised that this would improve the patient's health and quality of life. Similarly, Belsky (1994) hypothesised that self-hypnosis would be associated with an improved sense of well-being which would lead to better physiological control. This suggests that the authors consider some form of self-management to be an important factor to better health and improved quality of life, which would imply that the individual is coping better with their CF. The outcomes for both studies were mostly significant.

The results from Belsky (1994) have identified four significant outcomes (improved locus of control, self-concept, reduction in trait anxiety and improved lung function). This would imply that self- hypnosis has a positive impact on health and therefore coping. However, the results need to be considered with caution, as the sample size

for the total study reported twelve participants, with seven in the control condition. There could be further bias in the intervention around how much practice individuals were doing at home. There is considerable variability reported in practice (which is self-reported) of between twenty to sixty hours for the first and third month combined. In Bartholomew (1997) two outcomes are significant (self-management and health status), self-efficacy is found to be significant in children but not in adolescents. Quality of well-being was reported not significant. This study was found to have high risk of bias in four out of seven areas (random sequence generation, allocation concealment, detection bias, possible type 1 bias). It is difficult therefore, to use these findings to assess why the intervention has been effective as some of the results could be the result of bias.

Taylor (2003) also has a different theoretical unpinning. Written self-disclosure allows people to put their cognitions and emotions into words, therefore enabling them to make sense of their experiences. Through the disclosure process individuals are able to simplify complex experiences and develop a sense of control. This organisational process allows individuals to regulate their emotions. Therefore allowing their physiological and emotional energy to be freed and utilised by the body more effectively. The findings of this study reported no significant effects. One limitation to note in this study is that individuals were asked to complete two written sessions at home which could lead to variability in that individuals may not have spent twenty minutes writing about their distressing experience.

Similarly to Bartholomew (1997), Christian (2006) uses an educational cognitive behavioural approach. The theoretical background for the intervention suggests that the contexts of school and peers relationships impact on how children with CF perceive their chronic illness. If children hide their CF from friends, do not participate

in activities and do not adhere to treatment they have a negative experience which will negatively affect their health and quality of life. Therefore, they are likely to have negative coping strategies. This study was the most robust intervention with the least amount of bias reported. Although Christian (2006) found no significant effect on physiologic health status or functional health status but significant difference for some of the variables and sub variables in psychosocial adjustment (perceived illness experience, children's loneliness scale, scholastic competence, social acceptance, behavioural conduct, and global self-worth) were found. Isolation from others with CF is usually recommended to prevent cross infection. Therefore, it is often difficult for people with CF to socialise with others who have the same chronic illness as them. One part of this study was a group intervention, allowing individuals to meet other children with CF who may have common life experiences to form a CF peer group. The children would get involved in developmentally appropriate activities related to the module content within the group session. The positive outcomes reported related to loneliness and social acceptance could have been improved because of the group element of this intervention.

Additional analysis

No additional analysis was carried out for this review.

DISCUSSION

Summary of evidence

This review has investigated the quality of current research of psychosocial interventions in improving coping with cystic fibrosis, it has tried to identify whether in addition to standard care, the effects of the psychosocial interventions provide significant benefits for the psychological and physical wellbeing of individuals with CF and tried to compare the effectiveness of the psychosocial interventions in improving coping in people with cystic fibrosis. There is a lot of variability across the four studies with the participants, interventions, methodology, outcomes which made comparability challenging. Therefore, conclusions that are drawn from this review are limited. Three studies found some significant outcomes (Christian, 2006; Bartholomew, 1997; Belsky, 1994). However, two studies (Bartholomew, 1997; Belsky, 1994) were found to have several biases and therefore the outcomes may have been influenced.

Only one study (Christian, 2006), had a robust methodology (with a strong quality assessment rating) and with a low risk of bias. This educational intervention found improved coping by teaching children life skills for managing their CF. Without any additional studies that show similar findings, it is difficult to conclude what elements of this intervention are effective and if it is generalisable.

Limitations

Outcome level limitations

A limitation of this review is that the outcomes measured, the populations, the

interventions were very different, making comparison difficult. For instance, for pulmonary function, although three studies were using the outcome FEV1 (Christian, 2006; Taylor, 2003; Bartholomew, 1997), in Bartholomew (1997) the FEV1 was expressed as a percentage of the predicted mean for age, sex, and height using Knudson reference standards) and the last study (Belsky, 1994) used peak expiratory flow rate (PEFR).

Study level limitations

Four studies, involving a total of 336 participants were included in this review. The major limitation is having large enough samples. In this review two of the studies had small sample sizes (Taylor, 2003; Belsky, 1994). Only one study carried out a power calculation to ensure they had enough participants to detect statistical significance (Christian, 2006).

The assessment of the risk of bias identified that the quality of the studies varied; two of the articles identified higher risks of bias in comparison to the other two studies, which either had mostly a low risk of bias (Christian, 2006) or had a mix of an unclear risk and low risk of bias (Taylor, 2003). There was a high risk for selection bias (Bartholomew, 1997; Belsky, 1994), detection bias (Bartholomew, 1997), reporting bias (Belsky, 1994) and type one bias (Bartholomew, 1997).

Review limitations

One of the limitations for this review is the restriction placed on the inclusion of interventions reported in English only. This was because there were no resources or facilities to translate the articles into English. However, this may mean that other studies that were not reported in English may have been eligible for the review but were not included.

22. Conclusions

Four psychosocial interventional studies around coping with CF were identified. In this review only one study (Christian, 2006) was identified to have a robust methodology (with a strong quality assessment rating) and with a low risk of bias. This educational intervention found improved coping by teaching children life skills for managing their CF.

The author has concluded that due to the lack of studies in the area, no substantive conclusions can be made around the efficacy of psychosocial interventions in coping with CF. This highlights the urgent need for more high quality research to be conducted to assess the efficacy of psychosocial interventions to support coping with CF.

23. Funding

There was no funding for this review.

This review was completed as part of the Professional Doctorate in Health Psychology programme at the University of the West of England (UWE), in accordance with the British Psychological Society (BPS) requirements.

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Appendix 1. Review protocol

Review Objectives:

The objectives of this review are:

1. To investigate the quality of current research of psychosocial interventions in improving coping in people with cystic fibrosis.

2. To identify whether in addition to standard care, the effects of the psychosocial interventions provide significant benefits for the psychological and physical wellbeing of individuals with CF.

3. To compare the effectiveness of the psychosocial interventions in improving coping in people with cystic fibrosis.

P - individuals diagnosed with cystic fibrosis (CF) (Including children, adolescents, and adults) of any age and any ethnic background Including studies conducted in outpatient, community setting or at home Excluding studies conducted in an inpatient setting, patients due for transplant/transplant recipients or CF patients with other co-morbidities (such as CF related diabetes)
I - any psychosocial intervention Excluding interventions looking at nutrition, improving only physical health and adherence to treatment
C - against normal/usual care
O - any coping measures related to the individual diagnosed with CF (for example quality of life (QoL), anxiety, depression, social support, psychosocial adjustment) Excluding coping measures related to siblings, parents or caregivers
S - any study design with a comparator (for example RCTs, Quasi-experimental)

Appendix 2. Search strategy: AMED, CINAHL Plus, MEDLINE, PsycARTICLES, PsychINFO (EBSCOhost)

Search ID #	Search Terms
S1	cystic fibrosis
S2	mucoviscidos*
S3	S1 or S2
S4	psycho*
S5	intervention
S6	therap*
S7	counsel*
S8	transactional
S9	person cent*
S10	client cent*
S11	S5 OR S6 OR S7 OR S8 OR S9 or S10
S12	cope*
S13	coping measure
S14	scale
S15	wellbeing
S16	"well being"
S17	life satisfaction
S18	mood
S19	acceptance
S20	stress
S21	depress*
S22	psychological adjustment
S23	S12 OR S13 OR S14 OR S15 OR S16 OR S17 OR S18 OR S19 OR S20 OR S21 OR S22
S24	S3 AND S11 AND S23
S25	S3 AND S11 AND S23 (limiters: English Language; Human; Language: English; English Language; Human; English)

Appendix 3. Study eligibility criteria form

The abstract is to be retained if the study includes:

Study eligibility criteria	NS		LH	
	Yes	No	Yes	No
P - individuals diagnosed with cystic fibrosis (CF) (Including children, adolescents, and adults) of any age and any ethnic background Including studies: * conducted in an outpatient, community setting or at home Excluding studies: * conducted in an inpatient setting * patients due for transplant/transplant recipients * CF patients with other co-morbidities/condition complications (such as CF related diabetes)				
I - any psychosocial intervention Excluding interventions: looking only at nutrition improving only physical health adherence to treatment				
C - against normal/usual care				
O - any coping measures related to the individual diagnosed with CF (for example quality of life (QoL), anxiety, depression, social support, psychosocial adjustment) Excluding * coping measures related to siblings, parents or caregivers				
S - any study design with a comparator (for example RCTs, Quasi-experimental)				

Appendix 4. Risk of bias reasons of judgement

Bartholomew (1997)		
Bias	**Author's judgement**	**Support for justification**
Random sequence generation *(selection bias)*	**High risk**	The authors reported that "The intervention was to be implemented as a part of medical care. Therefore, it was not possible to randomize subject within sites." (Bartholomew, 1997, p.654)
Allocation concealment *(selection bias)*	**High risk**	Because the intervention was part of the medical care, participants or investigators enrolling participants could possibly foresee assignments.
Blinding of participants and personnel *(performance bias)*	**Unclear risk**	There was insufficient information reported to permit judgement on performance bias.
Blinding of outcome assessment *(detection bias)*	**High risk**	Because the intervention was part of the medical care, there is likely to be bias due to knowledge of the allocated interventions by outcome assessors
Incomplete outcome data *(attrition bias)*	**Unclear risk**	The number of participants did not seem to add up for the groups.
Selective outcome reporting? *(reporting bias)*	**Low risk**	The outcomes stated in the report are reported.
Other bias	**High risk**	The authors state "Of some concern in the study is the possibility of Type I error due to lack of control for multiple comparisons. The study featured three models of effect and outcome measures: parent, adolescent, and child. Each of these models included approximately 10 comparisons and had 4-5 statistically significant results. There is some likelihood, therefore of at least one spurious finding in each model, but little likelihood of four or more Type I errors in each model." (Bartholomew, 1997, p.663)

Belsky (1994)		
Bias	**Author's judgement**	**Support for justification**
Random sequence generation *(selection bias)*	**High risk**	The two groups were not randomised. The author stated that "Two groups were formed on the basis of comparability of age and clinical ratings." (Belsky, 1994, p.284)
Allocation concealment *(selection bias)*	**High risk**	Since the groups were not randomised, it is likely that investigators and/or participants could foresee assignments.
Blinding of participants and personnel *(performance bias)*	**Unclear risk**	Insufficient information to permit judgement.
Blinding of outcome assessment *(detection bias)*	**Unclear risk**	No information was provided on the blinding of the outcome assessors.
Incomplete outcome data *(attrition bias)*	**Unclear risk**	No information was reported by the author of dropouts.
Selective outcome reporting? *(reporting bias)*	**High risk**	Some outcomes are reported incomplete (sample size, means and standard deviations)

Christian (2006)		
Bias	**Author's judgement**	**Support for justification**
Random sequence generation *(selection bias)*	**Low risk**	It was described by the authors that "Children were assigned randomly to intervention and control groups using a computer-generated randomization plan." (Christian, 2006, p.302)
Allocation concealment *(selection bias)*	**Low risk**	Participants and investigators enrolling participants could not foresee assignment because of the computer generated randomisation.
Blinding of participants and personnel *(performance bias)*	**Unclear risk**	Insufficient evidence by the author to permit judgement.
Blinding of outcome assessment *(detection bias)*	**Low risk**	It was reported that "group assignments were unknown to the CF clinics and research assessment team." (Christian, 2006, p.302).

Christian (2006)		
Bias	**Author's judgement**	**Support for justification**
Incomplete outcome data *(attrition bias)*	**Low risk**	The authors state "there was no attrition in the sample" (Christian, 2006, p 305). They also report that "all children completed the baseline and three follow- up assessments" (Christian, 2006, p.301).
Selective outcome reporting? *(reporting bias)*	**Low risk**	All outcomes stated in the report are reported.

Taylor (2003)		
Bias	**Author's judgement**	**Support for justification**
Random sequence generation *(selection bias)*	**Unclear risk**	Insufficient information is provided around the sequence generation.
Allocation concealment *(selection bias)*	**Unclear risk**	Insufficient information is provided around the allocation concealment.
Blinding of participants and personnel *(performance bias)*	**Unclear risk**	Insufficient information is provided on the blinding of the allocated interventions by participants and personnel.
Blinding of outcome assessment *(detection bias)*	**Low risk**	The authors state "The physician and the clinic staff were not informed as to whether individual patients were in the WSD or the SMC condition." (Taylor, p.11).
Incomplete outcome data *(attrition bias)*	**Low risk**	The data was reported for all 39 participants.
Selective outcome reporting? *(reporting bias)*	**Low risk**	All outcomes stated in the research have been reported.

Appendix 5. Study characteristics

Bartholomew (1997)	
Method	Quasi-experimental pretest-postest nonequivalent comparison group design. To test the efficacy of the Cystic Fibrosis Family Education Program, a cystic fibrosis self-management program on improving participant's knowledge, self-efficacy, self-management behaviour, health and quality of life.
Participant	Sample agreeing to participate N=199 patient (aged from under 1 to 18 years; mean = 8.6 years) and primary caregiver dyads from two metropolitan CF centres (USA). Patients and caregivers were recruited by their healthcare providers. 92.5% of caregivers were female, middle classed (calculated by the Hollingshead Index) and 85% of caregivers were married. Intervention group n=2 died (during the 2.5 years of study) n=7 from each site declined to complete the post test Intervention centre n=104; usual care comparison centre n=95
Intervention	Educational and problem solving intervention (part of medical care). The intervention was based on the social cognitive theory constructs of self-efficacy, outcome expectations, social reinforcement and behavioural capability (Bandura, 1988; Parcel, 1981). Families in the intervention group were taught a general strategy of problem solving that involved appraising a situation for specific problems and then planning solutions. Social cognitive theory intervention methods of goals setting, reinforcement modelling, skill training and self-monitoring were translated into specific strategies and organised into learning activities for parents and for each of three age groups: early childhood, middle childhood, and adolescence. Uses a self-paced print format with independent learning strategies for parents and children. The program includes instructional modules for respiratory care, nutrition and malabsorption, communication and coping. Intervention time of 12-18 months per patient and post intervention measurement approximately 1.5 to 2.5 years post enrolment

Bartholomew (1997)	
Outcomes	**Measures of cognitive variables:** **Knowledge test:** multiple choice questionnaire, test of knowledge of CF self-management, (53 items for caregivers; 51 items for adolescents; 45 items for children) **Self-efficacy scales:** measures confidence in managing various aspects of medical treatment, symptom and behaviour monitoring, communication and coping (29 items for caregiver; 20 items for adolescents; 11 items for children - read to the 7-8 year old children by the examiner) **Outcome expectations scales**: measures expectations for positive outcomes as a result of carrying out self-management activities. (8 items caregivers and adolescents) **Measures of behavioural variables:** **Self-management questionnaire for Cystic Fibrosis**: 51 item self-report questionnaire to assess monitoring and treatment **Problem solving/Coping scale**: assesses the caregiver's ability to appraise and solve problems in moderately stressful situations with five scenarios, questions and probes about CF-related and non-CF related conflicts. **Means-end problem solving**: measures problem solving in adolescents. Stories were scored for three elements: steps toward a goal, obstacles to goal attainment, and recognition of time necessary to reach a goal. **Measures of health and quality of life variables:** **Health status:** (1) NIH score: used to evaluate patients physical health status (pulmonary function tests, chest x-ray, pulmonary symptoms and complications, and physical examination, weight, activity, and attitude). (2) Pulmonary function: measured by FEV1 (3) Brasfield (Birmingham) roentgenogram (x-ray score): used to quantify the severity of chest x-ray changes associated with CF (4) Weight and (5) height measures: weight and height of each patient was expressed as a standardised deviation from that predicted for the child's gender and age. **Vineland adaptive behaviour scales**: measured adaptive behaviour in three domains: communication, daily living skills and socialisation via structured interview with the caregiver. **Child behaviour checklist**: parent completed checklist of behaviour problems for children 4-16 years. **Quality of well-being scale:** assesses physical functioning combining items from mobility, physical activity, and social activity subscales. **Impact on family scale**: 21 item scale measuring the effect of the child's medical condition on the family system. **Parenting stress index**: 120 item Likert scale designed to identify parent- child systems under stress. The 54 item Parent Domain total scores was used.

