Exploring the role of music therapy in the lives of children with Neuronal Ceroid Lipofuscinosis (Batten Disease)

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Abstract

Background: Batten disease is a rare neurodegenerative condition that affects approximately 150 – 200 children and young people in the UK. Children experience gradual loss of skill over time, and most do not live past their adolescent years. Present research focuses upon clinical trials in enzyme replacement therapy, however, leading research in the field recognises the need for improved education strategies. Although under-researched, music has been recognised as a key support mechanism for individuals affected by Batten disease. Research advances in neuroscience and music therapy suggest that music interventions are successful for neurodegenerative conditions. As yet, research has not investigated the impact of music or music therapy for childhood conditions.

Aim: The project aims to explore the role of music therapy for children affected by Batten disease, focusing on how children respond to long-term music therapy, and how this can be measured. Secondly, the project aims to explore how elements of music therapy can be transferred into educational environments through a novel music education program, 'Music Speaks'. Finally, the project aims to capture experiences of professionals who support affected children in educational contexts and gather their experiences of delivering the Music Speaks program.

Methods: The project involved a three-phase mixed methods design involving observations over a four-year period; development and validation of a prototype music therapy assessment measure; and the creation and trial of a music education program for schools, Music Speaks. Semi-structured interviews captured the experiences of professionals, challenges and systemic structures surrounding support and education for children affected by Batten disease.

Findings: Psychometric measures confirmed strong face validity and inter-rater reliability of a new music therapy outcome measure. It was observed that music therapy facilitated sustained and, in some cases, improved functioning over time. The Music Speaks program created short-term improvements in expressive language. Professionals observed how music helped preserve skills, where in the absence of music, the skills of children with Batten disease might otherwise deteriorate. Interview data outlined how Music Speaks could enhance children's self-expression and communication and complement pre-emptive teaching approaches to preserve the child's independence in the future.

Conclusion: The project's findings create a dialogue between the experiences of those in education and the observed impact of music, supporting the potential use of music therapy interventions for children affected by Batten disease. The research provides preliminary evidence to support the development and review of educational guidance and policy to incorporate music therapy interventions for childhood neurodegenerative diseases. Further research could optimise music interventions considering implementation, adaptability, transferability, and feasibility in other contexts, to support the use of music in wider care pathways for children with Batten disease.

Table of Contents

List of Figures

Acronyms and Abbreviations

BAMT	British Association of Music Therapy
BDFA	Batten Disease Family Association
CLN	Ceroid Lipofuscinosis Neuronal
CMT	Chiltern Music Therapy
CMTOM	Chiltern Music Therapy Outcome Measure
COVID-19	Corona Virus Disease
DBS	Disclosure and Barring Service
DNA	Deoxyribonucleic Acid
EDO	Education Development and Observation Tool
EHCP	Education Health and Care Plan
EthOS	Ethnographic Observation System
HCPC	Health and Care Professions Council
JNCL	Juvenile CLN3 Batten Disease
LSA	Learning Support Assistant
LSD	Lysomal Storage Disease
MATADOC	Music Therapy Assessment Tool for Awareness in Disorders of Consciousness
fMRI	Functional Magnetic Resonance Imaging
MRC	Medical Research Council
MiDAS	Music in Dementia Assessment Scale
MIND	Music in Neurodegeneration
MUSTIM	Music Speech Stimulation
NCL	Neuronal Ceroid Lipofuscinoses
NICE	National Institution of Care and Excellence
NINDS	National Institute of Neurodegenerative Disorders and Stroke
NMT	Neurologic Music Therapy
NPI	Neuropsychological Inventory for dementia
NRP	Neurological Restorative Program
PE	Physical Education
PMLD	Profound and Multiple Learning Disability
RCT	Randomised Control Trial
RSC	Rhythmic Speech Cueing
SATS	Standard Assessment Tests
SD	Standard Deviation
SEN	Special Educational Needs
SOI	Sounds of Intent
TA	Teaching Assistant
TOMs	Therapy Outcome Measure
UBDRS	Unified Batten Disease Rating Scale
UK	United Kingdom
US	United States
VI	Visual Impairment

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Lastly, I want to pay dedication to the children affected by Batten disease in the UK, US and around the world. It has been a privilege to get to know you all and witness your astounding musical selves and identities. Your songs and the memories from this research will be carried with me, especially those children who are no longer with us.

It is for you that I dedicate this research.

Declaration

I declare that the research contained in this thesis, unless otherwise formally indicated within this text, is the original work of the author. The thesis has not been previously submitted to this or any other university for a degree, and does not incorporate any material already submitted for a degree.

Signed:

Dated: 23rd June 2022

Prelude

"Where words leave off, music begins."
— Heinrich Heine

This is the story of Emma.

A developing ''typical' 12-year-old girl who has learnt to walk, talk, think and express herself competently. Suddenly, and without warning, her family notice she is bumping into things and becoming confused. In less than two years, she is blind and has little speech, understanding or voluntary movement left. By the time she is diagnosed with 'childhood dementia' (also known as Batten disease), Emma's parents have watched their daughter deteriorate before their eyes to a devastating degree. The family are living on borrowed time, not knowing when she will eventually pass away.

But there is another story.

A story of opportunity, hope and existence. Whilst Emma is visibly losing her ability to speak, when she listens to familiar music, her speech is transformed. Emma can recite and sing words perfectly to a song that are in tune and in time with the music. A window of clarity emerges. The family have fleeting moments where she is back with them, cognitively present and they hear her voice again.

Emma's story is just one of many that illustrate the power of music.

When all else is lost, music still resides.
Perhaps we do not think of music as a medicine, but it has the power to 'find a way in'.

This is the story of a research journey that endeavours to unlock the potential of music for children with dementia.

Chapter 1: Introduction

"Music gives a soul to the universe, wings to the mind, flight to the imagination and life to everything"
— Plato

1.1. Rationale

Dementia in childhood (or Batten disease) is used to describe a group of rare neurodegenerative diseases that are usually diagnosed in childhood (Institute of Neurological Disorders and Stroke [NINDS], 2020). Typically, children affected experience vision loss, which is followed by a gradual deterioration of communication skills, memory loss, cognitive confusion and physical decline (Mole & Cotman, 2015). Generally, children with the disease have a shortened life expectancy; most individuals die in childhood, whilst others live later into their teenage and early adult years (NINDS, 2020). There is currently no confirmed curative treatment.

As a researcher and clinician, my experiences are the reason I decided to pursue doctoral research training, which offered an opportunity to help children with Batten disease. Firstly, my clinical experiences as a Music Therapist in children's palliative care settings was where I first witnessed the impact of music, where I saw music bring families of very sick children together. Music appeared to elicit positive responses, even just a glimpse of a smile, in devastating circumstances. In line with music therapy research in the palliative care sector (Ibberson, 1996; Pavlicevic, 1999), music therapy in this context provided a space for children to express themselves through the vehicle of music, regardless of ability, age or diagnosis.

My later work in the field of brain injury, as a Neurologic Music Therapist, provided the context within which my experiences were expanded and challenged. In my neurological work, music appeared to create meaningful and functional change to patients' lives. In situations where a patient's skills had been lost due to a brain injury or neurological impairment, music could apparently repair and compensate. Research within the field seemed to show that music could be the catalyst to help children to rehabilitate their speech, movement and cognitive skills (Bringas et al., 2015). When I became a researcher with the University of Roehampton I noticed that, whilst music was being used in everyday contexts to help children with Batten disease feel better, some children who could not speak maintained the ability to sing. Opportune moments were presented whereby children who were otherwise 'locked-in', were suddenly present and engaged in music (Ockelford, Atkinson, & Herman, 2019; von Tetzchner, Elmerskog, Tøssebro, & Rokne, 2019). The story of 'Emma' was taken from a reflective passage I wrote at the outset of my time as a researcher with the University of Roehampton (Ockelford et al., 2019).

Throughout my journey in clinical practice and research, I have recognised the value of reflection, critical analysis and a desire to dive deeper into how, where and why knowledge exists. From a critical realist position (Bhaskar & Danermark, 2006), I understood that Emma's story had influencing social factors, perspectives, experiences, underlying mechanisms contributing to her version of reality, and it was crucial to uncover these. Forging a dialogue between my clinical experience and research practice, these experiences drove my desire to understand more about the importance of music for children affected by Batten disease; understanding how, where and why music had such an influential impact on these children when all else was seemingly lost. This project, therefore, aims to explore the influence of music for children with Batten disease. Framed in the context of genetics, diagnosis, interventions and prevalence, this thesis begins by defining and explaining Batten disease. This chapter then briefly outlines the care and education pathway, and experiences of children, relatives and professionals affected by this disease, expanding upon the research rationale. It finally defines how this project seeks to deepen understanding in this area of research.

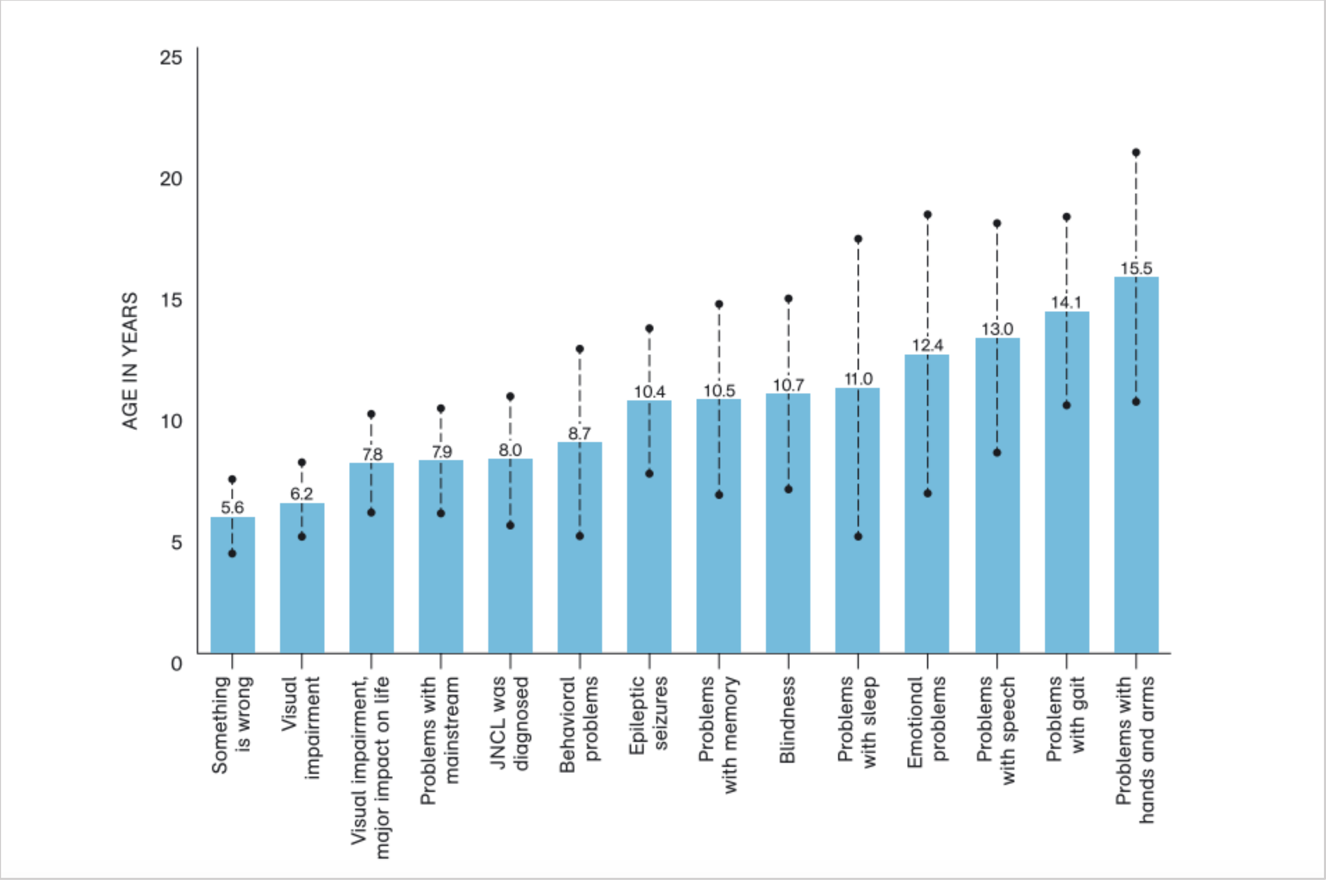
1.2. What is Batten Disease?

Batten disease is the most common form of Neuronal Ceroid Lipofuscinoses (NCLs), which belong to a family of lysosomal storage diseases (LSDs) and is caused by a genetic mutation (Ragbeer et al., 2016). Gene mutation is a regular process that happens in the human body and is often caused by an alteration in the DNA sequence that creates the body's genes. Many mutations go unnoticed, and in some cases, can be beneficial in adapting and introducing diversity to humans, however, some mutations are catastrophic (NINDS, 2020). The genetic mutation present in Batten disease affects a cell called the lysosome. The lysosome ordinarily acts as the 'recycle bin' in a human cell, which is crucial in facilitating the breakdown of waste proteins into smaller components to be discarded or recycled (Mole & Cotman, 2015). In Batten disease, the functioning of the lysosome is impaired, and consequently, there is an eventual build-up of protein that causes significant repeated damage to the delicate neurological system (NINDS, 2020). Although NCL diseases are rare, childhood-onset variants are the most common neurodegenerative disorders in children (Mole & Cotman, 2015). With the inevitable increase of damaged cells over time, individuals with Batten disease suffer continual progressive neurological deterioration and eventual impairment, and Batten disease results in early death in childhood or early adulthood (Adams & Mink, 2013; Mole & Cotman, 2015).

1.2.1. Clinical Presentation

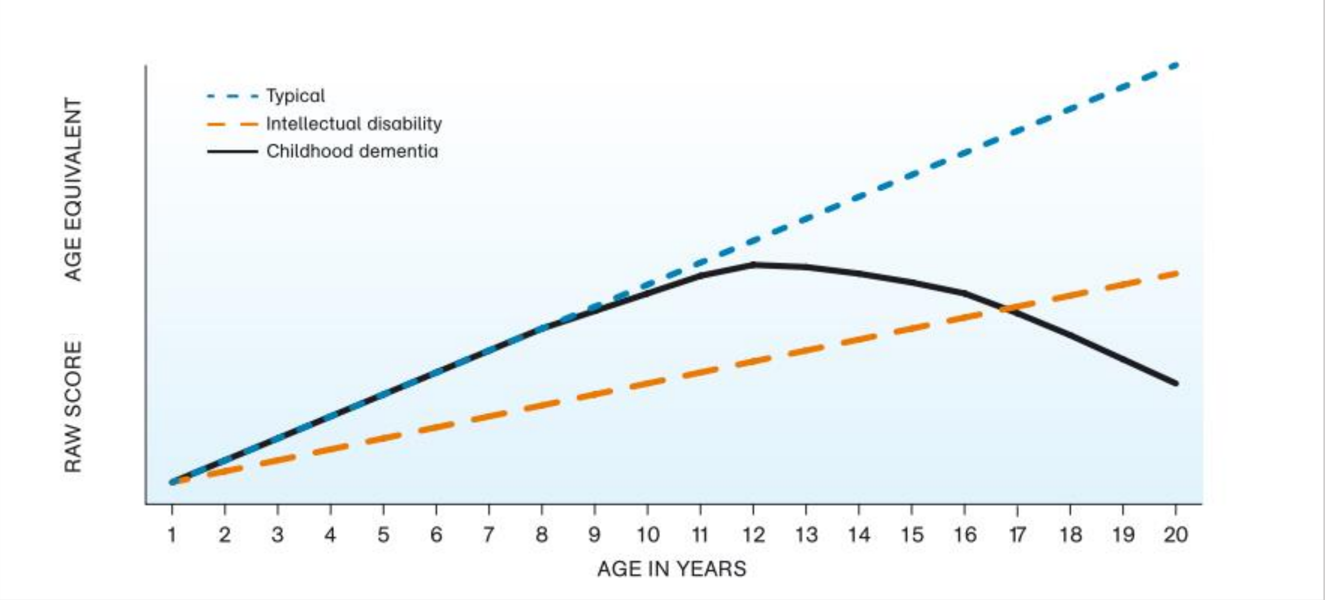
The journey for a child with Batten disease is entirely unique. In most cases, children present as neurotypically developing until parents start to notice that something is wrong (von Tetzchner et al., 2019). For sufferers of Batten disease and other NCLs, accounts suggest that symptoms can either appear suddenly and without warning, or the disease can progress slowly and the child appears to be in a state of relative stability (Bills, Johnston, Wilhelm, & Graham, 1998). Children with the disease can exhibit similar characteristics to adults with other neurodegenerative conditions such as Parkinson's or Alzheimer's disease (Mole & Cotman, 2015). Symptoms typically include seizures, visual impairment, personality or behaviour changes; and eventually a decline in speech, communication, cognitive and physical skills (Bills et al., 1998). Although children with the diagnosis display similar traits, these can all progress at very different rates, and children with Batten disease present as a heterogeneous population. Figure 1.1 visually represents the onset of symptoms experienced by children with a diagnosis of Juvenile CLN3 Batten disease (JNCL), which is the variant usually diagnosed in early teenage years. As can be seen in this figure, the onset of symptoms for children follows a steady trajectory, however the variance in age of onset is considerable. For children, the disease is unpredictable from one person to the next and the path of deterioration differs between children, representing the diverse nature of this population.

Figure 1.1: Parent and staff reported data on the average age (and standard deviations) of symptoms for children and young people with Juvenile Batten disease (cited in von Tetzchner, Elmerskog, Tøssebro, & Rokne, 2019:27)



For children with JNCL, as seen in Figure 1.1, symptoms of visual impairment and confusion can occur later in childhood after some or almost all knowledge acquisition has been established. Those diagnosed with JNCL mostly experience a gradual deterioration of skill into early adult years, rather than a static, unchanging impairment, thus making this condition different from childhood learning disabilities or other conditions that are present from birth. This impact can be seen in Figure 1.2, where children with JNCL display ‘typical’ presentation and are on par with peers up until the ages of eight or nine, when initial symptoms begin to occur. However, as deterioration sets in, skills diminish gradually into early adult years.

Figure 1.2: Representation of developmental patterns in neurotypical developed children, those with additional needs, and children with Batten disease (adapted from Shapiro & Klein, 1994:16)

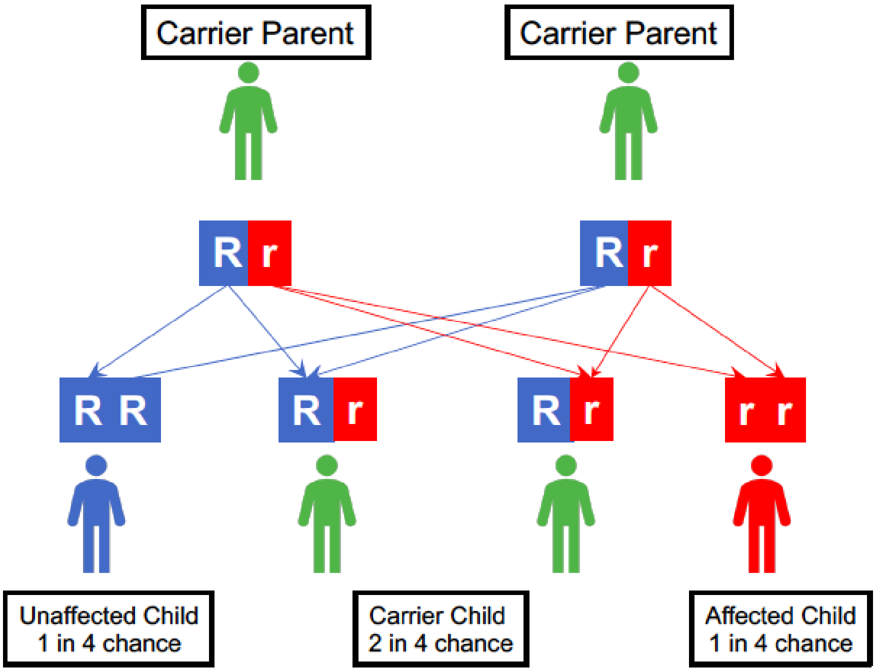


This rare and unusual developmental regression presents additional challenges for a child’s emotional and behavioural state. It is not uncommon for young people with JNCL to experience mood disturbances, anxiety, psychotic states (such as explosive, unprovoked laughing and/or crying), and hallucinations (Bills et al., 1998). Researchers and educators recognise this process can directly lead to perceivably challenging behaviour, as young people experience frustration, depression and demotivation when faced with their diminishing skills and knowledge and the condition (Bills et al., 1998; B. Johnson & Jochum, 1996). The landscape of deterioration for affected children is, therefore, complex and vast.

1.2.2. Prevalence

It is not known for certain how many people have Batten disease worldwide, but by some estimates there are approximately 100 – 150 children, young people and adults currently living with a Batten disease diagnosis in a given year in the UK (BDFA, 2014). Most variants of Batten disease are inherited as autosomal recessive disorders. This means that both copies of the gene are abnormal (one from each parent) with neither working properly (NINDS, 2020). As can be seen in Figure 1.3, both biological parents of a child with Batten disease will be carriers of the disease but physically unaffected by it. The figure provides a visual representation of how genetic inheritance carries the Batten disease gene from parents to their children. There is a 1 in 4 chance of a child being affected by Batten disease, however, due to the inherited nature of the disease, a diagnosis can occur in more than one person in a family. Often, once one sibling has been diagnosed, genetic testing can then identify if other children in the family will be affected, and subsequently, a secondary diagnosis of Batten disease is much quicker and more reliable (Adams et al., 2014).

Figure 1.3: Genetic inheritance of Batten disease



Although Batten disease is sometimes described by the age at which symptoms appear (infantile, late infantile, juvenile) the disease is more generally classified by the gene that causes the disorder (NINDS, 2020). Each gene is called CLN (ceroid lipofuscinosis, neuronal) and is given a different number designation as its subtype. The different gene mutations, signs and symptoms range in severity and progress at different rates. The main forms of Batten disease are summarised in Table 1.4.

Table 1.4: Diagnosis and symptom presentation of eight common types of Batten disease (adapted from NINDS, 2020; 2-5)

Disease Variant	Age on Onset	Symptoms	Prognosis
CLN1 disease (Infantile or Juvenile onset)	Before the age of 1 (infantile) or 5 or 6 (Juvenile onset)	Developmental skills such as standing, walking, and talking are not achieved or are gradually lost. Seizures develop and sight is lost. Some may eventually require full-time care and need a feeding tube.	Most affected children die in early to mid-childhood (infantile) or teenage/ adult years (Juvenile onset).
CLN2 disease (Late infantile and Later onset)	Developmental delay begins around the age of 2 (late infantile) or around 6 or 7 (later onset)	Seizures develop followed by loss of walking and talking. Brief, involuntary jerks in a muscle or muscle group then develop, and most will eventually require full-time care and will need a feeding tube. Later onset starts with the loss of coordination or ataxia.	Most affected children die between the ages of 6–12 years or teenage years (later onset).
CLN3 disease (Juvenile onset, [JNCL])	Symptoms appear around the ages of 4-7.	Initial symptoms include rapid vision loss, learning and behaviour problems, cognitive decline, seizures, speech disorders and motor difficulties. Dependence on caregivers increases.	Most children die between the ages of 15 and 30.
CLN4 disease (adult onset)	Symptoms appear in early adulthood.	Difficulty in movement and dementia symptoms.	Age of death varies.
CLN5 disease (late-infantile onset)	Symptoms appear after the first few years of life.	Seizures begin first, followed by vision loss and learning difficulties. Some may need feeding tubes and become fully dependent on care.	Most children live into late childhood or teenage years.
CLN6 disease (late infantile & adult onset)	Symptoms appear after the first few years of life.	Developmental delay, changes in behaviour, seizures and loss of walking, talking and vision.	Most children die during late childhood or in the early teenage years.
CLN7 disease (late-infantile onset)	Symptoms appear after the first few years of life.	Initially develop epilepsy along with sleep disturbance. Loss in walking and speech.	Most children with the disorder live until their late childhood or teenage years.
CLN8 disease (late-variant onset)	Symptoms are present around the ages of 2 and 7.	Loss of vision, cognitive difficulty, behavioural problems of lack of muscle control.	Life expectancy is uncertain; some children have lived into their second decade of life.

1.3. Batten Disease Research

Understandably, families faced with Batten disease often seek curative solutions. Recent research in the field offers the possibility of pharmacological interventions through enzyme replacement therapy (Lewis, Morrill, Conway-Allen, & Kim, 2019). This treatment has been provisionally accepted into standard care in the UK (NICE, 2018b), yet it is appropriate for only one type of disease variant, and not suitable for treating all children with Batten disease. Additionally, there are significant barriers associated with clinical trials of curative treatments in this arena, which include the ethical use of control groups, the lengthy duration of clinical trials, the diversity of the population and the difficulty with delayed diagnosis (Augustine, Adams, & Mink, 2013). Considering the challenges of curative trials, evidence suggests that more immediate research into alternative treatments based around care, therapy and education could yield meaningful results and better quality of life for children currently living with Batten disease. The primary aim of this research explores how music could support elements of educational and therapeutic support for children affected by Batten disease.

There is little research into how the experiences and wellbeing of the children, or those who support them, are affected over the course of the disease. Children with the disease often lack the cognitive capacity, memory or communication skills to be able to converse with a researcher, which limits the possibility of hearing the child's voice in research (Scambler, 2004). There are a presently only a small number of accounts from educators who have worked with children with Batten disease (Bills et al., 1998; Elmerskog et al., 2019; B. Johnson & Jochum, 1996; von Tetzchner et al., 2019), and an even smaller number of research studies exploring the experiences of those who support children in educational settings (von Tetzchner et al., 2019; von Tetzchner, Fosse, & Elmerskog, 2013). Capturing the experiences of the individuals themselves, or those who support them, could enable research to begin to understand fully the unique impact of the disease for each child. Rather than focusing on complex, lengthy, and restrictive curative solutions, then, it is important for research to consider lived experiences in research both with regards to children with Batten disease, and those who support them. With this in mind, this research project considered how the lived experience of those who support a child with the disease, could provide a holistic picture as to the impact of music for children with Batten disease.

1.4. The Care and Education Pathway

A diagnosis of Batten disease in childhood is often delayed, due to many childhood conditions sharing similar symptoms. Conclusive diagnosis can be challenging, especially in infants and toddlers where functional skills are difficult to observe (T. Johnson et al., 2019). A delayed diagnosis can often mean specialist care and education are considered and implemented after symptoms have already emerged and a deterioration has begun. In the UK, there is no standardised health or education guidelines or policies with recommendations for children with Batten disease. Since a curative treatment for children with Batten disease is yet to be achieved, after diagnosis the focus is on symptom management and therapy to support children in maintaining a reasonable quality of life throughout the disease's progression (Lewis et al., 2019; NINDS, 2020). Typically, pharmacological treatments are offered to help reduce seizure activity, anxiety, depression, and muscle stiffness (NINDS, 2020). According to NINDS, interventions such as physiotherapy and occupational therapy are also offered to maintain function for as long as possible. Support groups are additionally available to help children and their families to cope with the disease (BDFA, 2014). With different types of support targeting specific symptoms, care interventions are often incomplete in achieving holistic care (Augustine et al., 2013; NINDS, 2020).

In terms of education, children often remain in mainstream education in the UK until the point where they require a specialist curriculum or medical support throughout the day (von Tetzchner et al., 2013). At this point in the disease's progression, symptomatic treatment and palliative care prevail (Scambler, 2004). When children are in the final stages of the disease, many families draw on the support of local palliative care pathways, local hospices or respite care. However, funding streams for hospices and respite care in the UK are variable and inconsistent (Scambler, 2004; Together for Short Lives, 2019); therefore access to this care and respite is not always guaranteed. By focusing on educational and therapeutic support for children with Batten disease, this research project aims to deepen understanding in this area providing support for national care pathways to recognise the importance of standardised health and educational approaches and resources.

The absence of such guidance in educational approaches and inconsistent care systems present challenges in the care and education sector for affected children, and consequently influence the experiences of the children, their families and the supporting education specialists (von Tetzchner et al., 2019). Early research demonstrated a need for a greater understanding and knowledge in Batten disease care and education, and a small pool of researchers and educators in the 1990's presented education and therapy recommendations for affected children (Bills et al., 1998; B. Johnson & Jochum, 1996; Shapiro & Klein, 1994). Recent research across Europe and the US has, however, still identified gaps in education, supportive care and therapy for children with Batten disease (Elmerskog et al., 2019; von Tetzchner et al., 2013). Research has suggested that an adapted, personalised and cohesive educational framework could lead to improved maintenance of skill, less frustration and improved emotional wellbeing for the child, which in turn, could improve the situation for children and their families (von Tetzchner et al., 2013). Whilst researchers recognise the importance of quality of life, care and education (Elmerskog et al., 2019; von Tetzchner et al., 2013), systematic research in this area is lacking, and more clinical, evidence-based research is needed in order to fill the gap in care and education knowledge. Research needs to build on existing knowledge and explore the contextual and educational experiences for affected children, their families and their educators. The research project, therefore, aims to contribute knowledge to the field by considering the experiences of educators and professionals and by exploring a personalised and cohesive educational program for affected children.

Lastly, emerging evidence suggests an opportunity exists where a child with Batten disease has the potential to learn new skills, access long term memories and increase cognitive capacity (von Tetzchner et al., 2019). This concept is echoed in clinical trials where early implementation of drug treatment in the early stages of apparent ‘typical’ development can potentially halt or alter the progress of deterioration before irreversible neurological damage has occurred (T. Johnson et al., 2019). Evidence suggests, therefore, that research into pre-emptive approaches to education and therapy (i.e. introducing interventions before deterioration has begun) could impact positively on the overall quality of life for affected children. Drawing upon research into music and neuroscience, this research project considers how musical interventions could complement this pre-emptive approach aiming to provide evidence to support the use of music in educational approaches for children with Batten disease.

1.5. The Importance of Music

Educators appear to agree that an optimised education for children with Batten disease would address pre-emptive learning, active participation, the teaching of life skills, adaptation and compensatory techniques, and could also utilise more under-recognised activities such as music (Elmerskog et al., 2019). Accounts from educators and parents in particular, describe the positive influence of music and music therapy in facilitating increased communication, improved memory retrieval and clearer speech and language (Elmerskog et al., 2019; von Tetzchner et al., 2019). Explorative research in the adult neurology sector suggests a positive impact of music on the brain, evidencing how music can physically alter the brain make-up (Särkämö et al., 2014; Sihvonen et al., 2017), and bypass damaged areas of the brain to facilitate improvements in speech, emotion, movement and memory (Harris, Leenders, & de Jong, 2016; Salimpoor, Benovoy, Larcher, Dagher, & Zatorre, 2011; Samson, Dellacherie, & Platel, 2009; Volpe, Signorini, Marchetto, Lynch, & Morris, 2013). Similarly, music therapy research has demonstrated a positive impact of music groups and music-based home programs on emotional wellbeing and quality of life for adults with dementia (Baker, Grocke, & Pachana, 2012; Sixsmith & Gibson, 2007). Although adult neurology studies can offer relatable theory, systematic research into Batten disease is lacking, and there is a gap in current knowledge around music and its impact on language and/or memory for children with Batten disease. This research project, therefore, aims to explore the musical experiences of children with Batten disease, their responses to music interventions, the benefits of pre-emptive education approaches and how these might be translated into a music-based support resource for educational settings.

Lastly, research must address the challenge of a heterogeneous population and the absence of appropriate assessment tools for Batten disease, particularly in the music therapy sector. Although recent developments have provided more robust assessment measures for brain injury, Huntingdon’s and dementia populations (Magee, Siegert, Taylor, Daveson, & Lenton-Smith, 2016; McDermott, Orrell, & Ridder, 2015; O’Kelly & Bodak, 2016), research into validated methods of assessments for music therapy and neurodegenerative childhood conditions is currently non-existent. The absence of a relevant and comparable assessment for the Batten disease population currently prevents effective music-based interventions from being investigated effectively. This research project thus aims to explore relevant and appropriate assessment measures, to support research developments in this arena.

1.6. Overview

Research on Batten disease has primarily focused upon clinical trials in enzyme replacement therapy (Lewis et al., 2019). Yet clinicians and educators have recognised the need for improved strategies, research and education provisions for those affected by Batten disease (Augustine et al., 2013; Bills et al., 1998; B. Johnson & Jochum, 1996). Although under-researched, music is recognised as a key support mechanism for individuals with JNCL (Elmerskog et al., 2019; von Tetzchner et al., 2019), and research in the adult neuroscience sector suggests that music can have a positive impact on many areas of neurological functioning (Harris et al., 2016; Salimpoor et al., 2011; Samson et al., 2009). By capturing the lived experiences of those who support children with Batten disease, research could more holistically examine the potential role that music can play in enhancing their quality of life. As appropriate assessment tools for children with Batten disease are lacking (Augustine et al., 2013) and underdeveloped for music therapy, research also needs to develop new robust and systematic assessment tools to assess how music can be of benefit for children who are deteriorating.

1.7. Aims of the Doctoral Research

Given the overview above, this doctoral research aims to contribute positively to the dearth of evidence presently available in relation to music and its importance for children affected by Batten disease. The project firstly, sought to ascertain the appropriateness of a new music therapy outcome measure for children with Batten disease, the Chiltern Music Therapy Observation Measure (CMTOM, Atkinson, 2018;

see Appendix 1). To provide a clinical perspective of change over time, the project drew upon the Hamburg Clinical Rating Scale (Kohlshutter, Laabs, & Albani, 1988) to capture the clinical presentation of children with Batten disease over three years. Using the new music therapy outcome measure, the project then aimed to capture functional responses in music therapy over three years, with a focus on communication, wellbeing, cognition, mobility and behavioural responses. It has been suggested that music can be beneficial for children with JNCL (Elmerskog et al., 2019; von Tetzchner et al., 2019), and the project, therefore also aims to ascertain how responses in music therapy change over time for these affected children. Focusing on the JNCL variant of Batten disease, the project aims to translate elements of music therapy into an education context, exploring the perceived outcomes in response to a bespoke music education program ‘Music Speaks’. Importantly, the project aims to capture the experiences of educators and therapists supporting children with JNCL, and their experiences of using the Music Speaks program in a UK school setting.

The three research aims are therefore as follows:

1. To examine the ways in which children affected by Batten disease respond to music therapy, focusing on social, emotional, physical and cognitive outcomes.
2. To explore the ways in which children affected by Juvenile Batten disease respond to a daily music education program within their school setting.
3. To illuminate the experiences of educators and therapists who have supported children affected by Juvenile Batten disease using the Music Speaks program, within UK education settings.

Table 1.5 presents the research objectives aims, related research questions, methods, and data collection, assessment and analysis processes.

Table 1. 5: Overview of the research matrix mapping objectives, questions, approaches, data collection and analysis method

Research Phase	*Research Aims*		*Research Questions*	*Methods*	*Assessment Measures*	*Data*	*Analysis*
1	To examine the ways in which children affected by Batten disease respond to music therapy, focusing on social, emotional, physical and cognitive outcomes.	*1.1*	For children with Batten disease, how can functional outcomes be tracked and measured in music therapy?	Case study design	CMTOM	Questionnaire data Rater reliability data	Descriptive Statistics Cohen's (K) Intra-class Correlation
		1.2	From a clinical perspective how do functional outcomes (social, physical and cognitive) change over time for children with Juvenile Batten disease?	Case study design	Hamburg	Quantitative Hamburg data	Content Analysis Descriptive Statistics
		1.3	How do functional outcomes (social, physical and cognitive) change in music therapy over time for children with Juvenile Batten disease?	Case study design	CMTOM	Quantitative CMTOM data	Content Analysis Descriptive Statistics
2	To explore the ways in which children affected by Juvenile Batten disease respond to a daily music education program within their school setting.	*2.1*	Can the Music Speaks program be delivered effectively in a formal education framework?	Case study design	Program Recording Sheets	Program Delivery Task Completion Free Text	Descriptive Statistics
		2.2	How do communication outcomes for children with Juvenile Batten disease change in response to the Music Speaks program?	Case study design	CMTOM Coding	Quantitative CMTOM Data	Descriptive Statistics
3	To illuminate the experiences of educators and therapists who have supported children affected by Juvenile Batten disease using the Music Speaks program, within UK education settings.	*3.1*	What are the experiences of educators and therapists supporting a child with Juvenile Batten disease in an educational context?	Semi-structured Interviews	Coding	Interview transcripts	Thematic Analysis
		3.2	What are the perceptions of the Music Speaks program from those using it in educational contexts?	Semi-structured Interviews	Coding	Interview transcripts	Thematic Analysis

1.8. Research Questions

1.8.1. Phase One

The first phase of the research enquiry aimed:

1. To examine the ways in which children affected by Batten disease respond to music therapy, focusing on social, emotional, physical and cognitive outcomes.

The research aim was addressed through three research questions, firstly including all types of Batten disease, and later focusing on the Juvenile variant of the disease:

- 1.1. For children with Batten disease, how can functional outcomes be tracked and measured in music therapy?
- 1.2. From a clinical perspective how do functional outcomes (social, physical and cognitive) change over time for children with Juvenile Batten disease?
- 1.3. How do functional outcomes (social, physical and cognitive) change in music therapy over time for children with Juvenile Batten disease?

The dearth of measures available in the context of paediatric neurodegeneration and music therapy led this research project to first identify a suitable measure for tracking and measuring responses and tendencies in music therapy. This phase included measures to explore the validity and reliability of the Chiltern Music Therapy Outcome Measure (CMTOM). The CMTOM provides a framework of assessment for an in-depth and regular analysis of a patient's presentation within a music therapy context. It can track and monitor a patient's presentation in relation to music-based skills that fall within the functional areas of speech, behaviour, cognition, movement and wellbeing. The development of the CMTOM is described further in chapter four. For the purposes of this research, this measure was examined solely for its suitability in the context of tracking functional responses in music therapy for children with Batten disease; namely speech, cognition, physical skills, emotional wellbeing and behaviour (research question 1.1).

The research project then drew upon data from a preceding study (Music in Neurodegeneration [MIND]), which considered the impact of music therapy for individuals with Batten disease (Ockelford et al., 2019). The MIND research, for which I was lead researcher, implemented a case study design, which tracked 12 individuals with Batten disease in their music therapy and music lessons over three years. Originally, this project's video data were analysed using two measures: the Hamburg Clinical Rating Scale (Kohlshutter et al., 1988) and the Sounds of Intent

[SOI] framework, a music education assessment tool (Vogiatzoglou, Ockelford, Welch, & Himonides, 2011). This research project assessed annual data from the Hamburg Clinical Rating Scale (Kohlshutter et al., 1988) which provided a clinical perspective on each child's motor, speech and cognitive presentation. The scoring system is used to track the progress of decline for children with Batten disease diagnosis (National Institute of Care and Excellence, 2018b; Johnson *et al.*, 2019), and was completed with the parents of the children in the research project (research question 1.2). The latter measure from the preceding study, the SOI, focused solely on capturing the children's musical skills in the MIND research study. For the purposes of this doctoral research, therefore, the video data were re-analysed to observe the functional tendencies, behaviours and outcomes identified in the CMTOM measure and how these changed over time with a specific focus on children with JNCL (research question 1.3).

To achieve triangulation (Guion, Diehl, & Mcdonald, 2002), data collection in this phase included data generated through multiple perspectives from researcher visits, clinical assessments and practitioner-videoed sessions. Videos were taken at each researcher visit and Music Therapists also contributed video recordings of their music therapy sessions.

Drawing upon these two assessment measures enabled the research enquiry to ascertain responses to music therapy and functional changes from two distinct perspectives: a clinical perspective and a music therapy perspective. The Hamburg scale highlighted how tendencies of deterioration are generally tracked and monitored within a standard clinical pathway. Using the CMTOM measure enabled the research to assess functional outcomes within a music therapy setting and compare how these outcomes changed and differed to the clinical rating scale over time. Utilising longitudinal data, Phase One aimed to capture measurable information over a period by developing measures, triangulating perspectives and observations, and capturing responses that are present in music therapy sessions for those affected by Batten disease, specifically JNCL.

1.8.2. Phase Two

The second phase of the research project aimed:

2. To explore the ways in which children affected by Juvenile Batten disease respond to a daily music education program within their school setting.

In response to the second research aim, Phase Two focused on whether observed changes in language functionality could be translated to an educational context. This phase firstly set about devising a music therapy education program, Music Speaks, for educators supporting affected children. The research aim was addressed in the following two research questions:

2.1 Can the Music Speaks program be delivered effectively in a formal education framework?
2.2 How do communication outcomes for children with Juvenile Batten disease change in response to the Music Speaks program?

By applying research theory and practices of previous researchers in the field (Thaut, 2014; von Tetzchner et al., 2019), Music Speaks combined elements of Neurologic Music Therapy for everyday use in a UK school setting children (described further in chapter four). Within a case study design, five individuals with JNCL trialled the Music Speaks program within their school settings. Following training sessions with educators, Music Speaks was delivered daily by learning support staff and education professionals over a six-week trial period. Recording sheets were completed by educators and therapists delivering the program, which presented data about program delivery, timings, attendance, and program completion. This data aimed to present information about how the program could be delivered in a school setting by non-musical professionals, relating to the first research question 2.1.

This phase also aimed to examine the functional responses to the Music Speaks program to determine whether music therapy outcomes could be translated into an educational context. During the trial, videos from educators delivering the program were collected and assessments were made from the video data. A focus on music and language was specifically important in this phase of the research and therefore, the CMTOM observation assessment was adapted to capture the communication responses over the trial period (research question 2.2). The two sub-phases of this second research phase aimed to generate contextual information regarding functional responses to music in an educational environment. This phase focused on translating functional outcomes observed in music therapy into a bespoke music education program Music Speaks, and then observing how children responded to this daily program.

1.8.3. Phase Three

Phase Three aimed to capture the experiences, perspectives and perceptions of the educators who delivered the Music Speaks program in a school setting, aiming:

3. To illuminate the experiences of educators and therapists who have supported children affected by Juvenile Batten disease using the Music Speaks program, within UK education settings.

This phase aimed to explore these through the following research questions:

3.1 What are the experiences of educators and therapists supporting a child with Juvenile Batten disease in an educational context?
3.2 What are the perceptions of the Music Speaks program from those using it in educational contexts?

In a case study design, seven semi-structured interviews were conducted with educators and therapists from Phase Two, firstly exploring their experiences and perceptions of education within the field of Batten disease, and secondly exploring their perception of the impact of the Music Speaks program trial. Interviews gathered multiple first-hand accounts from the education sector (Learning Support Assistants and an Inclusion Lead), the therapy sector (Music therapists) and the Visual Impairment sector (VI teams). These were transcribed and analysed using inductive thematic analysis. Codes were generated from the texts, to propagate themes and sub-themes, representing the experiences and perceptions of educators and therapists (research question 3.1). Additionally, analysis focused on the experience of delivering the Music Speaks program (research question 3.2).

In summary, the systematic yet explorative three-phase structure of this research project contributes in-depth knowledge on this subject by building on past literature and theory alongside the contextual reality of those people supporting individuals with JNCL within education settings. To explore the experience of music for children with Batten disease in a holistic way, the doctoral project applies researcher experience and reflection, neuroscientific theory and principles of music therapy, whilst capturing empirical observations and participant experience. Ultimately, it is expected that, in the future, this three-phase project will forge a pathway for larger-scale research, investigating the effectiveness of music therapy and music education programs to improve functional outcomes and wider care pathways for those living with or supporting Batten disease and other neurodegenerative conditions.

1.9. Thesis Overview

This doctoral narrative is divided into eight chapters.

Following this introduction, chapter two explores the nine influencing theoretical areas that have influenced the direction of this research: current clinical research for children with Batten disease; the lived experience of children and their families; current education and therapy knowledge; a pre-emptive learning approach; the impact of music for Batten disease; the neuroscience of music; the impact of music in adults with neurological conditions; music therapy and relevant assessment and outcome measures in the field.

Chapter three outlines the epistemological journey in the research examining the critical realist standpoint (Bhaskar & Danermark, 2006) and its influence on the project design and methodology.

Chapter four defines the methodology employed over the three research phases, which includes the development and validation of the CMTOM measure and observations of children in music therapy; development of the Music Speaks program; and the experiences and insights from education staff supporting affected children. Chapter four describes in detail the methods used in each of the three distinct phases, setting out the data gathering methods, participants, assessment measures and methods of analysis for each phase.

Chapter five outlines the results and findings from Phase One, relating to the quantitative analysis of data gathered over the three-year observation period, with a particular focus on the impact of music on the key functional areas of communication, cognition and physical outcomes in music therapy, alongside findings from the validation of a bespoke music therapy measure.

Chapter six presents results from the quantitative analysis conducted in Phase Two, relating to the implementation of the Music Speaks education program.

Chapter seven presents findings from Phase Three, which involved interviews with educators who delivered the Music Speaks program in support of the children affected by JNCL. Findings detail the thematic analysis of the transcribed interviews here.

Chapter eight concludes the research enquiry by opening a discussion into the observations, perspectives and systemic mechanisms, which are present throughout the three phases of the research. The chapter highlights the observed importance of music in education and music therapy, diversity of the population and the lived experiences of those who support a child with Batten disease. Focusing on existing knowledge gaps in the field, the discussion considers the impact of current care, education and pre-emptive learning for children, as well as the underlying neurological impact of music interventions. Chapter eight also considers the feasibility and reliability of the bespoke measures and programs developed in the research, addressing measurement, outcomes, adaption and implications for children with Batten disease. Moreover, this concluding chapter highlights potential application of findings to other neurodegenerative conditions in both child and adult populations, and the impact of other music programs available to staff, educators and families of those affected by neurodisability and dementia.

Chapter 2: A Review of Literature

"Where words fail, music speaks"
– Hans Christian Andersen

2.1. Introduction

Whilst the impact of music for childhood dementia is a relatively under-researched area, there is a small pool of research that frames the landscape of experience, education, care and treatment interventions for children with Batten disease. From pharmacological treatment to education theories and therapeutic accounts, this literature review begins by drawing upon a small group of relevant studies that provide the context within which the research aims of this project are placed. Exploring the neurological impact of music and music therapy, particularly for those with neurological conditions can additionally offer relatable theories. Thus, this review interrogates nine key areas of literature and research that have influenced the direction of this research project. The scoping and review of literature for this thesis began in 2016 and ended in March 2020 and as such, research published after this date were not included in this literature review. The key areas are summarised as follows:

- Clinical Research for Batten disease
- The Lived Experience of Children and their Families
- Education and Therapy Knowledge
- Pre-emptive Learning
- The Impact of Music for Batten Disease
- The Neuroscience of Music
- The Impact of Music in Adults with Neurological Conditions
- Music Therapy
- Assessment and Outcomes

This chapter begins by exploring the current landscape of pharmacological treatments for Batten disease and the associated limitations of clinical research trials. Drawing upon lived experience, the chapter then explores what it is like for families receiving a diagnosis of Batten disease, their experiences navigating through care pathways in the UK, and the challenges associated with a lack of support or knowledge of the rare disease (Scambler, 2004; von Tetzchner et al., 2019). The chapter then presents studies exploring first-hand accounts into the challenges of supporting and caring for children with the disease in an educational context. In contrast to research into pharmacological interventions (Lewis et al., 2019), first-hand accounts from those supporting affected children place an important focus on experiences, perspectives, and challenges faced by those supporting children in educational contexts (Bills et al., 1998; B. Johnson & Jochum, 1996; Scambler, 2004; von Tetzchner et al., 2019). These first-hand accounts provide insight into past and present challenges in support pathways, providing a contextual understanding from the perspectives of families, and those in the education and care sector. The chapter then focuses on more recent research into the impact of recent bespoke education strategies, pre-emptive learning approaches and studies linking together music, education and therapy for children with Batten disease (Elmerskog et al., 2019; von Tetzchner et al., 2019). This chapter then explores research concerning the impact of music for children with Batten disease, drawing upon a pool of researchers who have been beginning to investigate this phenomenon over the past decade (Elmerskog et al., 2019; von Tetzchner et al., 2019).

Although research relating directly to the neuroscientific impact of music and Batten disease is absent, adult studies in music and neurology can offer relatable insight. Building on foundational arguments relating to the physical impact of music, this chapter explores research into music and neuroscience (Alluri et al., 2012; Särkämö, Altenmüller, Rodríguez-Fornells, & Peretz, 2016; Sihvonen et al., 2017). The literature review explores how these theories and findings facilitate an understanding of the impact of music on neurological diseases and neurodegenerative illness (Harris et al., 2016; Salimpoor et al., 2011; Samson et al., 2009; Särkämö et al., 2014). Evidence from the music therapy field is also discussed in relation to Batten disease, drawing in particular, upon evidence from the neurodisability and brain injury sectors (Baker et al., 2012; Bowell & Bamford, 2018; Bringas et al., 2015; Lim et al., 2013; Straube, Schulz, Geipel, Mentzel, & Miltner, 2008). Lastly, the chapter concludes with an exploration into relevant music and music therapy assessment and outcome measures for children with Batten disease (Augustine et al., 2013; Enderby & John, 2019; Vogiatzoglou et al., 2011; von Tetzchner et al., 2013).

2.2. Clinical Research for Batten Disease

As yet there is no confirmed cure for Batten disease. However, clinical trials in the field are working towards pharmacological treatments which can slow or halt the progression of the disease. Clinical trials are currently focused on enzyme replacement therapy (Lewis et al., 2019), and although recent international trials have demonstrated some success here, the treatment is only relevant to one of the 14 types of the disease (currently CLN2). There are currently no other treatments that can slow or halt the progress of the disease for other types of NCLs (NINDS, 2020), and it is unclear whether single variant clinical trials can be generalisable across the other forms of Batten disease (Augustine et al., 2013). It is also crucial to acknowledge that, whilst the disease presents with overall deterioration, the heterogeneous and complex nature of the population means every child's experience is unique, and it is difficult to comparatively track, measure and monitor presentation over time (Augustine et al., 2013).

Additionally, the complex and fragile clinical presentation of a child with Batten disease universally makes research extremely challenging. The core challenge faced by researchers is centred around the ethics of investigating a cure with a fragile and vulnerable population. Treatments that use a control or placebo groups are, in general, unethical and often not an option. Additionally, it is imperative that any treatment is rigorously vetted and tested on animals with the disease before human testing can begin (Kohan et al., 2011). Moreover, deterioration associated with Batten disease is primarily focused within the brain, which is a comparatively difficult area to access with drug treatments, and clinical trials in this sector are therefore considerably challenging for rare disease therapy development (Augustine et al., 2013). Given the small sample sizes, multiple variants and barriers in applying the vectors, they are time-consuming, costly and complex. The lengthy research protocols associated with clinical trials mean that many children with Batten disease today will not receive an approved curative treatment in their comparatively short lifetime. For treatment to be most effective, early intervention is key, which is often before diagnosis of the disease (Kohan et al., 2011). Considering the challenges of curative trials, evidence suggests that immediate research into alternative treatments such as care, therapy and education could yield meaningful results and better quality of life for children currently living with Batten disease (von Tetzchner et al., 2019). The restricted specificity of clinical trial research combined with the diversity of the population highlight the need for further research into alternative holistic interventions (such as education or therapy strategies), applicable to all variants of Batten disease.

In Batten disease research, first-hand accounts from affected children themselves are absent. Past research has identified the challenges with gathering this type of data from the children themselves – most children are not told they have the disease and therefore interviewing these children is challenging and unethical. Additionally, many teenagers with the disease lack the cognitive capacity, memory or communication skills to be able to converse with a researcher, which makes interviews impossible to conduct (Scambler, 2004). For this reason, research must employ a variety of data collection methods from multiple perspectives to build a comprehensive picture of the research topic from multiple dimensions. Perspectives and experiences of families and practitioners (educators, therapists and specialists) can provide important insights into the child's experience. Professionals supporting and educating children with Batten disease witness the daily experiences of the child in a school environment. Researchers also acknowledge that the experiences that families go through are so extreme, that many suggest families have first-hand experience of the disease, albeit indirectly (Scambler, 2004). This makes the experiences of educators and carers valuable in providing insight into a child's day-to-day experience living with the disease. Representing the experiences of families and practitioners, the following section therefore, provides some insight into the experiences of the child, their family and the practitioner.

2.3. The Lived Experience of Children and Their Families

From the point of diagnosis, the unique life of a child with Batten disease presents complex challenges, and in most cases, these challenges often stem from before the eventual diagnosis of Batten disease is reached (Scambler, 2004). Firstly, from before the point of diagnosis, research has demonstrated that parents felt they had to “clear the road themselves” (von Tetzchner et al, 2019: 483) when it came to the early stages of diagnosis of their child. In a research study exploring parents’ lived experiences, Scambler (2004) found that contact between the family, the health and medical sectors tends to be limited, strained or negative in the early phase of the disease, and many were left feeling dissatisfied, daunted and ill-equipped to know how to manage the news of the diagnosis. Post-diagnosis many families additionally experienced social isolation whilst supporting a child with such a rare condition (Scambler, 2004). Psychological and emotional challenges were also present in many parental accounts, with parents specifically reflecting upon their initial feelings of grief, bereavement and loss, which were felt from the moment of diagnosis (Scambler, 2004). Recognising the long and drawn-out journey of Batten disease, many reflected on the dichotomy of grief and loss alongside the need to try and instil a quality of life, not solely a quantity of life, for their child (Scambler, 2004).

Bills and colleagues (1998) identified that, in contrast to a developmental condition present from birth and like adult dementia, a child with Batten disease is aware of a physical deterioration and developmental regression, which creates intense emotional and psychological challenges for the child. Combined with changes in behaviour and personality, this can lead to experiences of ongoing psychological trauma and distress (Bills et al., 1998). Researchers recognise that the disease can lead to perceivably challenging behaviour, as young people experience frustration, depression and demotivation when faced with their diminishing skills and knowledge of the condition (Bills et al., 1998; B. Johnson & Jochum, 1996). Accounts from parents have highlighted the need for information, support and knowledge that considers both the wellbeing of the families and the affected child from the early stages of diagnosis and beyond (Scambler, 2004). Evidence suggests that there is a significant gap in support, health and wellbeing services for newly diagnosed families to support them throughout the journey of the disease.

Trauma and distress are also present when parents are faced with their child's diminishing and regressed faculties (Bills et al., 1998; Scambler, 2004). One mother of a 26-year-old adult with Batten disease, expressed that “the work [of caring for her son] is financially, emotionally, and physically draining” and that her son is “no longer able to communicate to the outside world, something that is frustrating to himself and his family" (Hanson, 1993 cited in B. Johnson & Jochum, 1996: 304). The communication breakdown was identified as the most distressing element of a child's deterioration for parents with an affected child (Scambler, 2004). Families are often unable to hold a two-way conversation, have meaningful interactions or read facial expressions when motor skills have completely diminished, causing further psychological trauma and emotional distress for families and children (B. Johnson & Jochum, 1996; Scambler, 2004). Family experiences like these suggest a need for greater knowledge, expertise, or guidance around how communication could be extended for longer. Therefore, research in this area could add significant, meaningful value for those supporting a child with Batten disease.

There is little research concerning the wellbeing of those who support children with Batten disease in educational settings. Yet research in the adult sector suggests that wellbeing can be impacted by caring for an individual with dementia. Previous research highlights the complexities of supporting adults with dementia, recognising the frustrations around lack of resources and time, lack of knowledge in others, and the importance of staff wellbeing (Edberg et al., 2008; Kitwood, 1997). Researchers have previously cautioned the danger in professionals or educators handling the education of a child with Batten disease alone, which can lead to distress and burnout (von Tetzchner et al., 2013). Adding further psychological stress and ethical complication to the situation, many parents, educators and professionals disagree about whether the child should know about their diagnosis of Batten disease. Though children perhaps recognise their deterioration, research suggests that often parents chose not to tell their child the explicit truth about the condition, going against advice from medical professionals (Scambler, 2004). This disagreement between parents and professionals can complicate approaches, strategies and psychological care for the child, where there needs to be a consideration for respecting both the needs of the child and the family's wishes (Scambler, 2004). This perhaps adds to an already complex situation where clear strategies and care approaches are in some cases misaligned. Research here highlights the need to understand the lived experiences of those caring for and supporting a child with Batten disease. There also needs to be a unified framework of guidance for carers and professionals that considers the complex ethical considerations that are present for children with Batten disease and their families.

Although research identifies gaps in medical support and knowledge, parent support organisations, such as the UK Batten Disease Family Association (BDFA), offer support and valuable resources to families and children (von Tetzchner et al., 2019). The BDFA offers research funding, parent advocacy, family support, education and care planning advice, and psychological support to families and siblings of affected children. Despite this support, there are limitations in accessing this form of advocacy (von Tetzchner et al., 2019). Unlike other areas, such as Scandinavia, the UK does not have a central registry system for all children with a Batten diagnosis.

The BDFA therefore relies heavily on medical practitioners signposting families towards their support services. If professionals are unaware of the BDFA, some families are perhaps not signposted towards this crucial support (von Tetzchner et al., 2019). The BDFA reflect that, although support groups like theirs exist, and social media can go some way in combating isolation and bringing families together, they firmly believe they cannot solely support all aspects of a family’s needs and thus actively campaign for improvements to national services and care (von Tetzchner et al., 2019).

Research here, focusing on the lived experience of families and carers, highlights the trauma and distress throughout multiple points of a child’s life, which begins with complicated, traumatic, and sometimes incorrect, diagnosis (Scambler, 2004). From early experiences with medical professionals and the medical sector, Scambler’s research crystalises the importance of positive early experiences that could work towards decreasing the distress experienced by families and thus children. More specifically, there is a dearth of evidence relating to strategies to support a child’s communication, emotional wellbeing, and psychological wellbeing, and there is a need for improved information, support and care at the point of diagnosis and throughout the journey of deterioration (Scambler, 2004). Crucially, there is little evidence or guidance to support the emotional wellbeing of parents, families and carers of affected children. Research here has illustrated that the absence of such support can further inflate feelings of isolation, distress, anxiety and wellbeing for those who are living through the traumatic experience of Batten disease with their affected child (Scambler, 2004). The impact of current guidelines, recommendations and education and care pathways for children affected by Batten disease in the UK is explored further in the following subsection.

2.4. Education and Therapy Knowledge

Current UK guidance around education and therapy for Batten disease is limited to either providing a medical overview of the condition (NINDS, 2020), or a list of current clinical pharmacological trials (NICE, 2018b). In most cases, the care pathway for a child with Batten disease centres around symptom management and often involves multiple professionals at different points in their care (Augustine et al., 2013; NINDS, 2018). For example, seizures are managed by medication, occupational therapy and physiotherapy address motor impairments, behavioural symptoms are addressed within psychiatric care, and specialist education services support visual and cognitive impairments. Currently, there are no health recommendations (such as guidance from NICE or the NHS) regarding education and care support for children with Batten disease. With professionals working in isolation, interventions are often unable to achieve total symptom control, and education and therapies often lack cohesion or longevity (Augustine et al., 2013). The absence of standardised guidance demonstrates a need for further research into holistic and joined-up care strategies, which could consider other interventions such as music therapy or arts-based interventions.

Early research in 1996 within US mainstream education recognised challenges in the education system, noting an insufficient evidence base for successful education strategies (B. Johnson & Jochum, 1996). This highlighted the need to adapt and plan for a constant new ‘normal’, and researchers recognised the challenge of an unpredictable disease progression that can be simultaneously sudden, gradual or unchanging at different stages of a given child’s journey (B. Johnson & Jochum, 1996). It was identified that a lack of educator knowledge and understanding of Batten disease was a significant challenge for those approaching education for affected children (Bills et al., 1998). More than twenty years later, research still echoes many of these challenges in the education sector, with educators expressing difficulty defining educational goals, completing educational plans, or implementing appropriate interventions for their student, due to the unpredictable nature of the disease (Elmerskog et al., 2019). Accounts here however, reference care systems in the US or Scandinavian countries and there is a significant lack of research exploring the situation in the UK.

Research conducted by Von Tetzchner and colleagues (2019) in Norway echoed this lack of support. Evidence suggested that individual experiences and support from family advocacy organisations generally formed the basis of expertise and knowledge in the education sector, rather than government or health sector information (von Tetzchner et al., 2019). In their work with Norwegian children with Batten disease, researchers identified an absence of any consistent educational or therapeutic approach. To understand this further, they launched a multinational project that asked parents and educators (N=192) about the care, quality of life, education and therapeutic input for a child or young person they supported with Batten disease (von Tetzchner et al., 2019). The project spanned six countries (UK, Norway, Denmark, Finland, US and Germany) and 18 different organisations (comprising syndrome societies, advocacy groups and educational consultation services), as well as schools, academic and medical institutions focusing on the educational and social care needs of individuals with JNCL. Across the international participants and diverse educational systems, significant worldwide issues in relation to a general lack of therapy and education resources were identified. It was found that educators lacked adequate resources and information to build sufficient competence to support affected children (von Tetzchner et al., 2019). For newly-diagnosed families, parents drew specifically on the support of other parents in the same situation and relied on small, overstretched parent advocacy groups, in the absence of medical or government initiatives (von Tetzchner et al., 2019). With regard to therapy services, parents of children with JNCL identified that under half of the children with Batten disease (38%) had access to speech and language therapy services and fewer (35%) had access to supportive therapies such as music therapy (von Tetzchner et al., 2019). The study highlighted societal constraints that influence a child’s care pathway, illustrating inconsistencies in numerous areas of a child’s education and therapy.

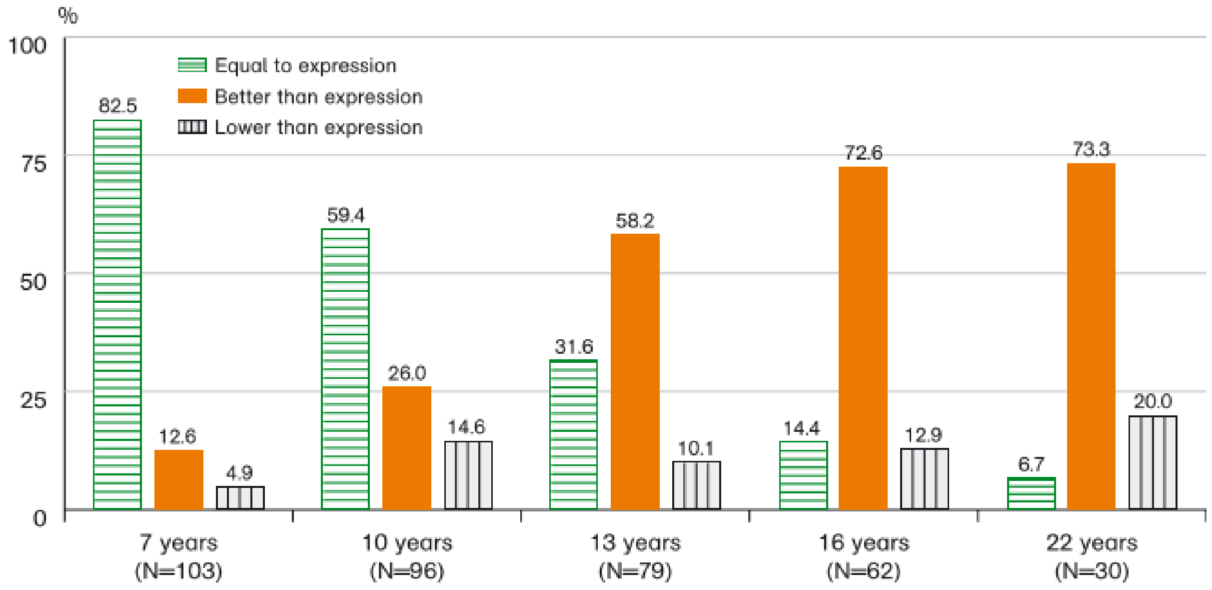
Whilst the study makes initial recommendations around education and care approaches for children with Batten disease, there is still a need for more research to be carried out considering the impact of education and dissemination with professionals in the UK and in Europe. The limited research in this sector demonstrates that the current provision for specialist education and therapy for children with Batten disease is restrictive. Gaps in education and therapy support are potentially forcing families to rely upon pharmacological interventions that have not yet been proven to be effective for every variant of the condition (Lewis et al., 2019; von Tetzchner et al., 2019), and more needs to be done in the research sector to offer an alternative. Findings from more recent research have also unearthed key ideas regarding the theory of pre-emptive learning for children with Batten disease, which are discussed below.

2.5. Pre-emptive Learning

Considering the inevitable decline of a child with Batten disease, early research cautioned against educators focusing on shortened life expectancy or palliative care, rather than the child’s potential (Bills et al., 1998). Other researchers also recognised that declining function could create the misconception that individuals were more ‘vegetative’ than they are, and in some cases, education support plans have simply aimed to keep a child happy rather than considering alternative ways to engage with the child (B. Johnson & Jochum, 1996). Recognising challenges in the adult dementia sector, Kitwood (1997) similarly identified the tendency of care staff to simply manage behaviour, depersonalise patients or, at best, poorly meet the psychological needs of adults in care, which further amplified feelings of despair, depression, vegetation, exhaustion and apathy in adults with dementia. Bills’ work with children in the US with Batten disease importantly observed that children lost their motivation to interact if subtle attempts were missed and not responded to, which in turn bred passivity in the child’s communication (Bills et al., 1998). Another study highlighted that professionals did not acknowledge the need to address education approaches for children with an unpredictable future, where one professional commented that “it feels wrong to make plans for the future when nobody knows what is going to happen” (von Tetzchner et al., 2019:132). This research sheds light on the experiences of those living with or those who support, adults and children with dementia. It highlights the need for a closer understanding of the capabilities of every child throughout their journey living with Batten disease.

The work of von Tetzchner and colleagues additionally unearthed important evidence with regards to a child’s understanding and comprehension. From survey data, parents reported that they believed their children were able to understand just as much as they were able to verbally express (von Tetzchner et al., 2019). As can be seen in Figure 2.1, 82.5% of parents and staff in their research (n = 85/103) felt that their child's language comprehension was on par with their expressive language in the early stages of the disease. However, as the disease progressed and expressive communication declined, most parents and staff (73.3%, n = 22/30) felt that their child's language comprehension remained intact. This indicates that in the early stages of adulthood, despite a child's ability to largely understand what is communicated to them, their decreasing expressive communication leaves them 'locked in'.

Figure 2. 1: Parent and staff perceptions regarding speech comprehension and expression for individuals with JNCL over time (percentage of survey responses shown here) (von Tetzchner et al, 2019:100)



Von Tetzchner et al (2019) went on to suggest that despite declining communication skills, more can be offered in the education and therapy sector to pre-empt the decline and deterioration of cognitive skill. A natural history study over 15 years, which explored cognitive skills for children with JNCL, indicated that the children experienced initial modest gains in some cognitive skills (albeit to a lesser extent than healthy peers) before previously attained skills decline (Elmerskog et al., 2019). Whilst deterioration was inevitable for the individuals in this research, there appeared to be a window of opportunity where learning could still take place before the onset of decline occurs. This was an important concept for educators and professionals supporting individuals with the disease to comprehend in terms of pre-emptive learning. Identifying this window of opportunity through early education and therapy interventions could facilitate improvements or maintenance of skills learned earlier on in a child's life.

Educators from past and present research concur with the notion of challenging misconception and championing a proactive learning approach that pre-empts expected declines in skills and abilities (Bills et al., 1998; Elmerskog et al., 2019; von Tetzchner et al., 2019). For children and young people with Batten disease, as capabilities decline and regress, the last thing a child learns is generally the first to be lost (Bills et al., 1998; B. Johnson & Jochum, 1996). By instilling a pre-emptive approach, children and young people with Batten disease could be given the opportunity to learn and maintain practical skills for longer, such as important independent life skills. Pre-emptive learning could involve, for example, introducing devices or technology to enhance mobility, communication and independence (i.e. the use of a cane, braille or other communication aids) whilst learning is still possible. This could hold value for the child in times of transition such as change of schools or residential services. Yet, it is not instinctual for educators to work backwards and teach in reverse (B. Johnson & Jochum, 1996), and the real challenge is in refining a unified approach that potentially contradicts the traditional method of mainstream teaching which focuses on development, achievement, and progress. Pre-empting interventions and implementing support, education and therapy strategies earlier on could not only facilitate improvements for the child but could provide guidance and support for educators.

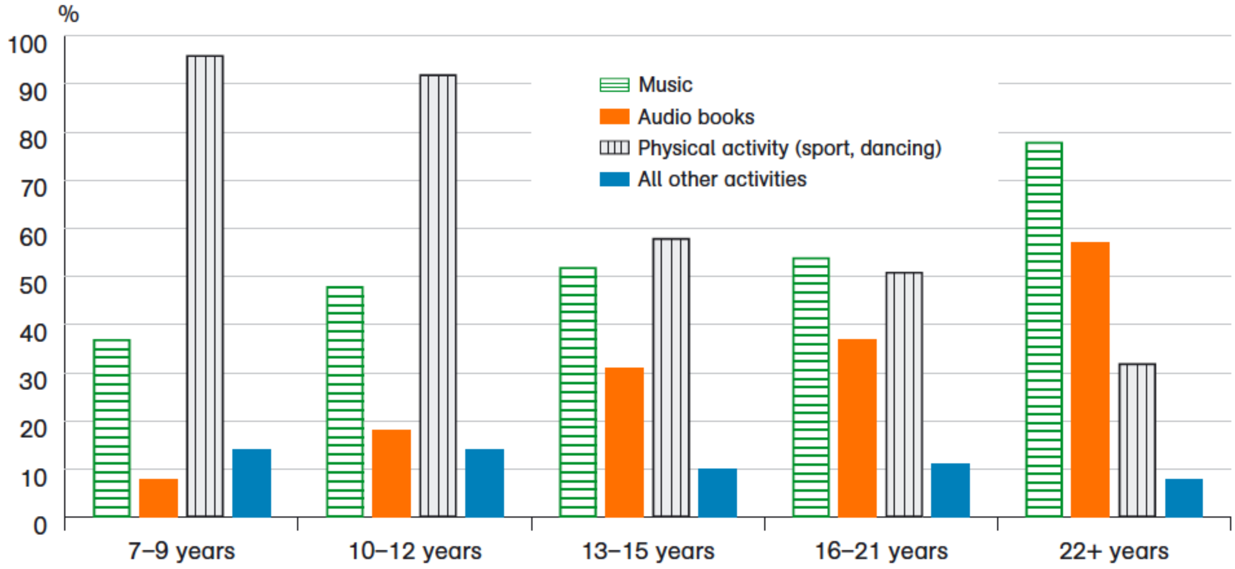
Research reviewed in this section has explored experiences from the perspective of practitioners, parents and researchers (Bills et al., 1998; B. Johnson & Jochum, 1996; Scambler, 2004; von Tetzchner et al., 2019), and has unearthed key inconsistencies and gaps in the educational knowledge, understanding and approaches for children with Batten disease (Augustine et al., 2013; Elmerskog et al., 2019; von Tetzchner et al., 2019). Research has suggested the importance of pre-emptive learning and harnessing the unique potential window of learning possibility (B. Johnson & Jochum, 1996; von Tetzchner et al., 2019). The next subsection explores the importance of music for children affected by Batten disease.

2.6. The Impact of Music for Children with Batten Disease

There are recorded accounts where music has been identified as important to for the wellbeing of a child with Batten disease (Elmerskog et al., 2019). It is suggested that for children with the Juvenile variant of Batten disease (JNCL) music can support four key areas of an individual's quality of life, comprising memory, movement, mood and language, and current research indicates that the importance of music increases with age (von Tetzchner et al., 2019). This section focuses on the recent worldwide research project by von Tetzchner and colleagues (2019), which explored the perspectives of parents and professionals supporting children with JNCL and the importance of music in their lives.

Von Tetzchner's survey (2019) found that children had a particular interest and affinity for music. Recognising the importance of music for children with Batten disease, the study presented a section of the survey dedicated to the experiences and influences of music. Figure 2.2 depicts the importance of music in comparison to other activities in the children's lifespan, derived from parental responses. Parents reported that physical activity declined with age, whereas the importance of music increased from around 36% in the age bracket 7–9 to around 50% at ages 13-15 and 16–21, and nearly 80% among those above the age of 22. These substantial changes from childhood to adolescence, and into young adulthood perhaps reflect the fact that the enjoyment of music does not rely on intellectual capacity or physical skills. A similar impact arose with the use of audiobooks (another auditory activity), and was reported to increase gradually from less than 10% at 7–9 years to over 50% above the age of 22 (von Tetzchner et al., 2019).

Figure 2. 2: Percentage of participants for whom music, audiobooks, physical activities and all other activities were viewed as important at different age levels (von Tetzchner et al., 2019: 344)



Research into music therapy provision found that 86% of parents and 78% of education professionals reported that music therapy had a high impact for individuals they supported with Batten disease (von Tetzchner et al., 2019). Furthermore, parents reflected on the specific ways in which music could facilitate social interaction, comfort, learning and understanding, stimulation and relaxation. Findings suggested key areas where music has a positive impact on a young person's life, memory, movement, mood and language (von Tetzchner et al., 2019), which are explored in detail in the following four subsections.

2.6.1. Music and Memory

Von Tetzchner's survey (2019) uncovered evidence that suggested for children with Batten disease familiar music sparked memories, and promoted comfort and wellbeing (von Tetzchner et al., 2019: 356). Parents endorsed this concept, noting that listening to music helped children to remember, for example:

> "If she went to that particular concert, she would remember what the group were wearing or doing during a particular song. She will also remember what we did before the concert or any funny things that happened. This is really important for us as parents, as it helps not only with her communication but also her long-term memory" (von Tetzchner et al., 2019: 357).

Accounts from parents in this survey drew clear links between music, memory and language, and preliminary evidence suggested that musical experiences helped children to connect with experiences, memories and enhance communication.

2.6.2. Music and Wellbeing

Identifying the positive impact of music and wellbeing, early research by Bills et al. (1998) suggested that the practice of listening to self-selected favourite music is important for emotional wellbeing and self-regulation, and can offer relief at times of psychological distress or emotional anguish. Evidence suggests that wellness and emotional wellbeing is further disrupted by the negative experience of loss and diminishing social interactions with others, placing importance on activities to improve wellbeing, and reduce anxiety, uncertainty and frustration (Bills et al., 1998; B. Johnson & Jochum, 1996). Parents in von Tetzchner's survey described how music could change a child's mood, enhance wellbeing and facilitate relaxation:

> "Music was very important to her. She liked music for relaxation, as well as music for singing along (which was functioning for an astonishingly long time). Music also evoked strong emotions, grief as well as joyfulness. Music therapy at school was one of her favourite subjects. She always liked music while being together with others, at school, with the family or on organised vacations. Music brought about a high level of wellbeing. She played her instruments, guitar and drums, and she enjoyed singing. Soft music was used to calm her down the last years she was alive (to prevent anxiety and restlessness)" (von Tetzchner et al., 2019: 346).

