



The potential role of music
to enhance the lives of
children and young people with
Neuronal Ceroid Lipofuscinosis
(Batten disease)

A report on a three-year
research project undertaken
by the Applied Music
Research Centre at the
University of Roehampton

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Funded by The Baily Thomas Charitable Fund with support from The Amber Trust and The Linbury Trust

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WITH THANKS

to the families, children and young people who took part in the research:

Aahil, Amelia, Eva, Freddie, Jenson, Laura, Lily, Louie, Luke, Olivia, Ollie, Storm, Yanna

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The Applied Music Research Centre specialises in using music theory to explore musical development across the spectrum of neurodiversity, with particular foci on special musical abilities and needs; to develop strategies and resources for those working in music education, music psychology, music therapy and community music; and to gather evidence for the importance of music in fostering cognitive and social skills and promoting well-being. See www.roehampton.ac.uk/research-centres/applied-music-research-group/
Chiltern Music Therapy is a not-for-profit organisation based in Chesham that provides music therapy and community music to people of all ages.

SCHOOLS

Fountaindale School, Mansfield; Linden Lodge School, Wandsworth; Ravensbourne School, Romford; The Redway School, Milton Keynes; Stephen Hawking School, Tower Hamlets; Stubbin Wood School, Shirebrook; Vernon Primary School, Poynton

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The Baily Thomas Charitable Fund www.bailythomas.org.uk funds research into learning disability and supports those affected by it through making grants to voluntary organisations working in the field.

The Amber Trust www.ambertrust.org is a registered charity created to help blind and partially sighted children who have a talent or a love for music.

The Linbury Trust www.linburytrust.org.uk is a charitable trust founded in 1973 by Lord Sainsbury of Preston and his wife Anya, Lady Sainsbury, that makes grants to organisations and towards causes including the arts, education, the environment and heritage, and in the areas of medicine, social welfare and developing countries.



FOREWORD

This report gives evidenced hope. It leaves me feeling optimistic. Why? Because it is academically sound, eminently practical, and, above all, it puts the needs and aspirations of the research participants – young people with childhood dementia – at its heart. It offers parents and professionals new insights and ideas to enrich the lives of those they care for and work with.

There is relatively little academic, evidence-based research in this highly specialised field of disability. Ockelford, Atkinson and Herman, supported by an eminent steering group, have produced a rigorous piece of research that will surely influence educational and therapeutic practice in the field of Batten disease and beyond. It is part of a wider body of work undertaken at the Applied Music Research Centre, University of Roehampton, that has forged an innovative approach in which music is seen as an indispensable element in the lives and education of people with complex needs.

For most of us, the experience of sound starts very early – in the womb – and so our understanding of music can develop before the genetic consequences of conditions such as Batten disease take hold. Music can also be one of the last effective ways of communicating as we near the end. So, while research into the place of music in all of our lives is important, it is nothing less than crucial for those with childhood dementia, who will lose their sight, their capacity to move and the ability to understand much of what is happening to them. Through all this, it seems, the brain's capacity to engage with music, at some level, remains intact.

Unlike many academic studies, the report's findings and recommendations will be of immediate, practical value to parents and carers, professionals in the field, researchers and policy makers. The language and tone of the text make it accessible to all these groups. However, it is the child-centred approach which lights me up most brightly. The research is based on the needs and wishes of the children and young people whose lives it explores. It starts and ends with them. It recommends building on what they can do rather than viewing them as an intractable knot of deficits. In other words, it is respectful and empowering. I commend it to you.

Ian Bruce

Professor Ian Bruce CBE CCMi FCIM DSocSc

Vice President: Royal National Institute of Blind People

President: Centre for Charity Effectiveness, Cass Business School

Grandfather of two young children



KEY FINDINGS AND RECOMMENDATIONS

This project is the first in a proposed series called 'MIND' ('Music in Neurodegenerative Disease') led by the Applied Music Research Centre at the University of Roehampton that explores the potential role of music in the lives of people who have a neurodegenerative disease. Here the focus is on children and young people with Neuronal Ceroid Lipofuscinosis (NCL), commonly known as Batten disease. The principal source of evidence for the research is 13 case studies of children and young people in the UK with NCL, undertaken over three years. In addition, data are derived from a European survey that examined the implications of one variant of Batten disease ('CLN3') for learning and teaching.

The key findings are as follows:

1. Music can play a unique role in enriching the lives of children and young people with Batten disease. There are a number of reasons why this should be so:
 - a) The intuitive capacity to understand music declines more slowly than other cognitive abilities.
 - b) For young people with the juvenile form of Batten disease, listening to music becomes ever more important as a leisure pursuit as the disease advances and other recreational options diminish.
 - c) Even in the very late stages of neurodegeneration there is evidence that the facility to engage with music as a listener remains.
 - d) Active engagement in music, including the chance to learn to play an instrument, should be encouraged as early as possible.
 - e) When children and young people no longer have the physical capacity to play conventional instruments, active participation in music-making should be sustainable through gesture-based music technology.
 - f) For young people who could once speak, but in whom the capacity to use language expressively is declining, 'micro-songs' (short songs that incorporate functional language) can sustain the capacity to communicate through words longer than would otherwise be the case; at the same time, certain of the techniques used in neurologic music therapy can aid respiratory control and assist with controlling the pace of speech.
 - g) Playing, singing and song writing (in particular) can offer young people a medium through which to access and articulate their feelings when other channels of emotional expression are occluded.
 - h) Favourite pieces of music can be remembered when other forms of recollection diminish, and songs can be used to stimulate the recall of associated memories that would otherwise be inaccessible.
 - i) Sound and music can be used symbolically to convey information about activities, places and people, potentially supporting children and young people's understanding, communication and choice-making.
 - j) Music has the capacity to aid children and young people in regulating their emotions, and produce a sense of well-being.
 - k) Music offers a unique form of social activity in which children and young people can engage with family members and friends, even in the most advanced stages of the disease.
2. However, there are certain barriers that may prevent music fulfilling as complete a role as it could in the lives of children and young people with Batten disease:
 - a) Music teachers are likely to lack the experience of working with blind pupils and so find it difficult to adapt their usual teaching strategies, particularly those who favour a notation-based approach.
 - b) Teachers and carers who are not music specialists may lack the confidence to use the musical strategies (such as micro-songs and sound symbols) that would enable children and young people to sustain communication for longer.
 - c) None of the music technology that is currently available that can be operated through gesture offers a readily affordable solution to accessibility for those with limited movement.

It is recommended:

1. that this report be disseminated to families, parents, teachers, clinicians, lobbyists, service providers and policy makers (and re-circulated periodically, since populations and post-holders change);
2. that resources and strategies showing how to use music to enhance the lives of children and young people with Batten disease, that are suitable for parents and carers (who may be non-music-specialists), be created and made freely available, both in online and physical form;
3. that information and advice for music teachers who are asked to work with a blind pupil be generated and disseminated; and
4. that music technology companies be approached with a view to creating gesture-driven devices or software that are readily affordable, yet potentially capable of producing sophisticated musical results.



1 INTRODUCTION

- 1.1 This research study, 'MIND, Phase 1', is the first to address the potential role of music in enhancing the quality of life of children and young people with Batten disease, which causes childhood dementia, in a systematic way. The findings have significant implications for this small but extremely needy group by offering strategies to their therapists, teachers and families that will permit them to sustain communication when words alone no longer function, to give children and young people a medium through which to articulate their feelings when other channels of emotional expression are occluded, to enable memories to be accessed that would otherwise be lost, and, above all, to help maintain a sense of well-being in an increasingly confusing and frightening inner world. There are implications too for those working with and caring for adults of working age and older people with dementia.
- 1.2 The research team worked closely with the Batten Disease Family Association (BDFA)¹ in the UK, which was a partner in a survey that examined the implications of Batten disease for learning and teaching in a number of European countries.² There was enthusiastic support too from the Batten Disease Support and Research Association (BDSRA)³ in the USA. Through these networks, it is hoped that the lessons learnt in the UK from the current study will have a wider geographical impact. Moreover, the resources that will be created in response to the recommendations set out in this report will potentially benefit children and young people with other forms of neurodegenerative disease, such as juvenile Tay-Sachs disease, Sandhoff disease and Canavan disease – forms of illness that collectively represent around a quarter of admissions to paediatric neurology services (Dyken and Krawiecki, 2004).

2 BATTEN DISEASE

- 2.1 Batten disease, embracing what are formally known as the Neuronal Ceroid Lipofuscinoses (NCLs), comprises several different genetic life-limiting neurodegenerative conditions that share similar features (Mole, Williams and Goebel, 2011). Symptoms include blindness, epilepsy with severe seizures, myoclonic jerks of limbs, difficulties in sleeping, the decline of speech, language and swallowing abilities, and the deterioration of fine and gross motor skills that eventually result in the complete loss of mobility. Those with the disease are also likely to suffer hallucinations and memory loss and, unsurprisingly, exhibit challenging behaviours (Bills, et al., 1998). The NCLs are sometimes described by the child's age at the time symptoms begin to appear ('infantile', 'late-infantile' or 'juvenile'), though their classification is based on the gene that causes the disorder. Each gene is called CLN (ceroid lipofuscinosis, neuronal), and the fourteen subtypes of the disease that have currently been identified are given a different number in order to distinguish them (CLN1 to CLN14). It is important to recognise, even within a single genetic category, that the course of the disease can vary in severity and result in different rates of functional decline.⁴ The NCLs are, fortunately, extremely rare: according to the National Institute of Neurological Disorders and Stroke in the USA, Batten disease occurs in around two to four of every 100,000 live births,⁵ and the BDFA estimates that there are between 100 and 150 affected individuals in the UK, with 11–17 new diagnoses being made each year.
- 2.2 Of the 13 participants in the MIND study, five variants of Batten disease were represented: CLN2, CLN3, CLN5, CLN6 and CLN8. The following short descriptions are derived from information provided by the UCL NCL resource⁶ and the US National Library of Medicine: Genetics Home Reference.⁷ More detailed accounts are to be found in the second edition of *The Neuronal Ceroid Lipofuscinoses (Batten Disease)*, edited by Sara Mole, Ruth Williams and Hans Goebel,⁸ and the 'Genetics of the Neuronal Ceroid Lipofuscinoses', by Sara Mole and Susan Cotman.⁹

¹ www.bdfa-uk.org.uk

² Funded by the European Commission through Erasmus+ Grant No. 2014-1-NO01-KA200-000388.

³ www.bdsra.org

⁴ Even monozygotic twins with Batten disease can manifest significant developmental differences, as the case of Freddie and Louie, reported in this study, shows (see Figures 15 and 16).

⁵ www.ninds.nih.gov/Disorders/Patient-Caregiver-Education/Fact-Sheets/Batten-Disease-Fact-Sheet

⁶ www.ucl.ac.uk/ncl-disease/ncl-resource-gateway-batten-disease

⁷ www.ghr.nlm.nih.gov

⁸ Published by Oxford University Press in 2011.

⁹ Published in *Biochimica et Biophysica Acta*, 1,852 (Issue 10, Part B), 2,237–2,241.

CLN2 disease, late-infantile

The CLN2 gene, found on chromosome 11, normally produces an enzyme called tripeptidyl peptidase 1, which breaks down proteins. In the late-infantile form of CLN2 disease, this enzyme is insufficiently active. Developmental delay becomes evident as children approach their third birthday; they develop seizures and gradually lose the ability to speak and to walk. Myoclonic jerks typically begin around the age of four or five. By the time they are six, most children are completely dependent on their caregivers, and most die before they are 12.

CLN3 disease, juvenile

The CLN3 gene, found on chromosome 16, directs the production of a protein called battenin, which is located in the membranes of lysosomes and endosomes, the parts of cells that digest and recycle materials. Most children with CLN3 disease, which is also known as Juvenile Neuronal Ceroid Lipofuscinosis or 'JNCL', and is the most common type of NCL in Europe and the US, have a missing part in the gene, which means that they are unable to produce battenin. Rapid loss of vision occurs between the ages of four and seven. Children enter a period of slow cognitive decline and typically start having seizures around the age of 10. In their teenage years, the ability to move diminishes, with increasing slowness, stiffness and loss of balance. Their capacity to speak is lost, and young people become more and more dependent on their caregivers. Most die between the ages of 15 and 30.

CLN5 disease, late-infantile

The CLN5 gene lies on chromosome 13 and encodes for a lysosomal protein whose function is as yet unknown. Children progress normally for the first few years of life before they start losing skills and typically develop behaviour problems. Seizures and myoclonic jerks usually begin between the ages of six and 13. Vision deteriorates and is eventually lost altogether. Children develop learning difficulties with particular problems in concentration and memory. Most die in late childhood or early adolescence.

CLN6 disease, late-infantile

The CLN6 gene, located on chromosome 15, directs the production of the protein linclin. The protein, whose function has not yet been identified, is found in the membranes of cells, largely in the endoplasmic reticulum (a structure that contributes to recycling). Symptoms vary, but typically start after the first few years of life and include developmental delay, changes in behaviour and seizures. Children also develop myoclonic jerks, problems with sleeping, and vision loss. They eventually become unable to walk or talk. Most die in late childhood or early adolescence.

CLN8 disease, late-infantile

The CLN8 gene, which is located on chromosome 8, encodes a protein (also called CLN8) that is found in the membranes of cells, mainly in the endoplasmic reticulum. Its function has not yet been identified. Children who are affected begin showing symptoms between the ages of two and seven, including vision loss, cognitive problems, treatment-resistant epilepsy, myoclonic jerks, changes in behaviour and unsteadiness to the point where they can no longer walk or stand unassisted. Life expectancy varies, though most children die before they are 10.

- 2.3 Currently there is no cure for Batten disease, although a clinical trial over 48 weeks of enzyme replacement treatment using Cerliponase Alfa for children with CLN2, halted their decline in 65% of cases ($N = 23$), with a further 22% showing a slower progression of symptoms than would otherwise have been expected.¹⁰ This result notwithstanding, specialist symptom management and therapy continue to be essential for the children with all forms of the disease in order to maintain as good a quality of life as possible. The current study explored one element of what is sure to be a multidisciplinary package – music – that may occur in the context of education or therapy, or as a leisure pursuit.

¹⁰ See www.brineura.co.uk/results/ and www.nice.org.uk/guidance/gid-hst10008/documents/final-evaluation-determination-document

3 ANECDOTAL EVIDENCE AS TO THE IMPORTANCE OF MUSIC IN THE LIVES OF THOSE WITH BATTEN DISEASE

3.1 Prior to the research reported here, no systematic study of music in the lives of those with Batten disease had ever been undertaken, in what is a highly specialised area. However, numerous anecdotal accounts of parents, and the reports of teachers and therapists, had suggested that music may play a crucial and often under-utilised role. For example, here is a mother's description of her nine-year-old twin boys (featured in the current study), who had been struggling to cope with the progressive deterioration of their faculties and were supported to use music for emotional self-regulation:¹¹

It was Freddie and Louie learning to love music ... that made the difference. The family spend most weekends seeking out music events and [they] now benefit from music therapy which brings not only great pleasure to the boys' lives, but helps their learning and development and enables them to reach out to the world.



Freddie and Louie

3.2 The power of music to evoke reminiscences and to enable children to share these memories with others is evident in Laura's music therapy report, written when she was 15 years old:

Her music therapy sessions have always been rich, emotionally vibrant and creative times. She has been able to use the sessions to express the emotions she is feeling at the time, as well as for the reflection and remembering ... It's been good that she's felt able to reflect on her life... singing about the events she can remember.¹²

This approach continued to play an important part in subsequent therapy sessions.

3.3 Crucially, it seems that the brain's capacity for processing music may remain, at least in some cases, until the very end of life. For example, a five-year-old, in the latter stages of the condition was reported to receive her music therapy in bed.

It is the one therapy that she is still responding to with smiles and clear enjoyment, providing a lovely time for the family to have together.¹³

¹¹ Account taken from a programme published by The Amber Trust for a concert at the Theatre Royal Haymarket, November 2014.

¹² Report to The Amber Trust, July 2014.

¹³ Email communication with The Amber Trust, May 2014.