Belsky (1994)	
Method	Control clinical trial (CCT) The study used a pre and posttest design, repeated measures and control group. A pilot study that assessed the effects of self-hypnosis on psychological and physiological functioning of children ages 7 to 18 with Cystic Fibrosis.
Participant	12 children with CF aged 7-18 years attending a local CF clinic (USA). Recruited by a letter of invitation to participate in the study. Population of interest: N=30. 16 responded, 4 expressed interest but did not volunteer (2 planned to move and 2 because of time constraints) Total sample: n=12. Control group: 5 girls and 2 boys. Control and experimental groups were matched for age and clinical severity.
Intervention	Parents and children in the experimental group had 3 appointments in a 2 week period during which time the children were taught a hypnotic technique. The hypnotic technique is fully described in the article.
Outcomes	**CF illness ratings:** completed by the physician at baseline, the Shwachman- Kulzcycki system standard form is based on 100 maximum points measuring four categories: general activity, physical examination, nutrition, and radiographic findings. **Other illness variables:** included each patient's average PEFR, height, weight, and number of hospital admissions during 2 years prior to the study. **Parents' measures:** (1) 'Impact on Family Scale' (Stein & Reissman, 1980) measures family functioning-the impact of chronic illness on family life (2) 'Child Behavior Checklist' (Achenbach & Edelbrock, 1983) parents' perception of behavioural problems and competencies; rating of the parents' assessment of the child's illness (The Parents' Questionnaire) (3) Parents also provided a record of schooldays missed by their child for 3 years before the pretest. **Children's measures:** (1) Locus of Control (LOC) was assessed with the Nowicki Strickland (1973) LOC for Children and with the Children's Health Locus of Control (HLOC) (Parcel & Mayer, 1978), a 20-item area specific measure of expectancies regarding locus of control and prediction of health related behaviour. (2) Self-concept was assessed with the Piers Harris Children's Self-Concept Scale (1964). (3) Anxiety was assessed with the State Anxiety Inventory for Children STAIC (Spielberger 1973).

Christian (2006)	
Method	Randomised controlled trial (RCT) 2-group, experimental, repeated-measures design (baseline, 3, 6, and 9 months intervals) with randomly assigned participants to intervention and usual care group. To test the effectiveness of an intervention to improve psychosocial adjustment, functional health, and physiologic health in children (8-12 years of age) with CF by teaching them life skills for managing their chronic illness in their everyday lives.
Participant	Number of randomised participants: N=116 children with CF aged 8 -12 years receiving healthcare from one of four university based CF centres in North Carolina (USA). Potential participants were recruited from children with CF Population of interest: N=128 parents and children with CF. 12 declined because of lack of time Intervention group: N=58; Control group 'usual care': N=58 59 boys and 57 girls Ethnicity: n=102 White; n=8 Native American; African American n=3; Hispanic n=2 and Asian n=1
Intervention	Delivered in two sequenced phases; children in the problem-solving group received individual, tailored intervention during one home visit and a small structured group session (conducted approximately 2 weeks after individual home visit). Educational problem-solving and social skills intervention (N=58) versus usual care (N= 58). Intervention was designed to support children (8 - 12 years) with the following specific problems:1. making sense of their CF diagnosis and constructing their own personal history of CF; 2. explaining CF-related differences; 3. dealing with teasing about CF; 4. keeping up with peers during physical activity. The intervention contains 4 modules. In every module there was a focus on one of the 4 mentioned areas of problems (1-4). The intervention team received detailed information about the intervention protocol and was trained and supervised conducting the intervention

Outcomes	5 questionnaires which are developed for children were read aloud by research assistant to the children. The following questionnaires were used to assess different constructs: (1) **Psychosocial adjustment** assessed by (Perceived Illness Experience Scale, Eiser *et al.*, 1995, 35 items) (2) **Children's Loneliness Scale** (Asher *et al*., 19,16 items) (3) **Social Support Scale for Children** (Harter, 1986,16 items) (4) **Self-Perception Profile for Children** (Harter, 1985, 30 items) (5) **Functional Health Status** assessed by (Functional Disability Inventory Walker & Greene, 1991, 15 items) (6) **Physiological Health Status**: Pulmonary function (FEV1), Height and weight. - BMI.

Taylor (2003)	
Method	Randomised controlled trial (RCT) Compared written self-disclosure intervention to standard care (wait-list control condition) To evaluate the effectiveness, feasibility, and acceptability of the written self- disclosure interventions when applied in paediatric chronic disease health care.
Participant	N=39 adolescents and young adults (ages ranging from 15 to 29 years) from two CF centres located at two children's hospitals in the southern eastern part of the USA Patients were recruited during routine consecutive patient visits from two CF paediatric clinics from February 2000 to May 2000. Population of interest: N=81 (met eligibility) 70 patients were recruited (11 refused participation citing time limitations) Dropouts: n=14 (receiving lung transplant n=3; failing to complete all three writing sessions n=5; failing to return to clinic for follow-up assessment n=6) Excluded from data analysis because of unusual health care utilization patterns: n =17 Ethnicity: Caucasian n=35; African American n=4 Initial eligibility criteria included: (1) a diagnosis of CF; (2) age at least 15 years; (3) enrolled in the CF clinic for at least 9 months; (4) physical and mental ability to complete the research protocol as judged by the project staff and CF physician or nurse; (5) willingness, to write for 20 min on 3 separate occasions, over a 5-day period and completion of self-report, measures on two occasions over the next 3 months; and (6) access to a telephone on the days that the participants were to write at home. Intervention group (WSD) n=18, control group (SMC) n=21

Intervention	Written self-disclosure intervention 3 sessions written self-disclosure intervention, adapted from Pennebaker's laboratory- based protocol (first one in a private room within the CF clinic, two home-writing sessions prompted by a telephone call each 20 minutes) Instructions were in accord with those of Pennebaker (1989, 1993, 1997) and required that participants write about their "deepest thoughts and feelings about the most distressing experience of their entire life for a period of 20 minutes" (Pennebaker 1997)
Outcomes	**Health care utilization** assessed by: number of (1) outpatient visits and (2) inpatient hospitalizations days **Physiological disease severity**: (1) FEV1 and (2) BMI. **Subjective health status**: (1) 'The Patient Health Questionnaire' (PHQ; Spitzer *et al.*, 1994; Spitzer, Kronke, Williams *et al*. 1999): providing information on perceived symptoms of (a) depression; (b) anxiety; (c) somatic complaints and (d) psychological distress. (2) 'The Stressful Life Events Scale' (10 items, including psychosocial complaints common among health care seeking populations (e.g., difficulties with family support, problems with significant others, and financial concerns); (3) SF-12 (Ware, Snow, Kosinski, & Gandek, 1993) self ratings of the severity and frequency of 12 physical and mental health problems, as well as of their impact on the patients' overall perceived health status **Feasibility and acceptability**: modified version of the 'Visit Specific Satisfaction Questionnaire' (VSQ; Ware & Hays,1988)

Appendix B – Research diary extracts

04/12/2013 – UWE research day

Points to consider:
Theory suggestion – self regulatory theory
Quantitative work – use illness perception questionnaire, use questionnaire to inform qualitative interviews
Could send out questionnaire via CF Trust

Action points:
Look up self-regulatory theory
Look up illness perception questionnaire
Find out who the PPI person is for the research site
E-mail Julian about supervisory team preferences
Consider stakeholders in research

Reflection:
I found the research day to be really useful. In preparation for the day, it made me begin to consider my research idea. It was good to discuss these with the cohort and get support from staff in building on my ideas.

Learning point:
To continue reading and talking about my research with others to help shape my research.

11/12/2013 – CF psychosocial adult regional services meeting

Points to consider:
D.O.B. against date of diagnosis on port CF
UKPPCF group – NR interested in late life
Genetically diagnosed vs clinical diagnosis?
Consider who was told about the diagnosis e.g. was the diagnosis held from the patient and they found out later in life? (therefore late diagnosis?)
Jiscmail information stored for late life diagnosis (files area)
Consider age for late life diagnosis – 10+ adolescents? / 20+ adults? / 40+ later adults? – Eysenck's development of age/life stages
Adjustment from diagnosis – how many years?
Possible interest in research from other centres

Action points:
Look up standards of care for late life diagnosis
X will contact and find out numbers of late life patients from everyone from different centres
Consider diagnosis – clinical vs genetic
Consider who was told about diagnosis – clear inclusion/exclusion criteria
Look at files on Jiscmail
Consider defining age groups
Consider years since diagnosis/requesting that information

Reflection:
Really useful meeting. Adult CF diagnosis is a 'hot' topic and a very relevant area. Limited practical research knowledge. Research in the area is much needed.

15/01/2014 – Research supervision with DH

I was really pleased with my supervisory team because of the research experience held by my supervisors. For me it was important to have a team which was strong academically and had experience in conducting and publishing research. I was unsure of what to expect for the first meeting, but I thought it went well. We covered quite a few points and ideas. My previous experiences of research/supervisory relationships have not been as good. I think this was partly due to the supervisory relationships being more a teacher-student relationship and so the research process has been more directed rather than guided. I felt that I had an active role in leading the meeting which made me feel respected as a professional and more of an equal, which made me feel empowered and in control of my research.

05/11/2014 – Research supervision with DH

Review of RD1 form.

Decisions made following discussion with supervisor:
Time since diagnosis (1 year +) ethically appropriate and patients have developed some sense of what it is like to life with condition.
Age of participant at diagnosis (18+ years) – a late diagnosis of CF could be any age after the first few years. Interested in the experiences of adults – what is it like to receive a diagnosis for a childhood disease?

06/04/2015 – Research reflection

Have struggled getting into doing research tasks and keep avoiding them. Meetings with supervisors have helped to get me to do work by next meeting. Doing work hasn't been difficult – issue of having to include everything in ethics form and making it perfect stops me progressing.

09/04/2015 – Meeting with research and development research officer - JP

Discussion of REC and R&D approval process, GCP training and data retention.

05/05/2015 & 06/05/2015 - NVivo course notes and reflection

Points to note:
Be clear where you are accessing information – i.e. working on data set at home, icloud – security?
How am I answering the research question? – what is my research question?

What demographic information might I need to know?
Consider name of interview files e.g. INV-001-ENGM-16
What are your coding units? How do you code your data? - Sentences, paragraphs

Thinking:
The course was really helpful
Can see the potential use of NVivo for managing data and more

Feeling:
Pleased that I was told about the course
Lucky that I managed to get on the course as I was 7th in the waiting list
Overwhelmed → so much to think about

Take away:
NVivo can really help to manage the data

10/05/2015 – Research reflection

Feeling angry and frustrated. Taking longer to get everything together for ethics than planned. Things keep getting added in → I'm being pedantic over little things. Useful to have a tight/solid protocol, thinking about eventualities.

18/06/2015 – Research reflection

Feeling good/positive. Finally managed to complete protocol. Getting work done/having the time to do work. Disappointed that I should have done IRAS form sooner. Feeling worried - will I get ethics back in time for participants to show expression of interest.

23/06/2015 – Research reflection

Tried to contact patient advisor for CF – no response. Looked up PPI at NHS site unable to use as research doesn't come under those services. Redirected to research design service. Research design service unable to help as research is not funded. Feeling pleased with work can see progress. All seems to be coming together.

04/08/2015 – Virtual REC review meeting

Prior to review felt very nervous, not knowing what questions to expect.

What went well:
Could answer the questions
Was not as bad as I thought

Not so well:
Did not understand the first question asked by reviewer. Asked for clarification but still did not understand, panicked and rambled. In the future, should ask for clarification again and not be afraid of not understanding.

14/08/2015 – Research reflection

Found out that application had an unfavourable opinion – unsure what this means. Disappointed and frustrated by this – not knowing what's happening and why as had been very thorough. Annoyed that I was in a rush to have REC approval and went for a sooner virtual appointment, perhaps if I waited for a face-to-face appointment this wouldn't have happened.

18/08/2015 – Research reflection

Received a letter from the committee giving an unfavourable opinion. Felt very angry with the outcome as it was clear the committee did not understand my study. I thought and had expected my application to get a favourable review. I spent a lot of time on each form/protocol to be as clear as possible – putting in as much information as possible. Having had conversations with R&D and supervisors, I thought the process would be straight forward. The committee would ask a couple of questions to show they have read the forms but I would get a favourable opinion. I am annoyed that I have put in a lot of hard work for what feels like nothing. I will be further delayed than planned. R&D read my application and protocol and understood it fine. Some of the questions the committee asked were not clear and therefore my answers were a bit waffley and woolly. Whilst putting my comments together in response to the committee, it is hard to be objective and separate my personal thought and feelings without responding to the committee and being sarcastic with my responses.

21/08/2015 - Meeting with research and development research officer - JP

Having met with R&D, I feel a lot calmer and hopeful about the process.

22/08/2015 – Research reflection

Committee comments made me rethink my recruitment strategy. What would I do if patients did not come forward?

Decided to incorporate a follow-up telephone call. Had to consider if it was in line with GCP are people consenting to receive a telephone call. Would a telephone call and saying no to a person be harder than ignoring a letter? It would be hard for me to know if they have ignored the letter because of lack of interest or because they are interested and have not got round to calling. Have included a statement in invite letter to say they may get a telephone call and that they can opt out of this.

By splitting the study, I don't need to address a lot of the comments as they were regarding the healthcare professional part of the study

30/09/2015 - REC review meeting

Was not feeling well on the day of the meeting. Asked for supervisor to attend the meeting with me as did not want a repeat of the previous review meeting. Panicked at the time when a question was asked as I did not know how to handle it and

referred to supervisor. In retrospect, it would have been better if I attempted to answer the question myself but having already been rejected it was easier for supervisor to answer the question. Felt overwhelmed by the panel and number of people.

24/11/2015 – Progression viva 1

I felt a little nervous before attending the progression viva and had anticipated it to be a grilling. I was pleasantly surprised that the time went by really quickly. I enjoyed having the opportunity to talk about my research. I found the feedback from the members of staff to be really helpful.

21/01/2016 – Reflections of interview 1

Thoughts:
Will I get to the participants house ok

Feelings:
Excited to be finally doing an interview
Uncertain – never done an interview in a participant's home not sure what to expect
Confident – experience of working with patients. Have to make sure does not turn into a therapy session

Post interview thoughts:
Maybe I have too many questions on the interview schedule
I've got a lot of transcribing to do
Interview content is all relevant

Post interview feelings:
Quite tired

First time I have done an interview at someone's house. Participant and their partner were very welcoming and home environment was very relaxing. Participant was very articulate and clearly passionate about wanting to see improvements. I was a similar age to the participant's children, I wonder if the participant responded to me in that way. Participant quite openly spoke to me about her experiences.

28/01/2016 – Reflections of interview 2

Wonder how patient could relate to me being a female from an Asian background in late twenties under half the age of the participant. Participant wanted the consent process to go very quickly, seemed to come across as annoyed. Felt exhausted after the interview. Concerned about sound quality of recording due to room used. Different experience from first interview as to second interview. Unsure about my questioning style? Had an opportunity to ask more questions in comparison to the first interview. First interview seemed to be more free flowing.