Parents and staff also recognised that music could be used to distract or divert during moments of personal care or pain management, or to help reduce anxiety in everyday activities like travelling in the car (von Tetzchner et al., 2019).

2.6.3. Music and Language

One symptom of Batten disease impacts the production of language (Hanson, 1993; Bills et al., 1998; von Tetzchner et al., 2019), where speech often speeds up, develops rapid stuttering, impaired articulation and dysarthria which eventually renders the speech unintelligible (Elmerskog et al., 2019). For children with Batten disease, the capacity to sing appears to remain intact, sometimes for years, beyond the point where speech is difficult or impossible to understand (von Tetzchner et al., 2019). This was reflected in perspectives from parents and educators who suggested a positive impact of music on speech. For example, one young girl spoke more clearly when singing what she wanted to say, and for another child, her “lyrics came out clearly, even though her speech was so little, stuttering and slow” (von Tetzchner et al., 2019: 348). Findings suggest that, despite declining language, children’s singing was at its best, and on par with neurotypical peers, even in their adolescent years (von Tetzchner et al., 2019).

2.7. Summary

Although research in the Batten disease field is limited, accounts demonstrate a general lack of support, policy or guidance for families, educators and carers who support children affected by Batten disease. Whilst research focuses on clinical trials, many families are left with little standardised support for their child. Recent evidence suggests that pre-emptive learning could help children harness their learning potential before deterioration sets in; however further systematic evidence is needed to create evidence-based practice for educators to draw upon (Elmerskog et al., 2019). The large sample size of the survey and geographical spread by von Tetzchner (2019) offers a wealth of information from varying countries, education, care and therapy systems, and provides one of the first opportunities to share the experiences and perceptions of parents and professionals from around the world. There is persuasive evidence from this study to suggest that music could have an overall positive impact for children with Batten disease through supporting memories, providing a functional scaffolding for language, supporting movement, and most importantly, fostering general wellbeing.

Considering evidence from this section of the literature review, this research project aims to explore the impact of music interventions for children affected by Batten disease. Addressing the gap in educational support for professionals, the project explores how a musical support resource could benefit educators and children affected by Batten disease. Given the evidence supporting the importance of lived experience, the research project implements a multidimensional approach involving the perspectives of educators, therapists, and clinicians to provide in-depth empirical evidence alongside research observations to paint a holistic picture as to the impact of music for children with Batten disease.

Evidence from relatable studies involving adults living with dementia can offer theories that explain why music has an impact for children with Batten disease and neurodegeneration. The next section interrogates research and theory relating to music and neuroscience, music and neurological conditions, music therapy and assessment and outcome measures.

2.8. The Neuroscience of Music

Pioneering work in the field of neuroscience has shed light on how music has a unique impact on the brain. Theories relating to the physical impact of music progressively offer novel scientific information, which contradicts previously established theories concerning music and the brain. With specific relevance to Batten disease, the research presented in this subsection offers insight into why music has an impact on those with neurological conditions.

Theories relating to music and the brain have become firmly established and refined across multiple disciplines. Historically, music psychologists have challenged the perception that music is processed in one location of the brain (Clynes & Nettheim, 1982; Patel, 2010) discovering that certain pathways and processes in the brain are linked with elements of music connected with hearing, senses, language, emotion and memory (Krout, 2007; Patel, 2010). There have also been links made between the autonomic nervous system and the physiological and emotional responses to music (Krout, 2007; N. Schneider, Schedlowski, Schürmeyer, & Becker, 2001). With advances in sophisticated neuroimaging, examinations have been able to pinpoint precise neurological processes that respond to isolated musical elements such as volume, pitch, rhythm and timbre (Alluri et al., 2012).

In a ground-breaking study looking at musicians' neurological processing of Tango music under fMRI, researchers found that multiple areas and neuronal networks in the brain were all actively involved in the processing of musical information (Alluri et al., 2012). Crucially, those areas governing cognitive functioning, emotion, movement and sensory information were simultaneously stimulated and activated. Although limited to participants with musical training, findings suggested that early musical training and exposure could have a physical impact on the brain. Further research, involving healthy neurotypical participants, suggested that merely engaging in musical activities could improve neuronal connectivity, promote neural plasticity, and stimulate grey and white matter changes in the brain by activating multiple neuronal networks (Sihvonen et al., 2017). Advances in this area suggest that the active process of listening to or playing music can induce multiple responses in the brain. Theories here provide a foundation of research to suggest that music can have an automatic global response on several areas of human function and could offer an explanation as to why some populations (such as children with Batten disease) display unique responses to music despite neurological impairment.

Research has also demonstrated the physiological impact of familiar music in the brain. For a neurotypically developed person, familiar music activates the areas that specifically govern memory (the anterior cingulate and medial prefrontal cortex), suggesting strong links between familiar music and memory (Särkämö et al., 2016). Furthermore, research has found that familiar music that is perceived as pleasurable can activate the dopaminergic mesolimbic system (an area regulating memory, attention, executive functions, mood, pleasure and motivation) and release the 'feel-good' hormone (Salimpoor et al., 2011). There is evidence to suggest that meaningful or familiar music could physically alter levels of neurotransmitters in the brain ultimately impacting mood, motivation, and a sense of pleasure or wellbeing. Placing these theories in the context of children with Batten disease, this research could explain why these children recall and connect with familiar songs. Evidence here demonstrates the need for further research to explore how, why, and where musical memory is facilitated, for children with Batten disease, by familiar music. Research into the impact of musical training on childhood development concluded that the experience of encoding music at an early age in childhood (such as learning a musical instrument) could significantly improve the development of functional skills such as auditory memory, speech perception, motor skills, language development and spatial reasoning (Hallam, 2006). Considering how evidence suggests that early music training may enhance neurological functioning in childhood, it could be suggested that early musical experiences could be of benefit for children with Batten disease before deterioration sets in. Further in-depth research is of course needed in this area.

In summary, the research outlined here provides insight into how music stimulates multiple parts of the brain simultaneously (Alluri et al., 2012; Krout, 2007; N. Schneider et al., 2001; Sihvonen et al., 2017) and creates neurological changes in the brain (Salimpoor et al., 2011). Currently, neuroscientific research linking music and Batten disease is non-existent, however, childhood neuroscientific research could explain how and why music impacts those affected by Batten disease (Hallam, 2006). It is possible that if music listening or active music-making can facilitate neuronal activity, then music could bypass diseased or damaged areas of the brain and stimulate neuronal connections in non-damaged areas of the brain (Thaut & Mcintosh, 2010). Metaphorically speaking, music could be the vehicle by which a traffic jam on a motorway is bypassed using an alternative route, over-riding the impairment so that functioning can resume (a concept explored later in the chapter). Familiar music could also be the key to accessing memories and experiences, acting as a memory scaffold for individuals with neurodegenerative disease. There is a need for further research to explore how familiar music might offer a meaningful context within which memories, senses and experiences could be accessed and connected simultaneously to facilitate recall, maintain memories and improve feelings of wellbeing for children with Batten disease. Expanding on the connection between music and functioning, the next section explores research around the impact of music on emotional wellbeing, memory and language, providing insight into how music can be beneficial for individuals affected by neurological deterioration and dementia.

2.9. The Impact of Music in Adults with Neurological Conditions

Despite the lack of evidence on the impact of music for Batten disease, there is a wealth of research studying the effects of music for adults with dementia and other neurodegenerative diseases. Previous research has unearthed the physical impact of music on the brain for neurotypically developed adults and those with a neurological impairment. In a randomised controlled trial, a study investigated the effects of listening to music daily for stroke patients (Särkämö et al., 2014). Results showed promising outcomes, where listening to music was seen to improve cognitive skills and language abilities by reorganising neuronal connections. This effect was greater than that observed for the participants who listened to audiobooks or a control group. This evidence suggests that music has a unique ability to impact physical changes in the brain and in some cases, rewire and reorganise damaged connections to improve functioning. If listening to music can have a positive impact for those who have suffered a stroke, it is feasible that similar re-organisation of damaged neuronal connections could be possible for children with Batten disease.

2.9.1. Emotional Wellbeing

There are many instances where music has been found to improve wellbeing for adults with dementia. In a study capturing the experiences of dementia patients, Sixsmith & Gibson (2007) found that music could bring enjoyment and pleasure, stimulate memories of past events and activities, and give people the opportunity to experience positive emotions from their past. Interview data from this study reflected that being involved in music-related activities made people feel happy and more ready to express their feelings, physically and emotionally. It was observed that even whilst discussing the topic of music with researchers, participants with dementia were visibly brighter, smiled or laughed, and in several cases, began to sing. Participants noted that music made them feel emotionally uplifted, more relaxed and less agitated. According to care staff, music also provided social opportunities for shared participation in meaningful activities with others, helped engage patients with non-verbal forms of communication and manage problematic behaviour and agitation. Although this research was based on only 26 patients, the study contributes key first-hand experiences of those living with dementia. Concerning

Batten disease, interview methods which captured rich, in-depth accounts and experiences could add value to understanding the lived experiences of affected children, and how music positively impacts wellbeing. Later research also suggests that the emotional effect of familiar music could provide comfort in a situation of confusion and disorientation for adults with dementia (Sihvonen et al., 2017).

In an All-Party Parliamentary Group commissioned report from 2017, several music-based projects and trials were found to alleviate symptoms of adult dementia, demonstrating how group singing, listening to music and playing can enhance wellbeing and quality of life for affected individuals and their carers (All-Party Parliamentary Group on Arts Health and Wellbeing, 2017). Other experts in the music and dementia field also suggested that “music can provide a true lifeline for those both with and without dementia by promoting social connection, restoring a sense of self and bringing joy even in the most challenging of times” (Bowell & Bamford, 2018:7). The positive effect of music for dementia is nationally recognised, and government guidance has recommended therapeutic interventions, such as music and dance, for those who suffer from symptoms of agitation as part of dementia (NICE, 2018a). Moreover, in the late stages of dementia, NICE advises that group activities such as music and other arts activities can help with stimulation and wellbeing, and aid reminiscence (NICE, 2019). Unfortunately, no such guidance exists for children with Batten disease, which again demonstrates the need for improved education and care policy guidelines for the neurodegenerative children’s sector.

2.9.2. Musical Memory

Whilst deteriorating memory can be the most common symptom of Alzheimer’s, growing evidence suggests that musical memory remains relatively preserved. Strong links between musical memory and language have been demonstrated in individuals with dementia who recognise and respond emotionally to familiar songs even in late stages of Alzheimer’s disease (Jacobsen et al., 2015; Särkämö et al., 2016), specifically remembering the tune and lyrics from familiar songs (Baird & Samson, 2009; Baird et al., 2017). One study demonstrated that networks associated with musical memory deteriorated more slowly than other areas of the brain in dementia (Jacobsen et al., 2015), explaining why people affected by neurodegenerative conditions may still be engaged with music despite showing symptoms of decline. Given that dementia is a symptom of Batten disease, findings such as this could have significant relevance to the Batten disease population, again providing evidence to explain why music is still able to be recognised even in later stages of the disease.

Remarkably, research has found that music can facilitate learning of new information in Alzheimer’s patients, where patients could learn and recall a new song within a short time frame (Baird et al., 2017; Samson et al., 2009). In a research study investigating the effects of music workshops and musical teaching, people with dementia could consistently recite workshop songs they had been taught up to 4 months later (Samson et al., 2009). Other research has also demonstrated the ability for adults with dementia to learn new information, evidencing the potential for both memory retention and memory recall (Baird et al., 2017). One case study of a 91 year old person living with dementia showed that after systematically teaching the individual a new song, they were able to recall song melody and lyrics up to 24 hours and 2 weeks later (Baird et al., 2017), which supported theories that the potential for musical learning can be preserved in adults with dementia (Jacobsen et al., 2015; Samson et al., 2009). Whilst the case study design of the research limits generalisations across the population, the study presents a persuasive exploration into the learning capabilities of patients with severe dementia, which demonstrates the need for more research in the field. If memory and learning remain intact for adults with dementia, for children with Batten disease, memory could be fostered through musical means, such as teaching them new songs or musical activities.

The wealth of evidence presented here suggests that for adults with symptoms of dementia, music, memory and emotion are inherently linked (Baird et al., 2017; Cuddy, Sikka, & Vanstone, 2015; Särkämö et al., 2016; Sixsmith & Gibson, 2007), and that music could incite new learning, long-term memory retrieval and autobiographical memories (Baird et al., 2017), as well as inducing positive feelings (Sixsmith & Gibson, 2007). Research here though is based on adult populations, and further studies with childhood populations could contribute further knowledge to this field. With a specific focus on patients with neurodegenerative conditions, key research also demonstrates the impact of music in maintaining language, which is explored subsequently.

2.9.3. Music and Language

Accounts from as far back as the 18th century indicate a noticeable impact of music on language, where singing surpasses language ability. One such account by Olof Dahlin (1745), demonstrated this phenomenon in a stroke patient who could reportedly only verbalise ‘yes’ in response to questions, but remarkably could sing in tune with the correct words to numerous familiar hymns. Dahlin documented this at the time:

> “Those who stutter, even if they cannot manage ten words in a row without interruption when they speak, however, can sing without impairment and with sense” (Dahlin, 1745 cited in Sihvonen et al., 2017; p. 10).

In past decades, research has identified the impact of music for Parkinson’s patients, (Harris et al., 2016). Assessors in this study, could identify speech dysprosody and impairment when listening to the speech of Parkinson's patients in comparison to neurotypical patients. However, assessors could not detect the difference between the Parkinson's patients and control group when listening to audio samples of them singing. It was as though singing eradicated speech difficulties, overriding language impairment and improving intelligibility for the participants with Parkinson's. Although not directly investigating children with Batten disease, affected children experience significant speech impairments akin to Parkinson's patients (Mole & Cotman, 2015; von Tetzchner et al., 2019). Evidence suggests that music could have a similar positive impact on a child's speech and communication, providing foundational research from which to explore similar programs for children with Batten disease.

In summary, the findings from adult research studies are important to consider, given that Batten disease and adults patients with neurological impairments present with similar symptoms and clinical presentation. The assessment measures used in the studies also offer valuable information. Rather than using neuroimaging techniques, the studies described here drew upon alternative assessment methods to track and monitor participant functionality (speech dysprosody analysis, Unified Parkinson's Disease Assessment Scale, melodic analysis, speech analysis and Speech Intelligibility Test). Neuroimaging techniques are largely invasive and difficult to administer without intrusive methods such as anaesthetic, and the clinical non-invasive, observation methods employed in these studies could be adopted for use with children affected by Batten disease to explore how they connect and respond to music.

Generally, research suggests that music has a unique impact on wellbeing (Sixsmith & Gibson, 2007), memory retrieval and learning of new information (Baird et al., 2017; Samson et al., 2009), and language improvements (Brotons & Koger, 2000; Harris et al., 2016). Theories outlined here could provide a neuroscientific explanation as to how children with Batten disease, who are seemingly unresponsive or unengaged, connect, engage with and benefit functionally from musical input. Considering the fact that individuals with Batten disease have significant challenges associated with behavioural presentation, memory loss, language deterioration and wellbeing (Bills et al., 1998; B. Johnson & Jochum, 1996; Mole & Cotman, 2015), key theories in the field of adult neurodegeneration could have significant application for individuals affected by Batten disease.

The following section considers how music therapy evidence can offer further insight into the impact for patients with neurological impairment, dementia and neurodegenerative conditions.

2.10. Music Therapy

Music therapy is an Allied Health and Care Profession that combines music with a psychological clinical supportive therapy intervention, to help people whose lives have been affected by injury, illness or disability through supporting their psychological, emotional, cognitive, physical, communicative and social needs (BAMT, 2017). A typical music therapy session might include playing instruments, receptive listening, singing, musical activities, song writing or improvisation (an example session structure can be found in Appendix 5). Training to be a Music Therapist entails a Masters level training at one of the accredited higher education facilities in the world. Training traditionally incorporates theories of attachment, psychodynamic therapy, developmental approaches, reflective practices, and concepts of therapeutic attunement. Since its conception in the 1950s, the profession has seen an evolving landscape of approaches (O'Kelly, Fachner, & Tervaniemi, 2016) and more recently, music therapy training courses have recognised the importance of neuroscientific theories with several institutions teaching the principles of neuroscience. Research in the music therapy sector has begun exploring the benefits of music therapy versus other more mainstream musical activities (such as community choirs and community music projects). Evidence suggests that it is the unique attributes of a music therapist that can create open reflective spaces in group situations, and support individuals (particularly adults with dementia) more closely in music therapy when other verbal interventions are inaccessible (Pedersen, 2017). Other researchers suggest how combined music approaches could be embedded into care pathways, suggesting a blended approach of community music interventions (such as community choirs or music listening) and music therapy in later stages of illness, particularly for adults with dementia who have lost speech (Odell-Miller, 2018).

For patients with neurological impairment, music therapy has a demonstrated positive impact (Lim et al., 2013; Sihvonen et al., 2017; Thaut, 2014), with one approach known as Neurologic Music Therapy (NMT), showcasing significant results for patients with neurodisability. Research in the music therapy and NMT field, contributes significant insight into the potential benefits for children with Batten disease, both from a psychological and functional perspective. This is explored in the following subsection.

2.10.1. The Impact of Music Therapy

Research in the music therapy field is broad and diverse, and evidence draws on multiple approaches for the different settings it serves (acute care, rehabilitation, education, community support, residential and palliative care). The recent advancements in neuroimaging have made it possible to physically observe neurological changes that occur in response to music, and consequently understand how music-based interventions can have a biomedical impact (Alluri et al., 2012; Sihvonen et al., 2017). Music therapy clinicians are therefore turning to the principles and theories of neuroscience, to evidence the physical and neurological impact of music therapy. When combined with the emotional, cognitive and behavioural outcomes observed in patient populations, the music therapy evidence-base is strengthening every year (O'Kelly et al., 2016).

Research into the effects of music therapy interventions for adult dementia has shown positive and robust results. Although the duration and the strength of the effect was unclear, in a Cochrane review of music-based interventions, randomised control trials (RCTs) suggested improved symptoms of depression and behavioural symptoms, as well as improved wellbeing and quality of life in adult patients with dementia (van der Steen et al., 2018). Other Cochrane reviews of RCTs for older people with dementia found that music, delivered as part of reminiscence therapy, was seen to assist in maintaining cognitive and physical functions for longer than would otherwise be the case (Woods, O'Philbin, Farrell, Spector, & Orrell, 2018). In the UK, research has suggested that interventions such as music therapy are regarded as beneficial for dementia (Bowell & Bamford, 2018; J. Schneider, 2018), and other studies have found that individual music therapy has a positive effect on neuropsychiatric symptoms of dementia for individuals in care home settings (Hsu, Flowerdew, Parker, Fachner, & Odell-Miller, 2015). Research has also identified the impact of music therapy on facilitating speech content and fluency for patients with dementia (Brotons & Koger, 2000). There are varied types of music therapy interventions delivered in the research here however (for example, delivering music as part of wider treatment such as reminiscence therapy, or other targeted music therapy interventions), challenging the ability to generalise findings. Additionally, the fact that there are some uncertain findings (van der Steen et al., 2018), and that the research involved adult populations, limits the relevance of the research to Batten disease. Nonetheless, the preliminary findings provide evidence supporting the effectiveness of music therapy for people with symptoms of dementia, offering a strong foundation from which further research into childhood dementia could be explored.

2.10.2. Music Therapy Home-Based Programs

It is important to consider other factors that influence the impact and uptake of music therapy services, such as accessibility, assessment and outcomes. For context, research has shown that worldwide dementia services are costly and increasing over time, with the majority of support being focused on family and social care rather than medical needs (Baker et al., 2019). Access to music therapy for adult patients is limited by availability, and there is a need for music therapists to share skills with carers and musicians in advisory and supervisory roles, to expand the benefits of music for individuals with dementia (Bowell & Bamford, 2018; J. Schneider, 2018).

Considering the wider involvement of music in dementia care pathways compared with music therapy interventions, there is evidence to suggest that, in the absence of music therapy services, other types of music-based interventions may be beneficial for those with adult dementia (Baker et al., 2019; J. Schneider, 2018). Current theories suggest specifically that music therapy could be considered for adults in the late stages of dementia and complemented by more mainstream music practices, such as choirs and community groups in the early stages of the disease (Odell-Miller, 2018). This model could perhaps be beneficial for those with Batten disease.

Music Therapist, Felicity Baker (2012) recognised the benefits of equipping caregivers to use music at home through music therapy home programs, focusing on the importance of the quality of life for both patient and their caregiver (Baker et al., 2012). Potentially unearthing a cost-effective and sustainable home intervention, music listening, singing and movement for the individual and their caregiver improved the mood of both individuals and improved spousal relationships, satisfaction with caregiving, and general caregiver wellbeing (Baker et al., 2012). As a potentially beneficial intervention in the Batten disease community, ongoing studies into music listening (Baker et al., 2019) could be relevant to improving the quality of life and wellbeing for children affected by Batten disease, as well as their families and caregivers. Other music therapy researchers have focused on singing-based therapy programs for Parkinson’s patients. Research aimed to investigate the effect of a weekly singing program, ‘ParkinSong’, to address the costly, drill-like nature of speech therapy programs for Parkinson’s disease (Tamplin, Morris, Marigliani, Baker, & Vogel, 2019). In a randomised control trial with 75 patients, weekly ‘ParkinSong’ sessions were seen to improve voice, speech, respiratory strength, and voice-related quality-of-life outcomes in comparison to control groups receiving speech and language interventions. Furthermore, the program was also seen to decrease the severity of voice problems, and lessen negative feelings about patients’ voice issues (Tamplin et al., 2019). It is possible that, for children with Batten disease, a similar complementary approach could be beneficial, which focuses on longer-term sustainable home or school-based music programs in early stages of diagnosis and incorporating music therapy in later stages of the disease.

2.10.3. Neurologic Music Therapy

Combining the theory of music therapy with neuroscientific evidence, the practice of Neurologic Music Therapy (NMT) has shown growing success for patients with neurological impairments. NMT draws upon the application of music to achieve functional changes for patients (Thaut, 2014), and research has demonstrated robust evidence that shows that there are positive changes in brain function and behaviour with this intervention (Thaut, 2014). NMT is an additional qualification for Music Therapists accredited by the University of Colorado (Thaut, 2014) and once training has been completed, therapists must evidence their work in the field through a fellowship program to practice under the title of a Neurologic Music Therapist. The approach utilises 18 techniques that combine music with other disciplines (physiotherapy, occupational, and speech therapy) to target specific functional goals in the areas of cognition, memory, speech and communication and physiology (Thaut, 2014). This approach has shown success for patients with neurodegenerative conditions such as Parkinson's and Alzheimer's disease, as well as for patients who have survived a stroke or other brain injuries (Thaut, 2014). Whilst the approach has not been directly applied to the Batten disease population, shared symptoms with adult neurodegenerative conditions suggest a potential unexplored impact for children with Batten disease.

A Cochrane review in 2017 assessed elements of NMT for brain injury rehabilitation, demonstrating improvements in functional outcomes, including movement, speech, cognition, sensory perception, and quality of life (Magee, Clark, Tamplin, & Bradt, 2017). Although focused on adult brain injury populations, the research is particularly relevant to the Batten disease population given that recent research has recognised similarities between the motor impairments (such as agitation and involuntary movement traits) of Batten disease and traumatic brain injury (Ostergaard, 2018). Research into paediatric brain injury has also shown that a combination of targeted and frequent NMT interventions improve children's outcomes compared to standard neuro-restoration interventions and therapies (Bringas et al., 2015). Children on a Neurorestorative Program (NRP) in this study, were compared to an experimental group of children who received additional intensive NMT interventions (primarily focused on attention training techniques). Results in the experimental group showed changes in brain plasticity occurring in the frontal region of the brain, measured by increased positive responses in the latency range. The 4-8 week music therapy intervention significantly enhanced children's motivation and social participation, which were assessed on clinical assessment scales. Findings from the research are limited by the small sample size (N = 34), but the use of blind assessors, control measures and randomisation increase the fidelity of these research findings. Although the participant diagnoses included a range of neurological impairments (cerebral palsy, Rett's syndrome, or spinal cord lesions), recruitment criteria excluded children with a diagnosis of neurogenerative disease, highlighting a need for research relating to the effectiveness of general NMT programs for children with degenerative disease.

Alongside research into the functional impact of music on neurological impairments (Baird et al., 2017; Jacobsen et al., 2015; Särkämö et al., 2016), NMT research in the brain injury arena could offer insight into appropriate, meaningful and translatable approaches for children with Batten disease. In the past decade, elements of NMT have been isolated and explored for various forms of brain injury. Two techniques in NMT, rhythmic speech cueing (Lim et al., 2013; Thaut, 2014), and musical speech stimulation (Straube et al., 2008; Thaut, 2014), are explored in more detail hereafter, with consideration to their relevance for children with Batten disease.

Rhythmic Speech Cueing (RSC)

One of the key principles of NMT, rhythmic entrainment, taps into the brain’s ability to unconsciously entrain to rhythm whilst naturally processing and responding to music (Sihvonen et al., 2017). Rhythmic entrainment is most commonly understood where one may unconsciously tap along to music or instinctively march in time. It is a unique physical effect that happens largely automatically. Rhythmic entrainment creates a close connection between the auditory and motor system, and in NMT, the concept of rhythmic entrainment has also been applied to communication to facilitate improvements in speech impairments (Thaut, McIntosh, & Hoemberg, 2015).

A technique called Rhythmic Speech Cueing (RSC) utilises rhythmic entrainment to initiate and control the rate of speech through cueing and pacing. To achieve the entrainment effect, a Music Therapist uses a physical pulse (hand tapping, use of a metronome or drum) to prime speech patterns into a metered structure (Thaut, 2014). Research has demonstrated the positive impact of RSC with stroke survivors demonstrating observed improvements in symptoms of dysarthria, aphasia or speech fluency (Lim et al., 2013). Although based on a small sample size (N = 21), the findings from Lim’s study are relevant to children with Juvenile Batten disease. For children with JNCL, communication impairments appear later in the disease, causing frustration, depression and demotivation (Bills et al., 1998; Scambler, 2004). There is evidence, therefore, to suggest that programs incorporating RSC could help support a child’s deteriorating communication skills.

Musical Speech Stimulation (MUSTIM)

A second approach targeting language, Musical Speech Stimulation (MUSTIM), also harnesses the impact of rhythm and entrainment in music by using musical rhymes and chants to stimulate speech (Thaut, 2014). MUSTIM utilises anticipation in musical phrases to facilitate phrase or sentence completion in melodic phrases or songs. For patients with aphasia, research has shown that singing familiar songs helps with word completion (Straube et al., 2008). Although the research lacks certainty around the causes for improved singing in aphasic patients, this technique could have significant relevance for the Batten disease population in that it crosses multiple clinical areas of memory and language simultaneously. As an under-researched area of NMT, further research is needed to ascertain the effectiveness of MUSTIM in adult or child populations, and more specifically the impact of MUSTIM on children affected by deteriorating communication or neurodegenerative disease.

2.10.4. Summary

The demonstratable impact of music therapy and NMT research supports previous neuroscientific perspectives into how music is processed in the brain (Alluri et al., 2012; Harris et al., 2016; Sihvonen et al., 2017). There is evidence to suggest that music therapy-based home programs could improve the quality of life for adults with dementia, and offer a cost effective support resource for carers (Baker et al., 2012). Practice-based research demonstrating the effectiveness of NMT, combines theory from music and neuroscience research with music therapy practice to deliver music-based rehabilitation programs for patients with neurological impairment (Thaut, 2014). Furthermore, specific isolated components of NMT practice can enhance functional rehabilitation for adults, and in some cases, paediatric brain injury patients or those with neurodisability (Bringas et al., 2015; Lim et al., 2013; Straube et al., 2008). What is missing, however, is evidence as to whether NMT can support patients with neurodegenerative conditions, such as Alzheimer's or Batten disease, where the focus of therapy is on maintenance, stasis, and compensation rather than improved outcomes over time. As with other populations, a sequence of NMT techniques could provide an opportunity for affected children to practice and maintain their speech.

Considering the evidence reviewed in this section of the literature review, this research project begins an investigation into the impact of music therapy interventions. Beginning first with one-to-one music therapy sessions, the project explores how functional responses and skills change in music therapy over time. Considering the possibility of music as part of the care and education pathway, the project explores the use of music therapy-based programs for children in their school environments, incorporating elements of NMT practice to support the maintenance of communication and speech.

Although research in the music therapy and NMT sector demonstrates the positive impact of music therapy, there are challenges in quantitatively evidencing the impact of such interventions for children with Batten disease. The last subsection explores existing assessment and outcome measures in the field, and how future research can contribute to developing these further.

2.11. Assessment and Outcome Measures

In Batten disease research, assessment measures lack generalisability across all disease variants and more appropriate measures are needed to capture the diversity of the population (Augustine et al., 2013; von Tetzchner et al., 2013). There are consistent challenges in quantifying psychological therapies, or interventions that support emotional improvement and wellbeing, and although new advancements in neuroimaging provide insight into the action mechanisms evident in music therapy sessions (O'Kelly et al., 2016), assessment measures are still under development concerning the neurodegenerative population (O'Kelly & Bodak, 2016). Research for adults with dementia has shown that music-based assessment measures (in comparison to standard clinical tests) are able to capture additional responses for individuals that other assessments exclude (Hsu et al., 2015). Other studies have illustrated the possibility of assessing the impact of music therapy for neurological conditions such as Huntingdon's disease (O'Kelly & Bodak, 2016), patients with Disorders of Consciousness (Magee et al., 2016) and dementia (McDermott et al., 2015), but none are appropriate for tracking the impact of music therapy with paediatric populations. Research has shown that there are significant neurological differences in the processing of music between adult and paediatric populations, and adult-based assessments and treatments (in particular brain injury populations) should not be automatically applied to child populations (Wilke, Schmithorst, & Holland, 2003). To gather meaningful data concerning outcomes in music therapy, research must first develop appropriate assessment measures for childhood neurodegenerative conditions, which, in turn, could significantly contribute value to research in the field and assess the effectiveness of the music therapy interventions.

In the music education and therapy sectors, there are other assessment measures that are relevant to children with additional needs and visual impairment. Firstly, the Sound of Intent (SOI) framework (Vogiatzoglou et al., 2011) has been developed as a music education measure to track children's musical abilities (where more conventional music examination frameworks are inappropriate). The measure aims to track musical abilities and engagement in the three domains of Interactive, Proactive and Reactive areas with each domain containing a six-point subscale scoring system. The framework, trialled previously for children with Batten disease (Ockelford et al., 2019), offers a systematic framework to capture musical abilities for children. However, the measure is firmly rooted in a music education framework, and focuses on musical outcomes (playing, repertoire, performance and musical techniques) rather than broader functional skills in response to music (mobility, cognition, recall, self-expression etc). It is important that assessment measures employed in this project and in future research, focus on capturing these latter responses to music and music therapy, given the influencing theory and context surrounding the impact of music on functional skills, namely wellbeing, memory and language (Baird et al., 2017; Harris et al., 2016; Sixsmith & Gibson, 2007).

Other functional therapy measures, such as mobility scales (Harvey, Morris, Graham, Wolfe, & Baker, 2010), speech and dysarthria assessment scales (Enderby, 1980), or intelligence testing (Kaufman, Flanagan, Alfonso, & Mascolo, 2006), can assess individual elements of a child's functionality. However, none are able to capture multiple areas of functionality simultaneously, and many need to be adapted for use with the Batten disease population (Ragbeer et al., 2016). A more holistic framework, the Therapy Outcome Measure (TOMs, (Enderby & John, 2019) assesses the impact of an intervention (such as music therapy) using a framework that encompasses four domains of Impairment, Activity, Participation and Wellbeing. The tool is intended for use before and after an intervention, or at consistent review points in a patient's care. Whilst it focuses on functional responses to interventions (an important consideration in this research project), recent validation studies in community adult populations indicate the measure is not the best fit for capturing sensitive, small improvements in patient groups who present with multiple pathologies and symptoms, and therefore, it is recommended that is used alongside other clinical recording measures (Caldwell, Twelvetree, & Cox, 2015).

The absence of an appropriate and relevant assessment in the music therapy field highlights a need for development work in this area, calling for appropriate measures to observe and track responses in music therapy for children with neurodegenerative diseases. Given the current disjointed and isolated care and therapy approaches for Batten disease (Augustine et al., 2013), a unified assessment measure could align health professionals and therapists to capture similar responses in therapy or education work. Through an appropriate assessment measure, the value, cost-effectiveness, and impact of music therapy input could also be robustly evidenced, offering the opportunity for music therapy to be considered in care and therapy support packages for affected children. It is an aim of this research project, therefore, to explore and identify an appropriate method by which responses in music therapy can be captured.

2.12. Conclusion

The review of literature in chapter two has unearthed key evidence and theory relating to the reality and experience of those affected by Batten disease, the importance of educational approaches and guidance, music and neuroscience, music therapy, and music-based assessment measures. Key learning points are summarised as follows:

1. The effectiveness of pharmacological interventions for children living with Batten disease is limited, calling for improvements in education and therapy support (Lewis et al., 2019; von Tetzchner et al., 2019)
2. Current health policy, care, support and education pathways are lacking for children affected by Batten disease (Scambler, 2004; von Tetzchner et al., 2019)
3. Music stimulates multiple parts of the brain simultaneously (Alluri et al., 2012; Krout, 2007; N. Schneider et al., 2001; Sihvonen et al., 2017), creates neurological changes in the brain (Salimpoor et al., 2011) and could bypass diseased or damaged areas of the brain (Thaut & Mcintosh, 2010).
4. Evidence suggests the importance of music on functional skills for affected children (von Tetzchner et al., 2019)

5. Music can influence wellbeing, speech and memory for patients with neurological conditions (Harris et al., 2016; Salimpoor et al., 2011; Samson et al., 2009).
6. Home programs and music therapy, including Neurologic Music Therapy, could be an effective intervention for brain injury and neurodisability patients (Baker et al., 2012; Bringas et al., 2015; Lim et al., 2013; Straube et al., 2008)
7. Outcome measures in music therapy need to be developed to incorporate neurodegeneration in children (Augustine et al., 2013; Magee et al., 2016; McDermott et al., 2015; O'Kelly & Bodak, 2016).

Research has shown that music holds significance for individuals with Batten disease (Elmerskog et al., 2019; von Tetzchner et al., 2019). It is feasible that the positive impacts of music for adults with dementia or neurodegenerative conditions could be mirrored in those affected by Batten disease. To date, however, there is a lack of systematic evidence to sustain this hypothesis. Building on the evidence from the literature review, this research project began by addressing the limited assessment measures in the field for Batten disease (Augustine et al., 2013; von Tetzchner et al., 2013), by developing and evaluating a music therapy assessment measure for children affected by the disease. The research project then used data from a clinical assessment scale to explore the differences in functional outcomes for children with Batten disease from a clinical perspective (Kohlshutter et al., 1988). Thereafter, the project, aimed to explore the functional responses and skills of children with Batten disease in music therapy sessions. Based on previous literature in the music therapy (Harris et al., 2016; Salimpoor et al., 2011; Samson et al., 2009) and the Batten disease arena (Elmerskog et al., 2019; von Tetzchner et al., 2019), this first phase focused on communication, mobility, wellbeing, cognition and behavioural responses in music therapy sessions.

Considering the evidence establishing the impact of NMT for children with neurodisability (Bringas et al., 2015), and the positive impacts of music-based home programs (Baker et al., 2012; Tamplin et al., 2019), it could be suggested that creating a bespoke music education program could benefit children with Batten disease in educational environments. Furthermore, evidence indicating the effectiveness of early intervention and pre-emptive learning for affected children (Elmerskog et al., 2019) suggests that educational music and language-based programs, implemented in the early stages of the disease, could help maintain communication and language skills for longer. Whilst, in theory, embedding early interventions could be beneficial, the true challenge is successfully executing a proactive education strategy that is accessible, realistic and sustainable for all involved (von Tetzchner et al., 2019). Therefore, any school-based music therapy program must be accompanied by specialist training, guidance and education as well as consideration given to the experiences and resources of professionals supporting affected children. Thus, drawing on literature concerning the use of music-based home programs (Baker et al., 2012; Tamplin et al., 2019) and the impact of NMT for neurodisability (Bringas et al., 2015; Lim et al., 2013; Straube et al., 2008), the second phase of the project aimed to translate the outcomes observed in music therapy sessions, into a school-based program for children with Batten disease, Music Speaks.

Crucially, this literature review has identified a general limitation in the breadth and quantity of research into the experiences of those affected by Batten disease, including the child themselves, and their parents, carers, educators, and therapists. To examine the full landscape of the research topic means traversing beyond the empirical evidence observed in neuroscientific studies and randomised control trials. From the position of a critical realist, experiences, perspectives and contexts demonstrate that the research cannot simply analyse the effect of music for affected children in a linear cause and effect fashion, but needs to consider interlinking concepts, challenges and absences that generate connected and influencing mechanisms (Bhaskar & Danermark, 2006; Porter et al., 2017). For example, the lack of guidance and inconsistency of resources at an institutional, governmental and international level (Augustine et al., 2013; NINDS, 2020; Together for Short Lives, 2019; von Tetzchner et al., 2019) highlight the need for this research to deepen the understanding of these challenges, framing empirical evidence within the societal constructs which influence the lived experience of those affected by Batten disease. Whilst research reviewed in this literature review has presented independent theories on the impact of music, no research has yet drawn links with these societal causal mechanisms and contexts. The positioning of critical realism offers a framework through which multiple levels can be explored (Fletcher, 2017), ultimately ascertaining what is happening, how, in what contexts, and to whom.

The third phase of this research, therefore, captured the experiences and perceptions of education and therapy professionals who support children with Batten disease in an educational context. Drawing on literature relating to educational approaches (Elmerskog et al., 2019; von Tetzchner et al., 2019), the experiences of carers (Kitwood, 1997; Scambler, 2004), and the current health and care pathways for children with Batten disease (NINDS, 2020; von Tetzchner et al., 2019), interviews with professionals aimed to understand the various influencing mechanisms, observations, perceptions and contexts, contributing a holistic picture of the influence of music interventions for children with Batten disease. Ultimately, findings in this doctoral research contribute towards a wider enquiry into the effectiveness of music therapy for children with Batten disease and the translatability of its perceived impact into other education and therapy contexts. Chapter 3 outlines the epistemological standpoints that provide the framework for the research approach and methods implemented in this doctoral project.

Chapter 3: Epistemology

"Music is… a higher revelation than all wisdom & philosophy"
- Ludwig van Beethoven

3.1. Introduction

Many music therapy researchers acknowledge that research approaches are influenced by the epistemological and ontological standpoint of the researcher, yet very few studies reference the researcher's epistemology (Edwards, 2012). This makes it an important concept to discuss and explore within this doctoral research project. In the social sciences, some researchers suggest that more pluralistic approaches are the most effective way of developing an exploratory enquiry which can be achieved through methodological designs that combine multiple perspectives from blended qualitative and quantitative research methods (Edwards, 2012; Mcevoy & Richards, 2006; Tashakkori, Teddlie, & Sines, 2012). Having previously approached research from a positivist stance (during earlier undergraduate training in psychology and music), the consideration of epistemological positioning has matured throughout the doctoral training. This chapter critically reflects on the epistemological journey I have taken on this research, considering how this has influenced the pluralist methods used.

3.2. Epistemology

Prior to this doctoral research, my degree in psychology and music led me to approach research from a logical positivist approach, believing that this enabled access to verifiable, clearly defined, empirical data (Edwards, 2012; Tashakkori et al., 2012). The positivist and quantifiable approach offered value by reducing ambiguity, improving clarity and offering robust approaches to exploring abstract paradigms (Edwards, 2012). This was also echoed in my experiences working as a Music Therapist, where funding for services depended wholly on robust, evidenced outcomes, quantifiable results and measurable improvement or change. The profession of music therapy is often underestimated, undervalued and consequently underfunded, driving the demand for more cost-effective, evidence-based quantifiable data from the research sector (Bruscia, 1995). Music therapy beneficiaries however, come from a diverse heterogeneous population varying in age, diagnosis, cultural background, economic background and therapy need. Epistemology needs to drive research to delve beyond surface level patient outcomes data and incorporate individual experiences, therapy needs, and societal circumstances (Edwards, 2012). Many researchers recognise that therapy and healthcare research demands an approach that goes beyond the confines of a purist qualitative or quantitative design (Bruscia, 1995; Edwards, 2012). It is essential to deeply understand the experiences of those fragile populations accessing music therapy, and the influencing factors that impact the therapy and healthcare they have access to. This is in combination with quantifiable information about outcomes, impact and therapeutic progress. Other music therapy and Batten disease research studies have recognised how mechanisms and constructs can influence a research enquiry (Porter et al., 2017; Scambler, 2004). Within this doctoral project I also recognise the importance of my researcher experiences, values and perspectives that have influenced the direction of the research enquiry (Fletcher, 2017).

The research strategy, therefore, considers how experiences and perspectives influenced and connected with the quantitative data that were collected from the research observations. Simply put, it is not just their lived experiences of participants, but the reality of their experience – their ‘lens’; their perspectives of education; and the support and social systems – that contribute towards an understanding of the impact of music in their lives. The position of critical realism offers this type of pluralist positioning that allows for deeper insight into the lived experiences of both the participants and the researcher – valuing both fidelity and quantifiable data (Fletcher, 2017; Porter et al., 2017). This project therefore takes a critical realist approach.

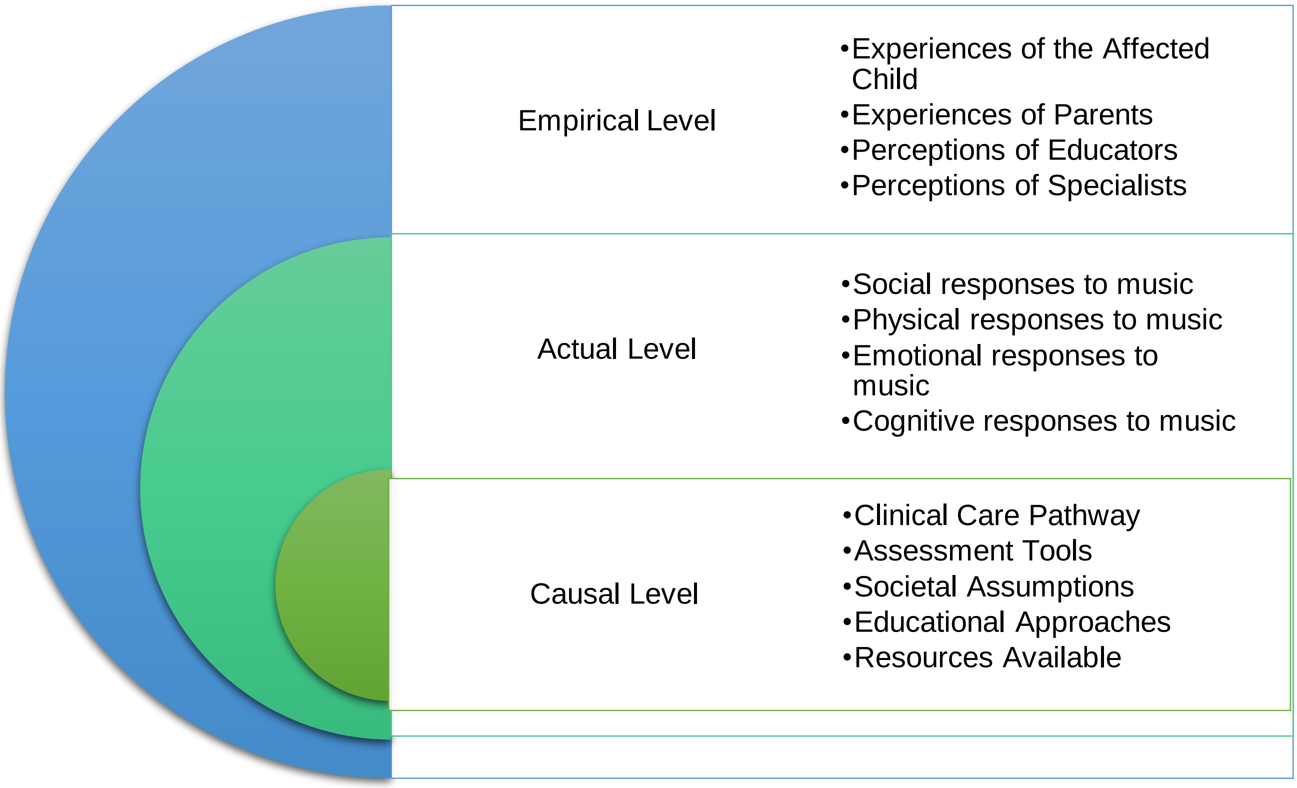
3.2.1. Defining Critical Realism

Recognising the relevance of both a realist ontology and a relativist epistemology, critical realism appreciates that unobservable constructs can cause observable realities (Bhaskar & Danermark, 2006; Fletcher, 2017). Critical realism emphasises that research into social paradigms must simultaneously investigate the underlying constructs behind the realities (Danermark, Ekstrom, Jakobsen, & Karlsson, 2017). The critical realist position proposes that reality is measurable, but this reality is only measurable within the parameters of the causal social structures and influences linked to the participants, populations, cultures and researchers themselves (Danermark et al., 2017). Moreover, critical realism acknowledges the role, the perspective and the ‘lens’ of the researcher, and the participants, as well as external and social factors at play (Porter et al., 2017). Allowing the nature of constructs and interactions to be investigated case-by-case without alliance to a specific methodology, critical realism offers plurality, flexibility and inclusivity (Archer et al., 2016). It accepts and utilises mixed methodologies, holistic theories, concepts, and approaches, allowing researchers to acknowledge and recognise the multi-dimensional perspectives within a research topic (Tashakkori et al., 2012). Research from a critical realistic standpoint therefore allowed an exploration into the unique impact of music for children with Batten disease, as well as simultaneously building an understanding of the lived experiences of the children, those who support them, and the researcher.

The methodological framework of the research project began by mapping out the key overarching and connected concepts and mechanisms considering the empirical evidence alongside social events and causal mechanisms which could influence the experiences of those living with Batten disease (Danermark et al., 2017; Fletcher, 2017; Porter et al., 2017). Specifically, considering the existing neuroscientific literature around the observable benefits of music and dementia (Baird et al., 2017; Harris et al., 2016; Thaut, 2014), with underlying perspectives, experiences and societal constraints of those living with and supporting children with Batten disease (Elmerskog et al., 2019; Scambler, 2004; von Tetzchner et al., 2019), enabled the project to explore and represents multiple aspects of the research topic.

Figure 3.1 presents a visual map of these initial overarching mechanisms and concepts that were unearthed from the literature and theory. Observations here identify *outcomes* (actual level) and behaviours in the presence of music and within music therapy (i.e. the ability to sing when language has been lost). *Contextual influences* (at a real, causal level) relate to inconsistent education and therapy approaches (von Tetzchner et al., 2019), undependable care systems (Together for Short Lives, 2019), societal assumptions (Bills et al., 1998; B. Johnson & Jochum, 1996; Scambler, 2004), and inconsistent assessment or resources (O’Kelly & Bodak, 2016; von Tetzchner et al., 2019), which could present contrasting realities for those affected by Batten disease. Thereafter, the literature additionally suggests that the *experiences* (empirical level) of children with Batten disease, their carers, families and educators, could contribute unique perspectives to the understanding of the disease, unearthing unheard narratives and observations of the research phenomenon, alongside influencing contexts, mechanisms and factors (Kitwood, 1997; Scambler, 2004; von Tetzchner et al., 2013). This is further married with my professional clinical experience as a Music Therapist and researcher, explored in the following reflexivity section.

Figure 3. 1: Identified interactions of mechanisms relating to the impact of music for a child affected by Batten disease



3.2.2. The Value of Reflexivity

Adopting the perspectives of a critical realist meant paying attention to the influence of my researcher position. In accordance with multiple reflective practice techniques, the research process recognised the influencing ‘lens’ I hold as both clinician and researcher, and the underlying importance and value of relationships that were present in all dimensions of this research journey (Atkins & Murphy, 1993). There have been key influential research projects and professional roles that have provided me with a foundation of learning and experience in supporting children with Batten disease (Elmerskog et al., 2019; Ockelford et al., 2019; von Tetzchner et al., 2019). Diving deeper into researcher perspectives meant critically interrogating how the multiple and fluid roles I held in different contexts and research projects influenced my research approach, and the connection with populations I have worked with. The vulnerable and fragile nature of the participants involved in the research evoked multiple feelings, thoughts and reflections in me as the research process progressed. Writing a reflective journal was fundamental to the research process, allowing the multiple roles, thoughts, feelings and ideas to be documented, explored and reflected upon (Edwards, 2012). My training as a Music Therapist also supported this reflective process, acknowledging my thoughts, ideas and feelings about the fragile participants and the sensitive research topic (Hewitt, 2017). Aligning with other music therapy research and social research methods (Edwards, 2012; Guion et al., 2002; Onwuegbuzie & Leech, 2005), reflective practices were just one aspect which added strength and plurality to the project (discussed further in chapter eight).

Fundamentally, my research position as a critical realist recognises that multiple perspectives could connect and influence multiple mechanisms, structures, processes, experiences and observations which, thereafter, contribute to and shape an existence of knowledge and truth. Knowledge is out there waiting to be discovered, but the reality of this knowledge goes beyond observable events – it connects more meaningfully to underlying causal structures and perspectives that are present in research phenomena (Danermark et al., 2017). The full landscape of the research topic for children with Batten disease therefore involved emergent and inductive processes, which allowed a dialogue to form between the complex and multiple layers of data.

Chapter 4 discusses the research phases in more detail, outlining the methodological approach, strategies and interventions, and providing an overview of data collection and methods of analysis.

Chapter 4: Methods

"Musical training is a more potent instrument than any other, because rhythm and harmony find their way into the inward places of the soul"
— Plato

4.1. Methodological Approach

The research implemented a flexible, mixed methods design, utilising the strengths of both quantitative and qualitative data collection methods and analysis to meet the needs of the research aims (Bruscia, 1995; Tashakkori et al., 2012). This design enabled the research enquiry to gather systematic data about the effects of music for children affected by Batten disease, whilst also collecting a wider representation of the impact, challenges and social realities of the affected children, workers and families. Furthermore, this project was able to draw upon insights and theory relating to causal societal and educational mechanisms and perspectives that also influenced the research enquiry.

Firstly, as identified in the literature review chapter, existing research and evidence into the impact of music therapy and education strategies for affected children are limited. Therefore, this doctoral research used an inductive research approach to formulate and construct concepts and themes from the raw data (Tashakkori et al., 2012; Thomas, 2006). Secondly, a small-scale case study approach was used to enable this research to gather and formulate multi-dimensional perspectives. This was carried through each of the phases of data collection. To begin with, following an exploratory design, the research project focused on the manifestation of the phenomena (unique musical abilities observed in affected children) through longitudinal assessments. This evolved into the employment of semi-structured interviews, to enable an in-depth exploration in perspectives, concepts and themes around the lived experiences of supporting a child with JNCL, and professionals' perceptions of delivering the program. This inductive approach concluded with a review of the empirical data and themes, which helped construct a dialogue linking the mechanisms, constructs, observations and experiences from each research phase (Bhaskar & Danermark, 2006). Finally, in line with music therapy and social research methods (Edwards, 2012; Guion et al., 2002; Onwuegbuzie & Leech, 2005), the research incorporated an array of measures, such as triangulation, longitudinal observations, inter-rater reliability, face validity, and the use of reflexive techniques, to add strength and robustness to the conclusions of this research. The following section outlines the methods for each of the three phases of the research, detailing participants, interventions, assessment measures, data collection measures, and analysis methods.

4.2. Methods

The project followed three distinct phases that explored the following research aims:

1. To examine the ways in which children affected by Batten disease respond to music therapy, focusing on social, emotional, physical and cognitive outcomes.
2. To explore the ways in which children affected by Juvenile Batten disease respond to a daily music education program within their school setting.
3. To illuminate the experiences of educators and therapists who have supported children affected by Juvenile Batten disease using the Music Speaks program, within UK education settings.

The sequential nature of the methodological design enabled the research to draw upon inter-related quantitative data and qualitative data collected from each phase, which informed the subsequent research phases. Additionally, themes and concepts highlighted in the final interview phase helped synthesise the research data within the three dimensions of *empirical, actual* and *causal* level evidence (Fletcher, 2017; Scambler, 2004), facilitating a dialogue into different levels and perspectives. The methods from the three phases are defined in the following subsections.

4.2.1. Phase One

To explore responses to music therapy, the research project aimed firstly to explore how music therapy assessment measures could enable outcomes to be tracked in neurodegenerative populations. With regards to appropriate music therapy assessment measures already used in the sector, existing tools focused on other adult neurological conditions such as Huntingdon’s disease (O’Kelly & Bodak, 2016), patients with Disorders of Consciousness (Magee et al., 2016) and dementia (McDermott et al., 2015); none of which were appropriate for use with the paediatric population. Other measures excluded the possibility of measuring functional outcomes, focusing solely on music development (Vogiatzoglou et al., 2011). Therefore, this research focused on exploring a prototype music therapy assessment measure (CMTOM) to track functional responses in music therapy sessions for children affected by Batten disease (research question 1.1).

The first stage of Phase One aimed to determine:

i. the appropriateness of the measure for children affected by Batten disease (through a music therapy practitioner questionnaire);
ii. the face validity of the measure (again through the music therapy practitioner questionnaire); and
iii. reliability of the measure for use in music therapy sessions (through an inter-rater exercise).

Subsequently, to explore responses in music therapy, Phase One used data I collected from September 2016 and March 2019 as the lead researcher from a study investigating the influence of music for children affected by Batten disease ('MIND', Ockelford, Atkinson, & Herman, 2019). The MIND study collected and analysed data on children in their music therapy sessions using the Sounds of Intent framework (Vogiatzoglou et al., 2011), but did not include any analysis of music therapy assessment data. Data collected from the Hamburg Clinical Scale (Kohlshutter et al., 1988) provided a clinical perspective on how these functional responses changed over time (research question 1.2). The data collected from the CMTOM assessment measure from the preceding study were then analysed by two assessors to understand functional responses in music therapy, how these changed over time (research question 1.3) and how these scores sat alongside scores from the clinical measure. Within this first phase of this doctoral project, the three research questions were addressed using data from the same 12 participants over the three years. The participants and intervention are outlined first, before defining the differing assessment measures and stages of analysis for each research question.

Participants

Using a voluntary response (convenience) sampling strategy (Navarro Sada & Maldonado, 2007), twelve children aged between 3 and 17 years (SD = 4.2) with Batten disease (identified as female [n = 5] and male [n = 7]) were recruited through the Batten Disease Family Association (BDFA, www.bdfa-uk.org.uk), using an email campaign in 2016. Due to the fragile and rare nature of Batten disease, there were no specific exclusion criteria with the recruitment of participants. Recruitment aimed to include any child between the ages of 0-18 with a confirmed medical diagnosis of Batten disease (see Appendix 3 for staff information consent form and Appendix 4 for parent/ guardian information consent form). From the fourteen variants of Batten disease presently known, the participants represented five different variants of Batten disease. Table 4.1 represents the participants in this first phase. As the Batten disease population in the UK is very small, to protect anonymity, identifying information relating to participants' gender and age has been removed from this table.

Table 4. 1: Participants from the first phase of the research

Participant	Disease Variant	Notes
1	CLN6	Participated for all 3 years
2	CLN2	Participated for 1 year
3	CLN5	Participated for all 3 years
4	CLN3	Participated for all 3 years
5	CLN5	Participated for all 3 years
6	CLN3	Participated for all 3 years
7	CLN2	Participated for 1 year
8	CLN3	Participated for all 3 years
9	CLN2	Participated for all 3 years
10	CLN2	Participated for all 3 years
11	CLN2	Participated for all 3 years
12	CLN8	Participated for all 3 years

Music Therapists (N=11) and one music practitioner were involved in the delivery of music therapy sessions and music sessions over the three years. One music practitioner was involved in this phase as they were already supporting one of the participants in this research phase. Their sessions were similar in content to sessions delivered by the Music Therapists, but the music practitioner was a music teacher rather than a credited Music Therapist. All practitioners were sourced through the Amber Trust (https://www.ambertrust.org/). Ahead of the music therapy sessions taking place, practitioners submitted evidence of their music therapy qualification (where relevant) and a copy of their DBS check. They were sent supplementary materials which included consent forms, a background to the research, articles on education strategies for children with Batten disease, a film schedule detailing which weeks to record sessions throughout the year, and a suggested session plan (see Appendix 5).

Intervention

Participants received weekly music therapy sessions from September 2016 to June 2019 from registered independent Music Therapists and a music teacher. Sessions were delivered either at their home or in an education setting following the academic calendar (average 120 sessions per year, per participant). In line with music therapy practice, sessions lasted up to an hour (variable on the child's presentation at each session time) and focused on a suggested session guiding framework to facilitate speech and language, cognition, creativity, movement and wellbeing (see Appendix 5). Music therapy sessions did not follow a set protocol, however as new resources emerged, these were shared with practitioners (such as new technological devices or activities). Although a suggested session plan was supplied to music therapists and practitioners, in line with the principles of music therapy practice (HCPC, 2013), practitioners set their own therapeutic objectives and activities depending on the child, their preferences and their abilities. Sessions, therefore, differed in terms of songs, musical content, activities and instruments.

Research Question 1.1

> *For children with Batten disease, how can functional outcomes be tracked and measured in music therapy?*

Assessment Tools

Chiltern Music Therapy Outcome Measure

Considering the complexity of the population and the absence of appropriate tools for children affected by Batten disease, this first phase aimed to explore the appropriateness of the Chiltern Music Therapy Observation Measure (CMTOM, Atkinson, 2018, Appendix 1) for identifying and tracking functional responses in music therapy sessions. The CMTOM was developed within my role as a Music Therapist alongside other Chiltern Music Therapy practitioners, to capture behaviours and skills in a music therapy session. The tool is intended for multiple populations, but specifically enables skills to be tracked over time, so that it can be used with neurodegenerative populations. Focusing on observations in the domains of Communication, Behaviour, Emotional expression, Cognition and Mobility, the measure uses a 5-point rating scale to track the prevalence of observed skills or behaviours in sessions. Table 4.2 shows an example of the communication domain from the CMTOM measure.

Table 4. 2: Coding scores for observed instances of skills or behaviours in the communication domain of the CMTOM measure (Atkinson, 2018)

Observations	**DATE**
Mark as 0 (never), 1 (rarely), 2 (sometimes), 3 (often) or 4 (consistently)	**Score**
Communication & Social interaction	
Appropriate eye contact or tracking of visual stimuli	
Appropriate use of gesture	
Vocalisation (any sound)	
Verbalisation (use of speech)	
Singing	
Awareness of others	
Ability to interact non-verbally / verbally	
Interaction with staff	
Ability to Initiate interactions	
Behaviour / music to therapist appropriate?	
Notice, tolerate, accept, aware of others	
Ability to participate / join in	
Sharing emotions, thoughts and ideas	
Being able to think about others - show empathy	

Assessment Measures

To initially capture an element of construct validity of the measure (Navarro Sada & Maldonado, 2007; Wuensch, 2021), face validity was deemed an appropriate domain to examine agreement between professionals as to whether the CMTOM measured the construct of functional responses in music therapy over time. To determine the face validity of this measure for the participant group, the questionnaire was designed to be completed by practitioners in the study (N = 6). The ten-item questionnaire aimed at gauging feedback on the appropriateness and relevance of the measure for children with Batten disease (see Appendix 6). Items in the questionnaire related to whether (and to what extent) the CMTOM measured communication, cognition, mobility and wellbeing responses in music therapy. The questionnaire also determined the extent to which the assessment measure needed training to use, whether it would be recommended to other practitioners, its use for retrospective analysis and the suitability of the measure alongside other assessment tools.

Items were worded to indicate that agreement with them represented a high value in the measure attribute (Wuensch, 2021), for example, agreement with the statement “The observation tool measures adequate elements of speech, language and communication” indicated a high level of agreement that elements of communication and language were being measured in the CMTOM. The questionnaire included Likert scale responses for each item to ascertain the level of agreement (ratings as follows; A=Strongly Disagree, B=Disagree, C=No Opinion, D=Agree and E=Strongly Agree).

Additionally, to further strengthen the appropriateness and reliability of the measure, the CMTOM was used by two assessors on the research team (including me), simultaneously over 3 months. To strengthen the reliability of ratings, an inter-rater reliability exercise was conducted. After one jointly rated video session of a participant using the CMTOM, an initial agreement was reached between the assessors. Videos (N = 182) were then anonymised and numbered, and ten randomly selected videos were selected for assessment by both researchers. Ratings were compared to achieve coefficiency of the CMTOM between the two assessors. The use of an inter-rater exercise and practitioner questionnaire aimed to examine whether and how the CMTOM was an appropriate measure in tracking and monitoring functional outcomes and responses in music therapy sessions for children affected by Batten disease.

Analysis

The first steps of analysis drew upon the practitioner questionnaire data and the inter-rater reliability exercises to assess the validity and reliability of the CMTOM measure, with a specific focus on relevance for the Batten disease population (research question 1.1.). Responses were scored respectively using the following scoring system to indicate criteria of agreement:

1 (A) = agreeing indicates the respondent is very low in the measured attribute
2 (B) = agreeing indicates the respondent is below average in the measured attribute
3 (C)= agreeing does not tell anything about the respondent’s level of the attribute
4 (D) = agreeing indicates the respondent is above average in the measured attribute
5 (E)= agreeing indicates the respondent is very high in the measured attribute

Analysis of the scores focused on descriptive statistical data on the number of respondent scores per item. From a clinical perspective, for the measure to be used in subsequent phases of the research project, an overall majority of responses to questions (80% or over) would need to score 4 or 5 on the scoring system.

Data from the inter-rater reliability exercise were analysed using the Cohen's Kappa and Intraclass Correlation Coefficient tests, to determine consistency among assessors. Criteria for successful agreement in Cohen's Kappa was defined by statistical guiding literature (Landis & Koch, 1977) whereby < 0.00 = Poor Agreement, 0.00-0.20 = Slight Agreement, 0.21-0.40 = Fair Agreement, 0.41-0.60 = Moderate Agreement, 0.61-0.80 = Substantial Agreement, and 0.81-1.00 = Almost Perfect Agreement. Intra-class Correlation Coefficiency ratings were determined by guiding statistical literature (Cicchetti, 1994) whereby below 0.40 = Poor Agreement; 0.40 to 0.59 = Fair Agreement; 0.60 to 0.74 = Good Agreement; and above 0.75 = Excellent Agreement. For the measure to be used in subsequent phases of the research, the research project deemed that almost perfect or excellent agreement was needed to be reached between assessors.

Research Question 1.2

> *From a clinical perspective how do functional outcomes (social, physical and cognitive) change over time for children with Juvenile Batten disease?*

Assessment Tool

Hamburg Clinical Rating Scale

To gather a clinical perspective on a child's functional skills over time, the standardised Hamburg Clinical Rating Scale for Batten disease (Kohlshutter et al., 1988) was used every 12 months to record participants' clinical presentation. The Hamburg scale is a tool by which clinicians yearly monitor and assess a child's deterioration against an individual index of severity, effectively tracking the rate at which each child is regressing. The Hamburg Scale covers the five main functional domains of Vision, Intellect, Language, Motor Function and Seizure activity and can offer insight into a child's regression over time. The Hamburg scale has since been adapted to provide baseline clinical scores (CLN2 clinical rating scale) to assess functional skills in other research studies (Lewis et al., 2019). Previous comparison studies, however, found that the Hamburg scale and CLN2 rating scale can both adequately track functional change over time in clinical studies, with near perfect agreement between both measures when compared with natural history data (Wyrwich et al., 2018). Table 4.3 illustrates how the ratings in each domain relate to patients' clinical functionality.

Table 4. 3: Single coding scores possible for each of the five major clinical domains for JNCL (taken from Kohlshutter et al., 1988:868)

Domain	Functional State	Coding Score
Vision	Normal	3
	Poor, but orientation good	2
	Poor, orientation difficult	1
	Blind	0
Intellect	Normal	3
	Abstract reasoning (maths) has become difficult	2
	Dementia clearly evident	1
	Apparent total loss	0
Language	Normal	3
	Minor difficulties recognised	2
	Hardly understood	1
	No verbal contact	0
Motor Function	Normal	3
	Slight handicap recognised	2
	Mostly wheelchair, some mobility preserved	1
	Immobile, bedridden	0
Epilepsy (only Grand Mal)	No Seizures	3
	1-2/ year	2
	<1/month, <12/year	1
	>Or equal to 12/year	0

Rather than the measure being completed by medical professionals, the Hamburg Scale was completed by the research team in collaboration with parents. Providing a complementary perspective to the CMTOM assessment data, researchers used the Hamburg measure to track general functional skills (focusing on speech, movement and cognition) each year to provide a picture of clinical change over three years. The use of the measure was supported with guidance from clinicians who were involved in the steering group from the previous research study, from which the data were taken (Ockelford et al., 2019). Acknowledging the limitations of this approach, the project recognises the potential biases of using researcher and parent-reported data, as well as the implication of lacking formal training or experience in using the scale. However, considering the lack of appropriate measures for this population and the wide use of the tool in Batten disease research (Augustine et al., 2013; Lewis et al., 2019; Ockelford et al., 2019), it was deemed an appropriate tool for use with this project.

Analysis

The second stage of analysis drew upon scores obtained from the Hamburg Clinical Scale to assess the video data. Descriptive statistics (mean scores and standard deviations) enabled the research to capture a clinical perspective on each child's functional state and how this changed over time. Analysis initially focused on overall participant mean scores in each of the functional domains (communication, cognition and mobility) excluding the domains related to vision and seizure activity (t was determined that these skills could not be influenced by music). Finally, individual participant data were extracted focusing on participants with a diagnosis of JNCL. This was based on theory from previous research relating explicitly to the impact of music for this specific variant of Batten disease (Elmerskog et al., 2019; Scambler, 2004; von Tetzchner et al., 2019).

Research Question 1.3

> *How do functional outcomes (social, physical and cognitive) change in music therapy over time for children with Juvenile Batten disease?*

Assessment Measures

Research Visits

I, as the lead researcher, made research visits with each of the participants once every school term (roughly three in each year; nine in total for each participant). In line with naturalistic data collection methods in mixed methods research (Tashakkori et al., 2012), research visits took place in the natural setting of the session location and were video recorded for later quantitative analysis using the CMTOM assessment measure. Visits took place for the total duration of each session and adopted a participatory nature where I sometimes took part in the music therapy sessions. The Music Therapists also video recorded their sessions with participants. These were made at alternating points in each school term to offer balanced observations throughout each year (i.e. sessions from Music Therapists were scheduled to be filmed several weeks after researcher visits occurred).

The use of the CMTOM provided an opportunity for a detailed and consistent assessment of each child within sessions over three years, (in comparison to the clinical data captured using the Hamburg Scale measure). By rating assessments across the five functional domains, the measure tracked changes in these domains over time. All video recordings taken during the researcher visits and submitted by Music Therapists were rated using the CMTOM assessment measure allowing for retrospective assessment of each video used. This process enabled triangulation of data collection points, enabling data to be submitted from two complementary perspectives (Guion et al., 2002): myself as the researcher and the Music Therapist or music teacher. This sought to enhance the fidelity and credibility of the data collected and gather varying perspectives from a therapy context.

Analysis

The third stage of analysis in this first phase focused on the CMTOM assessment scores generated from the assessment of the video data taken by the researcher visits and practitioner videos. Analysis was performed on all data on an intention-to-treat basis, regardless of research completion. A total of 182 videos (an average of 15 per participant) were assessed. Descriptive statistics (mean scores and standard deviations) enabled the research to plot the responses in the key areas of communication, behaviour, physical skills, cognition and overall wellbeing over time. Data were extracted on individual subcategories of each domain and mean scores were calculated across all participants to capture an overall impression of functional responses over time for all participants. Furthermore, baseline, midpoint and endpoint mean scores were extracted for each subcategory across all participants to see a more holistic picture of change over time. Thereafter, individual participant mean scores were calculated for each of the domains to show changes in functional responses in overall communication, wellbeing, behaviour, cognition and mobility. Data were then extracted for participants with the JNCL subgroup to observe changes over time in each domain for this group. Interpretations highlighted differences and similarities between the data collected using the two measures. This enabled the research to capture the perspectives from a clinical standpoint (Hamburg Scale data) alongside a music therapy standpoint (CMTOM) and examine how these differ for each child in the research.

4.2.2. Phase Two

Phase Two focused on exploring how responses in music therapy were transferrable to an educational context. This phase initially developed a school program, Music Speaks, which incorporated elements of NMT to target speech and language outcomes. The second part of this phase then observed how functional communication skills of participants in response to this daily program. The participants and intervention in this second phase are described below, before detailing the assessment and analysis for each research question.

Participants

Using a voluntary response (convenience) sampling strategy (Navarro Sada & Maldonado, 2007) through the BDFA, five participants were recruited for the second phase using an email campaign in 2019, sent out to all families associated with the charity. The participant pool for this phase was intentionally small-scale and focused on a case study design (see Appendix 7 for the parent participant information consent form and Appendix 8 for the staff information consent form). Table 4.4 details the participant sample in this second phase of the research, along with the educational context and the participating education and therapy staff members. Participants identified as male (n = 2) and female (n = 3) were recruited, and ages ranged from 9 to 19 years old (SD = 4.5). The children attended mainstream education settings (n = 3) and specialist education colleges (n = 2).

Table 4. 4: Participants from the second phase of the research

Participant	Disease Variant	Setting	Participating Professional/s
1	CLN3 (JNCL)	Mainstream Primary School Setting	2 Learning Support Assistants
2	CLN3 (JNCL)	Mainstream Primary School Setting	3 Visual Impairment specialists
3	CLN3 (JNCL)	Mainstream Primary School Setting	2 Visual Impairment Specialists 1 Inclusion Lead
4	CLN3 (JNCL)	Special Education College Setting	1 Learning Support Assistant
5	CLN3 (JNCL)	Special Education College Setting	1 Music Therapist 1 Learning Support Assistant 1 Student Music Therapist

Previous research (Elmerskog et al., 2019; Scambler, 2004; von Tetzchner et al., 2019) and initial findings from Phase One, suggested the specific impact of music for the JNCL population of Batten disease (CLN3 variant). The participants, therefore, were purposively selected based on a confirmed diagnosis of JNCL. With a sole focus on JNCL, this approach may also have allowed for more generalisable observations, given the heterogeneous nature of the whole Batten disease population. Inclusion criteria also specified attendance at a specialist centre, school or mainstream education provision, and an age range of 0-19 years of age. Two participants were also involved in the preceding study (Ockelford et al., 2019), and their data were included in Phase One. Exclusion criteria discounted children above the age of 19, children who were in full-time care at home, or children with another form of CLN diagnosis. This is because the research phase was focused on trialling the program in education settings which included children in school or college settings, rather than adult residential settings or in their own homes. Due to the pre-emptive nature of the Music Speaks program, the research also excluded any children who had lost speech already or those who were not able to communicate through verbal means. This was assessed in a preliminary meeting with the referrers and educational specialists at each school.

Using convenience sampling (Navarro Sada & Maldonado, 2007) school headteachers and SEN leads who supported the five children, were contacted by phone and email, to introduce the research and invite professionals to participate in the program trial. Phase Two included any type of professional who supported the child in a school setting, which meant that there was a broad range of the types of professionals recruited. The implications of this are discussed further in chapter eight. All school and education settings consented to taking part in the research, delivering the program and taking part in a subsequent interview about their experience (N = 13). These included Learning Support Assistants (n = 4), a Speech and Language therapist (n = 1), Music Therapist (n = 1), student Music Therapist (n = 1), Visual Impairment specialists (n = 5) and an inclusion lead (n = 1).

Intervention Development

Music Speaks Program Development

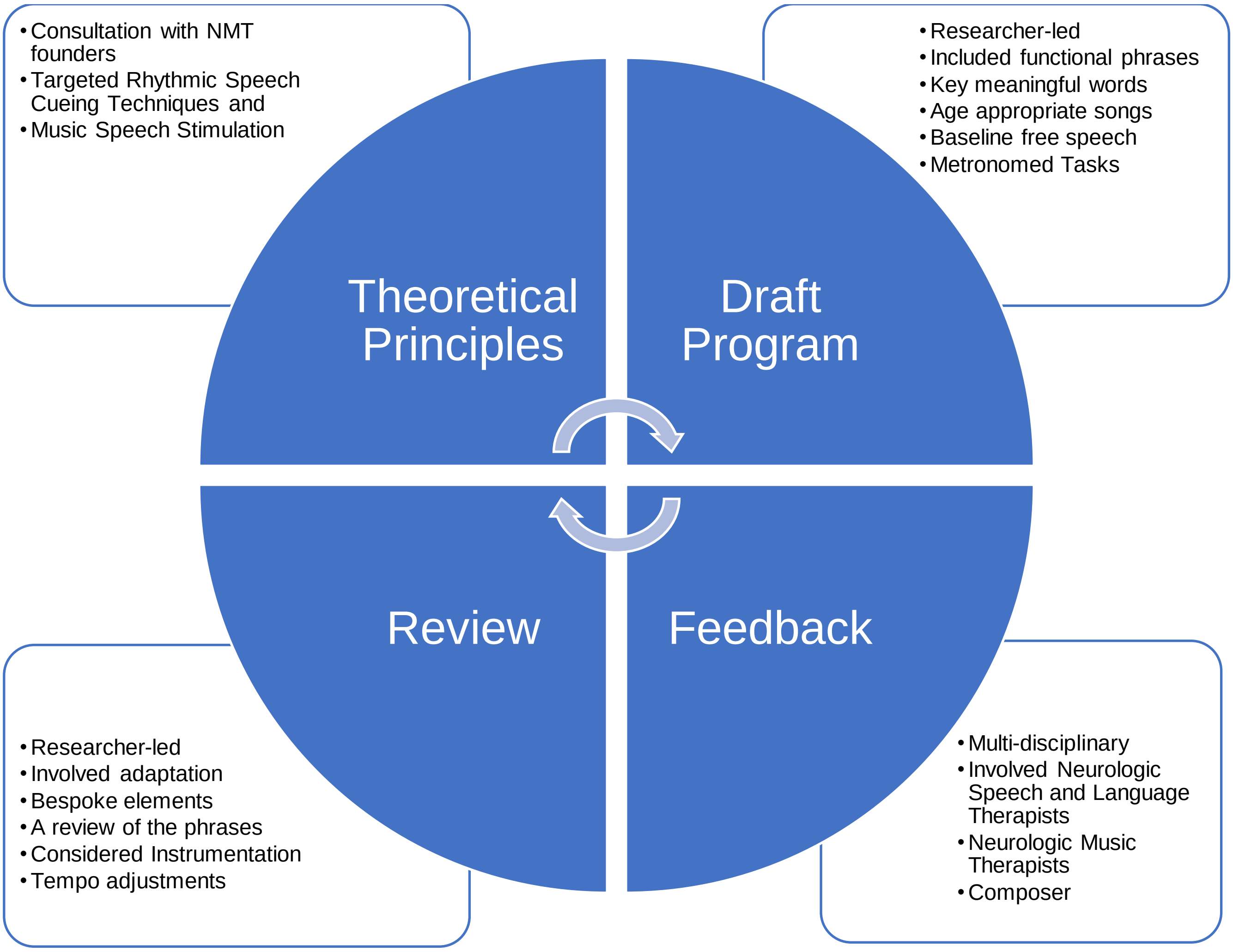
As a trained Neurologic Music Therapist (NMT), I developed the Music Speaks program (see Appendix 9) under the NMT approach (Thaut, 2014); incorporating the key techniques of Rhythmic Speech Cueing (RSC) and Musical Speech Stimulation (MUSTIM). For the purposes of this research project, the development stages focused on ideas generation, development, feedback, and revision. This more inductive process allowed the research to flexibly explore the potential transferrable elements of music therapy experiences and responses (research question 2.1), whilst considering additional perspectives from experienced clinicians. The program did not involve stages of feasibility and evaluation, as it was felt that the constraints of randomised, systematic trial designs would contradict the ethical principles of this research design as mentioned previously (Skivington et al., 2021). Therefore, data collected focused on observed responses of children carrying out the program, and later experiential accounts from interviews of professionals delivering the program. The topic of further evaluation and development of the program is discussed further in chapter eight (see section 8.7.2).