4 HYPOTHESIS UNDERPINNING THE CURRENT STUDY

- 4.1 These observations and others, based on conversations with parents and professionals, and drawn from the written reports received by The Amber Trust in relation to its Music Awards scheme, suggested two things. These were that, in children and young people with Batten disease:
- a) the ability to process musical sounds, to respond to music emotionally, and to make music (assuming that the barriers caused by motor impairment can be circumvented) appear to remain intact after other capacities, such as the ability to speak, have been lost; and
 - b) music can be used to nurture other cognitive abilities – as it can in all of us (see, for example, Schellenberg, 2016) – though in the current context it may fulfil particular roles such as aiding memory, accessing language when speech is no longer functioning, and fostering well-being.
- 4.2 These assertions led to the hypothesis that music may potentially play a unique role, educationally, therapeutically and recreationally, in the lives of some (if not all) children and young people with Batten disease.

5 RESEARCH QUESTIONS

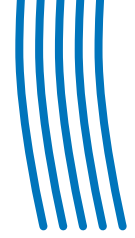
- 5.1 The hypothesis was used to generate the following research questions (RQs):
1. To what extent and in what ways do the level and nature of musical interests and musical abilities vary among the population of children with Batten disease?
 2. Do musical abilities decline as other cognitive abilities are lost, and, if so, what is the nature of the correlation between them, or does the capacity to understand, appreciate and make music take a different course?
 3. To what extent and in what ways can music be used as an intervention to promote:
 - a) the maintenance of expressive language when speech is in decline (or has ceased);
 - b) a day-to-day understanding of what is occurring, where, with whom and when;
 - c) the recall of important, long-term memories;
 - d) emotional regulation and well-being; and
 - e) social inclusion, particularly in the later stages of the disease?
- 5.2 The method by which each of these questions was addressed, and the results pertaining to them, are set out below.

6 DESIGN OF THE STUDY

- 6.1 It was decided to tackle the research questions in two ways. The first was through a survey, which was to have been distributed to families in the UK through the BDFA network. In the event, a wider opportunity opened up through the European Erasmus+ study, which the research team were able to utilise to their advantage. The second approach originally entailed 12 case studies. This was considered to be an appropriate number as it was small enough to enable detailed data to be gathered while nonetheless representing around 10% of the population of children and young people with Batten disease in the UK, with a spread of genetic variants, ages, social and cultural backgrounds and geographical locations. In the third year of the project, a further young person was recruited, who attended a special school close to the university, as it was felt that she would particularly benefit from the strategies for language maintenance that were being developed and tested. These 13 children and young people made up the 'core' participants in the study. See Table 1.



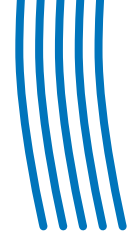
Jenson with his line of bells



Participant	Age at onset of study	Male or female	Genetic variant	Level of functioning	Engagement with music
Amelia ^{1,2}	3	Female	CLN2 Late-infantile	Vision still functional in most everyday contexts No issues with motor function No language difficulties No seizures	Receives bi-weekly music sessions at school Participates actively in singing and instrument play
Ollie ^{1,2}	5	Male	CLN2 Late-infantile	Low vision Minimal mobility No expressive language Seizures occurring frequently	Receives bi-weekly music sessions at school Most engagement with music is reactive, although some proactive and interactive participation occurs using music technology
Aahil	5	Male	CLN6 Late-infantile	Low vision Uses a wheelchair, some mobility preserved Significant language difficulties No seizures	Receives weekly music therapy sessions at school Participates actively in instrument play Readily engages with music
Yanna	5	Female	CLN2 Late-infantile	Low vision Uses a wheelchair, some mobility preserved Minimal expressive language Seizures occurring infrequently	Receives weekly music therapy sessions at school Participates actively in instrument play
Storm	6	Male	CLN2 Late-infantile	Low vision Walks with support Moderate language difficulties Seizures occurring infrequently	Receives weekly music therapy sessions at school Participates actively in singing and instrument play
Jenson ³	8	Male	CLN8 Late-infantile	Blind Total loss of mobility No expressive language Seizures occurring frequently	Receives weekly music therapy sessions at school Primarily engages reactively, although some proactivity is possible using music technology
Eva	9	Female	CLN2 Late-infantile	Blind Uses a wheelchair; minimal independent motor function No expressive language Seizures occurring frequently	Receives weekly music therapy sessions at school Primarily engages reactively

¹Left the project in Year 2 ²Siblings ³Passed away at the end of Year 3

Table 1. The research participants.



Participant	Age at onset of study	Male or female	Genetic variant	Level of functioning	Engagement with music
Freddie ⁴	10	Male	CLN5 Late-infantile	Blind Uses a wheelchair, some mobility preserved No expressive language Seizures occurring infrequently	Receives weekly music therapy sessions at school Participates actively in instrument play Readily engages with music
Louie ⁴	10	Male	CLN2 Late-infantile	Blind Uses a wheelchair, some mobility preserved No expressive language Seizures occurring infrequently	Receives weekly music therapy sessions at school Participates actively in instrument play Highly engaged by music
Luke	12	Male	CLN3 Juvenile	Blind Normal motor function Minor expressive language difficulties No seizures	Receives weekly music therapy sessions, music lessons at school, private piano instruction and weekly music technology support sessions at home Is working on recording an album Highly motivated by music
Olivia	15	Female	CLN3 Juvenile	Blind Uses a wheelchair, some mobility preserved Significant language difficulties Seizures occurring frequently	Receives weekly music therapy sessions at school Participates actively in instrument play
Lily ⁵	16	Female	CLN3 Juvenile	Blind Uses a wheelchair, some mobility preserved Significant language difficulties Seizures increasing in frequency	Has weekly music sessions at school as part of a group Highly motivated to participate through singing
Laura	17	Female	CLN3 Juvenile	Blind Uses a wheelchair with some mobility preserved Significant language difficulties Seizures occurring frequently	Receives weekly music therapy sessions at her residential home Participates actively in singing and instrument play Highly engaged by music

⁴Monozygotic twins ⁵Joined the project in Year 2

Table 1 (cont.). The research participants.

6.2 The range of participants by age and CLN variant is summarised in Figure 1. The skewed nature of the distribution is a reflection of the different ages at which variants manifest themselves and progress.

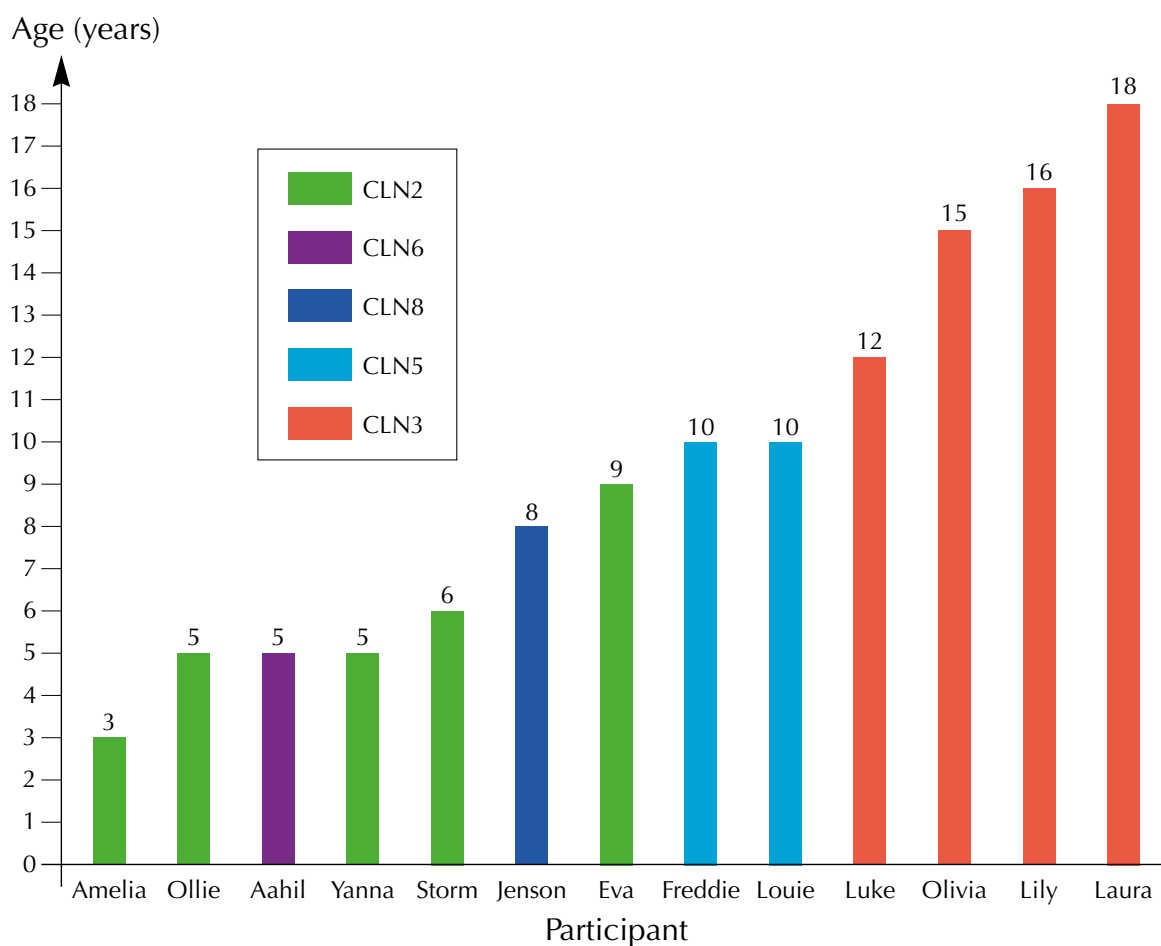


Figure 1. The range of research participants by age (in years) at the beginning of the study and their CLN variants.

- 6.3** Following requests from their families, two other ‘associate’ participants, Leo and Annabelle, were included in the dissemination of resources and information about the progress of the research. All the families were offered support from The Amber Trust, which guaranteed the ongoing funding of music provision (whether in the form of therapy, education or recreation) for the participants in the research.¹⁴
- 6.4** A case study methodology was adopted since, given the dearth of previous research in this area, the investigation was necessarily exploratory in nature. Moreover, RQ3 demanded qualitative data that were best gathered through detailed observation and discussion with families, professionals and the children and young people themselves. In contrast, RQ1 and RQ2 required the acquisition of quantitative information pertaining to participants’ global levels of functioning and their capacity for engaging with music. These were derived from the same observations and discussions, which were analysed using the Hamburg Scale (Kohlschütter, Laabs and Albani, 1988) and the Sounds of Intent framework of musical development (Vogiatzoglou, et al., 2011).
- 6.5** The Hamburg Scale was selected on the advice of Ruth Williams, Consultant Neurologist at the Evelina London Children’s Hospital, who sat on the project steering group, as providing sufficient data for the project without entailing the relative complexity of the Unified Batten Disease Rating Scale. Although originally conceived in the context of JNCL (CLN3), the Hamburg Scale can be applied equally to all the variants, and does not require clinical expertise or special training to administer. It records changes in four major functional areas: vision, intellectual capacity, expressive language and motor skills. Scores range from 3 (normal function) to 0 (total loss of function) with unit decrements representing milestones in the loss of function (for example, losing the ability to walk or crawl). There is also a four-point scale to record the frequency of grand mal seizures. The complete scale is shown in Table 2.

¹⁴In the course of the study, from spring 2016 to spring 2019, The Amber Trust offered Music Awards to 36 children and young people to have music therapy sessions or music lessons or to purchase instruments – around a third of the UK population with Batten disease. In this time, four of the children and young people died.



Yanna and Storm
explore the piano and guitar

Problem	Functional state	Coding score
Vision	Normal	3
	Poor, but orientation good	2
	Poor, but orientation difficult	1
	Blind	0
Intellect	Normal	3
	Abstract reasoning (mathematics) 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
	Minor difficulties 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
	>12 / year	0

Table 2. 'Single coding scores possible for each of 5 major clinical problems in JNCL patients' (from Kohlschütter, Laabs and Albani, 1988, p. 868).

6.6 The Sounds of Intent framework of musical development was created over the last two decades by a core research team comprising Adam Ockelford at the University of Roehampton, Graham Welch and Evangelos Himonides at the UCL Institute of Education, and Sally Zimmermann at the Royal National Institute of Blind People. The team worked with practitioners and professionals from the special needs sector (including music therapists and teachers, speech and language therapists, and teaching assistants) to ascertain whether it was possible to identify a music-developmental trajectory that was common to children and young people with profound and multiple learning difficulties. The initial method was inductive, or 'bottom up': detailed observations were subject to rich description followed by analysis and comparison. Could some apparently disparate musical activities reasonably be regarded equivalent in cognitive terms, for example? Were there certain ways of engaging with sound and music that

necessarily preceded others? Over a period of five years a model was built up and refined in a series of iterations that took account of children's musical interactions in a wide range of educational, therapeutic, social and cultural contexts, and it was extended to include those whose learning difficulties were less profound.

- 6.7 A second stage of the Sounds of Intent research was to subject the provisional model of musical development to deductive ('top down') scrutiny using 'zygonic' theory, which seeks to explain how music makes sense (Ockelford, 2017). In particular, the theory sets out three types of musical structure that are held to exist in a hierarchy: organisation at the level of events (notes and chords), groups (motifs and riffs) and frameworks (scales and metres). It was found that these three forms of structure, which make increasing demands on cognition, mapped readily onto three of the developmental milestones that had been identified through observation. It emerged that there are two distinct steps in musical development that precede the capacity to process musical structure: a stage before auditory perception starts, and a purely perceptual phase, in which pattern and predictability do not yet register. Moreover, it appeared that there is a further, advanced level of musicality, beyond an understanding of tonal and metrical frameworks, in which the social and emotional communicative capacity of music are understood (and which can be reflected in expressive performance). Hence, the evolving Sounds of Intent framework crystallised into six levels of musical engagement, listed here with their descriptors:

Level 1 – 'before hearing starts to develop'

Level 2 – 'music matters'

Level 3 – 'sounds interesting'

Level 4 – 'copy me, copy you'

Level 5 – 'whole songs, in time and in tune'

Level 6 – 'musical maturity'

- 6.8 The research team's observations indicated that children and young people engaged with sound and music in three ways: 'reactively' (through listening and responding), 'proactively' (creating sounds and music on their own) and 'interactively' (making sounds and music with others). Combining these three domains with the six levels of engagement resulted in 18 headlines of musical participation, which were represented as a series of concentric circles, with Level 1 innermost and Level 6 outermost. See Figure 2.