08/04/2016 – Research reflections and reflections of interviews 3 to 10

Wanted to get people booked in when they expressed interest in the study but this has meant that I have not had much time to reflect in-between interviews. It has been a really busy month with the service evaluation project, work and preparing for Uganda trip. Worries of am I collecting good data. Thoughts – what am I going to do with all this data? Noticing similar points coming up in interviews.

Interview 3:
Harder to initially build rapport in comparison to the first interview – even though it was a home visit. The experiences of the participant were really sad.

Interview 4:
Participant was annoyed that their CF had not been picked up sooner. Unsure how much I got from the interview, as the participant was not as forthcoming as the others had been. This interview was significantly shorter in length in comparison to the others.

Interview 5:
Does not tell anyone about their CF. Embarrassed about the cough.

Interview 6:
Participant kept stressing the importance of exercise. Did not feel I was able to connect with the participant as I had done with others. Felt I had to keep prompting for answers but this may have been because the participant did not have much to say or because the participant needed to get away quickly after the interview.

Interview 7:
Mentioned coughing up phlegm being embarrassing - no one else has done so. Participant was easy going to talk to. Possibly because I was a similar age to the participant. Participant was really reflective of experiences that they had gone through – really interesting interview and understanding the participants experience.

Interview 8:
Had already established rapport with participant prior to interview as this booked face-to-face rather than over telephone or e-mail.

Interview 9:
Limited impact of condition – considers self very lucky

Interview 10:
Surprised to hear that the patient did not experience symptoms and therefore having CF had no impact. My assumptions of having a chronic condition would be that maybe having it might. Although I have asthma which is considered a chronic condition but I do not really think about it. Maybe the symptom severity has an impact on condition. The more the symptoms, the more the condition is on your mind. Most of the conversation was about fertility. Interesting to hear how the diagnosis came about. Not being masculine by not fathering a child naturally? Participant was very open, seemed to be able to build a rapport quite quickly. Considers self-lucky. Difficult to ask some of the questions as they did not apply when the participant reported no impact of the condition.

07/06/2016 – Reflections of interview 11

Had not done an interview for quite a few months, felt quite relaxed, was not worried, felt confident in my skills. Had built a rapport with participant via e-mail and phone calls. Was quite a quick interview in comparison to the others. Concerned for time as participant had to get away quickly. Main point that came across from interview 'having a positive attitude'.

07/07/2016 – Transcription notes from interview 3

Being in control is important
Downward comparison – least I'm not like others
Injustice – getting access to treatment
How the family responds to CF
Comparison to being diagnosed with cancer
Feelings of guilt – unable to do things prior to diagnosis
Wasting life before diagnosis
My illness is responsible for my problems
The puzzle has come together
Accepting help from others – wanting it and not wanting it challenges independence

22/07/2016 - Reflections of interview 12

Had not carried out an interview or done any research for a while – feel completely out of the research mind. Was running late for the interview so felt quite stressed on route and initially when starting the interview. Built rapport with the participant very quickly. Exercise – engagement in this came up a lot. Similar to a previous participant. Did not appear that diagnosis seemed to have a huge impact. Worries of life expectancy – this has come up a few times. Unsure if I am getting anything new in the interviews.

26/07/2016 - Notes taken during interview 13 and reflections post interview

Asthma – smoker – had a child – gave up smoking – went to doctor with bad chest infection

Requested chest x-ray. Radiologist asked questions – regarding chest pain. Diagnosis appointment got moved a month earlier

CF was previously mentioned – "it was a shock"

Missing pieces to the puzzle – explained other health issues

Acceptance – denial → attitude "I've got CF it doesn't define me" – "CF stops me" "I was over doing it" – certain limitations, drug compliance

"haven't got time"

Angry → GP
→ with mum

Wanted information → what type of info → the more I know the more I could help
Feeling – anger gone → was useful to a point

Apprehension – my life's going to be different

Reflection
Participant was chatty, felt like we connected, participant was very open. Unsure if there is anything new coming up – have I reached saturation? (decision made to stop interviews and concentrate on transcribing). Felt very tired as the end of the interview. Participant had a lot to say and so a lot of concentration needed.

31/07/2016 – Transcription notes from interview 4

Participant was talking about feelings of neglect from lack of diagnosis
How can the CF have been missed by the health professionals
Disbelief of being diagnosed

05/08/2006 - Transcription notes from interviews 4 and 5

Interview 4:
Doing daily things are hard
CF stops me from travelling/going abroad – worry of something happening
No longer have the energy to do things
CF is a struggle
Downward comparison to children diagnosed with CF
CF has changed my life

Interview 5:
Sceptical of the medical professionals – missed CF
Relief of diagnosis (initial)
Felt depressed about diagnosis – death
Burying head in the sand – denial
Acceptance of chronic condition
What things does the participants miss out on because of CF – engaging in relationships
Frightened of inpatient admission
Treatment is intrusive
Recognise importance of doing medication
How does family respond to CF

18/08/2016 - Transcription notes from interview 7

Confusion over hospital appointment – participant was told about diagnosis, was not expecting it, had anticipated further tests
Did not hear anything after being told about CF– no information given, counsel

patients? (possible recommendation)
Disengaged with service – false diagnosis/blaming of service
Did not engage in treatment
Does not like hospital admissions “if in hospital you must be sick” – relationship to CF? – does not think that she’s that bad
Ignored my CF
CF impacts my life

10/10/2016 – Transcription notes from interview 10

It had been quite a while since I had done the interview so when I was listening back and transcribing it was hard to remember the interview. Participant reported not being affected by diagnosis – more infertility. In interview I was surprised by lack of impact of CF – feels awkward listening back to it as I had made an assumption

20/10/2016 - Transcription notes from interview 8

Having a doctor who takes you seriously when being diagnosed same as interview 2
Beliefs around where CF comes from
Exercise is important
Phlegm – coughing
Diagnosis reaction

03/11/2016 - Reflections of interview 13

Failure from GP
Description of own CF in adulthood – strong
Symptoms make sense

Participant was very chatty – meant I did not have to ask may questions similar to interview 11. Noticed I tended to ask more closed questions. Did not ask about future – time thing or afraid? Was uncomfortable asking about sex life – personal area

Reflection to ask in next interview:
Hospital admission experience:
What can be done to make it better?
Would it have been better to know your first experience would be like this
Embarrassment of the phlegm
Effect on intimate relationships
Future

11/11/2016 – Reflections of interview 14 and 15

Interview 14:
Found it harder to get participant to expand on answers – was not as chatty as some of the other participants.

Interview 15:
The participant was easier to have a conversation with.

24/11/2016 - Reflections of interview 16

A lot of what was said was similar to other interviews. Built good rapport with participant.

30/01/2017 – Research reflections

"missing piece of the puzzle" – Patient is the expert in their own health

Story of diagnosis
Looking for answers
Finally got answer hoping it will solve things
Leads to more questions

"riding the wave with the patient"

20/02/2017 - Progression meeting 2

Reviewed introduction and methods chapter.

Not really sure of direction of research – what's novel? Feeling like I am going through the fog. Need to trust in myself in that the process works.

05/03/2017 - Research reflections

Making sure that I am consistent with coding. Do not want to miss any of my coding – do not want to miss anything.

20/03/2017 – Reflections from coding process

Initial codes created and then added as I reviewed other files. Has become hard to keep track of names of codes - might be some duplication/worded differently

06/04/2017 - Reflections from coding process

From looking at interview 7

Recommendations

managing people's expectations

* participant had thought they were going for further tests
* was instead given a diagnosis
* did not think she would have CF
breakdown in communication???

delivering bad news?
* another participant similar experience
* person was matter of fact and went to get appointments diary

is it because the service are not used to doing this

** look at delivery of bad news literature
** stages of grief literature

Ideas
diagnosis exp
* sickly child
* dismissal by GP
* diagnosis process often long
* shock or relief

*get on with it and cope fine

or

* don't accept diagnosis

* hospital admissions are difficult

Stages of grief

13/04/2017 - Reflections from coding process

improvement for service provision

Talk through like a journey

people have various degrees of symptoms

GP - ignored
Get diagnosis

Diagnosis
- clinicians just being there with the person
- having an appointment booked close by
- anything that the patient needs
- in a timely manner

Information needs
- answer any questions
- access to appropriate information
- not to overload the patient - booking them in medical management etc.
- what is your understanding of CF? what have you taken from what I've said?

- Do not belittle someone - their symptoms and how they feel are valid

21/04/2017 - Reflections from coding process

Family worry about CF
- worry about family this makes you ill

CF does not make you unwell it's the other stuff - finances, etc.

- practical help from family was beneficial (picking children to and from school)
- practical help from friends coming along to appointments

No one tells you what's going to happen when're unwell

Reflection: picking up work

Can change the way I'm coding

Can't quite remember the codes

Complacent with coding

27/04/2017 - Reflections from coding process

Getting the diagnosis
information needs to be met
patients need time to process the diagnosis - being respectful

Treatment
giving appropriate feedback / timely - or managing expectations
patient knows with annual review will get thorough check over - happy for team to identify any problems (trust in team) - otherwise once they can manage their condition they are happy to just get on with life
most feel supported with their teams

Recommendation
providing tailored care, asking questions to patients about what they would like to know, how much they would like to know

having appropriate information allows you to be in control of CF and manage it

24/06/2017 – Coding reflections

Initially coded the first two transcripts and went through them with two other people. Used that as a basis to code transcripts and added codes along the way. Problem with this ended up with 254 nodes which was too many.

You have good days and bad days - put this under the code CF is unpredictable

Reviewed you can't have CF - all codes okay
New node created support vs unsupported put all support related codes under this heading
Changed name of I don't need support to I'm independent
Unsupported moved into unsupported by CF team
Feeling supported code moved into supported vs unsupported
Support networks moved out of feeling supported
Feeling supported node deleted
Nodes (CF team, family, friends, work) moved out of support networks.
Support networks deleted
Reviewed work node everything okay

05/08/2017 – Coding notes

Coping
- reviewed get on with things

Moved coming to terms with it into acceptance
CF is mild node deleted
Relief to be diagnosed moved to emotions around diagnosis
Dismissal by HCP and Ignorance from HCPs moved into experiences of seeing HCPs
New node created - fear
Fear of being alone, fear of health declining and fear of the unknown moved into fear node
First admission was scary moved into admission
Changed downward comparison to there are others worse off
Could have done more for health moved into taking care of health
This is who I am moved into identity
Expected it was CF moved into Diagnosis experience
New node created - The future
planning for the future, planning for the inevitable, uncertainty for the future, and uncertainty moved into this node
Treatment and treaded like a leaper moved into taking care of health
Rudeness from staff moved into negative experiences of HCP's. Rudeness from staff deleted
Wasn't looking after self moved into taking care of health

17/08/2017 – Coding notes

I can now be looked after moved into diagnosis experience, name changed to getting the right treatment
Attention seeking name changed to the one bit of normality left, moved to coping
I didn't choose this it just happened moved to coping
Get a break going to hospital moved to admissions
Health belief moved into diagnosis experience
Difficult explaining to others changed to illness spoils it for others, moved to coping
Resentful moved to coping

Everyone's different changed to everyone's CF is different moved to coping
I'm invincible changed to thought I was invincible
Other health problems worse than CF
Frustration node deleted
Poor health - one reference moved to health before diagnosis in diagnosis experience, other reference moved to it's not as bad as you think, poor health deleted
Cancer is known changed to CF is unknown
Depression node deleted
Having something to prove moved to coping
Negatives of CF moved into coping
I'm not that sick deleted
Get the right treatment moved to diagnosis experience
Get the right treatment and getting the right treatment combined

10/09/2017 – Coding notes

At point of diagnosis
- there are preconceived ideas of what it means to have CF
- Thin people/childhood disease
- People ca not relate to it
- Make diagnosis difficult to accept
- Limited relatable information makes it difficult
- Online or verbally told by staff seems to be the main source of information
- Told by people not to read the internet
- At loss as to where to go
- If told by staff will forget that information
- Knowledge is important to help understand condition

- sickly child
- turned away from GPs
- no one thought it was CF
- at diagnosis - double edged sword
  - relief of being diagnosed
  - reduction in life expectancy
  - lost faith in medical professionals
- engaged vs non engaged
- information is limited

- Beliefs about CF prior to diagnosis deleted
- Beliefs about CF expanded to include beliefs prior to diagnosis
- It's life limited / variations of CF created
- Understanding of CF before diagnosis deleted
- CF information reviewed
- Health belief delete
- CF is unknown changed to people don't know about CF
- If only we knew my diagnosis deleted
- Not a big deal deleted
- I got the answer deleted

- Tried lots of things deleted
- Pre diagnosis node created, health before diagnosis moved into node
- Not taken seriously deleted
- Nothing's wrong with me deleted

- CF doesn't stop me doing anything deleted
- Having to put self first deleted

***Pre-diagnosis, exp's of seeing HCP's
***Diagnosis exp - diagnosis a double-edged sword
***Diagnosis exp - control
*** Diagnosis exp - route to diagnosis
***Living with CF - managing the challenges of CF
***Living with CF - impact of CF

03/11/2017 - Coding notes

Feeling defeated deleted
Seriousness of the condition deleted
Negatives of CF deleted
This is who I am deleted
CF has taken its toll deleted
CF affects romantic relationships renamed to CF affects relationships (CF affects relationships moved over and deleted)
Treated like a leper deleted
Others know when I'm unwell deleted
Difficult to ask for help deleted

Appendix C – Reflective chapter

Learning is defined as a "process whereby knowledge is created through the transformation of experience" (Kolb, 1984, p.38). According to Kolb, effective learning involves a person progressing through four cyclical stages: 1) being actively involved in a new situation or experience (concrete experience), followed by 2) observation of and reflecting on the experience (reflective observation of the new experience), which leads to 3) modifying or forming new ideas following the reflection (abstract conceptualisation), which are then 4) tested out and applied, resulting in new experiences (active experimentation). I have chosen to use Kolb's model to reflect on my experiences and learning from undertaking the research element of the Professional Doctorate in Health Psychology programme.

My research journey started many years before joining the doctorate programme. After completing my undergraduate psychology degree, I searched for relevant work experience and made contact with a local Clinical Psychologist working within a regional CF centre. Unbeknown at the time, this opportunity marked the start of my personal and professional development as a Health Psychologist. The idea to undertake research within the area of CF in adulthood had begun to formulate a few years prior to joining the doctorate programme. Following a review of the adult CF literature and anecdotal evidence from healthcare professionals working with this population group, it was identified that this was an important area to undertake research. With limited academic support, I struggled to develop and take the research idea forward. When applying for the doctorate programme, I saw this as the perfect opportunity to develop my research ideas further with the support of my supervisors.

Researcher's knowledge and experience

Prior to this research project, I had limited experience in conducting qualitative research. My only experience of undertaking qualitative research was from my MSc in Health Psychology, six years before starting to interview for this study, in which, IPA was used as the method of analysis. Looking back, my research skills were at the time limited due to my lack of experience and confidence in interviewing people and understanding of qualitative research. Since undertaking my Master's research, I gained substantial experience of working in clinical roles providing therapeutic support to patients with chronic and/or long-term health conditions including CF. These roles allowed me to develop my knowledge, skills and confidence in undertaking qualitative research as specified by Braun and Clarke (2013). I developed the expertise to quickly build a rapport and trust with an individual and interview them on a personal topic, to ask questions which allowed me to explore and understand an individual's meaning, to listen to what was being said whilst reflecting on what they might mean and to reflect on my own experiences and be aware of my assumptions. At the time of collecting the data for this research, I felt confident in my ability to interview participants and explore their 'world'. Prior to this research, I had no previous experience of using TA as a research method. To build on my research skills, I attended a training session delivered by Victoria Clarke on TA (who co-developed the method of TA used within this research) and read key papers by the authors.

Research process reflections

Throughout my research journey, I faced several challenges and opportunities to reflect and learn from my experiences, which are discussed below.