Figure 4.5 illustrates the program development stages and the professionals involved at each stage. The Music Speaks program was developed to positively influence elements of speech and communication through the application of short therapy activities. In the development stages, the project enlisted the support of several expert professionals, who reviewed the program for technicality, coherence and relevance for Batten disease children. Initial consultation involved the Director of the Neurologic Music Therapy course at Colorado State University, who provided insights and feedback on the transferrable principles of NMT. Thereafter, the program development involved a specialist Neurologic Speech and Language Therapist who reviewed the program under typical speech and language therapy goals for this population. The musical structure and technicalities of the program were then reviewed by a colleague, who was a composer and a Music Therapist. Finally, the program was reviewed by two Neurologic Music Therapy clinicians who have specialist experience working with neurodegenerative conditions. The review stage crucially gave a perspective on certain practicalities and elements of recording and delivering the program. For example, through the feedback stages, it was deemed more appropriate to use song-writing software and pre-recorded midi instruments to create the program songs rather than live recordings. This is because midi song versions allowed each song’s tempo to be manipulated depending on the needs of each child, following the principles of Rhythmic Speech Cueing techniques (Thaut, 2014), to match each child’s speech pattern and pace. Furthermore, through this feedback stage, bespoke elements were deemed to be important to the program’s content, allowing for the option of adaptation for the participants involved, (specifically concerning the RSC component, where each program could be differed in speed according to the child’s needs). Additionally, parts of the program were individualised (topics of family, activities and preferred food/drink), and were thus bespoke for each participant.

The eventual program structure included speaking and singing tasks that focused on meaningful topics for each child, covering communication of basic needs, family members, preferred activities, and feelings. These topics were selected based on the guiding principles of NMT activities which target meaningful or functional phrases for individuals (Thaut, 2014) and were discussed with the neurologic speech therapists in subsequent feedback stages. Each session within the program lasted around ten minutes and included speaking tasks (narrated by me, the researcher), before and after the main musical component, to assess how speech skills were impacted.

All musical activities and instructions were recorded by me, the researcher to keep consistency in the verbal directions given. The delivery of the program was supported by a national initiative called the ‘iPod Pharmacy’ (Chiltern Music Therapy, 2020). All tracks (spoken tasks and musical songs) in the Music Speaks program were loaded onto individual iPods and given to each participant (these were available to keep beyond the course of the research).

Figure 4. 5: The four iterative development stages of Music Speaks Program and core elements of each stage



Intervention Delivery

Following the recruitment of the participants, the researcher visited each of the education settings. The purpose of this visit was threefold: to meet the members of the education team delivering the program, to meet the child, and to deliver an implementation training session on the Music Speaks program. Given the broad spectrum of professionals who were involved, the training session aimed to increase the fidelity of the program delivery, to ensure that all practitioners were delivering the program in the same way. The training session lasted approximately two hours, and covered the following elements:

- A background to music therapy and NMT,
- An introduction to the program,
- An overview of the program development,

- Guidance on how to make a video recording,
- A practical walkthrough of the program, the equipment and the materials,
- Set-up on the EthOS app (EthOS, 2020)

The EthOS program was chosen as it provided a secure platform within which professionals could upload video material of their child. Each professional had a unique log in and could view instructions about the data collection requirements. Professionals were only able to view content uploaded by themselves and were not able to see videos of other children taking part in the program trial.

The participants were asked to launch the program in the week following the training session and deliver it daily to students over 20 weeks. A program pack was given to participants in a hardcopy which detailed the program tasks, the song lyrics, a tick sheet and comments section (see Appendix 9). They were asked to complete data recording sheets which detailed the frequency, times, dates, and completion of each task in the program. Recording sheets included whether each task was achieved by the student and record any notes on the day (i.e. presentation of the child on that day, or feedback on tasks in the program).

During the period of data collection in Phase Two (January to July 2020), schools closed because of COVID-19 and as per government advice, face-to-face data collection ceased on 18th March 2020. As a result, the trial period was reduced from the intended 20 weeks to six weeks. A data collection retrieval protocol (see Appendix 10) was drawn up to support the process of transferring the data that had been collected securely and safely in the current climate of COVID-19, to ensure no data were lost. The re-opening of schools in June 2020, meant that the data retrieval protocol could be followed for schools that were back open.

The program was delivered in the same room and where possible, by the same education staff (up to three) and at the same timeslot in the child's school timetable, to reduce other influencing factors and firmly position research findings within an education context (for example rather than delivering the program at home, or in other therapy sessions). Video data were collected by the professionals once a week and submitted through the online research portal, EthOS. A filming schedule of pre-selected filming dates ensured that all schools were filming sessions on the same days each week to establish an element of consistency in data collection (see Appendix 11). This element of consistency aimed to facilitate fidelity of video observations (i.e. video data were taken at consistent times for all children rather than educators subjectively selecting videos to submit). Videos in this phase aimed to track and observe speech and communication skills in response to the Music Speaks program and ascertain the extent to which the observed responses in the music therapy context of Phase One could be transferred into daily educational contexts.

Research Question 2.1

> *Can the Music Speaks program be delivered effectively in a formal educational framework?*

Assessment Measures

To answer the first research question relating to the delivery of Music Speaks in schools, educators were asked to complete data recording sheets. These sheets were included in the daily program pack and incorporated the time and date of program delivery, completion of each task (using checkboxes) as well as an additional notes section. The purpose of the recording sheets was to capture data relating to the practical elements of educators delivering the program in an education setting (for example, quantitative data represented how often they delivered the program, at what time of the day, and which tasks were completed). These data enabled the research to explore the frequency of program delivery, and whether this was achievable for all participants, as well as how many tasks were achieved each day for each participant. Additionally, the notes section of the recording sheets provided the opportunity for educators to describe any considerations, challenges or issues in delivering the program, and note any reflections or feedback over the six weeks.

Analysis

Initial analysis in this second phase focused on data extrapolated from the Music Speaks recording sheets. Numerical data gave an indication as to the prevalence, frequency of delivery and task completion, as well as supplementary information relating to the context and feedback on the daily program. As in Phase One, analysis was again performed on an intention-to-treat basis regardless of program trial completion. Data were interpreted using descriptive statistics to indicate the mean frequency and time of program delivery, and total achieved tasks across the participant group. Supplementary information was extracted from recording sheets and organised into topics to capture data relating to program delivery from the professionals’ perspective. Notes were grouped into similar topics and contributed additional insight into influencing variables each day (i.e. a child’s absence, fatigue, or mood) as well as additional notes (such as the need for prompts or resources, the child’s response to each task or comments on the tasks and songs themselves). There were no predefined success criteria for this phase relating to completion of tasks or program delivery. This is addressed in the discussion chapter (see section 8.7.2).

Research Question 2.2

> *How do communication outcomes for children with Juvenile Batten disease change in response to the Music Speaks program?*

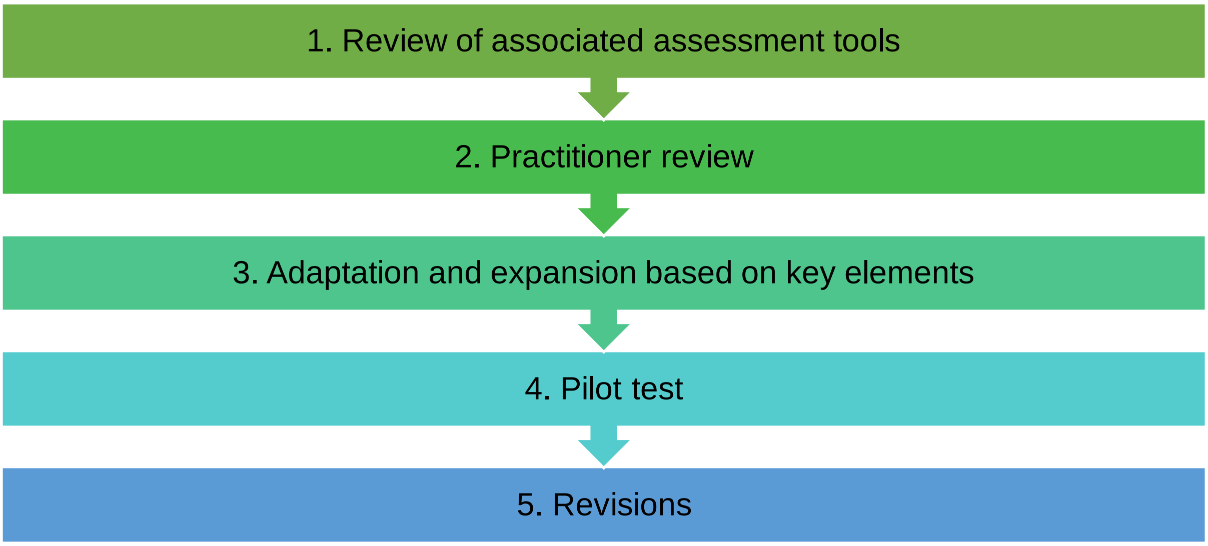
Assessment Measures

CMTOM Communication Subscale Development

It was imperative that the assessment measure used in this phase incorporated key elements of speech and language function. As described in chapter two, assessment measures for use with children affected by neurodegenerative conditions are lacking, and none are appropriate for use in observational settings or in music therapy. The two most relevant assessments for the dysarthric tendencies of affected children are the Frenchay Dysarthria Assessment (Enderby, 1980), which requires specific speaking, eating, drinking and swallowing activities to be carried out, and the TOM's assessment (Enderby & John, 2019) which is a holistic measure intended for use over a longer period. Furthermore, the Hamburg Clinical Scale (Kohlshutter et al., 1988) used in Phase One is a holistic measure intended for assessment over longer periods and was therefore inappropriate for short term interventions like the Music Speaks program. Two specific Batten disease assessment measures were also considered in this phase. The Unified Batten Disease Rating Scale (UBDRS, Adams *et al.*, 2007), which only contained one speech and language element 'speech clarity' and did not provide enough detailed assessment criteria; and the Education Development Observation (EDO) which, although described in research as an appropriate tool for intervention settings (von Tetzchner et al., 2019), has not yet been validated in the sector. Using the EDO in its entirety would have involved assessing 11 subcategories including vision, communication, literacy, social life, gross motor function, fine motor function, physical activity, behaviour, memory and attention, independence and autonomy, interests, and equipment. To maintain validity of the EDO, assessments would have needed to use all scale domains, and for this phase of research, it was not possible to use the whole measure as an assessment tool for communication.

Therefore, to capture the essential elements of speech and language, and to align closely with the methodological approaches in Phase One, adapting the communication section of the CMTOM was considered as an appropriate solution. The outcome measure did not, however, incorporate speech and language elements that were specific to JNCL, and therefore the doctoral project implemented stages of development, adaptation, and refinement of the subscale. Figure 4.6 depicts this process of development.

Figure 4. 6: Development Process for the CMTOM Communication subscale assessment



As can be seen in Figure 4.6, the development process incorporated a review of associated assessment tools which included measures from speech and language, music therapy, Batten disease, and neuropsychology sectors. Appendix 12 details the assessment materials, authors, and notes about their relevant communication items. The development then gathered and compared all associated and transferable elements of each scale which were applicable to the Batten disease population. For example, elements of expressive speech were incorporated from other assessment tools, such as articulation, volume or use of words (Enderby, 1980; von Tetzchner et al., 2019). From reviewing other measures, it was clear that there were many dimensions of communication that could be measured (i.e. non-verbal communication, word-finding, or vocalisations). Based on previous theory relating to the effects of music and expressive language and speech however (Harris et al., 2016; Lim et al., 2013; von Tetzchner et al., 2019), this research was interested in exploring communication responses in the expressive language domain specifically, and therefore the subscale included elements of expressive communication, language activity and receptive communication rather than elements of non-verbal communication.

The CMTOM communication subscale was shared for review with two reputable practitioners (one Neurologic Speech and Language therapist and one Neurologic Music Therapist). These practitioners had over 20 years combined experience in their sectors and had specialist experience working with paediatric neurological conditions, specifically children affected by Batten disease. Gathering feedback from the neurological music therapy and speech and language therapy sectors offered two complementary clinical perspectives on the use of the subscale as an assessment measure, according to their experiences supporting children with

Batten disease. Following the clinician feedback, the following adaptations were made to the scale:

1. Addition of prosody items such as ‘Intonation’, ‘Stress’, ‘Rhythm of speech/ Pacing’
2. Addition of ‘Use of Communication Aids’
3. Omitted ‘Awareness of Others’ and instead added ‘Responds to Familiar Voice’ and ‘Responds to Own Name’
4. Definition of ‘Ability to Interact’ to include ‘Turn-taking behaviours’, ‘Vocal imitations/ play’, ‘Use of Gestures’, ‘Use of Actions’, ‘Intensive Interactions’

Through a process of cross-referencing, expansion, inclusion, and integration, 33 items were formed into the first version of the CMTOM communication subscale (see Appendix 13a).

The draft CMT Communication subscale was then trialled with one video from Phase Two data collection to check its relevance to the data before final revisions were made. As a result of trialling the new subscale, several revisions were made based on the video data that were received in this phase. Firstly, given that none of the participants used communication aids or communicated using gestures during the Music Speaks program sessions, these items were omitted from the subscale. Secondly, items relating to interactive elements of communication (i.e. Use of Conversation, Turn-taking Behaviours, Initiating Interactions) were also eliminated, given that the Music Speaks program only required participants to copy and repeat phrases or join in with singing a song. Given that the program did not include elements of music play, the items relating to music playing were also omitted (i.e. use of actions and intensive interaction play). Lastly, three elements of receptive communication were included in the subscale to assess some basic elements of communication response (such as Word Recall, and the Ability to Respond and Participate). The final version of the CMTOM Communication Subscale can be found in Appendix 13b.

Analysis

This stage of analysis drew upon the video data submitted by educators delivering the Music Speaks program (N = 23). Videos were analysed using the CMTOM communication subscale assessment measure. Descriptive statistics (mean scores and standard deviations) enabled the research to capture responses to each of the Music Speaks program sessions delivered over the six-week period, in the areas of expressive and receptive communication. Videos were rated on a scale of 0-4 (0=Never, 1=Rarely, 2=Sometimes, 3=Often, 4=Consistently), and CMTOM ratings were recorded after task one (baseline) and after task four (endpoint). Three items were reversed scored (‘Stutters and Stammers’, ‘Repetition’, ‘Dysarthria or

Mumbling'). There was a total of 68 data collection points across the whole participant pool. Data were analysed across all participants to capture an overall impression of change over time, as well as individual participant-level data focusing on single items in subcategories of each domain.

4.2.3. Phase Three

Phase Three of the research design focused on gathering experiences and perspectives from educational staff and therapists supporting children with Batten disease. This final stage of data collection focused on capturing empirical evidence to enrich the data and findings from the previous two research phases. The rationale here was to draw together a dialogue into challenges, experiences and perspectives from educators, and explore perceptions of those using the Music Speaks program. Participants, interview methods and analysis are subsequently described below.

Participants

At the recruitment stage in Phase Two, participants were given the option to participate in an interview following the completion of the Music Speaks program delivery. Of the 13 participants who delivered the six-week Music Speaks program in Phase Two, seven professionals agreed to take part in an interview. Table 4.7 details the participants interviewed in this research phase, who included Learning Support Assistants (n = 2), Visual Impairment specialists (n = 3) an Inclusion Lead (n = 1), and a Music Therapist (n = 1). The interviewees supported four of the five children from Phase Two with JNCL (the professionals supporting participant four had left the education setting at the time of interviews), in a mainstream primary school (n = 3) or a special education college (n = 1). Children's ages ranged from 8-18 years of age (SD = 4.5, see Table 4.4), and most professionals were based in the North of England and East Midlands, (Yorkshire, Derbyshire and Northamptonshire, and Wales). All children were alive at the time of the interview.

Table 4. 7: Phase Three interviewees, their occupations, roles and associated child participants

Participant	Job Title	Role	Setting	Notes
1	Learning Support Assistant	Provide one-to-one support for the child in the classroom	Special Education College Setting	Supported child participant five
2	Music Therapist	Functional and psychological support through music therapy practice	Special Education College Setting	Supported child participant five
3	Visual Impairment Specialist	Provide one-to-one support for the child in the classroom	Mainstream Primary School Setting	Supported child participant two
4	Visual Impairment Specialist	Provide one-to-one support for the child's Visual Impairment needs	Mainstream Primary School Setting	Supported child participant three
5	Inclusion Lead	Provide support for all pupils in the school through effective inclusion.	Mainstream Primary School Setting	Supported child participant three
6	Visual Impairment Specialist	Provide one-to-one support for the child's Visual Impairment needs	Mainstream Primary School Setting	Supported child participant three
7	Learning Support Assistant	Provide one-to-one support for the child in the classroom	Mainstream Primary School Setting	Supported child participant one

Drawing upon the perspectives from four types of professionals (education, therapeutic, vision specialist and inclusion) offered the opportunity for triangulation of perspectives and data collected (Guion et al., 2002). This sought to enhance the fidelity and credibility of the data collected and gather the rich perspectives of the classroom setting, therapy contexts, visual impairment specialisms and inclusive education. Due to the unexpected Covid-19 pandemic, Phase Three was adapted to conduct online or phone interviews rather than face-to-face meetings. Although all participants had agreed to an interview in the initial recruitment stages, as the interview was held virtually online, follow-up verbal or written consent was obtained, and the format of online interviews was outlined and explained.

Data Collection

All interviews were conducted remotely either by video call (n = 3) or by telephone call (n = 4) in November 2020, and each participant took part in a one-on-one interview with the researcher. Participants received an email outlining the time and date of the interview and a link to the virtual conferencing space. The interviews all took place over two weeks (eight months after the Music Speaks trial had stopped). Interviews lasted between 25 minutes and one hour, and the average length of the interview was around 42 minutes. Interviews were recorded using a mobile audio recording device or using the in-app Teams recording function. Interviewees were not sent an interview schedule beforehand.

At the time of interview, all interviewees had stopped supporting the child when the UK schools and education settings closed as a direct impact of the COVID-19 pandemic in March 2020. Interviews, therefore, all opened with a recap of the research project, its aims, interests, and my background as a Music Therapist. Each interview also addressed ethical issues (data storage and protection, consent to record, and confidentiality) which included the specific limitations of using the audio or virtual interview platform and the impact on confidentiality. For example, the situation of the pandemic lockdown meant that both interviewer and interviewee were in their respective houses with other members of their household, and interviews were conducted where possible in private rooms with closed doors. Content discussed in the interviews was a retrospective reflection of their time supporting the child in school (research question 3.1), and their experiences running the Music Speaks program from February to March 2020 (research question 3.2). Example interview questions can be found in Table 4.8 and the full interview schedule can be found in Appendix 14. The interviews were semi-structured allowing for a more informal conversation between me and the interviewee, ensuring the flexibility for new topics to emerge and be discussed (Fletcher, 2017). There were also points in the interview where ideas or comments were clarified or reframed by the interviewer.

Table 4. 8: Example interview questions and corresponding interviewer probes

Interview Question	Interviewer Probe
1. Can you tell me about your role in supporting [NAME]?	Are there approaches that you have found to be most impactful? Can you describe any challenges they encounter?
2. How do you think Music Speaks has impacted the child you support?	How has the child’s engagement with the program changed over time? Did you see any changes in the child’s communication?

Following the interviews, video and audio recordings were immediately transferred onto the secure University One Drive data storage platform and removed from individual devices.

Research Questions 3.1 and 3.2

> *What are the experiences of educators and therapists supporting a child with Juvenile Batten disease in an educational context?*
>
> *What are the perceptions of the Music Speaks program from those using it in educational contexts?*

Analysis

A total of seven interviews were transcribed and analysed using an inductive thematic analysis approach (Braun & Clarke, 2006). In line with previous research projects, inductive, thematic analysis enabled the research to simultaneously be driven by the raw data as well as incorporating into themes existing theory, insights and literature (Scambler, 2004).

Interviews were transcribed with the support of an analysis program ‘F4 Transkript’. In contrast to naturalist approaches to transcription (whereby every utterance is captured in detail), the process of transcription involved transcribing the interviews clean verbatim, omitting conversational utterances (stutters, pauses, repetitions or non-verbal language), correcting grammatical errors, and included language associated with regional accents (Oliver, Serovich, & Mason, 2005). Transcription also involved conscious decisions, selections and exclusions to define the key concepts and themes in the interview data.

The thematic analysis focused on the core stages of data reduction, organisation, and interpretation (see Table 4.9 for the detailed stages of analysis).

Table 4. 9: Table illustrating the thematic analysis stages in research Phase Three

Analysis Phase	Detailed Stages
Data Reduction	1. Familiarisation with data
	2. Transcription of interview material
	3. Summative notes on each interview transcript
Organisation	4. Initial codes assigned to summary notes
	5. Codes organised by emerging protothemes.
	6. Protothemes refined into themes and subthemes
Interpretation	7. Thematic mapping of themes and subthemes with data extracts
	8. Review of themes and re-familiarisation of material
	9. Defining final themes and subthemes

4.3. Ethics

There were several ethical implications of conducting research with such a fragile and palliative population of children with Batten disease. Seven key areas were identified as presenting a potential risk to participants in Phase One of the research through the ethical assessment and approval process at the University of Roehampton. Phases Two and Three, which related to the program development and interviews, were subsequently granted ethical approval (see Appendix 2 for ethics approval letters). The doctoral research project began at the University of Roehampton before transferring to the University of Brighton in 2020, and therefore ethical clearance was applied for and granted from the University of Roehampton and reviewed and approved at the University of Brighton.

Ethical issues were linked primarily to the fragile medical condition of participants, knowledge relating to their diagnosis, emotional distress of all parents and professionals taking part, as well as any safeguarding risks associated with therapeutic work. To mitigate against these risks, practitioners were required to conduct initial visits (except for those who already worked with a child before the project), which presented the opportunity to complete in-person risk assessments, gain specific knowledge about the child they were supporting, their condition and the context of their care and education. From conversations with the BDFA, it was also deemed inappropriate to implement any kind of control groups or delayed start measures, as ethically, families should have immediate access to any beneficial therapy. Therefore, the project’s case study design allowed all participants to access the therapeutic input at the same time.

There were no serious adverse events in this research project, no safeguarding incidents and no participants withdrew from the project. As per the University of

Brighton's safeguarding procedures, data will be archived once the doctoral project has been completed and kept for ten years following project completion, on a secure, encrypted hard drive. Identified ethical considerations, health and safety risks and mitigating solutions associated with the phases of the research are subsequently outlined.

4.3.1 Vulnerable Populations

All children in the research phases were in fragile health and wellbeing and were potentially in the palliative care stages of their life. The fragility of participants was a key ethical consideration, and due care was taken to mitigate additional risks to the children taking part. Dependent on the health and wellbeing of the child, parents, guardians or professionals had the option of accepting therapeutic input where appropriate. If children were too ill when visits were scheduled, the visits were rescheduled. All practitioners in the three phases held an up-to-date DBS (Disclosure and Barring Service) check, as well as specialist training, experience or expertise with fragile children or those in palliative care settings. Importantly, all practitioners and researchers in the project followed on-site health and safety guidelines and completed school-specific safeguarding and risk assessments (which included measures like sterilising equipment etc). The Music Therapists, music practitioner and researcher, on the first meeting with parents or health professionals, carried out an initial risk assessment, to ensure the safety of all involved for each child in each setting. To protect the psychological wellbeing of participants and their families, potentially sensitive content of sessions was handled only by trained professional music therapy practitioners and in accordance with the Health and Care Professions Council Standards of Proficiency for Arts Therapists (HCPC, 2013).

4.3.2. Consent

Children participating in the research were under the age of 18 at the start of their participation, and therefore required parent or guardian consent to take part in the research project. For participants who had cognitive capacity, verbal consent to take part was obtained from their parent or guardian in addition to completed parent or guardian consent forms. Consent specifically covered participation in music sessions, as well as the video recordings of sessions and the sharing of data in research subsequent research documentation. Parents and guardians were given the option to opt-in or opt-out of each consent item and had the option to blur faces on video or photograph materials. All parents consented to their child receiving the research interventions and five opted to blur the faces of their children. Details of the research were presented in an information pack, and all participants were given the option to withdraw from the project (for the duration of the research project) without affecting the child's access to music therapy sessions. All data were anonymised to protect the identity of the children and adults taking part. Throughout the research, if participants withdrew from a phase (two participants withdrew from Phase One), consent to use anonymised data already collected was obtained.

4.3.3. Emotional and Psychological Distress

Due to the fragile and degenerative nature of children affected by Batten disease, the wellbeing and potential emotional distress of parents, guardians, carers and professionals was a key consideration for the research project. As with the measures employed to protect the child participants in the research project, the Music Therapists, music practitioner and researcher handled sensitive information and content of sessions as per the HCPC Standard of Proficiency for Arts Therapists (HCPC, 2013) and only relevant data were shared with the research team. The Music Therapists, music practitioners and researchers drew upon their own experiences of working with children and families affected by degenerative disease, working under guidance from the BDFA psychological support as to appropriate ways to support families who were dealing with high levels of anxiety and emotional distress.

4.3.4. Safeguarding

To safeguard participants, practitioners and researchers followed University of Roehampton safeguarding policies as well as the on-site safeguarding procedures for each school or care setting. The designated safeguarding lead in each setting was identified should any incidents arise. A parent, professional or other family member were always present when the Music Therapist or music practitioner was visiting, and sessions only took place at school, the child's home or in a health setting and never in an alternative private space. No safeguarding incidents arose throughout the research project.

4.3.5. Health and safety

Due to the low immunity and complex medical state of children affected by Batten disease, the research needed to implement infection control measures to protect children's fragile health. The University of Roehampton and site-specific health and safety policies were followed for each setting, and an initial risk assessment for each child identified issues around the health and safety of participants or the Music Therapists and music practitioner. The visiting Music Therapist, music practitioner and researcher were aware of the fragile nature of the child's health and accordingly adhered to infection control procedures such as washing hands before any interaction with the child, and any musical instruments used were sterilised before and after use. Musical instruments with small parts or which come in contact with mouths were not used (i.e. instruments which could harbour infectious bacteria or be a choking hazard).

4.3.6. Medical Context and Sensitive Information

The medical diagnosis of each child in some cases had not been disclosed to the affected child, family members, or supporting professionals, therefore due care and diligence had to be taken to handle this sensitive and confidential information. To protect sensitive information, the Music Therapists, music practitioner and researchers were mindful of participant medical history and aware of the degenerative nature of the child when liaising with parents, professionals or the child themselves, keeping a record of which information had been disclosed to whom. The Music Therapist, music practitioners and researcher always followed parent or guardian guidelines if talking to the child about their condition. The Music Therapists, music practitioners and researchers made themselves aware of and respected the cultural differences of the families taking part. Additionally, there was sensitivity and respect of cultural or religious matters when selecting session content or material.

In the first phase of the research under the previous research project (Ockelford et al., 2019), a breach of confidentiality arose where a sibling of an affected child potentially became aware of their siblings' condition. Due to an incorrect email address given by a parent, an email was mistakenly sent to the sibling of the affected child, which presented information relating to Batten disease and risked the sibling finding out about the affected child’s condition (previously undisclosed). In accordance with the University of Roehampton safeguarding procedures, the parents were notified immediately, and the email was recovered without being opened. A thorough check of all contact information was thereafter conducted immediately to ensure all information was correct and appropriate, and the incident was recorded following the University of Roehampton’s confidentiality protocol.

4.3.7. Lone Working and Unfamiliar Environments

In some situations (such as school holidays, or if the child was in a hospital or a hospice) the practitioner and researcher visited the child in an alternative setting outside of the education and care environment. In these situations, the University of Roehampton’s Lone Working policy was followed to ensure the safety of the child and the practitioner. Music Therapists and music practitioners were given dates, times and locations of visits, and shared these with the team. Emergency contacts were given and kept on the university database. Mobile phones were always carried during visits and parents and professionals confirmed beforehand that visits could take place. The Music Therapists, music practitioner and researcher carried photo identification and a copy of their DBS certificate with them during each visit.

An initial risk assessment of each location was recorded by the researcher, Music Therapists and music practitioner to ensure safety in all settings.

4.4. Conclusion

This chapter described in detail the methodological approaches and inductive methods implemented throughout the three phases of research. The methods outlined each research phase design, data collection methods, procedures and analysis for each of the three phases. In Phase One quantitative data was collected, which focused on questionnaire data and quantitative assessment measures (CMTOM and Hamburg assessments). Data analysis undertaken in Phase One was based on descriptive statistics and related to the child's functional responses to music therapy and how this sat alongside standard clinical perspectives of a child's functionality. Phase Two built upon the observations in Phase One, and led to the iterative conceptualisation and development of the Music Speaks program. In this phase, quantitative data was collected via recording sheet data and delivery, and provided an indication as to how the program was delivered in a school setting. Subsequent analysis of video data using the CMTOM communication subscale captured the communication responses to the Music Speaks program for the five children taking part. Phase Three focused on qualitative interview data, utilising inductive thematic analysis methods to formulate concepts around the experiences and perceptions of educators supporting an affected child and how they perceived the Music Speaks program.

Ultimately, the three distinct phases of this research's methodological approach are distinctly interlinked with Phase One which influenced the direction of the proceeding phases. In accordance with a critical realist research position (Porter, 2015; Scambler, 2004), this approach allowed for an agile and cyclical approach to research design, facilitating an emergent dialogue between observations, contexts and experiences, for those affected by childhood Batten disease.

The following chapters of the thesis present the results gathered in Phases One, Two and Three. Each proceeding chapter is divided in accordance with each phase for ease of reference.

Chapter 5: Results from Phase One Music Therapy Observations

"The most exciting rhythms seem unexpected and complex, the most beautiful melodies simple and inevitable"
— W.H. Auden

5.1. Introduction

This chapter outlines the findings from the data analysis conducted in Phase One, which assessed the appropriateness, validity and reliability of the CMTOM assessment measure and examined how children with Batten disease responded to Music Therapy. A summary of how each research question has been approached is set out below:

1.1 For children with Batten disease, how can functional outcomes be tracked and measured in music therapy?

Data from practitioner questionnaires captured perspectives on the appropriateness and validity of the measure for children with Batten disease receiving music therapy. An inter-rater reliability exercise aimed to determine whether an agreement between two different assessors using the measure could be reached.

1.2 From a clinical perspective how do functional outcomes (social, physical and cognitive) change over time for children with Juvenile Batten disease?

Analysis drew upon the mean Hamburg Clinical scale scores taken yearly over three years to present the clinical perspective of each child's functionality and presentation, with a focus on the JNCL disease subgroup.

1.3 How do functional outcomes (social, physical and cognitive) change in music therapy over time for children with Juvenile Batten disease?

Data captured functional responses in music therapy using the CMTOM assessment measure and analysis focused on the mean CMTOM scores, taken from video observations over three years, to identify functional responses in music therapy. This allowed this research project to plot the change of these over time with a focus on the JNCL subgroup.

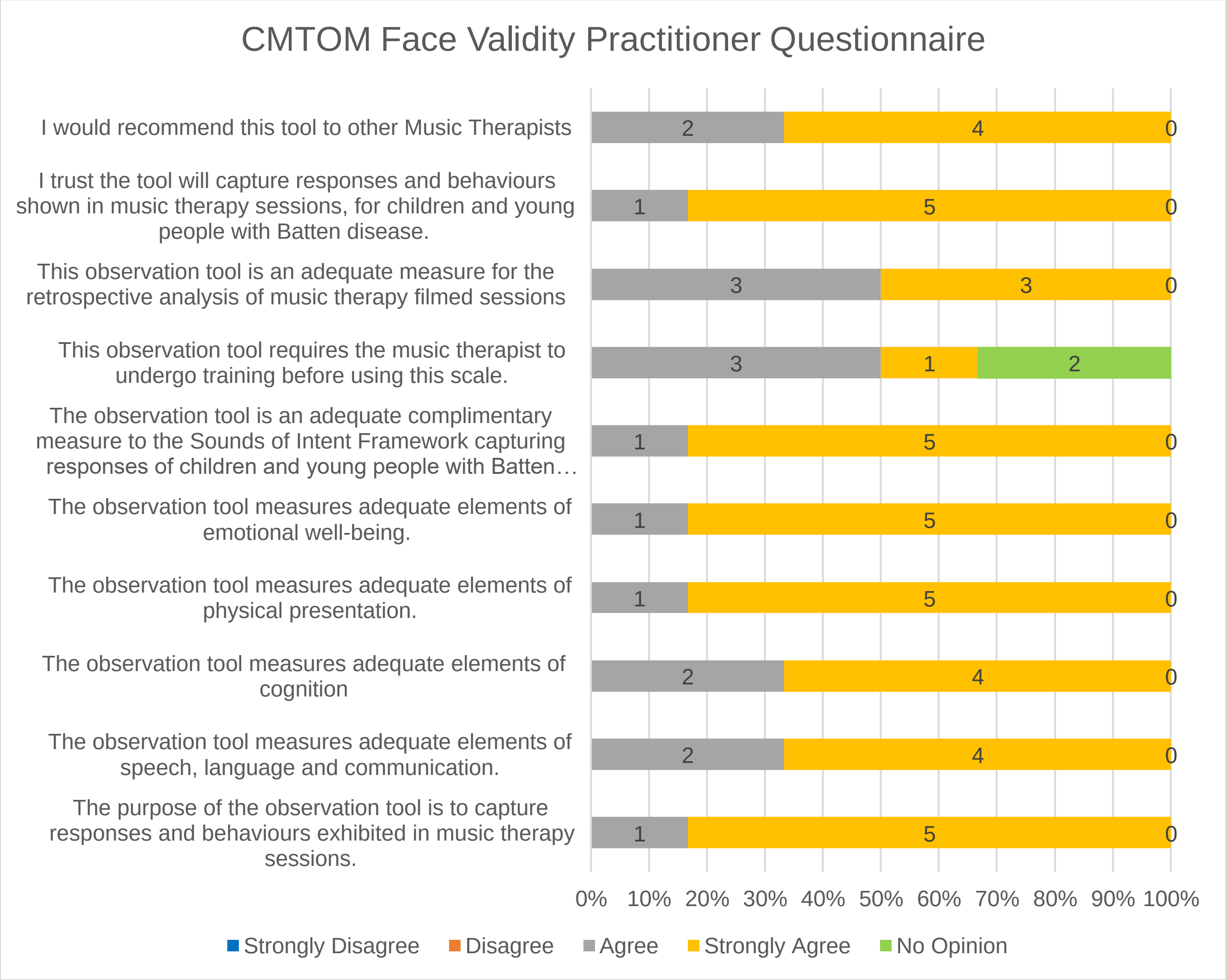
To answer each of the research questions in turn, the analysis from the Hamburg rating scale and the CMTOM assessment measure have been presented separately. It was important that the two measures are not presented comparatively, but instead provide two distinct perspectives on a child’s change over time. Sadly, three of the children from this phase have since passed away. Their data still included in this chapter with permission of their families.

5.2. Research Question 1.1: For children with Batten disease, how can functional outcomes be tracked and measured in music therapy?

5.2.1. Face Validity of the Chiltern Music Therapy Outcome Measure

Music Therapy practitioners from Phase One of the research (N = 6), completed a ten-item questionnaire based on the appropriateness and relevance of the CMTOM for use with individuals affected by Batten disease. Figure 5.1 plots questionnaire responses, illustrating the responses to each questionnaire item.

Figure 5. 1: Questionnaire responses, exploring face validity of the CMTOM measure



Results showed that practitioners strongly agreed that the CMTOM was a trustworthy tool for capturing responses and behaviours in music therapy sessions for children with Batten disease (n = 5/6), and strongly agreed that the CMTOM was an adequate tool in measuring emotional wellbeing (n = 5/6), physical presentation (n = 5/6), speech (n = 4/6) and cognition (n = 4/6). Practitioners strongly agreed (n = 4/6) that they would recommend the tool to other practitioners but agreed that (n = 4/6) the tool required the Music Therapist to undergo training before using the scale.

Results from these questionnaire data suggested strong face validity of the CMTOM measure as an adequate, and trustworthy tool for capturing responses in music therapy for children with Batten disease. Findings here suggest the measure could be recommended to other practitioners, but those using the measure, may require training before use. Based on the positive findings from this phase, it was demonstrated that the CMTOM could be used for subsequent observation and monitoring of children in their music therapy sessions, to capture functional responses (wellbeing, mobility, speech, and cognition). Subsequent reliability exercises sought to strengthen this measure further for it to be used in the subsequent stages of Phase One.

5.2.2. Reliability of the Chiltern Music Therapy Outcome Measure

An inter-rater reliability exercise was implemented to examine the agreement between the two researchers using the CMTOM assessment measure in Phase One. Inter-rater reliability was measured through an exercise which randomly selected ten session videos for two assessors to rate using the CMTOM measure. Analysis used the Cohen’s Kappa coefficient (k) and Intraclass Correlation Coefficiency tests to determine consistency between the two assessment scores. The results of the inter-rater analysis showed Kappa = 0.84 with $p < 0.001$. This result is statistically significant and indicates almost perfect agreement. The intraclass Correlation Coefficiency test produced a score of .957 with a 95% confidence interval at .950-.963, indicating excellent agreement between the two assessors with optimum consistency.

Findings from the statistical tests conducted on the dual assessor exercise indicated strong agreement between the assessment of the selected videos. This suggests that there is a high level of internal consistency in the measure, denoting its value in assessing observations across multiple participants with multiple assessors.

5.2.3. Summary

Based on the findings of the face validity and reliability exercises conducted, it was deemed appropriate that the CMTOM measure could be utilised for subsequent phases of this research project. In the context of music therapy, the measure could adequately capture and track observations of functional responses to music therapy for children with Batten disease, therefore, no changes were made to the measure or the intended use for the project. There were several limitations and considerations associated with the validity exercises conducted in this stage, which are discussed in more detail in chapter eight. However, as per validity findings, the project ensured the CMTOM measure was only used by assessors who had been trained in using the measure and would only be used for retrospective analysis of video data collected during researcher observation visits.

5.3. Research Question 1.2: From a clinical perspective how do functional outcomes (social, physical and cognitive) change over time for children with Juvenile Batten disease?

To provide a perspective on the clinical presentation over the three years, the Hamburg Clinical Rating scale was used to score three functional domains (communication, cognition and mobility) for the participants in the research. A member of the research team completed the Hamburg scores each year with the child’s parent. Data collection from the Hamburg rating scale yielded one reading every year of the project (except for participants who did not complete the research project) and therefore results were analysed using yearly assessments for the 12 participants (N = 34 total assessments). Some participants did not complete the full three years of the first research phase (n = 3/12). Due to the fragile nature of the participants involved there were no completion requirements for data to be included and analysis was performed on an intention-treat-basis basis regardless of research completion. Table 5.2 outlines each participant, diagnosis and the number of corresponding CMTOM and Hamburg assessments taken throughout their involvement in the research (identifying characteristics have been omitted from this table). CMTOM scores were taken after each video observation was made (from researcher visits, and practitioner videos), and averaged 15 assessments over the project (range = 5-19).

Table 5. 2: Participants' total recorded Hamburg and CMTOM scores

Participant	Disease Variant	No. of Hamburg Assessments	No. of CMTOM Assessments
1	CLN6	3	18
2	CLN2	2	10
3	CLN5	3	16
4	CLN3	3	18
5	CLN5	3	18
6	CLN3	3	13
7	CLN2	2	5
8	CLN3	3	19
9	CLN2	3	14
10	CLN2	3	15
11	CLN2	3	19
12	CLN8	2	17

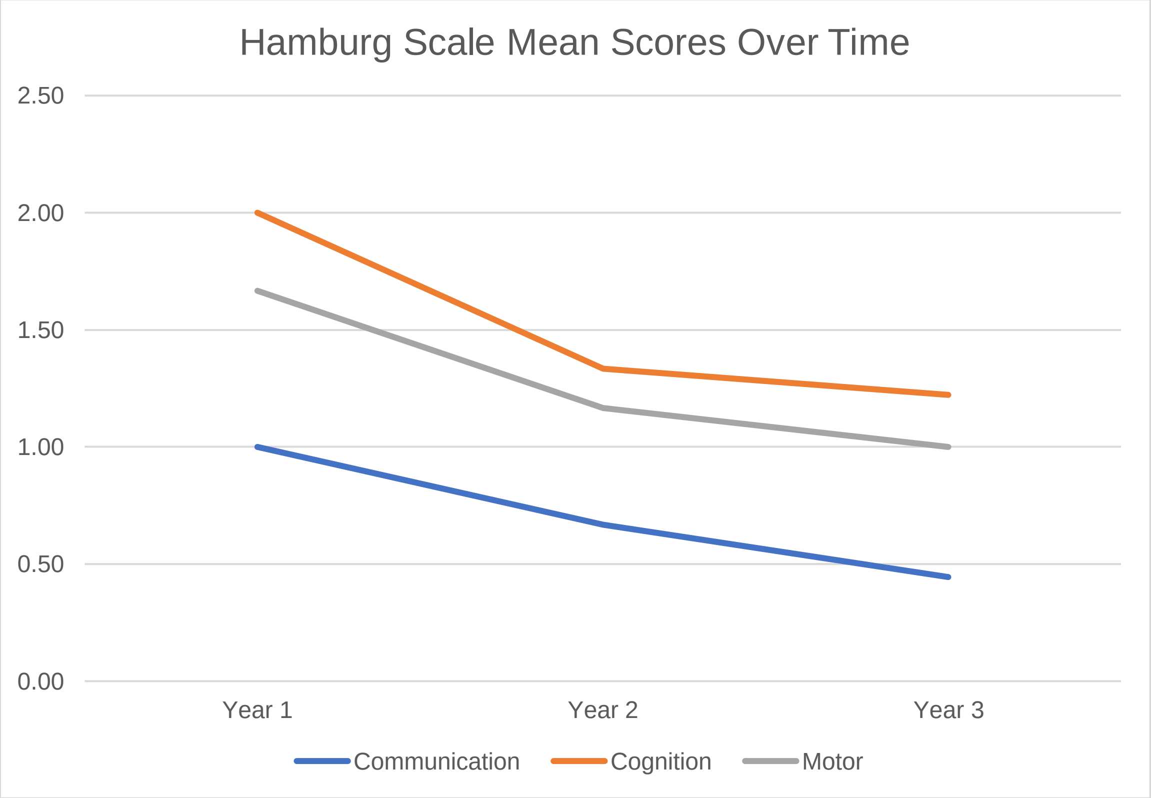
Taking mean ($\bar{x}$) and standard deviation (SD) scores, data analysis captured how these figures changed over the three years. Table 5.3 represents the values for the Hamburg scores across three years. As can be seen from Table 5.3 in communication, Hamburg scale mean scores decreased from a score of 1 (SD = 0.95) to 0.4 (SD = 0.53). In the area of cognition, Hamburg scale mean scores decreased from a score of 2 (SD = 0.95) to 1.22 (SD = 0.83). In the domain of mobility, an area with expected marked deterioration for participants in the later stages of the disease, mean scores according to the Hamburg scale decreased from 1.67 (SD = 0.99) to 1 (SD = 0.71).

Table 5. 3: Mean and standard deviation Hamburg Scale ratings across three years in communication, cognition and motor skills

	$\overline{x}$	SD
Hamburg Communication Year 1	1	0.95
Hamburg Communication Year 2	0.67	0.65
Hamburg Communication Year 3	0.4	0.53
Hamburg Cognition Year 1	2	0.95
Hamburg Cognition Year 2	1.33	0.89
Hamburg Cognition Year 3	1.22	0.83
Hamburg Motor Year 1	1.67	0.99
Hamburg Motor Year 2	1.17	1.03
Hamburg Motor Year 3	1	0.71

Plotting out these results illustrated a marked trajectory of deterioration in all areas of the Hamburg clinical scores across three years. Figure 5.4 illustrates the mean Hamburg scores taken from each year of Phase One.

Figure 5. 4: Mean Hamburg Scores in communication, cognition and mobility over three years

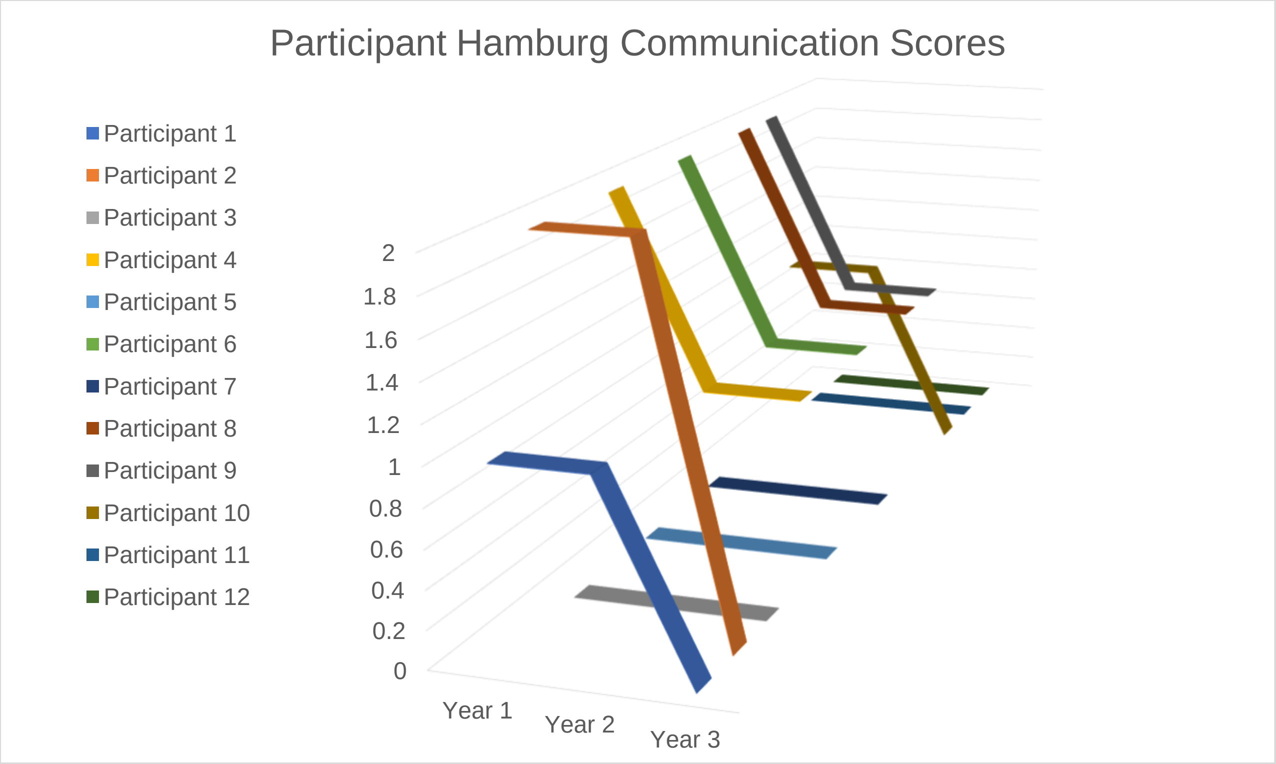


As can be seen in the mean Hamburg scores for participants over three years, there was deterioration in all three functional areas of communication, mobility and cognition.

5.3.1. Communication

Drilling down further into individual participant data represented a more complex picture of deterioration across all three domains. Figure 5.5 visually plots out communication scores over time for each participant in the first phase. Since many of the participants had identical Hamburg scores, the plot graph illustrates the data in a 3D format so that all participant plot lines are visible.

Figure 5. 5: Participants' Hamburg communication scores over three years



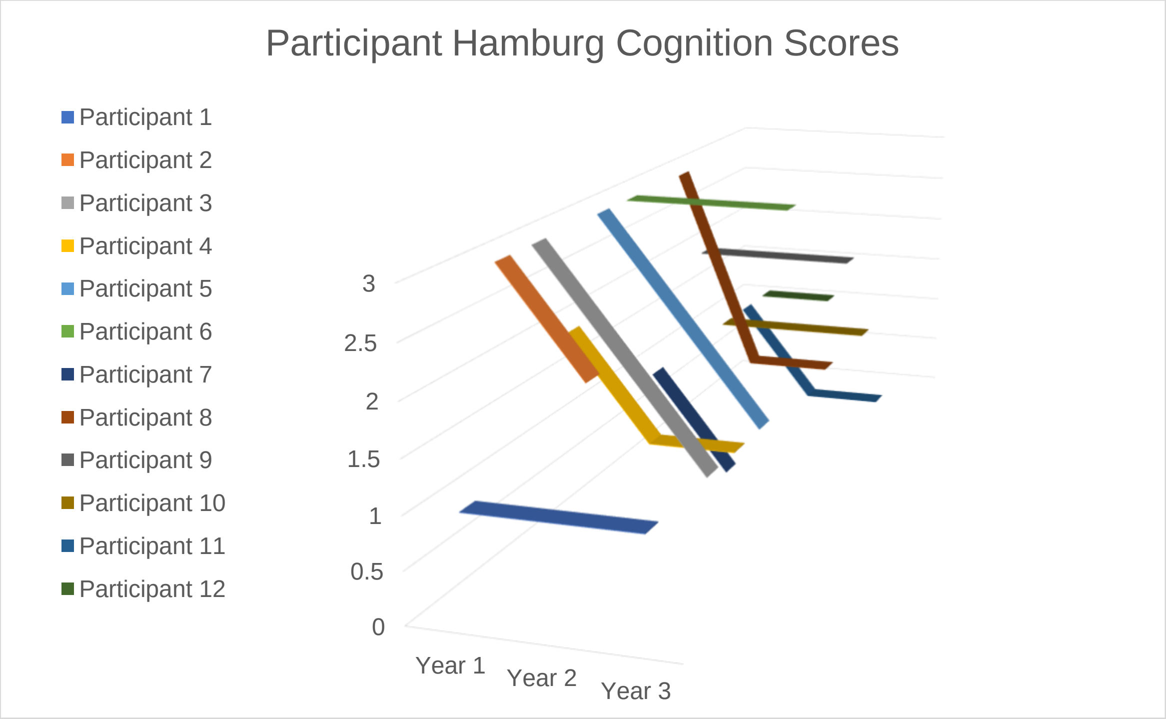
As can be seen from the plot graph, some participants had the lowest level of communication score (0) from the beginning to the end of Phase One (n = 5/12), other participants showed deterioration in communication scores over time (n = 3/12) and others showed deterioration in the first year followed by maintained communication thereafter (n = 4/12). The overall trajectories here indicated some level of deterioration in all participants over the three years.

Building on previous literature relating to the presentation of children with JNCL (Elmerskog et al., 2019; von Tetzchner et al., 2019), and in line with research question 1.2, further exploration into this subgroup offered a representation of functional skills from a clinical perspective. As can be seen in Figure 5.5, mean Hamburg scores for participants with JNCL showed an initial period of deterioration followed by a period of maintained communication skills thereafter (participants four, six and eight). However, this pattern was also seen in participants with alternative diagnoses (i.e. participant nine who has the CLN2 variant). Inconsistent patterns here represent the diverse variability in scores making generalisations challenging between participants.

5.3.2. Cognition

Exploring the cognition scores in further detail also showed varied patterns. Figure 5.6 illustrates the plot graph of participants' cognition scores. As can be seen in this figure some participants, showed deteriorating cognition scores (n = 4/12), some participants showed initial deterioration followed by maintain cognition scores (n = 3/12), and others displayed consistent stable cognition scores throughout (n = 5/12). Results here indicate variable patterns in cognition scores, with both consistent and inconsistent deterioration and stability observed.

Figure 5. 6: Participant Hamburg cognition scores over three years

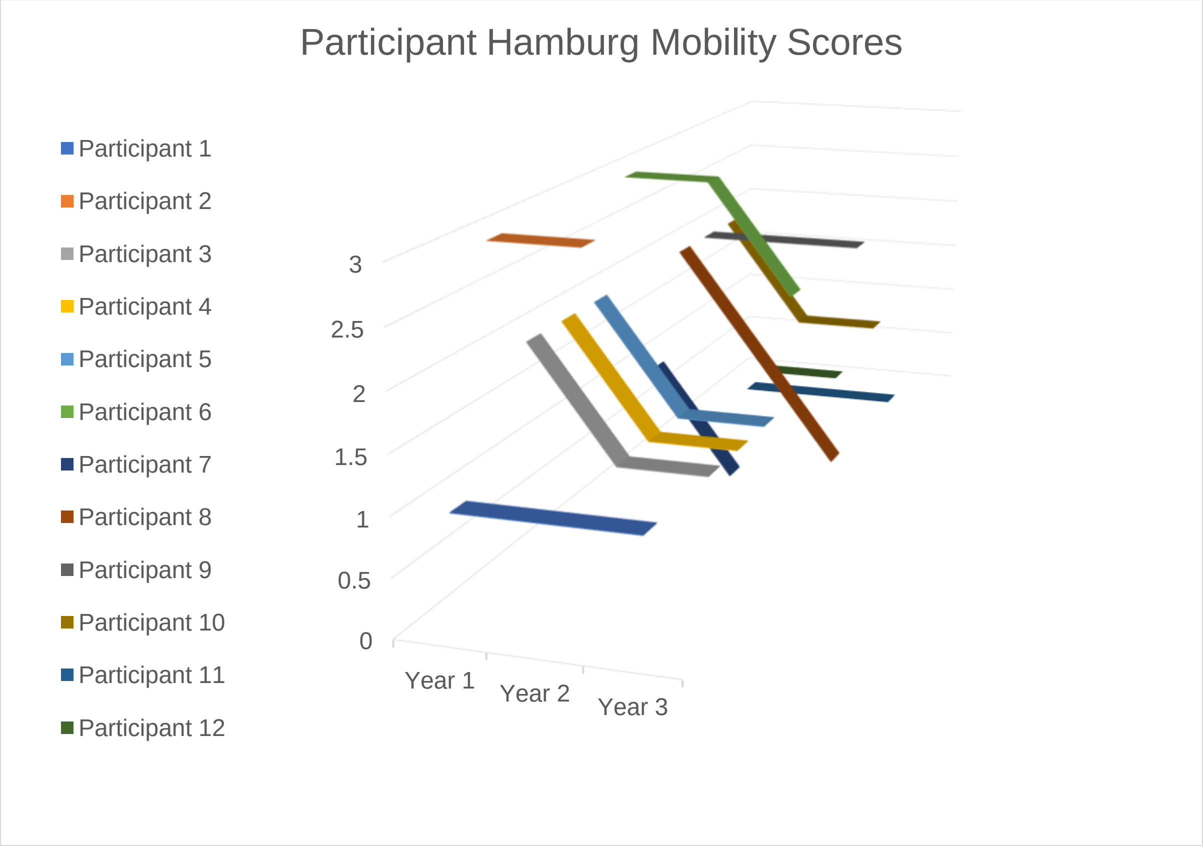


Examining data in the subgroup of participants with a diagnosis of JNCL offered a similar perspective of change over time for this participant group. As can be seen in Figure 5.6, mean Hamburg scores for participants with a JNCL diagnosis showed one participant with maintained cognition skills (participant six) and two participants showed initial deterioration followed by stable cognition skills (participants four and eight). As with data from the communication domain, data here showed variable patterns across the participants. Alongside participant six, other participants also showed maintained cognitive responses (participant one (CLN6), nine (CLN2), ten (CLN2) and twelve (CLN8)) and there were no unified patterns that were unique to the JNCL subgroup. Again, results demonstrate the challenge with generalisations by diagnosis and show no clear or consistent pattern for children affected by the Juvenile variant of Batten disease.

5.3.3. Mobility

Finally, in examining the mobility scores at a participant level, again, there were variable patterns observed. Figure 5.7 illustrates mobility scores for each participant.

Figure 5. 7: Participant Hamburg mobility scores over three years



As can be seen in this plot graph, participants either showed deteriorating mobility scores (n = 3/12), maintained mobility scores (n = 5/12), or initial deterioration followed by maintained scores (n = 4/12). Data here demonstrate wide variability in mobility scores, with inconsistent and consistent deterioration over time as well as maintained mobility.

Examining data from the subgroup of participants with a diagnosis of JNCL offered a similar perspective on change over time for this participant group. As can be seen in Figure 5.7, mean Hamburg scores for one participant with a JNCL diagnosis showed an initial period of deterioration followed by a period of maintained mobility skills thereafter (participant four), one participant showed stable mobility followed by deterioration (participant six) and one participant showed consistent deterioration (participant eight). Again, with the previous domains, inconsistent, variable patterns of deteriorating, or maintained mobility were shown in many participants across different variants of the disease, making subgroup generalisations challenging. Subsequent analysis focused on the CMTOM scores recorded within the three-year observations, to provide an indication of functional responses to music therapy (communication, behaviour, wellbeing, cognition, and mobility) and how these changed over time.

5.4. Research Question 1.3: How do functional outcomes (social, physical and cognitive) change in music therapy over time for children with Juvenile Batten disease?

Analysis of the CMTOM measure scores focused on descriptive statistics, to identify functional responses to music therapy and how these changed over three years. The use of descriptive statistics over other statistical analysis measures was deemed appropriate for the data collected, based on the small case study sample, the heterogeneity of the population, and the range of diagnoses and ages in the participant pool. A total of 182 session videos (an average of 15 per participant) were assessed by two researchers (me and another researcher officer). Both researchers using the CMTOM measure had received previous training in the tool before the research project. Scores were recorded for each of the CMTOM domains of Communication, Behaviour, Emotional expression, Cognition and Mobility.

5.4.1. Communication

The first category of communication consisted of 14 subcategories which were rated on a scale of 0-4 (responses observed 0=Never, 1=Rarely, 2=Sometimes, 3=Often, 4=Consistently). Appendix 15 shows participant mean scores and standard deviation values of the 14 communication subcategories over 19 data collection points (three years). Illustrating these data in a plot graph provides the opportunity to visually see how communication responses in music therapy change over time. Figure 5.8 plots mean communication scores each year in the 14 subcategories.

Figure 5. 8: Participants mean communication scores across time by subcategory

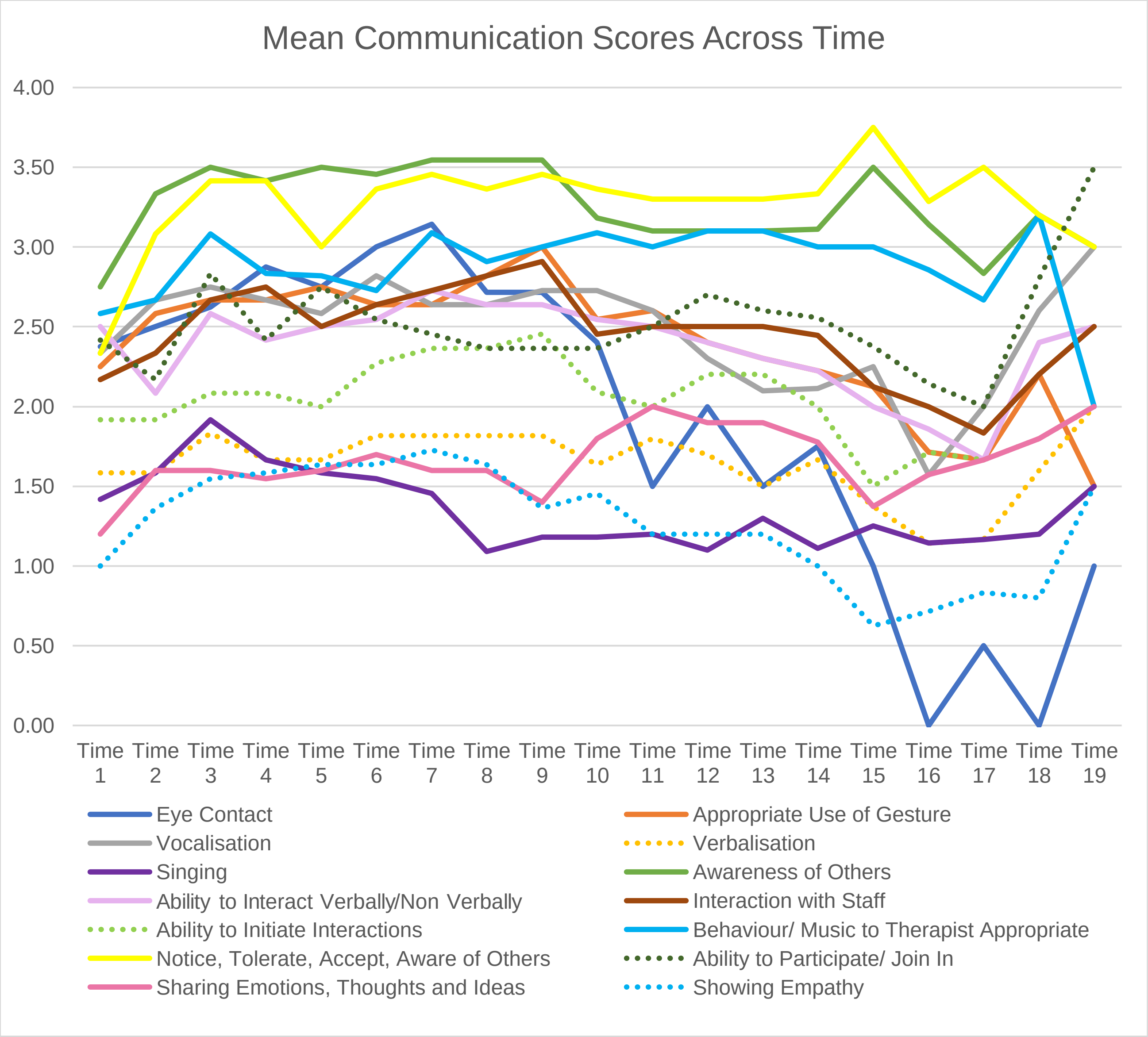


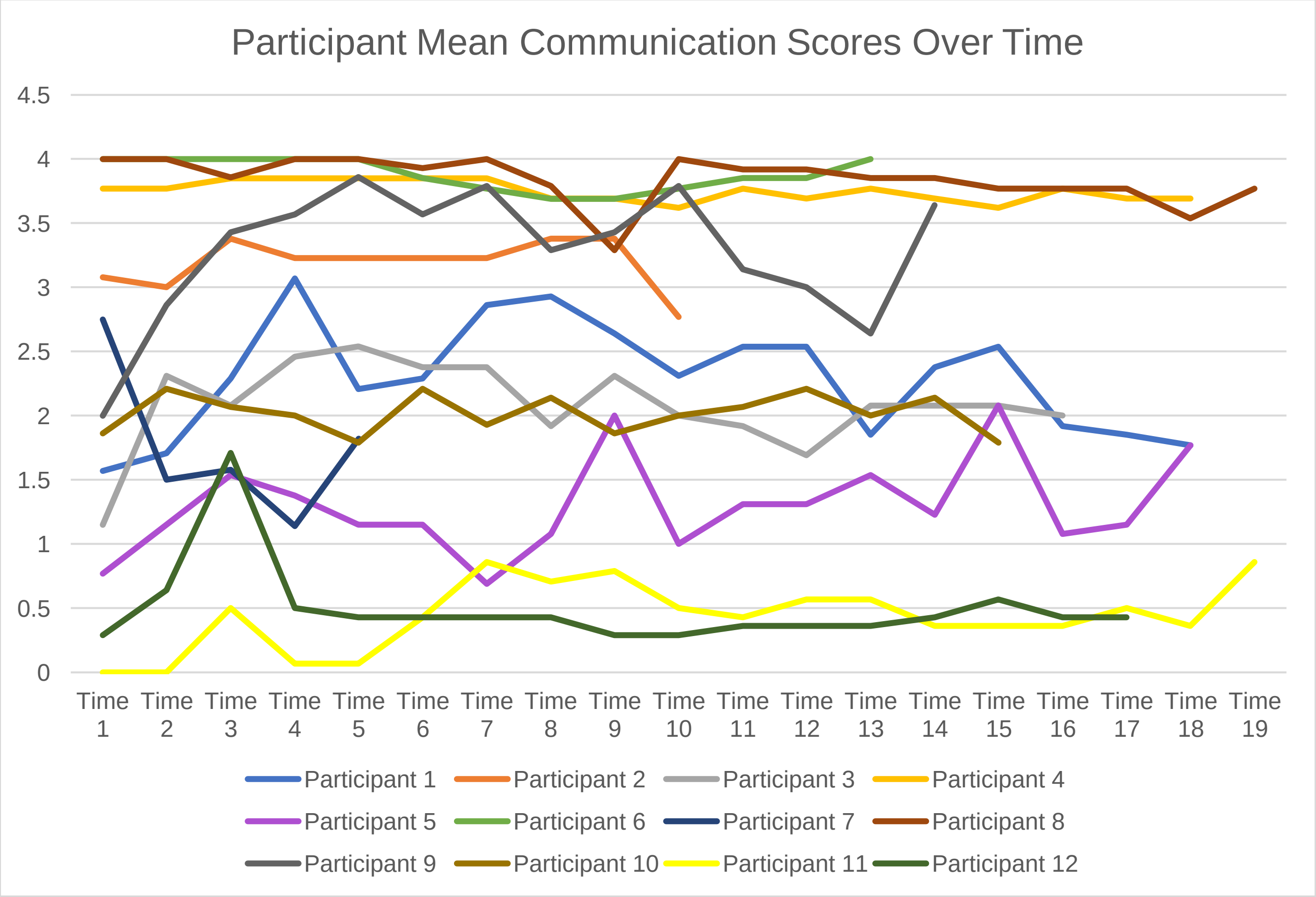
Figure 5.8 illustrates varied and undulated patterns across communication mean scores for participants over the 14 categories. From time points 1-14 (roughly years 1 and 2) however, the trajectories although varied (range = 1.00-3.5), show some stability over time, and some steady patterns can be seen in the plot graph. From time points 15-19 however, the scores showed a greater variance, with some participants showing sharp deterioration, others showing increased responses and others showing stability. This perhaps represents the variance of deterioration for each participant and indicates that in year three, participants began to show greater deteriorating responses.

To explore a broader overview of the data, time points were simplified to demonstrate change over each year. Mean scores from timepoints 1, 10 and 19 show how mean scores changed across baseline, mid-point, and endpoint stages.

Subcategories of ‘Vocalisation’, ‘Verbalisation’, ‘Showing Empathy’, ‘Sharing Emotions, Thoughts, and Ideas’ and ‘Ability to Initiate Interactions’, showed consistently increasing scores over time (n = 5/14). Although showing variable midpoint data, there were several subcategories that showed increased scores from baseline to endpoint data collection (n = 5/14). Subcategories of ‘Eye Contact’, ‘Behaviour to Music Therapist’ and ‘Appropriate Use of Gesture’ showed decreased scores over time (n = 3/14). The use of eye contact and gesture could perhaps reflect the nature of deteriorating visual impairments. One subcategory ‘Ability to Interact Verbally/Non-Verbally’ remained the same from the baseline to the endpoint of data collection.

Further analysis was conducted on communication scores for each participant in Phase One Figure 5.9 illustrates how the mean communication scores changed for each participant over the three years. Within this figure, it is also possible to see where participation stopped for some participants (for example participants two and seven only had limited CMTOM scores out of a total 19 time points as they left the research project in year two).

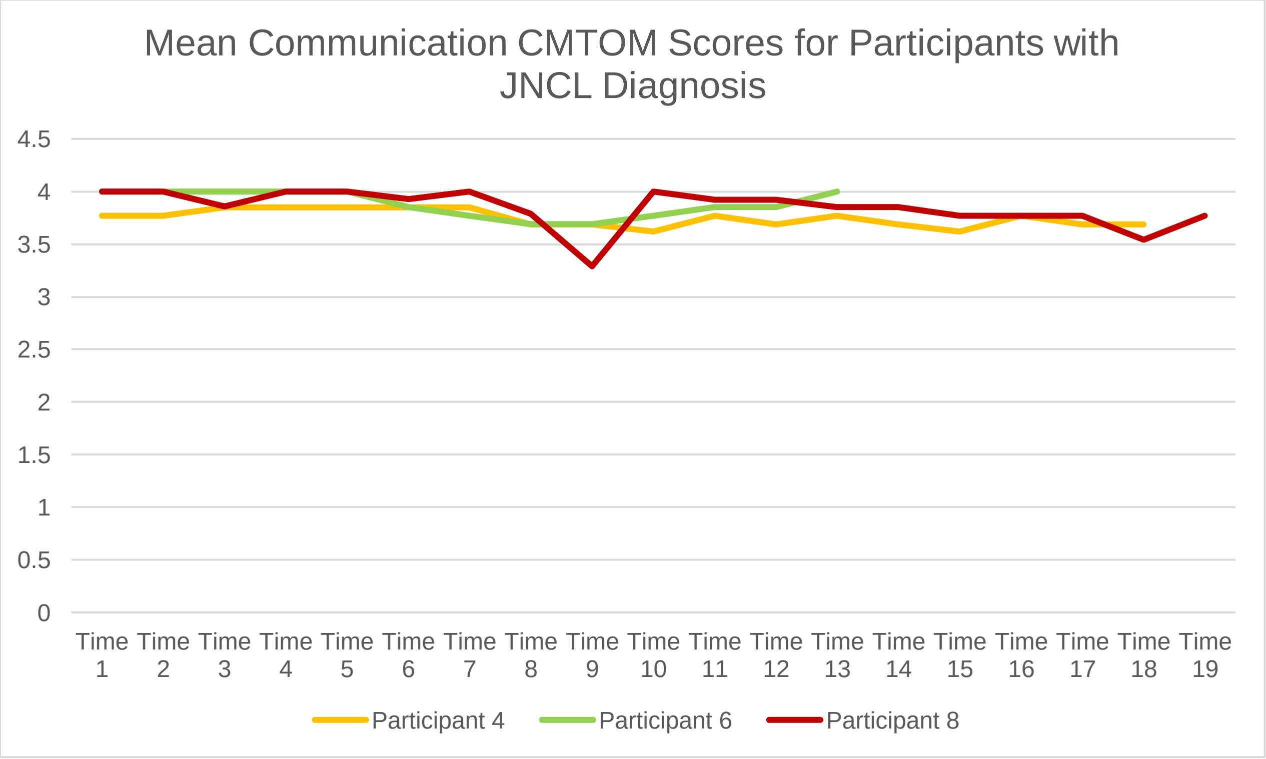
Figure 5. 9: Mean communication scores over 19 data collections points (three years), for each of the 12 participants



With mean scores spanning the entire range from 0-4, communication changes for each participant, mirror the heterogeneity of this participant sample and perhaps the Batten disease population in general. It could also be seen that some participants showed slight deterioration in responses (n = 3/12), some showed a plateaued response (n = 6/12), and in other cases there was a slight improvement in recorded responses (n = 3/12), indicating varied communication responses in music therapy over time. Data here, however, showed nine participants with an upward or steady pattern in their mean communication scores over time, suggesting that in music therapy, there is a chance that communication responses could be increased or maintained.

To analyse change over time for participants with a JNCL diagnosis, individual mean scores were extracted for the three participants with this diagnosis in this subgroup. Figure 5.10 shows mean communication scores for participants with JNCL diagnosis and how these change over time.

Figure 5. 10: Mean communication CMTOM scores for participants with JNCL diagnosis



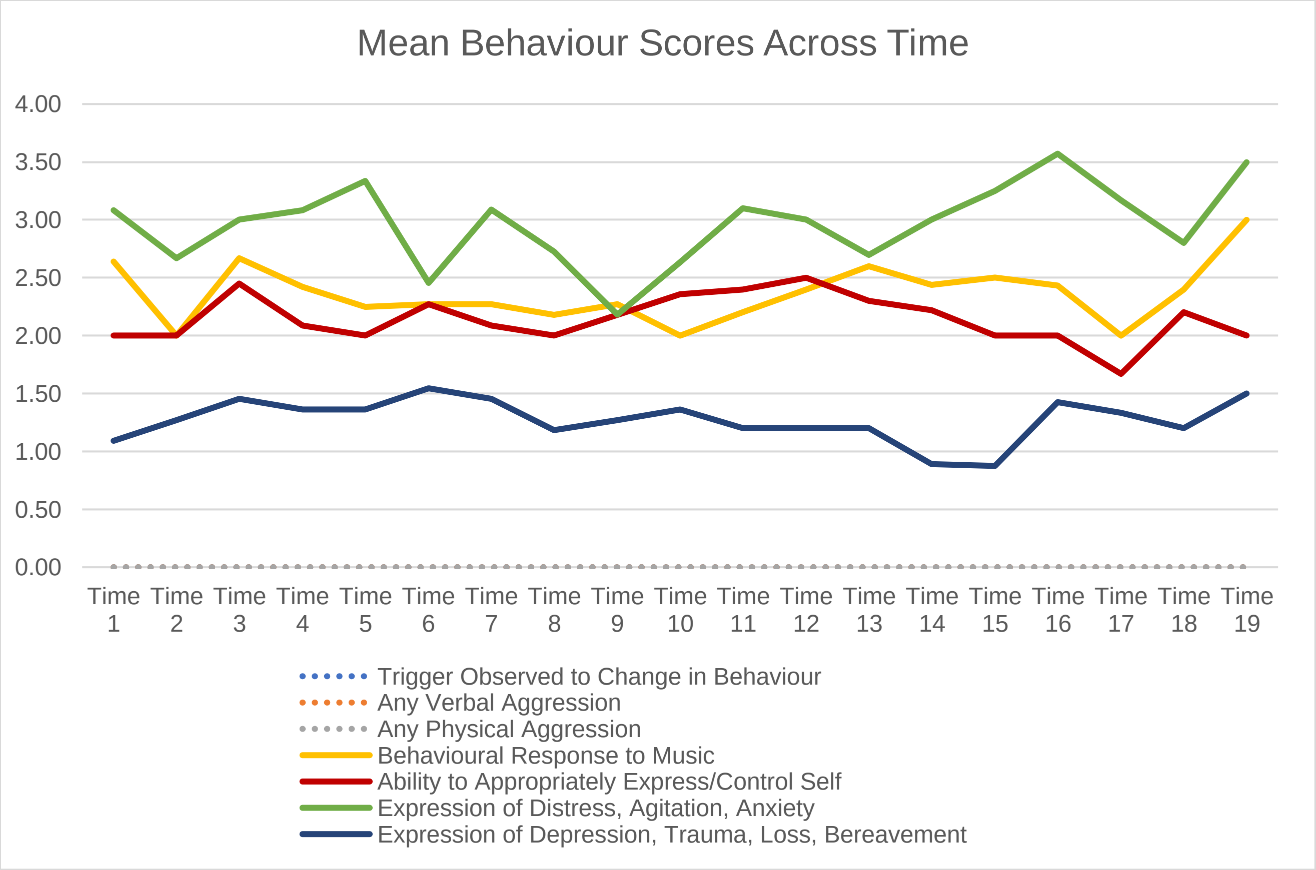
As can be seen from Figure 5.10 there is a distinct plateau effect observed in the individual data for the JNCL subgroup. Of the three participants, one participant showed maintained communication mean scores from baseline to endpoint (participant six) whilst the other two participants showed very slight deterioration over time (participants four and eight). Whilst there were some time points that show lower mean scores (participant eight, time point 9), there is an overall maintained picture of change in communication responses.