Luke plays the harmonica



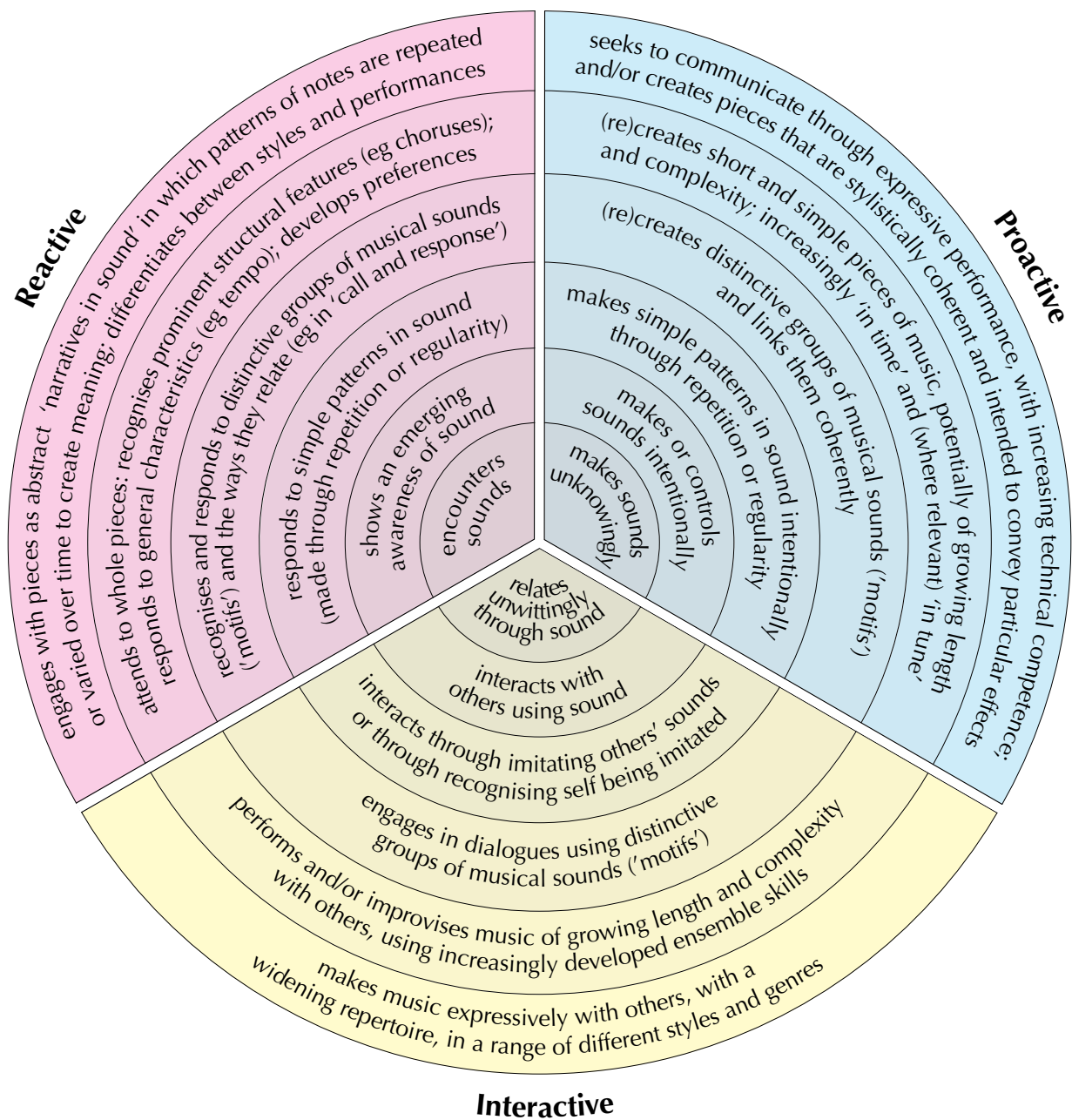


Figure 2. The Sounds of Intent framework of musical development.

6.9 Each of these 18 segments was subdivided into four 'elements', 'A', 'B', 'C' and 'D', which enabled detailed assessments and child-centred curriculum planning to take place. 'R' stands for 'Reactive', 'P' for 'Proactive' and 'I' for 'Interactive'. So 'R.1.C' means, 'Reactive, Level 1, Element C', for example, 'P.3.A' means 'Proactive, Level 3, Element A', and 'I.6.D' means 'Interactive, Level 6, Element D'. The elements are shown in Figures 3a, 3b and 3c.

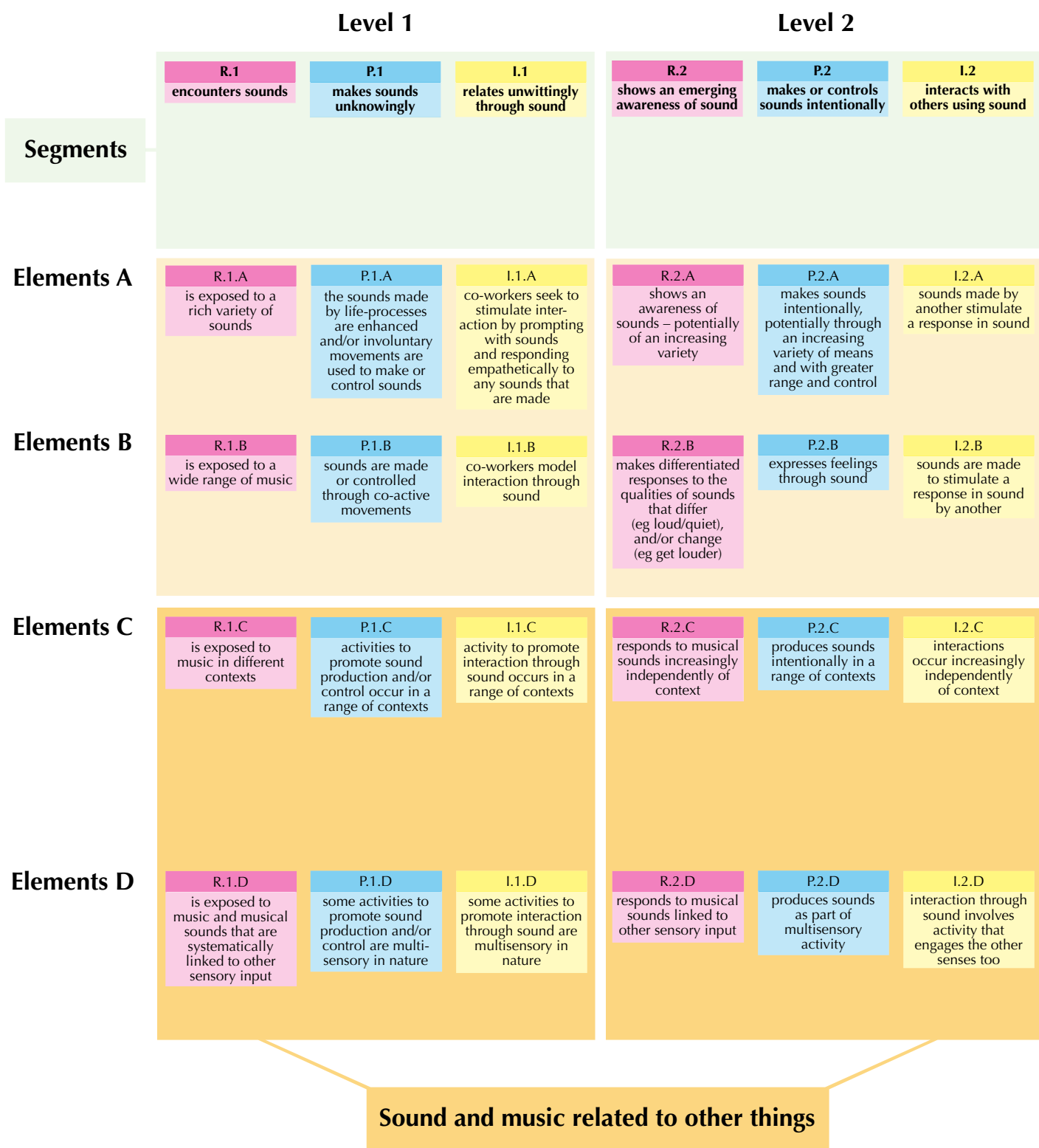


Figure 3a. The elements pertaining to levels 1 and 2 of the Sounds of Intent framework of musical development.



Figure 3b. The elements pertaining to levels 3 and 4 of the Sounds of Intent framework of musical development.

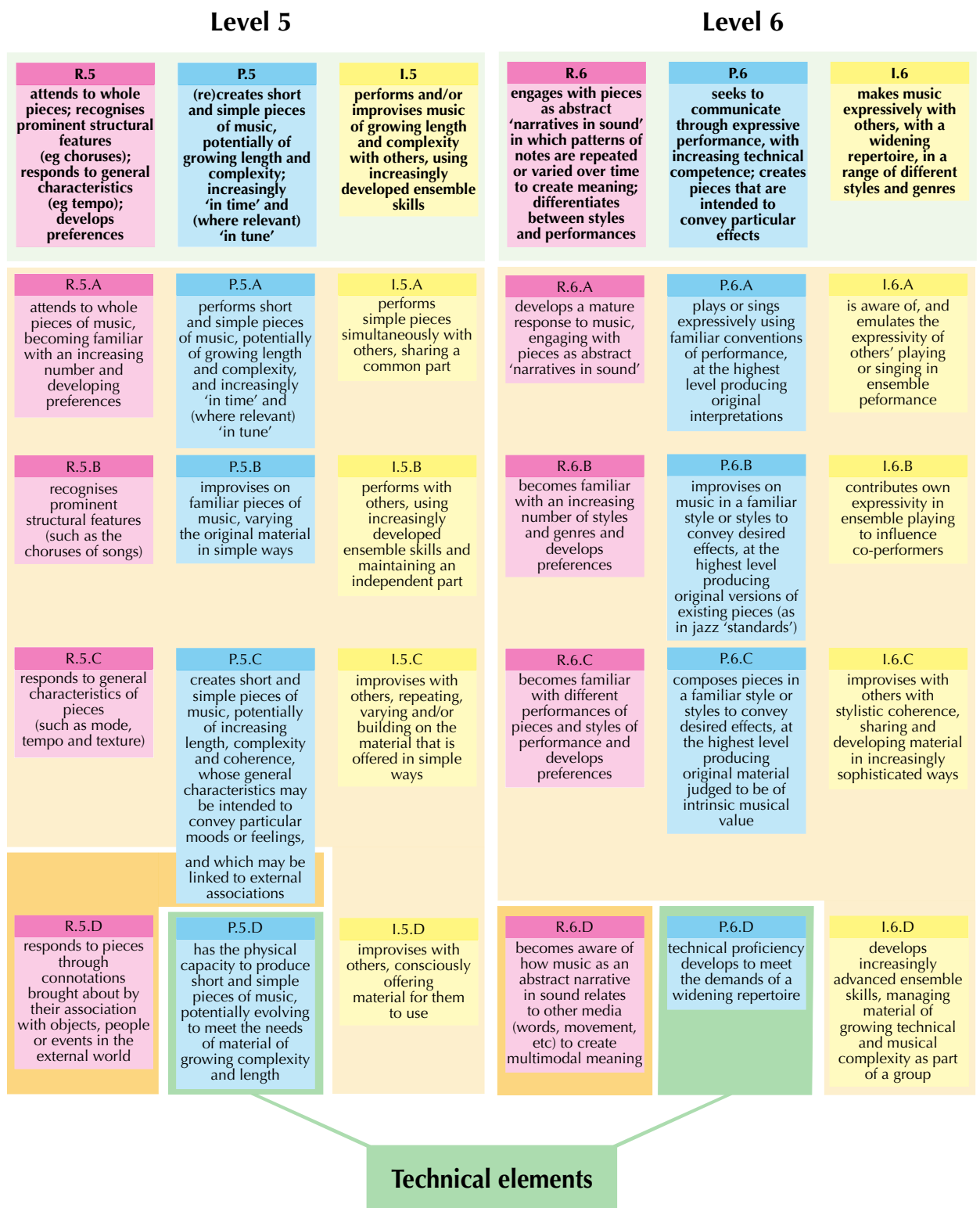


Figure 3c. The elements pertaining to levels 5 and 6 of the Sounds of Intent framework of musical development.

6.10 The roles and contributions of those involved in the MIND project were related to one another as follows (see Figure 4):

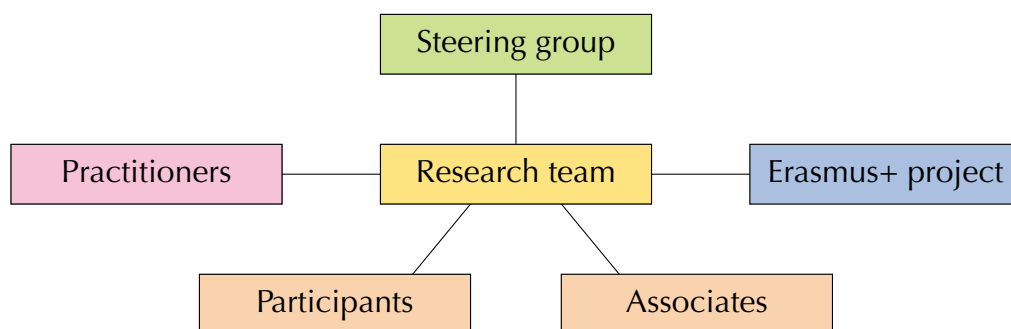


Figure 4. Organigram of the contributions of those involved in the research project.

6.11 The research was an initiative of the Applied Music Research Centre (AMRC) at the University of Roehampton, and was led by Adam Ockelford, with the majority of the fieldwork undertaken by two Research Officers: Rebecca Atkinson, a doctoral student at the AMRC and a director of Chiltern Music Therapy, and Katya Herman, a researcher at Chiltern Music Therapy, who covered Rebecca's maternity leave for a year. The project was guided by a steering group of parents and professionals (listed on p. ii), who met five times in the course of the research. In addition, there were visits to centres with particular expertise in treating, educating and caring for children and young people with Batten disease, including the Evelina London Children's Hospital, Great Ormond Street Hospital, UCL Great Ormond Street Institute of Child Health, The Royal Blind School in Edinburgh, and Heather House, run by SeeAbility, in Hampshire. Eleven music therapists, five other music practitioners, and nine teaching assistants were directly involved in the project, working with the participants and their families over the 32 months in which data were collected. Weekly sessions were scheduled during term time, with 36 planned to take place each year. However, due to illness and medical appointments, an average of 25 sessions were actually delivered.

6.12 Sessions took place at school, at home, or, in one case, at a residential centre. Members of the research team visited each participant once a term, and observed their engagement with music and the work of their therapist or teacher. These occasions provided the opportunity for gathering data first hand, to take videos for subsequent analysis, and to discuss and demonstrate strategies that were being developed and tested as part of the research. In addition, parents and practitioners were asked to make short recordings of their children in action, engaged in both musical and everyday activities, and to upload the clips using EthOS (a secure, online video app), to which the research team had access. This was intended to increase the amount of video available for analysis, as well as enhancing the ecological validity of the results by recording the children and young people in sessions when researchers were not present. The practitioners met with the research team twice, for training, feedback, discussion and the sharing of ideas.

6.13 The MIND study lasted for three years. The first four months were taken up with identifying families through the BDFA who indicated a willingness to take part in the study, finding suitable music therapists and teachers who were prepared to contribute to the project, establishing the steering group, setting up the EthOS account and planning the first round of visits to children's homes and schools. The period of observation lasted for 32 months, and qualitative evidence pertaining to RQ3 was amassed and analysed throughout. This informed the generation of strategies and resources that were tested and evaluated in a cyclical process of research, development and testing (see Figure 5).

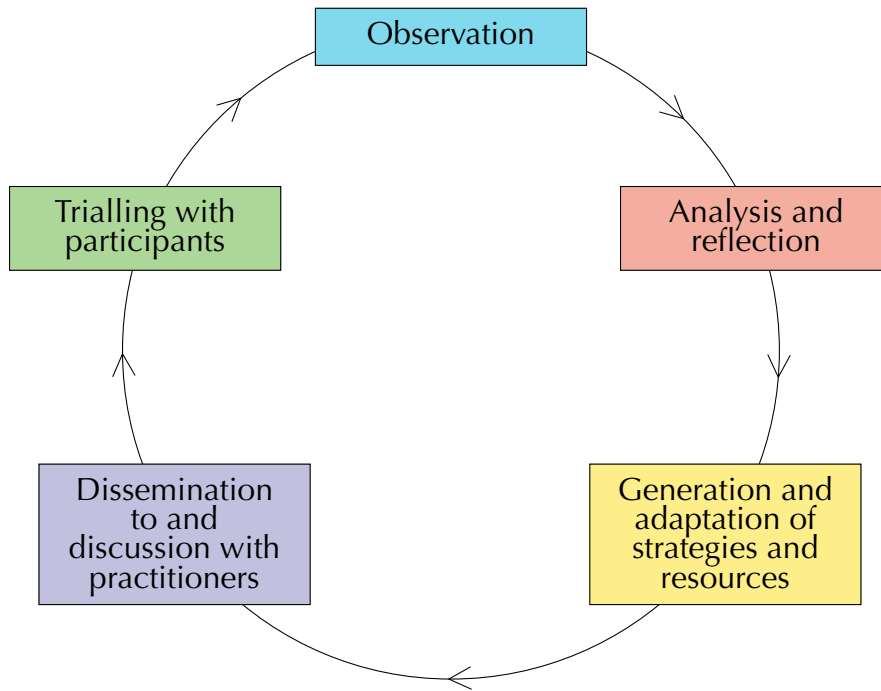


Figure 5. The cyclical process underlying the generation and adaptation of strategies and resources.