NHS ethics process

Applying for NHS ethics was a process I had never undertaken before. From speaking to others who had gone through the experience, I was advised that it can be an arduous process. At the time of completing the ethics application, I felt mentally exhausted after handing in my professional skills module. Overall, I found the ethics process overwhelming and at times struggled with knowing the level of detail to include. When having my ethics application reviewed, I opted to attend a virtual committee meeting rather than waiting for a face-to-face meeting. In retrospect, this was not the best decision as my initial application received an unfavourable review. I was devastated by this news as it felt like a major setback in my project timeline. I was able to learn from this experience and when I resubmitted my application, I received a favourable opinion. Whilst it was challenging, I found the ethics process beneficial and conducive to the development of my research. It forced me to think through potential issues and to consider what I would do to manage such situations.

Patient and public involvement

Patient and public involvement (PPI) is defined as research that is carried out ‘with’ or ‘by’ members of the public rather than ‘to’, ‘about’ or ‘for’ them (INVOLVE, 2012). Involvement refers to when members of the public are actively involved in the research, for example, as members of a steering group or commenting and developing patient information leaflets or other research materials (INVOLVE, 2012). Incorporating PPI within research is invaluable with many benefits, such as, providing a different perspective, improving the quality of the research through making the language and content of information provided more appropriate and

accessible and making the research more relevant through helping to reshape and clarify the research (INVOLVE, 2012). Following good research practice, I attempted to incorporate PPI involvement from the early stages of the research process by contacting the CF patient expert advisor for the service. However, this was met with limited success. Following this, I contacted the PPI officer at the local NIHR research design service for advice on incorporating PPI within the research project. An advert was distributed amongst the research design services e-mail mailing list asking for people to contribute by commenting on research documents such as the participant information sheet and consent form. As this research was self-funded, there were no finances available to reimburse any time costs. Unfortunately, I had no response from the advert, which was frustrating. Due to time constraints, I had to progress with my research without PPI involvement. However in retrospect and for future research, I would begin the process by referring to guidance documents such as the UK standards for public involvement (2019) and briefing notes for researchers: public involvement in NHS, public health and social care research (INVOLVE, 2012), which offer practical support on what good public involvement in research looks like and how to best involve members of public within the research. I would look to establish a patient steering group[8], inviting people diagnosed with cystic fibrosis in adulthood or childhood to be involved. Individuals could come from the CF centre in which the research took place or the invite could be extended further with support from the CF Trust or other CF centres. The group could be involved in all aspects of the research process from shaping the research question, reviewing documents such as the

[8] The patient research group would have to be a virtual group due to potential cross infection risks.

information sheet and interview questions, involvement in the data analysis and informing potential implications.

Recruitment process

As it had taken longer than anticipated to get the necessary ethical approval for the research study, I was keen to send out the invitation letters to participants as soon as possible. This meant sending out the letters just before the Christmas holidays. Although at the time I thought that this might not be the best for the recruitment outcomes, I decided to send out the letters anyway. In hindsight, I may have had a better initial response rate if I had sent the letters out in the New Year. After sending out the follow-up letter, many of the participants at the interview had informed me that they recalled receiving the initial invitation letter, however, as they were busy at the time with the holidays they had put the letter aside and forgotten about it. For future research projects, it is important to consider the timing of when invitation letters are sent out to potential participants to maximise the recruitment possibilities.

In one of the interviews, a participant had been recruited following a conversation with a member of the CF team. Within the interview, the participant had talked about her low literacy levels and how she often ignored paperwork if it seemed too complicated. This made me reflect on the recruitment process used for this research in that the participant was willing to participate, however, may have been put off by the letter and information sheet that was sent to her. When undertaking research in the future, I would need to think about how the research documents are presented so that it does not deter participants.

Interview experiences

As previously mentioned, I went into this research with a lot of experience of meeting people for the first time, tasked with having to build a rapport very quickly and having discussions around sensitive topics. Despite this, I felt nervous about my initial interview. I did not know if the interview questions and the schedule would work. Whilst it would have been ideal to pilot the interview schedule and attempts were made to do so, but as mentioned this was met with no success. I could have piloted the interview schedule but due to the small pool of potential participants available, I decided against this. I was unsure of how successful I would be with the recruitment of the study and could not afford to use one of the interviews as a pilot. In addition to this, my initial interview was my first experience of interviewing in a participant's home, therefore it was an environment that was unfamiliar to me. As I settled into the interview, I found that as time went on I quickly became more confident with my interview approach. Whereas before I was more rigid to sticking to the interview schedule, as it progressed, it became more of a guide.

The environment in which the interview takes place is important as it can impact the conversation and therefore the knowledge created. I noticed that the interviews that were carried out in the participants' homes tended to last longer than the ones in the hospital. This may have been for numerous reasons. For example, those that I had interviewed at home may have had more to share about their experiences, they may have been more comfortable in their home environment, they may have been less reserved individuals, or because those who were interviewed at the hospital may have parked their car in the hospital car park and were concerned about parking charges. Once again, as my research project was self-funded it meant I was unable

to reimburse travel costs, whereas most research projects usually budget for and offer this.

When carrying out the interviews, at times, I was very aware of my demographics (being a young, female and British Asian researcher) and how these may have influenced the interviews. Although I have had experiences with interviewing male participants, I found it easier to build a rapport with the females enabling them to share personal stories. Often the conversations with the women were dominated by the participant speaking with little need for prompting or follow-up questions from me, whereas with the male participants it seemed to involve a more formal interview question answer style dialogue. There may have been a number of reasons for this for instance, the women participants may have been chattier than the males, they were able to relate with me as another female or the impact of CF may not have been as significant for the male participants and therefore they did not have much to share in their interviews. My first interview with a male participant was with an individual who was twice my age, in this particular interview, I was very aware of the relational dynamics and how different they were between the participant and I. This experience may have clouded my other interview experiences with men. In some of my interviews, I was aware of the age difference between myself and the participants and therefore, how open participants may have been with me. In one interview a comment was made about my age and how young I was. This made me wonder if this was a barrier to participants and what they were able to share with me. At the beginning of all my interviews, I tried to find common ground with all of the participants to help build a rapport and encourage participant disclosure.

As previously mentioned, it was important to establish the right balance between maintaining control as the researcher whilst allowing the interviewee thinking space to generate novel insights within the interview. At times this was a challenge, particularly with chattier participants who had negative experiences. It was difficult to bring these participants back to the topic or to get a word in to ask follow-up questions or move the interview on. I was grateful for the time interviewees gave me and so I wanted them to benefit from the interview by having the opportunity to be heard and tell their story whilst also being mindful of the time and my interview schedule. I balanced the control and flexibility through a number of ways such as, asking broad open-ended questions, going off topic discussing something raised by the participant and then coming back to the topic guide, as well as asking participants at the end of the interview if they had anything else that they wanted to add which had not been covered or discussed. The last method was particularly useful in one of my interviews in which I had thought I was bringing the interview to a close. Usually, at this point, most interviewees would tend to say that they had felt everything had been covered. However, this participant began to share her experiences of how frightened she had felt on her first inpatient admission. Often when I was debriefing at the end of the interviews and shared back a summary of the interview, participants would make points or share experiences related to the interview questions.

Moving from a practitioner-based role to a researcher role was at times a challenge. During some of my interviews in which participants had particularly difficult experiences or were upset, it was hard to sit with what participants were telling me continuing my role as a researcher to collect information and not intervene as I would

with my ‘practitioner hat’ on. At times, I did notice myself start to engage with the participant as I may do so in a therapeutic setting but had to stop myself from doing so.

Following one of the interviews, a participant shared information with a member of staff that she reflected on after our interview, which she asked to be passed onto me. The participant talked about how CF had an impact on her sex life through the need to cough up phlegm. This was a topic area that had not been raised before by any of the other participants. Whilst I was interested to explore this area further with other participants, I found it very difficult to raise the topic and to discuss participants’ sex life in subsequent interviews as talking about sex can be considered taboo. I worried that participants might become uncomfortable, embarrassed or unwilling to speak about it therefore, impacting the relationship created. In retrospect, this was a missed opportunity as it may have had an impact on other’s lives. Research by Anderson (2014) found that participants with a childhood CF diagnosis had described the effects of CF on their sex lives including decreased desire or motivation to initiate sex, tiredness, the effect of coughing on sexual activity, how medical touch affected intimate touch, performance anxiety, men’s concern regarding the amount of ejaculation produced, infection risk and adaptions made to accommodate symptoms of CF. Through initiating the conversation with participants about it, it would have given them permission to choose to talk about it further or to decide to decline to answer questions.

Although initial recruitment to the study was slow, it quickly picked up and I found that within a short period I had several participants interested to take part. I was keen

to book the participants in once they had expressed interest but this meant that I was unable to transcribe and review my interviews in between. Most of my interviews were carried out in quick succession with little time to reflect on them in between. Ideally, I would have preferred to carry out a few interviews and then begin the initial analysis phase. This may have changed the focus of my line of questioning.

Of the eleven interviews carried out in the participant's home, five interviews involved having another family member, either a partner, children or a parent around at the time. A further three interviews involved having a dog present. In one of the interviews, the participant's mother sat in for part of it, which may have impacted on what the participant shared at the time the questions were asked. In one of the hospital interviews, the participant's partner sat in towards the end of the interview and contributed to the conversation. It was interesting to hear comments from the partner's perspective. Although this research was limited to interviewing patients and exploring their experiences, it might be interesting for further research to hear the perspective of other family members. On some occasions, the interview was briefly disrupted as a family member came into the room but for the majority of the time, they were in a different room in the house. Interviews that took place with dogs present were challenging as they would make a noise for attention which was distracting for the participant and I. Although the disruptions were minor, they temporarily affected the flow of conversation, whereby either the participant or I had forgotten our train of thought.

Data analysis and writing up

Transcribing and data analysis was another demanding area within the research process. Transcribing my own interviews was beneficial as it enabled me to stay close to the participants' voice, which was in line with my epistemological and ontological stance. However, some of the interviews were very lengthy, making transcribing a time-consuming task. In the beginning, it was difficult to know what level of detail to include within the transcription, for instance, utterances, pauses, space fillers and unrelated conversation. Due to time pressures, I decided for a few of my interviews to be transcribed for me. Whilst this was a tough decision to make, I felt this was best for ensuring the project moved forward in a timely manner. I also reminded myself that it is common practice within research settings for transcription to be carried out by a third-party. To stay close to the data, I reviewed the completed transcripts to check for accuracy.

Whilst I was excited to begin the initial data analysis phase, I had never used a data analysis programme to analyse my data before. I undertook training to develop my knowledge and competence in using the programme. I found that using NVivo made the coding process much simpler in comparison to doing it manually. However, I made a novice error of creating too many codes and in cases duplicating codes. When it came to synthesising the data, I found I was drowning in it and would get caught up with minutia detail. Deciding which areas to focus on and include in the final write up was difficult. Supervision from my supervisors was beneficial in guiding me through the process.

Conclusion

In summary, undertaking this research has been one of the most challenging experiences I have endured to date. It has been physically and mentally exhausting and pushed me to my utmost limits. Throughout the process, I have been continuously tested on my ability to prioritise and manage competing demands against my perfectionist behaviour traits, often at the expense of my health and wellbeing. Based on Kolb's theory, each element of the research journey has been a learning curve with high and low points allowing me to develop from the experiences. No doubt my confidence and competence as a researcher and professional have increased. This journey reminds me that I have barely scratched the surface and much to learn about qualitative research. I have particularly enjoyed undertaking the interviews, developing themes from the data, having the opportunity to present the stories of those diagnosed with CF in adulthood and seeing the research come to fruition. I have learnt immensely throughout this process about myself both professionally and personally and I will carry these experiences forward into the future.

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Appendix D – Participant interview schedule

Image redacted for anonymity reasons

University of the West of England BRISTOL

PARTICIPANT INTERVIEW SCHEDULE

*** For telephone and video call interviews: Have you been well since the time you sent back the consent form and today's interview?**

Diagnosis

1. **Can you tell me about how you came to be diagnosed with CF?**

Prompts:

- How old were you at the time of diagnosis?
- How long has it been since diagnosis?

2. **What was it like for you to be diagnosed?**

- Do you remember how you felt?
- What thoughts went through your mind?
- What did you know about CF at the time?
- Who told you about the diagnosis - GP/CF team?
- Did you think anything was wrong prior to diagnosis? - How did you make sense of that?
- How did having a diagnosis of CF change (or did it change) the way you managed your physical health?

Impact of CF

3. **Can you tell me what impact CF has on your life?**

- relationships - family/friends/CF team
- work
- health

- treatment/medication?
- social life
- your future?
- Has the impact of CF changed over time? If so, how?

Coping with CF

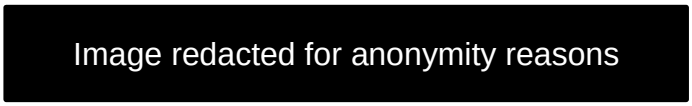
4. **How do you cope with and manage your CF?**
   - What do you do well /what works for you?
   - What do you find difficult?
   - How do you manage the difficult things?/How have you tried?
5. **What are your hopes and fears for the future?**

Support

6. **What support have you had with your CF? by whom? and when?**
7. **What support do you wish you had/have?**
8. **Has the help you needed changed over time and in what way?**
9. **If we could change things to help patient's in the future, what should we do on the basis of your experience?**
   - What might it look like?

Summary

10. **To me the main points from this interview have been...(summarise main points), have I missed anything out?** I'm going to do a few more interviews, would you mind if i got back to you if necessary.



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BRISTOL

Demographic information:

The information in the table will be noted throughout the interview as it is disclosed by the participant. Anything that is not covered during the interview will be asked by the researcher at the end of the interview.

Gender:	Male	Female	Trans*		
Ethnicity:					
Age of participant:					
Level of education:	GCSE's	A Levels	Undergraduate	Masters	Postgraduate
Employment:	Part-time	Full-time	Not working - looking for work	Not working - due to health	Other
Job role:					
Changes in work due to health:					
Relationship status:	Single	Married	Divorced	Widowed	Other
Age at diagnosis:					
Time since diagnosis:					
Family:	children? (has a CF diagnosis affected having children or not)				

Field notes:

Appendix E – Participant invitation letter

[redacted]

Letterhead and information on this page redacted for anonymity reasons

[Insert Date]

Dear [Insert Patient's Name],

I am writing to inform you about a research study being carried out within the [redacted] Cystic Fibrosis Service, which you may be interested in.

Along with this letter, please find attached an information sheet that will tell you about the research study, what it would involve, the benefits and drawbacks and how to take part in the research.

If you are interested in being involved with the research, please get in touch with Nisha Sharma before the end of November 2015 and let her know the best time to contact you and if you have a preferred and an alternative contact method (e.g. mobile phone number, land line number) by either:

- Telephoning the CF admin office on [redacted]
  OR
- E-mailing the CF admin team on [redacted] with the subject heading 'Research', addressed for the attention of Nisha Sharma.

If the recruitment for this study does not go as planned and we have not heard from you, we may contact you again by letter to see if you are interested in the study. If you are not interested in taking part or do not wish to be contacted, please let us know.

If you have any questions about the research, again please do not hesitate to contact Nisha or myself on the above contact details.

Yours sincerely,

Dr [redacted]
Chartered Clinical Psychologist

Encl. Participant Information Sheet - Version 1.5 27.08.2015

Participant Invitation Letter - Version 1.5 28.08.2015

Appendix F – Participant information sheet

Image and information on this page redacted for anonymity reasons

University of the West of England
BRISTOL

PARTICIPANT INFORMATION SHEET

IRAS Project ID: 189640

Cystic fibrosis diagnosis in adulthood: Patients' views.

We would like to invite you to take part in a research study, which is part of an educational qualification for the lead researcher. Before you decide whether to take part, it is important for you to understand why the research is being done and what it will involve. Please take the time to read the following information carefully and discuss it with others if you wish.