Data from the two assessment measures show discrepancies in communication scores. According to the Hamburg Clinical scores, some participants showed the lowest recorded scores over three years, where, no functional skills were recorded at all (participant three (CLN5), five (CLN5) and (CLN2) seven). For these same participants, the CMTOM mean scores were higher (there were no ‘0’ mean scores recorded at all), and the trajectories for these participants showed maintained communication responses over the three years. Data seem to demonstrate that the CMTOM measure was able to identify functional skills and maintained communication responses in affected children, in comparison to the Hamburg scale which showed global deterioration or non-existent function. The next section of analysis looks at behavioural responses recorded on the CMTOM for each of the 12 participants in music therapy.

5.4.2. Behaviour

The behaviour category consisted of seven subcategories which were rated on the same assessment scale of 0-4. Appendix 16 shows participant mean scores and standard deviation values of the seven behaviour subcategories over 19 data collection points (three years). As with the communication domain, the data were visually illustrated to provide the opportunity to observe how behavioural responses in music therapy change over time. Figure 5.11 plots mean behaviour scores each year in the seven subcategories.

Figure 5. 11: Participants mean behaviour scores across time, by subcategory



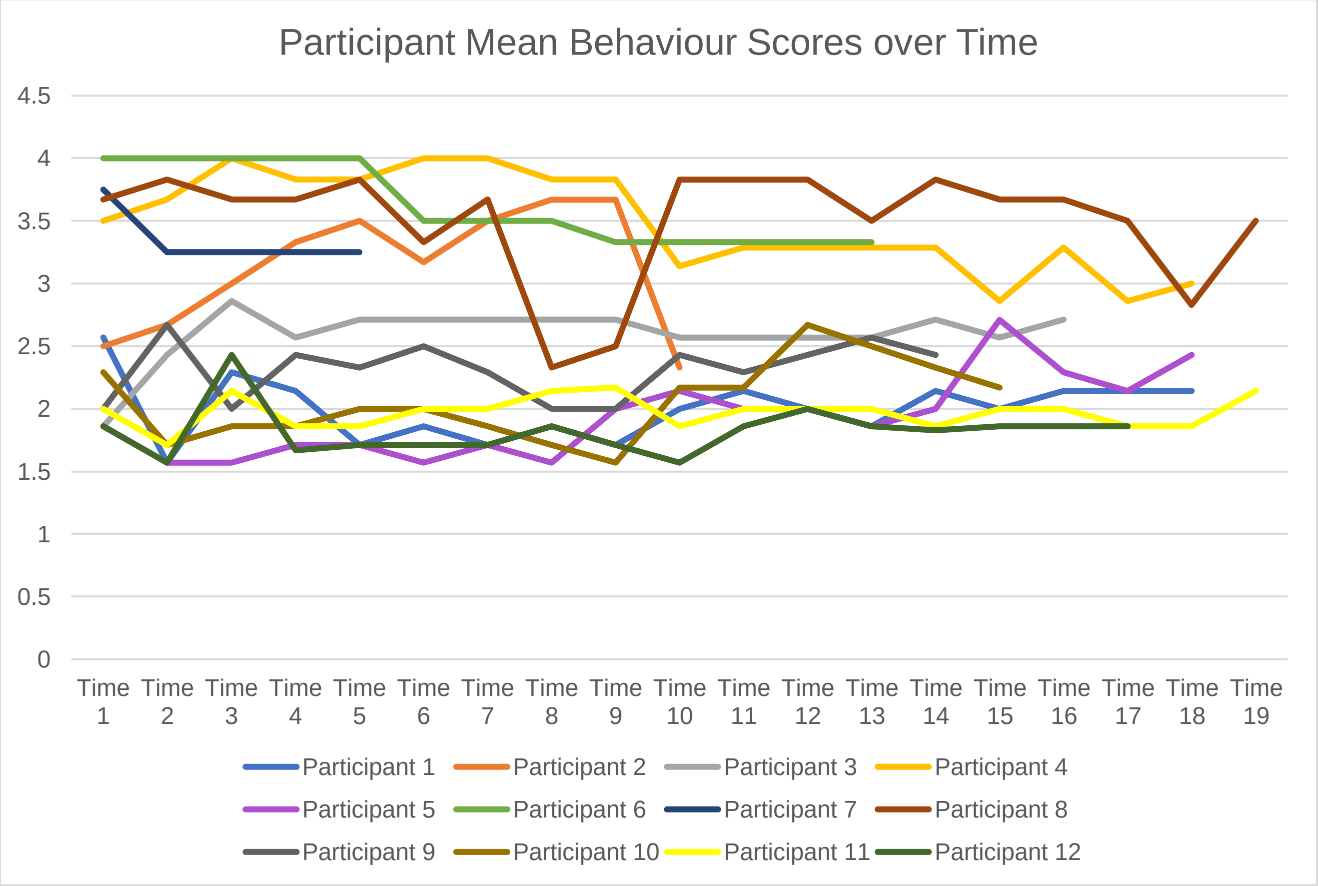
According to mean scores across all participants, Physical Aggression and Verbal Aggression produced mean scores were '0' over the three years. The subcategory 'Triggers Observed to Change Behaviour' (i.e. variables such as seizure activity, mood disturbances, fatigue, illness etc) also consistently scored '0' throughout the three years, suggesting changes in behaviour were due to influences within the music therapy session itself, rather than external influences. These three subcategories are represented as dotted lines in the plot graph overlapping one another at the bottom of the graph. As with the previous communication domain, mean behaviour scores varied between the remaining four subcategories (range = 0.88-4.00). Across the remaining subcategories, there was a stable pattern observed with regards to the 'Ability to Express/Control Self', 'Expression of Depression, Trauma, Loss Bereavement', 'Behavioural Responses to Music', and 'Expression of Distress, agitation and Anxiety'. It is worth noting that in accordance with the principles of music therapy (BAMT, 2017), expression of both negative and positive feelings is seen as a 'positive' response in music therapy, where it is seen as a positive outcome that a child can fully express themselves.

Taking a broader approach to the analysis, behavioural responses across three distinct time points showed a similar picture reviewing time points 1, 10 and 19. One subcategory 'Expression of Depression, Trauma, Loss or Bereavement' showed consistently increased scores over time (n = 1/7). Despite fluctuations in mid-point data, some subcategories showed increased scores at the endpoint in comparison to baseline data. These were: ‘Behavioural Responses to Music’ and ‘Expression of Distress, Agitation, Anxiety’ (n = 2/7). One subcategory, ‘Ability to Appropriately Express/Control Self’, remained unchanged from baseline to endpoint data (n = 1/7).

The data demonstrated overall stable or increased positive behavioural responses over time (i.e. self-expression of positive and negative feelings), and absent negative behaviour (i.e. behavioural triggers, verbal or physical aggression). The results suggested a potential for regulation and stability of behavioural responses in music therapy, and that perhaps music therapy could help reduce negative behavioural responses.

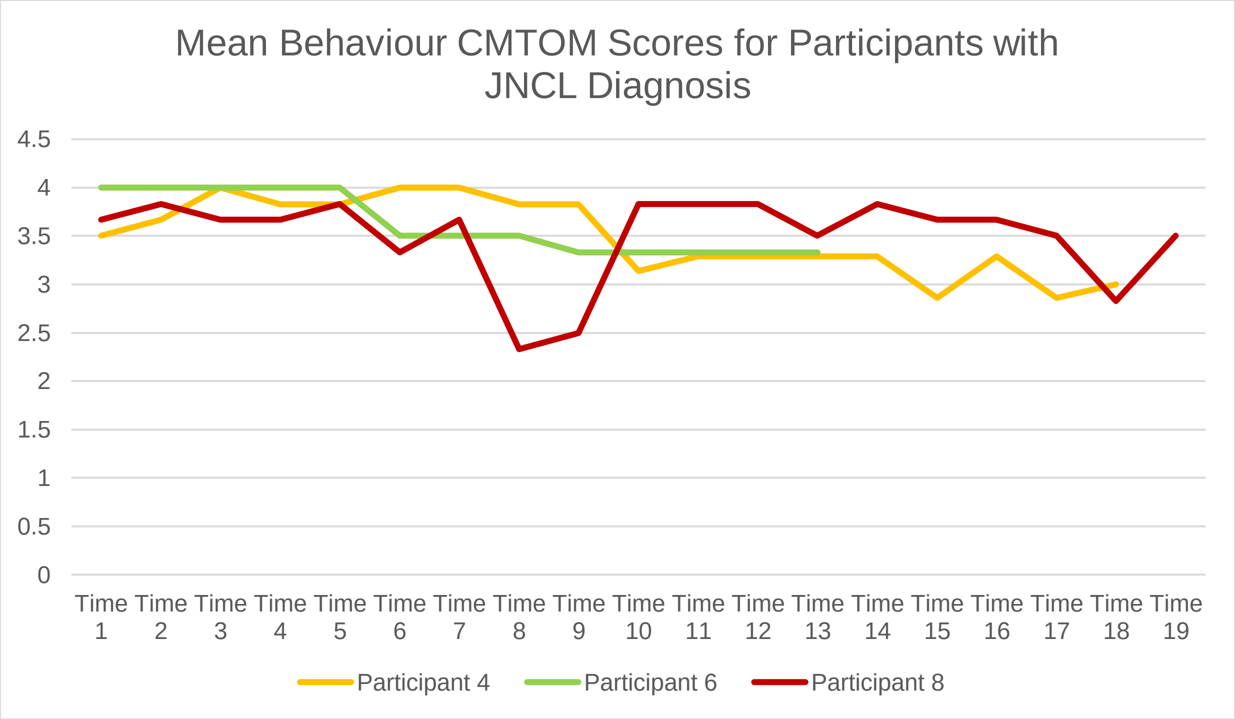
Further analysis was conducted on mean behaviour scores for each participant in Phase One. For mean participant scores, the subcategories ‘Physical Aggression’ and ‘Verbal Aggression’ were reverse scored where they reflected ‘negative’ behavioural responses in music therapy sessions. Responses observed for these categories were scored as 0=Consistently, 1=Often, 2=Sometimes, 3=Rarely, 4=Never. A score of 4 therefore, reflected a response ‘never’ occurring (which was deemed to be a ‘positive’ behavioural response). Figure 5.12 illustrates how the mean behaviour scores changed for each participant over the three years.

Figure 5. 12: Mean behaviour scores over 19 data collections points (three years) for each of the 12 participants



From the individual participant data, again there were variable mean behavioural scores observed over time. Participants in some cases showed decreased scores from at their endpoint in comparison to their baseline (n = 5/12) unchanged scores at their endpoint in comparison to their baseline to endpoint (n = 1/12) or increased scores at endpoint compared with their baseline (n = 6/12). From the baseline to the endpoint, values indicated mixed behavioural responses to music therapy by each participant. Whilst some children displayed increased or decreased behavioural responses from baseline to endpoint, most patterns fluctuated throughout. To analyse change over time for participants with the JNCL diagnosis, individual mean scores were extracted for the three participants in this subgroup. Figure 5.13 shows mean behaviour scores for participants with JNCL diagnosis and how these change over time.

Figure 5. 13: Mean behaviour CMTOM scores for participants with JNCL diagnosis

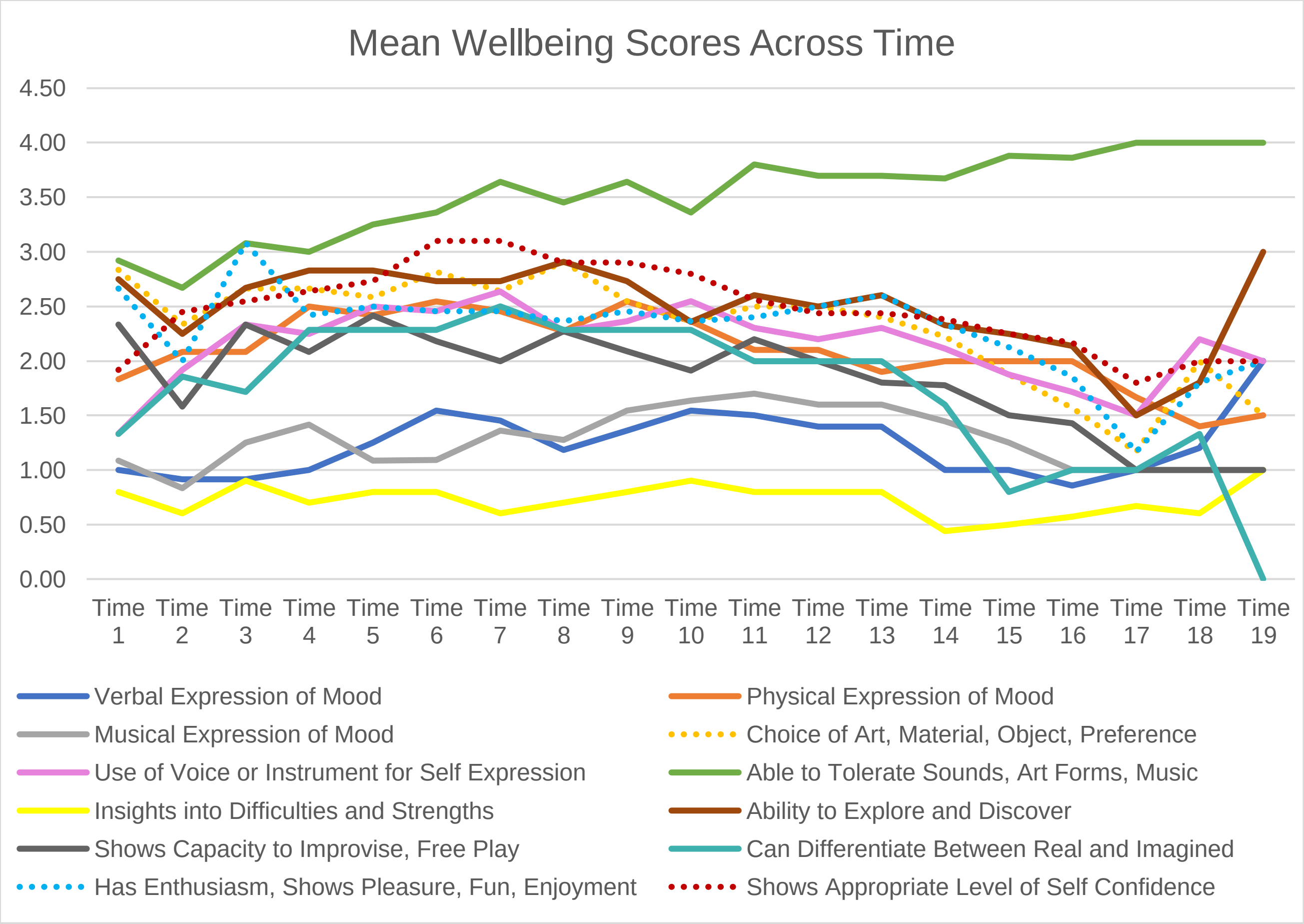


As can be seen from Figure 5.13 there is a slight downward pattern observed in the individual data for the JNCL subgroup and all mean scores for behaviour deteriorated slightly over time. In line with other domains there are some time points that show lower mean scores (participant eight, time points 8 and 9), however, the overall picture of change is one of gradually deteriorating behavioural responses in music therapy over time. The next section of the analysis focuses on patterns emerging in the wellbeing domain of the CMTOM measure.

5.4.3. Wellbeing

The wellbeing category consisted of 12 subcategories which were rated on the same assessment scale of 0-4. Appendix 17 shows participant mean scores and standard deviation values of the 12 wellbeing subcategories over 19 data collection points (three years). As with previous domains, the data were visually illustrated to provide the opportunity to observe how wellbeing responses in music therapy changed over time. Figure 5.14 plots mean wellbeing scores each year in the 12 subcategories.

Figure 5. 14: Participants’ mean wellbeing scores across time, by subcategory

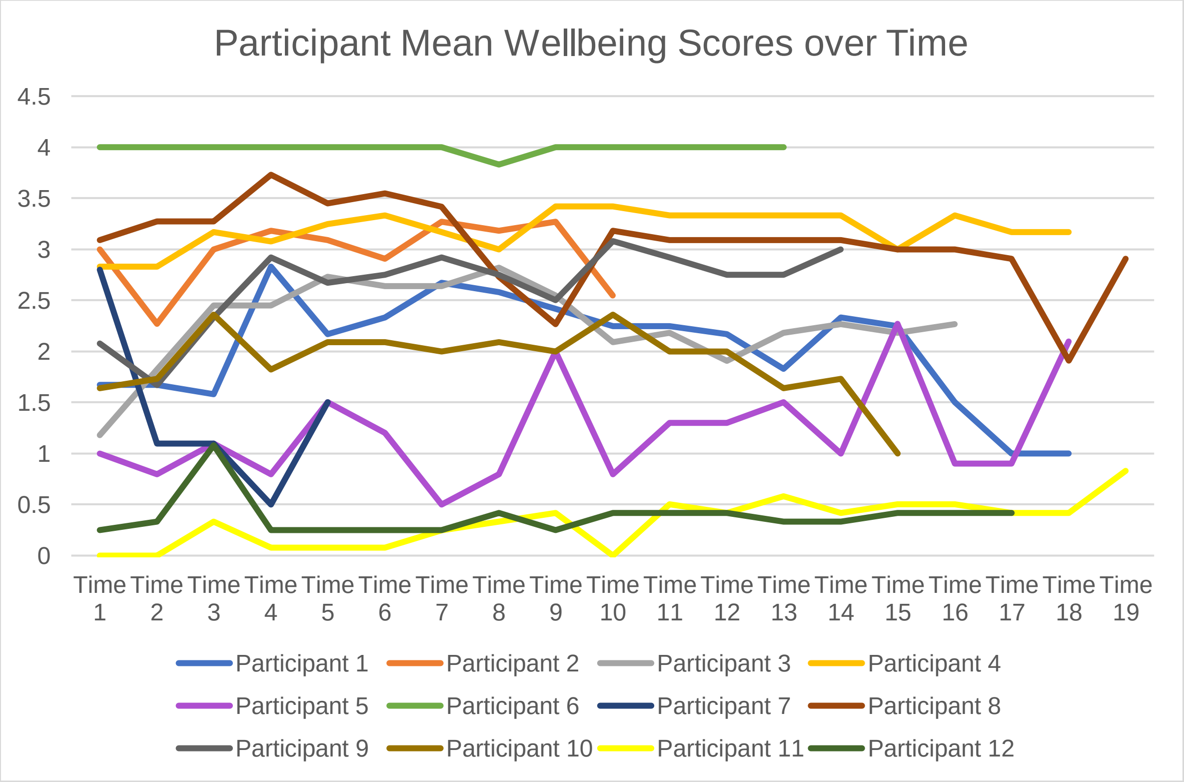


Wellbeing scores in this domain showed a wide variance (range = 0.00-4.00) and undulated patterns were observed for participants in each of the 12 categories. From time points 4-17 however, the participants’ data, although varied (range = 0.7-4.00), were relatively stable over time, and steady patterns can be seen in the plot graph. From early time points (1-4) and later time points (17-19), the trajectories show a greater variance, where some participants showed sharp deterioration, others showed increased responses and others remained in stasis. This suggests that wellbeing for each participant remained most stable in the middle portion of their therapeutic input when the therapy intervention and practitioner was familiar and consistent. Earlier variance in wellbeing patterns could be reflective of children getting used to the Music Therapist and music practitioner who, in most cases, was new to the child, and could also be reflective of the child becoming familiar with the music sessions. Later variance in wellbeing responses, could be because participants knew they were approaching the end of therapy, (although this was largely speculative as all children were given the option to continue receiving music therapy after the end of Phase One, and it is unknown in some cases, how much children understood their therapy duration). The unique plateau effect observed here, could indicate the importance of consistency and familiarity for children’s wellbeing.

Wellbeing responses across three distinct time points (1, 10 and 19) showed a contrasting picture). There was a more even spread observed in the responses which, depending on the domain measured, increased, decreased or remained the same over time. Three subcategories, 'Verbal Expression of Mood', 'Able to Tolerate Sounds, Art Forms, Music', and 'Insights into Difficulties and Strengths' showed consistently increased scores over time (n = 3/12). Despite fluctuations in mid-point data, some subcategories showed increased scores at the endpoint in comparison to baseline data; 'Musical Expression of Mood', 'Use of Voice or Instrument for Self-expression', 'Shows Appropriate Level of Self Confidence' (n = 4/12). The subcategories 'Physical Expression of Mood', 'Choice of Art, Material, Object, Preference', 'Shows Capacity to Improvise, Free Play', 'Can Differentiate Between Real and Imagined' and 'Has Enthusiasm, Shows Pleasure, Fun, Enjoyment' showed deteriorating scores from baseline to endpoint data (n = 5/12). The data here indicate that participants' wellbeing both increased and decreased in music therapy over time depending on the subcategory of wellbeing.

Looking more closely at individual participant wellbeing scores, further analysis was conducted on mean wellbeing scores for each participant. Figure 5.15 illustrates how the mean wellbeing scores changed for each participant over the three years.

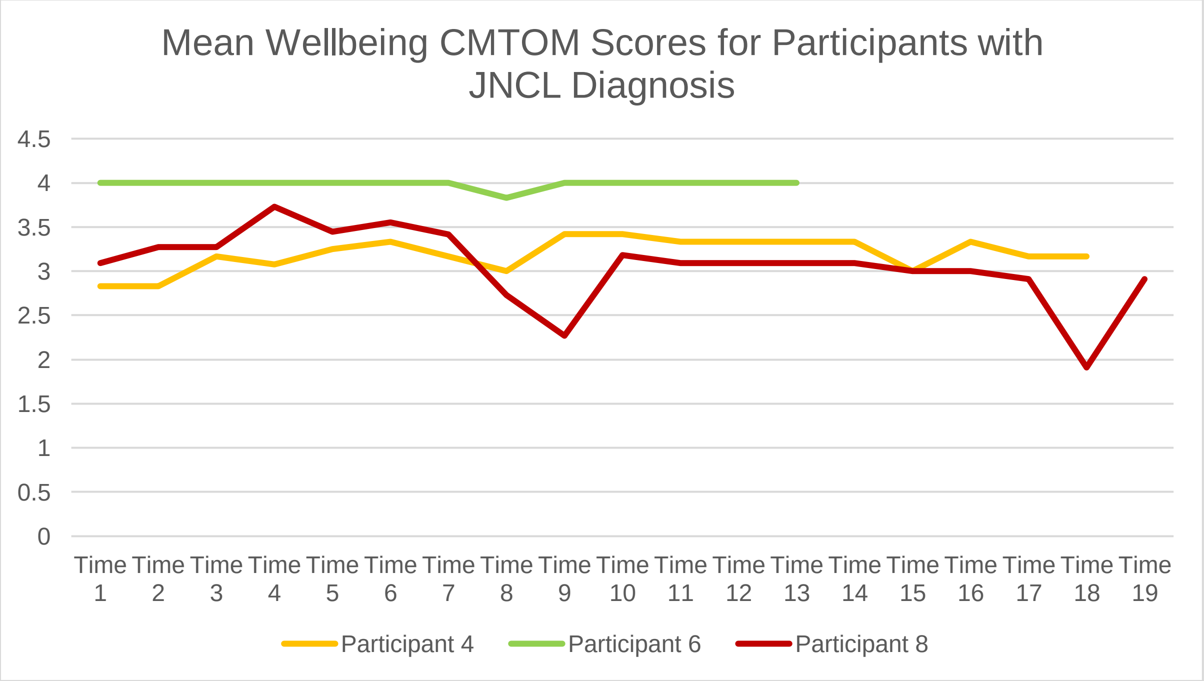
Figure 5. 15: Mean wellbeing scores over 19 data collections points (three years) for each of the 12 participants



From the individual participant data, most participants showed undulating patterns, and there was a wide variance in the range of scores for participants (range = 0.00-4.00). Participants in some cases showed decreased scores at their endpoint in comparison to their baseline (n = 7/12), unchanged scores at their endpoint in comparison to their baseline to endpoint (n = 1/12) or increased scores at endpoint compared with their baseline (n = 4/12). Despite observed plateau patterns in the overall participant mean data, the individual participant data demonstrated a variable picture of deteriorating, maintained or increased wellbeing responses from different participants over time.

As with previous domains, change over time was analysed for participants with JNCL, and individual mean scores were extracted for the three participants in this subgroup. Figure 5.16 shows mean wellbeing scores for participants with a JNCL diagnosis and how these change over time.

Figure 5. 16: Mean wellbeing CMTOM scores for participants with JNCL diagnosis

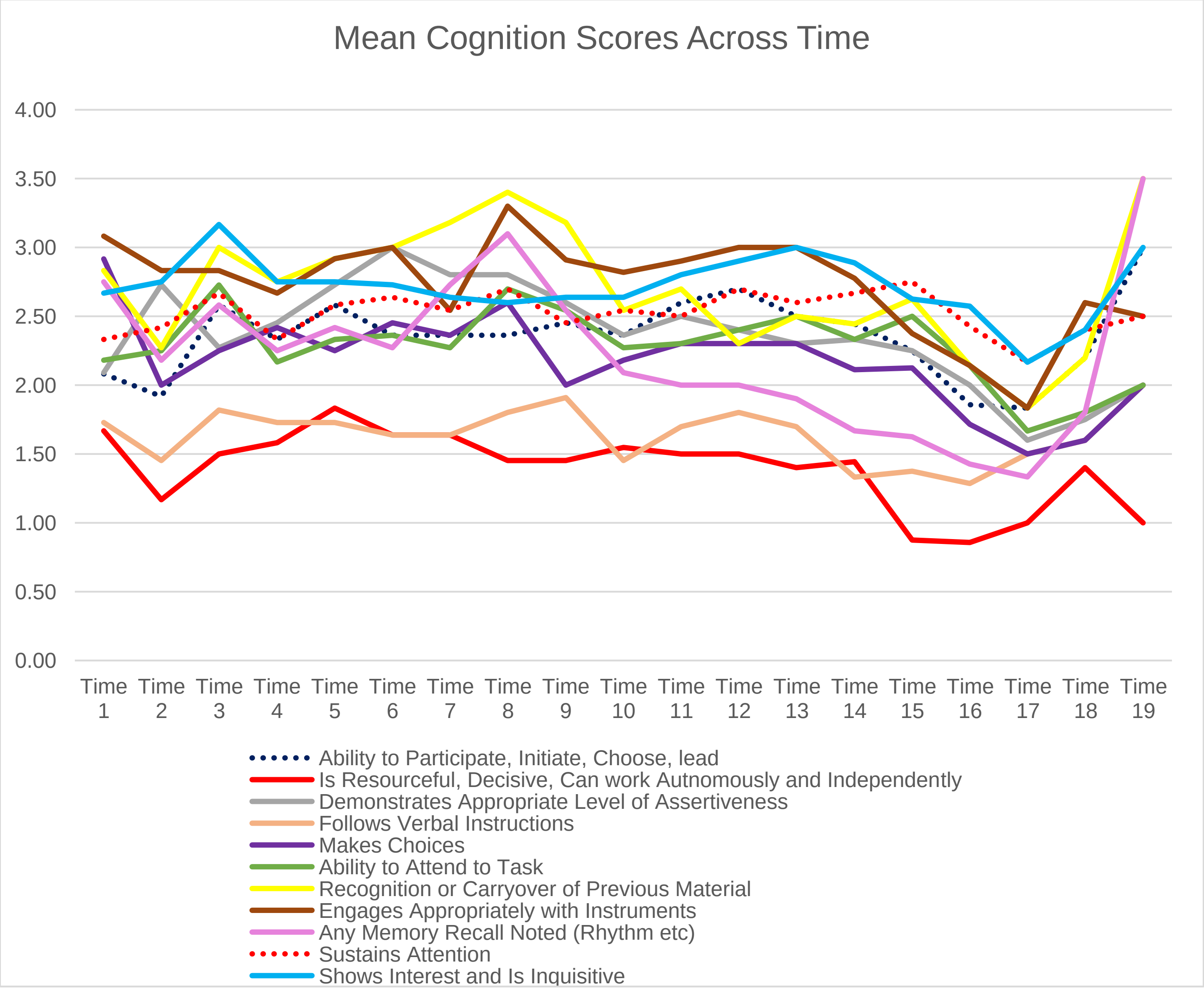


Again, as can be seen in previous domains, Figure 5.16 displays a steady pattern observed in the individual data for the JNCL subgroup. Of the three participants, two participants showed increased or maintained mean scores from baseline to endpoint data (participants four and six) and one participant showed slight deterioration in their mean wellbeing scores from baseline to endpoint (participant eight). In line with previous domains there are some time points which show lower mean scores (participant eight, time point 8 and 9), however, the overall picture of change is one of maintained wellbeing responses in music therapy over time. The next section of the analysis focuses on patterns emerging in the cognition domain of the CMTOM measure.

5.4.4. Cognition

The cognition category consisted of 11 subcategories which were rated on the same assessment scale of 0-4. Appendix 18 shows participant mean scores and standard deviation values of the 11 cognition subcategories over 19 data collection points (three years). As with previous domains, the data were visually illustrated to show how cognitive responses in music therapy change over time. Figure 5.17 plots mean cognitive scores each year in the 11 subcategories.

Figure 5. 17: Participants mean cognition scores across time, by subcategory

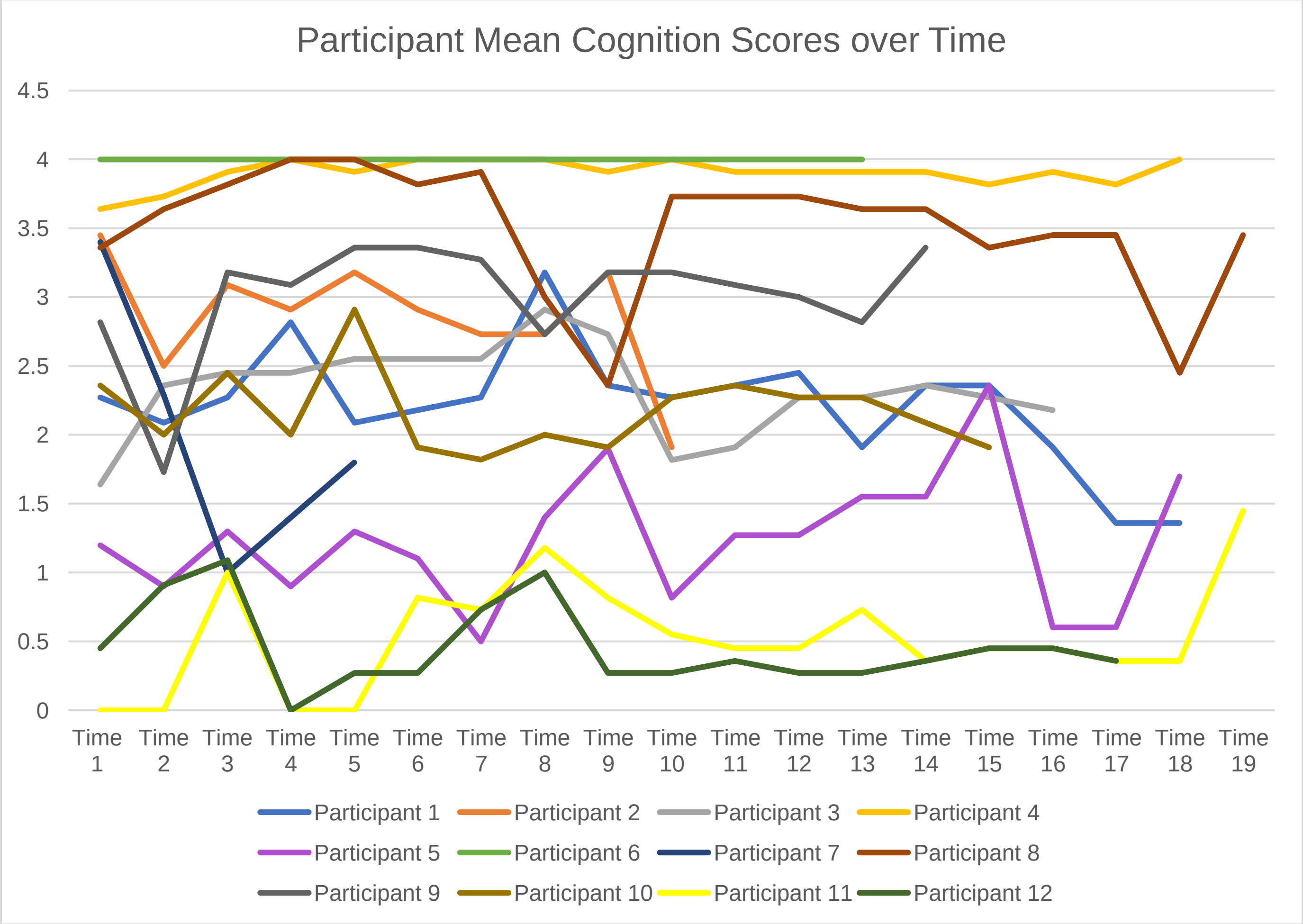


Cognition scores in this domain showed variable mean scores (range = 0.86-3.50) and as with other domains, undulated patterns were observed in participants over the 11 categories. From time points 2-18 however, the participants' data, although varied (range = 1.17-3.00), were relatively steady over time, shown in the stable patterns in the plot graph. In early time points (1-2) and later time points (18-19) the patterns showed a greater variance, where some participants showed sharp deterioration, others showed increased responses and others remained in stasis. As with the wellbeing domain, this maintained mid-stage response could be reflective of the impact of consistency and familiarity, which could yield more stable responses where therapy input is familiar and consistent.

Again, looking at the mean cognition values at three distinct time points 1, 10, and 19, represented a different picture. Observed responses at these three time points showed both increasing mean scores and decreasing mean scores. One subcategory, 'Ability to Participate, Initiate, Choose, lead'
showed consistently increased scores over time. Despite fluctuations in midpoint data, some subcategories showed increased scores from the baseline to the endpoint, namely: 'Follows Verbal Instructions', 'Recognition or Carryover of Previous Material', 'Any Memory Recall Noted (Rhythm etc)', 'Sustains Attention' and 'Shows Interest and Is Inquisitive' (n = 5/11). The remaining subcategories showed deterioration over time, namely: 'Is Resourceful, Decisive, Can work Autonomously and Independently', 'Demonstrates Appropriate Level of Assertiveness', 'Makes Choices', 'Ability to Attend to Task', and 'Engages Appropriately with Instruments' (n = 6/11). Data perhaps indicate that memory slightly improved over the three years of music therapy input, whereas independent skills (such as choice-making, engagement and assertiveness) decreased over time.

Looking more closely at individual participant cognitive scores, further analysis was conducted on mean scores for each participant. Figure 5.18 illustrates how the mean cognitive scores changed for each participant over the three years.

Figure 5. 18: Mean cognition scores over 19 data collections points (three years) for each of the 12 participants



From the individual participant data, there were observed fluctuating patterns in the mean cognition scores, and a wide variance in the range of scores for participants (range = 0.00-4.00). Participants in some cases showed decreased scores at their endpoint in comparison to their baseline (n = 4/12), unchanged scores at their endpoint in comparison to their baseline to endpoint (n = 1/12) or increased scores at endpoint compared with their baseline (n = 7/12). The data indicate cognitive responses from the participants were incredibly varied both in scores and trajectories over time. Although the overall mean cognition scores showed a stable period in the mid-stages of music therapy, individual participant data represent the heterogeneity of the participant pool and the variance in mean scores over the three years.

Individual participant data, however, for children with JNCL showed a different picture of change over time. Figure 5.19 shows mean cognition scores for participants with JNCL and how these change over time.

Figure 5. 19: Mean cognition CMTOM scores for participants with JNCL diagnosis

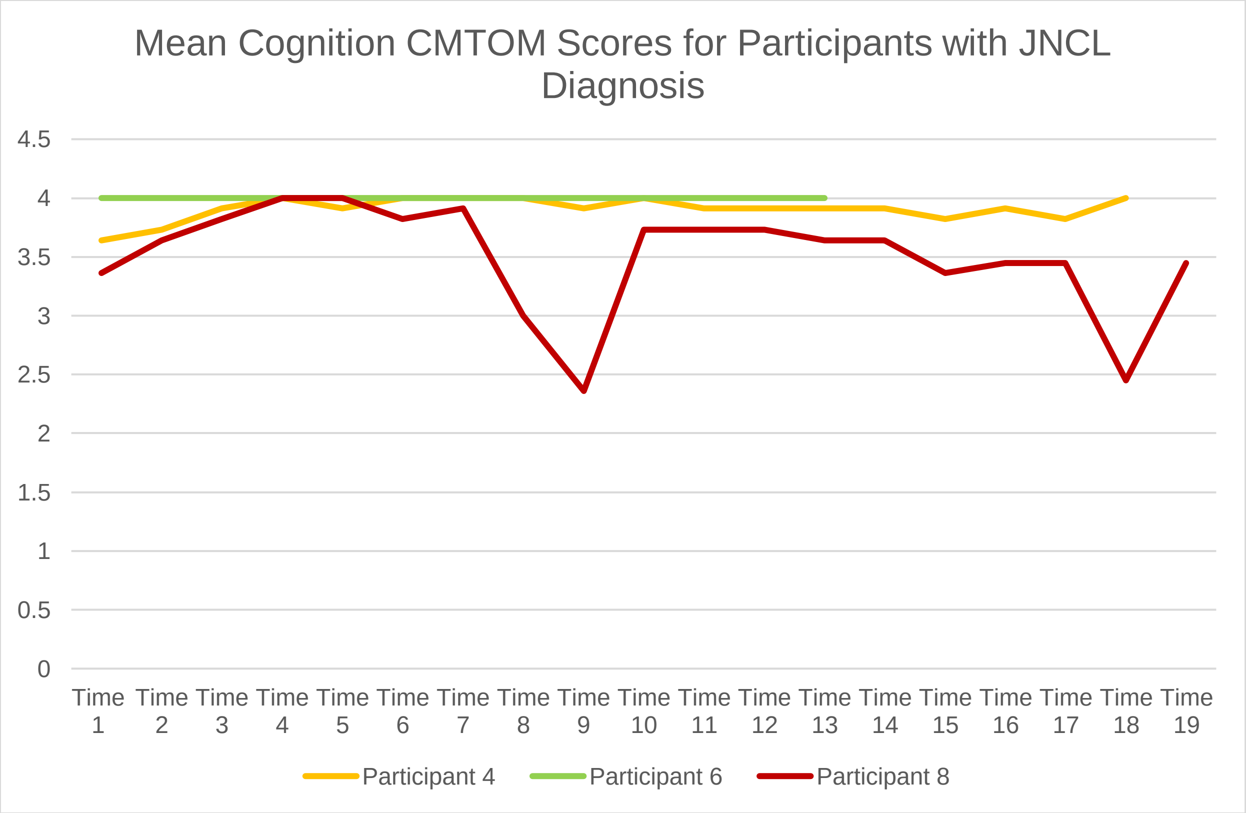


Figure 5.19 shows a plateau pattern observed in the individual data for the JNCL subgroup. Of the three participants, all participants showed increased or maintained mean cognitive scores from baseline to endpoint. As with the previous domains there are some time points that show lower mean scores (participant eight, time point 8 and 9); however, the overall pattern is maintained.

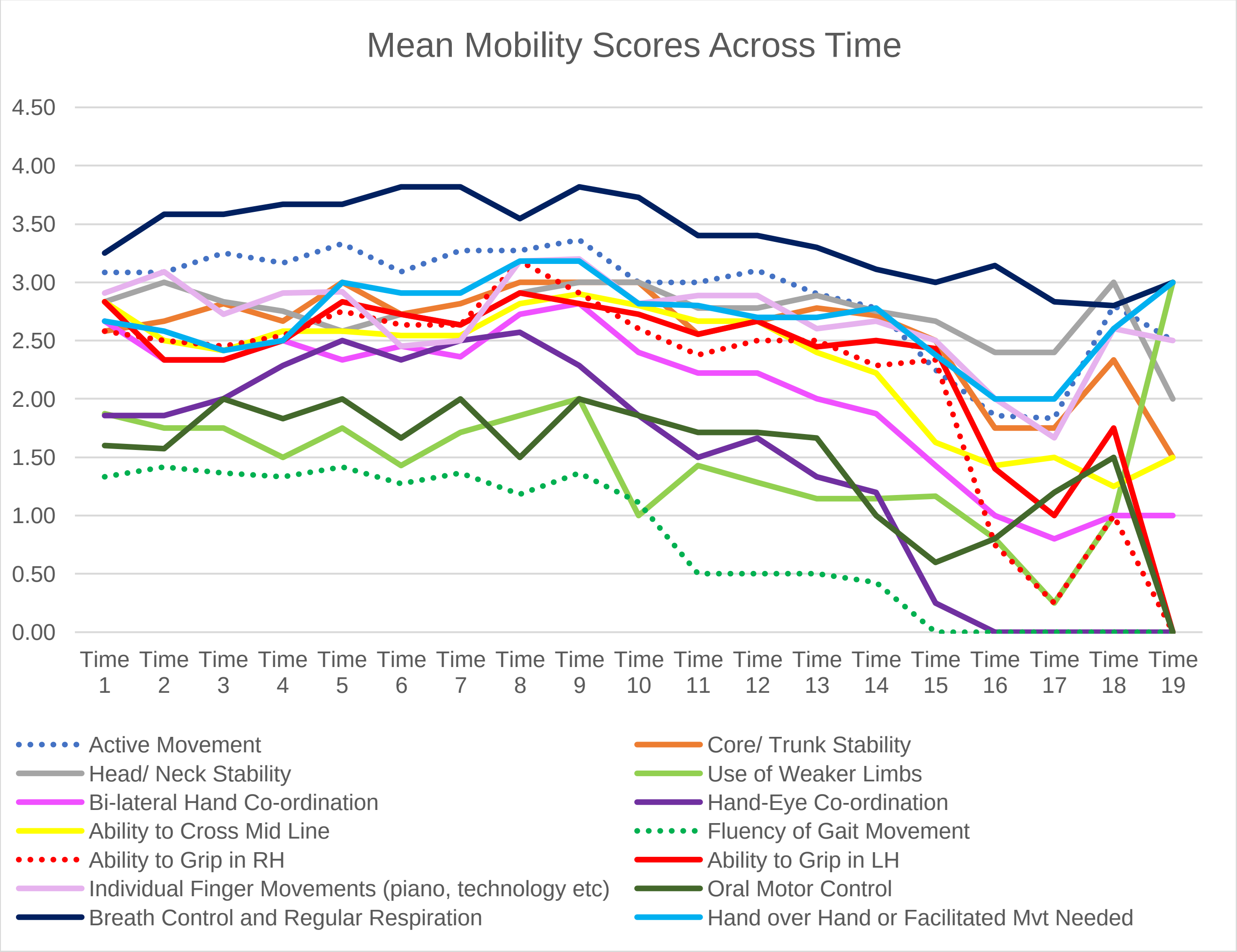
There were inconsistencies between the two assessment measures used. Hamburg means values in this category showed no clear or consistent pattern for children affected by the Juvenile variant of Batten disease. In contrast, trajectories according to the CMTOM scores for the JNCL subgroup displayed an overall plateau effect, suggesting that cognitive skills in music therapy could be maintained. Differences in patterns between the two measures suggest the CMTOM measure was able to identify the nuances of cognitive skills in children's responses to music therapy, in comparison to the Hamburg Scale which showed variable patterns across all participants. The final section of this analysis focuses on patterns emerging in the mobility domain of the CMTOM measure.

5.4.5. Mobility

The mobility category consisted of 14 subcategories which were rated on the same assessment scale of 0-4. In this domain, one subcategory 'Hand over Hand or Facilitated Movement' was reverse scored (it was deemed that a child requiring more hand over hand support would reflect less mobility in music therapy sessions). Responses observed for these categories were scored as 0=Consistently, 1=Often, 2=Sometimes, 3=Rarely, 4=Never. A score of 4 therefore, reflects a response 'never' occurring. Appendix 19 shows participant mean scores and standard deviation values of the 14 mobility subcategories over 19 data collection points (three years).

As with previous domains, the data were visually illustrated to provide the opportunity to observe how mobility responses in music therapy change over time. Figure 5.20 plots mean mobility scores each year in the 14 subcategories.

Figure 5. 20: Participants mean mobility scores across time, by subcategory



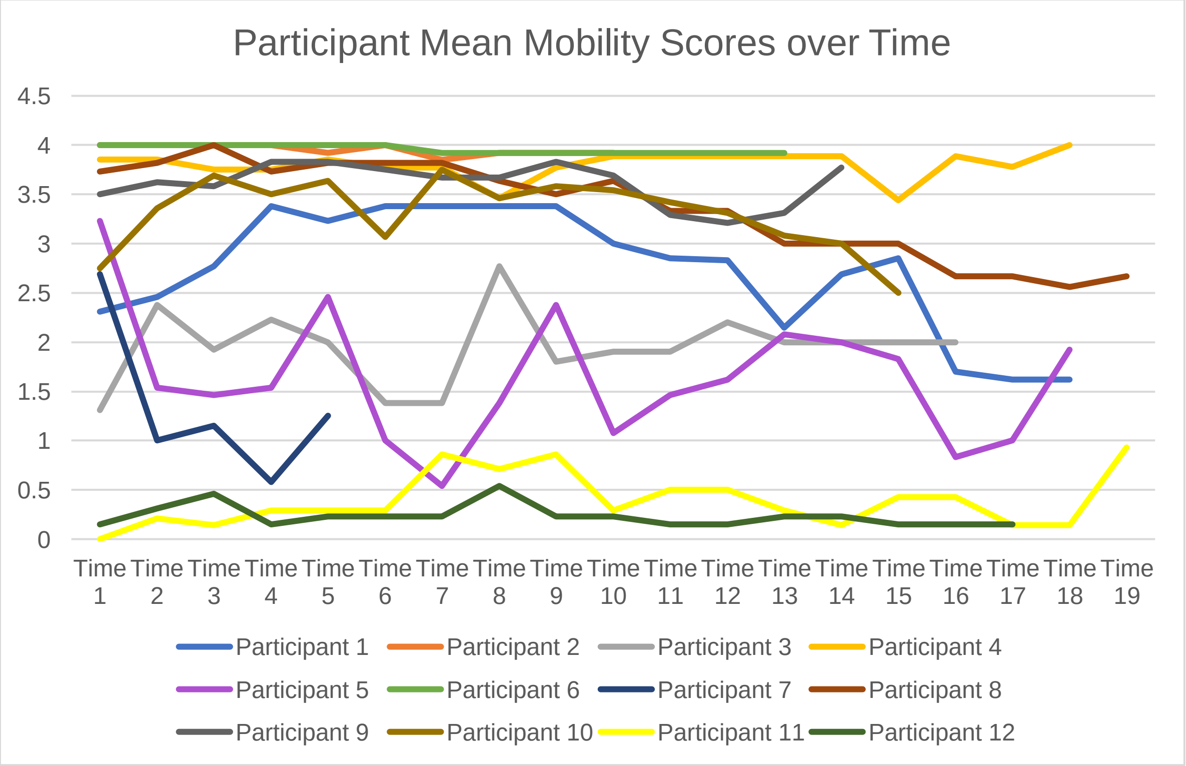
Mobility scores in this domain again showed a variance (range = 0.00-3.82) and as with other domains, fluctuating patterns were observed for participants over the 14 categories. From time points 1-9 however, the participants' data, although varied (range = 1.32-3.82), were relatively stable. At later time points (10-19) the data showed a more distinctive downward course of deterioration in mobility responses.

Although a short period of stasis was observed, the mean values here indicate overall deteriorating mobility responses.

Again, looking at three distinct time points 1, 10 and 19 in the mean mobility values indicated general deterioration across the participants' mobility responses. There were no subcategories in this domain, which consistently increased over time. Some subcategories showed increased endpoint scores when compared with baseline scores; 'Use of Weaker Limbs' and 'Hand over Hand or Facilitated Mvt Needed' (n = 2/14). The remaining subcategories showed deterioration across all data time points (n = 12/14). The subcategories showing small increases, namely use of weaker limbs and reduced hand over hand facilitated movement, perhaps indicated increased mobility in these areas. These findings, however, were inconsistent and contradicted deteriorating scores in the other subcategories, ('Active Movement', 'Core/ Trunk Stability', 'Head/ Neck Stability', 'Hand-Eye Coordination', 'Ability to Grip in RH', 'Ability to Grip in LH') suggesting that these latter data may more accurately reflect the deteriorative nature of Batten disease and the gradual loss of mobility over time.

Looking more closely at individual participant mobility scores, further analysis was conducted on mean scores for each participant. Figure 5.21 illustrates how the mean mobility scores changed for each participant over the three years.

Figure 5. 21 Mean mobility scores over 19 data collection points (three years) for each of the 12 participants



From the individual participant data, there were observed fluctuating patterns in the mean mobility scores, and a wide variance in the range of scores for participants (range = 0.00-4.00). Participants in some cases showed decreased scores from their endpoint scores in comparison to their baseline (n = 7/12), unchanged scores at their endpoint in comparison to their baseline to endpoint (n = 1/12) and increased scores at endpoint compared with their baseline (n = 4/12). Patterns indicate that participants' mobility was incredibly varied both in scores and trajectories over time. Although the overall mean scores across the 12 participants indicated a small period of stable mobility over time, the individual participant-level data demonstrated a complex and varied picture with overall deterioration. Individual participant data, for children with JNCL, also showed a similar picture. Figure 5.22 shows mean mobility scores for participants with JNCL diagnosis and how these change over time.

Figure 5. 22: Mean mobility CMTOM scores for participants with JNCL diagnosis

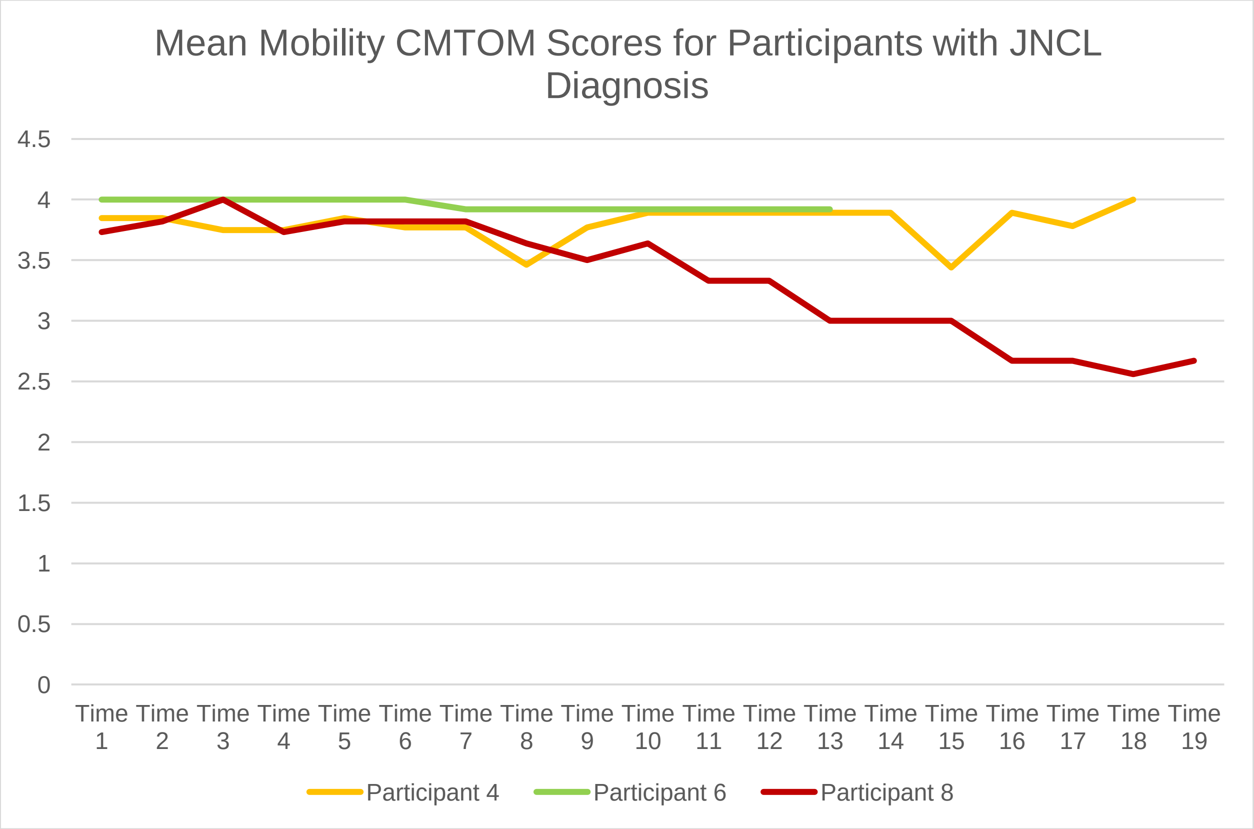


Figure 5.22 displays a plateau effect from time points 1-8 in the individual data for the JNCL subgroup. Of the three participants, one participant showed increased mean mobility scores from baseline to endpoint (participant four), one participant showed slight deterioration (participant six) and one participant showed marked deterioration over time (participant eight). There were some time points that showed lower mean scores (participant four, time points 8 and 16). Data indicate

the positive impact of music on movement for participants four and six, however the data here are variable and inconsistent between participants in this subgroup and therefore it is challenging to make deductions or interpretations about mobility in music therapy over time. Again, as with other overlapping domains between the two assessment measures, mobility scores showed differences. Hamburg scores for participants' mobility showed variable patterns of deteriorating or maintained mobility in the JNCL subgroup. In contrast, CMTOM mean mobility scores showed a general plateau effect with regards to JNCL subgroup, suggesting mobility skills could be maintained in music therapy over time. The different data between the two measures perhaps demonstrates the ability for the CMTOM measure to identify and detect consistent trajectories with regards to children's mobility, especially for the JNCL subgroup.

5.4.6. Summary

Generally, across the domains of communication, behaviour, wellbeing, cognition and mobility, mean scores for all participants showed a period within the therapy where stable patterns were observed. Results here indicated that in some circumstances, responses in music therapy could be stable and maintained for a period in the mid-stages of therapy input. Drilling down into individual participant data, however, highlighted the fluctuating and heterogeneous nature of the participant sample. The inconsistent results at individual participant-level demonstrate the complexity and challenge of generalising data across the different types of diagnosis, age and disease progression for children affected by Batten disease. The stable patterns in the JNCL sub-group, however, indicate maintained responses in music therapy in communication, cognition and wellbeing domains. The lower scores at time points 8 and 9 for participant eight, suggest the child had lower functional skills during these weeks than other weeks over the three years, perhaps due to external factors such as mood, fatigue or engagement (a concept explored further in Phases Two and Three). Preliminary evidence suggests a positive impact of music for this smaller population of affected children and further research in subsequent phases examine this response in more detail.

5.5. Overview

The two distinct perspectives utilised in Phase One (music therapy and clinical) have unearthed important differences in how affected children are perceived, observed, and monitored. Overall, findings indicate there are different levels of understanding as to a child's functional abilities according to the contrasting perspectives. It is important to note that the age range of participants across all three phases of the research (range = 3-17 years) contributed to the diversity of the group, and as such, developmental differences could have impacted functional responses or communication skills (for example the verbal skills of a three-year-old, would vastly differ from that of an 18-year-old). Nevertheless, there were seemingly marked differences in deterioration according to the two measures.

Specific responses such as singing abilities, non-verbal interactions, vocalisations and self-expression skills could be identified using the CMTOM measure in a music therapy context but excluded or missed from the Hamburg scale. The rate of decline was also distinctly different between the two perspectives. Specifically, CMTOM scores in some cases showed a period of stable responses during mid-stages of therapy, in comparison with downward trajectories in these domains on the Hamburg scale. This suggests that in the context of music therapy (and from the perspective of a music therapy-based observation measure), functional skills of cognition, mobility, wellbeing and behaviour could be maintained in mid-stages of therapy support and functional responses perhaps deteriorate to a lesser extent than is indicated in skills measured from a clinical perspective. Communication responses could also slightly increase or be maintained within a music therapy environment, in comparison to deteriorating communication from a clinical perspective. Findings therefore indicate the possibility that varying perspectives (music therapy versus clinical) give a different picture of a child’s functional responses and skills. The implications of this could potentially mean that measuring children on standard clinical scales is more likely to give professionals a surface level and rudimentary assessment of how the child is deteriorating throughout their condition. In contrast, tracking and monitoring a child within a music therapy context could potentially provide a more comprehensive assessment of a child’s functional presentation (further addressed in the discussion chapter).

By extracting data relating to participants with JNCL, patterns in the majority of the CMTOM scores showed maintained responses in music therapy over time, particularly in the domains of communication, cognition and wellbeing. Preliminary evidence suggests that weekly music therapy could help maintain language skills, memory, attention, participatory skills and general emotional wellbeing. However, when exploring these responses from a clinical perspective, patterns were more variable, where some participants showed periods of deterioration and others showed maintained skills according to their mean Hamburg scores. Distinct differences between the two measures further demonstrate the possibility that different measures present a different perspective of a child’s functional skills. Within a music therapy context and assessment framework, affected children demonstrate a more consistent picture of maintenance in comparison to the clinical perspective.

Positive trajectories in the communication responses particularly highlight the need to expand this line of enquiry in subsequent research phases. In contrast to the other subcategories in the CMTOM, overall mean communication scores, individual participant-level data and JNCL mean scores showed stable or slightly increased responses over time. Of the 14 subcategories within the communication domain, 78% of all areas showed increased or maintained trajectories over time (n = 11/14), and 75% of participants similarly showed maintained or improved communication responses over time (n = 9/12). Initial evidence suggests that music therapy may have a positive influence on communication responses, for children with Batten disease. This demonstrates the need for further exploration into the context of music and communication, to further examine this response in other contexts and from alternative perspectives.

5.5.1. Learning Points

In response to the research questions, findings from Phase One are summarised in the following learning points:

1.1. For children with Batten disease, how can functional outcomes be tracked and measured in music therapy?

- Observed functional outcomes of children affected by Batten disease in music therapy relate to communication, mobility, wellbeing, cognition and behaviour.
- The CMTOM measure offers a reliable and valid measure for use in subsequent phases of research with further research needed to evidence this further.

1.2. From a clinical perspective how do functional outcomes (social, physical and cognitive) change over time for children with Juvenile Batten disease?

- Functional skills according to a clinical rating scale show marked deterioration for the whole participant group in all three areas of communication, cognition and mobility.
- Functional skills according in the JNCL subgroup showed variance and inconsistencies.
- Music Therapy assessment measures could offer a deeper understanding of a child’s functional responses in comparison to standard clinical tests.
- The divergent nature of Batten disease demonstrates a need for further research to explore individual responses to music and music therapy.

1.3. How do functional outcomes (social, physical and cognitive) change in music therapy over time for children with Juvenile Batten disease?

- Long-term music therapy interventions could facilitate maintained functional skills for a period, for children affected by Batten disease.

- For children with a diagnosis of Juvenile Batten disease variant of Batten disease, communication, wellbeing and cognitive responses in music therapy could be maintained over time.
- Communication responses in music therapy could be maintained or in some cases improved over time.

In pursuit of the second research aim, Phase Two of the research aims to explore whether such positive responses in communication for children with JNCL can be observed in an educational environment. Based on the periods where communication skills were maintained and improved for children in music therapy, the next phase of the research aimed to examine whether these responses can also be seen in response to a daily music-based education program.

Chapter 6: Results from Phase Two Music Speaks Program Trial

“Music expresses that which cannot be put into words and that which cannot remain silent”
— Victor Hugo

6.1. Introduction

Further to Phase One, Phase Two aimed to ascertain the extent to which these responses from music therapy could be transferred to educational environments, through a music education program, Music Speaks. This phase of the doctoral research is significant as it has relevance to its potential ‘real-world’ application in ongoing care and education for affected children. If the unique responses to music and music therapy seen in Phase One can be transferred to educational environments beyond music therapy, then this could mean the practical application of lower-cost support programs for affected children.

This chapter sets out the data that was collected from the delivery of the Music Speaks music program in five schools in the UK, and the functional communicative responses from children affected by JNCL, who trialled the program. The program was designed in line with the guiding principles of NMT (Thaut, 2014) and with feedback from several professionals (Neurologic Music Therapy trainers, Neurological Speech and Language Therapists and Neurologic Music Therapists). The intervention was developed following the process outlined in section 4.2.2.

A summary of how each research question has been approached is set out below:

2.1. Can the Music Speaks program be delivered effectively in a formal education framework?

Data was retrieved from the Music Speaks recording sheets, which captured the date and time of delivery, the professional’s details, program task achievement, and other supplementary information. The subsequent analysis here relates to the number of times and when the program was delivered. Professionals’ comments provide contextual information relating to the participant’s responses to the program which included themes of engagement, fatigue, staff prompting and program adaptation.

2.2. How do communication outcomes for children with Juvenile Batten disease change in response to the Music Speaks program?

Quantitative data from videos of participants taking part in the program were analysed, focusing on mean CMTOM communication subscale scores tracking communicative responses for children over the six-week trial period. Average mean scores in the participant pool, as well as individual-level participant data were generated, drawing upon supplementary information provided by professionals delivering the program.

6.2. Participants

Five children affected by JNCL were recruited for this phase of the research, and all participants engaged with at least one Music Speaks program over six weeks. Table 6.1 outlines the participant records from Phase Two data collection which includes the number of videos submitted for the research and the number of completed programs on record.

Table 6. 1: Participants, data record sheets and video submissions from the Music Speaks program trial

Participant	Setting	Number of completed program sessions	Number of videos submitted	Recording sheets retrieved
1	Mainstream Primary School Setting	6	6	No
2	Mainstream Primary School Setting	2	2	No
3	Mainstream Primary School Setting	26	6	Yes
4	Special Education College Setting	1	1	No
5	Special Education College Setting	26	8	Yes

The ages of participants ranged from 9-18 years, with three participants attending mainstream schools, and two participants attending a specialist college setting. All children had a diagnosis of JNCL. As can be seen from the table, there were three missing data recording sheets for participants in the research project. Due to the onset of the COVID pandemic in March 2020, data collection for all research was immediately suspended. School closures across the UK, meant that all schools stopped delivering the program in March 2020, and this meant that participants had either just commenced the program delivery or had only completed a few weeks (ranging from one week to six weeks). A COVID-19 data retrieval protocol was drawn up (see Appendix 10) however, despite this preparation, many schools remained closed to non-keyworker children. From March to September 2020, three professionals left the schools and in other cases, professionals who worked at the school (but did not participate), could not locate the recording sheets. Data here relating to the program delivery therefore only includes the recording sheets which were retrieved from just two of the five participants in the research project. Video data were received from all participants (although in varying frequencies), and the five participants are described in more detail below.

Participant One

Participant one attended a mainstream primary school in the UK. They were diagnosed with JNCL several years before joining the research program. From meeting with the child before the program started, it was noted that their speech was seemingly unimpaired but was beginning to speed up. They were registered as visually impaired, and presented as focused, alert, attentive and able to hold a conversation. The child was referred to the research program as they were connected to an existing music scheme in the UK aimed at supporting children with additional needs and music therapy. Batten disease symptoms were in the early stages of onset for this individual.

Participant Two

Participant two attended a mainstream school in the UK when they participated in the research program trial. They had received a diagnosis of JNCL a year before participating in the research. From meeting with the child beforehand, it was noted that their speech intelligibility was good but was beginning to speed up. They were registered as visually impaired, and presented as focused, alert, attentive and able to hold a conversation. The child was referred to the program by the school. The child mentioned no particular interest in music or singing. Batten disease symptoms were in the early stages for this individual.

Participant Three

Participant three attended a mainstream primary school in the UK. The family received their diagnosis of JNCL whilst the child was attending primary school a few years before participating in the research project. From meeting with the child, it was noted that their general speech was intelligible, their focus was good, and the child was alert and present, and able to converse. This child was referred to the research program as they had an interest in music and dance. Batten disease symptoms were in the early onset stages for this individual.

Participant Four

Participant four attended a specialist college for young people with visual impairment needs. This individual participated in Phase One of the research project which began in 2016. When they joined the research at this time, they had a visual impairment, were cognitively focused, alert, and able to hold a conversation. Speech intelligibility was generally a challenge, and their speech was sped up and mumbled on occasion. They were referred to the research as they enjoyed song writing and playing music regularly. Over the five years, their speech intelligibility had declined slightly but mobility and cognitive functioning remained good. Batten disease symptoms were moderate for this individual at the time of the program trial.

Participant Five

Participant five attended a specialist college setting for young people with additional needs. The family received the diagnosis of JNCL when the child was in a mainstream primary school setting. At the time of joining Phase One of the research in 2016, the individual had good mobility and intelligible speech. They were referred to the research as they enjoyed singing and listening to music daily. They had regressed since Phase One, and during the program trial the individual was primarily fully dependent on care (using a wheelchair full-time, receiving peg feeds, had supported movement, and relied on communication aids), and responded with single words. Batten disease symptoms were in the progressed stages for this individual.

6.3. Research Question 2.1: Can the Music Speaks program be delivered effectively in a formal educational framework?

6.3.1. Delivery

With the consideration of the February half-term break and the reduced trial period, there were therefore a total of 29 possible school days within which the program could have been delivered. As can be seen from Table 6.1 only two participant recording sheets were obtained after the trial time, and program delivery ranged from one day to 26 days. It is worth reiterating that with the absent data recording sheets from participants one, two and four, analysis of program delivery was based on the number of videos submitted. It was assumed that a video submission meant that the program was delivered by professionals on the day the recording was made, but there may have been more frequent unrecorded programs delivered for these participants.

Taking the two participants with completed program records, the mean program delivery was 26 times out of a possible 29 program delivery days. These two participating schools, therefore, delivered and completed on average roughly 90% of the Music Speaks program with the participating children. Record sheets also captured the time of day that the program sessions were delivered in school. For participant three, the program was delivered at roughly the same time every day, ranging from 1.05pm to 1.25pm (average time of day was 1.12pm). For participant five, sessions were delivered in both the afternoon and the morning, ranging from 9.05am to 2.25pm, but most sessions were delivered in the morning (15 out of the 20 session times recorded).

Given the small data pool from which this data was drawn (n = 2), it is extremely challenging to draw any meaningful or generalisable observations about the Music Speaks program delivery. However, observing results from the two complete participant data sets suggests promising uptake and delivery of the program given that both participating schools were able to deliver roughly 90% of the full program with their respective children taking part. Program delivery times suggest that schools delivered the program at around the same time of day as one another.

6.3.2. Completion of Program Tasks

Within the Music Speaks program, there were four key tasks for the child to listen to and complete (see Appendix 9 for the full Music Speaks program). Tasks were as follows:

1. Repeat after me – Repeating a phrase
2. Sing along to a song
3. Repeat after me – Repeating a phrase along to a metronome
4. Repeat after me – Repeating a phrase

Within each of these tasks (except for task two) were three opportunities given on the instructions to complete the task. Recording sheets gave tick boxes for professionals to complete when they observed or heard the child completing each task. For analysis, if one attempt was completed for each task this was recorded as a task complete. During training sessions, professionals were told that the three allowances for task completion were not obligatory but rather opportunities to give the child more chances to complete the task. In some cases, for instance, once the child had completed the task the first time they moved on to the next speaking activity or song. From the data recording sheets, task completion was scored out of a possible total score of 4 for each program (see Appendix 20 for task completion scores for participants three and five).

For participant three, task completion was recorded in all the instances where the program was delivered, and there were no missing data. Of a total score of 104 tasks (based on 26 instances of program delivery) this child completed 72 tasks (or 69% of the total tasks) delivered in the program. Task completion varied over the six weeks but in all program sessions, the child was able to complete at least one task. For participant five, task completion was recorded in all but one program session (the first session on 31/1/20 was missing). Task completion again varied in the program trial and there were several weeks where the child was unable to complete any tasks.

As can be seen from Table 6.2, the mean task completion score for participant three was 2.8 (SD = 0.9) and an average of 69% of the total program tasks were completed (based on 26 recorded instances of program delivery). The mean task completion score for participant three was 1.7 (SD = 1.5) and an average 42% of tasks were completed (based on 25 recorded instances of program delivery).

Table 6. 2: Music Speaks task completion mean and SD scores for participants 3 and 5

	Participant Three	Participant Five
	Tasks Completed	Tasks Completed
Number	72/ 104	42/ 100
% Of Total Tasks Completed	69.2	41
$\bar{x}$ Completion Score	2.8/ 4	1.7/ 4
SD	0.9	1.5

6.3.3. Summary

In summary, the data presented here showed variable results. For the five participants who were recruited for this phase of the project, all schools were able to deliver at least one program for the participating children. However, due to the outbreak of the pandemic in 2020 and the absence of recording sheets, it was difficult to ascertain what the program delivery looked like for the other three participants. Based on the recording sheets collected from the remaining two participants, data indicated that program delivery was largely achievable with around 90% of the total program being delivered for both children. According to the professionals who completed the data sheets, in answer to the research question, “*Can the Music Speaks program be delivered effectively in a formal educational framework*?” the answer is provisionally ‘yes’ for the two individuals whose recording sheets were retrieved. Findings here, indicate that for both participants, the Music Speaks program was able to be delivered in school consistently at approximately the same time of day for a period of roughly six school weeks.

However, the data depicts a variable picture with regards to task completion scores. For both participants, this ranged from 42 to 72 tasks completed in the trial. The variation could have been due to the different presentation and age of each child. Participant three was in the early stages of their Batten disease journey and was largely independent, had good communication skills and good cognitive ability, whereas participant five was showing more progressed symptoms of the disease, and was dependent on care staff for most of their functional activities. Participant five had speech still intact, but they were largely unintelligible, and speech was marked by repetitive communication. The difference in the disease stages could have had an impact therefore on the tasks they were able to complete in the program. Data indicate that for children whose language and speech were still intact (as with participant three), the program tasks were perhaps more achievable than a child whose language has more significantly deteriorated (as with participant five).

6.4. Research Question 2.2: How do communication outcomes for children with Juvenile Batten disease change in response to the Music Speaks program?

This section of the thesis outlines the quantitative data relating to communication responses to the Music Speaks program and changes recorded during the six-week trial period. Using the developed CMTOM communication subscale (see Appendix 13b), video data were assessed and analysed by the researcher. Mean CMTOM subscale scores are presented to outline changes in communication outcomes, as well as professionals’ comments from the supplementary information on recording sheets.

6.4.1. Communication Responses to Music Speaks Program

To answer the research question relating to potential changes in communication responses, analysis of the data focused on descriptive statistics, to identify how communication responses to the program changed over the trial. The use of descriptive statistics was chosen over other statistical analysis measures due to the small case study sample size, the heterogeneity of the population, and wide range of ages in the participant pool (range 9-18 years of age), which would have made inferential analysis challenging. A total of 23 program session videos were assessed, and as can be seen in Table 6.1 video data ranged from one to eight submissions over the six-week period. Participants began using the program on different dates and therefore the submitted videos were organised by date into the six trial weeks. The data represent a perspective of individual session-by-session data, rather than an average cross-sectional mean score for each week.

Video submissions were analysed using the CMTOM communication subscale assessments. Scores were recorded for each of the 17 items on the CMTOM Communication subscale. Videos were rated on a scale of 0-4 (responses observed 0=Never, 1=Rarely, 2=Sometimes, 3=Often, 4=Consistently), and video assessments were recorded on the scale after task one (baseline) and after task four (endpoint). Three items were reversed scored (‘Stutters and Stammers’, ‘Repetition’, ‘Dysarthria or Mumbling’). There was a total of 68 data collection points across the whole participant pool.

Expressive Communication

Mean scores from video data were split into items relating to expressive communication and receptive communication. Expressive communication related to the subscale items of Stutters or Stammers, Repetition, Dysarthria/ Mumbling, Correct Use of Words, Breath Support, Pace/ Rhythm of Speech, Speech Clarity, Pitch, Volume, Articulation, Intonation, Stress, Presence of Words and Presence of Phrases. Expressive communication baseline mean and standard deviation values are represented in Table 6.3, for all participants across the trial period.