6.14 In relation to RQ2, baseline assessments using the Hamburg Scale and the Sounds of Intent framework were undertaken by the research team at the outset of the observation period in discussion with parents and the professionals who worked with participants. Thereafter, Sounds of Intent scores were taken termly (making eight in total) and Hamburg Scale ratings after 10 months and 22 months and at the end of the observation period (four in all), using information gleaned from direct observation, analysis of the video clips, and in dialogue with practitioners and children’s families.

6.15 The principal activities, events and milestones of the project are shown in Figure 6.

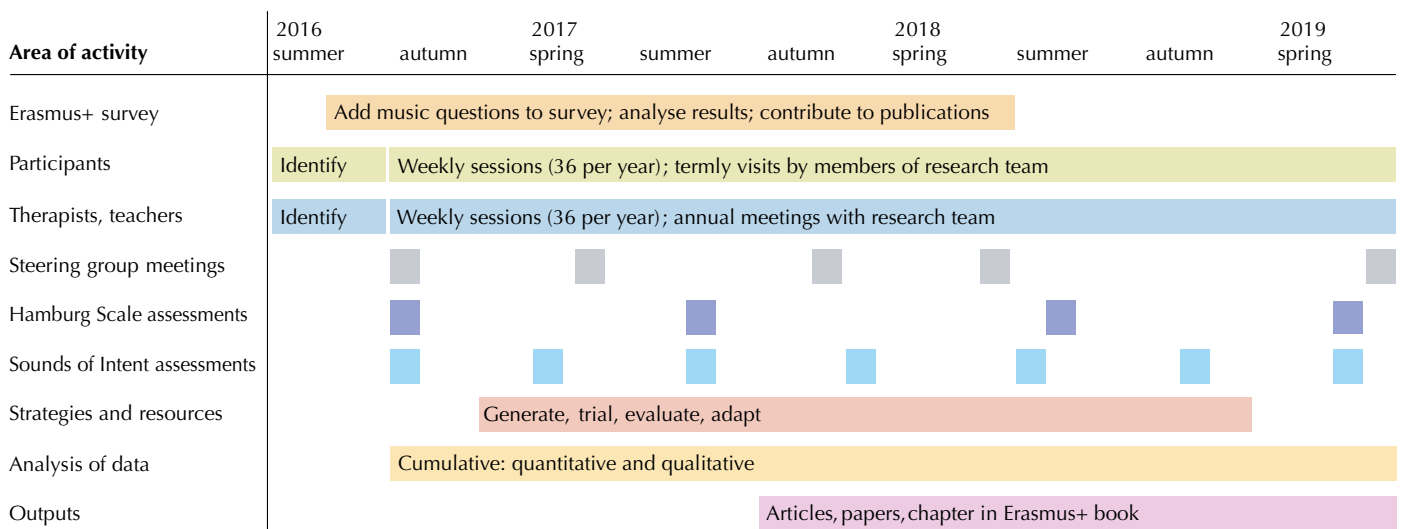


Figure 6. Gantt Chart highlighting significant activities, events and milestones in the project.

7 FINDINGS: RQ1

To what extent and in what ways do the level and nature of musical interests and abilities vary among the population of children with Batten disease?

- 7.1 Data relating to this RQ were gathered in two ways: first, through the incorporation of questions into the ‘JNCL and Education’ survey and interview study that were being organised as part of the international project ‘Juvenile Neuronal Ceroid Lipofuscinosis (JNCL) and Education’ (2014–2017), co-ordinated by Statped Midt in Norway; and second, by means of observation of the participants in the MIND study, and through discussion with them, their parents and teachers.
- 7.2 The JNCL and Education survey and interview study, funded by the Erasmus+ programme, elicited information from the parents of children and young people with CLN3 and the professionals who work with them, from six European countries – Denmark, Finland, Germany, Norway, England and Scotland – and the USA. The questions pertaining to music, listed in Appendix 1, were derived from the *Focus on Music* studies¹⁵, which examine the musical interests and abilities of children and young people with particular forms of visual impairment associated with different syndromes. There were 183 respondents to the JNCL survey, 110 parents and 73 professionals. Of these, 129 people (65 parents and 64 professionals) completed the music portion. There was some overlap in ratings, as several affected individuals were appraised both by their parents and associated professionals. The full results were published in 2019 in the edited collection *Juvenile Neuronal Ceroid Lipofuscinosis, Childhood Dementia and Education: Intervention, Education and Learning Strategies in a Lifetime Perspective*.¹⁶ A summary, ‘Overview of advances in educational and social supports for young persons with NCL’ is awaiting publication in *Biochimica et Biophysica Acta (BBA) – Molecular Basis of Disease*.¹⁷
- 7.3 The most striking finding of the Erasmus+ survey is that, as children and young people with CLN3 grow up, music is perceived by parents and professionals to gain dramatically in importance in their lives, at the expense of virtually all other activities, and particularly those that involve physical participation such as sport and dance. Audio books also gain greater prominence, though they lie some way behind music in their perceived significance. See Figure 7.

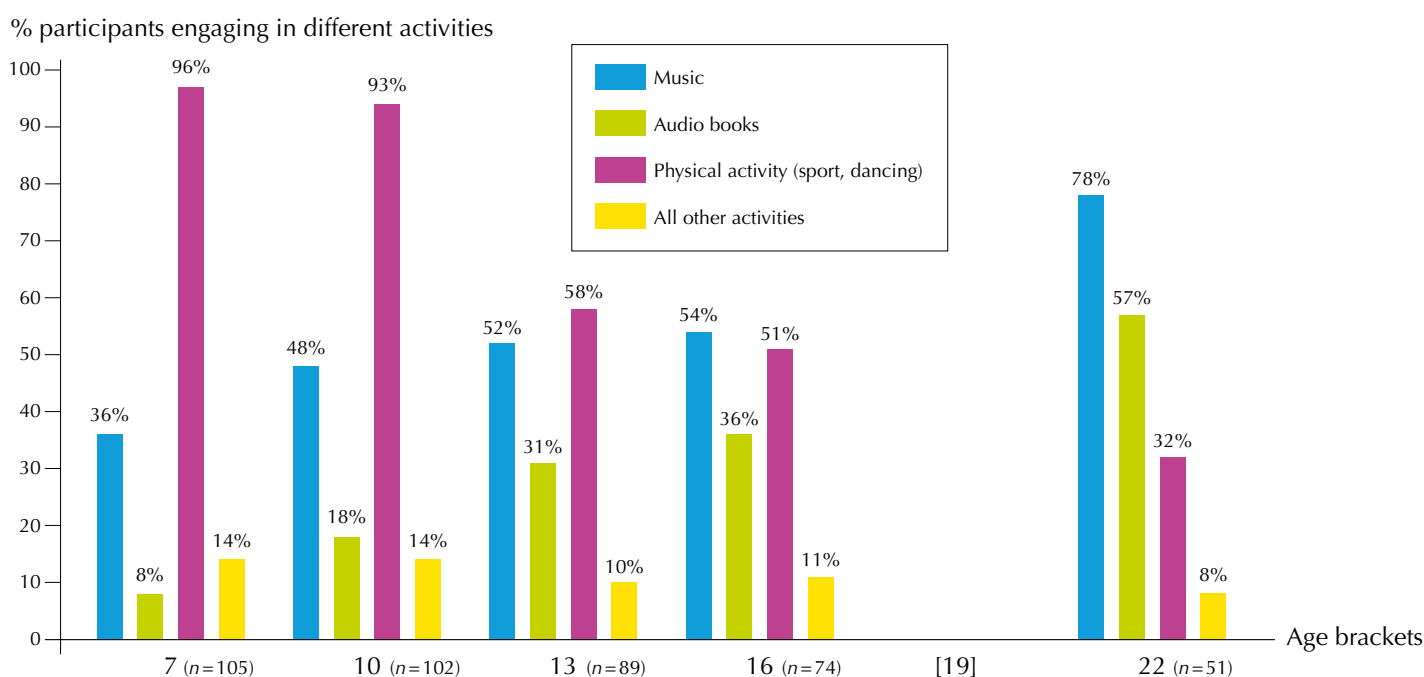


Figure 7. The importance of music for children with CLN3, at different ages, in relation to other activities.

¹⁵ See *Focus on Music: Exploring the Musical Interests and Abilities of Blind and Partially-Sighted Children and Young People with Septo-Optic Dysplasia*, by Adam Ockelford, Linda Pring, Graham Welch and Darold Treffert, 2006; and *Focus on Music 2: Exploring the Musicality of Children and Young People with Retinopathy of Prematurity* by Adam Ockelford and Christina Matawa, 2009; both published by the Institute of Education, London.

¹⁶ Edited by Stephen von Tetzchner, Bengt Elmerskog, Anne-Grethe Tøssebro and Svein Rokne and published by Snøfugl Forlag in Melhus, Norway.

¹⁷ Authors: Bengt Elmerskog, Anne-Grethe Tøssebro, Rebecca Atkinson, Svein Rokne, Barbara Cole, Adam Ockelford and Heather Adams.

- 7.4 How do we account for this marked change? Discussion with parents in the MIND project and at the steering group suggested that three factors were at play, related to participants' visual, cognitive and physical decline, which tend to occur in that order (Kohlschütter, Laabs and Albani, op. cit., p. 869). The great majority of activities in which children and young people engage rely to a greater or lesser extent on all three functional areas operating adequately. Hence as the capacities to see, think and move diminish, so do the opportunities for involvement in educational and recreational pursuits. However, as the results of studies using the Sounds of Intent framework (shown in Figure 2) demonstrate, in cognitive terms, even profound disability need not prevent the enjoyment of music (see Ockelford, et al., 2011). Moreover, observations in the reactive domain indicate that listening, which does not require physical activity, is as valid and meaningful a way of engaging with music as any other (Ockelford, 2008). And blindness – even when it is associated with learning difficulties – need be no barrier to active musical participation, understanding or appreciation (Ockelford, 2008). So music, with its capacity for differentiated engagement, is ideally suited for those with Batten disease, since the ways in which those with the condition can be involved in musical activities can change as neurodegeneration progresses.
- 7.5 This is shown in the responses of parents and professionals to the Erasmus+ survey. It was reported that around half of the children and young people with CLN3 played, or had once played, an instrument, with many trying two or three. The most popular instruments were the keyboard (including the piano), drums and other percussion, and the guitar. The majority of children started playing around the age of eight and continued for five years or so, typically never getting beyond the level of beginners. It seems that children began to engage more with active music-making as their eyesight deteriorated, but generally stopped when they could no longer read print music notation (and only one young person was reported to have attempted to learn Braille music). There was no mention of playing by ear, which may be a reflection of traditional attitudes to instrumental tuition that place a heavy reliance on reading music. When fine motor skills began to decline, playing accurately became increasingly difficult, and there were some reports of parents encouraging their children to switch to instruments that may be physically easier to manage. There were no references to gesture-based music technology, however. Children's singing was said to be at its best (and on a par with fully sighted peers) around the age of 12. And around one in five of children and young people were reported to make up (or to have made up) their own music, which occurred largely between the ages of 11 and 15. As formal music tuition declined in importance, music therapy, which requires neither music literacy nor technical precision, came to fulfil a more significant role in young people's lives. Thirty-six of those with CLN3 – around a third – were said to be having or to have had music therapy at some point in their lives ($n = 102$), and 86% of parents ($n = 38$) and 78% of professionals ($n = 19$) indicated that this had a positive impact, primarily as a source of stimulation and comfort. The median age for starting therapy was 14, and the mean duration of the intervention was eight years. In later adolescence and early adulthood, just listening to music was reported to become increasingly important, both as a social and recreational activity in its own right, and for stimulation (79%, $n = 107$), comfort (78%, $n = 106$), succour in certain situations (78%, $n = 103$) and for emotional regulation, particularly the practice of repeatedly playing favourite songs (91%, $n = 107$). Overall, 85% of parents reported that music had (or once had) a high or very high impact on the lives of their children.

8 FINDINGS: RQ2

Do musical abilities decline as other cognitive abilities are lost, and, if so, what is the nature of the correlation between them, or does the capacity to understand, appreciate and make music take a different course?

- 8.1 We begin by considering the findings of the MIND research project for the cohort of 10 participants who were observed for the full 32 months of the study: three with CLN2, three with CLN3, two with CLN5, one with CLN6 and one with CLN8. The Hamburg Scale results are presented first. Information regarding the participants' seizure activity has been omitted, as their epilepsy was often controlled (more or less effectively) through medication. Data were obtained by the Research Officers through observation (in person and on video), and in discussion with parents and music practitioners.
- 8.2 Participants were assessed using the Hamburg Scale at 0 (baseline), 10, 22 and 32 months (see Figure 6). The mean scores for intellectual capacity, motor skills, expressive language and vision are shown in Figure 8. There is sharp decline of 64% over the 32 months, equating to an average reduction in function of around 24% per year.¹⁸

¹⁸ Given that the Hamburg Scale comprises a set of discrete categorical sub-scales, such percentages should be treated cautiously, but are of value in indicating trends and for the purposes of comparison.

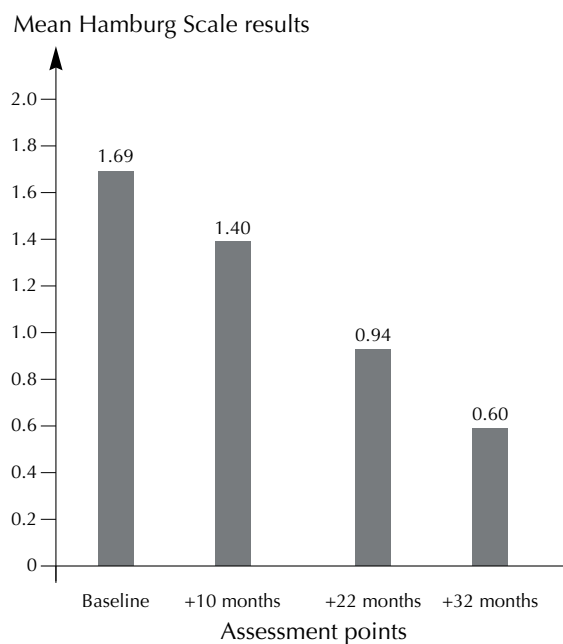


Figure 8. Mean results from the Hamburg Scale in relation to intellect, motor skills, language and vision ($n = 10$).

8.3 Domain by domain, the results break down as follows – see Figures 9 and 10 – with the greatest loss being that of vision (80% over the 32 months, which equates to 30% per year), followed by language (79%/30%), motor skills (58%/22%) and intellectual capacity (57%/21%). To put these figures in perspective: over the lifetime of the project, participants’ functional capacities ranged from ‘normal’ in one domain or more to what appeared to be total loss. That is to say, the levels of impairments within the group as a whole ranged from none to profound, with a weighting towards the latter; for most, the disease was at an advanced stage.



Eva plays the bells with her feet ...