After reading this information sheet, if anything is unclear or you would like more information, the researcher will be happy to discuss this with you. You are free to decide whether or not to take part in this study. If you choose not to take part, this will not affect the care you receive from the cystic fibrosis medical team.

Thank you for taking the time to read about the study.

What is the purpose of this study?
The aim of this study is to understand what it is like to be diagnosed with cystic fibrosis in adulthood and what support people might want.

Why have I been invited to take part and am I eligible?
We are inviting patients who have been diagnosed with cystic fibrosis in adulthood who receive their care from the [redacted] Cystic Fibrosis Service to take part in this study.

You are eligible to take part in the study if:
- You were diagnosed with cystic fibrosis in adulthood.
- It has been a minimum of a year since you were diagnosed with cystic fibrosis in adulthood.
- You are aged 18 or over.
- You are a native or fluent speaker of English.
- You are physically well at the time of the meeting with the researcher.

Do I have to take part?
No, it is up to you to decide whether to take part or not. If you are interested in taking part, you will have the opportunity to read the information, consider any questions you have and speak to or meet with the researcher who can give you further information and answer any questions you may have.

If you decide to take part you will be asked to read and sign a consent form, but you are still free to withdraw from the study at any time without giving a reason. If you decide not to take part in the study it will not affect your routine medical care in any way.

What will happen to me if I take part?

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If you agree to take part in the study, you will be contacted by the researcher to arrange a face-to-face interview. If this is not possible, alternative options including a telephone interview or video call (e.g. Skype, Facetime or Google Hangout) could be arranged instead. The interview is intended as an opportunity for you to talk about your experiences of being diagnosed and living with cystic fibrosis in adulthood. By participating in this research you will be asked to complete one interview with the researcher where you will be asked some questions. The interview will last approximately 60 to 90 minutes.

The face-to-face meeting will take place either in a private room at the [redacted] Cystic Fibrosis Service at [redacted] Hospital or your home, whichever is more convenient to you. If you decide to have the meeting at [redacted] Hospital, every effort will be made to co-ordinate the appointment with your next clinic visit. The telephone or video call interview would take place from a private room in [redacted] Hospital.

Will I get paid for taking part in this study?
There is no payment available for taking part in this study. However, as a token of appreciation for your time, at the end of the interview, you will be asked by the researcher if you would like to be entered into a prize draw for the opportunity to win a £25 Amazon gift voucher. You will need to let the researcher know within a week of participating in the interview if you want to be included. The researcher will record your contact details. The winner for the prize draw will be drawn after all of the data has been collected for the study. You will be notified by the researcher if you have been successful or unsuccessful. You do not have to enter the prize draw if you do not wish to.

What are the possible benefits of taking part?
There are no direct benefits to you as an individual in taking part. However, the information you provide may help us to better understand the impact of an adult diagnosis of cystic fibrosis and what it is like to live with the condition. As these issues have not been widely researched, it is hoped that this study will improve our understanding in these topics and may lead to improvement in care for those diagnosed with cystic fibrosis in adulthood.

What are the disadvantages of taking part?
We do not anticipate there being any disadvantages to taking part, but we do realise that this can sometimes be a difficult and emotional topic to talk about, so it is possible you may feel uncomfortable or slightly distressed by the conversation. If you do, you can chose not to answer any questions that you find difficult and you can decide to stop the interview at any point. After the interview, you can speak to Dr [redacted], Dr [redacted], or Dr [redacted] (Clinical Psychologists) about any issues that are raised by taking part in the research.

What should I do if I want to take part?
If you would like to take part in this study please contact Nisha Sharma on the [redacted] Cystic Fibrosis Service office number on [redacted] or e-mail [redacted] with the subject heading 'Research' addressed for the attention of Nisha Sharma by the end of November 2015. Please let us know the best way and time to contact you and if you have an alternative contact method (e.g. mobile phone number, land line number).

Image and information on this page redacted for anonymity reasons

University of the West of England BRISTOL

What will happen to my information?
Your interview with the researcher will be audio recorded and later typed up. This will be reviewed and analysed by the research team. The researcher will identify themes from all of the interview data. You will then be offered the opportunity to comment on the themes if you wish.

How will my information be kept confidential?
If you join the study, some parts of your medical records and the data collected for the study may be looked at by authorised people to check that the study is being carried out correctly. All will have a duty of confidentiality to you as a research participant and we will do our best to meet this duty. Any information that is disclosed by you, which is criminal in nature or any other information that is required by the law to be disclosed by the researcher, will be passed on to the relevant authorities.

Ethical and legal practices regarding data collection and storage will be followed and all information about you will be handled in the strictest confidence. The steps taken to ensure confidentiality are detailed below:

- Your consent to take part in the study will be recorded on a form that will include your full name. These forms will be stored in a locked filing cabinet in [redacted] or [redacted] which will only be accessible to the research team. A copy of the form will also be stored in your medical file.
- Interview data will be stored separately from the consent forms and will not include any personal identifying information. Your data will be stored using a unique, anonymous study identification number.
- A table linking your anonymous study identification number with your name will be stored on a password protected NHS computer which will only be accessed by the researcher.
- Any report of the study's findings and the quotes contained within it will contain no identifying information.
- If you opt to take part in the prize draw, a table with your name and contact details will be stored on a password protected NHS computer which will only be accessed by the researcher.

What will happen to the results of the study?
The information collected during this research study will be written up as part of a doctorate in Health Psychology for the researcher Nisha Sharma. A brief summary of the study findings will be posted on the [redacted] Cystic Fibrosis Service's website after September 2016. It is anticipated that the results of the research will be published in an academic journal. If you would like to be provided with a summary of the results please let the researcher know.

What happens if I change my mind?
You have the right to withdraw from the study without your legal rights or medical care being affected. Your participation is voluntary and you are free to withdraw your approval for the use of the interview data up to 28 days after the interview. If this is the case, please contact the researcher who will then destroy the interview data in the appropriate manner.

Who is organising and funding this study?

Image and information on this page redacted for anonymity reasons

The study is sponsored and organised under the guidance of the University of the West of England and is funded by the researcher.

Who has reviewed this study?
All research in the NHS is reviewed by an independent group of people, called the Research Ethics Committee, which is there to protect your safety, rights, wellbeing and dignity. This project has been reviewed and given a favourable opinion by the South Central – Hampshire B Research Ethics Committee and also the University of the West of England's Research Ethics Committee.

Who do I contact if I have any concerns?
If you have a concern about any aspect of this study, you should ask to speak to Nisha Sharma (Researcher) who will do her best to answer your question(s):

- Telephone: [redacted]
- E-mail: Nisha2.Sharma@live.uwe.ac.uk

If you remain unhappy, you can contact Professor Diana Harcourt (the Researcher's Supervisor) at the University of the West of England:

- Telephone: 011 7328 2192
- E-mail: Diana2.Harcourt@uwe.ac.uk

If impartial advice is required, please contact [redacted] Hospital's patient support services:

- Telephone: [redacted] (available 9:00 am - 4:00 pm Monday to Friday, with an out of hours answer phone).
- E-mail: [redacted]
- Address: [redacted]

Where can I get more information about this study?
The research team will be happy to provide further information about the study and answer any questions you may have. Please feel free to contact them on the contact details below:

Researcher: Nisha Sharma, Trainee Health Psychologist
Telephone: [redacted] E-mail: Nisha2.Sharma@live.uwe.ac.uk

Supervisory team:
Dr. [redacted], Clinical Psychologist
Telephone: [redacted] E-mail: [redacted]

Professor Diana Harcourt
Telephone: 011 7328 2192 E-mail: Diana2.Harcourt@uwe.ac.uk

Thank you.

Appendix G – Participant follow-up letter

[redacted]

Letterhead and information on this page redacted for anonymity reasons

[Insert Date]

Dear [Insert Patient's Name],

I am writing to follow up on my letter dated [Insert Date] about a research study being carried out within the [redacted] Cystic Fibrosis Service.

We have not heard from you and wanted to check whether you were interested in taking part in the study. If you are interested in being involved with the research, please get in touch with Nisha Sharma and let her know the best time to contact you and if you have a preferred and an alternative contact method (e.g. mobile phone number, land line number) by either:

- Telephoning the CF admin office on [redacted]
  OR
- E-mailing the CF admin team on [redacted] with the subject heading 'Research', addressed for the attention of Nisha Sharma.

If you are not interested in taking part or do not wish to be contacted, please let us know.

If you have any questions about the research, again please do not hesitate to contact Nisha or myself on the above contact details.

Yours sincerely,

Dr [redacted]
Chartered Clinical Psychologist

Participant Follow Up Letter - Version 1.1 28.08.2015

Appendix H – Recruitment poster

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University of the West of England
BRISTOL

Were you diagnosed with cystic fibrosis as an adult?

Do you receive your care from the [redacted] Cystic Fibrosis Service?

If so, you might be interested in taking part in a research study that is looking to improve the care provided to adult cystic fibrosis patients. Taking part involves sharing your experiences of being diagnosed and living with cystic fibrosis in adulthood.

For more information please contact:

Nisha Sharma
Tel: [redacted]
E-mail: [redacted]

Please include the subject heading of your e-mail 'Research' and address it for the attention of Nisha Sharma
Please let us know the best time to contact you and if you have a preferred and an alternative contact method (e.g. mobile phone number, land line number).

Recruitment Poster - Version 1.3 28.08.2015

Appendix I – Participant consent form

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University of the West of England
BRISTOL

CONSENT FORM

IRAS Project ID: 189640
Participant Identification Number:

Title of Project: Cystic fibrosis diagnosis in adulthood: Patients' views.

Name of Researcher: Nisha Sharma

Please read this information carefully before deciding whether to take part in this research. You will need to indicate that you have understood this information before you can continue.

PLEASE INITIAL ALL THE BOXES IF YOU AGREE WITH THE STATEMENT(S):

1. I confirm that I have read the information sheet dated 12.10.2015 (version 2.0) for the above study. I have had the opportunity to consider the information, ask questions, and have had these answered satisfactorily. ☐
2. I understand that my participation is voluntary and that I am free to withdraw at any time without giving any reason, without my medical care or legal rights being affected. I am free to withdraw my approval for use up to 28 days after the interview has taken place. ☐
3. I agree to take part in an interview with the researcher as part of this study and I am aware that some of the questions may be of a sensitive nature. ☐
4. I understand that the interview will be audio recorded. ☐
5. I understand that information collected about me during my participation in this study will be made confidential and stored securely and that this information will only be used for the purpose of this study. I understand that the recorded material, interview transcripts, and all of the study information that has been generated will need to be kept for a minimum of 5 years after the end of the study. ☐
6. I understand that my participation in this study will be recorded in my medical notes. ☐
7. I consent for the researcher to access and take information about me from my medical notes (e.g. FEV_1) for the purposes of this study. ☐
8. I understand that relevant sections of my medical notes and data collected during the study may be looked at by research supervisors from the University of the West of England, from regulatory authorities or from the NHS Trust, where it is relevant to my taking part in this research. I give my permission for these individuals to have access to this information. ☐
9. I consent for direct quotes gathered from my participation in this study to be used in any subsequent publication of this research. I understand that I will not be identifiable by these quotes. ☐
10. I agree to take part in the above study. ☐

Image and information on this page redacted for anonymity reasons

University of the West of England
BRISTOL

OPTIONAL

PLEASE INITIAL IN THE APPROPRIATE BOX:

	YES	NO
1. I would like to be entered into the prize draw for the opportunity to win a £25 Amazon gift voucher. I agree to be contacted if I have been successful or unsuccessful.	☐	☐
2. I am happy to be contacted by the research team to provide my feedback on the themes from the interview data.	☐	☐
3. I would like to be provided with a summary of the results from the research.	☐	☐

If you have any questions about this study please contact Nisha Sharma (Researcher) on [redacted] or e-mail Nisha2.Sharma@live.uwe.ac.uk.

It is unlikely that the questions in this study will cause distress but if you do feel anxious or have any concerns regarding the issues discussed, we recommend that you contact Dr [redacted] Dr [redacted] or Dr [redacted] (Clinical Psychologists) through the [redacted] Cystic Fibrosis Service office on [redacted].

If you have questions about any aspect of this study and your involvement as a participant, you may wish to contact Professor Diana Harcourt (University Research Supervisor) on 011 7328 2192 or e-mail Diana2.Harcourt@uwe.ac.uk.

Name of participant	Date	Signature

Name of researcher taking consent	Date	Signature

When completed: 1 for participant; 1 for researcher site file; 1 (original) to be kept in medical notes.

Appendix J – Example of an annotated transcript

Interviewer:

So just to start off with if you could tell me a little bit about how you came to be diagnosed with CF?

Participant:

Okay, I think I can't even remember the dates properly I think it was around about 2003 2004 um my sister up in [country name] was extremely unwell and very closed family where discussions were not held my mother had already died from ovarian cancer so with my father left he kept everything close to his chest anyway the one thing that was said was if your sister has to go back into hospital in [city name] it will probably be the last time she goes in um because the condition is that serous so the first question was well what condition because CF had never even been mentioned so that was in the January of whatever July 2003 I think and then she went back in in June and died at the age of forty three so then after that it was a case of right everybody in the family now needs to be tested and it just seemed to now be out on in the open and that was it so I've been seeing a lung specialist at the [hospital name] in [town name] for about three years prior to this umm because I had pneumonia at one stage um and he never mentioned CF or looked at it or anything so when [sister's name] died we said um "I need to be tested for CF" and he said "no you're too old we tested you for emphysema when you were in the [hospital's name] but you were too young for that" so I said "well if you test me for that why not test me for CF and knock it out of the equation" no no no not necessary so when [sister's name] died and I had

Comment [NS1]: Didn't remember when diagnosis happened – not significant?

Comment [NS2]: Sister was really unwell

Comment [NS3]: Family discussions not held

Comment [NS4]: Father kept everything close to his chest

Comment [NS5]: Sister's condition is serious

Comment [NS6]: CF not spoken about – family secret?

Comment [NS7]: Everybody in the family needs to be tested

Comment [NS8]: CF was now out in the open

Comment [NS9]: CF was never considered

Comment [NS10]: Needing to be tested for CF

Comment [NS11]: Too old to have CF

Comment [NS12]: Testing for CF is not necessary

1

her death certificate I went in and I said I'm obviously not too old she was three years older than me so do you think we should be tested now so still never got done still nothing was done about it and then the CF trust were in touch with my father up in [country name] and they said "have the rest of the family the two son's and a daughter been tested" he said "[participant's name] down south trying desperately to get tested and nobody's paying attention" so then they phoned me up the CF trust and advised me of [doctor's name] down here and then that was when it all took off and I was diagnosed with full blown CF my two brothers were carriers so that was the original story.

Comment [NS13]: Not taken seriously by healthcare professionals

Comment [NS14]: I'm not too old to have CF

Comment [NS15]: Diagnosed with having CF

Interviewer:

And how long did it take from that point to to get the diagnosis?

Participant:

Um I think it was round about the kind of later part of the year October November when I first came down here for a meeting

Comment [NS16]: Finding out about the diagnosis

Interviewer:

Mmm

Participant:

And then they said because of the funny situation was we lived at that point just in the [county name] county but our doctor was still in [county name]

because we are four miles we had moved four miles along the road so there was this [county name] [county name] border thing and um oh no you should go to [hospital name] and blah blah blah so amongst the kind of um officials there was a bit of arguing um not the team down here they said look just come down and we'll see you so they saw me and started the tests off I would have said the November December Christmas Eve I got test results saying that yeah look likely are the more tests need to be done but you know it wasn't looking great umm and then the final diagnosis was all done by the February the sweat test and all the rest of it and it was yes you've got it and then unfortunately I got a phone call from the [hospital trust name] trust people saying no you will need to be re-diagnosed at [hospital name] because your doctor is in [county name] and I just said "well actually I've not been diagnosed correctly for three years so just leave me where I am or else we'll need to take it further" and I've just been left here now so that's that so all in all I would say three four months max was what I did.