Table 6. 3: Expressive communication baseline and endpoint mean scores and SD values

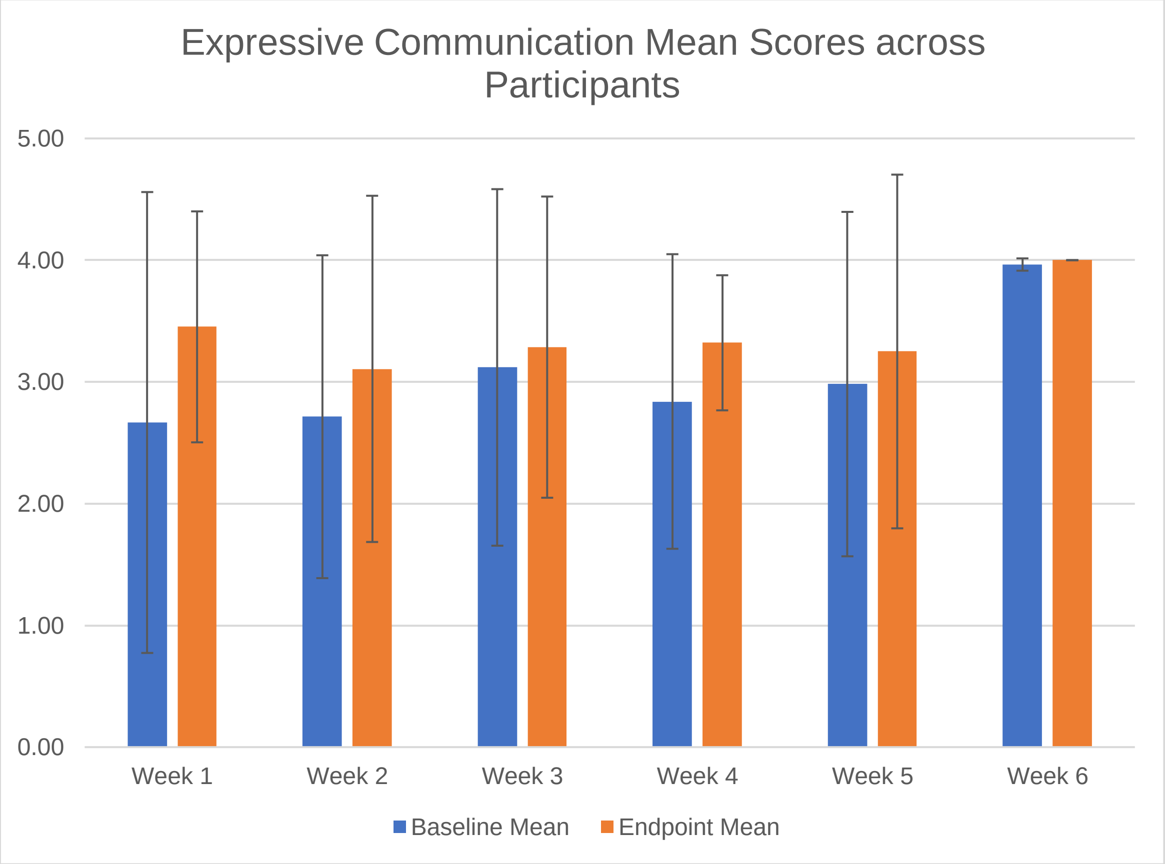
	Expressive communication across all participants					
	Week 1	Week 2	Week 3	Week 4	Week 5	Week 6
Baseline $\bar{x}$	2.67	2.71	3.12	2.84	2.98	3.96
Baseline SD	1.89	1.33	1.46	1.21	1.41	0.05
Endpoint $\bar{x}$	3.45	3.11	3.29	3.32	3.25	4.00
Endpoint SD	0.95	1.42	1.24	0.55	1.45	0.00
Change Score	0.78	0.40	0.17	0.48	0.27	0.04

As can be seen in Table 6.3, baseline and endpoint mean scores were variable over the six-week period (range = 2.67-4.00), with no constant observed change in the program trial. Baseline mean scores remained variable and there was no constant pattern of change (range = 2.67-3.96). Exploring endpoint scores illustrated a more stable pattern with less variance over the six-week period (range = 3.11-4.00). Overall, mean values for baseline and endpoint recordings over the trial remained relatively stable. It could have been expected that over time, children became more familiar with the program content producing higher scores in their expressive communication session-by-session. This appeared not to be the case, however, and there was no indication of expressive communication scores improving over the six weeks. This indicates that there was no longer-term change or improvement in expressive communication with repetition or familiarisation of the program, and responses were variable over time.

However, the mean change within each session of the program trial showed a more consistent pattern. Session by session, mean expressive communication scores increased from the baseline. Mean baseline scores increased on average by 0.36 points ranging from 0.04 to 0.78 across the trial six weeks (see appendix 21 for mean and standard deviation expressive communication scores for all participants). The increases in individual session baseline mean scores could indicate a short-term positive change in expressive communication in response to the Music Speaks program.

Figure 6.4 visually represents this data for mean baseline and endpoint expressive communication scores over six weeks for all participants. Depicting an undulated pattern of expressive communication scores, the graph illustrates inconsistencies across communication responses over six weeks for the five participants. It can also be seen that mean expressive scores increase each session from baseline to endpoint suggesting short-term change in response to each Music Speaks program.

Figure 6. 4: Participants mean scores and standard deviations for baseline and endpoint expressive communication across time



Receptive Communication

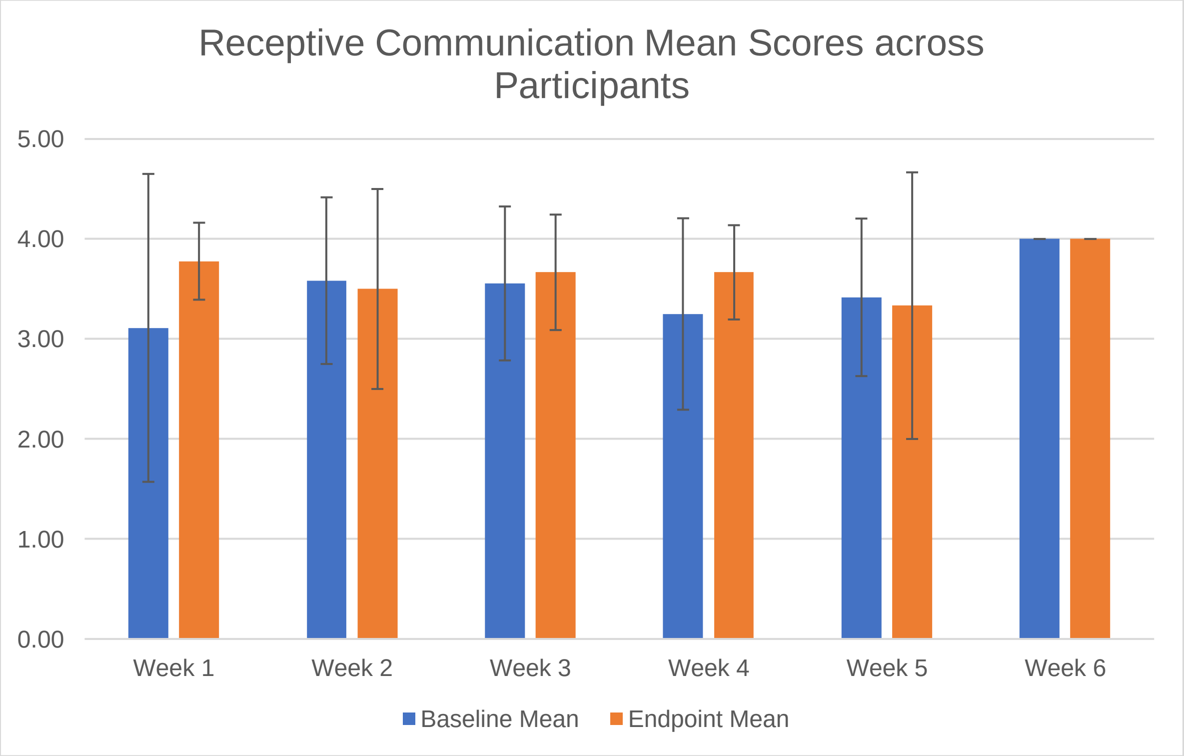
Analysis of the video data drew upon descriptive statistics from the receptive communication subdomain. Receptive communication related in this case to the three subscale items Word Recall, Responds to Familiar Voice, and Ability to Verbally Respond and Participate. The baseline means and standard deviation values in Table 6.5 are taken from assessments after task one (baseline) and after task four (endpoint).

Table 6. 5: Receptive communication baseline and endpoint mean scores and SD values

	Receptive Communication across all participants					
	Week 1	Week 2	Week 3	Week 4	Week 5	Week 6
Baseline $\bar{x}$	3.11	3.58	3.56	3.25	3.42	4.00
Baseline SD	1.54	0.83	0.77	0.96	0.79	0.00
Endpoint $\bar{x}$	3.78	3.50	3.67	3.67	3.33	4.00
Endpoint SD	0.38	1.00	0.58	0.47	1.33	0.00
Change Score	0.67	-0.08	0.11	0.42	-0.08	0.00

As can be seen from Table 6.5, receptive communication responses to the program were less varied (range = 3.11-4.00). As with expressive communication, there were no constant patterns of improvement in the data over time and showed more stable patterns in baseline and endpoints means (see appendix 22 for mean and standard deviation receptive communication scores for all participants). Individual sessional mean scores showed that three baseline session mean scores increased, one was unchanged and two decreased, suggesting there was no pattern of short-term improvements in receptive communication across the sample. In comparison to expressive communication, however, generally mean scores were higher. Figure 6.6 visually shows mean pre and post receptive communication scores over six weeks in the three subcategories.

Figure 6. 6: Participants means scores and standard deviations for baseline and endpoint receptive communication across time



As can be seen in Figure 6.6, the mean scores were generally higher than those recorded in the expressive communication domain. Results here suggest that receptive responses to the program showed no consistent change from baseline to endpoint or consistent overall communication changes over the six weeks.

6.4.2. Individual Participant Data Analysis

Considering the unique presentation of the children affected by Batten disease and the heterogeneity of participant data from Phase One, it was deemed appropriate to analyse the individual communication responses in the program trial. The following section presents the analysis of mean scores and standard deviation values for expressive and receptive communication by the individual participants. Each section also describes the analysis of scores from specific items from the communication subscale such as word recall, speech pacing, phrase completion and speech clarity, from baseline to endpoint. Visual Figures have only been included for those participants with notable score changes. These data were extracted to examine whether elements of the Music Speaks program specifically targeting these aspects of communication had an impact. Lastly, recording sheet comments were analysed here to help build a narrative of how each participant responded to the program daily.

Participant One

All six videos were submitted of the program sessions for participant one, which were delivered by two learning support assistants in the school. Although there were no recording data sheets retrieved for this participant, time stamps on the video indicate that sessions were delivered between 10.00am and 2.00pm with most sessions happening around 11.30am.

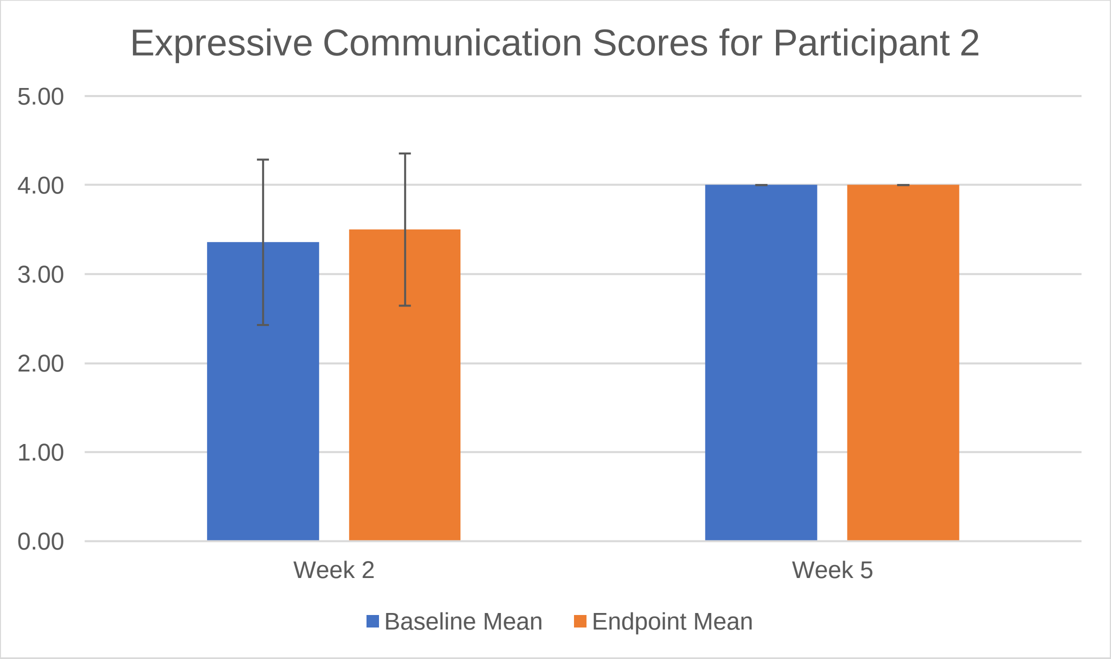
Expressive communication responses for participant one showed a static pattern. No change was recorded in their communication responses, and there was no variation in these scores over time (SD = 0 in all cases). Data here represents a high level of communication from this participant which remained at the highest mean score of 4.00 throughout the program trial. This suggests a possible ceiling effect, perhaps suggesting the need for a more sensitive assessment measure.

Receptive communication scores similarly remained unchanged and were at the highest mean value of 4.00 throughout (SD = 0). There was no change from the baseline to endpoint data collection points and no change was recorded over the six-week program. Investigating elements of expressive communication (such as ‘Stutters or Stammers’, ‘Pacing’, ‘Speech Clarity’ and ‘Dysarthria or Mumbling’, ‘Words’ and ‘Phrases’) also showed a static picture with mean scores remaining at the highest score of 4.00 throughout the program trial (SD = 0). Similarly, for the receptive communication items on the subscale such as ‘Word Recall’, ‘Responds to a Familiar Voice, and ‘Ability to Interact, Respond and Participate’, there were no patterns of change. All scores remained at the highest score of 4.00 throughout the program trial (SD = 0) and remained consistent throughout. At the time of the program trial, speech for this participant was unimpaired but could be a little fast at times. Data here perhaps indicates that the Music Speaks program had little effect on children who have little or no existing speech impairments, or a ceiling effect occurred whereby a more sensitive measure could have identified finer detailed changes to communication.

Participant Two

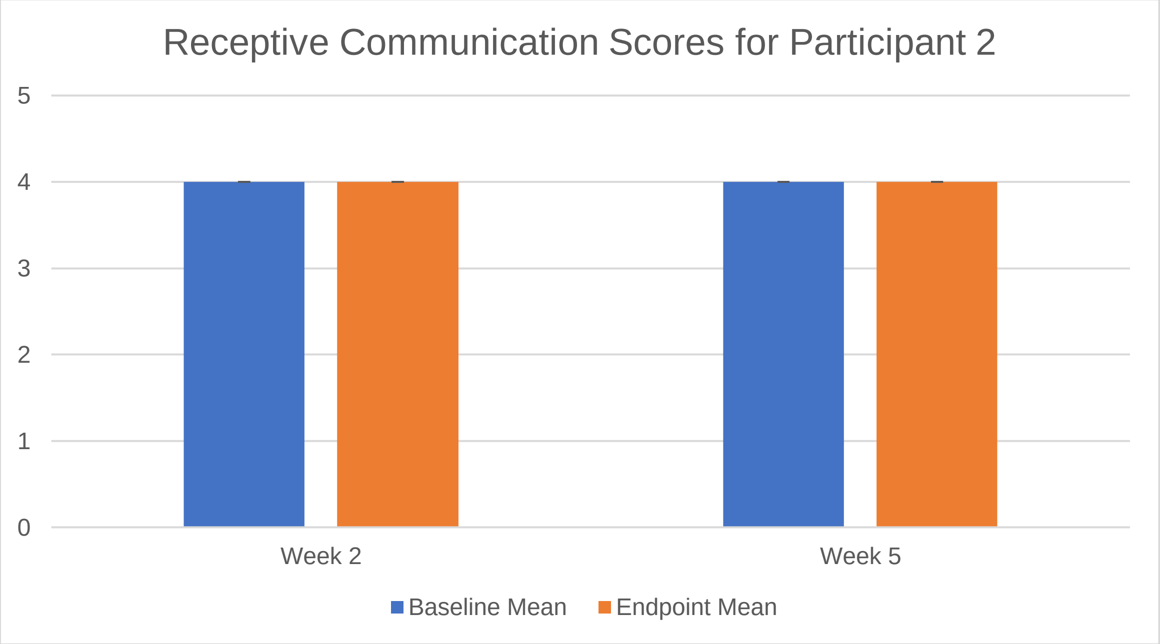
There were two videos submitted for participant two during weeks two and five of the program trial. Program sessions were delivered by three learning support assistants in their mainstream primary school. Although there were no recording data sheets retrieved for this participant, time stamps on the video indicate that sessions were delivered at 9.30am and 10.00am. Figure 6.7 illustrates mean and standard deviation values for expressive communication responses from participant two.

Figure 6. 7: Expressive communication means and standard deviation scores for participant two



As can be seen in Figure 6.7 there were variable scores recorded in week two (baseline SD = 0.93, endpoint SD = 0.85) compared to week five (SD = 0 at both baseline and endpoint) and overall mean expressive communication scores increase from week two to week five. The mean baseline score in week two increased slightly by 0.14, in comparison to week five where mean scores remained unchanged throughout ($\overline{x}$ = 4.00, SD = 0). Patterns here could suggest a slight improvement in communication over time, however, this is not definitive based on just two assessed sessions.

Figure 6. 8: Receptive communication means and standard deviation scores for participant two



As can be seen from Figure 6.8, receptive communication scores remained unchanged over the two recorded sessions remaining at the highest mean score of 4.00 throughout (SD = 0). There was no change from baseline to endpoint data collection points and no change recorded over the six-week program. Receptive communication scores were generally higher for this participant than expressive communication.

Investigating elements of expressive communication (such as ‘Stutters or Stammers’, ‘Pacing’, ‘Speech Clarity’, ‘Dysarthria or Mumbling’, ‘Words’ and ‘Phrases’) showed a similar picture. Individual item scores for elements of expressive communication showed a static picture, with mean scores remaining unchanged from baseline to endpoint in most areas (except for improved Stutters and Stammers in week two). There seemed to be an overall improvement whereby most scores in sessions increased by 2 points from week two to week five. Data could suggest the Music Speaks program may have improved elements of expressive speech from the beginning to the end of the program trial, however, this is not a definitive finding based on just two assessed sessions.

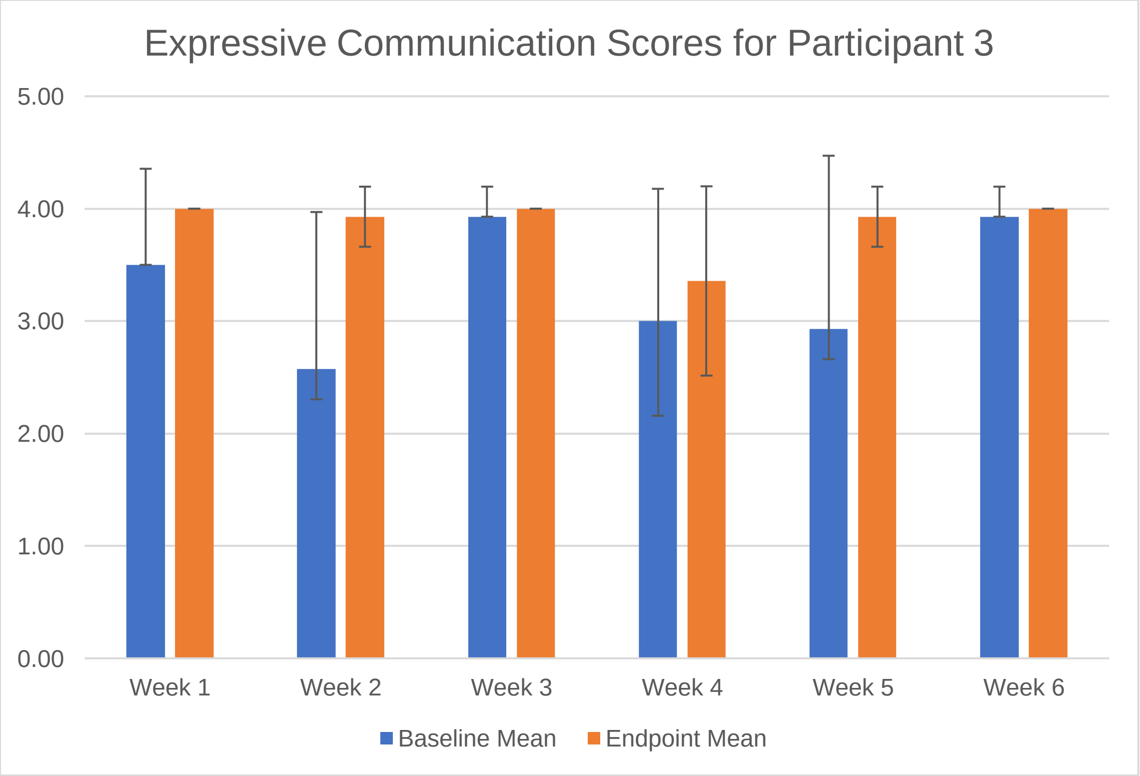
In looking into the receptive subscale items relating to ‘Word Recall’, ‘Responds to a Familiar Voice, and ‘Ability to Interact, Respond and Participate’ there were no patterns of change. All scores remained at the highest level of 4 throughout the program trial and remained static throughout. Data shows that receptive communication scores were generally higher from the beginning of the six weeks in comparison to expressive communication scores which increased over time.

At the time of the program trial, speech for this participant was again largely unimpaired but could be a little fast at times. Data here indicates that the Music Speaks program may have had a very small effect. Findings, however, are limited by the small data set.

Participant Three

All six videos were submitted of the program sessions for participant three, which were delivered by two visual impairment specialists in the school. Recording data sheets from this participant, indicate that the program sessions were delivered at roughly the same time every day, ranging from 1.05pm to 1.25pm (average time of day was 1.12pm). Figure 6.9 illustrates the mean scores and standard deviation scores in expressive communication responses for participant three.

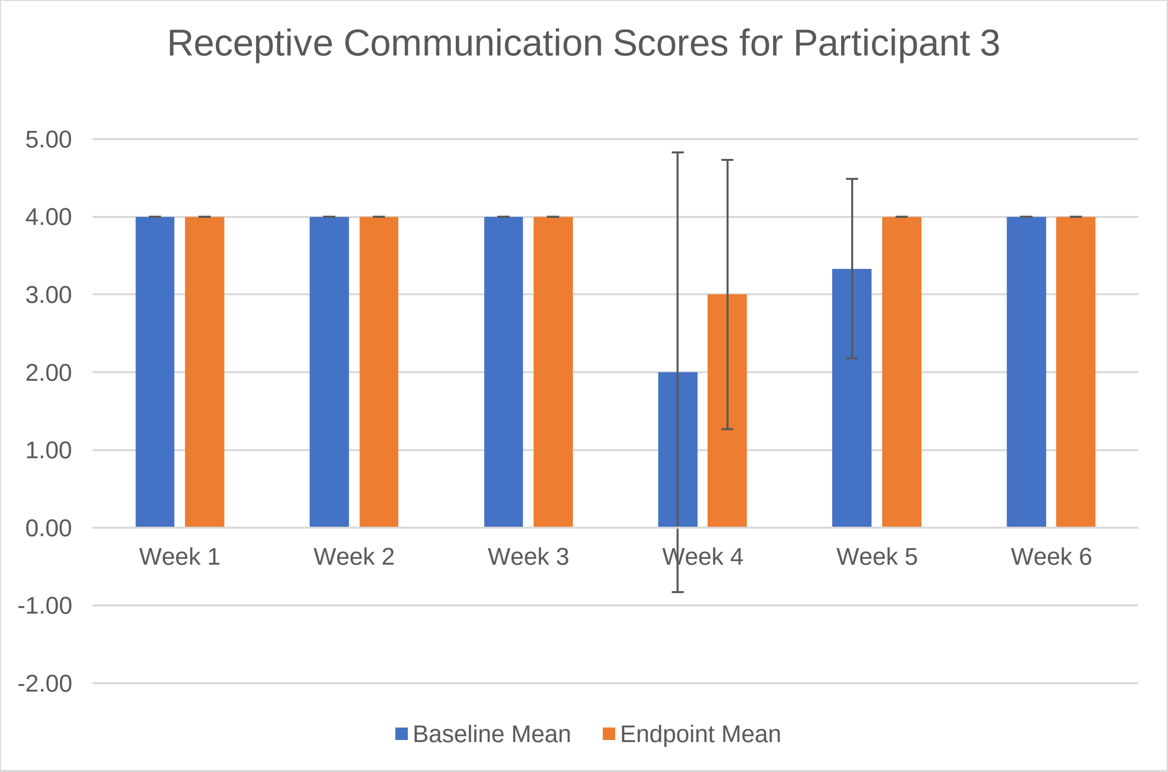
Figure 6. 9: Expressive communication mean and standard deviation scores for participant three



As can be seen from Figure 6.9, mean scores for expressive communication were variable from baseline and endpoint over six weeks (range = 2.57-4.00). The undulated patterns show inconsistent changes in communication responses. However, there is a more consistent pattern observed in the sessional data where, mean expressive communication scores increased from baseline to the endpoint (average 0.56 point increase, range = 0.07 to 1.36 across the trial six weeks). The increased weekly baseline means scores indicate a short-term positive change in expressive communication, which could be in response to the Music Speaks program.

Exploring receptive communication responses for participant three indicated a slightly steadier pattern. As can be seen from Figure 6.10, baseline and endpoint mean scores across the six weeks were more consistent, and weeks one, two, three and six were all static and remained unchanged from baseline to endpoint ($\overline{x}$ = 4.00, SD = 0). Weeks 4 and 5 showed more variable communication responses with mean scores ranging from 2.00-4.00. Again, within weeks four and five, baseline scores increased by 1.00 and 0.67 values for each week. Data here also show that receptive communication scores were generally higher over the six weeks in comparison to expressive communication scores.

Figure 6. 10: Receptive communication means scores and standard deviation values for participant three



Delving deeper into individual subscale item scores indicates a similar picture of variance. The expressive sub-items (‘Stutters and Stammers’, ‘Speech Clarity’, ‘Pacing’, ‘Improvement to Dysarthria’, ‘Words and Phrases’) either remained static or improved (except for week four) after each program session. Data from the recording sheets described how the child’s engagement was particularly impacted by fatigue throughout week 4 (sessions 15-19), summarised subsequently in the discussion of the supplementary information for this participant. Data echo the inconsistent patterns seen in the mean scores, but scores session by session showed greater instances of maintenance or improvement.

Similarly, receptive communication subscale items that included ‘Word Recall’, ‘Responds to a Familiar Voice, and ‘Ability to Interact, Respond and Participate’ echoed the mean data for this participant. There was little change recorded both session by session and in the program trial, with most responses receiving the highest possible score of 4. As with expressive communication scores, during weeks four and five, levels of participation were lower than other weeks, which may have been due to external factors (later discussed in the following subsection). Despite this, some scores still improved after each program session.

As there were data recording sheets for this participant, it was possible to analyse the comments recorded week by week. Comments related to the child’s engagement, the need for prompting, fatigue, and program adaptation.

Engagement

Generally, half of the comments on the recording sheets related to variable engagement whereby the child either did or did not engage with the program. There were four instances where professionals commented on how the child engaged by singing along to the program songs or completing the verbal tasks. Comments such as “Joined in parts of the song” (participant three, session 7) and “…did click fingers to the beat” (participant three, session 12), indicated that the child was sometimes engaged in the musical elements of the program and able to join in. Comments relating to positive engagement were also interspersed with comments relating to the child’s lack of engagement during the song section. Professionals particularly commented that the child “did not want to sing today” (participant three, session 6), and “…did not want to do the music session this morning (participant three, session 18) and “…didn’t want to sing, chatted throughout” (participant three, session 19). Comments about the child chatting throughout the song, perhaps meant that the child was not engaged and did not want to participate in singing. Despite a lack of engagement depicted in the comments here, score data showed the child was able to complete half of the verbal tasks in these sessions. According to comments, engagement with the songs was variable, however, even when the child did not participate in singing, score data demonstrate that the child was still able to complete some spoken tasks. This could suggest that the songs were not appropriate or preferential for this participant or that they preferred to engage with the verbal tasks. Comments relating to the child’s engagement were a common occurrence throughout the program trial suggesting that engagement and attention levels were variable throughout the program length.

Prompting

Recording sheet comments were related to the need to prompt the child to complete the tasks in the program. For example, on day four of the program trial, one professional commented that the child "…joined in with song intermittently, just needed some prompting for number 4, repeated once" (participant three, session 4) and that "Task 1: with prompts, did not want to sing today" (participant three, session 3). Comments here described how the child was able to complete the verbal tasks if staff prompted them. As this was the first week of the program trial, it was not expected that children would have learned the program material yet, and therefore it was highly likely that they would need to be prompted. According to the recording sheet data however, score data from these weeks indicated that all verbal tasks were completed accurately. Data here indicate that with additional prompting the child was able to complete the tasks.

Fatigue

Comments from the trial also mentioned how fatigue impacted the child's engagement levels mentioning one day that they "Did not want to sing, very tired due to storm last night" (participant three, session 6). Implying the child was lacking sleep, there was evidence to suggest that tiredness led to the child's aversion to singing. During week four of the program, comments again related to fatigue where one professional commented that the child was "Tired today!" (participant three, session 15). Score data from both sessions demonstrate that task completion was lower in these sessions suggesting that fatigue and tiredness impacted the child's ability to engage and complete the program tasks.

Program Adaptation

In later weeks of the program trial, comments related to adaptations that the child made to the program material. For example, one day a professional noted how the child "…added their own new sentence 'I want to see my cat' + original sentence" (participant three, session 13). The evidence here suggests that the child was able to expand upon the program material and create new program content adding variations of phrases for interest and perhaps humour. Similarly, on the following day, a professional commented how the child; "2: Joined in with song, 3: Changed part of the phrase today ~ Distracted as someone came to the door, 4: Did not want to participate today" (participant three, session 14). Suggesting variable engagement within the session, comments here suggest that the child created adaptations perhaps for interest if they had become too familiar with the program content. Score data for these sessions suggest that although their engagement was variable, the child was still able to complete most of the program tasks. There is evidence here to perhaps suggest that as the child became familiar or bored with the program content, they began making adaptations to the program to add variation and perhaps humour.

Data from the recording sheets for this participant, describe how the child engaged variably with the program over time. In some cases, engagement with verbal tasks was good, whereas singing was less frequent. In the early stages of the program trial, comments suggest that the child could complete tasks if the professionals added further prompts. Later in the program, comments suggest that the child became comfortable and confident making adaptations to the program.

Participant Four

There were no recording sheets submitted for this participant and only one video submission, therefore data are limited. The program session was delivered by a learning support assistant at 10.00am.

Expressive communication mean scores were lower than receptive communication means scores. There was also more variance in the expressive communication mean scores (SD = 1.48 at baseline, SD = 1.49 at endpoint) compared to the high mean scores of 4.00 for receptive communication at baseline and endpoint (SD = 0). As there was only one session, data only illustrate change within the session from baseline to endpoint. There were marginal changes to expressive communication mean scores (an increase of 0.07) but no recorded change to receptive communication. Although the video was submitted in week four of the trial, it is not known which program session this was for the participant (i.e. whether or not it was the first session), and therefore, it is difficult to understand the extent to which the child would have been familiar with the program content.

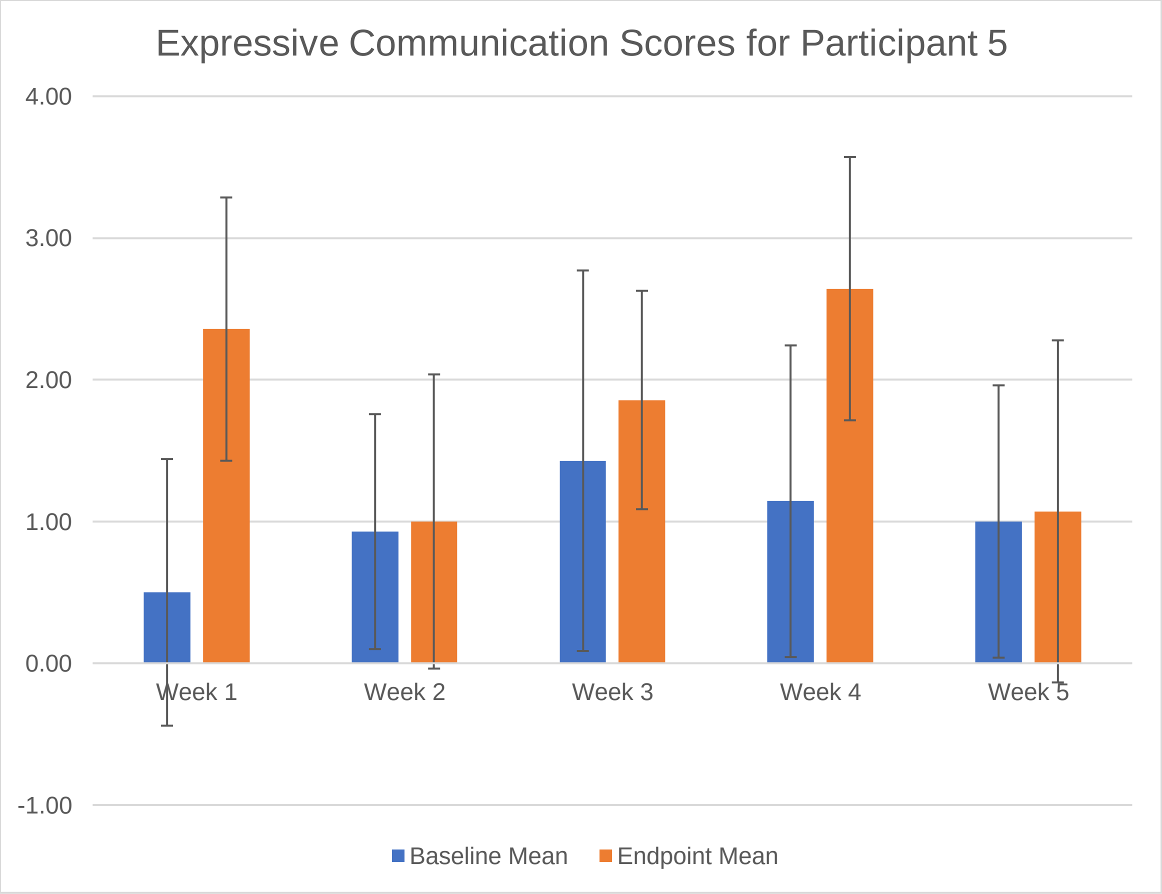
Investigating elements of expressive communication (such as ‘Stutters or Stammers’, ‘Pacing’, ‘Speech Clarity’, ‘Dysarthria or Mumbling’, ‘Words’ and ‘Phrases’) showed a similar picture. As with the mean scores, there was very little change in subscale item scores from the baseline (except for speech pacing, which increased by one point). Generally, the scores relating to expressive communication showed a variable picture overall; the participant showed consistent stuttering and stammering, yet speech was clear, and word and phrase production was high. All subscale items relating to receptive communication were scored the highest value of 4 which was maintained from baseline to endpoint.

Data here show that although expressive communication scores for this individual were variable, they generally remained the same from baseline to endpoint. Receptive communication scores were generally higher in comparison to expressive communication scores and again did not change from baseline to endpoint. As per the participant description, the speed of speech for this individual had increased since they joined the research program in Phase One, and they had moderate symptoms of the disease during the program trial, which perhaps explains the variable expressive communication.

Participant Five

There were five videos submitted of the program sessions for participant five, which were delivered by one learning support assistant, a Music Therapist, and a trainee Music Therapist in the school. Sessions were delivered in the afternoon and the morning, ranging from 9.05am to 2.25pm, with most sessions being delivered in the morning (15 out of the 20 session times recorded). Figure 6.11 illustrates the mean scores and standard deviation scores in expressive communication responses for participant five.

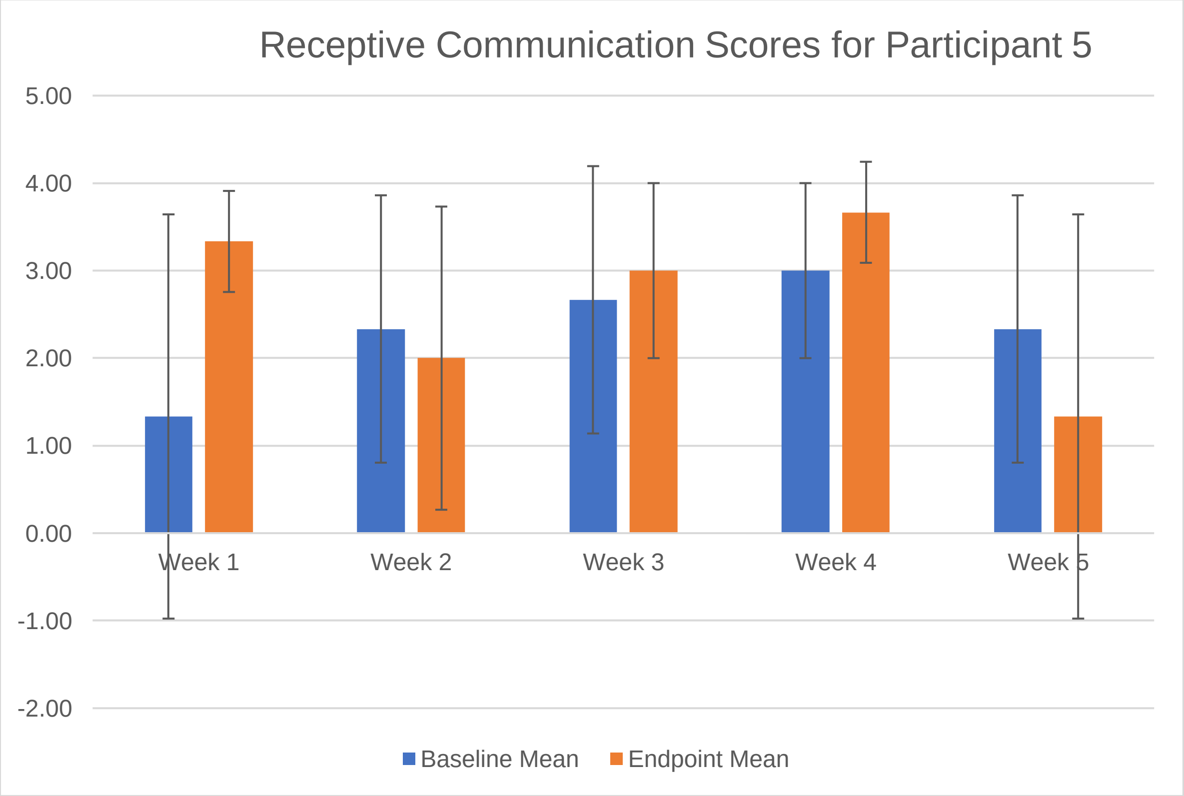
Figure 6. 11: Expressive communication means scores and standard deviation values for participant five



As can be seen from Figure 6.11, mean scores for expressive communication were generally lower than the other four participants (range = 0.50-2.64), and there were inconsistent patterns in the recorded mean scores over the six-week program trial. Data here indicate that there were no consistent changes to expressive communication in response to the trial. However, the sessional mean scores in the program trial showed a more consistent pattern. Session by session, all mean expressive communication scores increased from the baseline to the endpoint, increasing on average by 0.79 points ranging from 0.07 to 1.86, across the trial six weeks. The increased weekly baseline mean scores could indicate a short-term positive change in expressive communication in response to the Music Speaks program.

Exploring the receptive communication responses, however, depicts a variable picture of change. As can be seen from Figure 6.12, scores were again variable over the six weeks (range = 1.33-3.67) and there were no consistent patterns of change observed across the program trial. Data here indicate that the receptive communication did not consistently change in response to the program trial, however, mean scores were generally higher than expressive communication scores. Sessional mean receptive scores depict a variable picture, with some baseline line scores increasing (week one, three and four) and others decreasing (weeks two and five). The average change score was 0.33 (range = -1.00-2.00).

Figure 6. 12: Receptive mean communication scores and standard deviation values for participant five



Delving deeper into individual subscale item scores indicates a similar picture of both variance and change. The expressive sub-items of ‘Stutters and Stammers’, ‘Speech Clarity’, ‘Pacing’, ‘Improvement to Dysarthria’, ‘Words’ and ‘Phrases’ all showed inconsistent scores across the six weeks. However, except for ‘Pacing’ in week two and ‘Words’ in week three, all subscale items either remained static or improved session by session. Data echoed the patterns seen in the mean scores, whereby patterns over time showed inconsistencies, but scores session by session showed greater instances of maintenance or improvement. For this participant, the low scores in production of phrases perhaps that this level of speech was not achievable.

Receptive subscale items that included ‘Word Recall’, ‘Responds to a Familiar Voice, and ‘Ability to Interact, Respond and Participate’ also showed variance for this participant. Twelve weekly scores increased or remained static from baseline to endpoint, however, three scores decreased from baseline. Data here indicate that receptive communication could have increased each week in response to the program, however, scores are wide-ranging and variable over the six-week trial.

As there are data recording sheets for this participant, it was possible to explore how professionals’ comments related to the child’s communication responses week by week. As with participant three, generally, comments related to the child’s engagement, the need to prompt the child, and the impact of fatigue. There were no comments though for this participant related to the theme of adaptation as with participant three, perhaps due to the lower cognitive abilities of this participant.

Variable Engagement

During the program trial, there were several comments which related to the child’s engagement with the program. For example, in early sessions professionals commented how the “…song is too long to hold their concentration” (participant five, session 3), and in later sessions, professionals commented on how “…the instrumental feels too long in the song and they lose focus” (participant five, session 17). Comments here suggest that songs for this participant needed to be shorter in length to hold the child’s attention and engagement. Other comments in later weeks related to better engagement with initial spoken tasks, but a similar response to the program songs, describing how: “1. the word rest was repeated very clearly, 2. lost interest with the song and refused to join in” (participant five, session 12). Comments like this showed how engagement at the beginning in task one was good, but then declined during the song and later tasks. For this participant, evidence suggests that engagement may have been improved with shorter songs and program tasks.

Engagement with the program was variable and other comments described good engagement with the program songs, where later sessions described how the child was “…really engaged with the song :)” (participant five, session 15). Comments here relate to different program sessions and days of the week, and evidence suggests that responses to songs were variable session-by-session and were not specific to the type of song delivered. In later weeks, professionals commented on how the child “…enjoyed listening to the programme after their session with B” (participant five, session 23). This comment described how the school delivered the program after a music therapy session with their Music Therapist ‘B’ and good engagement was observed following this therapeutic intervention. Score data demonstrate that the child was able to complete most of the tasks from this session which could imply that a session with the Music Therapist beforehand helped improve the child’s engagement. Comments suggest a variable picture of engagement both within each program session and across the trial weeks, and perhaps imply that shorter tasks would have been more appropriate for this child. Data also suggest that music therapy sessions beforehand, facilitated better engagement before they participated in the program.

Need for prompting

Various comments from professionals described again how the child was able to complete tasks with the help of staff prompts. One professional commented that the child “…said word ‘food’ and ‘drink’ when asked to repeat, repeated ‘food’, ‘curry’, ‘water’, repeated ‘food’ and ‘drink’…didn’t repeat the whole sentence just key words. Really good day :)” (participant five, session 10). Prompting the child to repeat verbal tasks in this instance seemed to help the child to reproduce key words in phrases. Score data from this session indicated that all tasks were completed by the child with the help of this staff prompting. In later weeks, professionals again, observed how prompting helped the child to engage more; “…with staff prompts they were able to spontaneously say the odd word but with no staff prompts they did not repeat anything” (participant five, session 18). Comments here emphasised the importance of additional staff input and prompting to help the child remain engaged and respond to the program tasks. It is not known, however, the level to which the child was understanding and responding to the verbal tasks or simply repeating the words they heard in staff prompts.

Fatigue

Lastly, one comment related to the child’s fatigue. In session eight, one professional commented that the child was “…yawning through song, could see that they were listening to the song” (participant five, session 8) describing how the child was still engaged in listening even though they showed fatigue. The child was unable to complete any tasks for this week which suggests that the child’s engagement was impacted by fatigue.

Data for participant five here describes a narrative of variable engagement and response to the Music Speaks program. Comments offer context as to the reasons why there were lower scores in some sessions, which was due to a lack of engagement, fatigue, or how the program songs were too long. In other cases, factors such as staff prompting, or music therapy sessions perhaps helped improve engagement and task completion. There was an overall variable picture of engagement in the trial which perhaps echoed the progressed Batten disease symptoms for this individual.

6.5. Overview

Firstly, the refinement and development of the CMTOM communication subscale offered the opportunity to track and monitor participants' communication in more detail than previously captured. With further development, this subscale could offer a unique perspective in assessing participants with communication difficulties such as Batten disease or other childhood or adult conditions which affect speech.

Secondly, the data presented here illustrate a dynamic picture of children's communication responses to the Music Speaks program. Overall, mean results suggested that after participating in the program, children's expressive speech improved or maintained in their sessional mean scores. Individual participant scores supported this finding for two participants, where clarity of speech, pacing, dysarthria and stuttering improved in the short-term each session in response to the program. For other participants, scores were either maintained or marginally improved. Implications of these findings suggest that elements of the program (such as rhythmic pacing, phrasing, and metronomic speaking) could have had short term positive effect on a child's clarity of speech, and techniques such as rhythmic speaking or rhythmic tapping could help children slow their speech down and improve the intelligibility of their speech. Mean scores relating to expressive speech, generally varied over the six weeks, however, and there were no consistent patterns of change observed over time, suggesting there was no longer-term impact of the program. This finding illustrates the variable presentation of participants over the six weeks, further evidencing the heterogeneity of communication responses in the participant population. Some scores for participants (participants one and two) remained at their highest over the program trial. This may suggest a ceiling effect occurred or may have been reflective of the fact that these participants were in the early stages of their disease progression and their functional communication was better (later addressed in the discussion chapter see sections 8.5.6 and 8.7.2).

Receptive speech generally yielded higher scores for participants, suggesting that children's receptive communication was generally better than their expressive communication. Mean scores here were variable over the six weeks and no consistent pattern of change was observed over time. Session-by-session, mean scores varied where responses either improved or declined after each program session. This finding was also echoed in the individual participant data where scores varied considerably, following a variable pattern of change throughout the trial. This suggests that word recall, response to familiar stimuli or overall interaction were less influenced by the program session-by-session and over the six-week trial period. During the process of analysis, it was also important to mention that the filming dates of program sessions were specifically chosen to reflect different days of the week. From the data, weeks four and five represented Thursdays and Fridays, and children may have shown more fatigue towards the end of the week. The mean scores, however, did not show any difference in scores in these weeks suggesting that this did not have an impact on the ability to engage with the program on these days.

Qualitative data from recording sheets provided more context around how the child responded to program sessions and the variables which may have affected their ability to engage with the program (such as fatigue, engagement, the need for prompting, or the length of program songs). Program comments reflected the heterogeneity of the disease, alluding to the fact that a child’s engagement, presentation and participation were variable day-to-day. Although most of the recorded comments related to different sessions from those which were assessed using the CMTOM (referenced in the discussion chapter, see section 8.5.3), insights from professionals explain how each child responded on each day that the program was delivered, and important contextual information about the child at that moment on any given day. Factors such as program duration, musical environments and staff prompting were important considerations in how the program could be delivered to other children with varying communication skills in the future.

It is important to recognise that different types of professionals may have had different experiences of the Music Speaks program. Comments on the recording sheets regarding contextual influences or detailed observations were influenced, perhaps, by their experience, knowledge and understanding of the child, perhaps impacting how they expected the child to respond to the program. They would undoubtedly have had different perspectives to one another as to what constituted a task as being ‘complete’ and why or how the child was engaged with the program on each day. Furthermore, learning support assistants may have felt more inclined to prompt the child in the program, and this may not have been a technique that was adopted by everyone in the trial. Musical professionals, such as Music Therapists, may have felt more comfortable delivering the program. It is possible that the professionals who took part may have volunteered to be part of the project because of a pre-existing interest in music or based on a child’s affinity to music. Further development of the program could consider how different types of professionals’ access and deliver the program, and how implementation considers this factor. This is discussed further in chapter eight of this thesis.

6.5.1. Learning Points

In response to the research questions, findings from Phase Two are summarised in the following learning points:

2.1. Can the Music Speaks program be delivered effectively in a formal educational framework?

- From the recording sheets retrieved, schools were able to deliver over 90% of the program to the children participating in the program trial.
- Program sessions were delivered at roughly the same time daily.
- According to professionals, participants were able to complete 42% and 69% of the program tasks over the six weeks.
- According to professionals, engagement with the program varied and appears to have been influenced by factors such as fatigue, mood, a need for variation and prompting.
- A wider participant pool and improved electronic data collection methods are needed to explore feasibility further.

2.2. How do communication outcomes for children with Juvenile Batten disease change in response to the Music Speaks program?

- Expressive communication traits such as clarity of speech, pacing, dysarthria and stuttering improved after program session was delivered and completed by participants.
- There were no observed sustained patterns of change in expressive communication over the full six-week period according to the data.
- Receptive communication was generally better than expressive communication but remained variable each week and did not change markedly at the end of the six weeks according to the data.
- Further research is needed to investigate the full breadth of communication skills over a longer period with a wider participant pool and consider how different types of professionals may understand and implement the program.

Results from Phase Two indicated that elements of music therapy could be transferred and delivered in an educational context and the preliminary evidence suggests the program had some impact on a child's communication responses for those affected by JNCL (from the perspective of professionals working with the child and from researcher observations). The data here, however, only depict part of the whole narrative referencing program delivery, completion, brief staff comments, and observed responses from video data. Interviews with educators in the third phase help paint a more holistic picture of the educators' experiences, both in response to the program and more generally in an education context.

Pursuing the third research aim, therefore, the next phase of the research explores professionals' experiences. From the interview data analysis, the next chapter outline's the themes, subthemes and narratives from the professionals themselves, exploring challenges, considerations, and experiences of supporting a child with JNCL in an educational and therapeutic context, as well as their perceptions of delivering the Music Speaks program.

Chapter 7: Results from Phase Three Interviews with Professionals

"Music is the shorthand of emotion"
— Leo Tolstoy

7.1. Introduction

This chapter aims to develop the research enquiry further by exploring the experiences of professionals and educators who support children with JNCL. The data used in this phase were taken from interviews conducted with therapists, educators and visual impairment specialists, who took part in Phase Two of the project. The professionals supported four of the five children who participated in the program trial. A summary of the approach taken to each research question is set out below:

3.1. What are the experiences of educators and therapists supporting a child with Juvenile Batten disease in an educational context?

This phase aimed to answer this research question by analysing the day-to-day experiences of those people supporting a child with the disease, as well as capturing some of the broader considerations faced by professionals involved in education, care and therapy.

3.2. What are the perceptions of the Music Speaks program from those using it in educational contexts?

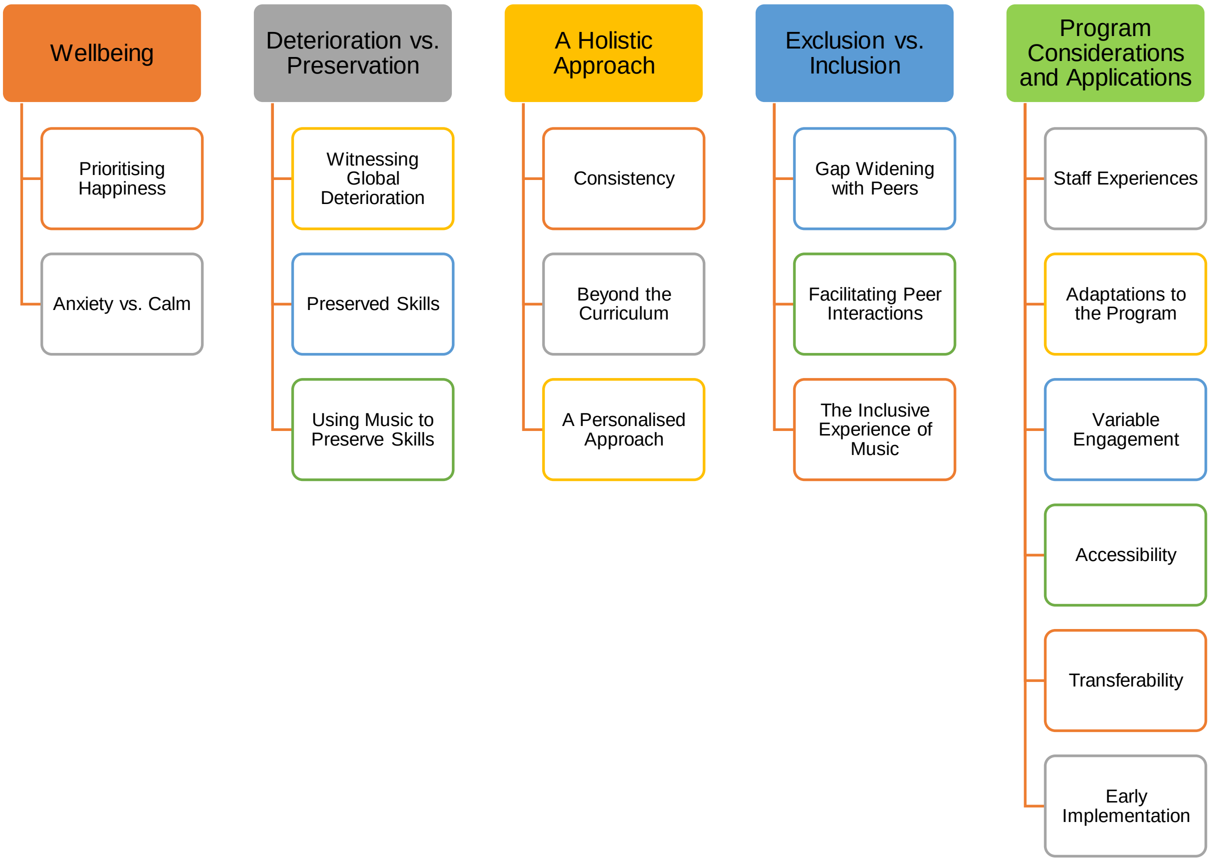
Data from professional interviews were also analysed with regards to their experiences and observations of delivering the Music Speaks program daily with children affected by JNCL.

This chapter begins with an overview of the key themes emerging from the data before exploring each theme in detail, drawing on interview quotations. For the purposes of anonymity of the children, references to gender have been replaced with gender neutral terminology.

7.2. Overview of Key Themes

Several topics were covered in the interview questions. Professionals were interviewed about the child they supported (including any challenges they face); their role (including any approaches and challenges in their work); the child's engagement with music; experiences of the Music Speaks program; and any general comments or reflections. Interview data were organised into five key themes and 17 subthemes. The five key themes were as follows: 1. Wellbeing; 2. Deterioration vs. Preservation; 3. A Holistic Approach; 4. Exclusion vs. Inclusion; and 5. Program Considerations and Applications. Figure 7.1 illustrates the five key themes and the respective subthemes.

Figure 7. 1: Thematic Map of Phase Three interview data analysis



Themes one to four cover different aspects of the professional's experience working in their role supporting a child with JNCL. Theme five relates to their experiences of the program trial.

In response to research question 3.1, themes one to four represent participants' observations, perceptions and descriptions of their daily experiences of education and care. Theme '1. Wellbeing' includes the professionals' experiences of both the child's wellbeing and their own wellbeing in their roles. The remaining three themes (two, three and four) represent ideas that sit on a spectrum. For example, professionals described the global deterioration that affected almost all areas of a child's life, in contrast with the recognised preserved skills, behaviours or personalities that prevail despite a child's deterioration ('2. Deterioration vs.

Preservation' theme'). In other instances, professionals reflected on the need to maintain consistency, routine and familiarity for the affected child versus the importance of adaption, flexibility and the need to go beyond the curriculum ('3. A Holistic Approach' theme). Finally, professionals described the developmental and emotional gap that emerged between the child and their peers, and specific instances where professionals stressed the importance of dedicated times away from the classroom, as well as facilitated peer interaction spaces, or inclusive activities in the classroom ('4. Exclusion vs. Inclusion' theme). Contained within these first four themes are also sections that refer to related experiences and perceptions of the Music Speaks program. For example, narratives described how the program could facilitate improved communication or self-expression skills, contribute to the sense of consistency and personalisation, or bring a sense of inclusion for the child.

In response to research question 3.2, Theme '5. Program Considerations and Applications', relates mainly to experiences of the program trial. 'Program Considerations and Applications' focuses on staff experiences of the program discussing how professionals felt valued and supported in their roles, and how the program supported them with a sense of empowerment, as well as an understanding of the wider benefits of music in education. The theme also discusses observed limitations, adaptations and considerations of the future use of the Music Speaks program in other contexts and with other children.

The five themes are united by the overall sentiment that the professionals went above and beyond the standard curriculum to enhance the school experience for the child they cared for. With consideration to the child's unique and changing needs, professionals demonstrated a deep understanding of what the child needed from them at the present time, and how their education could support them in the future. The themes also depict the importance of music and how particularly, elements of the Music Speaks program could contribute to enriching school experiences for the child.

7.3. Wellbeing

A common theme that echoed in all interviews was the notion of wellbeing and quality of life. All professionals expressed the importance of wellbeing and the role it could play in the child's daily experience of the education setting. Subsequently explored within the subthemes, professionals described how they saw their role as facilitating wellbeing, prioritising happiness, calm, and relaxation, and reducing anxiety for the child when they were at school and in their care. Narratives touched upon how music contributed to this and the subsequent positive impact for the child in the school setting.

7.3.1. Prioritising Happiness

On many occasions, professionals simply described their role as helping the child to have fun and be happy. One professional reflected that their job was to "…make them happy, you know, to make school life as happy as it could be" (participant six, Visual Impairment [VI] specialist) and another commented how "…that's what it's all about, is creating fun times and happy times for them whilst being at school" participant three, VI specialist). Professionals expressed in interviews how they prioritised a child's wellbeing and happiness at school. Perhaps by prioritising wellbeing, the academic side of the school environment was deprioritised by staff, and children had a different experience in school compared with their peers.

Others also felt that music was important to a child's sense of wellbeing and happiness. Professionals described the experience of seeing the child 'light up' in response to a familiar song: "I think it's a sense of their face just lighting up when they hear a song that they recognise and then they want to join in" (participant five, inclusion lead) and "…their face kind of lit up as soon as music was played" (participant two, Music Therapist). Comments suggest music had the potential to create a sense of clarity or connection where children 'lit up' to music and responded in a unique way, more so in some cases than anything else (discussed further in section 7.4.3). Others reflected on how they felt singing impacted the child's sense of wellbeing:

> "…they love music…they'll just suddenly just sing, burst out singing a song… And they'll be singing away, and like, they won't be in tune, but they're smiling and they're giggling…so, it brings out all their confidence and their abilities… they will just sing and it's lovely" (participant three, VI specialist).

Describing the immediate impact of singing, the professional observed how music could increase the child's wellbeing by facilitating happiness and encouraging confidence. There was a sense that the immediate benefits of singing could reduce the child's inhibition, and songs could facilitate self-expression through music. The same professional went on to describe their understanding of how music was important in creating a positive experience for children:

> "I didn't think that music would have such a positive impact on children with Battens. I've never worked with a child with Battens before until I met them, and I can see that music is a real, good positive experience. We can't go through the morning without listening to music" (participant three, VI specialist).

Surpassing their expectations, the professional here conveyed their recognition of how music had had a positive impact on an affected child, and how it had developed into an important part of their ongoing care. Professionals also saw the value of daily and repeated musical input, where regular times in the school day involved music listening for this child (the importance of which is further explored in the subtheme Consistency, see section 7.5.1). Others reflected that being part of the project has "…really made me think, as well, just how amazing music is. You know, it brings back a lot of memories for people. I think it just shows music's embedded in your memory" (participant four, VI specialist). Recognising the positive impact of music for the child, comments here emphasised how the research contributed to professionals' understanding of the importance of music, and the connection with memory and positive experiences.

Bearing in mind the focus here on wellbeing and happiness, professionals also described moments where the child was unhappy, anxious, confused, or scared and how their role supported and attempted to counteract this. It is important to clarify that, rather than this relating to a clinical diagnosis of anxiety, this label rather refers to attributes of anxiety that relate to confusion, fear, discomfort and disorientation, which are subsequently explored in further detail.

7.3.2. Anxiety versus Calm

In describing moments in the school day where the child was concerned, confused or anxious, professionals reflected on the impact this had for both child and professional, and how their role counteracted these experiences. Two professionals described the anxiety for one child they both supported:

> "…as time went on, they got very anxious. And they weren't very good with transition, moving from place to place…with their anxiety, and it was the moving and handling aspects as well…they felt scared, they'd jitter, yeah, they were scared. It's sad, really" (participant one, Learning Support Assistant [LSA]).
>
> "…was becoming a lot more confused. Just in their everyday life…confused by their routine, confused by their environment and kind of their sensory needs and they were becoming a lot more anxious…becoming anxious about kind of physically where they were and a bit disorientated" (participant two, Music Therapist).

In describing fear and anxiety, the first professional observed how certain activities in the child's day provoked distress. In moving locations, or being physically moved by staff, it was noted that perhaps this caused distress because of the uncertainty associated with changing location, or perhaps there was discomfort in being physically touched. As well as the physical reasons for the child's anxiety, data here also described the sadness and empathy experienced by professionals in response to the child's distress. "As time went on…" gives a sense of change over time, and how worsening anxiety was saddening for the professional caring for the child. The second professional observed how sensory and environmental experiences could feed into the child's anxiety, disorientation and confusion, describing how the child experienced their school environment and how this again worsened over time.

For another child, a sense of disorientation, anxiety and confusion in the school environment was also observed by professionals:

> "…they used to get very, very nervous about uneven surfaces just going from tarmac to grass, things like that, and their spatial awareness changed…they'd come out of the toilets in year six and just think, 'Oh, I'm not sure where I am. I'm not sure which way to go anymore'… and that must have been really scary" (participant five, inclusion lead).

Professionals explained how, in their opinion, confusion and disorientation could impact a child's experience of school recognising their feelings of fear and anxiety. Data here emphasised the sense of empathy and understanding that professionals had, seeing things from the child's perspective, and attempting to understand how the child experienced the school environment. A sense of change over time was also described, where professionals observed and understood how anxiety, confusion and disorientation worsened over time for the child they supported.

In response to the child's anxiety, confusion and fear, professionals in many cases discussed their approaches to reduce anxiety and induce calm, understanding how, for example "…they liked to feel secure. And they liked to hold their chair pommel in between the legs and know that they were safe in that space" (participant one, LSA). This suggests that grounding and physically holding onto something helped them feel safe and secure, where perhaps they were experiencing the sensory disorientation previously mentioned. Others also used grounding techniques where they "…kept saying that they were safe, reminding them where they were, and what we were doing" (participant two, Music Therapist). Self-expression was also described in relation to wellbeing, where one school were "…doing lots of work at school about strategies for feeling calm and places to go and communicate your feelings" (participant five, inclusion lead). The data here described how professionals employed grounding techniques to instil calmness, reduce anxiety and help ground the child and open up space for self-expression. Later explored in the 'Holistic Approach' theme, professionals here introduced the idea that they adapted and evolved with the child's changing needs rather than seeing their role as strictly educational.

Professionals described how music was involved in their approaches to help calm the child. One professional commented that "…music always settled them, always calmed them down" (participant three, VI specialist) and others spoke about how

"…we would often just go and listen to music, and that would get them to calm down, and we would sing together" (participant six, VI specialist). Music was talked about positively and fondly by professionals, as an activity that they could engage with alongside the child and was often the consistent medium by which calm could be achieved.

Often anxiety was described as a barrier to effective communication with the child, and others reflected on how this impacted the child's engagement:

> "…was all about their anxiety levels. And if they were through the roof, then you couldn't really talk but if you're nice and calm, you got a really good, a good day and some days you had really good conversations" (participant one, LSA).

According to this professional, the child's anxiety levels directly impacted their ability to converse and engage with the professional. The LSA emphasised the importance of their calm approach in reducing anxiety for the child to facilitate productive communication. The professional here described how their approaches and behaviours towards the child could influence the child's wellbeing, outlining the importance of the professional's response to anxiety. Other professionals described how their patient and calm approach could help with a child's communication, discussing the importance of a patient approach to communication:

> "…we won't butt in and try and finish their sentence for them. We'll just wait and wait patiently until they're finished…they'll still be trying to stutter and find the right words, but it's very important to let them finish what they want to say (participant three, VI specialist).

Another VI specialist echoed a similar patient approach in addition to slowing down, recalling how one child would say "'I can't get my words out'… and I'd say 'slow down, and you know, you can tell me your own time'", alongside knowing "…to slow down my speech with them" (participant six, VI specialist). Data here suggest that professionals employed strategies that involved modelling slowed speech in a patient and calm way to help nurture the child with slowing their speech and communication.

As well as recognising anxiety in the child, professionals expressed their own feelings of concern, where others described their response to the child's fragile condition:

> "So you're constantly…'are you ok? is everything ok' and if there's something different 'are you ok? Is everything ok?'. Because I would be absolutely petrified…I mean it's going to happen I know" (participant seven, LSA).

The professional here expressed their feelings of dread and fear in relation to the possibility of the child having a seizure at school. The child’s medically fragile condition was undoubtedly evoking fear for this professional, representing the anxiety that perhaps professionals experienced in their roles supporting a child with Batten disease.

In this first theme, a sense of wellbeing, happiness, and calm for the child in their care were described by professionals as the priority of their role. A deep level of understanding and empathy was demonstrated, as professionals could imagine the experience of school from the child’s perspective. Strategies mentioned here emphasised how professionals prioritised wellbeing and positive experiences, rather than curriculum teaching. Professionals identified the challenges in supporting a child who was experiencing anxiety, confusion or disorientation, and their feelings in response to this. The idea that music could facilitate wellbeing was mentioned consistently in all interviews with professionals. The idea that music could facilitate and preserve functional skills is further echoed in the following theme relating to deterioration and preservation.

7.4. Deterioration versus Preservation

When professionals were asked about the child they supported and the challenges they faced, all described the inevitable deteriorating condition of the child and the impact this had on many areas of the child’s life at school. Yet, professionals also described moments where they observed that certain skills or traits remained preserved for children, despite declining functional skills. Moreover, in many cases, music was observed to help foster and maintain skills such as physical movement, communication or orientation around the school environment, more so than other activities or therapy. Professionals also described how elements of the Music Speaks program were perceived to help preserve functional skills for the child, namely independent communication and self-expression. This section begins by exploring the topic of deterioration.

7.4.1. Witnessing Global Deterioration

At the beginning of interviews, when asked to describe the child they supported, professionals began by describing the many challenges brought on by Batten disease and the child’s declining skills. Areas of deterioration were identified in most areas of the child’s academic life, and included the impact of sight loss, cognition and memory, mobility, communication, writing, and fine motor skills. Reflecting on the decline over time, a sense of sadness was expressed from staff as they reflected on how the child used to be before they started deteriorating:

> “They were really, really bright. And then it all just… the Juvenile Battens disease took over…and it's just sad to see. They still had days where you could see that they wanted to engage. But it's just… it's disheartening. It's not nice to see, it's sad…they struggled (participant one, LSA).

The professional’s emotional experience emphasised the sadness and the disheartening nature of supporting a child with the disease, especially witnessing the child’s declining health. A sense of hopelessness is alluded to here, where the child was perceived as somehow still wanting to connect but restricted by physical abilities. Experiences here depict how professionals could feel demoralised in their roles as they observe how the disease overtakes the child they once knew. A sense of inevitability of the disease progression is present along with the sadness of witnessing decline over time.

Professionals often described how deterioration could impact the child’s confusion in the school environment. Sight loss, for example, was reflected on as a major disruption in the child’s ability to participate:

> “…due to their vision, they can't see very well at all, so they struggle to write…they struggle in PE now because there's too many kids running all around and being boisterous that they don’t know which direction they come in… So, it freaks them out a little bit” (participant three, VI specialist).

Talking empathetically, this professional understood how the deterioration of sight negatively impacted the child’s school experience. Professionals empathised with how children could feel overwhelmed and confused in environments that were perhaps unstructured and relied on being able to see those around them, recognising the distinct challenges associated with attending a mainstream school with a visual impairment. Others reflected on the impact of sight loss and how this impacted communication:

> “…because they couldn't see...they don’t get those, you know, the subtleties of conversations and things like that… they don’t see who's involved and who's not involved” (participant five, inclusion lead).

Describing small observations like this demonstrated how professionals recognised and deeply understood the different types of challenges for the child with a visual impairment, emphasising how their role and knowledge went far beyond the child’s education.

The notion of increased fatigue and cognitive deterioration was referenced in all interviews as a key factor that impacted activities both at the end of the day and towards the end of each school week. Professionals reflected that in the case of two children: “…they would tire, especially Friday” (participant four, VI specialist) and "…they do all their work, they get very, very, very, very tired…we noticed there was a deterioration towards the end of the week (participant seven, LSA). Professionals talked about how they responded to the child's fatigue, and deteriorating sight and physical abilities:

> "…it's their concentration skills, it's their mind, how they focus. And the fact that they're only able to focus for about twenty minutes and then that's it… so we did keep it short and sweet" (participant seven, LSA).
>
> "…when they couldn't access the keyboard that they had, when they still had some ideas, we used to write everything down for them" (participant one, LSA).
>
> "…other things such as PE - they would struggle with those and the mobility side of it. So, for PE, we'd do things that were a lot more gentle for them" (participant four, VI specialist).

In emphasising an in-depth knowledge of the child, professionals often spoke about how they responded to the child's deterioration by adapting the activities or their school day accordingly. In the case of these comments, this was through tailored activity sessions, transcription of the child's ideas, or less demanding physical activities. This theme of adaptation arose in many different areas of interviews, where professionals showed how they translated their understanding of the child's changing needs, into adaptations or strategies which ventured outside of the school curriculum (further discussed in section 7.5.3). The concept of fatigue was also referenced in the recording sheet comments from Phase Two of the program trial, where children's engagement in the program decreased when they were tired.

In exploring the area of communication, all professionals felt that deteriorating speech and language skills significantly challenged the child's ability to express themselves, communicate and be understood. Reflecting on how this had changed over time, professional described their experiences of supporting a child with deteriorating speech skills:

> "When I was working with them, at the beginning, I might have understood most of their sentence, although it was very, very fast. Whereas towards the end, the speech, they were speaking very fast, and I'd pick up on certain words, rather than much of the sentence" (participant four, VI specialist).

> “Their speech, lots and lots of stammering, stuttering, forgetting their words, and it became increasingly hard to understand what they were saying…it's worse when their anxious” (participant six, VI specialist).

> “…they’re on like a little hamster wheel and it goes round and round, and the conversations are repeated” (participant seven, LSA).

Professionals concurred with the difficulty in understanding and connecting with the child as their speech deteriorated noticing how the child’s speech sped up, was stuttered, stammered, or repetitive. Participant six believed the negative impact on speech and communication was a consequence of anxiety within the child, coinciding with other subthemes captured as part of the Wellbeing theme. From the responses of the professionals interviewed, the main barriers in understanding and communication between child and professional were as a result of the common symptomatic traits of Batten disease. Commonly, professionals concurred that the speed of the child’s speech often led to stuttering and stammering, and this had the greatest impact on their intelligibility, implying that the barriers in communicating with the child impacted how they supported the child.

In reflection of the journey of deterioration, professionals revealed the demoralising and inevitable nature of the Batten disease journey, demonstrating a deep level of understanding and empathy for the child’s experience. Accounts here revealed how sight loss, cognitive decline and deteriorating communication significantly challenged the child’s ability to connect with peers and participate in lessons. Data also revealed how the professionals’ experienced challenges with connecting, understanding and interpreting the child’s communication. Comments here suggested that professionals understood the importance of an adapted environment and curriculum to suit the changing and deteriorating needs of the child. Despite global deterioration, there were, however, instances where certain skills and interests were largely preserved for children. This is explored in more detail in the following subtheme.

7.4.2. Preserved Skills

Despite skills that were deteriorating, interviewees reflected upon aspects of the child’s presentation which they felt remained intact. These included elements of identity and personality, interests and hobbies, and memory for specific people and experiences. Professionals observed certain areas which remained unaffected by their memory loss, with reports of how a child could remember stories and narratives:

> “They can still read…their books for pleasure. And the books that they’re reading, if it's something fairy story that they enjoy, they will remember some of it” (participant six, VI specialist).

In recognising elements of the child’s intact function, professionals here identified how familiar, repeated and enjoyable interests or topics were, in their opinion, still accessed in the child’s memory and perhaps remembered more easily. Professionals described how people were retained in the child's memory:

> “…that was really important for them to, I don't know, to feel involved in something or have a link with a person. People are important to them, they link people with experiences… So, for example, they can tell you all the names of the teachers that have taught them since reception at school. So if you're walking down the corridor, they could say ‘yes, in ladybirds I had so and so, and in year one, I had so and so in year two’. So yeah, people are very important to them” (participant five, inclusion lead).

The interview data here suggested that, according to professionals, key people and related experiences were more easily accessed in the child’s memory. A sense of inclusion was also an important aspect of these observations, where professionals identified the importance for the child of connecting with others and feeling involved rather than excluded or isolated (a topic which is explored later in the Exclusion versus Inclusion theme).

In summary, professionals observed that there are many ways in which the child could still connect with memories, interests, and experiences despite deterioration. In the professionals’ experience, the child’s interests and information that was meaningful to them, seemed largely unaffected by the condition and, where skills allowed, these could still be expressed and accessed (i.e. a love of stories, people and experiences). Professionals went on to describe how music could help preserve certain skills, explored in the following subtheme.

7.4.3. Using Music to Preserve Skills

In addition to key memories, connections and interests that were seemingly unaffected by the disease progression, interviewees suggested that other functional skills improved in the presence of music. Professionals all commented on the unique way in which children responded to music being played. One professional commented on how music could facilitate functional responses:

> "…it's the one thing… that they responded to over the years that really brought them out of themselves. It kind of spontaneously promoted and provoked responses from them verbally and vocally and physically…" (participant two, Music Therapist)

According to the Music Therapist, music could elicit functional responses from the child beyond other activities at school. The concept of confidence and connection is also alluded to, where music is the one thing that connected with the child, encouraging engagement and response. Other professionals observed how music could help improve physical skills, speech, and cognitive functioning, where perhaps other activities could not. Concerning physical skills, a professional commented that "I noticed that if you said, you know, 'get down, get up', when it was actually to do with the music, to me, they could move a little bit better" (participant six, VI specialist). Witnessing the positive impact of music, professionals believed that physical activities could improve with music suggesting an impact on the child's functional skill.

Professionals commented on how they used music creatively and consistently to help preserve cognitive skills, specifically memory, learning and orientation during their care and education at school. For example, professionals reflected on how they used music markers throughout the day could help the child know which activity or room they were entering:

> "…we'd always start with a song. So, when they came into the door, they knew what was coming. They liked that. They like routines. Music helped them with feeling comfortable with routines and what was going to happen next. If they knew what was coming and what was expected, it helped them to just relax into the day" (participant five, inclusion lead).

Narratives here reveal how professionals understood the importance of using music to help support the child to understand what was happening next to reduce confusion and anxiety (see also section 7.3.2). Professionals described how, they used music as an auditory cue to help the child orient themselves in activities or different classrooms in their day (perhaps in the absence of a visual timetable which other children would have used). This approach is just one of many strategies that professionals described using, to help the child maintain their skills within the school environment that went above and beyond the standard curriculum teaching (see section 7.5.3).

All professionals concurred with the unique effect that music had on the child's language and communication skills. Professionals observed that music and singing helped reduce the stuttering and stammering symptoms often experienced by children with JNCL:

> “…their speech was often really quick, but when they were singing their speech would slow and would become really measured, so singing seemed to be a really good way of kind of, them having some control over their speech” (participant two, Music Therapist).