... and makes music on the iPad with her hand

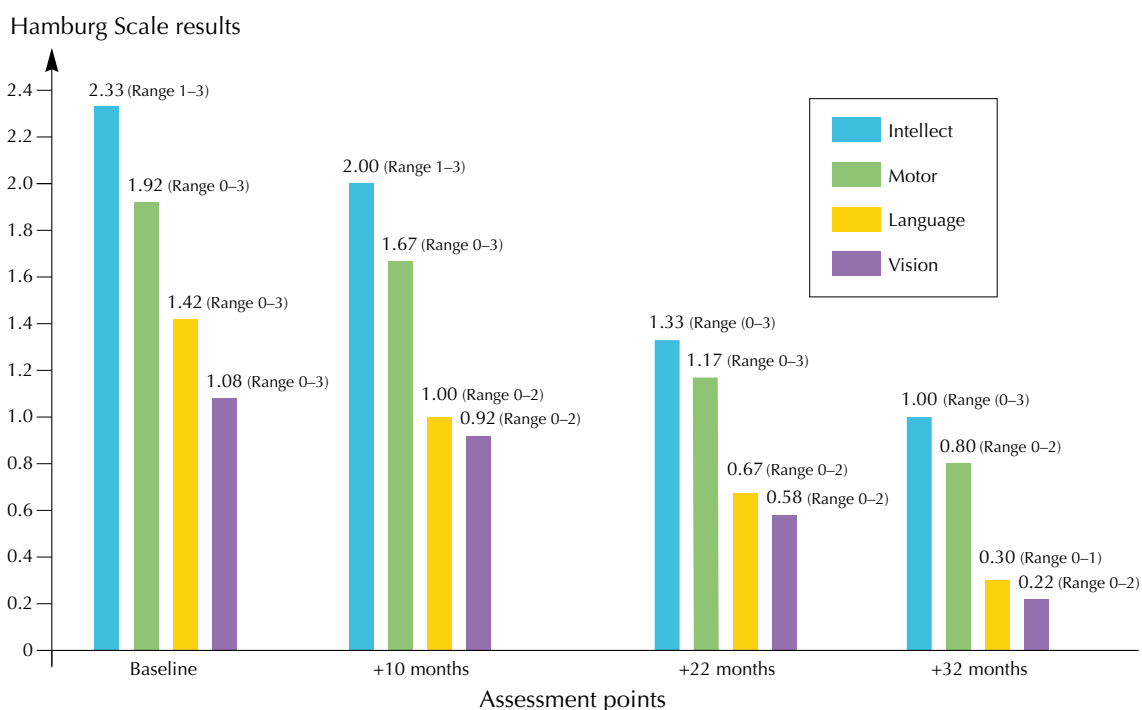


Figure 9. Mean results from the Hamburg Scale, domain by domain, over 32 months ($n = 10$).

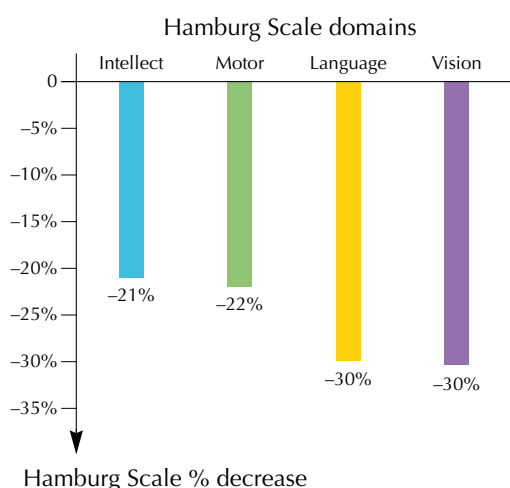


Figure 10. Average functional deterioration per year, domain by domain of the Hamburg Scale ($n = 10$).

8.4 Sounds of Intent levels were assessed eight times, at periods of four-and-a-half months (or as near to this as was feasible). Assessments were undertaken using the protocol available on the website (www.soundsofintent.org). This takes a score for engagement, ranging from one to five, and a score for consistency, which also extends from one to five, and multiplies them, giving a range per level (and per domain) of 1–25. The descriptors against which judgements are made vary from element to element, but all follow the same underlying pattern. For example, element P.2.B is defined as: ‘makes sounds intentionally, potentially through an increasing variety of means and with greater range and control’, see www.soundsofintent.org/sounds-of-intent?level=P2. The engagement criteria are:

1. Intentionally makes at least **one** type of sound
2. Intentionally makes at least **two** different sounds
3. Intentionally makes at least **three** different sounds
4. Intentionally makes at least **four** different sounds
5. Intentionally makes at least **five** different sounds or more

And the consistency descriptors are:

1. Given the appropriate opportunities, intentional sound-making is **rare** (occurring on around one in eight occasions or fewer)
2. Given the appropriate opportunities, intentional sound-making is **occasional** (occurring on around one in four occasions)
3. Given the appropriate opportunities, intentional sound-making is **regular** (occurring on around one in two occasions)
4. Given the appropriate opportunities, intentional sound-making is **frequent** (occurring on around three in four occasions)
5. Given the appropriate opportunities, intentional sound-making is **consistent** (occurring on around seven in eight occasions)

So a child who, for example, **frequently** makes **three** different sounds would be given a Sounds of Intent rating of $4 \times 3 = 12$. As a percentage of what is possible, this equates to $12/25 = 48\%$. In order to make a continuous scale, extending from Level 1 to Level 6, such percentages are normalised across a range from 0.0–1.0. Hence the child concerned (who is functioning at Level 2) would have a Sounds of Intent score of 2.48.

- 8.5** To set the scene, we begin by taking a global snapshot of participants' capacity for musical engagement among those who were observed for the full 32 months ($n = 10$). The mean of all Sounds of Intent scores across the three domains (reactive, proactive and interactive) was 3.90, with a range extending from 1.40 to 6.96; that is to say, participants' musical abilities varied from someone who is encountering sound and music, without making an observable response, to a young person who has a mature understanding of music, and is able to create or reproduce music expressively and persuasively. In other words, participants in the study exhibited the full range of potential for engagement with music. However, with an average age of 10, the mean musical capacity of the cohort was two Sounds of Intent levels below what would be expected neurotypically (Ockelford and Voyajolu, 2019).
- 8.6** Analysing musical engagement domain by domain yields the following results, based on the cohort ($n = 10$) who were observed for the full 32 months (see Figure 11). The range of each is from Level 1 to Level 6. The differences between average reactive and proactive, and reactive and interactive scores are highly statistically significant (using Tukey's HSD test, R v P, $p = .003$ and R v I, $p = .009$). There is no statistical difference between mean engagement in the proactive and interactive domains, however. The tendency of listeners' responses to music to be more advanced than their capacity to create new pieces, or to play or sing, on their own or with others, is found in both disabled and neurotypical populations (Cheng, Ockelford and Welch, 2009; Voyajolu and Ockelford, 2016). We can assume that this tendency is compounded in the musical development of those in the later stages of Batten disease, when severe motoric challenges constrain the ability to play instruments.

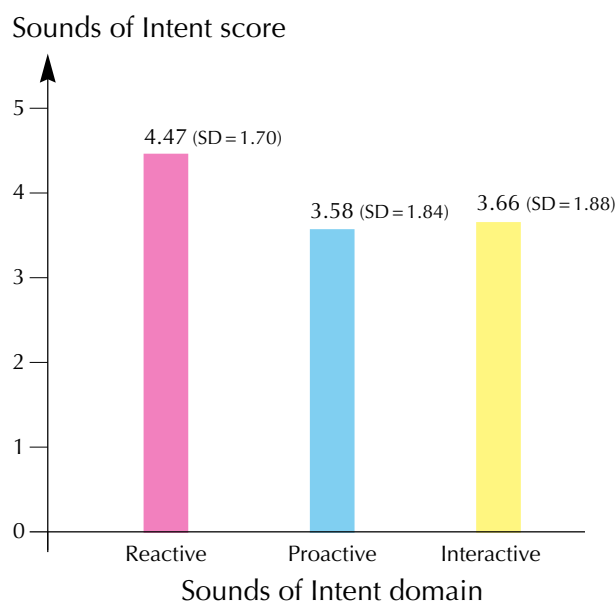


Figure 11. Mean Sounds of Intent scores by domain over the lifetime of the project ($n = 10$).

8.7 We consider next how participants' capacity to engage with music changed over the course of the 32 months of the project. Averaging their Sounds of Intent scores over all three domains (reactive, proactive and interactive) for each of the eight assessment points yields the following data (see Figure 12). A linear regression indicates a reduction in the ability to engage with music of around 2.3% a year – less than a tenth of the rate of decline of other skills and abilities (24%). However, neither this trend nor any of the differences between scores is statistically significant. That is to say, the fluctuations in the figures can be attributed to what is effectively random variation in the data, and it is reasonable to assume that there is no underlying pattern of change. Hence, while the cohort of participants as a whole shows a small deterioration in their capacity to engage with music in a period of just over 2½ years, it would not be safe to ascribe this to the progression of their disease. It would be necessary to gather more data over a longer period of time to be sure of what is occurring.

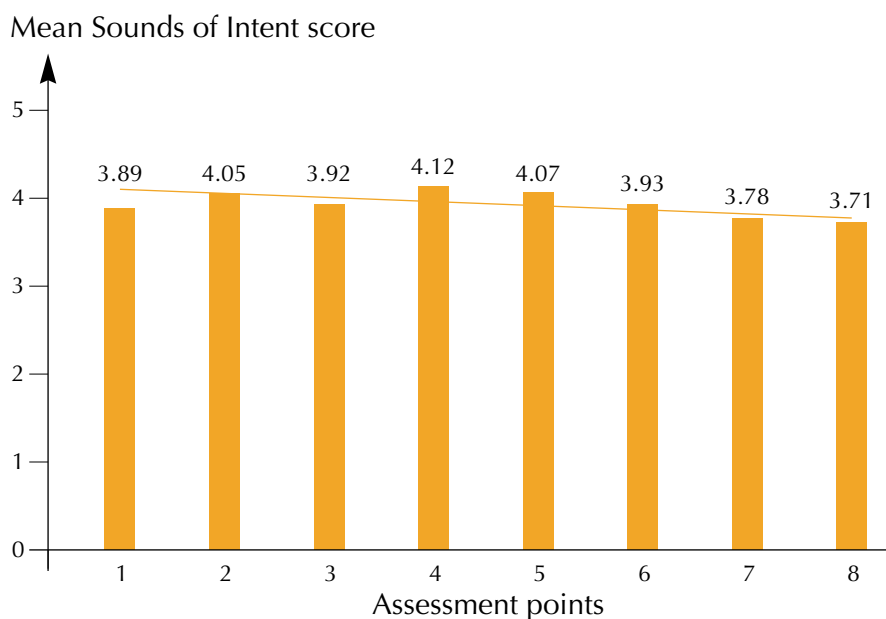


Figure 12. Changes in average Sounds of Intent scores over 32 months ($n = 10$).

8.8 Taking the Sounds of Intent scores domain by domain, it is evident that the majority of the general decrease in the capacity to engage with music occurs in the reactive domain, with minimal change occurring in the proactive and interactive domains, although none of the differences or trends within domains is statistically significant (see Figure 13). Despite this, it is of value to consider why a greater degree of reactive change was observed, since the result is arguably counterintuitive (we have already postulated that, with failing motor skills but intact hearing, participants' responses to music would show a slower decline than the capacity to perform or make musical sounds). One possibility is that musical reactivity fell at the greatest rate for the simple reason it had furthest to fall, although, as we shall see, given that responses to music can still be observed when ability to make organised sounds has disappeared completely, suggests that, at best, this can offer only a partial explanation. A further possibility is that participants' reactions to music and musical sounds will become harder to determine as their capacity to move and speak diminishes. Hence internal responses to music may conceivably persist, more or less unaffected by deterioration in other areas – it is the physical manifestation of these that becomes harder for observers to read (see Ockelford, 2012, pp. 75–84, and cf. O'Kelly, et al., 2013).

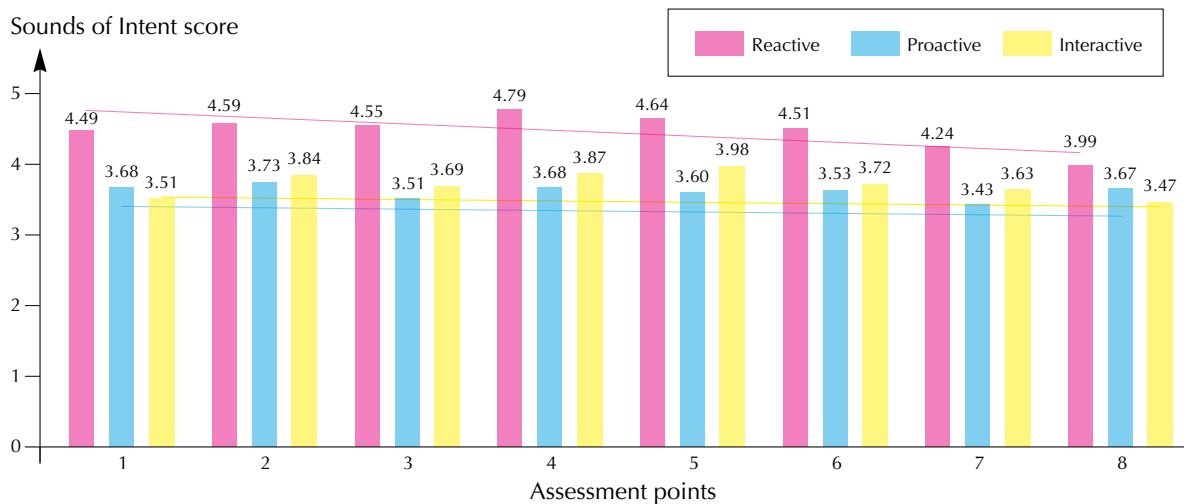


Figure 13. Changes in average Sounds of Intent scores across all three domains over 32 months ($n = 10$).

8.9 To summarise the findings pertaining to RQ2 up to this point: over a period of 32 months, there was a marked deterioration in the average day-to-day functioning of participants ($n = 10$), with a general loss of intellectual capacity, motor skills, language and vision of around 24% per year. In comparison, their ability to engage in music, reactively, proactively and interactively only fell by around a tenth as much, 2.3%, and this decline was not statistically significant. Yet it is also the case that some participants functioned musically at levels characteristic of children who are much younger, as in the first few months of life. Since (we can assume) their musical development would have progressed normally until the onset of the first CLN symptoms, which would have occurred at the earliest at 18 months, evidently there must have been a significant loss of musical capacity at some stage. How can we account for this apparent anomaly?

8.10 To better understand the issue, we can take a different approach and compare each child's average Sounds of Intent level over the 32 months of the study with their mean Hamburg score, using the sub-scales intellect, motor skills and language. (The vision sub-scale needs to be omitted for the purposes of this comparison, since having less vision is likely to enhance listening skills and musical engagement – see Ockelford, et al., 2006; Ockelford and Matawa, 2009.) A meaningful relationship between the two forms of data can be calculated by considering each value as a ratio of 'neurotypical' expectations. For Sounds of Intent levels, such expectations are based on participants' ages, using the sigmoidal trend of musical development in the early years identified in Ockelford and Voyajolu (2019), and extrapolating from it to obtain predicted levels in later childhood and adolescence (see Table 3). The Hamburg scores can be converted to ratios by dividing each by the maximum available score (which equates to having no symptoms of the disease). Given that three sub-scales are being used, each with a range of 0–3, this means that raw values have to be divided by nine.

Age	Sol level	Age	Sol level	Age	Sol level
0	2.00	7	5.25	14	5.95
1	3.20	8	5.38	15	6.03
2	3.90	9	5.49	16	6.11
3	4.40	10	5.59	17	6.19
4	4.70	11	5.68	18	6.27
5	4.90	12	5.77	19	6.35
6	5.10	13	5.86	20	6.43

Table 3. Age-related expectations in relation to Sounds of Intent levels (after Ockelford and Voyajolu, 2019).

8.11 The results are as follows. Over the 32 months of the project, participants' average capacity for musical engagement across the three domains (reactive, proactive and interactive) is consistently higher than their mean Hamburg score (intellect, motor skills and language sub-scales): 67% as opposed to 45% of neurotypical expectations. This difference is statistically significant, $t(11) = 5.43$, $p = .0002$. See Figure 14.

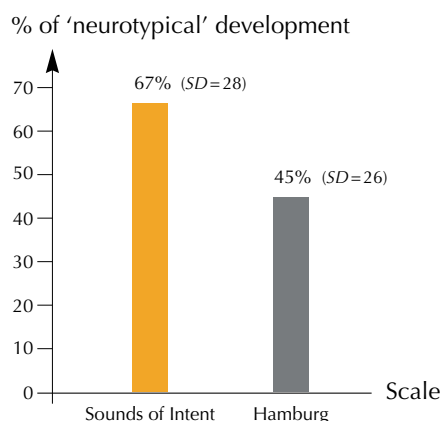


Figure 14. Relationship between mean Hamburg scores (intellect, motor skills and language) and Sounds of Intent levels (reactive, proactive and interactive), over the lifetime of the project, as percentages of what would be expected in neurotypical development, $n = 12$.