Comment [NS17]: Getting the diagnosis

Comment [NS18]: Re-diagnosis because of postcode lottery

Comment [NS19]: Time taken for diagnosis

Interviewer:

And do you remember much about around the time of being being diagnosed?

Participant:

Yeah yeah

Interviewer:

And what how did that feel for you at that time?

Participant:

I think it was more people around about me that were a bit wary about it when the initial tests were done I think the initial tests were blood tests before the Christmas and they'd said you know um we'll have these back before Christmas it was Christmas yeah I will always remember it was Christmas Eve and my husband was taking his daughter back home early afternoon and he said don't phone the hospital if you're gonna phone wait till I get back and I thought what rubbish I'll phone them anyway so I phoned up and that was when they said yeah well the initial test looked like you may have and I said "right ok" so I was a little bit kind of shocked I don't know if it was even shock it was like ok and then the sweat test and things would be done later so when [husband's name] came back to the house he said to me you've already phoned haven't you I think it was more his kind of shock I'm quite a positive person anyway for me when the final diagnosis came I remember sitting here in the quiet room and there was a lot of people in the room and I thought what's all this and [husband's name] sat there and he was a bit in shock and I just said look I said there's nothing to worry about I said life's terminal at least now I know what's wrong with me because for years I had suffered from as I said bronchitis, catarrh then I had the pneumonia I knew I wasn't feeling well and nothing was ever working seeing a lung specialist who told me one thing that was nonsense and just never getting to the bottom of anything the day I got diagnosed here as I said it was like a ton

Comment [NS20]: People around me were wary when initial tests were done

Comment [NS21]: Being told of suspected CF

Comment [NS22]: Shocked with diagnosis

Comment [NS23]: Reacting to other people's emotions

Comment [NS24]: sxPositive person coping strategy?

Comment [NS25]: Husband was shocked with diagnosis

Comment [NS26]: Wasn't worried about diagnosis

Comment [NS27]: Validation for illness

of weight lifted off my shoulders and I thought right now we know what we are dealing with doctors here are absolutely superb so you know how to deal with things tell me what I have to do and we'll take it from there

Interviewer:

Mmm

Participant:

It was the family round about that all seemed to go into spasm shock and oh my good grief what's happening and you know why we've just lost [sister's name] but for me it's just been ok now just get on with things so

Interviewer:

And how are the family now?

Participant:

Fine well they see what I'm like you know I'm so positive it hasn't stopped me doing anything touch wood I'm one of the I would say lighter sufferers you know I don't have a lot of the bad bits of the CF you know as I say the support from the people here is phenomenal so I don't have any concerns it's just you know it's been great so with that then the family see that and they just know that I'm in very good hands so

Interviewer:

Comment [NS28]: Relief to have a diagnosis – at least I know what's wrong with me

Comment [NS29]: The doctors know what they are doing

Comment [NS30]: Tell me what I have to do to manage my condition

Comment [NS31]: Just get on with things

Comment [NS32]: Family went into shock at diagnosis

Comment [NS33]: Positive attitude

Comment [NS34]: Light sufferer

Comment [NS35]: Well supported by CF team

Comment [NS36]: I am in good hands

Sounds like they are quite reassured by the care that you get

Participant:

Totally totally

Interviewer:

And tell me about you know you said [sister's name] um was very poorly what was it like growing up and how was she in terms of health wise?

Participant:

We were both kind of the same to be quite honest with you I was always kind of bigger and stature she was very very thin but she was a solicitor so she went to uni all the rest of it she hadn't been diagnosed in childhood either she was you know I think she was only diagnosed a few years before she died she but she was such a closed person she didn't like me at all she never spoke to the rest of the family very very closeted in everything so we really knew nothing about it but growing up as children well we both had the catarrh and the asthma then you know just the childhood colds up in [country name] and that was the way it was and we just carried on she went to uni straight from school very successful I went into business I was very successful you know that was just it so growing up it was just like childhood coughs colds and illness

Comment [NS37]: Sister and I were the same

Comment [NS38]: Sister was diagnosed a few years before she died

Comment [NS39]: Sister was a very closested person

Comment [NS40]: Having symptoms in childhood – asthma, cold

Comment [NS41]: Growing up we had childhood colds and illnesses

Interviewer:

So just the normal

Participant:

And that was yeah yeah

Interviewer:

I was just wondering whether you know the family ever thought anything well went on to investigate or you know take on

Participant:

No but when we went to the doctors back home in [country name] catarrh was always the thing which you never hear about these days or I don't and then a bit of bronchitis and that was it you know so cough medicines were always in the house but no that was just it that was you know I don't think my parents you know they were both professionals they never thought you trust what the doctors telling you

Comment [NS42]: Paren t's trusted what the doctors told them

Interviewer:

Yeah

Participant:

So do extra tests need to be taken no you've got bronchitis or catarrh go home and take the antibiotics and you'll be fine so that was that was the way it was done my younger brother is deaf and can't speak so I guess none of us

Comment [NS43]: You' ve got bronchitis

kids that [brother's name] was kind of more alright what's this we'll take him to the specialist down in [city name] and we'll do this and we'll do that see if there's anything with the deafness but because ours was just treated as common cold's by the GP there was no kind of then inclination that there was anything other than that

Comment [NS44]: Focus was on brother's deafness

Comment [NS45]: There 's nothing wrong with you

Interviewer:

And did you ever think yourself that there's you know anything else wrong or?

Participant:

No I just thought I was unhealthy unfit and unhealthy [laughs] and that was it but it never ever stopped me from doing anything yeah I would always have the cough wasn't actually till later on after I had been diagnosed but one of my cashiers 'cause I was a bank manager one of my cashiers in the bank said "oh I was going down Sainsbury's the other day and I knew you were in Sainsbury's" and I said "did ya" I said "how did ya know that I didn't see you" she said "well" she said "you must have been about four or five aisles up from me and I could hear your coughing" and then it dawned on me and I thought do you know what I've always had this cough so why would I notice it

Comment [NS46]: Healt h beliefs

Comment [NS47]: I thought I was unhealthy and unfit

Comment [NS48]: CF hasn't stopped me doing anything

Comment [NS49]: Didn' t notice cough

Comment [NS50]: Alwa ys had cough but didn't notice it

Interviewer:

Yeah

Participant:

And that goes way back to childhood so

Interviewer:

Yeah and did you notice I mean in terms of with the cough um I guess I'm trying to think what I was going to ask then um I don't know in terms of did you notice it yourself I know that you said you were't necessarily aware of it but anything that you picked up on I dunno

Participant:

The only things that used to I mean I was a ballet dancer

Interviewer:

Yeah

Participant:

And you know I did a little ballet tap in [area name] and got my teachers [inaudible] and I was very young then I went from that into water skiing so I was doing really active things

Comment [NS51]: Physically active

Interviewer:

Yeah

Participant:

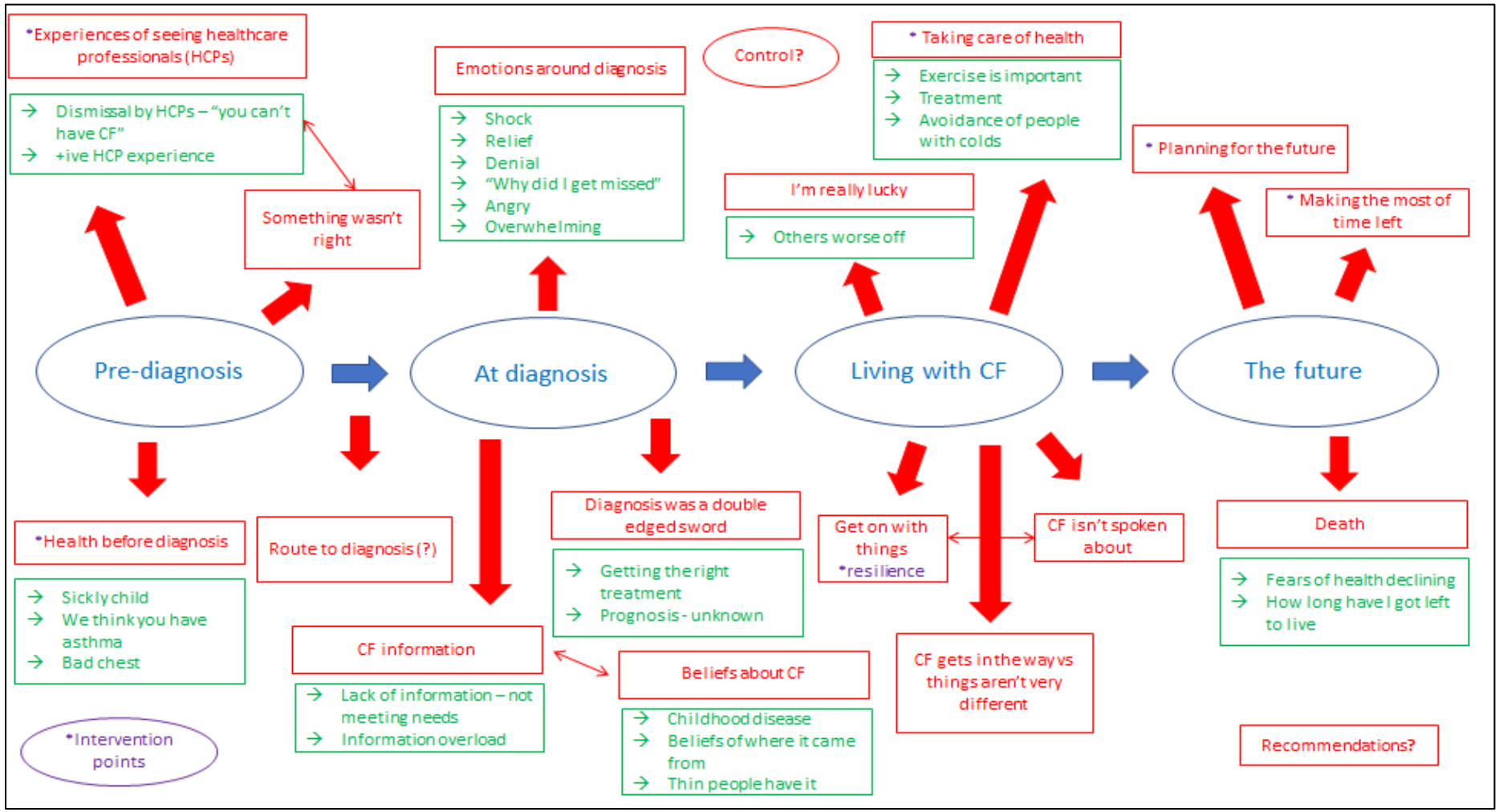
The only thing I would ever say is sometimes what always kind of made me think back to a childhood is when I used to cough sometimes if you didn't get your hand up to your mouth quickly enough and this green phlegm would come out of your mouth so quick now and I'd think it doesn't happen to anybody else when they're coughing why would and that's the only thing that I think as a child growing up I used to get embarrassed about was this cough and the phlegm just being instantly there

Comment [NS52]: No one else coughs up phlegm

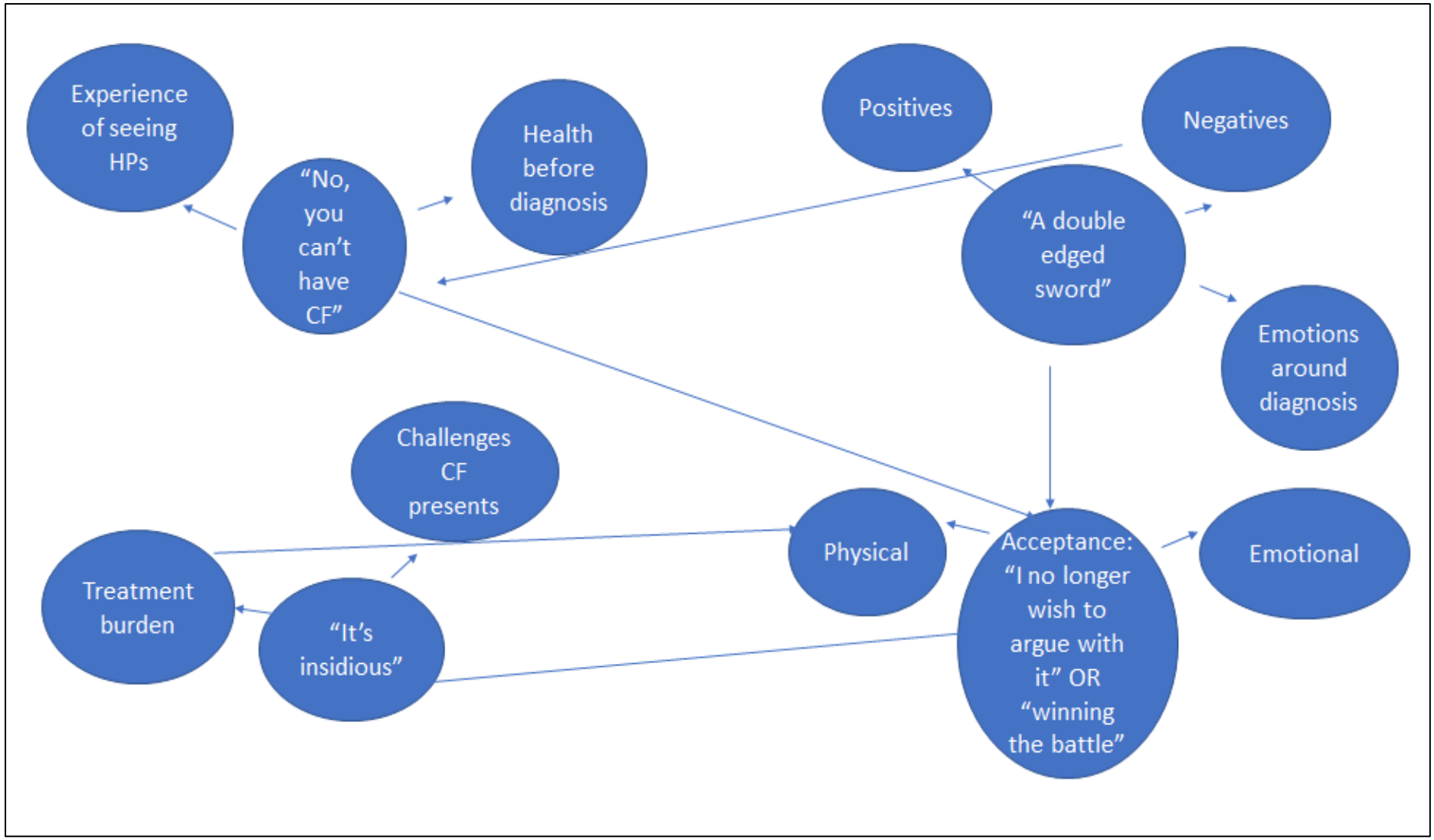
Comment [NS53]: Embarrassed by the phlegm

[End of interview extract]