Observing how the speed of speech was increased for this child, this Music Therapist felt that the act of singing physically slowed the child’s speech down and helped pace their speech. They indicated that music had the potential to be a tool in supporting the child’s communication, gaining control over their speech and slowing them down to the pace of the music. Others also agreed with how music had a controlling impact on the child’s speech and how lyrics were easily remembered:

> “…they used to struggle with their speech, but when they sang a song, there were no stutters or stammers, and they could sing the whole thing through…that was very interesting for us to see at school. Because sometimes they wouldn't be able to have a conversation with us, but if we put the music on, they could sing a whole song” (participant five, inclusion lead).

Emphasising the positive impact of music, professionals observed how music could actively control and change the child’s speech and communication, even when they were unable to converse with those around them. Considering professionals identified communication as one of the most challenging aspects of supporting a child, there is evidence to suggest music could help enhance communication for the child and those around them.

When asked if they wanted to conclude with any closing remarks, professionals commented on the overall importance of music therapy for children with Batten disease:

> “People might be reluctant to think, ‘Oh, no, music’s not gonna do anything’. But it really does. It has such a big difference…every child with Battens should have access to music… if they had the routine and they could put some music therapy in every single day, I think you're going to have a really, really good positive impact…music’s really, I think it's vital. It's vital for children with Battens” (participant three, VI specialist).

Recognising perhaps resistance in others with regards to the use of music, comments here champion the benefits of music and music therapy. Professionals suggest that all children affected by Batten disease could benefit from daily music therapy interventions, passionately describing how music is essential. Others concurred with the potential impact of music and therapy for primary education more generally:

> "…we knew the wider impact of music for other children, who joined in and wanted to be part of that. So, I think music is something we should be doing more of in primary schools and the link to therapy, and how we support children in all different areas of learning" (participant five, inclusion lead).

Data here described how professionals observed the unique effect of music for children affected by Batten disease; how they used music in their educational approach for the child; and how they believed that music therapy could support children more widely in schools.

7.4.4. Music Speaks Program

Whilst reflecting on the child's responses to the Music Speaks program, professionals observed instances where the program appeared to preserve skills relating to the child's speech, expression, and pacing. For example, professionals specifically recalled the unique effect of the metronomic speaking parts of the Music Speaks program, and how the child's speech slowed down in response to the tapped beat they heard (task three in the Music Speaks program, see Appendix 9):

> "…when they heard your sentences with the metronome, that's when they really perked up and sat up. And I think that is when they responded best? When they could hear the metronome and the beat in the background. They would just say it really, really, quickly, and yet when they heard the metronome, they slowed down to that beat (participant six, VI specialist).

Considering children were described as having significant stutters, stammers and difficulty remembering their words, professionals believed that techniques such as pacing could support a child to slow their speech down, helping them to communicate more coherently, and ultimately preserving their communication. For the same child, the inclusion lead described the effect of this pacing part of the program:

> "…you could almost see them just getting themselves into a calming zone, to get the words out… they were able to communicate more effectively. And I think it slowed their speech down. And you could almost hear them and see them trying to slow everything down to communicate their feelings. I think in terms of expressing feelings, it really helped" (participant five, inclusion lead).

Not only does the professional describe the positive impact of the metronome effect on preserving the effectiveness of the child's speech, but they also allude to the positive impact on the child’s expression of feelings. Here they described the child’s ability to self-regulate a sense of calm, to converse and express their feelings. The same professional described this in more detail:

> “…in terms of expressing feelings, [Music Speaks] really helped…they learned to say they ‘feel tired’ or they ‘feel hungry’, and those were things that they weren’t able to do maybe before, they just would get cross and frustrated” (participant five, inclusion lead).

They furthered this thought by explaining how the child linked the program phrases with other expressions of feelings:

> “…before we did the music sessions [Music Speaks] with them they would struggle to communicate how they were feeling and struggled to get their words out. Whereas something that I noticed, that was a positive impact was that they could say, ‘I feel tired’, for instance, and that would flow very freely from them… which could mean other things. But it just meant that actually, ‘I’m not feeling that great now and maybe need some time or something else’” (participant five, inclusion lead).

According to this professional, the program phrases appeared to help this one child in multiple ways. The short and accessible phrases helped foster the child’s self-expression. Also, the child used the learned phrases to represent an associated feeling, helping guide their self-expression skills. Although these observations only related to one child, it is plausible that the program could similarly help other children by simplifying communication and helping them to communicate their feelings more easily in an indirect manner.

Other professionals commented on how their experience of the program changed their understanding of how the program could preserve independent skills, emphasising the importance of communication in the child’s future:

> “…as their speech may decline, they’re able to then tap into their mind to think about the key phrases and give them that little bit of independence in the future” (participant four, VI specialist).

> “…it's in the back of their mind, hopefully they’ll be able to recall it, you know, slow down and get some of their words out… because there will be times when or they will need to, you know, say hungry, tired, hurting” (participant six, VI specialist).

> “…it's so important that these children are able to communicate as much as they can on their own” (participant five, inclusion lead).

There was a shared understanding about the importance of promoting independent communication and how the Music Speaks program could actively help children to speak more effectively in the future. Experiences here described how professionals witnessed a positive impact of the Music Speaks program for the child’s self-expression, and recognised that the program could help promote future independent communication.

In summary, the theme of Deterioration versus Preservation has been explored through its subthemes of global deterioration, preserved skills, and the role of music preserving skills. The emotional impact of the child’s deteriorating health and the challenges with supporting and communicating with the child in their care were described by many. In-depth knowledge and understanding of the child were also relayed, where professionals recognised areas where the child could express themselves, their identity, and their experiences. Furthermore, the interview data here identified how professionals observed changes in a child’s speech and other functional skills in response to the program. The next theme, ‘A Holistic Approach’ is explored through the subthemes of familiarity, the concept of venturing beyond the standard curriculum and the importance of a personalised adapted approach to education.

7.5. A Holistic Approach

When professionals were asked about their approaches to education for the child, all participants commented on a multi-faceted approach to education. They described providing both consistency and flexibility; keeping a routine yet constantly needing to adapt beyond the taught lessons in school. The initial subtheme in this section explores the role of consistency, familiarity and routine for the child in school.

7.5.1. Consistency

Professionals firstly reflected on familiarity and the importance of “…a familiar voice, so they weren't too anxious” (participant one, LSA). Professionals described how sounds and voices contributed to a sense of familiarity, which in turn helped reduce anxiety for the child. Others described how the child was more at ease with familiar members of staff:

> “With me, I just think they were more comfortable. When they were talking to me rather than other people. I still think that they stammer and stutter and couldn't get their words out a lot with other people. But I don’t know if that’s more because they were comfortable with me, or whether or not it was just because I knew to slow down my speech with them (participant six, VI specialist).

Evidence here suggested that according to the professional, familiar members of staff perhaps helped facilitate confidence in the child. Comments here again, described the importance of strategies and approaches employed by the professional, and how they modelled slowed speech with the child. Data here suggest that both consistency of staff and familiar approaches contributed to helping the child feel at ease, which in turn could have helped facilitate more coherent communication. Professionals talked about the consistency of strategies across the team of educators and professionals, for example, reflecting on a situation where staff members talked about aligning and transferring goals for the child:

> “…they were saying ’that's really interesting that you’re doing those kind of things’, and in parallel, they would talk about kind of things that they were doing in class…it kind of proved that the teaching assistants were kind of on the same page in a way, and they were thinking along the same lines…and, you know, they saw that it was kind of working and thinking, ‘Oh, what can I try in class?, you know, if they’re doing this in music therapy, maybe I could try something in class’…it is trying to carry that over, isn't it? into other environments, and to make it meaningful” (participant two, Music Therapist).

These comments demonstrated how the professionals considered the importance of consistency between teaching and intended outcomes between professionals, stressing perhaps the value of networking and consistent communication between professions. This implies that communication between professionals helped ensure that support for the child was aligned and complementary. Importantly, according to this Music Therapist, the activities that professionals observed in music therapy sessions led them to consider similar activities that they could implement in the classroom environment. Referencing carryover of activities and techniques, the Music Therapist here acknowledged the importance of transferring therapeutic activities from music therapy sessions into the wider school environment, recognising how transferring skills could perhaps increase the meaning for the child. This concept is further explored in section 7.7.5.

Others delved deeper into the concept of familiarity and consistency, describing their impression of routines, and how disruptions or alterations to the child’s timetable in the school day could be challenging:

> “…every morning, they had their own timetable, which had their routine on for the day, and if something was a bit different, or they weren't prepared for it, they’d find that very challenging” (participant five, inclusion lead).

Supporting the concept of familiarity and consistency, the practical importance of consistent routines and schedules were highlighted here, and how, in their opinion, disruptions caused challenges both for the child. All interviewees concurred with the idea that a routine was a central part of ensuring the child knew what to expect from their school day and how this, in turn, contributed to positive wellbeing.

One VI specialist explained their frustrations concerning the inconsistency of staff support at the school. Referencing inconsistent staff structures, they reflected on their experience of education systems and resources:

> “I have a big argument with our team at the minute because, before I came off, the school were asking ‘Please, can you look after them in the afternoon, they need a one to one’, and there's not enough staff for them to have one to one. So, the TA has two children that both require one to one. So, it's like one's not getting enough attention...but I was being sent to another school in the afternoons with a child that doesn't need one to one. And I used to say, ‘look, it's not fair, why can't I stay? Why can't I stay with them in the afternoons, because it's got continuity, and it's better for them?’ But they just wouldn’t budge. And it's so frustrating to think that this child has got an EHCP, but yet, nobody's willing to budge and help them” (participant three, VI specialist).

Recognising the importance of continuity and familiarity, frustrations here related to what professionals observed as inappropriate and inconsistent support systems for the child. Data here suggested that the school, the professional (an external county visual impairment specialist) and the EHCP support plan identified a need for extended support, continuity and one-to-one support. Yet higher education systems (perhaps wider council education systems) would not deliver this for the child. Data here emphasise the constraints of education and support systems which are perhaps inflexible and do not consider the changing needs of children requiring additional support.

Others touched upon the importance of a basic and consistent knowledge and experience of Batten disease, and challenges associated with the unfamiliar support staff when children were transitioning to mainstream secondary settings:

> “…the two ladies that took over from me, obviously have no idea…. Well, they haven't worked with a child with a visual impairment, and they certainly haven't worked with a child with Battens… it didn't help because one of the ladies kept on saying, ‘I don't know what you're saying, and I can't understand’ which made [the child] more anxious” (participant six, VI specialist).

Alluding to feelings of frustration, comments here stress the perceived importance of a basic familiarity of visual impairment and Batten disease which were lacking in this situation. Comments suggest that the lack of knowledge from the new supporting member of staff was, in this case, detrimental to the child’s wellbeing, creating anxiety in a situation where the child was attempting to communicate. Comments from the professional here also perhaps outline the challenges of transitions in mainstream education and the potential negative impact of settings which lack the consistency of specialist professionals who are familiar with Batten disease.

7.5.2. Music Speaks Program

Considering the importance of familiarity, consistency and routine, professionals described how they felt the program contributed positively to a child’s routine and how “…we did it every day, at the same time every single day. So, it was kind of in their routine” (participant three, VI specialist) and others commented that “…they liked that routine, they liked the structure in the day to hear the songs and to talk about them” (participant five, inclusion lead). Describing the importance of familiarity, consistency, and routine, the Music Speaks program seemed to positively support this. Recalling how they embedded the program into the existing school routine, professionals emphasised the positive impact this had for the child in their school day.

Other professionals described how they shared techniques learned from the program to help build consistency for children between support staff. In one instance where a child was transitioning to secondary school, the professional shared their new understanding of pacing, music and rhythm from the program to help both the child and the new professional to communicate more effectively:

> “I used to just tap on the table and say, ‘just talk slowly, and then, you know, the lady will know what you’re saying’, and they did try… even if it's just one person, just thinking, ’I just tap out a rhythm and help that child to talk’…some people just need, you know, a guidance… a programme like this to think ‘slow down, slow down’” (participant six, VI specialist).

Emphasising the importance of skill-sharing, imparting techniques and strategies appeared to be important in creating consistent approaches, helping others understand how they could support children affected by the disease. Comments here identified the potential uses of the program in helping guide other professionals to understand how they need to support and respond to the child’s communication. As yet, networking and shared resources are currently only available to families of affected children through the family advocacy charities. Data here suggested that a similar networking platform for sharing resources could be also beneficial for professionals.

The next subtheme highlights how professionals ventured beyond the school curriculum to address the needs of the child they supported. This involved aspects of care and medical tasks associated with the physical needs of the child; challenges with teaching a child in a mainstream education system; as well as creative ways in which professionals considered how their teaching could impact the child's future independence.

7.5.3. Beyond the Curriculum

Speaking about their role in supporting a child with Batten disease, one professional described carrying out multiple daily therapeutic interventions. They described the tick-box nature of their role:

> "…there's all these experts feeding me all this information and sometimes like, physio, tick! Music, tick! And you're trying…and we do a gardening club with them now, Tick! It becomes like, you just want to make sure that everything's right for them" (participant seven, LSA).

Shedding light on the issues of demands placed on professionals, comments here described the pressure of delivering different aspects of therapeutic exercises outside of their role as a learning support assistant. The professional here described how advice and activities from experts became a 'tick-box' exercise rather than perhaps a consideration of what was appropriate and right for the child in their care. Others echoed the demands of medical and physical care for the child, listing the different daily aspects of their support role:

> "Bring them into class, try and get them to engage with activities that we were doing, in class, registration, and then it was personal care …they were peg fed, so we did their feeds. Oral hygiene we tried to do daily, and we used to encourage drinks, and I did their music therapy with them, daily, and we used to play Harry Potter storybooks for them, music on the iPad…" (participant one, LSA).

Comments here again described how professionals' roles routinely involved caring, education and therapeutic tasks that were completed each school day. The activities and tasks described by the professional went beyond simply supporting the child in the classroom to access the curriculum and lessons like their peers. Medical interventions such as peg feeding, encouraging the child to drink, and managing their oral hygiene suggest that their role was more aligned with that of a personal carer. This professional also described activities tailored towards the child's wellbeing which involved listening to audiobooks or music, which again went beyond the typical classroom support assistant role.

Whilst reflecting on the challenges experienced by the child in school, one professional introduced the difficulties with the standard school curriculum. Describing how the child often lacked a sense of accomplishment and achievement at school, they talked about the impact of the standard school curriculum and assessment structure:

> “…we knew the academic side of things was becoming more and more difficult… them having a sense of fulfilment, experiencing success, and enjoyment, because I was very aware that their time in class was getting increasingly difficult for them… it was quite hard for them to access some of the…especially in year six when you’re getting more towards SATs and all of those kind of things. The English and maths focus becomes very big and we didn’t want to narrow their curriculum experiences, we wanted to keep them quite broad… it was, for our part, making sure that we were giving them opportunities for challenge and progress and attainment” (participant five, inclusion lead).

Although only relating to one school’s experience, comments here distinctly frame the challenge of the standard mainstream education, describing the difficulties associated with exams and curriculum teaching for a child with increasing needs. Data here described how this participant recognised the restrictive nature of the standard curriculum, which focused largely on topics that were inaccessible to the child and their level of academic skill. In response to this challenge, the professional alluded to bespoke approaches which adapted and broadened opportunities for the child so that they could still experience progress, achievement and continue to be challenged in the school environment.

Whilst describing their approaches to education, professionals described other ways in which their role consequently ventured beyond teaching the standard curriculum. Many of their experiences intentionally focused on teaching the child skills which would help preserve their independence in the future:

> “When they first started losing their sight, I think we painted some of the things different colours … so they’d help them to work out routes and maps and things like that…also lots of practice. They used to be on stairs, and things like that. We used to once a week, take them for a walk to the shops, so that they were able to navigate roads and crossings and feeling the bumps on the pavement and going into a shop and buying something” (participant five, inclusion lead).

> "…in the afternoons, [we would] do things that were going to be very valuable for them, so you know, cooking …doing the movement skills, doing the music therapy and continuing their Braille…thinking about what do they need right now, what can we help them with right now? And those life skills that they're going to need in the future" (participant five, inclusion lead).

Professionals widely described how they considered the child's future skills and ventured beyond the taught curriculum to achieve this. Data here emphasised the importance of repetition and practice for the child to understand skills such as mapping, orientation, and independent mobility. By preserving a child's skills and considering their future needs, it was demonstrated that professionals understood that the school environment needed to be more than curriculum teaching.

In venturing beyond the standard curriculum, many reemphasised the importance of teaching such life skills and independence activities pre-emptively and early on in a child disease journey, so that they could be maintained and recalled later in the child's life, and future-proof their independence. The Music Therapist stressed the importance of familiarity: "…how the familiarity…over time, could have an impact on them sustaining those skills" (participant two, Music Therapist). Actively revisiting and recording memories and experiences in memory books was also described:

> "Recording their voice into a book…so we try to capture as many memories and for their family as we could. And we used to have sessions every week where we played back the book and talked about memories that are important" (participant five, inclusion lead).

Creative methods, such as memory books, were used in this instance to help the child maintain and connect with memories and experiences, which professionals talked about using regularly. The memory books seemed to consider the child's family as well. Recording the child's voice into the book seemed to be a way that professionals could capture the child's identity and voice for the family, perhaps preserving memories of the child for their family in the future. Again, the strategies employed here demonstrate the unique way in which professionals ventured outside of the curriculum to address the needs of the child, their family and their future skills.

Going beyond education, data here illustrated how professionals recognised the limitations of mainstream education, and the need to actively encourage the preservation of independent skills which could support the child's future; ultimately considering what is needed now, what they are still able to do, and what they will need in the future. Exploring the concept of a holistic approach further, many interviewees described a flexible approach where they creatively adapted the curriculum to help the child engage and learn in an accessible way. This is outlined in the last subtheme of this section.

7.5.4. A Personalised Approach

There were many instances where professionals described how they developed approaches or strategies to offer an adapted and personalised approach to the child’s education. One professional described how the child was “…more or less taken off timetable” (participant six, VI specialist) suggesting that activities in the school day were bespoke to that child. Others agreed with the notion of flexibility, commenting that “sometimes you have to go with the flow…it's all about, you know, compromise” (participant three, VI specialist). This perhaps described the nature by which each child could present differently daily, and therefore professionals had to respond flexibly, and compromise on curriculum teaching.

Highlighting the importance of flexibility, all professionals talked in detail about how they adapted the curriculum to suit the child they were supporting. In lessons, the professionals talked about how they adapted the approach to learning whilst still retaining the topic or theme:

> “…while the other kids were writing biographies of the different astronauts, we were making rockets out of playdough and paper aeroplanes and seeing how far they went, trying to incorporate all the different things and fun things into their day” (participant three, VI specialist).
>
> “…everything would just be very much very heavily differentiated for them. And in literacy we'd often Braille things for them, that would be the same as the other students, but then we would tailor it to them” (participant four, VI specialist).

A common process was described by VI specialists here, where lesson topics were adapted to involve sensory, physical, or braille-based activities to be more accessible for the child. Comments here suggest that professionals drew upon creative adaptations to address the additional needs of the child, whilst attempting to maintain a connection to the lesson and the wider curriculum. It is perhaps because of their visual impairment that sensory and tactile activities were more widely accessible and enjoyable for children affected by Batten disease. In the absence of recognised standardised educational guidance for children with Batten disease, it could be suggested here, that sensory, tactile, arts-based activities could begin to offer children a way of accessing the curriculum and maintaining a connection with the lesson alongside their peers.

One professional talked about how the school “…basically just did a personalised curriculum and focused on the things that we felt were important for them” (participant five, inclusion lead). Linking personalisation with the child’s future needs, comments supported previous narratives about the importance of fostering wellbeing, life skills and independence beyond that which the curriculum could offer. All professionals described the importance of adapting lessons to suit the needs of the child; be it through sensory activities, creative crafts, or using Braille, they all described how they ventured away from the class lesson whilst keeping a connection to the overall topic as much as possible.

7.5.5. Music Speaks Program

When commenting on their experience of the Music Speaks program, professionals observed how the Music Speaks program supported this personalised educational approach, and that there was a sense of ownership and personal recognition:

> “I thought it was a really, really brilliant programme to be part of. I think they loved the personalisation of it. And they loved that it was all about them, that was really important to them” (participant four, VI specialist).

> “…there was enjoyment… there was routine…I think they enjoyed that the songs were personalised and about them, and it was their special time to sing” (participant five, inclusion lead).

Another participant described how the child would say “…it's my programme, it’s all about me” (participant four, VI specialist). The evidence here suggested that the program brought a sense of recognition and value through the fact that something special and bespoke had been created just for that child. This is perhaps in contrast to the standard lessons or activities in school which, according to previous comments made about the challenges of the mainstream education, limited opportunities for recognition, progress or achievement. The bespoke personalised elements of the program, which included the child’s family members, favourite activities, and food and drink preferences, seemed important to both the child and professional. In turn, this facilitated engagement and enjoyment for the child as they were given something that was exclusively for them and presented them with opportunities to engage in something meaningful to them.

Similarly, professionals talked about the importance of the program contributing to a wider personalised ‘package’ for the child:

> “…we just wanted something very personal for them, in their last year at school and this kind of fitting into that perfectly so we knew they had some music [Music Speaks program], they had some cooking, they had some movement, they had their time for them to read to younger children and this all became a lovely little afternoon package for them, which was important for us because we wanted their last year at primary school to be meaningful for them” (participant five, inclusion lead).

A personalised wellbeing package of favourite activities such as music, cooking, movement and reading were intentionally curated for this child. Observing the value in creating a school experience that was meaningful for the child, data here imply that wellbeing and independence activities were more valued by professionals than the standard curriculum in preparation for secondary school. Again, data support the notion that professionals considered their role to go beyond education for the child, creating practical and enjoyable activities for the child in their school environment.

Lastly, one professional described the importance of how the program enabled the child to have a sense of achievement, pride and ownership:

> “They remembered the words so that gave them a sense of, you know well-being, that gave them a sense of accomplishment…they'd be proud to tell them, so there was a sense of pride, a sense of ‘look what I’m doing’, a sense of ‘this is for me’” (participant seven, LSA).

Describing a sense of accomplishment and progress, according to professionals, the child here clearly felt a sense of pride and satisfaction, in remembering the program content and wanting to share their accomplishments openly with others. The program contributed to a sense of achievement and progress, which professionals perhaps identified as lacking in the mainstream education system for the child.

Subthemes here described a fine balance in the professionals’ approach to education for a child affected by JNCL. The importance of consistency, familiarity, and routine were emphasised, as well as frustrations around lack of resources or inconsistent support systems in the school environment, and how skill-sharing could support this. Professionals described their consideration of flexibility, adaptation and a personalised, bespoke approach for each child, also referring to how the Music Speaks program supported this consistent yet personalised approach. Narratives from professionals described approaches to education that were unique to the individual and their needs, and how they felt that Music Speaks could facilitate a sense of accomplishment, achievement, and progress for the child. Overall, the conversations here emphasised the importance of a bespoke approach to the child’s education, stressing the challenges associated with mainstream academic objectives, and a universal, one-size-fits-all model. The next theme, ‘Exclusion versus Inclusion’, similarly explores the many different educational approaches in school, commenting on the diverse ways in which children were either included and integrated into the standard curriculum or intentionally separated.

7.6. Exclusion versus Inclusion

Whilst discussing strategies and approaches to education, professionals described the dichotomy of inclusion and exclusion, and how they addressed aspects of this spectrum in different ways. In relation to topics such as developmental regression and social isolation, professionals observed ways in which the child became distanced from their peers. Likewise, professionals discussed the importance of dedicated separate spaces, recognising the benefits of separating the child from the classroom. The concept of inclusion was explored and how activities such as music could provide environments that integrated the child with their peers naturally. The first subtheme explores the concept of the developmental, emotional and social gap between the child and their peers.

7.6.1. Gap Widening with Peers

When professionals were asked about any challenges the child faced in their daily school lives, interviewees reflected on the emotional and developmental gap that they saw emerging between the child and their peers. One professional observed how “…the impact of having Battens was becoming more and more noticeable at school and the gap in learning was growing” (participant five, inclusion lead). Professionals talked about how they overcame the developmental gap with adaptations to the curriculum or personalised approaches (explored in the ‘Holistic Approach’ theme), but some highlighted the social challenges associated with developmental regression:

> “…some of the girls were starting to talk about music, like popular music, obviously, [the child] still wants to watch CBeebies…they loved it if I read Cinderella to them, if I read any of the nursery rhymes, they just loved it. But obviously year six, I mean some of the girls turned 11 in September/ October …that gap is getting very, very wide” (participant seven, LSA).

The professional here described the regressed nature of the child’s interests, which centred around younger-aged stories, characters or programs. In contrast to the interests of their peers, according to professionals, this regression distanced them from children of the same age. This could, in part, be due to more recent memories or interests deteriorating first, leaving earlier interests or preferences intact. It could also imply that with cognitive decline, simpler ‘younger’ stories and programs were more easily accessed and understood by the child as their disease progressed. Others concurred with this social and developmental gap between the child and their peers:

> “I think they have always had a love of music and dancing and singing … obviously their journey with the deterioration of Battens has had a significant impact on them - I think their emotional wellbeing and a sense of belonging and fitting in and being like their peers and their friends. And I think having this focus of the music therapy [Music Speaks] was something new and very personal to them” (participant five, inclusion lead).

A passion for music, singing and dancing was described here as negatively impacted by the child’s deteriorating condition. Professionals recognised the negative effects of children lacking a feeling of inclusion or connection with their peers, which in turn impacted on the child’s emotional wellbeing. However, in contrast to concepts of deterioration and exclusion, comments here suggest that the music therapy (or the Music Speaks program) brought something bespoke for the child, which had a positive impact on their school experience.

Other professionals also reflected on the gap widening between the child and their peers and a lacking confidence to initiate interaction with other children. One child would “…say hello, if they’re spoken to but they won't start a conversation” (participant three, VI specialist) and another “…didn't tend to approach the children for conversations, but they would engage in them if it was something of interest to them” (participant four, VI specialist). Initiating interaction and self-expression were seemingly challenging for children unless others spoke to them first, further demonstrating the sense of social isolation that children may have experienced in school environments with their peers.

In response to the developmental gap between the child and their peers, professionals recognised the need for alternative approaches to their educational environment. One professional described their conflict with separating the child from their peers, commenting that “I don't want them to leave the classroom because they’re part of that class and they’ve known those children a lot of years, and I don't want them to feel that they’re being taken out all the time” (participant seven, LSA). Professionals understood the value of integrating the child with their peers, and the need to support a sense of inclusion for the child. Others recognised the limitations of excluding the child from the classroom and how this impacted the relationship with their classroom teacher:

> “If they’re with people like me, they will, they’ll tell me and the other TA but they don’t really communicate that well with their teacher because they’re not in not in the classroom that often with them” (participant three, VI specialist).

The consequences of being taken out of the classroom seemingly had a negative impact on the child's relationship with their teacher. The unique relationship with support staff is also alluded to here, where the child only had the confidence to express themselves around familiar members of staff (supporting the previous theme of Consistency). For this child, the classroom teacher became someone who was unfamiliar to them, and consequently, someone they struggled to interact with.

In contrast, all professionals recognised the importance of having a dedicated space for the child away from the classroom, and the positive impact this had on their focus, learning and wellbeing. Professionals described the intimacy of the dedicated space:

> "…we had all the lights down. And we'd just do relaxing kid's stories, we did a little bit of peer massage, which they really loved, and it was just like a quiet time. We did sensory play… things like shaving foam with hands and feet in different things" (participant five, inclusion lead).

A dedicated space away from the classroom appeared to facilitate calm and relaxation, offering a space to foster wellbeing through sensory and peer activities. Other professionals described using the space when the child had "had enough" (participant three VI specialist), putting music on in those situations to facilitate calm, relaxation and recovery for the child. Whilst all professionals talked about the unique importance of the dedicated space, it is assumed that not all children or schools had access to rooms such as these. The experiences described here are perhaps unique to these schools, and possibly would not be replicable in every school supporting a child with Batten disease.

The subthemes here relating to the developmental gap and a dedicated space, depict one side of the exclusion/ inclusion spectrum. With an observed gap widening with peers and the importance of a dedicated 'safe' space, professionals described instances where schools recognised that the child's environmental and educational needs were distinct from their peers. Strategies were also widely recognised as being unique to each child and their school, and therefore may not be a universal experience for all affected children who attend mainstream settings. On the opposite end of the spectrum, professionals reflected on the need for inclusion and connection for children with their class and peers. The next subtheme focuses on this concept exploring the idea of intentional facilitated peer-to-peer interaction.

7.6.2. Facilitating Peer Interactions

All professionals described how they facilitated peer activities for the child both with their peers and with younger children in the school, recognising the challenges of exclusion. Drawing upon the child's interest in stories, acting and drama, professionals described how the child enjoyed interacting with younger children in school. One professional described this experience:

> "We did lots of music and movement with younger children, which they absolutely loved doing. They loved drama…they played with dolls' houses, and they talked. And again, there was a lot of younger children, and [other children] would talk to them, and they would talk to [other children]" (participant six, VI specialist).

Comments here outlined how the school, by facilitating sessions with younger children, embraced the concept of regressed developmental interests and needs of the child. Focusing on interaction, data here described how conversations were more easily achieved with younger children, perhaps because of the simplified language and communication involved. Creatively addressing peer interaction, this school clearly understood how they could support the child to feel included and connected with others in the school through arts-based activities with younger children.

Others described strategies to connect the child with same-aged peers through specific lunchtime groups, which aimed at developing peer-to-peer relationships and connections:

> "I think they sometimes felt a little bit isolated. It's still important that they had conversations and relationships with their peers. So, the lunchtime bit was where we could invite two different friends every day on a rota. They'd come and do braille … play dough, … Lego, so that [the child] could just have some time with their friends. They told us at the end of year six that the support at lunchtime with their friends had really made a difference, because I think they felt happier, that they were more included" (participant five, inclusion lead).

Comments here emphasised the value of spending time with the child's peers outside of classroom activities at lunchtime, recognising the importance of peer integration. With a focus on activities that were accessible to both the child and their peers, according to professionals, interventions seemed to help reduce isolation and facilitate interaction and connection with friends, which positively impacted the child's sense of inclusion and wellbeing. All professionals recognised the need for the child to feel included and integrated with their peers or with other children in the school who were younger or with those who had similar additional needs. It was implied that, without this creative support from the school, affected children may experience isolation and loneliness as the gap between their peers widened. There were also specific instances where professionals reflected on the benefits of music creating a naturally inclusive, shared experience, which is discussed further below.

7.6.3. The Inclusive Experience of Music

Professionals commented on their impression of the inclusive experience that music could bring, describing how activities like musical assemblies, could facilitate an inclusive experience for the child:

> “…we had singing assembly on a Tuesday, they loved that… because we've done the same songs, like you do, in school over time, they felt comfortable, they knew them, they could join, they felt part of a school. They just wanted to feel, like everybody does, part of the school” (participant five, inclusion lead).

Musical environments in the school such as singing together seemed to help the child to feel included with their peers, and professionals all spoke about the positive impact of shared musical experiences with the child. Others described the inclusive effect of shared music-making in class instrumental lessons:

> “I’m sitting there reading the music and I’ve got my hand on theirs and we’re playing it together… They’d sat with the crowd. You know, one little boy gets the cello for them, another little boy helps them with a chair, they have their own special cushion” (participant seven, LSA).

Music lessons seemed to have the same positive impact by naturally facilitating an environment where the child could connect with their peers without the need for forced or facilitated peer interaction time. Comments suggest that professionals recognised how playing music could support connections and relationships with their peers, where other children also adopted a supporting role for the affected child as well. From a social aspect, others commented on the benefits of listening to music with peers in school, and favourite music pieces were “…a talking point… that was something that they’re able to share with their peers and have a chat about and that was really, really important to them” (participant five, inclusion lead). Musical interests were described as a socially binding activity which could allow the child to connect with peers on the same level, despite the developmental gap between them and their peers.

7.6.4. Music Speaks Program

Professionals reflected on the inclusive shared experience of the Music Speaks program:

> “…brings you together and… we used to sit and talk…yeah, it was fun. We had fun with it…they’d try and sing some of the other days of the week, when I was working with them, so we listened to those as well? So, we didn't just listen to Mondays and Tuesdays, we listened to all of them really” (participant three, VI specialist).

The program was seemingly enjoyable for professionals, and in many cases, provided an opportunity to share positive moments and dedicated time together. It seemed that the inclusive aspects of music were also present in Music Speaks program. Considering the previous comments relating to the demands associated with caring and supporting the affected child, it is possible that the program session offered an opportunity to enjoy something pleasurable with the child.

Observations here illustrate the inclusive environment that music created for children in school – be it instrumental lessons, assembly times, sharing music with peers or enjoying the Music Speaks program together. Although professionals emphasised the challenges associated with the developmental and emotional gap widening between the child and their peers, many also described several situations where the child could feel integrated and included. It was implied that, without this facilitated peer time, children may have experienced isolation and distance from their friends and peers. From a professional perspective, alongside interventions that specifically facilitated peer interactions, musical environments facilitated a shared inclusive experience for the child. Much like the balance of educational approaches in the ‘Holistic Approach’ theme, the concept of inclusion and integration appeared also to be about balance. Many commented that it was important to facilitate peer activities to help the child interact, whilst also recognising the importance of a safe, dedicated space where the child could retreat to for relaxation and calm.

The next theme relates to the experiences of the Music Speaks program in response to the second research question. Whilst reflecting upon their experiences of the Music Speaks program trial, professionals commented on considerations, adaptions, and potential applications of the program for other children with Batten disease in school settings. The final theme, ‘Program Considerations and Applications’, explores this concept in more detail.

7.7. Program Considerations and Applications

Towards the end of each interview, the professionals were asked about how, in their experience, the Music Speaks program could be improved, adapted, or applied in other contexts or with other children. Professionals described their own experiences of running the program as well as mechanisms that they thought helped support them in their roles.

7.7.1. Staff Experiences of the Program

There were some instances where staff referred to their feelings, responses and personal experiences of supporting a child with Batten disease as well as being involved in the research project. Professionals also reflected on how they felt they were best supported in their roles, and in doing so, participants touched on feelings of empowerment, confidence, value, and recognition. In response to questions relating to challenges of their role, one professional talked about how encouragement and recognition helped them in their role of supporting the affected child:

> “…that helped a lot just, for them to say to me ‘you've done really well today’, and just that encouragement … Yeah, it was staff that got me along really and Mum. Well parents, they were very encouraging, and they used to write nice letters in a diary” (participant one, LSA).

A sense of recognition was alluded to here, with the professional indicating that it was helpful when other professionals acknowledged the pressure and challenge of supporting a child with Batten disease. Perhaps easing the pressure experienced in their role, comments described how recognition and support from parents were also important in helping professionals feel acknowledged, valued and recognised.

The Music Speaks program also seemed to support these feelings of value, and empowerment for the professionals involved. The Music Therapist reflected upon the observed impact for their colleague (the LSA) delivering the program:

> “…I think they coped really well with it. I think it gave them actually, a focus. And from me observing them doing it, I mean they seemed really organised doing it. How they spoke about kind of the routine of doing it with [the child], sounded positive you know? Them having that role, it seemed quite empowering for them” (participant two, Music Therapist).

It seemed that the LSA delivering the program here spoke positively with their colleague about organising and running each session of the program. The defined and purposeful role of participating in and delivering the program seemed to have a positive impact for the LSA. Comments here perhaps allude to the way in which a natural support system had emerged between these two professionals. There is evidence to suggest that having the role of delivering the Music Speaks program gave professionals a sense of purpose, focus, and routine, which led to them feeling empowered in their role. This perhaps contrasted previously mentioned feelings around anxiety and pressure with regards to the ‘tick-box’ and multidimensional pressures of a professionals’ supporting role.

Other professionals described how Music Speaks encouraged them to carry over music into lessons, activities or other areas of the child’s day. One professional described how:

> “I do think that it [Music Speaks] has helped them enjoy things, enjoy lessons, enjoy school a bit more, because we can incorporate music into all sorts of different topics and different subjects…we did listen to music beforehand, I didn't realise the impact that it would have on them (participant three, VI specialist).
>
> “I do think the little, even the short time and the few sessions that we had, I do think just by watching them and knowing the metronome and just the beats, obviously, I could see a difference, and therefore I used it further on” (participant six, VI specialist).

Professionals described their understanding of how music could benefit the child they supported in school. Participant three explained how they already used music before the research project, but the experience of Music Speaks helped them to consider how music could be incorporated into lessons and topics which could ultimately lead to a greater school experience for the child. Likewise participant six described how the program increased their understanding of how pacing and rhythm could impact the child’s speech. They described how this encouraged them, as a professional, to incorporate these elements of the program beyond the program trial.

Support from the researcher seemed to be instrumental in enabling the professionals to feel supported and confident in delivering the program:

> “…your meetings with us at school were really valuable as well, because we felt we could really talk to you about them, and you met them, which is lovely…I think without you, we couldn’t have done it” (participant five, inclusion lead).

> “We wouldn't have been able to do it if you hadn’t come up…I was much more comfortable actually having you there, and then we could talk through everything…We thought we were doing it, you know, for you” (participant six, VI specialist).

Comments from participant five suggest that alongside the training and meetings which were practically supportive, a sense of connection and relationship with me (as the researcher) was also important; professionals could talk about the child they supported and recognise the value of me meeting the child in person as well. The latter comments explained how they would not have been able to deliver the program without the support from me, perhaps as they would have lacked the confidence and the opportunity to run through everything, and my practical support was seemingly beneficial. Data here suggested that the connection to me as the researcher not only provided practical support but also gave them a sense of accountability; that they were delivering the program for me, rather than just for the child or for themselves. My support was seemingly important in improving their experience of taking part in the program.

Reflecting on their wellbeing, this first subtheme touched upon mechanisms that supported professionals to fulfil their role, and how Music Speaks could build confidence, empowerment, and an understanding of how they could use music in educational approaches. Professionals generally explained that with the guidance, training and ongoing support, they felt they could deliver the Music Speaks program confidently and consistently, implying that program delivery was not achievable otherwise. Having a connection to the researcher and knowing that they were always available to help if they needed, appeared to help them to feel supported and equipped. Perhaps for future application of the program, therefore, support, training, guidance, and researcher involvement are important considerations.

7.7.2. Adaptations to the Program

When considering the future application of the program, professionals made suggestions around potential adaptations to the content (aside from the existing bespoke content previously discussed in section 7.5.5). Firstly, professionals discussed how children learned the program content quickly, but how this also proved to be a benefit and a challenge. One professional reflected the positive impact of the program on the child’s recall:

> “I asked them just before I left, and I said ‘Can you remember all the songs that we used to do?’ I said, ‘What was Monday?’ And they could remember, they actually could remember what songs were for each day…which surprised me because I thought because it was quite a new thing in their mind. So obviously listening to the music, it definitely stayed in their mind (participant six, VI specialist).

Comments here described how the memory for the program songs surpassed the professional’s expectations of what the child would be able to learn and recall. A sense of learning is described where, contrary to the professional’s expectations, new information could be recalled by the child even though it had been taught recently. Professionals went on to describe how this over familiarity led to adaptations:

> “…we found that the songs became repetitive for them and that was…there was two sides to it. They remembered them really, really quickly, which was fantastic. But they also got bored of them as well and wanted to change the words” (participant seven, LSA).

The repetition of the program appeared to have both a negative and positive impact; for this child, learning and memory recall was speedy, yet the professional suggested that the repetition of the program material became too familiar and boring for them. Data here also aligned with recording sheet comments from Phase Two, where professionals described how children adapted the program after they became familiar with the content. This suggests that for children who were cognitively more able (as with this participant) the program needed more variation, flexibility and perhaps adaptation options.

Developing this concept further, when asked about how the program could be improved, professionals suggested more variation and flexibility. One professional mentioned the idea of program variation:

> “I think maybe a little bit more…variation? I think, that's only from my experience with them. As I said they got ‘Oh here we go again’ you know?…but they loved it. The first few weeks they were really enjoying it. They knew all the words. Um… but yea a little bit more variation” (participant seven, LSA).

The concept of adapting the program indicates that it should not be a static, repetitive resource for children, but something which requires adaption and development as each child progress through the tasks.

In some cases, professionals described how children were unresponsive to certain songs in the program (the negative feelings song), and how they adapted to this.

For one child, this was related to the slow tempo of the song where "…the music wasn't quite upbeat enough for them, it were quite slow, and there were a lot of instrumental parts in it…they then lost concentration" (participant one, LSA). In other cases, it was related to feelings of singing or practising negative phrases when the child felt contrasting positive emotions:

> "…they didn't like the sad one, because it … made them feel sad, when even though they were happy, they were listening to sad song. Instead of saying sad, they would go, 'No, I'm happy. I'm happy'. They'd always change the words" (participant three, VI specialist).

According to this professional, the prescribed format of the program (i.e. singing about sad feelings on a Tuesday) perhaps did not fit with the reality of the child's complex and variable emotions. It could be implied that when children are feeling positive, they prefer to engage with upbeat positive music. Again, the theme of adaptation (see section 7.5.2) is alluded to here, where the child felt confident changing the words of the song to suit their needs. Other professionals described how a sense of control over the program positively impacted the child: "…it was quite important for their control, and having things as they wanted them, and as they knew them. So, to them, that was their song" (participant four, VI specialist). It seemed that the program offered an activity where the child could have ownership in creating songs in the format that they wanted.

Control was also mentioned in relation to me and the research, where one professional described how the child felt a sense of ownership over the recorded materials that were being sent to me as the researcher:

> "…they'd say your name, 'Is Rebecca coming back?' Or 'is this for Rebecca?' 'Yes, it is. Yeah, we're sending this off'… and sometimes when we were choosing which video to share with you, they'd go 'not that one!' So, they were involved, they knew that you'd be seeing what they were doing…that was really important for them" (participant five, inclusion lead).

The child seemingly felt actively part of the project and had a sense of co-creating the material for analysis. Although professionals described how the child helped to select the videos, all-but-one of the submitted videos for this participant were as per the pre-defined filming schedule. This perhaps implies that this was a strategy employed by professionals to help the child feel involved and in control of their participation in the project. Relating to the theme of preserved skills and interests (see section 7.4.2), comments here also align with the sentiment that people and experiences were memorable for the child, and a sense of connection to people was important.

Suggesting adaptations to the program, professionals emphasised that a more flexible, co-created approach could be more suitable, as opposed to a set listening format of songs and phrases. For this reason, others commented on how they felt that the program could be most successfully delivered on a one-to-one basis with the child rather than in a group classroom format, for example. One professional mentioned how “I don't think you'd be able to deliver the programme in the classroom with other students” (participant one, LSA), suggesting that the program was best delivered in a dedicated space away from the classroom environment. Others described how they utilised the dedicated space (see section 7.6.1) to deliver the program:

> “You just need … A room to deliver it in? Coz not every single school has got, accommodating rooms that we could just go into? But I think there should be … provision made for something like this” (participant six, VI specialist).

Recognising variable school resources, data here suggested that, as there is no universal provision of space for children affected by Batten disease (or for others with additional needs), each child’s experience may be unique to them and their school. This is a consideration for future implementation of the program which may have an impact on how others can deliver the program in school environments in the future.

7.7.3. Variable Engagement

Professionals reflected that, in some cases, children refused to engage or express themselves in response to the Music Speaks program. Describing factors that impacted their engagement, they commented that sometimes the child “…would come in and they'd be like, ‘No, no, no, no, no’” (participant two, Music Therapist). All professionals commented that they felt this was in keeping with their general engagement in school, which was largely dependent on wellbeing, mood, and other factors such as fatigue. Others agreed that “…there were days where they weren't really interested, and there were days where they loved it, but that’s their condition I think overall” (participant seven, LSA). Similarly, another child’s engagement “…was dependent upon mood as to how they felt about doing things sometimes that they didn't always want to do” (participant five, inclusion lead). Mood, fatigue, engagement, and wellbeing were all described here as factors that impacted the child’s willingness or ability to engage with the program. In line with findings from Phase Two, this variable engagement was also consistently observed when children were taking part in the Music Speaks program, and therefore, is a key consideration in the future implementation of the program.

Professionals also expressed how engagement with the program was impacted by the fact that the child knew they were being filmed. One professional observed how "…the recording bit was a barrier for them, and that maybe inhibited them from joining in when we know they could have done" (participant five, inclusion lead). According to professionals, children felt less confident with the presence of the recording equipment or the knowledge that they were being filmed. Data suggest that perhaps filming children in activities could impact their confidence levels and their ability to join in or express themselves. More generally, evidence could suggest that filming children carrying out activities in school could impact their ability to engage confidently in the task or activity.

7.7.4. Accessibility

Whilst describing the program, professionals reflected on the accessibility of the program and the ability to deliver it as a non-musician. One professional described the experience of delivering the program:

> "…I don't think you need any kind of music abilities to be able to deliver that at all…I was a bit nervous to start with, because it's not something I've ever been involved in…But I was excited as well I think, that this is going to have such a positive impact, I'm going to be part of that. And I was really quite honoured to be part of it" (participant three, VI specialist).

Comments here reflected an apprehension from the professional about taking part in the program, recognising nervousness at the prospect of something new and not having musical abilities. However, the idea of creating a positive impact for the child highlighted anticipation and excitement for positive outcomes. The mention of honour also implied that this professional held the research in high regard and felt privileged to be involved, describing a sense of taking part in something which could, as a first-of-its kind project, could be impactful for the child.

Others reflected on the importance of accessibility in helping them deliver the program with the child explaining that the program was "…very simple for you to follow, and everything's pre-recorded, it's very structured and you just go through the process, and it doesn't take too long" (participant five, inclusion lead). Aspects of accessibility and a low demand program seemed important factors in helping the professionals deliver the program, which are key considerations for future program implementation.

7.7.5. Transferability

When asked about whether they used any parts of the program at other points in the day, professionals described how they introduced elements into their general communication with the child, suggesting the program could be transferred to other contexts. One professional believed that the key words and phrases in the program helped the child’s communication:

> “…key words, that's the positive, getting the key words out, and letting them say them, remembering them…’Drink’, ‘Water’. Just simple words, ‘Food’. I mean, we’d say ‘Crisps’? ‘Yes or no? Really key words, with them towards the end” (participant one, LSA).

Comments here suggests that professionals transferred elements of the program into other situations in the school day, describing how they recognised the value of using key words with the child to support elements of communication and how they carried this through to everyday situations like mealtimes. Data here also indicated how the program words and phrases helped professionals in their communication approaches with the child, using key words and repeated sentences to simplify their conversation. This evidence suggested that the Music Speaks program could go some way to reduce the communication challenges experienced by both the affected child and professionals. It is possible that elements of the program could be transferred to other areas of the child’s life to anchor essential communication with simple, key words to more widely help guide their communication with others.

Professionals suggested how other children with Batten disease or other difficulties could also benefit from the program in the future. One professional mentioned that “…it would be really useful for partial sight loss, and also I think it would be really, really useful, if someone was struggling with a stutter or speech” (participant seven, LSA) and another commented that “I think the other children that have difficulties with language, it would help them to be able to express their needs” (participant five, inclusion lead). Widening its application to more general difficulties with sight loss or speech here, professionals suggested that elements of the program could be applicable to children with other educational challenges. This supports the concept that the program had an element of transferability, where it could be delivered outside of the Batten disease population, with other children in other contexts.

7.7.6. Early Implementation

Lastly, all professionals believed that for the program to be successful for children with Batten disease, it relied on early implementation. One professional commented that “I think it's got to be done quite early on…so, it really is embedded in the early stages. So, it's really in their minds” (participant six, VI specialist). Others similarly echoed the importance of early implementation:

> “I wish we’d have been able to do it maybe earlier with them…maybe if we’d have had something like this in school earlier, it may have helped them in those moments when they particularly struggled, so before we knew they had Batten’s maybe” (participant five, inclusion lead).

Aligning with previous subthemes concerning pre-emptive teaching (see section 7.5.3), comments here suggest that programs like Music Speaks may have more success if implemented before the point of diagnosis when the child was beginning to experiences challenges with their communication. Others emphasised the value of early implementation describing their impression of how the program could support the child in the future:

> “…when they’ve left school and those songs, and those phrases and the meaning of those to them, they might recall those, in a year or so’s time, when they’ve lost all other kind of language or communication and they might be able to recall part of that song or a certain word, or phrase” (participant two, Music Therapist).

> “…giving them that independence in the future, if they need it, to be able to tap into those important phrases that they've learned through the songs…these are things, these are so important for the children. Needing to be able to communicate if they feel pain, and if they're hungry and need a drink” (participant four, VI specialist).

Comments here suggest that professionals believed that musical information (such as program phrases) could be recalled even when the child has lost the ability to communicate their needs. Demonstrating a consideration of the child’s future, professionals also stressed the importance of equipping a child now for their future, and how the program could feed into this learning, by embedding skills in the child’s memory to future-proof their communication. There is evidence here to suggest that professionals understood how meaningful teaching, based on the child’s essential needs in the future was an important aspect of education for the child. This concept of early implementation could have an impact on when the Music Speaks program could have the most benefit, such as in the early stages following an initial diagnosis, rather than in the later stages of the disease’s progression. This is discussed further in the concluding chapters; see section 8.7.2.

Considering adaptations to the program, professionals identified potential limitations of such a repetitive, restrictive program (with regards to specific, set songs each day), implying that a more agile program could better fit the changing needs of the child. Professionals observed variable engagement and a lack of confidence when children knew they were being recorded, which could have implications on expectations of engagement with the program in the future.

Experiences suggested that the accessibility of the Music Speaks program was important for non-musical professionals. Data, relating to transferability, suggested that the program was used in other situations to support communication for the child, and Music Speaks had the potential to support other children with communication difficulties or additional needs. Lastly, the concept of early implementation further emphasised the theory that pre-emptive approaches to teaching could help support the child from the early stages of diagnosis.

7.8. Overview

Describing the many considerations, challenges, and experiences of supporting an affected child in an educational context, professionals have illuminated the complex and rapidly changing landscape for children affected by JNCL. Whilst the interviews related to just five children in five mainstream schools and special education facilities in the UK, the narratives and experiences of these professionals offered valuable insight into the challenges they (and the child they support) faced in their daily school life.

Experiences described how professionals go above and beyond their educational remit to improve the wellbeing and quality of life for affected children. There was a distinct sense that above all else, the focus of their support should be around enjoyment, relaxation and creating positive experiences. Narratives described a deep connection to, and understanding of, the child they support, knowing how and when a child needed adaptations. Professionals also described the fondness and appreciation that children in their care have for music, and how they utilised this medium in many aspects of their support role. Others reflected on the positive impact that music had on preserving functional skills, sometimes developing skills for the child who was experiencing unpredictable change and deterioration.

One overriding narrative illuminated the experience of striking the right balance; how each professional had their approach or adapted strategy to support their affected child. Whilst professionals widely created adaptations in the child's curriculum, teaching life skills or implementing activities to integrate the child with their peers, all agreed that it was about a balanced approach. Finding ways for the child to have a safe space for relaxation, calm and a sense of wellbeing, for example, was widely acknowledged by all. Narratives described how approaches were unique to the individual and their supporting professional, emphasising that there was no 'one-size-fits-all' approach to their education.

Experiences described how the professionals felt about their role in supporting a child affected by Batten disease. Sadness and empathy were shared in response to them witnessing the child's deterioration, alongside a sense of pressure and fear in relation to the demands placed on the professionals in their roles. Feelings of frustration were revealed, relating to under-resourced systems and processes, and others around them who lacked knowledge of Batten disease. Narratives described how peer and family support, and researcher encouragement, gave professionals a sense of recognition and value in their role supporting the child in an educational environment.

Interviews described how the Music Speaks program could enhance elements of communication, such as pacing, memory and self-expression for the child. Describing the positive benefits for the child, comments also emphasised how Music Speaks supported elements of personalisation, consistency, accomplishment and ownership in the child’s school experience. Professionals were empowered and honoured to be delivering the program with the child, and in some cases, the program seemingly gave staff the confidence and understanding to introduce music into their educational approaches. Data evidenced the need for a flexible approach to the program that meets the changing needs of the child, and the importance of early implementation to help embed phrases and language in the child’s memory before deterioration begins.

Lastly, thinking beyond an educational role, narratives described a holistic consideration of the many needs that the child has, both now and in the future, and how their role contributed to this. There were instances where observations and narratives from professionals focused on different outlooks and priorities for the child in their care. VI specialists and learning support assistants, for example, focused first and foremost on the adaptations that they could make to the child’s existing education environment in the moment, describing specific strategies to help the child navigate the present environment they were in. The Music Therapist mentioned the transferability of the program into other areas of the child’s school day, commenting more on how they envisioned the program benefitting other functional areas of the child’s life. In contrast, the inclusion lead focused primarily on the child’s future, describing strategies and approaches to help the child develop and maintain independence. Despite differing priorities, all professionals emphasised the value in teaching life skills, pre-empting the child’s decline, and thinking about how they could support the child to lead an independent life in the future. Many believed that elements of the Music Speaks program complemented this pre-emptive approach, explaining how key phrases and words from the program could help support the child to communicate independently and effectively with those around them. Further research could continue to pursue this enquiry, considering how an understanding of these multiple perspectives could impact the future development and implementation of programs such as Music Speaks for musical and non-musical professionals.

7.8.1. Learning Points

Findings from Phase Three are summarised in the following learning points:

3.1. What are the experiences of educators and therapists supporting a child with Juvenile Batten disease in an educational context?

- Professionals supporting a child with JNCL see their role as sitting outside of the standard curriculum, going above and beyond to make adaptations where needed.
- Professionals prioritise wellbeing and quality of life for the child in their care, describing how music supported this.
- Many described a unique connection, understanding and knowledge of the child they support.
- Professionals expressed sadness, empathy and fear witnessing the inevitability of a child’s deterioration.
- Professionals experienced aspects of the frustration associated with the pressure of daily demands, lacking resources and systems, and those around them who lacked a basic knowledge of Batten disease.
- Professionals described how children could be isolated and excluded in a mainstream school system, and noticed how music and other interventions could facilitate inclusivity.
- Many professionals appreciate, value, and use music in everyday activities for the child they support and recognise the wider value of music therapy.
- Professionals acknowledge the importance of familiarity but consistently adapt the curriculum, striking a bespoke balance for the child.
- Many recognise the importance of peer and family support, encouragement, and empowerment in delivering their role.
- Professionals consider the future, thinking about how their support can build independence.

3.2. What are the perceptions of the Music Speaks program from those using it in educational contexts?

- Professionals observed a positive impact of the program on the pacing of speech, memory, and self-expression for the child they supported.
- They felt that the program gave the child a sense of accomplishment, ownership, and recognition having something exclusively for them alone.
- Professionals felt privileged taking part in the trial, and some developed the confidence to introduce music into educational approaches.
- The program offered a shared inclusive musical experience for the child and professional.
- An adaptable, flexible approach to the program would address the changing needs of the child, better than a static, rigid program.

- Implementing the program earlier could offer the opportunity for the child to learn and embed key phrases, making use of the phrases as a communication tool earlier on.
- Diverse perspectives illustrate the distinct outlooks and priorities of professionals, impacting considerations for future implementation of the program with other professionals.

The following chapter combines the themes from the interview data here with findings from the previous phases of the research, to provide an overview of overarching concepts and ideas. It draws upon observations of children in music therapy (Phase One), responses to the Music Speaks program (Phase Two) and the core six themes expressed by professionals supporting children with the Music Speaks program (Phase Three) to integrate the data with the observable and unobservable experiences and outcomes. Grounding the findings in theory, chapter eight provides a synthesis of concepts, mechanisms, experiences and structures of how music and music therapy are experienced by children affected by JNCL, and the professionals who support them.

Chapter 8: Discussion

"One good thing about music, when it hits you, you feel no pain"
- Bob Marley

8.1. Introduction

This chapter synthesises the findings from each of the three research phases, responding to the research questions described in section 1.8, and grounding the discussion in theory, policy and systemic contexts. From the positioning of a critical realist (Danermark et al., 2017), this chapter discusses how the research observations and perspectives interact with causal mechanisms, theoretical research, and the wider implications on policy, education and care pathways for children and adults with other neurodegenerative conditions. The chapter considers how future research could incorporate these findings to further optimise music-based programs for the benefit of children with Batten disease. It concludes with a reflective passage interrogating my personal experiences, values, responses and encounters throughout the doctoral journey and how they have ultimately shaped this research project.

8.2. Approaches to Education and Therapy

There is currently limited guidance for families and educators supporting a child with Batten disease. What does exist focuses solely on symptom management, physical support or improving quality of life through palliative care solutions later in the child's life (NINDS, 2020). Many families have negative experiences and lack the adequate support from national health services at the early point of diagnosis at the beginning of their journey with Batten disease (Scambler, 2004). In addition, there is a lack of appropriate and generalisable assessment tools that can capture the nuanced differences of children who have different variants of the disease (Augustine et al., 2013; von Tetzchner et al., 2013).

Research and guidance in the adult dementia sector is more developed, with early theorists emphasising the importance of inclusive, holistic care for adults with dementia (Kitwood, 1997). The NICE guidelines for individuals with adult dementia make reference to the importance of complementary psychological support therapies, as well as training and support for carers (NICE, 2018a). Yet no such guidance or health policy exists for Batten disease. This subsection firstly considers how evidence from the research project regarding the diversity of children with Batten disease, diverse approaches to education, lack of support resources and knowledge, and professionals' wellbeing could partially fill this gap in education and healthcare guidance for children with Batten disease.

8.2.1. The Diversity of Children with Batten Disease

Firstly, the need for standardised educational and therapeutic guidelines is highlighted by the fact that children with Batten disease are a diverse and heterogeneous population. Data from all three phases consistently highlighted the diversity of children affected by Batten disease, and how the needs of children vary throughout the course of their education. Children in the early phases of the research differed in terms of their functionality, disease variant, disease severity, stage of onset, and speed of deterioration, which was reflected in undulating patterns of change in music therapy over time. Even in later phases, where participants had the same disease diagnosis, the child's presentation varied day-to-day and week-by-week. Professionals observed how engagement was impacted by factors such as mood, fatigue and anxiety levels, emphasising the variability and changing needs of the participant group. As with other rare disease research, the data support previous studies recognising the challenges of observing children with variable disease journeys, functionality, severity, stage of onset, and speed of deterioration (Augustine et al., 2013; von Tetzchner et al., 2013).

Findings from later phases additionally presented a complex picture of how a child with Batten disease is seemingly 'locked-in'. Interview data demonstrated how professionals described the difficulty with understanding the child's speech and their needs, which presented barriers in communicating and interacting with a child. Yet, observations from Phase Two indicated that receptive communication scores were generally higher for children than expressive communication scores, suggesting that children were able to understand and comprehend more than they could express conversationally. Findings support early research which stressed that children with Batten disease are more aware than their vegetative presentation suggests (Bills et al., 1998; B. Johnson & Jochum, 1996). Later research in the sector also demonstrates that children with JNCL were believed by parents and professionals to have a better understanding of language than they were conversationally able to express, suggesting that children may have more awareness or capability than previously thought (von Tetzchner et al., 2019).

Research into adult dementia has similarly identified common misconceptions about an individual's capacity, which could have an impact on the way in which carers interact with the individuals they support (Kitwood, 1997). Recognising the communication challenges for adults with dementia, national guidance recommends training for carers in this area, as to how interactions can be adapted to support the individual (NICE, 2018a). There is no such guidance in the education sector that recognises the diversity of children with Batten disease or makes suggestions for adapted communication approaches. Findings from this research, therefore, suggest that guidance for educators and therapists could help support communication strategies guiding professionals on how to respond to the diverse and changing needs of the child and their communication difficulties.

Assessment measures in the project additionally highlighted the heterogeneous nature of Batten disease and the diversity of each child. Although recent research has developed bespoke assessment scales for different variants of Batten disease (Wyrwich et al., 2018), these are currently applicable to just one type of Batten disease and require further evaluation and refinement. Evidence indicated that the CMTOM music therapy assessment measure could potentially capture functional responses and behaviours not identified on the standard clinical assessment of the Hamburg Rating scale (Kohlshutter et al., 1988). It was perhaps the nuanced and detailed nature of the CMTOM scale that meant that it was able to pick up smaller, more frequent changes in children, in contrast to clinical scales which are broader and administered less frequently. Findings complement other research studies, demonstrating the possibility that symptoms of dementia are tracked and monitored differently in music therapy assessments compared to standard clinical tests (Hsu et al., 2015). Offering the research sector other assessment measures such as the CMTOM could provide the opportunity for clinicians and professionals to capture more nuanced changes for affected children, capturing the diverse skills that are preserved in the presence of music. Findings from this doctoral research, could help professionals understand how affected children comprehend information that is presented to them, and reconsider how they as professionals can help children engage with the world around them. The research also supports the need for education and care guidance to involve diverse, music-based activities to support improved communication between children, their staff and their carers (further explored in section 8.3.3).

8.2.2. Diverse Approaches to Education and Care

Interviews with professionals highlighted the ways in which they ventured outside of the standard curriculum to address the diverse needs of the child, prioritising wellbeing, life skills and their future independence. Evidence suggests that there is no universality in caring for a child with Batten disease, and each approach is tailored and bespoke for each child. Accounts from professionals highlighted their ability to adapt educational approaches, creating bespoke, personalised and inclusive activities to suit the changing needs of the child in their care. Perspectives from professionals also highlighted the challenges with current school systems for children with Batten disease, describing moments in the school day where children appear to be emotionally, socially and developmentally distanced from their peers. Data also shed light on how systemic challenges with mainstream education could exclude a child with Batten disease as their academic abilities decreased, and children lacked opportunities for achievement, progress or success in the standard curriculum. As such, data emphasise the need for an adaptable, inclusive and flexible approach to education and therapy to respond to the diversity of each affected child; supported by other professionals in the field (Scambler, 2004; von Tetzchner et al., 2019). Guidance in the adult dementia field explicitly recognises the importance of personalised strategies for individuals with dementia, suggesting how carers need to understand, adapt and respond to the diverse and changing needs of someone with dementia (NICE, 2018a). In the absence of such guidance for Batten disease, these research findings call for guidance to recognise the ways in which affected children need a unified, yet bespoke, personalised approach to education that sits outside of standard mainstream education.

Lastly, interviews with professionals highlighted how educators considered the future needs of the child, introducing many creative ways to encourage the child to practise independent skills for the future. Professionals additionally suggested that using the Music Speaks program could encourage children to practise and embed key communicative phrases that they could draw upon when their conversation is deteriorating, to help maintain independent communication in the future. Supporting the theory of a window of opportunity for pre-emptive learning (von Tetzchner et al., 2019), professionals also recognised the fast speed at which children learnt the content of the program and their ability to recall this months later.

Findings are supported by previous research exploring the impact of music and learning and memory retrieval. In the adult dementia sector, research has shown that despite memory loss, learning of new information (in this case new songs) is achievable through music (Baird et al., 2017). Research has also demonstrated that information retrieval is more effective within musical structures like songs than poetry (Samson et al., 2009). This could explain why children could learn the Music Speaks content quickly and remember it months later. Research findings suggest there is a window of learning opportunity for children with Batten disease (Elmerskog et al., 2019), and that they are perhaps able to retain new information, both in the short and long term, before their skills and memory have deteriorated. The unique effect of preserved musical memory could enable children with Batten disease to absorb and remember new information which is meaningful and useful for them (for example phrases that express hunger, pain, or favourite activities). Even in the absence of standardised guidance or policy, it seemed that all professionals understood this window of opportunity, adopting similar approaches to one another by teaching children the skills to benefit them in the future.

Findings emphasise a need for such approaches to be standardised to help others in the education sector and recognise how such approaches can support a child’s care pathway, to compliment medication and other symptom support. Unified educational guidance could help professionals to understand and recognise the significance and importance of pre-emptive teaching, that some educators currently fail to recognise when faced with the child's unpredictable future (von Tetzchner et al., 2019). Recommendations in adult dementia care recognise this approach, suggesting that carers address the future needs of the individual by considering adaptations and personalised strategies to support needs as they change (NICE, 2018a). Research findings therefore call for an expansion to existing policy and health guidance beyond symptom management and physical deterioration for affected children (NINDS, 2020). Guidance for educators could consider how targeted, personalised and meaningful music-based activities could benefit children with Batten disease in the future.

8.2.3. The Experience of Professionals

Lastly, the research tells the compelling story of what it means to support a child affected by Batten disease. Hearing from professionals about their experience of supporting a child with Batten disease for the first time, common challenges and considerations from their educational experiences emerged. Narratives described what is important in those situations of devastating deterioration, and why education, therapy and care can be challenging for both the child and the professional in mainstream education settings. Interviews with professionals give some insight into their personal feelings, highlighting the challenges they experienced in response to caring for a child with a fragile condition. Whilst witnessing years of deterioration, professionals expressed sadness, empathy and fear, reflecting on how the child's presentation declined rapidly during their time in school. Professionals also expressed anxiety and frustration around the pressures of having additional caring responsibilities, the lack of resources, and those around them who lacked a basic knowledge of Batten disease. The perspectives from professionals suggest how the fragile nature of the condition, inappropriate support systems, and lack of knowledge and guidance could impact their own wellbeing.

Previous research also highlighted the complexities of supporting adults with dementia, recognising the frustrations around lack of resources and time, lack of knowledge in others, and the importance of staff wellbeing (Edberg et al., 2008). Recognising the importance of value, recognition and peer acknowledgement, professionals in the current project suggested that support from those around them notably helped them in their roles which perhaps improved their wellbeing. Again, the evidence supports previous research reinforcing the importance of carer wellbeing for those caring for children with Batten disease (Scambler, 2004) and adults with dementia (Edberg et al., 2008; Kitwood, 1997).

In the adult sector, NICE guidelines explicitly address this issue, recommending that carers are given education, training and advice on the challenging aspects of supporting an individual with dementia, suggesting the importance of considering their own wellbeing (NICE, 2018a). The absence of such guidance for staff and carers in the context of childhood dementia suggests there is a gap in appropriate support, resources and training that is needed to deal with the complexities of supporting a child with Batten disease. By addressing this gap, educational guidance for the professional sector could provide a framework for education systems to recognise and improve support resources for schools who educate children with Batten disease. Findings also support the need for wellbeing support mechanisms and guidance for professionals and carers. Given the importance of peer support, evidence also suggests a need for professionals to have a space to recognise, value and support their peers in the field, perhaps in group support spaces with other educators who support children with Batten disease.

The next subsection considers how findings from the research, related to the impact of music, support the use of music therapy-based interventions in the care and education pathways for children with Batten disease.

8.3. The Impact of Music Interventions for Children with Batten disease

Research in the adult sector has offered relatable evidence supporting the use of music interventions for neurological conditions such as dementia or brain injuries. Yet when it comes to the processing of music, there are significant differences between adults and children with neurological conditions (Bower, Magee, Catroppa, & Baker, 2021), stressing the need for further research into the impact of music for children with Batten disease. In the last decade, researchers around the globe have begun to recognise the need for systematic research that explores educational and therapeutic approaches that go beyond pharmacological interventions for children with Batten disease (Elmerskog et al., 2019; Ockelford et al., 2019; von Tetzchner et al., 2019). Whilst primarily focusing on one type of Batten disease (JNCL), this doctoral research makes a unique contribution of evidence to this small and valuable pool of research. The following subsection discusses the ways in which music interventions impacted the children in the research (through music-based activities, music therapy interventions, and the Music Speaks program), and considers how music-based interventions could be embedded into wider care and education pathways for the benefit of children with Batten disease and their educators.

8.3.1. Preserving Functional Skills and Wellbeing

Evidence from the research highlighted how music had a unique and positive impact for children with Batten disease. Professionals shared instances where music could improve wellbeing, reduce anxiety and preserve functional skills (such as communication and mobility). Despite witnessing the overall deterioration of a child over time, professionals described how a child could ‘light up’ in the presence of music, often observing how music brought a sense of clarity for children.

Professionals also described how they, as non-musicians, used music to support children, introducing music-based activities to support inclusion and peer interactions in the school environment.

Presenting similar findings, past research has indicated that music has a unique ability to connect with individuals experiencing memory loss and confusion, and improve wellbeing (Sixsmith & Gibson, 2007). Research findings also support the concept that music can preserve key skills and musical memory in adults with Alzheimer's disease (Cuddy et al., 2015). Parents of children with Batten disease have also reported the importance of music for their child, describing instances where music could elicit functional responses (such as improved speech and communication) as well as moments of improved wellbeing and calm (von Tetzchner et al., 2019). Findings from this research further enhance these observations, suggesting the possibility that general music activities could be beneficial in preserving skills and increasing wellbeing for affected children, demonstrating the potential for music to improve health outcomes for those living with Batten disease.

8.3.2. The Impact of Music Therapy Interventions

The research highlighted how functional skills changed in the presence of music therapy sessions and music therapy-based programs. From observations of children in music therapy sessions, preliminary evidence suggested that music therapy positively influenced communication, mobility, cognition, wellbeing and behaviour for affected children. In particular, a child's wellbeing, speech and cognition in music therapy could seemingly be maintained over periods of time. For the population of children with JNCL, data indicated that cognition and wellbeing could be maintained, and communication slightly improved in music therapy over time. From interviews, professionals also advocated and championed the importance of music therapy interventions for children with Batten disease. Data from the six-week program trial demonstrated how Music Speaks had the potential to facilitate immediate improvements in expressive communication for children with a diagnosis of JNCL. Data suggested short term changes in the clarity of speech, pacing, dysarthria and stuttering for each child immediately after program sessions.

Similarly, professionals also witnessed these immediate improvements to speech pacing and self-expression, describing in interviews how, from their perspective, children visibly learned how to slow their speech down and pace their sentences. Although no sustained improvements were observed in children's expressive communication, professionals described how they felt that the program gave the child the confidence and tools to support their independent communication, suggesting how the program could help preserve their communication for longer. They described how Music Speaks could positively impact general self-expression, ownership and wellbeing. Similarly, findings from interviews suggest that the personalisation of the Music Speaks program was able to meet the child’s diverse interests, providing them with bespoke sessions that were able to give the child a sense of progress and attainment, which was otherwise lacking in their school environment.

Considering how the brain responds to music allows for a deeper understanding as to how and why children with Batten disease may have shown language improvements when taking part in music therapy and the Music Speaks program. Neuroscientific research evidences that music is processed in many different areas of the brain (Alluri et al., 2012), which govern different aspects of language, speech and other functional skills. In the presence of neuroscientific damage or deterioration, theories suggest that music allows new connections to be formed, bypassing damaged areas and allowing functioning to be preserved, re-developed or maintained (Sihvonen et al., 2017). Furthermore, the unique elements of music such as rhythm, pitch, melody and lyrics make it more successful as an intervention in neurorehabilitation than other activities such as listening to audiobooks (Särkämö et al., 2014). The ability for music to access, activate and redevelop areas of the damaged brain could explain why the music interventions in the research had an impact on functional skills. By simultaneously stimulating multiple areas of the brain, it is possible that music therapy sessions and the Music Speaks program can create momentary neurological change for children, allowing functional skills to change and improve. Further research is needed to explore the neuroscientific effects of this in more detail. Preliminary evidence from this doctoral research, however, provides a basis to support the use of music-based speech interventions for children with Batten disease or other neurodegenerative conditions, more so perhaps than other speaking therapies.

Research into the link between music and language could also explain why the researched music interventions had an impact on the child’s speech and communication. Previous research has demonstrated how regular music therapy sessions and workshops can facilitate improvements in language skills and memory over time, in comparison to learning information through oral means such as poetry (Samson et al., 2009). Based on the principles of rhythmic entrainment (Thaut, 2014), previous evidence indicated that the components of rhythm, pulse and tempo allow the brain to unconsciously sync in time with the beat by physically moving or singing in time. It is perhaps the pacing and singing elements of Music Speaks particularly that encouraged children to unconsciously slow their speech down to the rhythms and beats in the program, which in turn, improved the intelligibility of their communication. This supports evidence in the adult sector demonstrating the unique effect of singing for aphasia patients in comparison to speech and language therapies (Lim et al., 2013), and the positive benefits of singing programs for Parkinson’s patients, in comparison with painting, dancing or Tai Chi interventions (Tamplin et al., 2019). It is therefore possible that the brain’s automatic responses to music, rhythm and pacing are one of the reasons why programs like Music Speaks could be effective for children with JNCL.

Lastly, research exploring the physical impact of music on wellbeing and emotion can offer an explanation as to why music therapy or programs like Music Speaks enhanced wellbeing for children with Batten disease. Evidence from the neuroscientific sector suggests that the physical impact of music on ‘feel-good’ hormones (dopamine) can improve mood and wellbeing for people listening to and engaging with familiar music (Salimpoor et al., 2011). First-hand accounts from adults with dementia also describe how familiar music can lift mood, alleviate stress and agitation, and bring feelings of enjoyment and pleasure (Sixsmith & Gibson, 2007). This evidence indicates that the Music Speaks program may have had a positive impact for both the child and the professional by reducing stress and anxiety and increasing wellbeing. Additionally, the person-centred nature of music therapy (BAMT, 2017), was perhaps also able to respond to the changing needs of each child, helping maintain functional skills and wellbeing more effectively over time.

Evidence from the research uniquely demonstrates how music activities and music therapy interventions could have the ability to improve a child’s wellbeing, preserve declining skills, provide opportunities for inclusive and shared experiences, and potentially future-proof independent communication for affected children. Music therapy input could positively facilitate and maintain functional skills over time, and programs like Music Speaks could create short-term, immediate changes in communication outcomes, more so than other types of arts-based or speaking therapy interventions. Considering the mechanisms of neurological change, and the physical effects of music, findings suggest that music therapy-based interventions could improve health and wellbeing outcomes for children with Batten disease. The next subsection goes on to discuss how music interventions could support and develop wider care pathways for children with Batten disease.

8.3.3. Embedding Music into Care and Education Pathways

Considering the positive impact of music therapy interventions in the research, and the lack of national policy or guidance for Batten disease, it is crucial to consider how music could support wider care pathways for children with Batten disease and their educators. Firstly, interviews with professionals described how participation in the research project personally impacted them as professionals. From empowering them in their support roles to enhancing their use of music in education, professionals described how the Music Speaks program facilitated positive, shared experiences with the child. In many cases, professionals described how the program gave them the confidence to use music or program techniques more frequently in school activities for the child. The findings support recent research demonstrating how music therapy home or care-based programs for adults with dementia improved wellbeing and quality of life for individuals and their carers (Baker et al., 2012; Dowson, 2019). Similarly, music therapy care home programs have been shown to enhance an understanding of the impact of music for care staff, giving them the confidence to weave music into their own daily personal care tasks with the adult dementia residents in their care (Hsu et al., 2015). Later research also uncovered similar findings, where singing programs in care homes were positively received by care staff and gave staff, in some cases, the confidence to run their own music programs in the care home setting (Dowson, 2019). It is possible that programs like Music Speaks could improve wellbeing for professionals and facilitate confidence in delivering their own musical exercises. Including more music-based activities in their educational approaches and pre-emptive teaching methods could help maintain the child's functional skills for longer. Evidence from the research has shown that there is a lack of unified guidance or support in educational approaches, and professionals experience pressure in supporting a child with Batten disease. Given the fact that many already used musical activities with the child, it is possible that educators are receptive to new music-based resources or guidance to help formalise and guide their approaches.