8.12 Moreover, participant by participant, there is a close connection between musical and other-than-musical capacities; correlation coefficient $R = 0.86$, $p = .0003$ (see Figure 15). That is, both variables decrease together in a strong, robust linear relationship. With regard to the coefficient of determination, $R^2 = .75$. This means that 75% of the variation in levels of musical engagement can be attributed to the progressive impact of Batten disease. Other factors therefore must account for the remaining 25%. However, neither the age of participants nor their CLN variant is related in a consistent way to the capacity to engage with music. For example, participants with CLN2 appear across the full range of the distribution, while different ages are spread throughout, with no discernible trend.

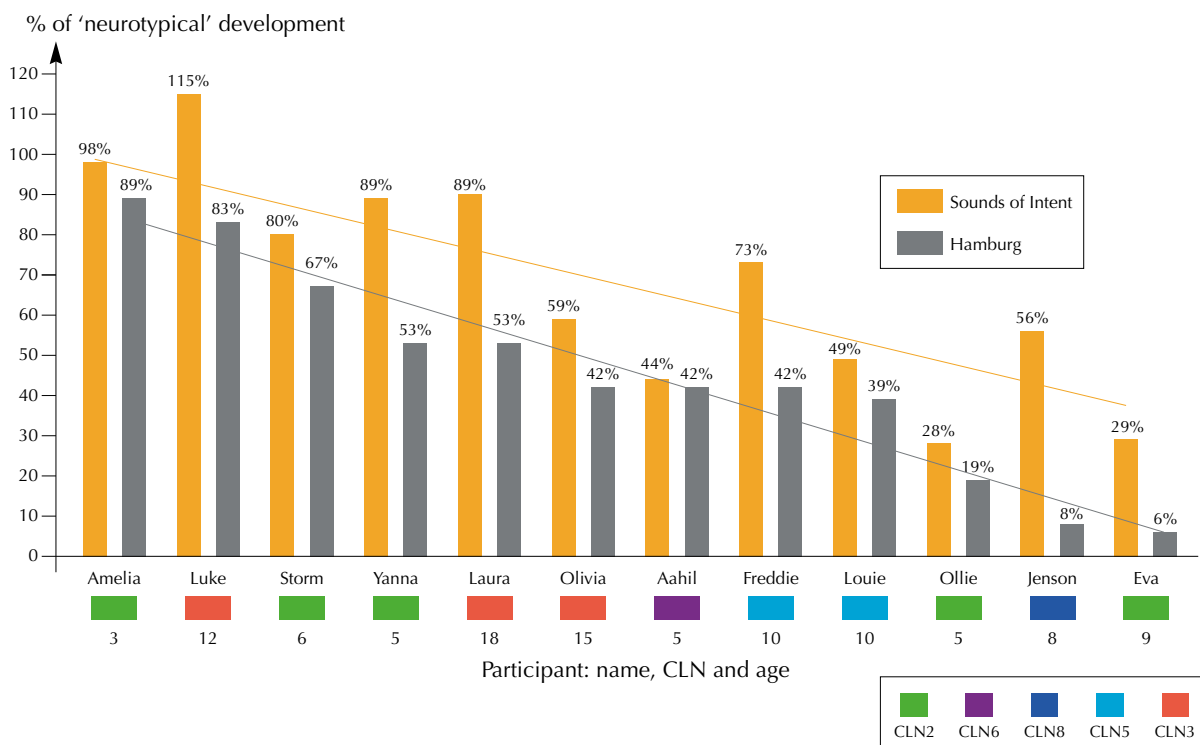


Figure 15. Relationship between Hamburg scores (intellect, motor skills and language) and general Sounds of Intent levels.

8.13 So we can conclude that other factors must be playing a part, in particular individuals' levels of musical interest and their motivation to engage with it, reactively, proactively and interactively. These impulses will in turn be heavily influenced by children and young people's living and learning environments at home and at school (or centre), including the capacity of family and carers to engage with them musically. Importantly, from the point of view of those planning music services, whether educational, therapeutic or recreational, the findings suggest that, despite the devastating and inexorable progress of the disease, interventions have the potential to make a real difference: in simple terms, around a quarter of a child or young person's capacity to engage with music is open to external influences. This can explain the apparent discrepancy between the data illustrated in Figure 15, which show levels of musical engagement declining more quickly than participants' Hamburg scores (albeit from a much higher starting point), and the longitudinal findings of the 32 months of the MIND study, in which levels of musical engagement show only a modest decline in contrast to the rapid deterioration of functioning in other areas (see Figures 8 and 12). While the end is inevitable, the trajectory towards it is not, and the musical interventions undertaken in the course of the MIND study appear to have had a considerable impact.

8.14 Finally, it is important to note that participants' levels of engagement with music, even when the disease is having a significant impact in terms of understanding, movement and vision, may potentially be higher than we would typically expect for young people of their age (for example, Luke – see Figure 15). Whatever a child or young person's level of musical engagement, the reactive domain dominates (see Figure 16), and in the most advanced stages of the disease it may be the sole avenue of musical participation that remains open.

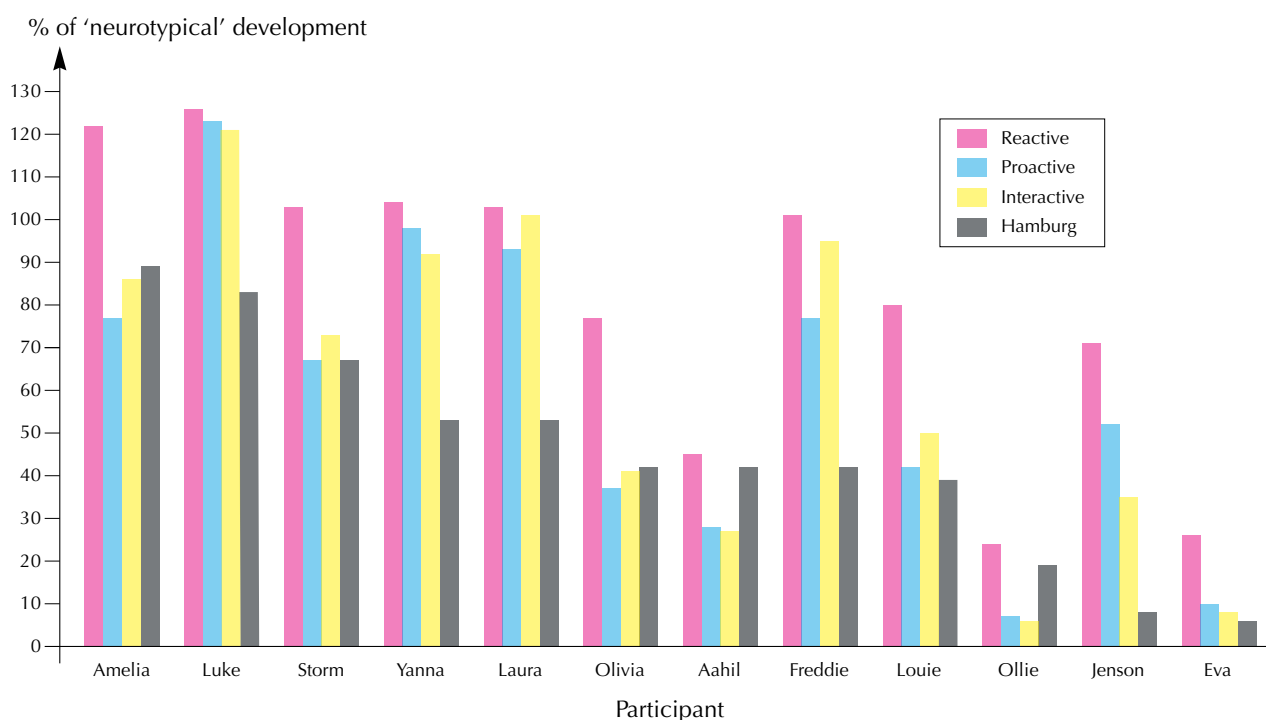


Figure 16. Relationship between mean Hamburg scores (sub-scales of intellect, motor skills and language) and participants' engagement with the three Sounds of Intent domains.

9 FINDINGS: RQ3(A)

To what extent and in what ways can music be used as an intervention to promote the continued use of expressive language when speech is in decline (or has ceased)?

9.1 Communication through language takes two forms – receptive and expressive – and parents participating in the Erasmus+ survey were asked to compare how their children’s abilities in these domains compared at different ages, as their disease progressed (Tøssebro and von Tetzchner, 2019, p. 100). During the early years in primary school, when the first symptoms of CLN3 usually become apparent, receptive and expressive language skills were rated as being comparable with most children. However, as they grew up, children and young people’s capacity to be understood gradually declined, and most had more than one problem with speech, with 75% of parents reporting issues finding the right word and 42% describing disorders of prosody and articulation resulting in unintelligible rapidity of verbal production and stuttering ($n = 111$) – see Figure 17. A narrowing of vocabulary, with repetition of the same words, was said to become prominent during the late teen years. Yet language is particularly important for people who cannot see, and so we would expect the consequences of speech apraxia to be even more severe than would otherwise be the case. And so it proved to be: in the survey 43% of parents ($n = 106$) reported that issues with expressive language had a ‘high’ or a ‘very high’ negative effect on their children’s social lives.

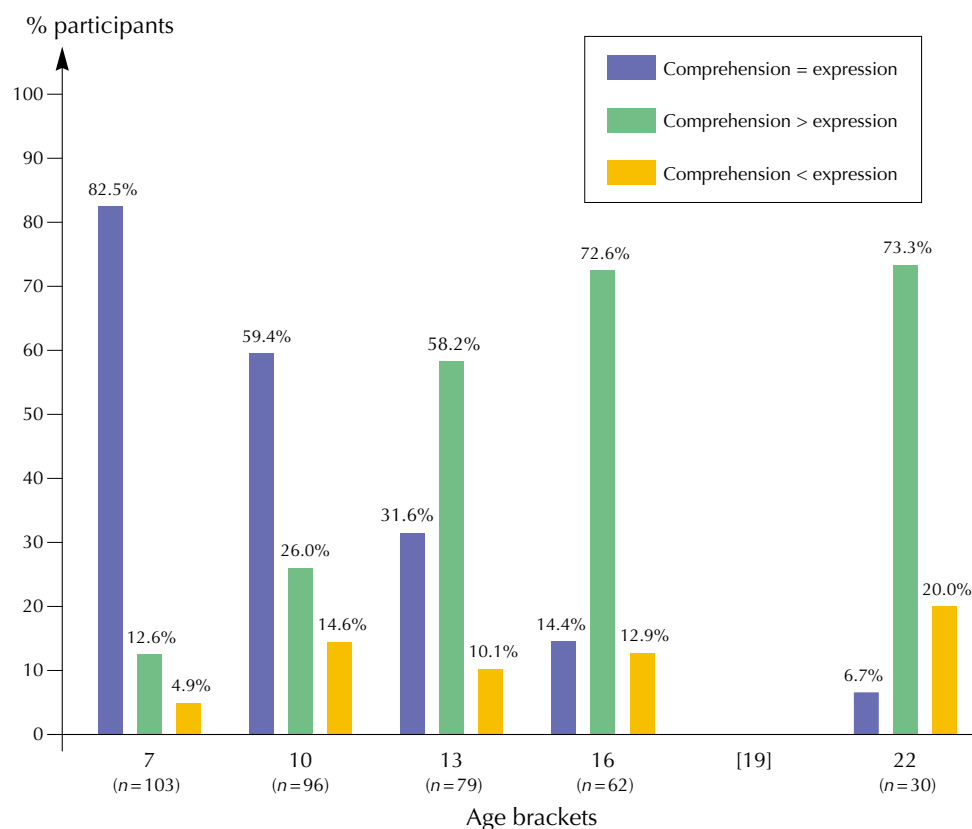


Figure 17. Parents’ perceptions of the changes in receptive and expressive language abilities as children and young people with JNCL grow up (after Tøssebro and von Tetzchner, 2019, p. 100).

- 9.2 Two main approaches were used to ascertain whether, and in what ways, music could support the maintenance of expressive language, when speech alone had declined or ceased. The first was the development and use of ‘micro-songs’. The second constituted activities based on the Neurologic Music Therapy techniques of Melodic Intonation Therapy, Oral and Motor Respiratory Exercises, Vocal Intonation Therapy and Rhythmic Speech Cueing (summarised in Thaut and Hömberg, 2014).
- 9.3 Micro-songs were developed by Adam Ockelford in the 1990s, and first published in *All Join In! A Framework for Making Music with Children and Young People who are Visually Impaired and have Learning Disabilities* (Ockelford, 1996). These recently appeared in a revised and expanded version called *Tuning In* (Ockelford, 2018), comprising 64 songs designed to promote wider learning and development, including movement, understanding, social skills and – in particular – language. This is achieved by consistently setting key words and everyday phrases to the same fragments of melody, thereby supporting learning and recall in a way that is especially effective in children whose verbal communication skills are limited. The songs are also of potential value to those who are unable to make speech sounds at all, since by reproducing the contour or rhythm of the melodic fragment, its associated word or phrase can be implied and potentially understood.
- 9.4 In neurological terms, micro-songs tap into the fact that language and music share some resources in the brain (Patel, 2010), and empirical research over several decades has suggested that in songs, music and language may be encoded together (Serafine, Crowder and Repp, 1984; Morrongiello and Roes, 1990; Schön, et al., 2010). However, music and language appear to have different routes ‘in’ and ‘out’ of the shared neural processing resources, and expert singers have the capacity to decouple the two forms of auditory communication (Wilson, et al., 2011). There is increasing evidence that a comparable effect can be caused by certain forms of dementia, when the capacity to speak is lost, but the ability to sing remains (see, for example, Gomez and Gomez, 2017; Baird and Thompson, 2019). Finally, it seems that the presence of a melody can increase phonetic recognition, and a tune can facilitate the learning and recall of attendant words, provided that the music repeats (Wallace, 1994). Repetition was key to the design of the micro-songs.
- 9.5 Thirty-nine of the micro-songs were felt to be of potential relevance to the children and young people in the MIND study. These are organised in five conceptual areas, as shown in Figure 18. An example of one of these, with motifs pertaining to ‘How are you feeling?’, is shown in Figure 19.



Figure 18. Map of the micro-songs that were available for use in the study.

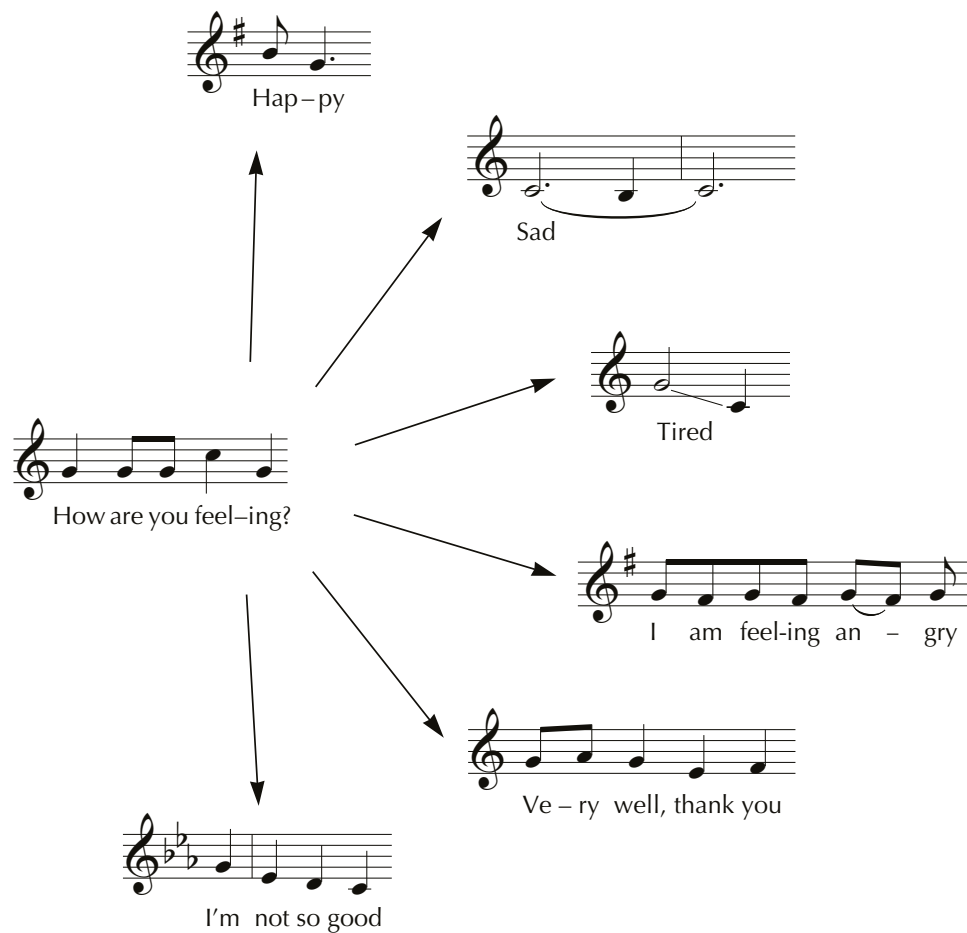


Figure 19. Examples of melodic motifs pertaining to 'How are you feeling?'