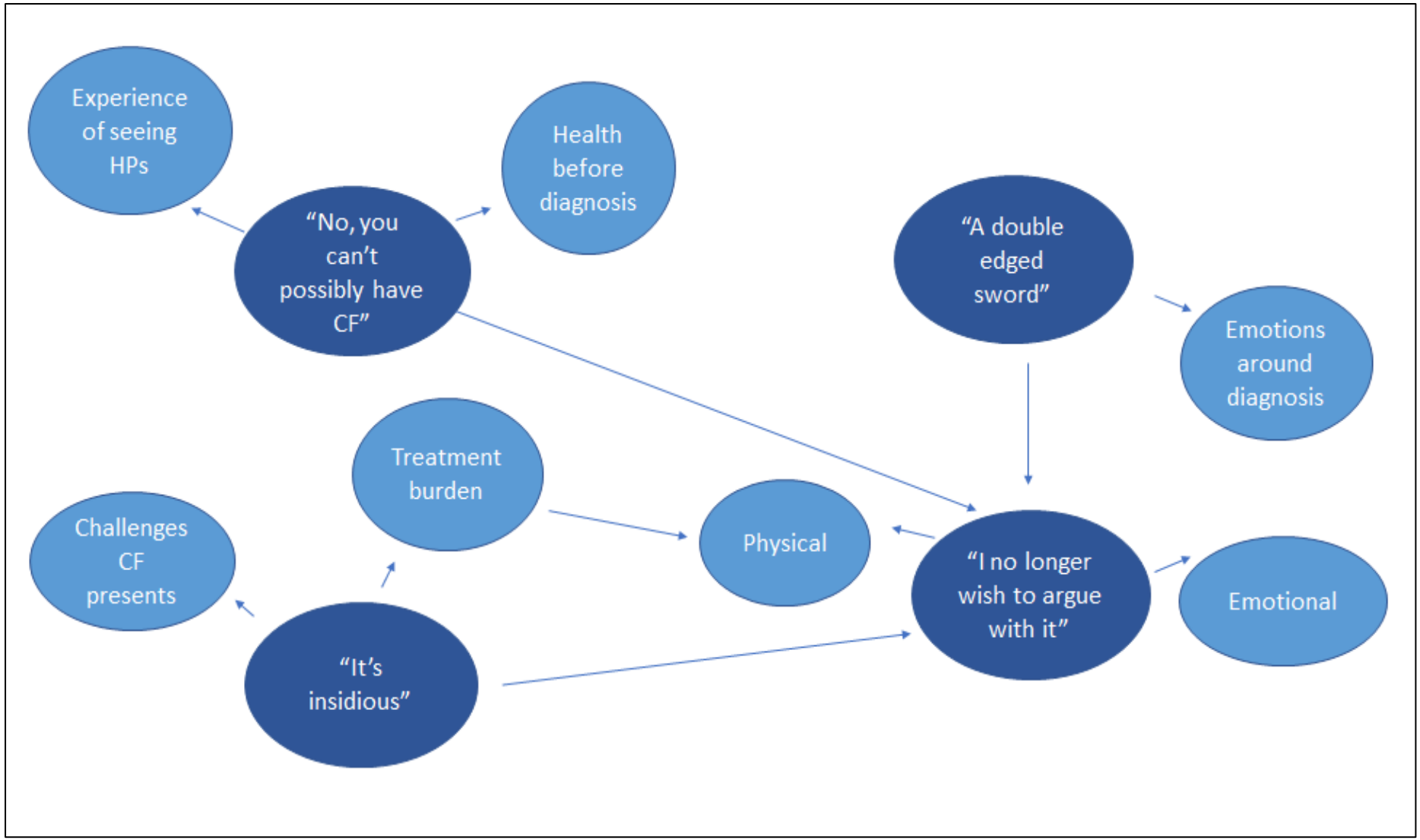
Appendix K – Thematic maps



Initial thematic map



Development of thematic map



Development of thematic map

Appendix L – A 15-point checklist of criteria for good thematic analysis (Braun and Clarke, 2006)

Adapted with permission of the copyright holder Taylor & Francis (www.tandfonline.com)

Process	Criteria	Application of criteria within this research
Transcription	The data have been transcribed to an appropriate level of detail, and the transcripts have been checked against the tapes for "accuracy".	*The interviews were transcribed verbatim and then rechecked against the audio for accuracy.*
Coding	Each data item has been given equal attention in the coding process.	*Each transcript was reviewed systematically through the coding process and equal attention was placed on the data items. The coding process was thorough, inclusive and comprehensive. NVivo was used to code and manage the data allowing for easy accessibility to extracts for each theme. The themes were generated based on ample examples of participant data. Once the themes were created they were checked with each other to avoid duplication and checked back with the original data set to ensure it represented what had been said. Themes were also checked against other themes to ensure that they were distinctive. All of the data set was reviewed to ensure that it fit under the theme for internal coherence and consistency.*
	Themes have not been generated from a few vivid examples (an anecdotal approach), but instead the coding process has been thorough, inclusive and comprehensive.	
	All relevant extracts for all each theme have been collated.	
	Themes have been checked against each other and back to the original data set.	
	Themes are internally coherent, consistent, and distinctive.	
Analysis	Data have been analysed – interpreted, made sense of - rather than just paraphrased or described.	*The results section describes to the reader the journey of adults diagnosed with CF, demonstrating a balance between analytic narrative and participant quotes to illustrate the point being made. Participant quotes included in the results section were matched to the analysis written*
	Analysis and data match each other – the extracts illustrate the analytic claims.	

	Analysis tells a convincing and well-organised story about the data and topic.	*around it. The analysis of data was not just paraphrased or described but rather a higher-level analysis was carried out. This is demonstrated in the discussion in the key finding around 'mismatch' and the observation around 'duality', which has been identified through analysis of the data.*
	A good balance between analytic narrative and illustrative extracts is provided.	
Overall	Enough time has been allocated to complete all phases of the analysis adequately, without rushing a phase or giving it a once-over-lightly.	*Each element of the analysis was given substantial time in which the researcher could analyse and reflect on the data generated.*
Written report	The assumptions about, and specific approach to, thematic analysis are clearly explicated.	*The type of thematic analysis used has been clearly explained and documented within this report demonstrating consistency between the reported method and the analysis carried out. An active position has been taken in the research process in which the themes have been constructed by following an iterative and systematic process. The report avoids using language such as 'the themes emerged from the data', instead referring to the themes as being 'identified'. As stated earlier, the epistemological and ontological position adopted for this research was a critical realist contextualist approach. This approach accepts multiple realities and recognises that there is some 'truth' to produce knowledge to make a difference. The language used in the results and discussion reflects the belief of multiple realities, 'truth' and wider application of the knowledge produced but also recognises that knowledge can only be partially accessed due to individuals' perceptions of reality being subjective, socially influenced and can vary over time.*
	There is a good fit between what you claim you do and what you show you have done – i.e., described method and reported analysis are consistent.	
	The language and concepts used in the report are consistent with the epistemological position of the analysis.	
	The researcher is positioned as *active* in the research process; themes do not just "emerge".	

Appendix M – Eight "big-tent" criteria for excellent qualitative research (Tracy, 2010)

Adapted with permission of the author

Criteria for quality	Description of the criteria	Application of criteria to this study
Worthy topic	The topic is relevant, timely, significant, interesting	*The conceptualisation of this research project formed from clinical practice, therefore demonstrating relevance. There was limited information available for individuals diagnosed with cystic fibrosis in adulthood. It is a growing area for CF departments within the UK and a driver within the NHS to listen to the patient voice in order to make service improvements (Patient Voice South, 2018). Therefore showing that the topic area was significant and timely.*
Rich rigor	The study uses sufficient, abundant, appropriate and complex theoretical constructs, data and time in the field, sample(s), context(s), data collection and analysis processes	*Participant interviews were between just under an hour to three hours in length, demonstrating that enough time was spent gathering interesting and significant data. This was also a testament that participants were keen to share their story. Rigorous data analysis and transparency were demonstrated by providing a description of the data analysis process followed.*
Sincerity	The study is characterised by self-reflexivity about subjective values, biases, and inclinations of the researcher(s), transparency about the methods and challenges	*A research diary was kept documenting the steps of the study as well as recording the researcher's influence and biases throughout the research process.*
Credibility	The research is marked by thick description, concrete detail, explication of tacit (nontextual) knowledge, and showing rather than telling, triangulation or crystallisation, multivocality, member reflections	*Transcripts were discussed with two individuals to get different perspectives on the participants' voice. The results include detailed descriptions of the themes which were illustrated with relevant quotes from the participant. Member reflections were carried out with the participants to seek their feedback on the data.*

Resonance	The research influences, affects, or moves particular readers or a variety of audiences through aesthetic, evocative representations, naturalistic generalisations, transferable findings	*The findings from the study resonate with other chronic health conditions.*
Significant contribution	The research provides a significant contribution: conceptually/theoretically, practically, morally, methodologically, heuristically	*The findings from the research provided recommendations to improve practice whilst also giving a voice to those diagnosed with CF in adulthood. It provided heuristic significance by making suggestions for future research.*
Ethical	The research considers procedural ethics, situational and culturally specific ethics, relational ethics, exiting ethics	*The necessary procedural ethics were carried out for the research project and ethical considerations and consent were considered and reported (see 3.7 Ethical considerations)*
Meaningful coherence	The study achieves what it purports to be about, uses methods and procedures that fit its stated goals, meaningfully interconnects literature, research questions/foci, findings, and interpretations with each other	*Careful consideration was given to the study's research design and method to ensure that it matched the aims and purpose of the project.*

Appendix N – Application of the quality framework for qualitative research (Meyrick, 2006)

Quality framework			Application of framework to this study
Researcher epistemological /theoretical stance		Objective	The researcher clearly stated the epistemological stance adopted for the research project
		Reflective	Being a qualitative researcher requires an active involvement within the research (Willig, 2013). Therefore, the subjective perspective of the research process was acknowledged. A reflective account in the form of a research diary was kept (see Appendix B Research diary extracts and Appendix C Reflective chapter for general reflections). This enabled a continuous review and scrutiny of the researcher's role in the research process. This discouraged any meanings that may have been imposed, therefore promoting the validity of the research (Willig, 2013).
Process	Methods	Aims/objectives clear	The aims and objectives for this research were clearly stated (see 1.9 Research aim and research questions).
		Methods appropriate to research question	Informed by the research aim, the appropriate methods were selected to answer the research question (see 2.0 Methodology and 3.0 Method).
	Sampling	Detail of sampling described	The sampling strategy was clearly described (see 3.1 Recruitment and 3.3 Participants).
		Represents groups	All eligible adults with CF were invited to take part in the study. Of those recruited and interviewed, there was no reason to believe that they were not representative of individuals diagnosed with CF in adulthood.
		Sampling strategy described	
	Data collection	Transparent – change in focus justified	See 2.3 Data collection and 3.2 Procedure for sufficient detail about how the data was collected and recorded.
		Systematic – analytic framework/process described	During the interviews, a semi-structured interview schedule split into areas of interest was used (see 2.6 Interview design).

		Data collection/analysis responsive to data	Detail about how the data was collected was recorded (see 2.3 Data collection and 3.2 Procedure).
Analysis		Transparent pathway data to conclusion	The analysis process was transparent and well documented (see 3.5 Data analysis).
		Systematic pathway described	The analysis process was clearly described (see 3.5 Data analysis)
		Complete, all cases, theories + deviance examined	The data derived from all of the interviews were included in the analysis. Deviant and contradicting statements were included and discussed in the analysis.
		Triangulation of method, source, sample, research	
		Internal validation Reflexivity relationship of researcher to data Qualitative audit trail data to conclusions	The researcher detailed the steps taken from data to the conclusion, provided information on the interview technique and reflected on the process.
		External validation Multiple coding	Two independent reviewers were asked to review and make notes on one transcript each. These notes were reviewed and discussed with the researcher to inform the coding of the data.
Results/conclusions		Findings grounded in data (illustrated)	The findings of the research are grounded by the data. This has been demonstrated by using a balance of participant quotes and narrative description within the analysis (see 4.0 Results)
		Respondent validation	Participants were given a summary of the results and asked for any comments or feedback.
		Applicability	
		Generalizable representative	The researcher has no reason to believe that the findings from the research were not representative of the experiences of others diagnosed with CF in adulthood.

Appendix O – Ethical approval

NHS
Health Research Authority

South Central - Hampshire B Research Ethics Committee
Level 3 Block B
Whitefriars
Lewins Mead
Bristol
BS1 2NT

27 October 2015

Address and information in this letter redacted for anonymity reasons

Dear Miss Sharma

Study title:	**Cystic Fibrosis (CF) diagnosis in adulthood: patients' views on their experiences and support needs.**
REC reference:	**15/SC/0567**
IRAS project ID:	**189640**

Thank you for your letter of 23rd October 2015. I can confirm the REC has received the documents listed below and that these comply with the approval conditions detailed in our letter dated 10 October 2015

Documents received

The documents received were as follows:

Document	*Version*	*Date*
IRAS Checklist XML [Checklist_23102015]		23 October 2015
Other [Protocol - Version 2.0 12.10.2015]	2.0	12 October 2015
Participant consent form [Participant Consent Form - Version 2.0 12.10.2015]	2.0	
Participant information sheet (PIS) [Participant Information Sheet - Version 2.0 12.10.2015]	2.0	12 October 2015

Approved documents

The final list of approved documentation for the study is therefore as follows:

Document	*Version*	*Date*
Copies of advertisement materials for research participants [Recruitment Poster - Version 1.3 28.08.2015]	1.3	28 August 2015
Copies of advertisement materials for research participants [Social Media Adverts - Version 1.1 10.05.2015]	1.1	10 May 2015
Covering letter on headed paper [15-ES-0122 Unfavourable Opinion Covering Letter Researcher Comments 27-08-15]	1	27 August 2015
Evidence of Sponsor insurance or indemnity (non NHS Sponsors only) [Indemnity letters UMAL EL PL 15-16]	1	20 July 2015
Interview schedules or topic guides for participants [Participant Interview Schedule - Version 1.3 22.08.2015]	1.3	22 August 2015
IRAS Checklist XML [Checklist_23102015]		23 October 2015
Letter from sponsor [NRES Committee NRES Committees [redacted] B]	1	01 September 2015
Letters of invitation to participant [Participant Invitation Letter - Version 1.5 28.08.2015]	1.5	28 August 2015
Letters of invitation to participant [Participant Follow Up Letter - Version 1.1 28.08.2015]	1.1	28 August 2015
Other [Summary CV for supervisor (student research) - [redacted] CV]	1	05 May 2015
Other [Evidence of Sponsor insurance or indemnity (non-NHS Sponsors only) - Indemnity letters UMAL PI 15-16]	1	20 July 2015
Other [Introduction to Good Clinical Practice - Certificate of Attendance 11.05.2015]	1	11 May 2015
Other [Telephone/Video Call Interview Consent Script - Version 1 03.05.2015]	1	03 May 2015
Other [15-ES-0122 Unfavourable Opin Let 18-08-15]	1	18 August 2015
Other [Protocol - Version 2.0 12.10.2015]	2.0	12 October 2015
Participant consent form [Participant Consent Form - Version 2.0 12.10.2015]	2.0	
Participant information sheet (PIS) [Participant Information Sheet - Version 2.0 12.10.2015]	2.0	12 October 2015
REC Application Form [REC_Form_01092015]		01 September 2015
Research protocol or project proposal [Protocol - Version 1.4 27.08.2015]	1.4	27 August 2015
Summary CV for Chief Investigator (CI) [Nisha Sharma Research CV 28 Aug 2015]	1	28 August 2015
Summary CV for supervisor (student research) [Diana Harcourt CV 01 June 2015]	1	01 June 2015

You should ensure that the sponsor has a copy of the final documentation for the study. It is the sponsor's responsibility to ensure that the documentation is made available to R&D offices at all participating sites.

15/SC/0567	Please quote this number on all correspondence

Yours sincerely

[redacted]

Mrs Siobhan Bawn
REC Manager

E-mail: nrescommittee.southcentral-hampshireb@nhs.net

Copy to: Mrs Leigh Taylor
[redacted]

NHS
Health Research Authority

South Central - Hampshire B Research Ethics Committee
Level 3 Block B
Whitefriars
Lewins Mead
Bristol
BS1 2NT

Telephone: 01173421386

10 October 2015

Address and information in this letter redacted for anonymity reasons

Dear Miss Sharma

Study title:	**Cystic Fibrosis (CF) diagnosis in adulthood: patients' views on their experiences and support needs.**
REC reference:	**15/SC/0567**
IRAS project ID:	**189640**

The Research Ethics Committee reviewed the above application at the meeting held on 30 September 2015. Thank you for attending to discuss the application.