Although developed with specific reference to children with Batten disease and JNCL, music therapy services and home or school-based therapy interventions such as Music Speaks could hold potential for other children with neurodegenerative conditions. Music Speaks perhaps offers an intervention that brings new ideas, approaches and ways of using music with the child, in the absence of music therapy or other support therapies. Without standardised guidance or access to music therapy, Music Speaks could potentially offer accessible and sustainable musical resources for affected children. Music therapy-based programs like Music Speaks, could support a child's communication skills and wider wellbeing at school, whilst also providing staff with much-needed resources and music-based approaches to meet the changing, unique needs of the child they support. Considering the psychologically distressing experiences of families at the stages of early diagnosis (Scambler, 2004), early music-based support services like music therapy and the Music Speaks program could help improve wellbeing and psychological support for children and their families as they begin to navigate their way through their Batten disease journey. Ultimately, research findings could provide content for standardised education, care and treatment guidance for professionals in the field and families, calling for unified guidance which recognises the importance of music-based support for children living with degenerative disease.

It is important to lastly consider how the wider role of music could complement existing dementia care pathways. With an ageing population, neurodegenerative conditions are generally increasing in prevalence over time, and whilst there are hundreds of diagnosed neurodegenerative diseases, many of them share symptoms and pathology (Przedborski, Vila, & Jackson-Lewis, 2003). With an anticipated increase of those living with neurodegenerative disease, there is consequently an increasing need for services which support the emotional, physical and social needs of individuals (Hsu et al., 2015; Odell-Miller, 2021; Przedborski et al., 2003), placing an importance on the need to expand therapy and care pathways and possible preventative sustainable approaches. Research into the use of sustainable, long-term music therapy-based home programs for adult dementia is expanding in the UK (Baker et al., 2019; Dowson, 2019; Hsu et al., 2015; Tamplin et al., 2019). Researchers are emphasising the need to formalise music as part of standard care for adults with dementia, embedding music interventions into standard care pathways (Odell-Miller, 2021). As with research into home programs for adults with dementia (Baker et al., 2012; Dowson, 2019; Tamplin et al., 2019) further development of Music Speaks could formalise a program which uses music to pre-empt decline, improve wellbeing and maintain independence for children with neurodegenerative disease. Considering sustainability, music-based home programs in early stages of diagnosis could provide longer-term accessible music-based activities, that reduce the need for intensive medical interventions later in their lifetime. Preventative music home-based programs could complement music therapy services later in the disease journey, offering children and adults with dementia, a more holistic and embedded music-based support pathway throughout their disease journey.

8.4. Summary

As there is no pre-existing research systematically exploring the experiences and influences of music for children affected by Batten disease, this research contributes unique findings to the field. The thesis set out to explore functional responses in music therapy; to explore the extent to which elements of music therapy can be transferred to an educational context and to analyse the lived experiences of those who support children affected by Batten disease. Yet the research unearthed much more than this. Findings have not only revealed the numerous ways in which music can connect with children affected by Batten disease but have also uncovered systemic issues with current care and education guidance for those who support children with Batten disease.

Firstly, the research confirmed that each child is entirely unique and different, bringing into focus the key priorities for supporting children, namely: adaptability; the present and future needs of the child; pre-emptive teaching; and an ultimate focus on the child’s happiness. The research also shed light on the wellbeing of those who support affected children, calling for education and guidance to consider, not only the needs of children with Batten disease, but also those of the people who support them. Shedding light on the issue of the limitations of existing assessment measures in the field, a novel assessment measure enabled the research to explore and identify functional responses of children with Batten disease that standard clinical assessments missed. Considering the diversity of children with Batten disease, further development of the CMTOM assessment measure could add valuable alternative assessment methods for future research in the Batten disease field. The use of an assessment measure like the CMTOM could capture more nuanced skills and behaviours within a musical framework that is applicable for children with all types of Batten disease.

Secondly, music therapy-based interventions in schools seemingly improved wellbeing and preserved key functional skills, indicating the potential for improving health and wellbeing outcomes for children affected by Batten disease. Music Speaks created positive experiences for both children and adults, with initial findings indicating the potential of the program to improve children’s communication through school-based activities. Potentially filling a gap in existing care and education approaches, Music Speaks could offer an accessible solution for professionals, therapists and schools to support a child with JNCL in an education context. Developing the Music Speaks program could contribute a valuable resource for those supporting children in care and education, to improve the wellbeing of children and those who support them. The research offers persuasive evidence to support the wider use of music therapy-based interventions to be introduced and embedded into care and education pathways for children with Batten disease, to address the diverse and changing needs of each child with sustainable music-based resources and activities.

The following subsections discusses limitations in the research project, and consider the benefits of the research methods used and potential further development research in the field.

8.5. Limitations

There are several limitations to this research enquiry. These primarily involve issues associated with the size of the research project, diversity of the interventions, limitations of cross-sectional data, missing or incomplete data sets, and the challenges of assessment measure development. These are explored in more detail in the following sections, with suggestions for future research explored subsequently.

8.5.1. The Nature of Small-Scale Studies

As previously mentioned, given the diversity of the Batten disease population, the small-scale case study design was the most appropriate strategy for this exploratory project. The case study design was largely dictated by the small population of children with Batten disease in the UK and a limited pool of children suitable for research participation. Recruitment of participants was conducted through convenience sampling and therefore may not have represented the wider population of children with Batten disease. The small sample size challenged the ability to make generalisations from the research findings, but equally allowed for data to capture the diversity within the population and the nuanced variability of each child, their disease progression and disease severity. Future research could focus on increased participants, broader groups of professionals and alternative contexts such as home-based program use. This concept is explored in the subsequent section of this chapter.

8.5.2. The Heterogeneity of Music Therapy Practice

It is crucial to recognise the limitations and challenges with the music therapy interventions used in Phase One of the research. Twelve practitioners were recruited to take part in the first phase, delivering weekly music therapy or music-based sessions with children with Batten disease. Aside from a generic suggested session structure (see appendix 5), practitioners created their own session content and set their own therapeutic objectives in line with the principles of standard music therapy practice (HCPC, 2013). It is not known how long each of the music therapy sessions lasted and this may have varied between participants. Even though there were regular researcher visits in sessions throughout the three years, there were only session guidance notes for practitioners to follow. As such, the sessions may have used different musical instruments, songs, music, and activities, creating diverse musical experiences for each child. There may also have been differences in those practitioners who were trained in NMT and some may have used the techniques in their practice.

In a study reviewing music interventions and therapy practices for brain injury rehabilitation (Magee et al., 2017), research also recognised the limitations of the diversity in music therapy practice. Differing approaches to music therapy can challenge the ability to make generalisations from the data as well as the possibility of replicating findings in future studies. Although making generalisations from the doctoral research is challenging, the naturalistic setting of music therapy sessions contributed ecological validity and practical utility for program roll out, acknowledging that in reality that there are diverse ways in which music therapy practitioners approach their practice. Furthermore, given the importance of personalisation of care for affected children, is it perhaps partly the bespoke person-centred approach to therapy that makes music interventions beneficial to wellbeing (Sixsmith & Gibson, 2007). Future research could aim to explore different types of music intervention protocol (structured and non-structured) to explore the extent to which changes in music therapy are dependent on individual practitioners, length of sessions, or approaches to therapy.

8.5.3. Cross-Sectional Data

It is important to note that cross-sectional data collected for this population brought additional challenges regarding the variability and unpredictability of a child's presentation day-to-day and week-to-week. In Phase One, for example, there were no consistent days where research observations took place, and therefore research visits for each of the twelve participants often occurred on different days of the week and different weeks in a school term. Similarly, in Phase Two, data submitted by video only related to one individual session in a week and, although they were all aligned to the same week, some were captured on different days for different participants. This variance was observed in the individual participant scores in both phases, where engagement varied within each session and throughout each week. It is therefore challenging to make deductions or cross-sectional comparisons about children's 'weekly' progress when observations differed between participants. Any changes in functional or communicative responses must therefore be treated as standalone sessional observations rather than weekly representative data.

In Phase Two it was not possible to draw comparisons between the CMTOM scores and the recording sheet comments. This was because most of the recording sheet comments related to different sessions from the videos which were assessed using the CMTOM subscale. Again, in some cases, the child's engagement varied over the week and within each program session, and data therefore could have differed considerably over just one week. This further reinforces the need for future research to address the robustness of data collection, the relevance and frequency of assessment tools used, and the integration of system change theories to strengthen the validation and feasibility of program interventions (discussed further in section 8.7).

8.5.4. Missing and Incomplete Data

In Phases One and Two, there were several instances of missing or incomplete data. For example, in Phase One, mean scores recorded in week one of the research drew on all 12 participants' scores, whereas the final two data collection points only captured five participants' scores and two participants' scores in the final week. Similarly, Phase Two video submissions per participant were variable (ranging from one to eight) and missing data impacted the range of sessions available for analysis. This meant that there was an uneven representation of participant data in this phase. For some weeks, for example, analysis was based on data from just three participants (weeks one, three, and six). This was also the case for the task completion and records of completion, which only featured two of the five participants involved in this phase of the research. Considering these unplanned impacts, future research should aim to improve data collection methods by perhaps utilising electronic recording sheets, or electronic data recording platforms, such as EthOS, so that data are recorded, stored securely and are retrievable in the event of unplanned interruptions.

8.5.5. The Impact of COVID-19

Phase Two of the research was significantly impacted by the COVID-19 pandemic where the overall length of the program trial reduced from 20 weeks to just six. The pandemic not only influenced the trial period length but also the availability of recording sheets and other video data that were lost during periods of lockdown. This impacted the full breadth of data that could be analysed. Having a wider pool of data could represent a more in-depth picture of how more children accessed the program, identifying how their communication responses change over a longer period. It is possible that continuing the program for longer or having access to all participant data may have produced different results. This concept is explored in the next section of the chapter. COVID-19 also impacted how interviews were conducted, moving the face-to-face format onto online virtual platforms. Some participants were unable to access the video conferencing platform, and often their interviews were conducted over the phone at the last minute. Those who encountered problems with virtual communication may have experienced additional stress and discomfort, which could have impacted the mindset of the professional on the day and the content of the interviews.

It is crucial to mention that, particularly given the medically vulnerable state of children affected by Batten disease, the presence of COVID-19 could have impacted wellbeing and anxiety levels of staff and children during the Music Speaks program trial. Research has demonstrated that schools and educators experienced the pressure of school closures, child welfare, and the challenges of remote learning in home environments during the pandemic (Moss et al., 2020). This further reinforces the importance of future research involving longer trial periods, pluralistic mixed data collection methods, a wider pool of children and professionals, as well as the feasibility of transferring the program into a home-based format, to strengthen the evaluation of the program in other contexts. It also suggests the need to collect rich, person-centred data that enables a fuller picture of the complex lives of participants to be explored.

8.5.6. Challenges with Assessment Measures

There were several limitations with the assessment measures used. Firstly, limitations were identified in the face validity and reliability tests carried out on the CMTOM assessment measure. Namely, in the face validity exercise, since all practitioners were also participants in the research delivering the music therapy intervention, they may have felt obliged to give positive scores to support the research or represent a more favourable perspective of the assessment measure.

There was also a limitation in the sample size (N = 6) and a larger number of respondents may have yielded different results. These first-hand perspectives from practitioners though, provide valuable insight into the use of the measure for those who support children with Batten disease in real life music therapy contexts, adding depth and ecological validity to the questionnaire response data.

The reliability tests additionally had limitations. The small number of videos assessed (n = 10/182) only represented 5.5% of the video data collected and may not have adequately been representative of the whole video data set. Additionally, using two assessors limited the findings, and a higher number of assessors may have yielded a more accurate representation of assessors' agreement. Again, both assessors (me included) were research officers in the research team, and reliability measures, therefore, can only be a measure of subjective agreement and interpretation (Onwuegbuzie & Leech, 2005). Furthermore, it is important to acknowledge that this first stage of Phase One largely focused on the assessor and practitioner responses in the validity and reliability exercises. Further validation studies could aim to assess the credibility and validity against other standardised interventions and assessment measures in the field, to optimise the measure's fidelity and reliability (Navarro Sada & Maldonado, 2007). Future work could also consider alternative stakeholder perspectives on the validation and reliability of the measure, for example, affected children themselves, other practitioners, parents or family members or other types of professionals supporting children affected by Batten disease (Skivington et al., 2021).

Secondly, in Phase One, the research utilised two different measures to capture responses from the perspectives of music therapy and clinical contexts. There was a notable variance in the number of times each assessment was conducted over the three years. The Hamburg scale yielded three ratings in total, in contrast to the CMTOM, which gathered an average of 15 ratings per participant. This meant that the CMTOM measure picked up more detailed data as it was executed more frequently, and sharp deteriorating scores in the Hamburg data could be exaggerated by the infrequent yearly use of the measure. Drawing comparisons between the two measures is therefore challenging. There is a need for future research to optimise initial findings from this research project and conduct further analysis into the appropriateness, relevance, frequency and robustness of assessment measures in the Batten disease research field.

The rapid and brief development of the CMTOM communication subscale brought challenges and limitations to the research. Whilst early phases of the research explored the validity and reliability of the CMTOM measure, the CMTOM subscale development did not involve such validation exercises. Further research, therefore, could aim to explore the robustness and appropriateness of this subscale with validation and reliability exercises. The professionals involved in the feedback stages of the subscale development were also involved in the Music Speaks program development and may have presented a more favourable perspective on music therapy education programs and promote positive research outcomes. Involving these professionals though gave important insight into how responses to the program could be translated and tracked with a suitable assessment measure, adding important continuity to the development of both the program and the subscale measure. Again, further validation exercises could draw upon blind inter-rater evaluation exercises to strengthen the validity of this subscale measure.

The disparity in the individual participant data from Phase Two additionally highlighted the need for a more nuanced assessment measure for this group of children. With some children maintaining the highest scoring on the CMTOM subscale throughout the whole six-week program trial, a possible ceiling effect could have occurred. Other participants scored lower on the CMTOM subscale but showed more variable change in response to the program week-by-week. This suggests a more sensitive scale could have picked up more detailed communication responses from the baseline to the endpoint for each participant. The program could have yielded different results if implemented at different stages in disease progression. The research reinforces the need for further development and evaluation of the measure and further exploration into alternative methods of implementation.

Lastly, throughout the development stages, there were significant limitations in measuring certain characteristics of language and communication that were relevant to the Music Speaks objectives. Research suggests that communication consists of many components of non-verbal gesture, eye contact, facial expression, exchange, interaction, as well as listening skills such as hearing, message retrieval, understandability and context (Miller, 2013). Using a measure that solely assessed speech production, expressive communication, and receptive skills such as word recall, therefore, omits potentially important contextual information about the full breadth of a child's communication skills. For the CMTOM communication subscale to be more widely applicable as a clinical assessment measure, development and evaluation stages should consider both verbal and non-verbal aspects of communication.

8.6. The Strength of Research Assessments and Methods

Despite the limitations outlined in this chapter, methodologically speaking, the approaches used in this research contributed a unique perspective on the impact of music for children with Batten disease. A central challenge in research for children with Batten disease lies in identifying appropriate measures that capture all elements of a child's functionality irrespective of age, diagnosis and disease progression (Augustine et al., 2013; von Tetzchner et al., 2013), emphasising a need for more frequent, detailed generalisable assessment tools in the field. The

CMTOM measure provided the opportunity to observe in closer detail responses to music therapy for affected children, beyond that which can be observed from the standard clinical perspective. Initial validation of the CMTOM measure suggests that it serves as a valuable tool in facilitating observation of children in music therapy sessions over short term and longer periods of time. Expanding upon the findings from this research could begin to address existing challenges with appropriate assessment measures for the Batten disease population. Strengths of the CMTOM measure could open-up the opportunity for more appropriate music-based assessment measures to be utilised for affected children.

The mixed methods approach enabled the analysis of multiple perspectives through diverse data collection methods that involved quantitative observational methods and qualitative interviewing. The diverse perspectives provided a deeper understanding and context as to how music had an observed influence on functional skills for children, their perceived impact of the Music Speaks program, and how their education experiences were impacted by systemic issues and challenges (Archer et al., 2016). Capturing in-depth narratives, observations and perspectives in the research demonstrates why mixed methods study designs are appropriate for this population.

Using an inductive approach enabled a dialogue to emerge between the empirical data (observations of children in musical contexts), the perspectives of professionals and the underlying causal mechanisms (Bhaskar & Danermark, 2006). Highlighting the uniqueness of each child, systemic issues around absent policy and guidance, and diverse approaches to education guided each follow-on phase of the research. For example, observations in Phase One initially suggested the importance and influence of music. While narratives and observations from other phases offered a contextual framing around how and why music may have such an influential role for children with Batten disease and the immediate need for music-based school resources for the child, leading to the development of the Music Speaks program.

By utilising a more holistic, ethical, and flexible research design than say a randomised control study (Augustine et al., 2013), the research offered an alternative way of researching children affected by Batten disease. Rather than using unethical comparative control groups, the case study approach focused on nuanced within-participant changes and unique perspectives, allowing the research to gather in-depth data for children and those who support them. This could pave the way for further research to adopt similar inductive case study research approaches to investigate the diverse needs of participants.

Exploring considerations for future research, the next subsection discusses potential future developments to improve the validity and robustness of measures and programs developed in this research.

8.7. Considerations for Future Research

Initial findings from the three phases have provided a platform from which future research can develop this project further. This section begins by outlining the possibility of further evaluative studies to strengthen the validity of the CMTOM assessment measure. Furthering the development of the Music Speaks program, comments and ideas presented by professionals regarding adaptations, improvements are then presented, with the concluding section discussing how further optimisation studies can explore valuable future developments to the program.

8.7.1. Validation of the CMTOM Assessment Measure

The development of the CMTOM and CMTOM communication subscale offered an opportunity for responses in music therapy and music sessions to be reliably tracked over time. Further research studies could aim to strengthen the validity of the CMTOM measure, piloting alternative methods of research design, data collection methods, and alternative communication criteria. Alternative methods could involve the use of natural history data to provide comparative data from children whose skills have been naturalistically tracked over time without additional interventions or medications. By collecting and analysing a wider sample of data over a longer period of time, future research could consider other measures to improve the robustness of findings and test-retest reliability such as using blind raters for inter-rater assessments. Further research could also explore alternative communication criteria, examining how other elements of receptive and non-verbal communication can be tracked and measured using the CMTOM subscale to widen the potential for using the tools with other communication disorders. Preliminary findings highlight the need to evaluate the assessment measure in other contexts (such as music therapy sessions at home or in residential care settings, music therapy sessions in acute care, or group music therapy sessions) or with other children with Batten disease. There could also be a consideration of structured music therapy protocol and how this may impact the assessment scores over periods of time. Validation exercises overall could aim to assess contextual relevance, external validity, concurrent validity and construct validity (Navarro Sada & Maldonado, 2007).

8.7.2. Development of the Music Speaks Program

Future research developing the Music Speaks program must first listen to the suggestions made by professionals, as to how the program could be adapted and developed. Professionals identified ways in which the program was perhaps restrictive and repetitive for some of the children taking part. Suggesting a program format that includes greater opportunity for adaptation, professionals introduced the idea of content flexibility (for example suggesting how songs on specific days of the week could be interchangeable and flexible); a suggestion that was also made in previous research exploring group singing programs for adults with dementia (Dowson, 2019). Professionals suggested the program may have a greater impact if it were implemented earlier on in the disease progression before a diagnosis of Batten disease and before a child's skills had begun to deteriorate. Complementing other research into music therapy-based programs (Baker et al., 2012), professionals lastly suggested the possibility of transferability outside of the program sessions, using musical elements from the program in other situations or with other children (for example those with other communication difficulties).

To optimise initial findings from the Music Speaks program, therefore, future research could aim to develop adaptability, implementation, transferability and feasibility of the program. The following sections explore these four key factors in more detail guided by the protocols of evaluating and developing complex medical interventions (Skivington et al., 2021).

Adaptation

To address adaptability of the program, rather than opting for a randomised controlled trial format or comparative study, further research could aim to involve mixed methods research approaches that integrate theory with evaluation (Skivington et al., 2021). Beginning with input from relevant stakeholders (professionals, families and perhaps children themselves), refinement of the program could initially repeat development iterations to address appropriateness and adaptability of the program content. Involving stakeholders in early refinement stages could help develop different versions of the program that include both structured and unstructured formats, varying lengths of the intervention, and different variations of content. Collaborating with stakeholders could also help future research consider and define reasonable criteria for the measurement of success. For example, stakeholders could help determine the extent to which communication and wellbeing outcomes are classified as improved, how data collection or assessment measures might capture and measure research outcomes, and the capacity to which the program is practical in other care or home-based contexts.

In line with other research into the lived experiences of adult dementia (H. Johnson, 2016), research methods such as Interpretative Phenomenological Analysis (IPA) could enable an in-depth exploration into the lived experiences of children, families and professionals. Although interviews with children themselves could raise ethical issues (concerning cognitive capacity, lack of memory, communication issues or knowledge about their condition), it could be possible to explore other methods of data collection from children with appropriate ethical consideration of consent and cognitive capacity. Future research for example, could aim to interview children with more general questions about their experiences of school and the Music Speaks program or use alternative data collection methods such as arts, crafts, play, photography or videography approaches (Blaisdell, Arnott, Wall, & Robinson, 2019). Using creative methods like this could gather reflective and meaningful accounts from children without the need for a spoken interview format, which some children with Batten disease may struggle to engage in.

Implementation

Addressing implementation, further research could aim to assess the impact of delivering the program at different points in disease progression to assess the effects of earlier or later implementation over a longer period. To explore the diversity of the population in more detail, a wider pool of participants would investigate the impact of the program for more children with all types of Batten disease variants (aside from those with JNCL). Success criteria relating to percentages of program delivery and task completion would also be pre-defined to increase the fidelity of findings. Considering the potential loss of data, studies could further address more robust data collection methods utilising electronic methods such as EthOS. Economic modelling at scale would also determine whether the intervention can be carried out at a reasonable cost now and into the future (Skivington et al., 2021).

Transferability and Feasibility

Follow-on studies could aim to evaluate the program in other contexts, beginning with the possibility of transferring the program into home settings or in other care settings. Transferability could also incorporate research involving different types of professionals, family members of affected children, and children with other types of neurodegenerative or communication disorders. Maintaining the positionality of critical realism, interviews could aim to develop feasibility for different types of professionals, capturing a deeper understanding of the level to which educational, system and practice change is needed (Skivington et al., 2021). Thereafter, research could explore how family members respond to the needs of a child, what their support priorities involve, and how this, in turn, could impact future considerations of program delivery and implementation at home.

In summary, utilising a process evaluation structure, follow-on research could ultimately consider and explore change theories relating to pre-emptive education (von Tetzchner et al., 2019), the influence of music as a change mechanism (Lim et al., 2013; Särkämö et al., 2014), and systemic theories relating to sustainable music-based programs (Baker et al., 2012). Framing future research in systemic change theory could offer insight into the implications of cost-effectiveness, scalability, and the longer-term impact of music therapy programs to improve the health and wellbeing outcomes for children affected by Batten disease and those who support them in education settings. Considering the wider impact for families, professionals and affected children, future research could provide further evidence to support the need to create and advise on the content for guidance for Batten disease, calling for policymakers to include childhood neurodegenerative disease in NICE guidelines, and to improve guidance and support for families and professionals from the point of diagnosis.

8.8. Reflexivity

Researching children with Batten disease has brought to light considerable ethical, emotional and psychological experiences for me as the researcher. The fragile and devastating nature of Batten disease has impacted the ways in which I have developed as a researcher and professional, and has ultimately influenced how the project has been carried out. This section therefore considers how these experiences have impacted and influenced the course of the research project and doctoral journey.

In line with other music therapy research and social research methods (Atkins & Murphy, 1993), I kept a reflective journal throughout the stages of research planning, data collection and interpretive analysis to critically interrogate my perspectives, experiences, and values. Reflective journaling allowed for reflection, self-awareness, and critical analysis to facilitate a deeper level of engagement with the data, the children and the professionals in the research (Atkins & Murphy, 1993; Edwards, 2012). My various roles, encounters, feelings and values are explored subsequently in further detail.

As I first met with families affected by Batten disease, I got to know them, understanding their individual circumstances, and learning about the prognosis from scientific and medical literature. As my understanding developed within the first few months, I recognised my feelings of disbelief, astonishment and compassion. The situations faced by families around the world felt unjust, and there was a feeling of disconnect; I could not fully understand how families felt, lived and managed day-to-day. As a parent, I recognised my empathy for their fragile and rare situation which became the main driver and motivation to, in some way, improve the lives of the children. As a researcher, I realised that my previous intentions to approach research from positivist positionings were exclusive and inappropriate (Edwards, 2012) and my epistemological position began to gravitate towards a critical realist standpoint.

These first experiences gave me insight into the unpredictable and uneven trajectory of loss that many children experienced. For most, the journey is turbulent and the psychological anguish for parents is consistently present. When children in the research passed away unexpectedly, I shared feelings of sadness, loss, compassion, and bereavement with families. Moments like these strengthened my relationships with those in the Batten disease community, and writing a reflective journal was fundamental during these times. Documenting, acknowledging and reflecting upon my thoughts and feelings allowed me to process the events of a child passing. As with other researchers in the field (Scambler, 2004), I found that these experiences ethically influenced the direction of the research, allowing me to recognise the immediate need for tools, resources and programs, which were useful or meaningful for the children now in their current situations, rather than years into the future.

Whilst working alongside care professionals and educators, I began to see another dimension to the world of those supporting children affected by the disease. In care and education settings I witnessed how opportunities for affected children seemed to deteriorate in synchrony with their medical condition. In most cases, I felt that professionals lacked a systematic approach to proactive or adaptive education strategies for affected children. This led me to consider the possibility that lacking standardised guidance or systems perhaps influenced the experiences of children and their professionals, which again guided me towards observing research from a critical realist perspective. I also developed a passion for advocacy in the research community, seeing the value in educating others on a little-known subject such as childhood dementia. Initially, my feelings of disconnect and a lack of understanding were replaced with a unified purpose; to advocate and improve the lives of children with Batten disease and educate others in the health and social care sectors.

My expertise in the field of neurologic music therapy and brain injury additionally had a part to play in the research process. Coupled with my passion to help and support, I recognised the value of exploring creative solutions to the issues and challenges faced by the children. It was apparent that financial circumstances, fragile health and the unpredictability of the disease in many ways rendered conventional music therapy services inappropriate for affected children. Witnessing the immediate and urgent needs of children and their families led me to focus on the possibility of finding a way to transfer elements of music therapy into accessible resources for educational environments. As with other research for children with Batten disease, there were moments of equipoise where the immediate needs of the child and their professionals needed to be balanced with the research intentions and outcomes (Augustine et al., 2013). The urgent need for music resources was at odds with the pressure to preserve the integrity and format of traditional music therapy practices. Exploring the idea that elements of music therapy could be transferred into programs run by professionals (who did not have music therapy training) could undermine the validity of the music therapy profession and challenge the importance of music therapy clinical training and accreditation. As with other researchers in the music therapy field, I acknowledged a cautious balance between preserving music therapy principles, and the need for low-cost, sustainable and accessible music-based interventions for vulnerable populations and their carers (Baker et al., 2019). Reflecting on this position of flux enabled me to consider how the perspectives of Music Therapists could be captured in research to offer a clinical perspective on the program's intentions and outcomes.

My experience as a clinician combined with my emersion in the Batten disease community also impacted the analytical stages of the research. Responses, observed behaviours and experiences were interpreted by me, which were influenced by my own experiences, intentions, and expectations of what the data would reveal (Braun & Clarke, 2006). For example, my experience as a clinician lent itself to tracking and monitoring behaviours from a clinical perspective, which perhaps allowed me to capture more detailed data in the early research phases research than another type of researcher or professional. As I became familiar with the children's responses, behaviours and communication skills over time, there were instances where I inferred meaningful or coherent communication from a child because of my familiarity with them and their speech. In later stages of the research project, my familiarity and connection with the professional participants increased over time, meaning I interpreted content from their interview data based on my knowledge of them and their inferred meanings. The process of reflective journaling allowed me to critically interrogate these influences considering their impact on the research outcomes. Ultimately, I believe my own values, relationships, understandings, and interpretations in the analytical process supported the process of gathering such unique, rich interpretations, concepts and ideas of others (Braun & Clarke, 2006; Fletcher, 2017).

In summary, throughout my doctoral training, I have been immersed in the Batten disease community on multiple levels. My role as a researcher initially facilitated a rapport with the families and children affected by the disease and allowed me to forge connections with other researchers, scientists, and family members in the international Batten disease community. My passion as a Batten disease advocate in the public domain positioned me alongside those championing Batten disease education, giving me insight into the societal and educational challenges associated with Batten disease. These experiences have ultimately influenced a journey of self-learning and discovery, which has led me from a positivist stance to the positioning of critical realism. From initial visceral feelings and reactions to becoming an advocate in the community, the reflective research experience has ultimately led to the conceptualisation and application of an innovative program which I hope will support those who need it most. As a pioneering researcher in this area of music therapy and Batten disease, I aspire to continue the research into subsequent projects, furthering the evidence around music therapy interventions for neurodegenerative diseases.

8.9. Concluding Remarks

Batten disease is a rare neurodegenerative disease, causing distress and devastation to the affected children, their families and those who care for them. With potential curative solutions being a long way from completion, there is an increasing need for developments to education, care and therapeutic interventions. As a first-of-its-kind study, this project has unearthed the value of music for affected children and their education professionals. Music interventions appear to have the ability to positively impact functional skills both in the short term and over longer periods of time for affected children. Yet the landscape of care and education is more complex and vast. Delving deeper into experiences highlighted the way in which children with Batten disease are diverse, emphasising the fact that any beneficial intervention must be flexible and bespoke, addressing both children’s current and future needs.

Music therapy-based programs could provide a much-needed resource for professionals and schools, tapping into the neurological, emotional and physical impact of music. With an absence of any health policy or educational guidance for children with Batten disease, educators are seemingly finding their own way without adequate support, and this needs to change. Policymakers need to recognise the immediate need for standardised guidance considering how music therapy-based interventions could be incorporated into care and education pathways for children with Batten disease. Embedding valuable music-based support could complement existing symptom support, offering a blended model of care to address the diverse and changing needs of the child, their families and educators throughout their disease journey.

In conclusion, I remember Emma.

Her story, and the stories of other children suffering with Batten disease, are the reason I started this research. Emma passed away a year before the research finished. But there are stories of opportunity, hope and existence for other children still to be told. Emma’s family and close friends continue to give voice to children with this devastating disease by telling her story, and by supporting other families with newly diagnosed children. I hope that this thesis will bring the distressing consequences of this rare disease into sharp focus, giving others the impetus and passion to improve the lives of those living with this heart-breaking disease. Parent and family advocacy charities around the world continue to raise awareness for Batten disease, raising much-needed funds for research in many new areas. I hope that my findings and resources in this area can offer valuable ways for educators, therapists and family members to connect with children through music, improving the quality of life for children who are living with the disease every day. I trust and believe that music can in some way help alleviate the harrowing symptoms of the disease, even if only for a single moment. I hope that music can become a support mechanism for children to hold onto parts of themselves and their world for as long as possible.

One of Emma's favourite songs was 'Part of This World' from The Little Mermaid (1989). I leave you with her favourite part, the words she would sing every week in her music therapy sessions:

> "Up where they walk, up where they run
> Up where they stay all day in the sun
> Wandering free, wish I could be
> Part of that world"

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Appendices

Appendix 1: Chiltern Music Therapy Observation Assessment Measure (Atkinson, 2018)

Observations	**DATE**
Mark as 0 (never), 1 (rarely), 2 (sometimes), 3 (often) or 4 (consistently)	**Score**
Communication & Social interaction	
Appropriate eye contact or tracking of visual stimuli	
Appropriate use of gesture	
Vocalisation (any sound)	
Verbalisation (use of speech)	
Singing	
Awareness of others	
Ability to interact non-verbally / verbally	
Interaction with staff	
Ability to Initiate interactions	
Behaviour / music to therapist appropriate?	
Notice, tolerate, accept, aware of others	
Ability to participate / join in	
Sharing emotions, thoughts and ideas	
Being able to think about others - show empathy	
Behaviour	
Trigger observed to changed behaviour?	
Any verbal aggression noted	
Any physical aggression	
Behavioural response to musical components noted?	
Ability to express / control self in an appropriate way	
Expression of feelings of distress, agitation, anxiety	
Expression of feelings of depression, trauma, loss, bereavement	
Emotional Expression	
Verbal expression of mood	
Physical expression of mood	
Musical expression of mood	
Choice of instrument / art material / object / preference	
Use of voice/ sound making tools for expressing self	

Able to tolerate sound(s), art form, types of media used	
Insight into difficulties & strengths	
Ability to explore and discover	
Shows capacity to improvise / free play	
Can differentiate between real and imagined	
Has enthusiasm, shows pleasure, fun, enjoyment	
Sense of Self	
Ability to participate, initiate, choose, lead	
Shows appropriate level of self confidence	
Is resourceful, decisive and can work autonomously / independently	
Demonstrates appropriate levels of assertiveness	
Cognition	
Follows verbal instructions	
Makes choices	
Ability to attend to task	
Recognition or carry over of previous material	
Engages appropriately with instruments	
Any memory recall noted? (rhythmic recall etc.)	
Sustains attention	
Shows interest and is inquisitive	
Physical Presentation	
Active movement noted?	
Core/trunk stability noted?	
Head and neck stability noted?	
Use of weaker limbs noted?	
Bilateral (both hands) coordination noted?	
Hand-eye coordination noted?	
Ability to cross mid-line - movement R-L or L-R	
Fluency of gait movements?	
Ability to grip in RH	
Ability to grip in LH	
Individual finger movements noted for use with piano, assistive or music technology?	
Oral motor control noted?	
Breath control and regular respiration noted?	
Hand-over-hand or facilitated movement needed to participate in music making?	

PMLD & SD clients only	
Tracking of visual stimuli	
Turning head towards visual stimuli	
Can focus on one visual stimulus	
Moves eye gaze or head towards auditory stimuli	
Can focus on one auditory stimulus	
Responses to musical stimuli	
Interactive response to musical stimuli	
Responses / activity to verbal commands	
Arousal observed during session	
Change in facial gesture or expression	
Change in eye direction or eye contact	
Change in physical movement	
Change in respiration	
Change in vocalisation	
Change in level of arousal	
Response to pulse / rhythm	
Response to melody / pitch	
Response to timbre	
Response to dynamics / intensity	
Response linked to form e.g. phrase, silence	
Response to tempo	
Response to mood / feel of music	
Vocalisation to musical stimuli	
Singing to musical stimuli	
Use of communication and gesture without prompts	
Made choices for object preference (looking towards one)	
Made choice between two objects	
Communicated choice between more than 2 objects	
Reflexive / non-purposeful movements made	
Purposeful movements made	
Attended to tasks	
Intentional responses	
Emotional responses	

Appendix 2: Ethical Approval

Phase One Ethics Approval Letters

Dear Rebecca,

Ethics Application
Applicant: Rebecca Atkinson
Title: Researching the role of music in enhancing the lives of children with neurodegeneration ('MIND') Phase One – Battens Disease
Reference: EDU 17/ 137
Department: Education

Under the procedures agreed by the University Ethics Committee I am pleased to confirm that your Department has approved your above application subject to the following conditions:

Comment:

I am pleased to confirm that the risk assessment for your project has been reviewed and approved by the Health, Safety and Environment Department.

Dear Rebecca,

Ethics Application (Amendment 02.18)
Applicant: Rebecca Atkinson
Title: Researching the role of music in enhancing the lives of children with neurodegeneration ('MIND') Phase One – Battens Disease
Reference: EDU 17/ 137
Department: Education
Original Approval Date: 20.10.17

Under the procedures agreed by the University Ethics Committee I am pleased to advise you that your Department has approved the minor amendment to your above application dated 02.02.18 and confirmed that all conditions have been met (but please see the minor condition below). We do not require anything further in relation to this amendment.

Minor Condition:

> Please proofread carefully the text at the top of the questionnaire. Please insert 'to' between 'the tool aims' and 'gather and measure characteristics'. This should read as 'the tool aims TO gather and measure…'

As this is only a minor condition it is assumed that you will adhere to this condition for approval and therefore we do not require a response

Please Note:

- **This email confirms that any conditions have been met and thus confirms final ethics approval for this amendment (it is assumed that you will adhere to any minor conditions still outstanding, therefore we do not require a response to these).**

- **University of Roehampton ethics approval will always be subject to compliance with the University policies and procedures applying at the time when the work takes place. It is your responsibility to ensure that you are familiar and compliant with all such policies and procedures when undertaking your research.**
- **If this project involves clinical procedures or administering substances it is a condition of Ethics approval that all relevant SOPs published on the department communities pages are fully complied with.**
- **Please advise us if there are any changes to the research during the life of the project. Minor changes can be advised using the Minor Amendments Form on the Ethics Website, but substantial changes may require a new application to be submitted.**

Phase Two Ethics Amendment Approval Letter

Dear Rebeca,

Ethics Application (Amendment 08/10.19)
Applicant: Rebecca Atkinson
Title: Researching the role of music in enhancing the lives of children with neurodegeneration (‘MIND’) Phase One – Battens Disease
Reference: EDU 17/ 137
Department: Education
Original Approval Date: 20.10.17

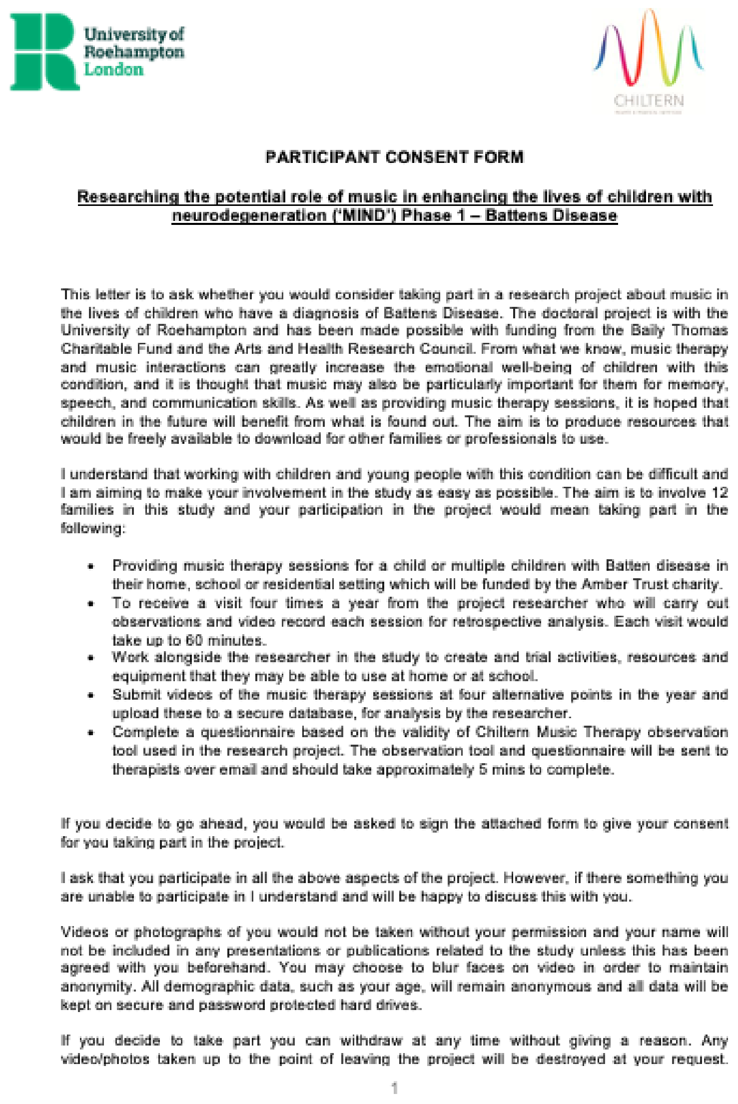
Under the procedures agreed by the University Ethics Committee I am pleased to advise you that your Department has approved the amendment to your above application dated 06.08.19 and the further revisions dated 23.10.19. We confirm that we have received the amendment form signed by your Director of Studies (minor condition in our email dated 27.08.19). We do not require anything further in relation to this amendment.

1)

Please Note:

- **This email confirms that any conditions have been met and thus confirms final ethics approval for this amendment (it is assumed that you will adhere to any minor conditions still outstanding).**
- **University of Roehampton ethics approval will always be subject to compliance with the University policies and procedures applying at the time when the work takes place. It is your responsibility to ensure that you are familiar and compliant with all such policies and procedures when undertaking your research.**
- **If this project involves clinical procedures or administering substances it is a condition of Ethics approval that all relevant SOPs published on the department communities pages are fully complied with.**
- **Please advise us if there are any changes to the research during the life of the project. Minor changes can be advised using the Minor Amendments Form on the Ethics Website, but substantial changes may require a new application to be submitted.**

Appendix 3: Phase One Practitioner Information and Consent Pack



University of
Roehampton
London

CHILTERN

PARTICIPANT CONSENT FORM

Researching the potential role of music in enhancing the lives of children with neurodegeneration ('MIND') Phase 1 – Battens Disease

This letter is to ask whether you would consider taking part in a research project about music in the lives of children who have a diagnosis of Battens Disease. The doctoral project is with the University of Roehampton and has been made possible with funding from the Baily Thomas Charitable Fund and the Arts and Health Research Council. From what we know, music therapy and music interactions can greatly increase the emotional well-being of children with this condition, and it is thought that music may also be particularly important for them for memory, speech, and communication skills. As well as providing music therapy sessions, it is hoped that children in the future will benefit from what is found out. The aim is to produce resources that would be freely available to download for other families or professionals to use.

I understand that working with children and young people with this condition can be difficult and I am aiming to make your involvement in the study as easy as possible. The aim is to involve 12 families in this study and your participation in the project would mean taking part in the following:

- Providing music therapy sessions for a child or multiple children with Batten disease in their home, school or residential setting which will be funded by the Amber Trust charity.
- To receive a visit four times a year from the project researcher who will carry out observations and video record each session for retrospective analysis. Each visit would take up to 60 minutes.
- Work alongside the researcher in the study to create and trial activities, resources and equipment that they may be able to use at home or at school.
- Submit videos of the music therapy sessions at four alternative points in the year and upload these to a secure database, for analysis by the researcher.
- Complete a questionnaire based on the validity of Chiltern Music Therapy observation tool used in the research project. The observation tool and questionnaire will be sent to therapists over email and should take approximately 5 mins to complete.

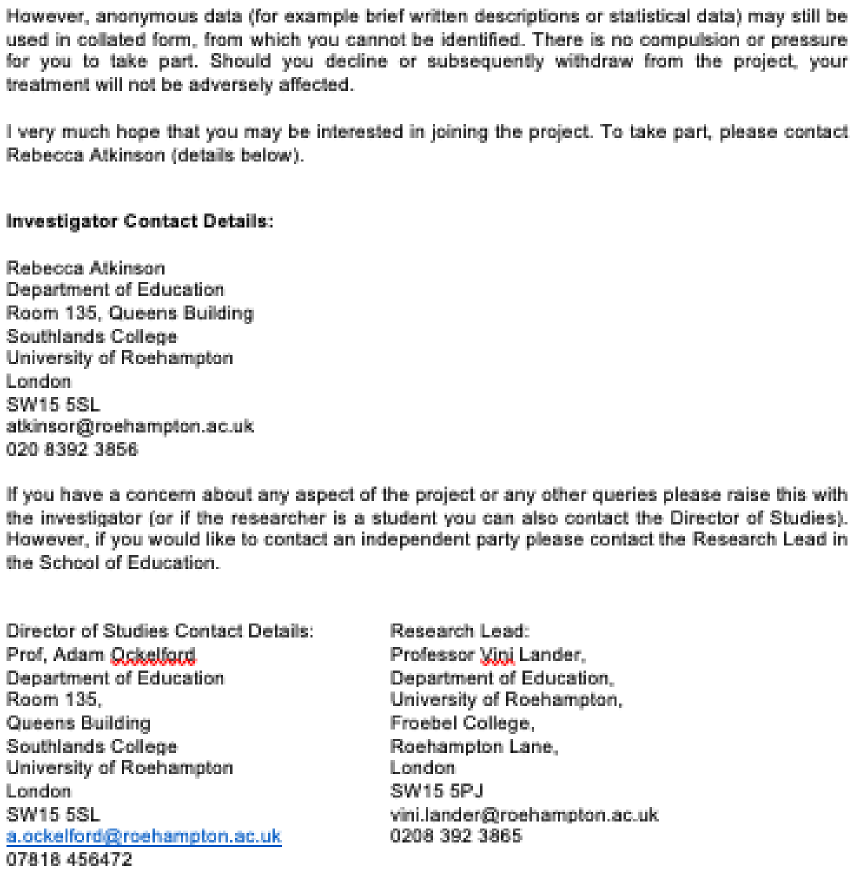
If you decide to go ahead, you would be asked to sign the attached form to give your consent for you taking part in the project.

I ask that you participate in all the above aspects of the project. However, if there something you are unable to participate in I understand and will be happy to discuss this with you.

Videos or photographs of you would not be taken without your permission and your name will not be included in any presentations or publications related to the study unless this has been agreed with you beforehand. You may choose to blur faces on video in order to maintain anonymity. All demographic data, such as your age, will remain anonymous and all data will be kept on secure and password protected hard drives.

If you decide to take part you can withdraw at any time without giving a reason. Any video/photos taken up to the point of leaving the project will be destroyed at your request.

1



However, anonymous data (for example brief written descriptions or statistical data) may still be used in collated form, from which you cannot be identified. There is no compulsion or pressure for you to take part. Should you decline or subsequently withdraw from the project, your treatment will not be adversely affected.

I very much hope that you may be interested in joining the project. To take part, please contact Rebecca Atkinson (details below).

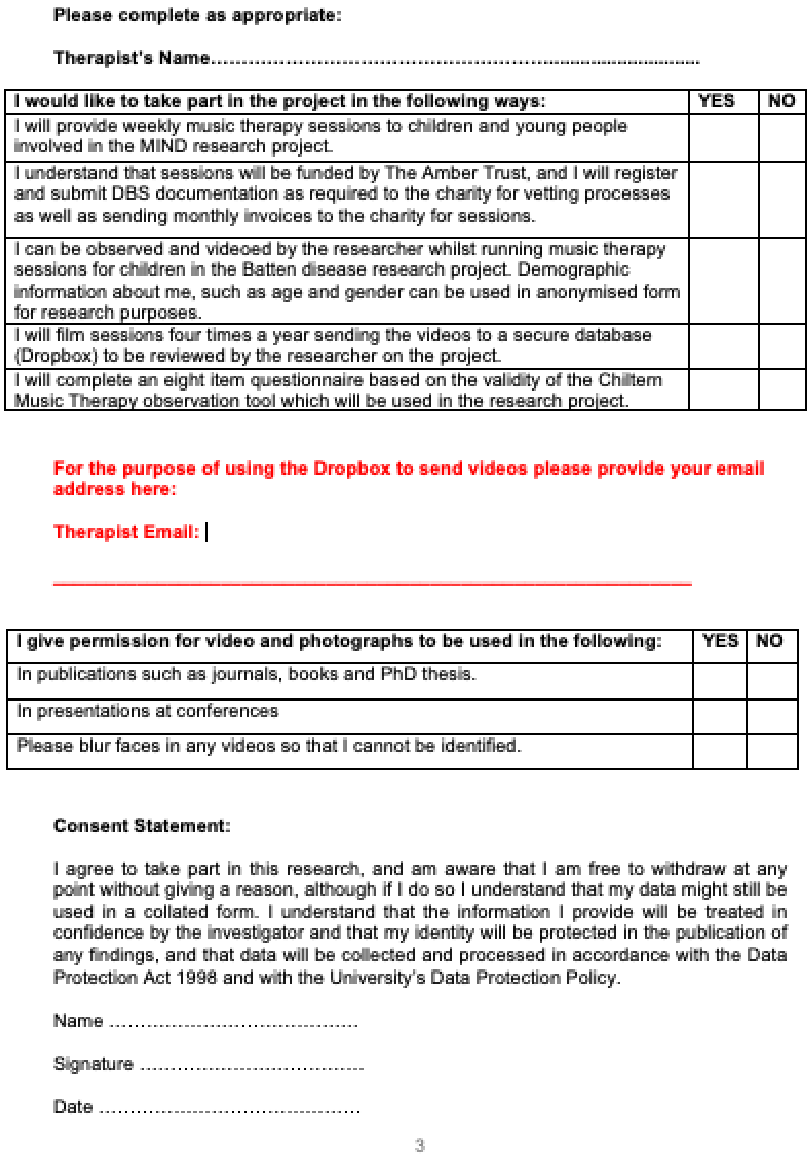
Investigator Contact Details:

Rebecca Atkinson
Department of Education
Room 135, Queens Building
Southlands College
University of Roehampton
London
SW15 5SL
atkinsor@roehampton.ac.uk
020 8392 3856

If you have a concern about any aspect of the project or any other queries please raise this with the investigator (or if the researcher is a student you can also contact the Director of Studies). However, if you would like to contact an independent party please contact the Research Lead in the School of Education.

Director of Studies Contact Details:
Prof, Adam Ockelford
Department of Education
Room 135,
Queens Building
Southlands College
University of Roehampton
London
SW15 5SL
a.ockelford@roehampton.ac.uk
07818 456472

Research Lead:
Professor Vini Lander,
Department of Education,
University of Roehampton,
Froebel College,
Roehampton Lane,
London
SW15 5PJ
vini.lander@roehampton.ac.uk
0208 392 3865



Please complete as appropriate:

Therapist's Name……………………………………………………………………………

I would like to take part in the project in the following ways:	**YES**	**NO**
I will provide weekly music therapy sessions to children and young people involved in the MIND research project.		
I understand that sessions will be funded by The Amber Trust, and I will register and submit DBS documentation as required to the charity for vetting processes as well as sending monthly invoices to the charity for sessions.		
I can be observed and videoed by the researcher whilst running music therapy sessions for children in the Batten disease research project. Demographic information about me, such as age and gender can be used in anonymised form for research purposes.		
I will film sessions four times a year sending the videos to a secure database (Dropbox) to be reviewed by the researcher on the project.		
I will complete an eight item questionnaire based on the validity of the Chiltern Music Therapy observation tool which will be used in the research project.		

For the purpose of using the Dropbox to send videos please provide your email address here:

Therapist Email: |

____

I give permission for video and photographs to be used in the following:	**YES**	**NO**
In publications such as journals, books and PhD thesis.		
In presentations at conferences		
Please blur faces in any videos so that I cannot be identified.		

Consent Statement:

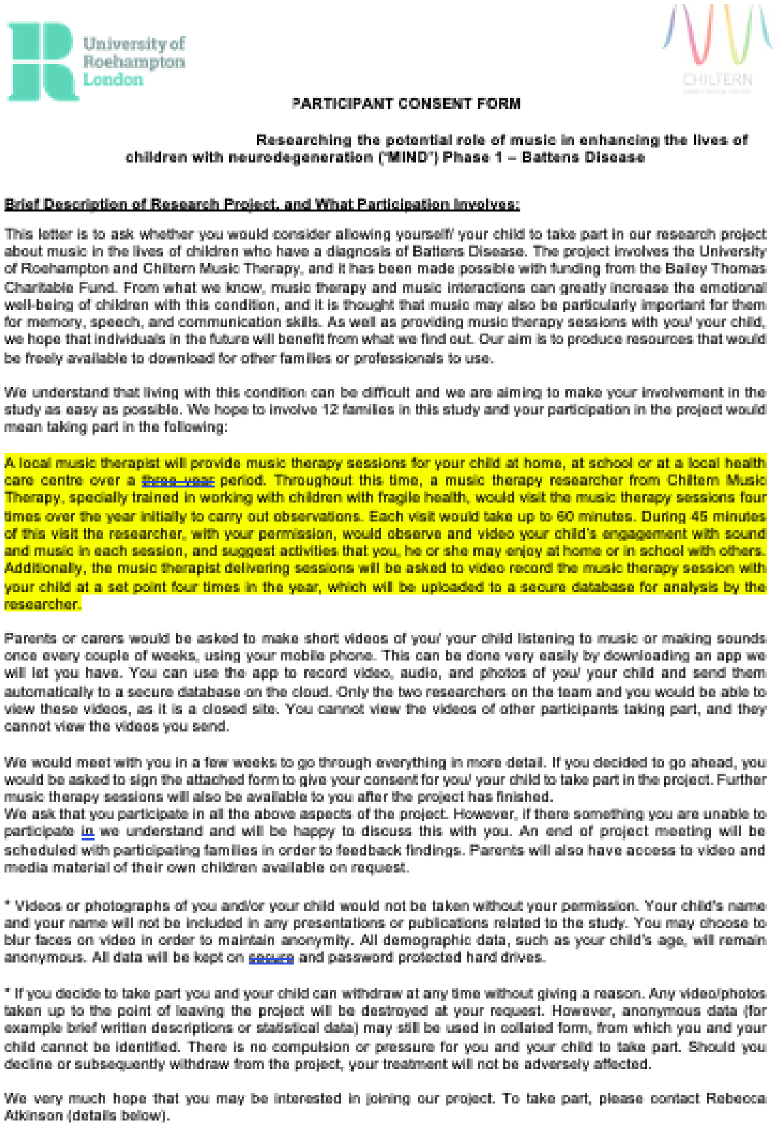
I agree to take part in this research, and am aware that I am free to withdraw at any point without giving a reason, although if I do so I understand that my data might still be used in a collated form. I understand that the information I provide will be treated in confidence by the investigator and that my identity will be protected in the publication of any findings, and that data will be collected and processed in accordance with the Data Protection Act 1998 and with the University's Data Protection Policy.

Name ……………………………………

Signature ………………………………

Date ……………………………………

Appendix 4: Phase One Parent Information and Consent Pack



University of Roehampton London

CHILTERN

PARTICIPANT CONSENT FORM

Researching the potential role of music in enhancing the lives of children with neurodegeneration ('MIND') Phase 1 – Battens Disease

Brief Description of Research Project, and What Participation Involves:

This letter is to ask whether you would consider allowing yourself/ your child to take part in our research project about music in the lives of children who have a diagnosis of Battens Disease. The project involves the University of Roehampton and Chiltern Music Therapy, and it has been made possible with funding from the Bailey Thomas Charitable Fund. From what we know, music therapy and music interactions can greatly increase the emotional well-being of children with this condition, and it is thought that music may also be particularly important for them for memory, speech, and communication skills. As well as providing music therapy sessions with you/ your child, we hope that individuals in the future will benefit from what we find out. Our aim is to produce resources that would be freely available to download for other families or professionals to use.

We understand that living with this condition can be difficult and we are aiming to make your involvement in the study as easy as possible. We hope to involve 12 families in this study and your participation in the project would mean taking part in the following:

A local music therapist will provide music therapy sessions for your child at home, at school or at a local health care centre over a three-year period. Throughout this time, a music therapy researcher from Chiltern Music Therapy, specially trained in working with children with fragile health, would visit the music therapy sessions four times over the year initially to carry out observations. Each visit would take up to 60 minutes. During 45 minutes of this visit the researcher, with your permission, would observe and video your child's engagement with sound and music in each session, and suggest activities that you, he or she may enjoy at home or in school with others. Additionally, the music therapist delivering sessions will be asked to video record the music therapy session with your child at a set point four times in the year, which will be uploaded to a secure database for analysis by the researcher.

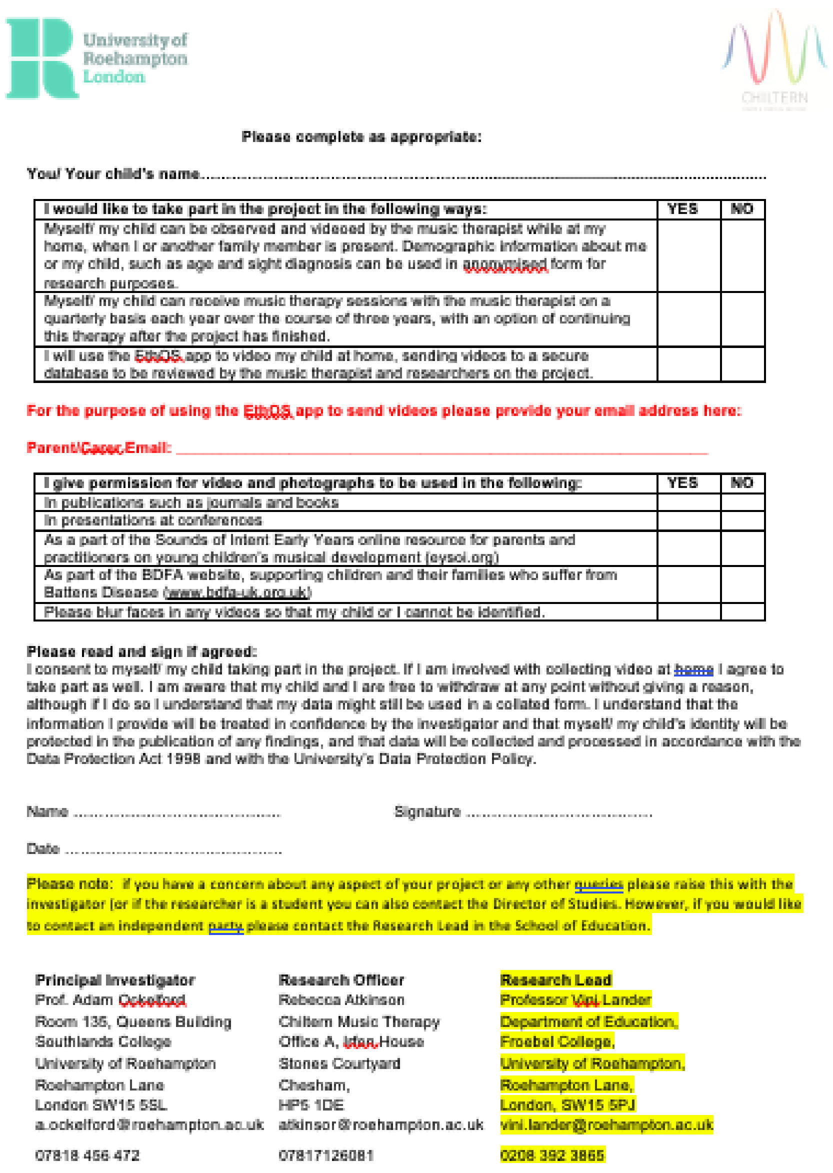
Parents or carers would be asked to make short videos of you/ your child listening to music or making sounds once every couple of weeks, using your mobile phone. This can be done very easily by downloading an app we will let you have. You can use the app to record video, audio, and photos of you/ your child and send them automatically to a secure database on the cloud. Only the two researchers on the team and you would be able to view these videos, as it is a closed site. You cannot view the videos of other participants taking part, and they cannot view the videos you send.

We would meet with you in a few weeks to go through everything in more detail. If you decided to go ahead, you would be asked to sign the attached form to give your consent for you/ your child to take part in the project. Further music therapy sessions will also be available to you after the project has finished.
We ask that you participate in all the above aspects of the project. However, if there something you are unable to participate in we understand and will be happy to discuss this with you. An end of project meeting will be scheduled with participating families in order to feedback findings. Parents will also have access to video and media material of their own children available on request.

* Videos or photographs of you and/or your child would not be taken without your permission. Your child's name and your name will not be included in any presentations or publications related to the study. You may choose to blur faces on video in order to maintain anonymity. All demographic data, such as your child's age, will remain anonymous. All data will be kept on secure and password protected hard drives.

* If you decide to take part you and your child can withdraw at any time without giving a reason. Any video/photos taken up to the point of leaving the project will be destroyed at your request. However, anonymous data (for example brief written descriptions or statistical data) may still be used in collated form, from which you and your child cannot be identified. There is no compulsion or pressure for you and your child to take part. Should you decline or subsequently withdraw from the project, your treatment will not be adversely affected.

We very much hope that you may be interested in joining our project. To take part, please contact Rebecca Atkinson (details below).



University of Roehampton London

CHILTERN

Please complete as appropriate:

You/ Your child's name...

I would like to take part in the project in the following ways:	YES	NO
Myself/ my child can be observed and videoed by the music therapist while at my home, when I or another family member is present. Demographic information about me or my child, such as age and sight diagnosis can be used in anonymised form for research purposes.		
Myself/ my child can receive music therapy sessions with the music therapist on a quarterly basis each year over the course of three years, with an option of continuing this therapy after the project has finished.		
I will use the EthOS app to video my child at home, sending videos to a secure database to be reviewed by the music therapist and researchers on the project.		

For the purpose of using the EthOS app to send videos please provide your email address here:

Parent/Carer Email: ____

I give permission for video and photographs to be used in the following:	YES	NO
In publications such as journals and books		
In presentations at conferences		
As a part of the Sounds of Intent Early Years online resource for parents and practitioners on young children's musical development (eysoi.org)		
As part of the BDFA website, supporting children and their families who suffer from Battens Disease (www.bdfa-uk.org.uk)		
Please blur faces in any videos so that my child or I cannot be identified.		

Please read and sign if agreed:
I consent to myself/ my child taking part in the project. If I am involved with collecting video at home I agree to take part as well. I am aware that my child and I are free to withdraw at any point without giving a reason, although if I do so I understand that my data might still be used in a collated form. I understand that the information I provide will be treated in confidence by the investigator and that myself/ my child's identity will be protected in the publication of any findings, and that data will be collected and processed in accordance with the Data Protection Act 1998 and with the University's Data Protection Policy.

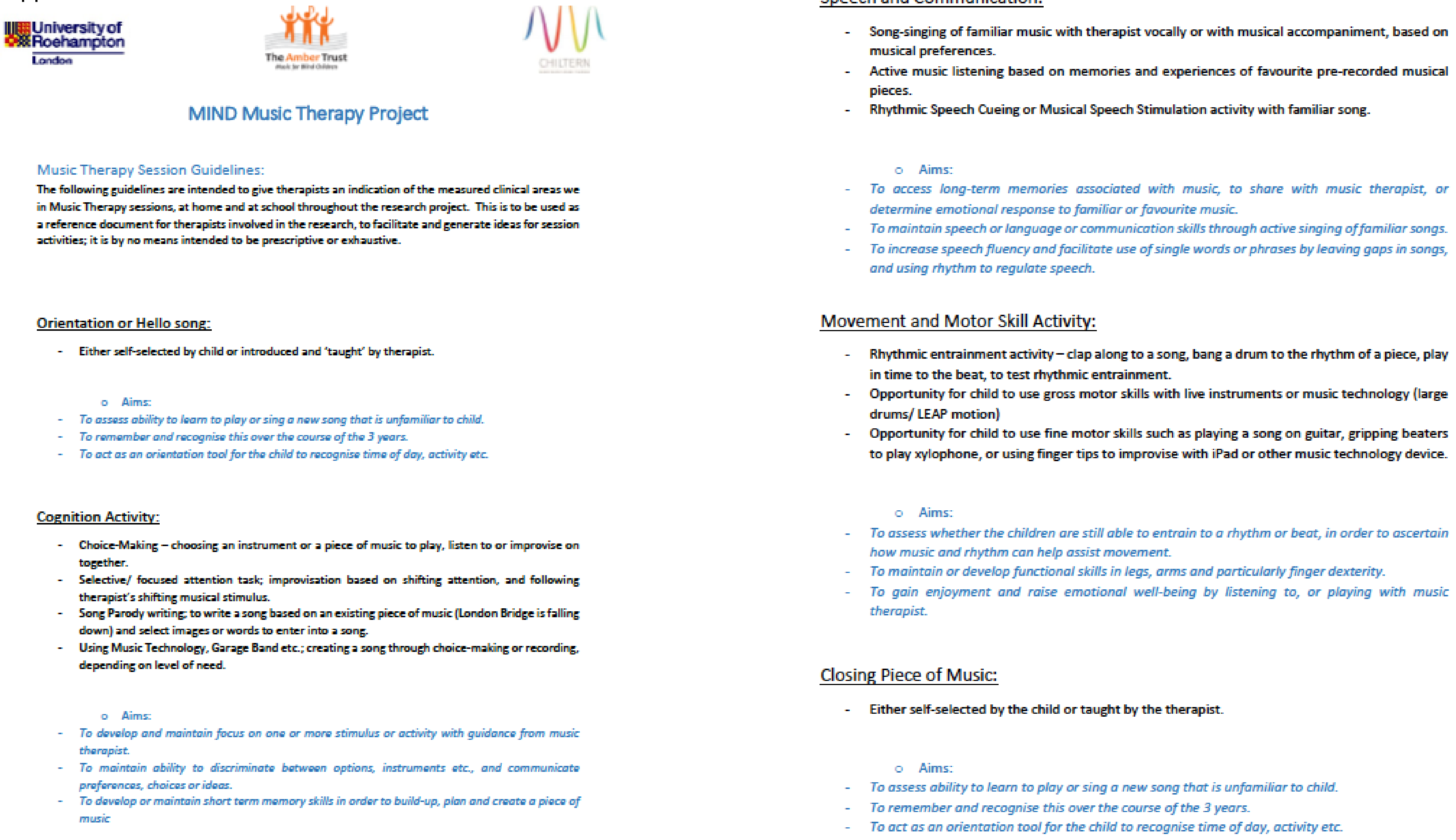
Name .... Signature ....

Date ....

Please note: if you have a concern about any aspect of your project or any other queries please raise this with the investigator (or if the researcher is a student you can also contact the Director of Studies. However, if you would like to contact an independent party please contact the Research Lead in the School of Education.

Principal Investigator	Research Officer	Research Lead
Prof. Adam Ockelford	Rebecca Atkinson	Professor Vini Lander
Room 135, Queens Building	Chiltern Music Therapy	Department of Education,
Southlands College	Office A, Idan House	Froebel College,
University of Roehampton	Stones Courtyard	University of Roehampton,
Roehampton Lane	Chesham,	Roehampton Lane,
London SW15 5SL	HP5 1DE	London, SW15 5PJ
a.ockelford@roehampton.ac.uk	atkinsor@roehampton.ac.uk	vini.lander@roehampton.ac.uk
07818 456 472	07817126081	0208 392 3865

Appendix 5: Session Guidance Notes



MIND Music Therapy Project

Music Therapy Session Guidelines:

The following guidelines are intended to give therapists an indication of the measured clinical areas we in Music Therapy sessions, at home and at school throughout the research project. This is to be used as a reference document for therapists involved in the research, to facilitate and generate ideas for session activities; it is by no means intended to be prescriptive or exhaustive.

Orientation or Hello song:

- Either self-selected by child or introduced and 'taught' by therapist.

  - Aims:
- *To assess ability to learn to play or sing a new song that is unfamiliar to child.*
- *To remember and recognise this over the course of the 3 years.*
- *To act as an orientation tool for the child to recognise time of day, activity etc.*

Cognition Activity:

- Choice-Making – choosing an instrument or a piece of music to play, listen to or improvise on together.
- Selective/ focused attention task; improvisation based on shifting attention, and following therapist's shifting musical stimulus.
- Song Parody writing; to write a song based on an existing piece of music (London Bridge is falling down) and select images or words to enter into a song.
- Using Music Technology, Garage Band etc.; creating a song through choice-making or recording, depending on level of need.

  - Aims:
- *To develop and maintain focus on one or more stimulus or activity with guidance from music therapist.*
- *To maintain ability to discriminate between options, instruments etc., and communicate preferences, choices or ideas.*
- *To develop or maintain short term memory skills in order to build-up, plan and create a piece of music*

Speech and Communication:

- Song-singing of familiar music with therapist vocally or with musical accompaniment, based on musical preferences.
- Active music listening based on memories and experiences of favourite pre-recorded musical pieces.
- Rhythmic Speech Cueing or Musical Speech Stimulation activity with familiar song.

  - Aims:
- *To access long-term memories associated with music, to share with music therapist, or determine emotional response to familiar or favourite music.*
- *To maintain speech or language or communication skills through active singing of familiar songs.*
- *To increase speech fluency and facilitate use of single words or phrases by leaving gaps in songs, and using rhythm to regulate speech.*

Movement and Motor Skill Activity:

- Rhythmic entrainment activity – clap along to a song, bang a drum to the rhythm of a piece, play in time to the beat, to test rhythmic entrainment.
- Opportunity for child to use gross motor skills with live instruments or music technology (large drums/ LEAP motion)
- Opportunity for child to use fine motor skills such as playing a song on guitar, gripping beaters to play xylophone, or using finger tips to improvise with iPad or other music technology device.

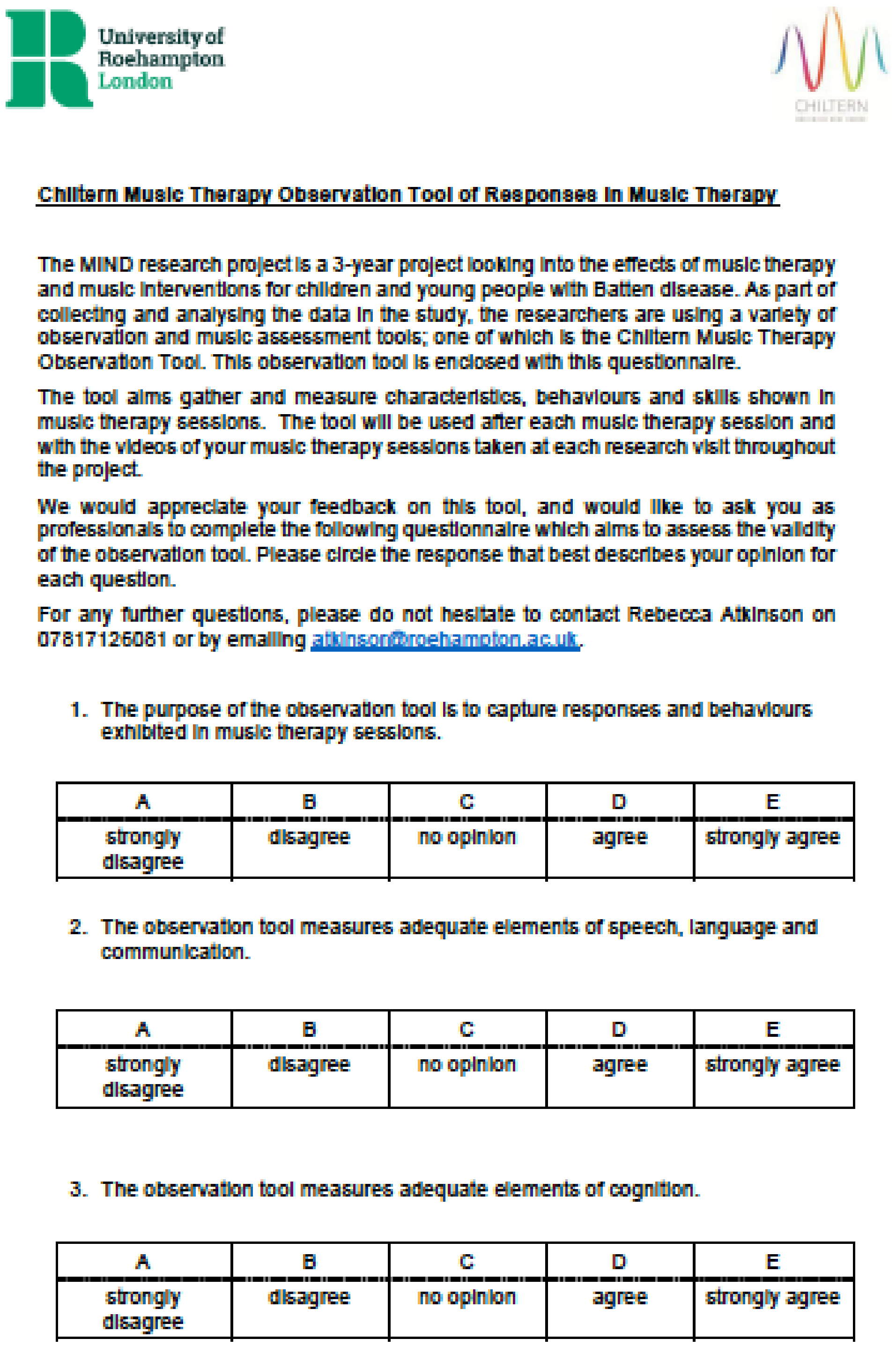
  - Aims:
- *To assess whether the children are still able to entrain to a rhythm or beat, in order to ascertain how music and rhythm can help assist movement.*
- *To maintain or develop functional skills in legs, arms and particularly finger dexterity.*
- *To gain enjoyment and raise emotional well-being by listening to, or playing with music therapist.*

Closing Piece of Music:

- Either self-selected by the child or taught by the therapist.

  - Aims:
- *To assess ability to learn to play or sing a new song that is unfamiliar to child.*
- *To remember and recognise this over the course of the 3 years.*
- *To act as an orientation tool for the child to recognise time of day, activity etc.*

Appendix 6: Face Validity Practitioner Questionnaire



University of Roehampton London

CHILTERN

Chiltern Music Therapy Observation Tool of Responses in Music Therapy

The MIND research project is a 3-year project looking into the effects of music therapy and music interventions for children and young people with Batten disease. As part of collecting and analysing the data in the study, the researchers are using a variety of observation and music assessment tools; one of which is the Chiltern Music Therapy Observation Tool. This observation tool is enclosed with this questionnaire.

The tool aims gather and measure characteristics, behaviours and skills shown in music therapy sessions. The tool will be used after each music therapy session and with the videos of your music therapy sessions taken at each research visit throughout the project.

We would appreciate your feedback on this tool, and would like to ask you as professionals to complete the following questionnaire which aims to assess the validity of the observation tool. Please circle the response that best describes your opinion for each question.

For any further questions, please do not hesitate to contact Rebecca Atkinson on 07817126081 or by emailing atkinson@roehampton.ac.uk.

1. The purpose of the observation tool is to capture responses and behaviours exhibited in music therapy sessions.

A	B	C	D	E
strongly disagree	disagree	no opinion	agree	strongly agree

2. The observation tool measures adequate elements of speech, language and communication.

A	B	C	D	E
strongly disagree	disagree	no opinion	agree	strongly agree

3. The observation tool measures adequate elements of cognition.

A	B	C	D	E
strongly disagree	disagree	no opinion	agree	strongly agree

4. The observation tool measures adequate elements of physical presentation.

A	B	C	D	E
strongly disagree	disagree	no opinion	agree	strongly agree

5. The observation tool measures adequate elements of emotional well-being.

A	B	C	D	E
strongly disagree	disagree	no opinion	agree	strongly agree

6. The observation tool is an adequate complimentary measure to the Sounds of Intent Framework capturing responses of children and young people with Batten disease in music therapy sessions.