- 9.6 It was decided to pilot the songs with Lily, who at the time of the research found producing comprehensible speech very challenging, though her strong desire to communicate was evident in her persistence in attempting to express her thoughts and feelings. Crucially, her ability to sing extended fragments of favourite tunes, with melodic contour and rhythm intact (indicating that she was functioning at Level 4 in terms of the Sounds of Intent framework), was maintained. The micro-songs were systematically introduced during weekly music sessions that were organised as an early evening leisure pursuit at the residential school for the blind that Lily attended, in a group of around half-a-dozen other teenagers with a range of severe and profound disabilities. Sessions were structured around songs of social greeting ('Hello', 'Who's sitting next to me?', and the like), songs to promote movement and understanding from **Tuning In**, songs about feelings (shown in Figure 20), children's songs, contemporary hits, and songs of relaxation and closure (including 'Now it's time to rest', 'Music has finished' and 'Goodbye'). The age-appropriateness of some of the early childhood melodies to which Lily would return time and again (particularly *Twinkle, Twinkle, Little Star*) was discussed at length by the staff concerned, and a satisfactory solution was reached whereby each rendition was nominally dedicated to a younger relative or friend.
- 9.7 The results of the year-long intervention were very promising. Lily learnt the micro-songs rapidly and retained them well, including over the breaks of several weeks brought about by school holidays. As well as facilitating social greetings such as 'hello' and 'goodbye', choices such as 'yes please' and 'no thank you' were made in the context of the music sessions. And Lily was particularly fond of expressing her feelings, and eliciting similar responses from all those in the group. She was almost invariably happy! Above all, it appeared that Lily was hugely motivated to engage with others by being able to communicate and participate fully in the socially inclusive sessions. It was evident that, for Lily, it was important that all of those present, including staff, immersed themselves in musical activities, whose content and direction were often determined by Lily herself.
- 9.8 In addition, short songs with open-ended answers were developed in the course of sessions, to meet Lily's communication needs of the moment. For example, in response to the question 'What would you like to eat, Lily?', answers such as 'crisps', 'pizza' and 'Weetabix' were given. 'What would you like to drink, Lily?' produced responses such as 'squash', 'water' and 'Coke'. See Figure 20.

10 FINDINGS: RQ3(B)

To what extent and in what ways can music be used as an intervention to promote a day-to-day understanding of what is occurring, where, with whom and when?

- 10.1** The effect of the cognitive decline that is a central feature of the NCLs, which alone would have an increasingly negative impact on individuals' day-to-day awareness and understanding, is compounded by the loss of sight that is also generally characteristic of the disease, since most information about the world is gleaned visually (Colavita, 1974; Spence, Parise and Chen, 2012; San Roque, et al., 2014). Moreover, the majority of educational strategies that seek to ameliorate the consequences of learning difficulties are visual in nature (Ockelford, 2013). In an effort to address this issue, in the 1990s, Adam Ockelford led a team working at the (then) Royal National Institute for the Blind in London that advocated the use of 'objects of reference' with visually impaired pupils and students who have severe learning difficulties (Ockelford, 1994). 'Objects of reference' had been devised in the 1960s by the Dutch pedagogical pioneer Jan van Dijk to facilitate learning and communication with children who were deafblind, whose number had risen sharply following an epidemic of rubella.
- 10.2** Objects of reference can be defined as items that have acquired symbolic meaning, and that can be identified through touch. At their simplest, they may comprise everyday things that form part of whatever is represented, such as a flannel used to indicate 'bath time', for example, or a key used to denote 'home'. Other symbols may be more abstract, and their meaning learnt through association. For instance, a particular scarf may come to represent a certain relative or friend. As well as having a recognisable texture and shape, objects of reference may have other sensory qualities that contribute to their identity, such as a colour or scent, or they may make a particular sound. 'Swimming' may be indicated by trunks smelling of chlorine, for example, and 'going on the swing' by a jangly chain.
- 10.3** The RNIB research team's main contribution to the conceptual development of communication systems using objects of reference was the realisation that the practicalities of functioning in everyday life require information about **what** is occurring, **where**, **with whom** and **when** (Ockelford, 2001). To achieve this, three categories of objects were established that pertained to **activities**, **places** and **people**. The fourth area – **time** – was conveyed through sequencing two objects or more, through which it was determined that a particular activity would occur before or after another.
- 10.4** Objects of reference quickly came to be accepted in generic special schools for pupils with learning difficulties across the UK as a means of facilitating interaction, learning and understanding for their pupils, particularly those who were visually impaired or who had visual processing difficulties. Their principal use in the context of pupils' receptive communication was to indicate what was happening next, with objects sometimes presented in series, forming tactile timetables; as expressive language, objects were most often utilised to support (forced) choice making. The use of objects of reference also became embedded in regional specialist schools that focused on educating blind and partially sighted children with additional disabilities, such as Linden Lodge School in Wandsworth, the WESC Foundation in Exeter, and the Royal Blind School in Edinburgh.
- 10.5** Since virtually all children beyond the early years who are in the advanced stages of Batten disease attend a special school, it is inevitable that objects of reference are among the strategies that are often considered to support their learning and communication, when the decline of cognitive, proprioceptive and fine motor skills makes the use of Braille problematic. However, the challenge of physically manipulating items and absorbing information through touch means that objects of reference can become increasingly difficult to use too. This issue is also faced by children with other special educational needs, particularly those with profound and multiple learning disabilities, whose movement is often very limited. A potential solution – at least in part – was suggested by Ockelford (1996; 2008), whereby sounds and music alone (without a tactile referent) could be made to function symbolically: so-called 'sounds of reference'. These can be used to denote **activities** (for example, a jingly bell to indicate that a music session is about to begin), **places** (for instance, windchimes placed at the entrance to a child's classroom), and **people** (who can indicate their presence non-verbally by using a particular personal soundmaker such as a small rainstick).

10.6 These approaches were all tried out with Lily in her music sessions at Linden Lodge School, using the [Tuning In](#) songs (see Figure 19), and all were successful in that limited context. Personal soundmakers were particularly helpful in indicating to Lily and the other participants in the group (who were all non-verbal and blind) who was present in the room. As with the micro-songs, the challenge proved to be embedding the approach across the school curriculum (and beyond, into life at home or residential centre). Some establishments, such as Stephen Hawking School in Tower Hamlets, where Yanna and Storm are pupils, already use particular pieces consistently to signal upcoming activities and to ease the potential anxiety that can exist during times of transition. For example, children come in to class every morning to the theme from [The Greatest Showman](#). Freddie and Louie's parents were observed using a similar approach at home, whereby different pieces of music were played at key times in the day, so letting the boys know what was about to happen as well as supporting their emotional regulation during activities that were not intrinsically enjoyable. Hence another recommendation arising from this report is that ideas for sounds of reference and guidelines for their use in everyday life should form part of the resources available to those teaching and caring for children and young people with Batten disease in its later stages. As with all augmentative communication strategies, the key is consistency in their use.

11 FINDINGS: RQ3(C)

To what extent and in what ways can music be used as an intervention to promote the recall of important memories?

11.1 To explore the potential impact of music on memory, the research team used information gathered from the Erasmus+ study from the accounts of parents and professionals to suggest techniques and activities to be implemented in the MIND research. Parents repeatedly stated that music could fulfil a special function in helping to revive memories for their children with Batten disease. For example, one parent noted that listening to music helped their daughter to recall special events:

If she went to that particular concert [she will 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.

Similarly, one professional reported that 'familiar pieces of music ... often sparked a memory of events from the past', while another observed that 'well known songs promoted his memory, comforted him and made him feel fine.' A third professional observed the following:

Music is one of her biggest interests, and she remembers several lyrics because she has sung them so many times ... the music helps with her memory.

One mother of a child in the study remarked:

One of the only days that she remembers what she's done at school is on a Wednesday after she's had 'music therapy with Beki'.

11.2 At the Royal Blind School in Edinburgh, 'memory books' are created for children and young people with Batten disease, to help maintain memories of favourite music bands, songs, occasions or events. These books are multisensory in nature and include objects associated with the musicians (tactile materials or 3D shapes), words from the songs in Braille, 'sound bites' of the songs on recordable units, and a CD of songs or associated sounds related to events, memories or significant trips. One 16-year-old with CLN3, who struggled to find the words for day-to-day objects and activities, could still remember the lyrics of entire songs. With the help of her learning support assistant, she constructed her own tactile books, which she continues to revisit as a way of recalling favourite memories.

From the start, she played an active part in the process: rehearsing a song that she chose herself, recording it and then burning it onto a CD She then selected the tactile materials that she deemed relevant to each song or artist and attached them to each page. For a Paloma Faith song for example, she chose a red flower to represent the fact that Paloma Faith always wears flowers in her hair; for Sam Smith, it was a piece of leather, as he always wears leather trousers. Finally, she put the song lyrics in Braille as according to her teacher 'she is much more motivated to use Braille if she is writing the words of her favourite song, and is also more motivated to participate in art if she is making things to put in her special book'. This fully interdisciplinary approach involving music, English, Braille, and art was motivating for her and as her disease progresses further, the book will be there to aid her in remembering her favourite songs, through listening to her own voice and by feeling the tactile materials that she has so carefully chosen.

11.3 Music memory books were introduced to two of the participants with CLN3 in the study, and it was suggested that such books could be created that were centred on their memories of holidays, family members or important events. Creating and perusing the books were introduced as activities that could be undertaken at school or at home, and which had the potential not only to engage individuals in their education, but could also be used as a personal legacy of a young person's identity, preferences and memories. As one parent put it, this allowed music to serve as a 'bridge' to a past reality, in the final stages of the disease. It was hoped that, in the future, the books would also allow memories and experiences to be transferred from one education or care setting to another, when young people were no longer able to 'tell their own story', enabling new carers to become familiar with an individual's preferences and important life events, and to give them some sense of their identity.

12 FINDINGS: RQ3(D)

To what extent and in what ways can music be used as an intervention to promote emotional regulation and well-being?

12.1 One of the most distressing elements of Batten disease is the emotional anguish that children and young people can experience, and evidence gathered prior to the MIND project suggested that self-selected music may promote emotional regulation (Bills, et al., 1998), as it can do in neurotypical contexts (Saarikallio and Erkkilä, 2007), and offer relief at times of psychological crisis. In the MIND project, data were obtained using the 'EthOS' app on mobile phones, which enabled parents to capture video recordings of their children engaging in musical activities. The videos were categorised, and these categories were recorded using the tools available within the software. Within each category, the frequency of consistent or reoccurring themes was ascertained. Analysis showed that participants and their carers most often used music for relaxation, stimulation, as a motivation for movement and for improving mood. Around 30% of the sequences in the videos ($n = 126$) depicted positive behaviours such as smiling, clapping or laughing in relation to music, or hearing favourite songs.

12.2 All the 13 families in the MIND project reported that they regularly use music at home to support their child's emotional well-being. For example, when it came to regulating their mood and energy levels, one parent reported that:

We use music all the time to manage his moods: Amy Winehouse before bed, reggae at breakfast to perk him up. Rock if he's grumpy! It just feels instinctively like the right thing to do.

Another observed that:

We usually use music to create a calm, relaxing environment ... but a fast song with a strong beat will usually get her to open her eyes.

Four parents commented on the inverse correlation between their children's eyesight deteriorating and their increased appreciation of music. In these cases, music had become an increasingly significant pastime at home:

He listens to a lot more music now, and when one of his favourite songs comes on his eyes light up and get bigger. You can tell he's really listening and concentrating.

Two parents observed the importance of having regular access to their child's favourite music:

He likes to have the iPad on all the time. With music and nursery songs, or Peppa Pig. It really calms him down, and he gets so upset when we try to turn it off. ... He loves having his iPad for music and stories. He likes having access to his familiar songs: nursery rhymes, Thomas the Tank Engine, Toy Story ... It really calms him down when he's agitated or in pain.

One parent reported a significant decline in her daughter's general engagement, over the past year:

She's much less engaged ... more withdrawn, but music and films are still the things that capture her attention more than anything else. They give her something she can do and enjoy when everything else is so limited.

12.3 The parents of the young people with CLN3 commented on their children's experience of social isolation, the social significance of music in their lives, and its importance for their sense of identity.

Music connects her with her old self. It's a way to say 'This is what I'm all about.'

She's very sociable, but it's harder now for her to have a two-way conversation. Music is still a space where she can be surrounded by people, which she loves.

One of the real stumbling blocks with his visual impairment is his complete lack of social life. He feels very isolated, especially when his brothers are busy or away. He finds it hard to communicate easily with strangers, and tends to clam up. He loves being able to share his music on Soundcloud or the physical CDs that we can burn for him. He's always saying: 'I must give so and so a copy of my album!' It gives him an identity. ... Music is the one thing that binds it all together.'

12.4 Another theme that emerged was the value of using music at school to support pupils' participation: one parent reported that his son's teacher has started to incorporate more music in the classroom, based on the activities and instruments his music therapist uses 'and now he's getting merits all the time for his participation in class.' Another commented that although her daughter is:

Always very sleepy and sedated, there are certain songs that really get her attention. The other day, someone played an Ed Sheeran song in assembly and she opened her eyes straight away! As soon as the song finished, she closed her eyes again. That afternoon, they used the same song when she was swimming, and she was able to keep her eyes open the whole time.

12.5 Data were also gathered from the Erasmus+ study. Here, parents and staff reported that music helped to stimulate children and young people, to comfort them and to help them express themselves. For example, parents reported that 'music helps her to go to sleep ... it has a calming effect', how 'her mood is lifted by her favourite music ... she refuses songs she doesn't like, music makes her happy' and how music 'could bring relaxation and be a diversion'. Similarly, professionals in the survey said that 'music is used as a calming mechanism throughout the school day; it is a source of comfort during periods of high anxiety', and 'sounds and music were of high significance ... they also could produce a high level of excitement; it could have a calming effect during critical situations; sometimes it helped to start actions or interactions and we played music during the times of care.'

13 FINDINGS: RQ3(E)

To what extent and in what ways can music be used as an intervention to promote social inclusion through active musical participation using assistive music technology?

13.1 Five computer programs and music technology products were trialled by children and young people in the project, with the aim of being able to identify devices that parents, relatives or school staff can utilise to engage in music activities with their children and young people.

13.2 'Boom Ball'

The simplest of the devices that were evaluated, this vibrating speaker unit was purchased for 12 of the participants in the study. The size of a ping pong ball, the device can be attached to tables, containers, wheelchairs, and furniture to enhance the volume and enable everyday objects to become resonant and, in effect, play music. The Boom Ball vibrates to the beat of the music it is playing, offering a multisensory experience. This appeared to help one participant sing in time to the music, as she felt the regular patterns of vibration on her finger tips.

13.3 'Leap Motion Controller'

The Leap Motion Controller¹⁹ is a small USB device that is designed to be placed on a flat surface, facing upward. Using infrared LEDs and tiny cameras, the device can pick up hand movements that can be used to play digital musical instruments. The device was trialled with one of the participants in the study, but with only limited success as it required sight to calibrate hand movements to specific functions or sounds in the program.