We plan to publish your research summary wording for the above study on the HRA website, together with your contact details. Publication will be no earlier than three months from the date of this favourable opinion letter. The expectation is that this information will be published for all studies that receive an ethical opinion but should you wish to provide a substitute contact point, wish to make a request to defer, or require further information, please contact the REC Manager Mr Rajat Khullar, nrescommittee.southcentral-hampshireb@nhs.net. Under very limited circumstances (e.g. for student research which has received an unfavourable opinion), it may be possible to grant an exemption to the publication of the study.

Ethical opinion

The members of the Committee present gave a favourable ethical opinion of the above research on the basis described in the application form, protocol and supporting documentation, subject to the conditions specified below. .

Conditions of the favourable opinion

The favourable opinion is subject to the following conditions being met prior to the start of the study.

1. *PIS page 1, under the heading, "Why have I been invited..", the word "all " should be removed from "We are inviting all patients who have been diagnosed with cystic fibrosis..". Not all patients will be invited as there are inclusion/exclusion criteria for the study.*

2. *PIS page 1, under the heading, "Do I have to take part?", the sentence, "Any decision you make…..medical care you receive" should be revised to "If you decide not to take part in this study it will not affect your routine medical care in any way".*

3. *PIS page 2, under the heading, "What are the possible benefits of taking part?", the sentence, "However, the information you provide will help us to…" should be revised to, "However, the information you provide may help us to…".*

4. *PIS page 2, under the heading, "What are the disadvantages of taking part", the sentence, "If you do, you can refuse to answer..." should be revised to, " If you do, you can choose not to answer..".*

5. *Limitations of confidentiality should be clearly explained in the PIS. The standard paragraph on confidentiality should be included. Details regarding standard section in the PIS can be found on the HRA website.*

6. *Optional sections of the Consent Form should have a Yes [ ] No [ ] initial boxes for the participants to select.*

7. *It is recommended that the prize draw should be limited only to participants in this revised version of the study so as not to include the health professionals who are being dealt with independently elsewhere.*

You should notify the REC in writing once all conditions have been met (except for site approvals from host organisations) and provide copies of any revised documentation with updated version numbers. The REC will acknowledge receipt and provide a final list of the approved documentation for the study, which can be made available to host organisations to facilitate their permission for the study. Failure to provide the final versions to the REC may cause delay in obtaining permissions.

Management permission or approval must be obtained from each host organisation prior to the start of the study at the site concerned.

Management permission ("R&D approval") should be sought from all NHS organisations involved in the study in accordance with NHS research governance arrangements.

Guidance on applying for NHS permission for research is available in the Integrated Research Application System or at http://www.rdforum.nhs.uk.

Where a NHS organisation's role in the study is limited to identifying and referring potential participants to research sites ("participant identification centre"), guidance should be sought from the R&D office on the information it requires to give permission for this activity.

For non-NHS sites, site management permission should be obtained in accordance with the procedures of the relevant host organisation.

Sponsors are not required to notify the Committee of approvals from host organisations.

Registration of Clinical Trials

All clinical trials (defined as the first four categories on the IRAS filter page) must be registered on a publically accessible database. This should be before the first participant is recruited but no later than 6 weeks after recruitment of the first participant.

There is no requirement to separately notify the REC but you should do so at the earliest opportunity e.g. when submitting an amendment. We will audit the registration details as part of the annual progress reporting process.

To ensure transparency in research, we strongly recommend that all research is registered but for non-clinical trials this is not currently mandatory.

If a sponsor wishes to request a deferral for study registration within the required timeframe, they should contact hra.studyregistration@nhs.net. The expectation is that all clinical trials will be registered, however, in exceptional circumstances non registration may be permissible with prior agreement from the HRA. Guidance on where to register is provided on the HRA website.

It is the responsibility of the sponsor to ensure that all the conditions are complied with before the start of the study or its initiation at a particular site (as applicable).

Ethical review of research sites

NHS Sites

The favourable opinion applies to all NHS sites taking part in the study taking part in the study, subject to management permission being obtained from the NHS/HSC R&D office prior to the start of the study (see "Conditions of the favourable opinion" below).

Summary of discussion at the meeting

Favourable risk benefit ratio; anticipated benefit/risks for research participants (present and future)

The Committee expressed concern with regard the possibility of using telephone interviews; telephone conversations of around 60 minutes could be quite tedious and onerous for the participants. There are also concerns about any of the participants being contacted by telephone or Skype and living alone. During the conversation if they get distressed, then how will that be managed and how would any immediate support be provided. Dr [redacted] explained that as a part of regional cystic fibrosis service she works as a clinical psychologist and a lot of therapeutic work is carried out over the phone. This study does not actually involve a questionnaire but a conversational interview. She further explained that there are a number of aspects that could help with this study. Firstly they work as psychologists and regularly work with people who are distressed. Secondly the potential participants will be screened on the day to ensure if they are suitable to be included in the study. She explained that participants will be able stop the interview at any point if they feel uneasy. Dr [redacted] assured the Committee that she has confidence in your abilities for dealing in such a situation with distressed patients, and in addition there will be back up support available in the form of Dr [redacted] herself, as well as two other colleagues from the cystic fibrosis team. She further added that the study only involves academic interviews and not therapeutic sessions and therefore they do not anticipate participants getting very distressed. If the participants are self-selecting to take part in the study given the information then they are more likely know what to expect and successfully finish the interviews. She added that Skype

video calling will have the advantage of allowing the researcher to see the participants and pick up any signs of distress in time and to stop the interviews if required.

The Committee asked Dr [redacted] in her previous experience whether most research interviews were face to face and if any were done using Skype. Dr [redacted] explained that in most of the research people choose to do face to face interview only. However, in therapy a number of people choose to do telephone interviews. The Skype option is only included as a back-up for those who are not able to attend. She however mentioned that they expect most of the interviews to take place face to face. She added that the target population in this area is fairly young and they are quite likely to be comfortable using Skype. However,if the participants are not comfortable with Skype then they will not chose it and they will be able to have face to face interviews. It is just an extra option instead of excluding those who are not able to travel.

The Chair asked Dr [redacted] how she would feel if the Committee recommended that the study be restricted only to face to face interviews. Dr [redacted] explained almost all the interviews are expected to take place face to face, and that interviews by telephone or Skype were only added to make the study more equitable, as explained above. It is quite a young population with a wide geographical spread and some of them may have disabilities, and therefore some of the participants might benefit from this. The Committee was satisfied with the explanation provided.

Other general comments

The Committee expressed some concerns with regard to the costs of the study. The Committee noted that even though the costs are not significant and are clearly laid out, all the expenses are being borne by the student. There are some other costs like travel expenses that do not seen to have been included in the study costs, and these could amount to a substantial expenditure. The Committee asked Dr [redacted] if any funding can be arranged to cover these costs or if funding cannot be arranged then could a guarantor be identified who could guarantee to cover the costs if the student is not able to cover these. Dr [redacted] replied that it is quite difficult to get any grant or funding for the PhD studies, and especially for psychological studies. The students do get some funding for clinical psychology studies however the health psychology studies are mostly self-funded. She explained that with regards to the travel costs, the study has been designed to fit around the clinical appointments of the participants and therefore the travel expenses and any costs to the participants would be minimal. She added that they will try and do most of the appointments either when the participants attend the hospitals or through Skype. Dr [redacted] agreed to mention in the application that there will not be much travel expense and therefore it will not be a major concern. The Committee asked you if when you joined this degree course you were alerted to the fact that there could potentially be some extra expenditures imposed for successful completion of the doctorate. You replied that when you joined the course you expected that in terms of delivering the project there would be some funding needed, but that you would have to raise this yourself, as this is a self-funded degree programme.

The Committee suggested that the [redacted] Medical Research Charity is a small charity that may be able to fund the study. Dr [redacted] thanked the Committee for the suggestion. The Committee offered to provide help in the form of a statement of ethical approval of the study if it was required by the student for raising funding to meet all expenses and successful execution of the study. You and Dr [redacted] thanked the Committee for the support.

Approved documents

The documents reviewed and approved at the meeting were:

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Copies of advertisement materials for research participants [Recruitment Poster - Version 1.3 28.08.2015]	1.3	28 August 2015
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Other [Telephone/Video Call Interview Consent Script - Version 1 03.05.2015]	1	03 May 2015
Other [15-ES-0122 Unfavourable Opin Let 18-08-15]	1	18 August 2015
Other [Research protocol or project proposal Protocol - Version 1.4 27.08.2015 - Tracked Changes]	1.4	27 August 2015
Participant consent form [Participant Consent Form - Version 1.5 27.08.2015]	1.5	27 August 2015
Participant consent form [Participant Consent Form - Version 1.5 27.08.2015 - Tracked Changes]	1.5	27 August 2015
Participant information sheet (PIS) [Participant Information Sheet - Version 1.5 27.08.2015]	1.5	28 August 2015
Participant information sheet (PIS) [Participant Information Sheet - Version 1.5 27.08.2015 - Tracked Changes]	1.5	27 August 2015
REC Application Form [REC_Form_01092015]		01 September 2015
Research protocol or project proposal [Protocol - Version 1.4 27.08.2015]	1.4	27 August 2015
Summary CV for Chief Investigator (CI) [Nisha Sharma Research CV 28 Aug 2015]	1	28 August 2015
Summary CV for supervisor (student research) [Diana Harcourt CV 01 June 2015]	1	01 June 2015

Membership of the Committee

The members of the Ethics Committee who were present at the meeting are listed on the attached sheet.

There were no declarations of interest

The Committee is constituted in accordance with the Governance Arrangements for Research Ethics Committees and complies fully with the Standard Operating Procedures for Research Ethics Committees in the UK.

After ethical review

Reporting requirements

The attached document "After ethical review – guidance for researchers" gives detailed guidance on reporting requirements for studies with a favourable opinion, including:

- Notifying substantial amendments
- Adding new sites and investigators
- Notification of serious breaches of the protocol
- Progress and safety reports
- Notifying the end of the study

The HRA website also provides guidance on these topics, which is updated in the light of changes in reporting requirements or procedures.

User Feedback

The Health Research Authority is continually striving to provide a high quality service to all applicants and sponsors. You are invited to give your view of the service you have received and the application procedure. If you wish to make your views known please use the feedback form available on the HRA website: http://www.hra.nhs.uk/about-the-hra/governance/quality-assurance/

HRA Training

We are pleased to welcome researchers and R&D staff at our training days – see details at http://www.hra.nhs.uk/hra-training/

15/SC/0567	**Please quote this number on all correspondence**

With the Committee's best wishes for the success of this project.

Yours sincerely

[redacted]

pp Professor Ron King
Chair

E-mail: nrescommittee.southcentral-hampshireb@nhs.net

Enclosures: *List of names and professions of members who were present at the meeting and those who submitted written comments*

"After ethical review – guidance for researchers"

Copy to: *Mrs Leigh Taylor*
[redacted], [redacted]
[redacted]

South Central - Hampshire B Research Ethics Committee

Attendance at Committee meeting on 30 September 2015

Committee Members:

Name	*Profession*	*Present*	*Notes*
Ms [redacted]	Patient and Public Involvement Officer	Yes	
Mr [redacted]	Consultant Urological Surgeon	Yes	
Mrs [redacted]	Pharmacist	Yes	
Ms [redacted]	Speech and Language Therapist	Yes	
Mr [redacted]	Senior Lecturer in Radiography	No	
Ms [redacted]	Contract Manager, Research and Innovation Services	Yes	
Ms [redacted]	Former Civil Servant	Yes	
Mrs [redacted]	Acute Oncology Clinical Nurse Specialist	No	
Professor [redacted] Chair)	Mathematician (Retired)	Yes	
Mr [redacted]	Chartered Engineer (Retired)	Yes	
Dr [redacted]	Course Leader, M.Sc. Clinical Exercise Science	No	
Dr [redacted]	Consultant Psychiatrist	No	

Also in attendance:

Name	*Position (or reason for attending)*
Mr [redacted]	REC Manager

University of the West of England
BRISTOL

Faculty of Health & Applied Sciences
Glenside Campus
Blackberry Hill
Stapleton
Bristol BS16 1DD

Tel: 0117 328 1170

Our ref: JW/lt

27th November 2015

Address and information in this letter redacted for anonymity reasons

Dear Nisha

Application number: HAS/15/11/041
Application title: Cystic Fibrosis (CF) diagnosis in adulthood: patients' views on their experiences and support needs.
NHS Application Number: 15/SC/0567

Your NHS Ethics application and approval conditions have been considered by the Faculty Research Ethics Committee on behalf of the University. It has been given ethical approval to proceed with the following conditions:

- You comply with the conditions of the NHS Ethics approval.
- You notify the Faculty Research Ethics Committee of any further correspondence with the NHS Ethics Committee.
- You must notify the Faculty Research Ethics Committee in advance if you wish to make any significant amendments to the original application.
- If you have to terminate your research before completion, please inform the Faculty Research Ethics Committee within 14 days, indicating the reasons.
- Please notify the Faculty Research Ethics Committee if there are any serious events or developments in the research that have an ethical dimension.
- Any changes to the study protocol, which have an ethical dimension, will need to be approved by the Faculty Research Ethics Committee. You should send details of any such amendments to the committee with an explanation of the reason for the proposed changes. Any changes approved by an external research ethics committee must also be communicated to the relevant UWE committee.

- Please note that any information sheets and consent forms should have the UWE logo. Further guidance is available on the web: http://www1.uwe.ac.uk/aboutus/departmentsandservices/professionalservices/marketingandcommunications/resources.aspx
- Please note that the University Research Ethics Committee (UREC) is required to monitor and audit the ethical conduct of research involving human participants, data and tissue conducted by academic staff, students and researchers. Your project may be selected for audit from the research projects submitted to and approved by the UREC and its committees.

Please note that your study should not commence at any NHS site until you have obtained final management approval from the R&D department for the relevant NHS care organisation. A copy of the approval letter(s) must be forwarded to Leigh Taylor in line with Research Governance requirements.

We wish you well with your research.

Yours sincerely

Dr Julie Woodley
Chair
Faculty Research Ethics Committee

c.c. *Di Harcourt*

Appendix P – University sponsorship letter

University of the West of England
BRISTOL

Faculty of Health & Applied Sciences
Glenside Campus
Blackberry Hill
Stapleton
Bristol BS16 1DD

Tel: 0117 328 1170

Our ref: JMA/CF

01 September 2015

Address and information in this letter redacted for anonymity reasons

Dear Sir/Madam

Study title: Cystic Fibrosis (CF) diagnosis in adulthood: patients' views on their experiences and support needs
Principal investigator: Nisha Sharma
REC Reference: 15/SC/0567

I am writing to confirm that the University of the West of England, Bristol ("UWE") has agreed to act as Research Sponsor in accordance with the Department of Health Research Governance Framework (2001) for the above research. UWE's acceptance of Research sponsorship is subject to ethics approval having been obtained.

UWE has made the following insurance arrangements for employees, and for students working under the supervision of a UWE employee, and where the project is included on an authorised UWE research register.

UWE has insurance cover for clinical trials up to £5m in the aggregate which includes cover for non-negligent harm. This cover is provided only when UWE (via Research, Business and Innovation) has approved projects with our insurers and they are then listed on our clinical trials register.

For research which is not deemed a clinical trial (i.e. not on UWE's clinical trials register):

- UWE's Professional Indemnity policy provides insurance cover for indemnity against legal liability for damages and claimant's costs and expenses arising out of any act, neglect, error or omission.
- UWE's Employers Liability Insurance is in place to protect UWE's employees if they are harmed whilst engaged on UWE business, should UWE be held legally liable.
- UWE's Public Liability insurance policy covers legal liability for third party personal injury, death, disease or illness to any person or loss or damage to third party property.

Details of the Employers/Public and Professional Indemnity policy covers are attached.

Yours faithfully

Prof Jennifer Ames
Associate Dean (Research and Innovation)

Encl