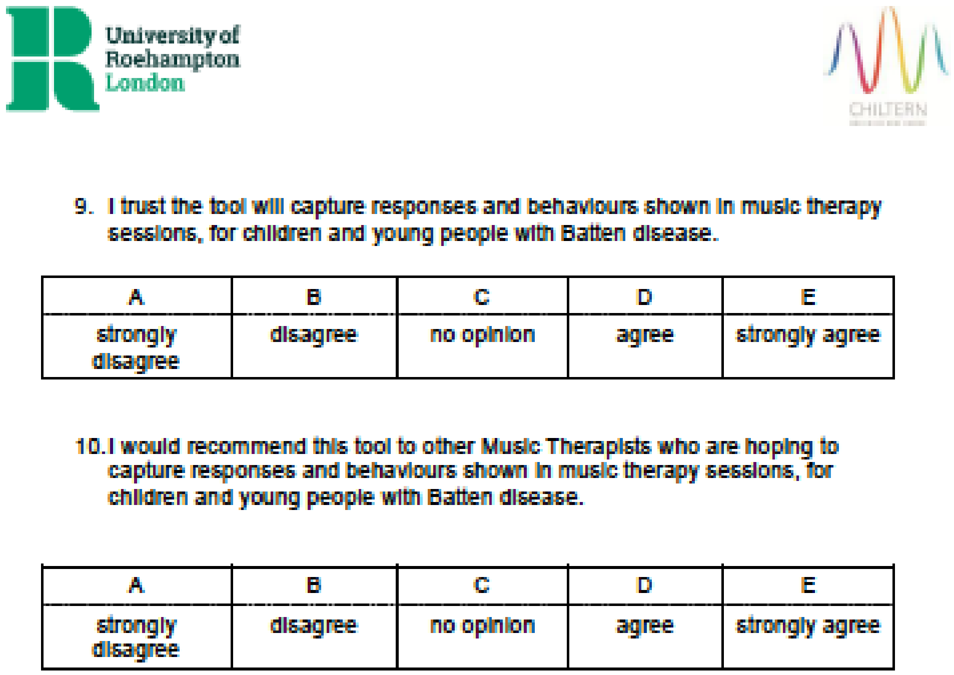
A	B	C	D	E
strongly disagree	disagree	no opinion	agree	strongly agree

7. This observation tool requires the music therapist to undergo training before using this scale.

A	B	C	D	E
strongly disagree	disagree	no opinion	agree	strongly agree

8. This observation tool is an adequate measure for the retrospective analysis of music therapy filmed sessions

A	B	C	D	E
strongly disagree	disagree	no opinion	agree	strongly agree



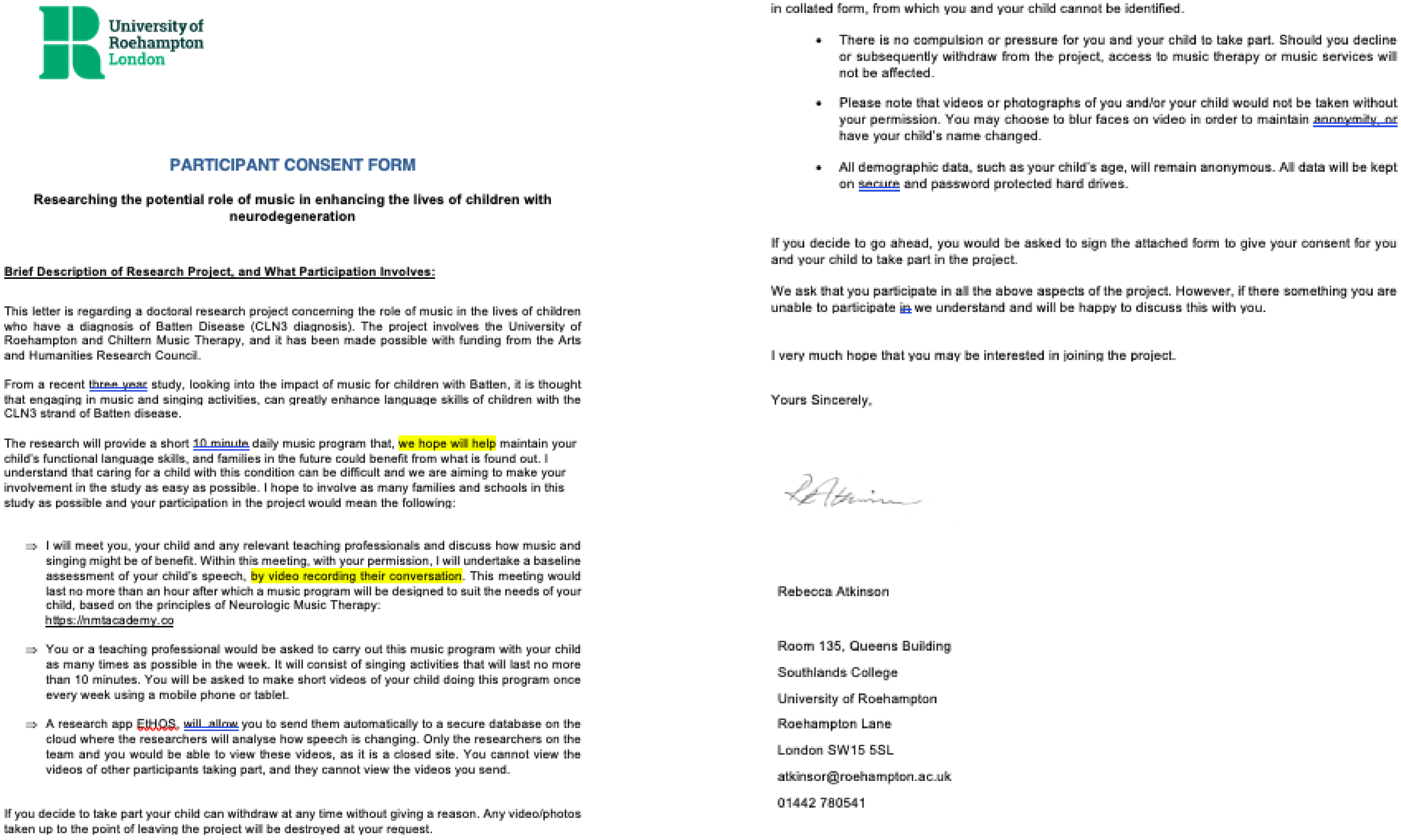
9. I trust the tool will capture responses and behaviours shown in music therapy sessions, for children and young people with Batten disease.

A	B	C	D	E
strongly disagree	disagree	no opinion	agree	strongly agree

10. I would recommend this tool to other Music Therapists who are hoping to capture responses and behaviours shown in music therapy sessions, for children and young people with Batten disease.

A	B	C	D	E
strongly disagree	disagree	no opinion	agree	strongly agree

Appendix 7: Phase Two Parent Information and Consent Form



University of Roehampton London

PARTICIPANT CONSENT FORM

Researching the potential role of music in enhancing the lives of children with neurodegeneration

Brief Description of Research Project, and What Participation Involves:

This letter is regarding a doctoral research project concerning the role of music in the lives of children who have a diagnosis of Batten Disease (CLN3 diagnosis). The project involves the University of Roehampton and Chiltern Music Therapy, and it has been made possible with funding from the Arts and Humanities Research Council.

From a recent three year study, looking into the impact of music for children with Batten, it is thought that engaging in music and singing activities, can greatly enhance language skills of children with the CLN3 strand of Batten disease.

The research will provide a short 10 minute daily music program that, we hope will help maintain your child's functional language skills, and families in the future could benefit from what is found out. I understand that caring for a child with this condition can be difficult and we are aiming to make your involvement in the study as easy as possible. I hope to involve as many families and schools in this study as possible and your participation in the project would mean the following:

- ⇨ I will meet you, your child and any relevant teaching professionals and discuss how music and singing might be of benefit. Within this meeting, with your permission, I will undertake a baseline assessment of your child's speech, by video recording their conversation. This meeting would last no more than an hour after which a music program will be designed to suit the needs of your child, based on the principles of Neurologic Music Therapy: https://nmtacademy.co
- ⇨ You or a teaching professional would be asked to carry out this music program with your child as many times as possible in the week. It will consist of singing activities that will last no more than 10 minutes. You will be asked to make short videos of your child doing this program once every week using a mobile phone or tablet.
- ⇨ A research app EIHOS, will allow you to send them automatically to a secure database on the cloud where the researchers will analyse how speech is changing. Only the researchers on the team and you would be able to view these videos, as it is a closed site. You cannot view the videos of other participants taking part, and they cannot view the videos you send.

If you decide to take part your child can withdraw at any time without giving a reason. Any video/photos taken up to the point of leaving the project will be destroyed at your request.

However, anonymous data (for example brief written descriptions or statistical data) may still be used in collated form, from which you and your child cannot be identified.

- There is no compulsion or pressure for you and your child to take part. Should you decline or subsequently withdraw from the project, access to music therapy or music services will not be affected.
- Please note that videos or photographs of you and/or your child would not be taken without your permission. You may choose to blur faces on video in order to maintain anonymity, or have your child's name changed.
- All demographic data, such as your child's age, will remain anonymous. All data will be kept on secure and password protected hard drives.

If you decide to go ahead, you would be asked to sign the attached form to give your consent for you and your child to take part in the project.

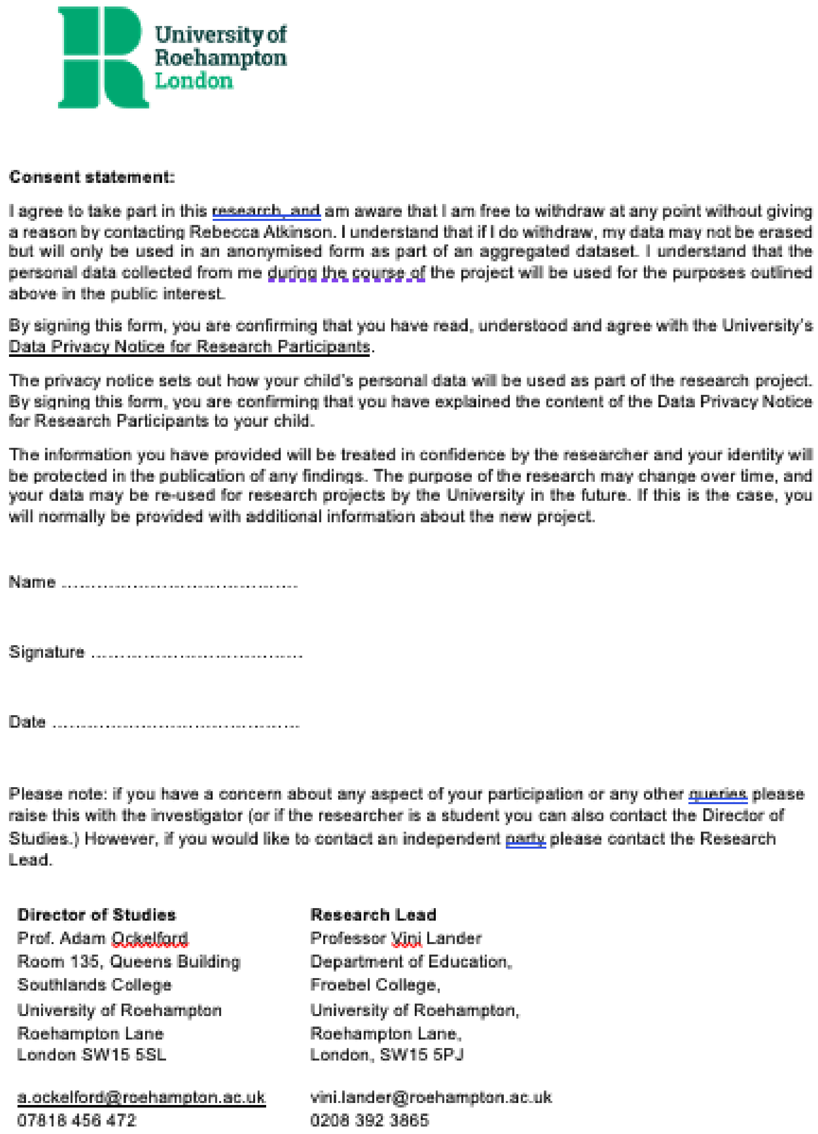
We ask that you participate in all the above aspects of the project. However, if there something you are unable to participate in we understand and will be happy to discuss this with you.

I very much hope that you may be interested in joining the project.

Yours Sincerely,

Rebecca Atkinson

Room 135, Queens Building
Southlands College
University of Roehampton
Roehampton Lane
London SW15 5SL
atkinsor@roehampton.ac.uk
01442 780541



University of
Roehampton
London

Consent statement:

I agree to take part in this research, and am aware that I am free to withdraw at any point without giving a reason by contacting Rebecca Atkinson. I understand that if I do withdraw, my data may not be erased but will only be used in an anonymised form as part of an aggregated dataset. I understand that the personal data collected from me during the course of the project will be used for the purposes outlined above in the public interest.

By signing this form, you are confirming that you have read, understood and agree with the University's Data Privacy Notice for Research Participants.

The privacy notice sets out how your child's personal data will be used as part of the research project. By signing this form, you are confirming that you have explained the content of the Data Privacy Notice for Research Participants to your child.

The information you have provided will be treated in confidence by the researcher and your identity will be protected in the publication of any findings. The purpose of the research may change over time, and your data may be re-used for research projects by the University in the future. If this is the case, you will normally be provided with additional information about the new project.

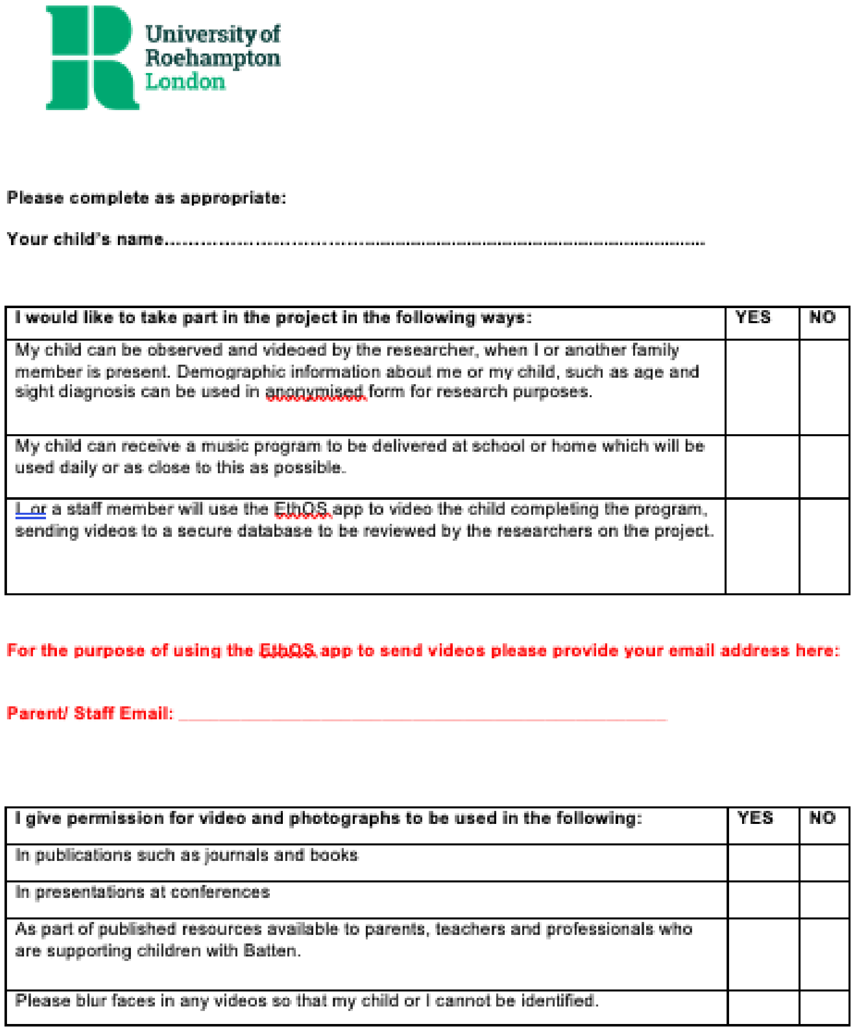
Name ……………………………………

Signature ……………………………….

Date ……………………………………..

Please note: if you have a concern about any aspect of your participation or any other queries please raise this with the investigator (or if the researcher is a student you can also contact the Director of Studies.) However, if you would like to contact an independent party please contact the Research Lead.

Director of Studies	**Research Lead**
Prof. Adam Ockelford Room 135, Queens Building Southlands College University of Roehampton Roehampton Lane London SW15 5SL	Professor Vini Lander Department of Education, Froebel College, University of Roehampton, Roehampton Lane, London, SW15 5PJ
a.ockelford@roehampton.ac.uk 07818 456 472	vini.lander@roehampton.ac.uk 0208 392 3865



University of
Roehampton
London

Please complete as appropriate:

Your child's name………………………………………………………………………………………

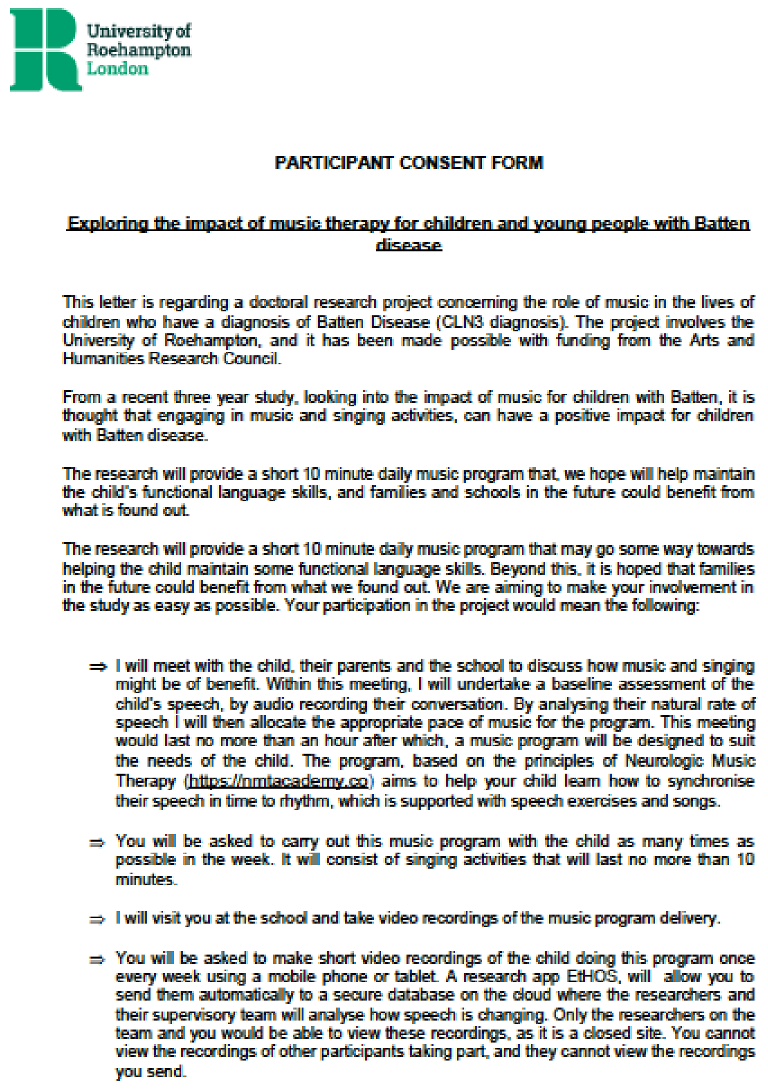
I would like to take part in the project in the following ways:	**YES**	**NO**
My child can be observed and videoed by the researcher, when I or another family member is present. Demographic information about me or my child, such as age and sight diagnosis can be used in anonymised form for research purposes.		
My child can receive a music program to be delivered at school or home which will be used daily or as close to this as possible.		
I or a staff member will use the EthOS app to video the child completing the program, sending videos to a secure database to be reviewed by the researchers on the project.		

For the purpose of using the EthOS app to send videos please provide your email address here:

Parent/ Staff Email: ____

I give permission for video and photographs to be used in the following:	**YES**	**NO**
In publications such as journals and books		
In presentations at conferences		
As part of published resources available to parents, teachers and professionals who are supporting children with Batten.		
Please blur faces in any videos so that my child or I cannot be identified.		

Appendix 8: Phase Two Staff Information and Consent Form



PARTICIPANT CONSENT FORM

Exploring the impact of music therapy for children and young people with Batten disease

This letter is regarding a doctoral research project concerning the role of music in the lives of children who have a diagnosis of Batten Disease (CLN3 diagnosis). The project involves the University of Roehampton, and it has been made possible with funding from the Arts and Humanities Research Council.

From a recent three year study, looking into the impact of music for children with Batten, it is thought that engaging in music and singing activities, can have a positive impact for children with Batten disease.

The research will provide a short 10 minute daily music program that, we hope will help maintain the child's functional language skills, and families and schools in the future could benefit from what is found out.

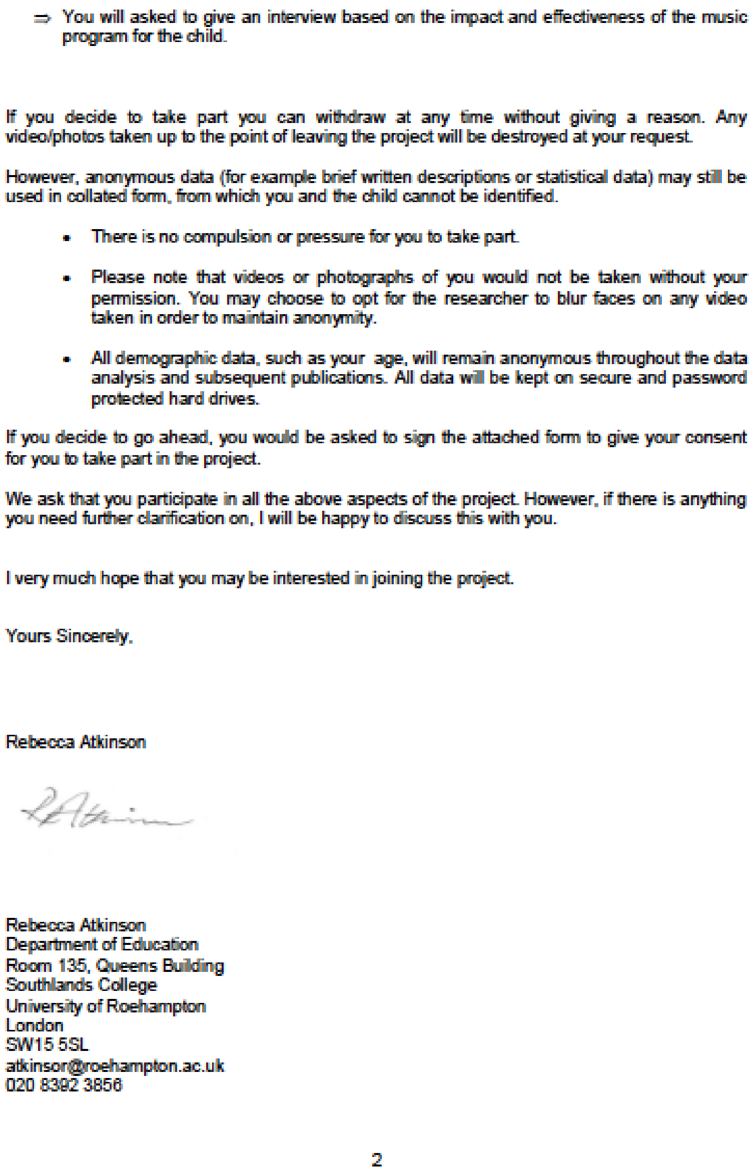
The research will provide a short 10 minute daily music program that may go some way towards helping the child maintain some functional language skills. Beyond this, it is hoped that families in the future could benefit from what we found out. We are aiming to make your involvement in the study as easy as possible. Your participation in the project would mean the following:

⇒ I will meet with the child, their parents and the school to discuss how music and singing might be of benefit. Within this meeting, I will undertake a baseline assessment of the child's speech, by audio recording their conversation. By analysing their natural rate of speech I will then allocate the appropriate pace of music for the program. This meeting would last no more than an hour after which, a music program will be designed to suit the needs of the child. The program, based on the principles of Neurologic Music Therapy (https://nmtacademy.co) aims to help your child learn how to synchronise their speech in time to rhythm, which is supported with speech exercises and songs.

⇒ You will be asked to carry out this music program with the child as many times as possible in the week. It will consist of singing activities that will last no more than 10 minutes.

⇒ I will visit you at the school and take video recordings of the music program delivery.

⇒ You will be asked to make short video recordings of the child doing this program once every week using a mobile phone or tablet. A research app EtHOS, will allow you to send them automatically to a secure database on the cloud where the researchers and their supervisory team will analyse how speech is changing. Only the researchers on the team and you would be able to view these recordings, as it is a closed site. You cannot view the recordings of other participants taking part, and they cannot view the recordings you send.



⇒ You will asked to give an interview based on the impact and effectiveness of the music program for the child.

If you decide to take part you can withdraw at any time without giving a reason. Any video/photos taken up to the point of leaving the project will be destroyed at your request.

However, anonymous data (for example brief written descriptions or statistical data) may still be used in collated form, from which you and the child cannot be identified.

- There is no compulsion or pressure for you to take part.
- Please note that videos or photographs of you would not be taken without your permission. You may choose to opt for the researcher to blur faces on any video taken in order to maintain anonymity.
- All demographic data, such as your age, will remain anonymous throughout the data analysis and subsequent publications. All data will be kept on secure and password protected hard drives.

If you decide to go ahead, you would be asked to sign the attached form to give your consent for you to take part in the project.

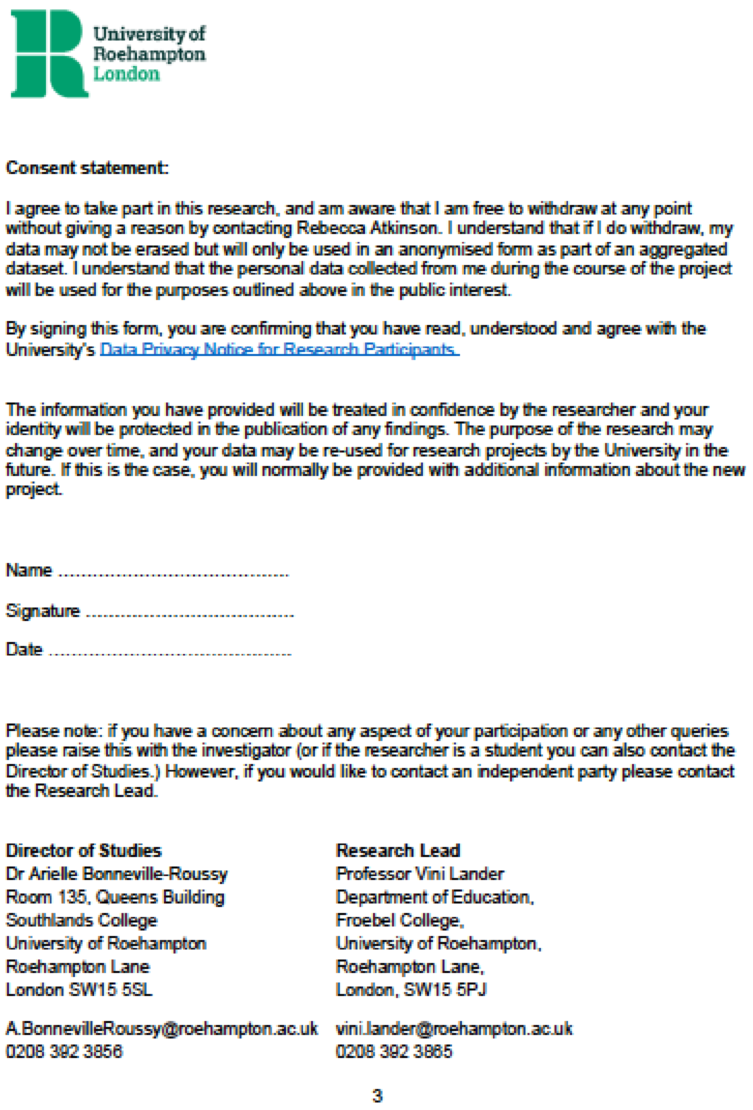
We ask that you participate in all the above aspects of the project. However, if there is anything you need further clarification on, I will be happy to discuss this with you.

I very much hope that you may be interested in joining the project.

Yours Sincerely,

Rebecca Atkinson

Rebecca Atkinson
Department of Education
Room 135, Queens Building
Southlands College
University of Roehampton
London
SW15 5SL
atkinsor@roehampton.ac.uk
020 8392 3856



University of Roehampton London

Consent statement:

I agree to take part in this research, and am aware that I am free to withdraw at any point without giving a reason by contacting Rebecca Atkinson. I understand that if I do withdraw, my data may not be erased but will only be used in an anonymised form as part of an aggregated dataset. I understand that the personal data collected from me during the course of the project will be used for the purposes outlined above in the public interest.

By signing this form, you are confirming that you have read, understood and agree with the University's [Data Privacy Notice for Research Participants.]()

The information you have provided will be treated in confidence by the researcher and your identity will be protected in the publication of any findings. The purpose of the research may change over time, and your data may be re-used for research projects by the University in the future. If this is the case, you will normally be provided with additional information about the new project.

Name ……………………………………

Signature …………………………………

Date ………………………………………

Please note: if you have a concern about any aspect of your participation or any other queries please raise this with the investigator (or if the researcher is a student you can also contact the Director of Studies.) However, if you would like to contact an independent party please contact the Research Lead.

Director of Studies
Dr Arielle Bonneville-Roussy
Room 135, Queens Building
Southlands College
University of Roehampton
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London SW15 5SL

A.BonnevilleRoussy@roehampton.ac.uk
0208 392 3856

Research Lead
Professor Vini Lander
Department of Education,
Froebel College,
University of Roehampton,
Roehampton Lane,
London, SW15 5PJ

vini.lander@roehampton.ac.uk
0208 392 3865

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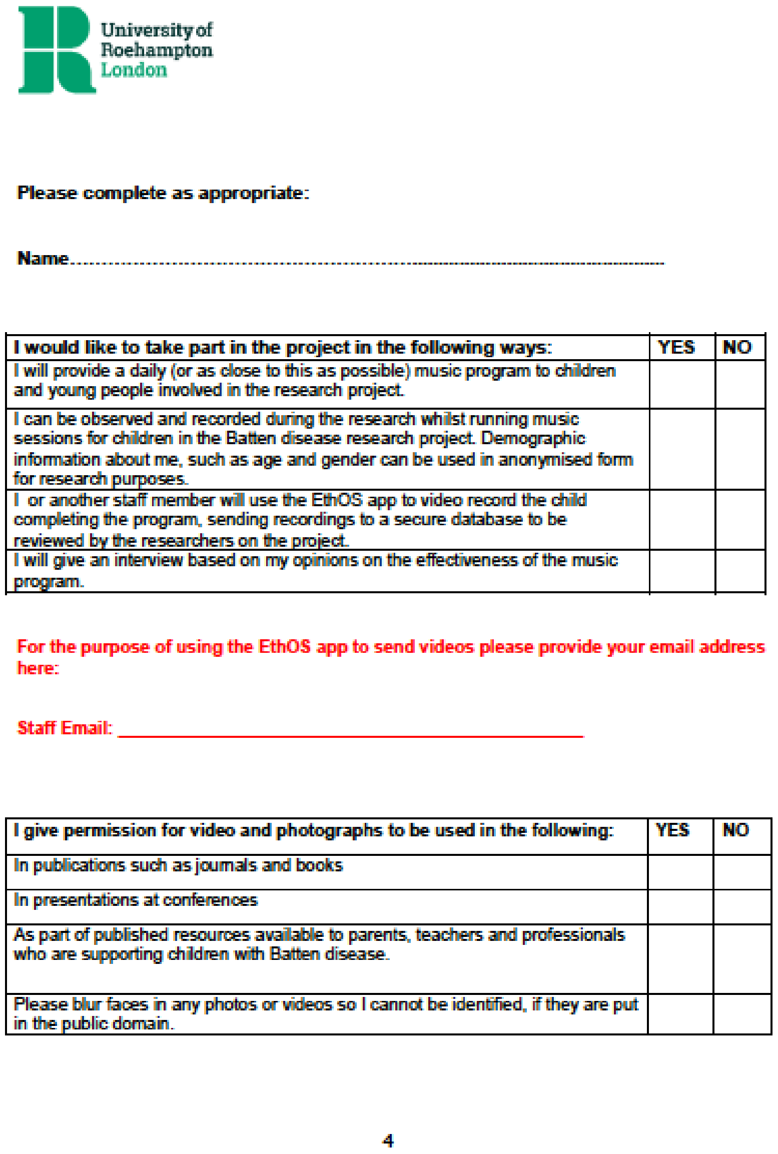


University of Roehampton London

Please complete as appropriate:

Name……………………………………………………………………………………………

I would like to take part in the project in the following ways:	**YES**	**NO**
I will provide a daily (or as close to this as possible) music program to children and young people involved in the research project.		
I can be observed and recorded during the research whilst running music sessions for children in the Batten disease research project. Demographic information about me, such as age and gender can be used in anonymised form for research purposes.		
I or another staff member will use the EthOS app to video record the child completing the program, sending recordings to a secure database to be reviewed by the researchers on the project.		
I will give an interview based on my opinions on the effectiveness of the music program.		

For the purpose of using the EthOS app to send videos please provide your email address here:

Staff Email: ______________________________

I give permission for video and photographs to be used in the following:	**YES**	**NO**
In publications such as journals and books		
In presentations at conferences		
As part of published resources available to parents, teachers and professionals who are supporting children with Batten disease.		
Please blur faces in any photos or videos so I cannot be identified, if they are put in the public domain.		

4

Appendix 9: Music Speaks Program

MUSIC SPEAKS PROGRAM

University of
Roehampton
London

OVERVIEW

The following daily exercises have been designed to help support speech and language through music for the young person you are working with. The following activities aim to work on aspects of speech, communication, articulation, and pace of word formation. Please record this Program once a week when requested and upload using the EthOS research platform.

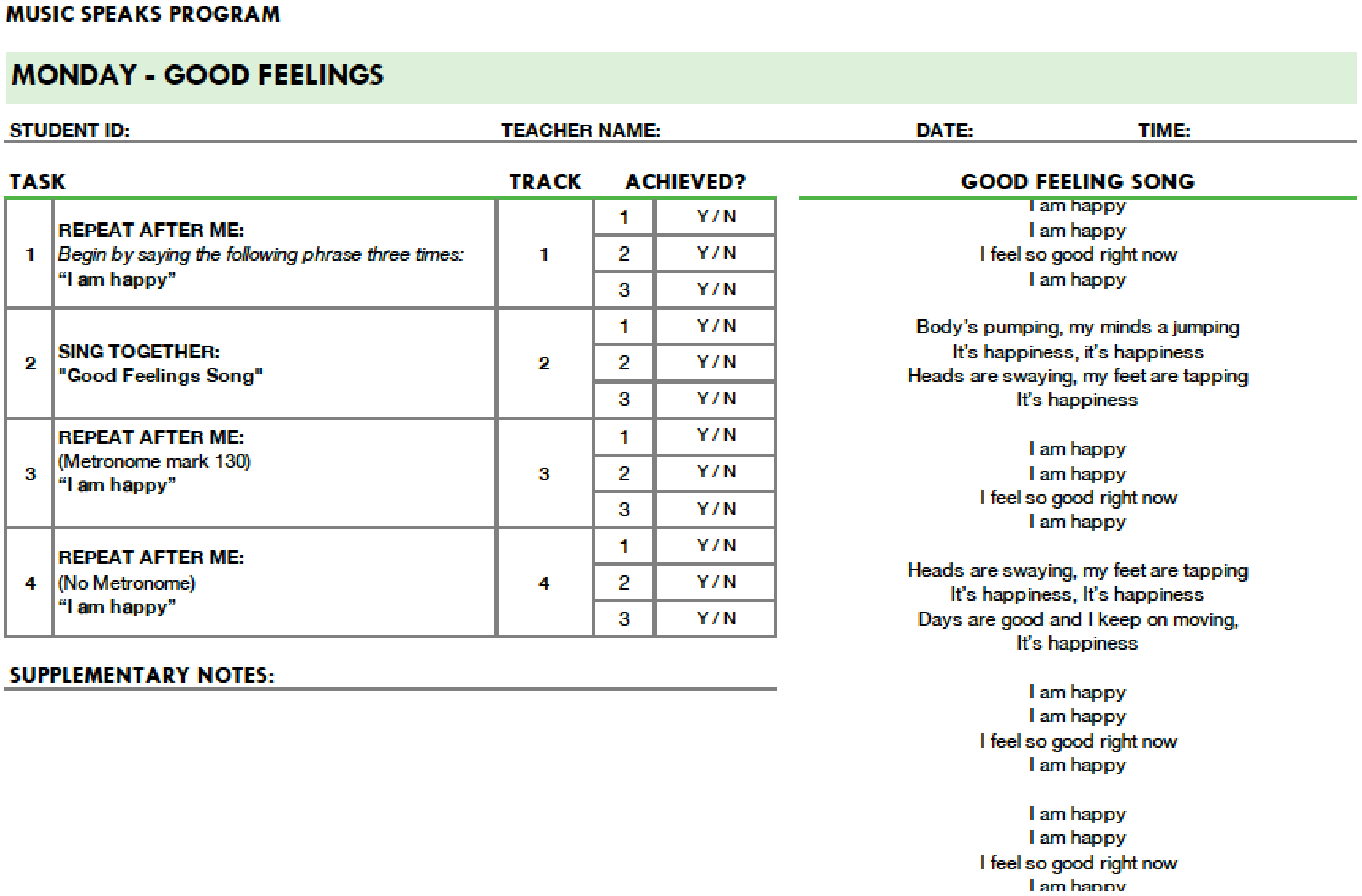
NEUROLOGIC MUSIC THERAPY

Neurologic Music Therapy 'NMT' is defined as the application of music to cognitive, sensory, and motor dysfunctions (Thaut, 2014). It is based on the neuroscience model that music perception and production has an influence on functional changes in the brain. The music and language activities outlined in this programme use some of the techniques used in NMT as highlighted in the list below and activities here are given as an example of the more extensive activities and tasks that can be carried out in music therapy sessions.

TECHNIQUES USED IN THIS PROGRAM

Rhythmic Speech Cueing (RSC)
Music Speech Stimulation (MUSTIM)

*For more infromation about NMT please visit https://nmtacademy.co/home/clinic/



MUSIC SPEAKS PROGRAM

MONDAY - GOOD FEELINGS

STUDENT ID: **TEACHER NAME:** **DATE:** **TIME:**

	TASK	TRACK	ACHIEVED?	
1	**REPEAT AFTER ME:** *Begin by saying the following phrase three times:* **"I am happy"**	1	1	Y / N
			2	Y / N
			3	Y / N
2	**SING TOGETHER:** **"Good Feelings Song"**	2	1	Y / N
			2	Y / N
			3	Y / N
3	**REPEAT AFTER ME:** (Metronome mark 130) **"I am happy"**	3	1	Y / N
			2	Y / N
			3	Y / N
4	**REPEAT AFTER ME:** (No Metronome) **"I am happy"**	4	1	Y / N
			2	Y / N
			3	Y / N

SUPPLEMENTARY NOTES:

GOOD FEELING SONG

I am happy
I am happy
I feel so good right now
I am happy

Body's pumping, my minds a jumping
It's happiness, it's happiness
Heads are swaying, my feet are tapping
It's happiness

I am happy
I am happy
I feel so good right now
I am happy

Heads are swaying, my feet are tapping
It's happiness, It's happiness
Days are good and I keep on moving,
It's happiness

I am happy
I am happy
I feel so good right now
I am happy

I am happy
I am happy
I feel so good right now
I am happy

MUSIC SPEAKS PROGRAM

TUESDAY - BAD FEELINGS

STUDENT ID: **TEACHER NAME:** **DATE:** **TIME:**

TASK		TRACK	ACHIEVED?	
1	**REPEAT AFTER ME:** *Begin by saying the following phrase three times:* **"I need to rest, I feel tired"**	1	1	Y / N
			2	Y / N
			3	Y / N
2	**SING TOGETHER:** **"Bad Feelings Song"**	2	1	Y / N
			2	Y / N
			3	Y / N
3	**REPEAT AFTER ME:** (Metronome mark 90) **"I need to rest, I feel tired"**	3	1	Y / N
			2	Y / N
			3	Y / N
4	**REPEAT AFTER ME:** (No Metronome) **"I need to rest, I feel tired"**	4	1	Y / N
			2	Y / N
			3	Y / N

SUPPLEMENTARY NOTES:

BAD FEELING SONG

I feel tired, I feel tired
I don't feel so good
I feel tired
I need to rest, I feel tired

I feel pain, I feel pain
I don't feel so good
I feel pain
I need to rest, I feel pain

I feel tired, I feel tired
I don't feel so good
I feel tired
I need to rest, I feel tired

MUSIC SPEAKS PROGRAM

WEDNESDAY - ACTIVITIES

STUDENT ID: **TEACHER NAME:** **DATE:** **TIME:**

	TASK	TRACK	ACHIEVED?	
1	**REPEAT AFTER ME:** *Begin by saying the following phrase three times:* **“Let's play music, let's play music”**	1	1	Y / N
			2	Y / N
			3	Y / N
2	**SING TOGETHER:** **"Activities Song"**	2	1	Y / N
			2	Y / N
			3	Y / N
3	**REPEAT AFTER ME:** (Metronome mark 120) **“Let's play music, let's play music”**	3	1	Y / N
			2	Y / N
			3	Y / N
4	**REPEAT AFTER ME:** (No Metronome) **“Let's play music, let's play music”**	4	1	Y / N
			2	Y / N
			3	Y / N

SUPPLEMENTARY NOTES:

ACTIVITIES SONG

Let’s go dancing, let’s go dancing
Take me now let’s go
Let’s go dancing, let’s go dancing
Take me now let’s go

I’m up and ready, and I’m feeling steady,
There’s no stopping me,
It’s so much fun, yeah with everyone,
It’s where I want to be,

Let’s play music, Let’s play music
Take me now let’s go
Let’s play music, Let’s play music
Take me now let’s go

I’m up and ready, and I’m feeling steady,
There’s no stopping me,
It’s so much fun, yeah with everyone,
It’s where I want to be

Let’s go dancing, let’s go dancing
Take me now let’s go
Let’s go dancing, let’s go dancing
Take me now let’s go

Let’s play music, Let’s play music
Take me now let’s go
Let’s play music, Let’s play music
Take me now let’s go

MUSIC SPEAKS PROGRAM

THURSDAY - PEOPLE

STUDENT ID: **TEACHER NAME:** **DATE:** **TIME:**

	TASK	**TRACK**	**ACHIEVED?**	
1	**REPEAT AFTER ME:** *Begin by saying the following phrase three times:* **“I want to see Mummy”**	1	1	Y / N
			2	Y / N
			3	Y / N
2	**SING TOGETHER:** **"People Song"**	2	1	Y / N
			2	Y / N
			3	Y / N
3	**REPEAT AFTER ME:** (Metronome mark 120) **“I want to see Mummy”**	3	1	Y / N
			2	Y / N
			3	Y / N
4	**REPEAT AFTER ME:** (No Metronome) **“I want to see Mummy”**	4	1	Y / N
			2	Y / N
			3	Y / N

SUPPLEMENTARY NOTES:

PEOPLE SONG

I want to see Mummy
I want to see Mummy
I want to see Mummy
Let’s go and her now

Being friends together is just so much fun,
I’m making memories with you
Being friends together is just so much fun,
We’re making memories us two

I want to see Daddy
I want to see Daddy
I want to see Daddy
Let’s go and see them now

Spending time together means so much to me
I’m making memories with you
Spending time together means so much to me
We’re making memories us two

I want to see Jasmine
I want to see Jasmine
I want to see Jasmine
Let’s go and see them now

I want to see Uncle Dean
I want to see Uncle Dean
I want to see Uncle Dean
Let’s go and see them now

MUSIC SPEAKS PROGRAM

FRIDAY - FOOD AND DRINK

STUDENT ID: **TEACHER NAME:** **DATE:** **TIME:**

TASK		TRACK	ACHIEVED?	
1	**REPEAT AFTER ME:** *Begin by saying the following phrase three times:* **"Can I have some food? Can I have a drink?"**	1	1	Y / N
			2	Y / N
			3	Y / N
2	**SING TOGETHER:** **"Food and Drink Song"**	2	1	Y / N
			2	Y / N
			3	Y / N
3	**REPEAT AFTER ME:** (Metronome mark 110) **"Can I have some food? Can I have a drink?"**	3	1	Y / N
			2	Y / N
			3	Y / N
4	**REPEAT AFTER ME:** (No Metronome) **"Can I have some food? Can I have a drink?"**	4	1	Y / N
			2	Y / N
			3	Y / N

SUPPLEMENTARY NOTES:

FOOD AND DRINK SONG

There's a rumble in my tummy and I want to eat,
Let's get something now for a treat
There's a rumble in my tummy and I know what to do,
Let's go to the kitchen, yeah me and you

Pizza I like to eat, Pizza
Can I have some food?
Pizza I like to eat, Pizza
Can I have some food?

I'm feeling so thirsty and my mouth is dry
"I know what you need" yeah, I hear you cry!
I'm feeling so thirsty and my mouth is dry,
A drink is what I need to satisfy

Lemonade I like to drink, Lemonade
Can I have a drink?
Lemonade I like to drink, Lemonade
Can I have a drink?

Pizza I like to eat, Pizza
Can I have some food?
Pizza I like to eat, Pizza
Can I have some food?

Lemonade I like to drink, Lemonade
Can I have a drink?
Lemonade I like to drink, Lemonade
Can I have a drink?
Right now!

Appendix 10: Covid-19 Mitigation and Data Retrieval Protocol

Research Adaptation Considerations

Data collection protocol

Video Data		Data Sheets	
	Options		Options
1	Upload to EthOS app/ website	5	School to scan and send the password protected sheets over email and password to be sent in a separate email to student email address
2	Check school status around using an alternative data transfer platform (One Drive, We transfer, G Drive)	6	Rebecca to email over template electronic data sheets for data to be transferred onto, password protected and emailed back
3	Arrange an online meeting with IT or school personel to arrange transfer	7	Data sheets are sent recorded delivery to university (no identifying information)
4	Physically visit the school to transfer videos using airdrop method or cable to mac	8	Physically retrieve the data sheets from schools adhering to social distancing and COVID specific health guidance.

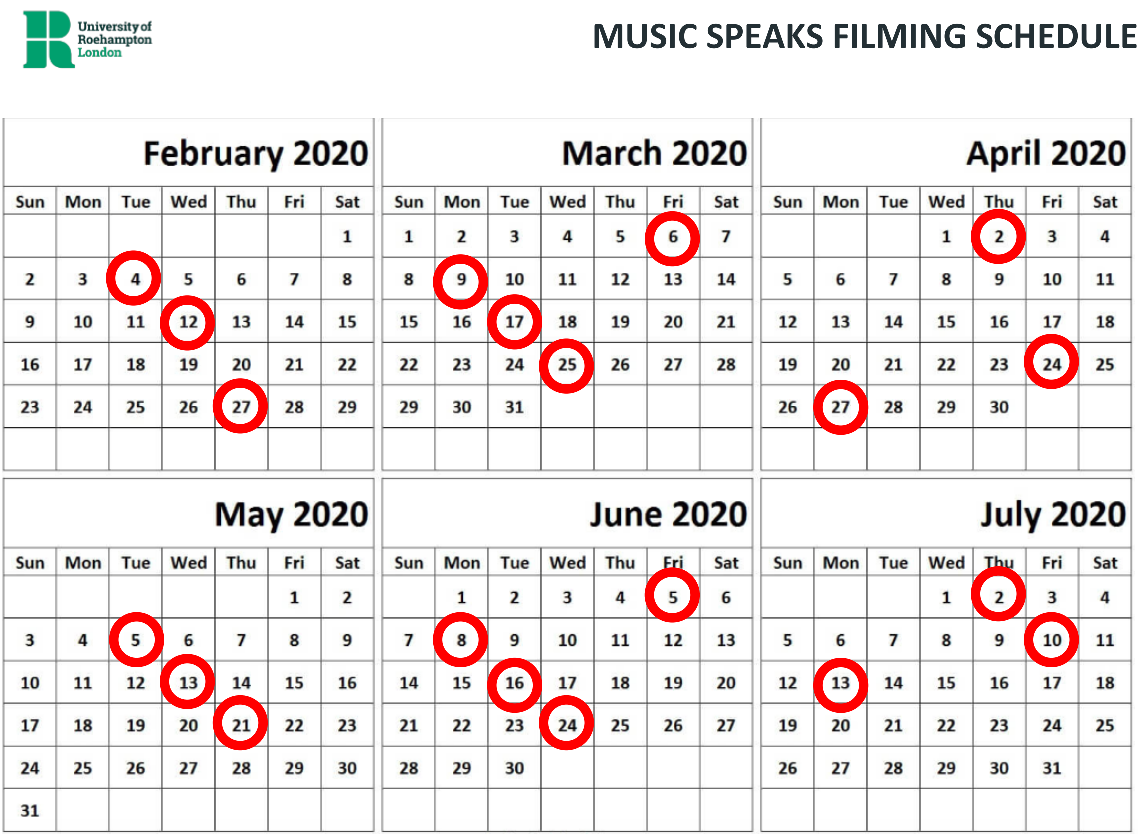
Holding Virtual Interviews

Considerations	
1	Is the platform secure and encrypted?
2	Can the interview space be made confidential?
3	Can the interview be recorded?
4	Where is the data stored?
5	Can parents and staff be interviewed?
6	Insufficient technology

Solutions	
1	In this case, the most appropriate platform would be Zoom
2	Use of headphones by both interviewer and interviewee.
2	Where possible a dedicated room to be used at home or in school
2	Disclosures made around compromised confidential spaces if interviewer is at home or interviewee is at home or in an open space.
3	Using Zoom would enable the interview to be video and audio recorded. Ensure participants are given the option of turning their camera off.
4	As per the ethics application, video data will be stored on an encrypted, password protected hard drive in anonymised folders.
5	No - at this point only written consent for staff has been obtained
5	Follow up verbal/ emailed consent would need to be obtained to check participants are still happy to be interviewed
6	Face to face or phone interview to be a second preference
6	Record the interview using audio recorder or for a phone interview using 'tapeacall' or other app.

Appendix 11: Music Speaks Filming Schedule

A visual calendar detailing the program filming schedule in Phase Two of the research. The schedule was given to educators and therapists as a reminder of which program session dates were to be filmed and uploaded onto the EthOS application.



University of
Roehampton
London

MUSIC SPEAKS FILMING SCHEDULE

February 2020

Sun	Mon	Tue	Wed	Thu	Fri	Sat
						1
2	3	[✓ 4]	5	6	7	8
9	10	11	[✓ 12]	13	14	15
16	17	18	19	20	21	22
23	24	25	26	[✓ 27]	28	29

March 2020

Sun	Mon	Tue	Wed	Thu	Fri	Sat
1	2	3	4	5	[✓ 6]	7
8	[✓ 9]	10	11	12	13	14
15	16	[✓ 17]	18	19	20	21
22	23	24	[✓ 25]	26	27	28
29	30	31				

April 2020

Sun	Mon	Tue	Wed	Thu	Fri	Sat
			1	[✓ 2]	3	4
5	6	7	8	9	10	11
12	13	14	15	16	17	18
19	20	21	22	23	[✓ 24]	25
26	[✓ 27]	28	29	30		

May 2020

Sun	Mon	Tue	Wed	Thu	Fri	Sat
					1	2
3	4	[✓ 5]	6	7	8	9
10	11	12	[✓ 13]	14	15	16
17	18	19	20	[✓ 21]	22	23
24	25	26	27	28	29	30
31						

June 2020

Sun	Mon	Tue	Wed	Thu	Fri	Sat
	1	2	3	4	[✓ 5]	6
7	[✓ 8]	9	10	11	12	13
14	15	[✓ 16]	17	18	19	20
21	22	23	[✓ 24]	25	26	27
28	29	30				

July 2020

Sun	Mon	Tue	Wed	Thu	Fri	Sat
			1	[✓ 2]	3	4
5	6	7	8	9	[✓ 10]	11
12	[✓ 13]	14	15	16	17	18
19	20	21	22	23	24	25
26	27	28	29	30	31	

Appendix 12: Review of Associated Assessment Tools

Parts 1 and 2 of the CMT Communication Subscale Development

Assessment Measure	Author	Related Speech Items	Notes
Frenchay Dysarthria Assessment (FDA)	(Enderby, 1980)	Time Pitch Volume Words Sentences Conversation	Requires eating, drinking, swallowing tasks
Speech Intelligibility Rating Scale (SIRS)	The Ear Foundation, 2020	Pre-recognisable words Intelligibility rating	Applicable for deaf populations
Therapy Outcome Measure, Dysarthria version (TOMs)	(Enderby & John, 2019)	Impairment Activity Participation Wellbeing/Distress Dysarthria	Long-term, holistic assessment measure
Unified Batten Disease Rating Scale (UBDRS)	(Adams et al., 2007)	Speech Clarity	Not detailed enough for communication elements
General language impairments for JNCL	(von Tetzchner et al., 2019)	Stuttering Wrong words Word finding Mumbling Short sentences Limited vocab/ repetition Weak voice, difficult to hear Calling attention with speech Uses single words Intelligibility Unclear, unintelligible No longer comprehensible No speech production	Expressive language Observed Speech

Education Development Observation Tool (EDO)	(von Tetzchner et al., 2019)	As above plus: Need for repetition/ clarification Articulation Use of signs/ gestures body language Technical/ Electronic Aids/ objects of references 1-20, 21-100, above 100 words used.	
Neuropsychological Inventory for Dementia (NPI)	(Cummings, 1997)		Emphasis on behavioural symptoms
Music in Dementia Assessment Scale (MiDAS)	(McDermott et al., 2015)		Behavioural based assessment for adults with dementia Development/refinement process includes peer review
Music Therapy Communication and Social Interaction Scale (MTCSI)	(Bell, Perry, Peng, & Miller, 2014)	Social Interaction Vocalisation	More appropriate as a general tool for music therapy assessment.

Appendix 13a: CMTOM Communication Subscale (version 1)

CMTOM Communication Subscale	Date		
Mark as 0 (never), 1 (rarely), 2 (sometimes), 3 (often) or 4 (consistently)	Pre	During	Post
Communication & Social interaction			
Stutters or stammers			
Repetition			
Dysarthria Mumbling			
Correct Use of Words			
Breathe Support			
Pacing			
Speech clarity			
Pitch			
Volume			
Articulation			
Intonation			
Stress			
Rhythm of speech			
Verbalisation (use of speech)			
Words			
Phrases			
Conversation			
Use of Communication aids			
Appropriate eye contact or tracking of visual stimuli			
Appropriate tracking of auditory stimuli			
Vocalisation (vocal play/ imitation)			
Responds to Familiar Voice			
Responds to own name			
Use of gesture			
Ability to musically interact, respond and join in			
Ability to vocally interact, respond and join in			
Ability to initiate interactions			
Behaviour / music to therapist appropriate?			

Appendix 13b: CMTOM Communication Subscale (final version)

CMTOM Communication Subscale	Date		
Mark as 0 (never), 1 (rarely), 2 (sometimes), 3 (often) or 4 (consistently)	Pre	During	Post
Elements of expressive communication			
Stutters or stammers			
Repetition			
Dysarthria/ Mumbling			
Correct Use of Words			
Breathe Support			
Pacing/ Rhythm of speech			
Speech clarity			
Pitch			
Volume			
Articulation			
Intonation			
Stress			
Words			
Phrases			
Receptive Communication			
Word Recall			
Responds to Familiar Voice			
Ability to verbally respond and participate			

Appendix 14: Interview Schedule

Music Speaks Interview

Introduction

- ☐ Intro and recap of researcher – Rebecca, MT and research, NMT application
- ☐ Recap of research and aims – Importance of music, impact of music education program, support with communication
- ☐ Ethical issues (video recording consent, confidentiality, (limitations of), data protection)
- ☐ Q's?
- ☐ Start video recording

Opening

1) Could you tell me about the young person you support?
    - ☐ What do you think are the challenges they face?

2) Can you tell me about your role in supporting [NAME]?

    - ☐ Are there approaches that you have found to be most impactful?
    - ☐ Can you describe any challenges they encounter?
    - ☐ How have you dealt with these?

3) Can you tell me how [NAME] engages with music?

    - ☐ What musical activities do they engage with (i.e. singing, listening, playing)?
    - ☐ How do you think music supports their communication?
    - ☐ How do you think music supports their well-being?

Music Speaks

4) Can you describe the Music Speaks Program?

5) What has been your experience of the program?
    - ☐ Was it appropriate for [NAME]?
    - ☐ Can you tell me about how the training supported you to deliver the program?
    - ☐ Challenges?
    - ☐ Positives?
    - ☐ Are you still using it?

6) How do you think Music Speaks has impacted the child you support?

    - ▯ How has the child's engagement with the program changed over time?
    - ▯ Did you see any changes in the child's communication?
        - ☐ Within each session of the program (start to finish)?
        - ☐ Over the 6 weeks of doing the program?
        - ☐ At other times in the school day?

7) What, if anything, did you learn from using the program?

8) How, if at all, might the program be useful for other children with a diagnosis of Batten disease?
    - In your opinion, what would be the perceived benefits, if any?

9) How do you think the program could be improved?

    - Would there be anything that would change?
    - What, if anything, do you think was missing from the program?

10) To what extent do you think this program is feasible for non-musical staff to deliver the program?

Closing

11) Is there anything else you'd like to tell me that we haven't covered today?

Appendix 15: Average Participant Mean Communication Scores Across 19 Time Points

	Time 1	Time 2	Time 3	Time 4	Time 5	Time 6	Time 7	Time 8	Time 9	Time 10	Time 11	Time 12	Time 13	Time 14	Time 15	Time 16	Time 17	Time 18	Time 19
Eye Contact	2.38	2.50	2.63	2.88	2.75	3.00	3.14	2.71	2.71	2.40	1.50	2.00	1.50	1.75	1.00	0.00	0.50	0.00	1.00
Appropriate Use of Gesture	2.25	2.58	2.67	2.67	2.75	2.64	2.64	2.82	3.00	2.55	2.60	2.40	2.30	2.22	2.13	1.71	1.67	2.20	1.50
Vocalisation	2.33	2.67	2.75	2.67	2.58	2.82	2.64	2.64	2.73	2.73	2.60	2.30	2.10	2.11	2.25	1.57	2.00	2.60	3.00
Verbalisation	1.58	1.58	1.83	1.67	1.67	1.82	1.82	1.82	1.82	1.64	1.80	1.70	1.50	1.67	1.38	1.14	1.17	1.60	2.00
Singing	1.42	1.58	1.92	1.67	1.58	1.55	1.45	1.09	1.18	1.18	1.20	1.10	1.30	1.11	1.25	1.14	1.17	1.20	1.50
Awareness of Others	2.75	3.33	3.50	3.42	3.50	3.45	3.55	3.55	3.55	3.18	3.10	3.10	3.10	3.11	3.50	3.14	2.83	3.20	3.00
Ability to Interact Verbally/Non-Verbally	2.50	2.08	2.58	2.42	2.50	2.55	2.73	2.64	2.64	2.55	2.50	2.40	2.30	2.22	2.00	1.86	1.67	2.40	2.50
Interaction with Staff	2.17	2.33	2.67	2.75	2.50	2.64	2.73	2.82	2.91	2.45	2.50	2.50	2.50	2.44	2.13	2.00	1.83	2.20	2.50
Ability to Initiate Interactions	1.92	1.92	2.08	2.08	2.00	2.27	2.36	2.36	2.45	2.09	2.00	2.20	2.20	2.00	1.50	1.71	1.67	1.80	2.00
Behaviour/ Music to Therapist Appropriate	2.58	2.67	3.08	2.83	2.82	2.73	3.09	2.91	3.00	3.09	3.00	3.10	3.10	3.00	3.00	2.86	2.67	3.20	2.00
Notice, Tolerate, Accept, Aware of Others	2.33	3.08	3.42	3.42	3.00	3.36	3.45	3.36	3.45	3.36	3.30	3.30	3.30	3.33	3.75	3.29	3.50	3.20	3.00
Ability to Participate/ Join In	2.42	2.17	2.83	2.42	2.75	2.55	2.45	2.36	2.36	2.36	2.50	2.70	2.60	2.56	2.38	2.14	2.00	2.80	3.50
Sharing Emotions, Thoughts and Ideas	1.20	1.60	1.60	1.55	1.60	1.70	1.60	1.60	1.40	1.80	2.00	1.90	1.90	1.78	1.38	1.57	1.67	1.80	2.00
Showing Empathy	1.00	1.36	1.55	1.58	1.64	1.64	1.73	1.64	1.36	1.45	1.20	1.20	1.20	1.00	0.63	0.71	0.83	0.80	1.50

Appendix 16: Average Participant Mean Behaviour Scores Across 19 Time Points

	Time 1	Time 2	Time 3	Time 4	Time 5	Time 6	Time 7	Time 8	Time 9	Time 10	Time 11	Time 12	Time 13	Time 14	Time 15	Time 16	Time 17	Time 18	Time 19
Trigger Observed to Change in Behaviour	0.00	0.00	0.00	0.00	0.00	0.00	0.00	0.00	0.00	0.00	0.00	0.00	0.00	0.00	0.00	0.00	0.00	0.00	0.00
Any Verbal Aggression	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00
Any Physical Aggression	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00
Behavioural Response to Music	2.64	2.00	2.67	2.42	2.25	2.27	2.27	2.18	2.27	2.00	2.20	2.40	2.60	2.44	2.50	2.43	2.00	2.40	3.00
Ability to Appropriately Express/Control Self	2.00	2.00	2.45	2.09	2.00	2.27	2.09	2.00	2.18	2.36	2.40	2.50	2.30	2.22	2.00	2.00	1.67	2.20	2.00
Expression of Distress, Agitation, Anxiety	3.08	2.67	3.00	3.08	3.33	2.45	3.09	2.73	2.18	2.64	3.10	3.00	2.70	3.00	3.25	3.57	3.17	2.80	3.50
Expression of Depression, Trauma, Loss, Bereavement	1.09	1.27	1.45	1.36	1.36	1.55	1.45	1.18	1.27	1.36	1.20	1.20	1.20	0.89	0.88	1.43	1.33	1.20	1.50

Appendix 17: Average Participant Mean Wellbeing Scores Across 19 Time Points

	Time 1	Time 2	Time 3	Time 4	Time 5	Time 6	Time 7	Time 8	Time 9	Time 10	Time 11	Time 12	Time 13	Time 14	Time 15	Time 16	Time 17	Time 18	Time 19
Verbal Expression of Mood	1.00	0.92	0.92	1.00	1.25	1.55	1.45	1.18	1.36	1.55	1.50	1.40	1.40	1.00	1.00	0.86	1.00	1.20	2.00
Physical Expression of Mood	1.83	2.08	2.08	2.50	2.42	2.55	2.45	2.27	2.55	2.36	2.10	2.10	1.90	2.00	2.00	2.00	1.67	1.40	1.50
Musical Expression of Mood	1.08	0.83	1.25	1.42	1.08	1.09	1.36	1.27	1.55	1.64	1.70	1.60	1.60	1.44	1.25	1.00	1.00	1.00	1.00
Choice of Art, Material, Object, Preference	2.83	2.33	2.67	2.67	2.58	2.82	2.64	2.91	2.55	2.36	2.50	2.50	2.40	2.22	1.88	1.57	1.17	2.00	1.50
Use of Voice or Instrument for Self Expression	1.33	1.92	2.33	2.25	2.50	2.45	2.64	2.27	2.36	2.55	2.30	2.20	2.30	2.11	1.88	1.71	1.50	2.20	2.00
Able to Tolerate Sounds, Art Forms, Music	2.92	2.67	3.08	3.00	3.25	3.36	3.64	3.45	3.64	3.36	3.80	3.70	3.70	3.67	3.88	3.86	4.00	4.00	4.00
Insights into Difficulties and Strengths	0.80	0.60	0.90	0.70	0.80	0.80	0.60	0.70	0.80	0.90	0.80	0.80	0.80	0.44	0.50	0.57	0.67	0.60	1.00
Ability to Explore and Discover	2.75	2.25	2.67	2.83	2.83	2.73	2.73	2.91	2.73	2.36	2.60	2.50	2.60	2.33	2.25	2.14	1.50	1.80	3.00
Shows Capacity to Improvise, Free Play	2.33	1.58	2.33	2.08	2.42	2.18	2.00	2.27	2.09	1.91	2.20	2.00	1.80	1.78	1.50	1.43	1.00	1.00	1.00
Can Differentiate Between Real and Imagined	1.33	1.86	1.71	2.29	2.29	2.29	2.50	2.29	2.29	2.29	2.00	2.00	2.00	1.60	0.80	1.00	1.00	1.33	0.00
Has Enthusiasm, Shows Pleasure, Fun, Enjoyment	2.67	2.00	3.08	2.42	2.50	2.45	2.45	2.36	2.45	2.36	2.40	2.50	2.60	2.33	2.13	1.86	1.17	1.80	2.00
Shows Appropriate Level of Self Confidence	1.92	2.45	2.55	2.64	2.73	3.10	3.10	2.90	2.90	2.80	2.56	2.44	2.44	2.38	2.25	2.17	1.80	2.00	2.00

Appendix 18: Average Participant Mean Cognition Scores Across 19 Time Points

	Time 1	Time 2	Time 3	Time 4	Time 5	Time 6	Time 7	Time 8	Time 9	Time 10	Time 11	Time 12	Time 13	Time 14	Time 15	Time 16	Time 17	Time 18	Time 19
Ability to Participate, Initiate, Choose, lead	2.08	1.92	2.58	2.33	2.58	2.36	2.36	2.36	2.45	2.36	2.60	2.70	2.50	2.44	2.25	1.86	1.83	2.20	3.00
Is Resourceful, Decisive, Can work Autonomously and Independently	1.67	1.17	1.50	1.58	1.83	1.64	1.64	1.45	1.45	1.55	1.50	1.50	1.40	1.44	0.88	0.86	1.00	1.40	1.00
Demonstrates Appropriate Level of Assertiveness	2.09	2.73	2.27	2.45	2.73	3.00	2.80	2.80	2.60	2.36	2.50	2.40	2.30	2.33	2.25	2.00	1.60	1.75	2.00
Follows Verbal Instructions	1.73	1.45	1.82	1.73	1.73	1.64	1.64	1.80	1.91	1.45	1.70	1.80	1.70	1.33	1.38	1.29	1.50	1.60	2.00
Makes Choices	2.92	2.00	2.25	2.42	2.25	2.45	2.36	2.60	2.00	2.18	2.30	2.30	2.30	2.11	2.13	1.71	1.50	1.60	2.00
Ability to Attend to Task	2.18	2.25	2.73	2.17	2.33	2.36	2.27	2.70	2.55	2.27	2.30	2.40	2.50	2.33	2.50	2.14	1.67	1.80	2.00
Recognition or Carryover of Previous Material	2.83	2.27	3.00	2.75	2.92	3.00	3.18	3.40	3.18	2.55	2.70	2.30	2.50	2.44	2.63	2.14	1.83	2.20	3.50
Engages Appropriately with Instruments	3.08	2.83	2.83	2.67	2.92	3.00	2.55	3.30	2.91	2.82	2.90	3.00	3.00	2.78	2.38	2.14	1.83	2.60	2.50
Any Memory Recall Noted (Rhythm etc)	2.75	2.18	2.58	2.25	2.42	2.27	2.73	3.10	2.55	2.09	2.00	2.00	1.90	1.67	1.63	1.43	1.33	1.80	3.50
Sustains Attention	2.33	2.42	2.67	2.33	2.58	2.64	2.55	2.70	2.45	2.55	2.50	2.70	2.60	2.67	2.75	2.43	2.17	2.40	2.50
Shows Interest and Is Inquisitive	2.67	2.75	3.17	2.75	2.75	2.73	2.64	2.60	2.64	2.64	2.80	2.90	3.00	2.89	2.63	2.57	2.17	2.40	3.00

Appendix 19: Average Participant Mean Motor Scores Across 19 Time Points

	Time 1	Time 2	Time 3	Time 4	Time 5	Time 6	Time 7	Time 8	Time 9	Time 10	Time 11	Time 12	Time 13	Time 14	Time 15	Time 16	Time 17	Time 18	Time 19
Active Movement	3.08	3.08	3.25	3.17	3.33	3.09	3.27	3.27	3.36	3.00	3.00	3.10	2.90	2.78	2.25	1.86	1.83	2.80	2.50
Core/ Trunk Stability	2.58	2.67	2.82	2.67	3.00	2.73	2.82	3.00	3.00	3.00	2.56	2.67	2.78	2.71	2.50	1.75	1.75	2.33	1.50
Head/ Neck Stability	2.83	3.00	2.83	2.75	2.58	2.73	2.64	2.91	3.00	3.00	2.78	2.78	2.89	2.75	2.67	2.40	2.40	3.00	2.00
Use of Weaker Limbs	1.88	1.75	1.75	1.50	1.75	1.43	1.71	1.86	2.00	1.00	1.43	1.29	1.14	1.14	1.17	0.80	0.25	1.00	3.00
Bi-lateral Hand Coordination	2.67	2.33	2.33	2.50	2.33	2.45	2.36	2.73	2.82	2.40	2.22	2.22	2.00	1.88	1.43	1.00	0.80	1.00	1.00
Hand-Eye Coordination	1.86	1.86	2.00	2.29	2.50	2.33	2.50	2.57	2.29	1.86	1.50	1.67	1.33	1.20	0.25	0.00	0.00	0.00	0.00
Ability to Cross Mid Line	2.83	2.50	2.42	2.58	2.58	2.55	2.55	2.82	2.90	2.80	2.67	2.67	2.40	2.22	1.63	1.43	1.50	1.25	1.50
Fluency of Gait Movement	1.33	1.42	1.36	1.33	1.42	1.27	1.36	1.18	1.36	1.11	0.50	0.50	0.50	0.43	0.00	0.00	0.00	0.00	0.00
Ability to Grip in RH	2.58	2.50	2.45	2.55	2.75	2.64	2.64	3.18	2.91	2.60	2.38	2.50	2.50	2.29	2.33	0.75	0.25	1.00	0.00
Ability to Grip in LH	2.83	2.33	2.33	2.50	2.83	2.73	2.64	2.91	2.82	2.73	2.56	2.67	2.44	2.50	2.43	1.40	1.00	1.75	0.00
Individual Finger Movements (piano, technology etc)	2.91	3.09	2.73	2.91	2.92	2.45	2.50	3.18	3.20	2.82	2.89	2.89	2.60	2.67	2.50	2.00	1.67	2.60	2.50
Oral Motor Control	1.60	1.57	2.00	1.83	2.00	1.67	2.00	1.50	2.00	1.86	1.71	1.71	1.67	1.00	0.60	0.80	1.20	1.50	0.00
Breath Control and Regular Respiration	3.25	3.58	3.58	3.67	3.67	3.82	3.82	3.55	3.82	3.73	3.40	3.40	3.30	3.11	3.00	3.14	2.83	2.80	3.00
Hand over Hand or Facilitated Mvt Needed	2.67	2.58	2.42	2.50	3.00	2.91	2.91	3.18	3.18	2.82	2.80	2.70	2.70	2.78	2.38	2.00	2.00	2.60	3.00

Appendix 20: Task Completion for Participants Three and Five in the Music Speaks program Trial

	Participant Three		Participant Five	
	Achieved	Not Achieved	Achieved	Not Achieved
03/02/2020	3	1	0	4
04/02/2020	4	0	1	3
05/02/2020	3	1	2	2
06/02/2020	4	0	0	4
07/02/2020	4	0	1	3
10/02/2020	2	2	2	2
11/02/2020	4	0	0	4
12/02/2020	1	3	0	4
13/02/2020	3	1	0	4
14/02/2020			4	0
17/02/2020				
18/02/2020				
19/02/2020				
20/02/2020				
21/02/2020				
24/02/2020	2	2	1	3
25/02/2020	3	1	1	3
26/02/2020	2	2	0	4
27/02/2020	3	1	3	1
28/02/2020	3	1	4	0
02/03/2020	1	3	3	1
03/03/2020	3	1	3	1
04/03/2020	3	1	0	4
05/03/2020			0	4
06/03/2020	1	3	2	2
09/03/2020			2	2
10/03/2020	2	2	3	1
11/03/2020	3	1	3	1
12/03/2020	3	1		
13/03/2020	3	1	4	0
16/03/2020	3	1		
17/03/2020	3	1	2	2
18/03/2020	3	1		
19/03/2020	3	1		

Appendix 21: Mean Pre and Post Expressive Communication Scores in Response to Music Speaks Program Trial

Expressive communication	Week 1 Baseline	Week 1 Endpoint	Week 2 Baseline	Week 2 Endpoint	Week 3 Baseline	Week 3 Endpoint	Week 4 Baseline	Week 4 Endpoint	Week 5 Baseline	Week 5 Endpoint	Week 6 Baseline	Week 6 Endpoint
Participant 1	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00
Participant 2			3.36	3.50					4.00	4.00		
Participant 3	3.50	4.00	2.57	3.93	3.93	4.00	3.00	3.36	2.93	3.93	3.93	4.00
Participant 4							3.21	3.29				
Participant 5	0.50	2.36	0.93	1.00	1.43	1.86	1.14	2.64	1.00	1.07		
Mean	2.67	3.45	2.71	3.11	3.12	3.29	2.84	3.32	2.98	3.25	3.96	4.00
SD	1.89	0.95	1.33	1.42	1.46	1.24	1.21	0.55	1.41	1.45	0.05	0.00

Appendix 22: Mean Pre and Post Receptive Communication Scores in Response to Music Speaks Program Trial

Receptive Communication	Week 1 Baseline	Week 1 Endpoint	Week 2 Baseline	Week 2 Endpoint	Week 3 Baseline	Week 3 Endpoint	Week 4 Baseline	Week 4 Endpoint	Week 5 Baseline	Week 5 Endpoint	Week 6 Baseline	Week 6 Endpoint
Participant 1	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00	4.00
Participant 2			4.00	4.00					4.00	4.00		
Participant 3	4.00	4.00	4.00	4.00	4.00	4.00	2.00	3.00	3.33	4.00	4.00	4.00
Participant 4							4.00	4.00				
Participant 5	1.33	3.33	2.33	2.00	2.67	3.00	3.00	3.67	2.33	1.33		
Mean	3.11	3.78	3.58	3.50	3.56	3.67	3.25	3.67	3.42	3.33	4.00	4.00
SD	1.54	0.38	0.83	1.00	0.77	0.58	0.96	0.47	0.79	1.33	0.00	0.00