¹⁹ www.leapmotion.com

13.4 'Beamz'

The Beamz²⁰ unit is a W-shaped table-top, laser-based music device, operated by using the hands to interrupt the beams. Connected via USB or Bluetooth to a computer, breaking the beam triggers instrumental (or other) sounds that automatically fit harmonically with other notes being played at that time. Beamz was tested by five individuals in the project who had a good awareness of cause and effect, but who had limited gross motor skills. Some children and young people with higher cognitive abilities enjoyed navigating their way around the console using their fingertips to determine where the lasers were situated and how sounds could be manipulated by movement. They also enjoyed recording their pieces of music and hearing these played back to them. Beamz was not as effective, though, with younger children in the project, who relish the tactile nature of instruments that they can pick up and hold. It was not a realistic option either for those who had limited voluntary movement.

13.5 'Soundbeam'

Soundbeam²¹ is an interactive MIDI system in which movement that breaks an ultrasonic beam at different points along its length is used to control multimedia hardware and software. Many schools and centres have a Soundbeam on site. The device can be time-consuming to set up, but among its advantages are the capacity to connect almost any movement (however small) to different musical outcomes and effects. This permits progressive differentiation, whereby the parameters of the beam can be adapted to meet the changing needs of users as their ability to move is increasingly constrained by the progression of their disease. So a pattern of notes that was once produced by a wave of the arm can later be made merely by raising a finger, for example. And the facility for selecting different sets of pitches as the musical outputs from the device enhances the potential for interaction with others – in the form of imitation, call and response, or even simple turn taking. Work is ongoing to assess the value of Soundbeam with children and young people in the later stages of Batten disease, although a similar system ('Optimusic') proved successful in enabling a young woman with the juvenile form of Tay-Sachs disease (which has similar symptoms to Batten disease) to participate in a sophisticated way in group music-making (imitating short motifs both rhythmically and melodically) when she no longer had the capacity to vocalise or play conventional instruments (Ockelford, 2013).

13.6 Music programs on the iPad

Programs on the iPad such as 'EchoString', 'Drum beats' and 'SoundPrism' were used successfully by participants who had some range of movement in their hands or arms, but for whom conventional instruments were too difficult to play. One parent commented on her son's enjoyment of 'Drum beats':

It has all the instruments that he likes. He loves the ocean drum: and when you tip the iPad, it makes the same sound. This is brilliant because he can't hold the real ocean drum as well any more, but the iPad he can still grip and tilt.

Similarly, a participant with profound physical disabilities was able to play music with her music therapist by making tiny but voluntary movements with her fingers on the 'EchoString'. By placing the iPad on an adapted stand and positioning the instrument conveniently for her, the two could improvise simultaneously. This was the first time that technology had been used to enable her to engage in playing along to the music that she still enjoyed hearing. These interactive programs allow children and young people to make sounds easily and help develop an awareness of how they can use small movements in their hands to make interactive sounds. However, they are too limited for those children and young people who have the cognitive ability to engage in more complex music activities such as song-writing or music improvisation. In such cases, programs such as 'GarageBand' and 'Launchpad' were used in sessions and beyond, to encourage young people to execute decision making and planning, and, above all, to give them enjoyment in creating their own music.

²⁰ <https://thebeamz.com/>

²¹ <https://www.soundbeam.co.uk/>

- 13.7 One of the participants, Luke, was highly motivated by music technology. The research team were able to observe him in classroom activities, music lessons and therapy sessions. He currently spends at least an hour every week recording his original music with his support teacher who visits him at home, where sessions cover complex tasks related to recording, mixing, looping, sampling and composing, all of which he accomplishes with no sight. Luke shows us how a passion for music technology combined with appropriate intervention and support can enable him to make and share music albums. The one challenge, according to his parents, is ‘finding the sweet spot’ in the level of complexity and accessibility in the technology he uses: ‘Beamz is too basic, and GarageBand/Pro Tools are too much for him.’ He has stated a preference for physical equipment and instruments (in addition to playing the piano, clarinet and saxophone, he is quite adept at using an eight-track mixer) but requires support with setting it up. As his father observes: ‘Once it’s ready, he’s very confident with all the knobs and dials.’
- 13.8 To sum up, it seems that both generic and specialist music technology have something to offer those with Batten disease, depending on their musical interests and levels of ability. All of the participants the research team worked with needed help in setting equipment up, and some required ongoing assistance to use it. Beyond the equipment that was tried, there appears to be a gap in the market for a device that would function rather like a small Soundbeam, with adjustable settings of beam length and resolution, but requiring only minimal set up in conjunction with an iPad or a similar device. It would potentially function to allow sophisticated control of musical parameters such as pitch and rhythm, as well as providing a more basic sense of cause and effect.

14 CONCLUSION

- 14.1 The MIND research project and the Erasmus+ study provide persuasive evidence that music can play a number of unique roles in the lives and education of those with NCL at different stages in the progression of the disease.
- 14.2 First, as listeners, music can provide the same sense of enjoyment and well-being that it offers all children and young people, wherever they exist on the continuum of neurodiversity. In fact, listening to music becomes more important as a leisure pursuit as the disease advances and other recreational options become more limited. An understanding of music appears to diminish more slowly than cognitive activity in other domains and, even in the final stages of neurodegeneration, the facility and propensity to engage with abstract patterns of organised sound seem, at some level, to be spared.
- 14.3 Second, participation in music-making is unconstrained through visual loss, though the decline in motor skills eventually precludes performance on conventional instruments. The use of gesture-based MIDI technology can extend the period in which active engagement is possible, however. And, as is the case with adult dementias, the ability to sing persists for some time after the capacity to speak has been lost.
- 14.4 Third, music is, above all, a social activity that can prevent the world of contact with others, including family and friends, from shrinking too much: sustaining communication when words alone no longer function, offering young people a medium through which to articulate their feelings when other channels of emotional expression are ruptured, and enabling memories that would otherwise be effaced to be accessed and shared.
- 14.5 In the view of the research team, it is vital that the potential place and function of music for those with Batten disease is made known to families, therapists, teachers and clinicians; that appropriate strategies are disseminated and resources made freely available that will enable music to be integral to children’s education, recreation and care from the time that a diagnosis is made. The four education strategies identified in the European JNCL project (von Tetzchner, et al., 2019) provide a useful framework for considering music interventions:
- **Hastened learning** (the learning of complex skills while it is still possible).
Give the children the opportunity to try playing whatever musical instruments attract their interest – teachers and parents should be flexible as a child’s motivations evolve. Developing the coordination required to play an instrument will have a wider neurological value.
 - **The precautionary approach** (proactive learning of skills that are not of importance today but will be tomorrow).
Focus on learning a wide repertoire of songs by ear; they will be remembered later in a child’s life, and the broader the initial base of knowledge, the more that will be retained. They are a legacy that can increasingly be drawn on as other areas of engagement decline.

- **Life fluency** (consolidation of life flow, to ensure that life moves in a desirable direction). Here, music can provide a source of stability and continuity as children and young people journey through the different phases of their disease. Ensure that the musical world of the child is there to support them through different settings. Keep playlists and recordings of special musical occasions. Encourage all those involved in children's lives to engage with them in their musical interests.
- **The participation/team/coaching/interdependence model** (to consider a person's possibilities/participation when he/she is supported by a good helper). Find musical friends for children (teachers, therapists, community musicians), who will share their musical experiences and scaffold their participation, whether as listeners (at concerts, as well as recorded music), performers (potentially in a range of social settings) or creators of new material (that can be shared online as well as among immediate networks of family and friends).

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APPENDIX 1

Music-related questions used in the JNCL questionnaire

- 1 Please consider the impact of music on the person's life. Use the categories below and choose the most appropriate alternative.
 - 1 = No impact
 - 2 = Low impact
 - 3 = Moderate impact
 - 4 = High impact
 - 5 = Very high impact
 - 6 = Don't know
 - 7 = Not relevant
- 2 Indicate the age when the person is/was offered music therapy for the first time.
- 3 Please indicate the number of years the person has been receiving/received music therapy.
- 4 Consider the degree to which music therapy has/had an impact on the person. Use the categories below and choose the most appropriate alternative.
 - 1 = No impact
 - 2 = Low impact
 - 3 = Moderate impact
 - 4 = High impact
 - 5 = Very high impact
 - 6 = Don't know
 - 7 = Not relevant
- 5 Please rate the following where
 - 1 = Not at all
 - 2 = Somewhat
 - 3 = To a moderate degree
 - 4 = To a high degree
 - 5 = To a very high degree
 - 6 = Don't know
 - a) The person is/was especially interested in sounds (eg vacuum cleaners etc.)
 - b) Music is/was important for the person as a source of stimulation
 - c) Music is/was important for the person as a source of comfort
 - d) Music is/was important for the person to facilitate communication
 - e) Music is/was important for the person to facilitate social contact
 - f) Music is/was used to develop comprehension (eg numbers or days)
 - g) Music is/was important for the person to highlight events and daily routines
 - h) The person has/had favourite music pieces or favourite music genres
 - i) Music is/was important in certain situations
- 6 Answer this question if the person plays/played any instruments. Please specify the instrument(s), at what age the person started playing the instrument(s), at what age the skills were at their best, and if relevant at what age the person stopped playing the instrument. If relevant, what level did the person reach on her/his principal instrument, compared to peers of the same age playing the same instrument?
- 7 Answer next question if the person used to sing. Estimate the person's approximate age when the singing was at its best. How good was the person's singing performance in comparison with peers of the same age?
- 8 Does/did the person read music in clear print?
- 9 Does/did the person read music in Braille?

- 10 Does/did the person at any time have perfect pitch? If 'yes' or 'don't know', please describe what the person can/could do (eg identify pitch by ear, identify what key a song is played in, keep the pitch while singing etc.).
- 11 Does/did the person make her/his own music? If yes, from what age? If yes, until what age? If yes, please describe.
- 12 Please consider whether learning music/music therapy at school has/had any positive effect on life outside school when the person is/was a student. Use the categories below and choose the most appropriate alternative.
- 1 = No impact
 - 2 = Low impact
 - 3 = Moderate impact
 - 4 = High impact
 - 5 = Very high impact
 - 6 = Don't know
 - 7 = Not relevant
- 13 Choose the most appropriate alternative to indicate whether music/music therapy at school has/had a positive impact on the person's life outside school after he/she has concluded school attendance/formal education.
- 1 = No impact
 - 2 = Low impact
 - 3 = Moderate impact
 - 4 = High impact
 - 5 = Very high impact
 - 6 = Don't know
 - 7 = Not relevant
- 14 Write down the three most important interests and activities for the person outside school/day centre at different ages. Write 'not relevant' if the person has not reached the actual age yet. Around seven years of age, 10 years of age, 13 years of age, 16 years of age and 22 years of age. Make comments if the person developed certain and long-lasting skills as a result of strong interests/favourite activities.

APPENDIX 2

Presentations at conferences

- Ockelford, A. (2015) 'Researching the potential role of music in enhancing the lives of children with neurodegenerative disease ('MIND'), Phase 1: Batten disease', paper presented at a Batten Disease Awareness Day, organised by the Batten Disease Family Association, at Linden Lodge School, London.
- Ockelford, A. and Atkinson, R. (2016) 'Exploring the potential role of music in the lives of children with Batten disease', paper presented at a Batten Disease Awareness Day, organised by the Batten Disease Family Association, at Linden Lodge School, London.
- Ockelford, A. and Atkinson, R. (2016) 'Initial findings of the European JNCL and Education project: music', paper presented at the JNCL and Education Conference, at the Royal Blind School, Edinburgh.
- Atkinson, R. (2016) 'Exploring the role of music to enhance the quality of life in children with Batten disease', paper presented at the Live Music Now and Royal Society of Medicine Conference, Examining the Utility of Music Interventions for Children with Learning Disabilities, Royal Society of Medicine, London.
- Atkinson, R. (2016) 'The potential place of music in the lives of children with Batten disease', paper presented at the BDFa Annual Conference, Staffordshire, UK.
- Atkinson, R. (2017), 'Music and Batten disease', paper and workshop presented at the BDSRA Annual Family Conference, Pittsburgh, US.
- Ockelford, A. and Atkinson, R. (2017) 'Music in the lives of those with JNCL', papers presented at the JNCL and Education Dissemination Days at Linden Lodge School, London, New College Worcester and the Royal Blind School, Edinburgh.
- Atkinson, R. (2017) 'Music in the lives of children with Batten disease', paper presented at the TECHNE Doctoral Researcher Conference, London.
- Atkinson, R. and Herman, K. (2018) 'Neurodegenerative conditions: exploring the role of music to enhance speech and cognitive functioning in children with Batten disease', paper presented at the Mary Kitzinger Trust Conference, University College, London.
- Herman, K. (2018) 'Update on the findings of the MIND project', paper presented at the BDRSA Family Conference, Nashville, US.
- Herman, K. (2018) 'The MIND music project: findings to date', paper presented at the NCL Conference 2018, Royal Holloway University, London.
- Ockelford, A., Atkinson, R. and Herman, K. (2018) 'Neurodegenerative conditions: exploring the role of music to enhance speech and cognitive functioning in children with Batten disease', paper presented the British Association for Music Therapy Conference, Music, Diversity and Wholeness, Barbican, London.
- Atkinson, R. (2019) 'Findings of the European JNCL and education project: music', discussion panel and workshop at the BDSRA Annual Family Conference, Denver, Colorado.

APPENDIX 3

Publications arising from the research

- Atkinson, R. (2017) 'Can music help improve memory, language and wellbeing for children with Batten disease?', Insight Online, London: Royal National Institute of Blind People, www.rnib.org.uk/insight-online
- Atkinson, R. (2017) 'Neurodegenerative conditions: exploring the role of music to enhance speech and cognitive functioning in children with Batten disease', BDFA Newsletter, Autumn 2017, London: Batten Disease Family Association, www.bdfa-uk.org.uk/bdfa-newsletter-autumn-2017/
- Elmerskog, B., Tøssebro, A.-G., Atkinson, R., Rokne, S., Cole, B., Ockelford, A. and Adams, H. (in press) 'Overview of advances in educational and social supports for young persons with NCL', *Biochimica et Biophysica Acta (BBA) – Molecular Basis of Disease*.
- Ockelford, A. and Atkinson, R. (2019) 'Music education and music in education', (in) von Tetzchner, S., Elmerskog, B., Tøssebro, A.-G. and Rokne, S. (eds), *Juvenile Neuronal Ceroid Lipofuscinosis, Childhood Dementia and Education*, Melhu, Norway: Snøfugl Forlag, pp. 343–358.

APPENDIX 4

Relevant organisations

The Amber Trust
64a Princes Way
London SW19 6JF
www.ambertrust.org

Batten Disease Family Association
209 City Road
London EC1V 1JT
www.bdfa-uk.org.uk

Batten Disease Support and Research Association
2780 Airport Drive, Suite 342
Columbus, OH 43219
www.bdsra.org

British Association for Music Therapy
24–27 White Lion Street
London N1 9PD
www.bamt.org

Chiltern Music Therapy
Office A, Irfon House
High Street
Chesham
Buckinghamshire HP5 1DE
www.chilternmusictherapy.co.uk

Contact (For Families with Disabled Children)
209 City Road
London EC1V 1JN
www.contact.org.uk

The Evelina London Children's Hospital
Westminster Bridge Road
London SE1 7EH
www.evelinalondon.nhs.uk/Home.aspx

Great Ormond Street Hospital
Great Ormond Street
London WC1N 3JH
www.gosh.nhs.uk

Jessie's Fund
15 Priory Street
York YO1 6ET
www.jessiesfund.org.uk

Linden Lodge School
61 Princes Way
Wimbledon Park
London SW19 6JB
www.lindenlodge.wandsworth.sch.uk

New College Worcester
Whittington Road
Worcester WR5 2JX
www.ncw.co.uk

The Royal Blind School
Gillespie Crescent
Edinburgh EH10 4HZ
www.royalblindschool.org.uk

Royal National Institute of Blind People
105 Judd Street
London WC1H 9NE
www.rnib.org.uk

Sebastian's Action Trust
The Woodlands
Upper Broadmoor Road
Crowthorne RG45 7FN
www.sebastiansactiontrust.org

SeeAbility Heather House
Tadley
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RG26 4QR
www.seeability.org

The WESC Foundation
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