

## 'Researching the potential role of music in enhancing the lives of children with neurodegeneration ('MIND'), Phase 1: Batten disease'

### Interim Report to the Baily Thomas Charitable Fund

June 2018

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#### 1. Overview

- 1.1 The second year of the three-year project is now complete, and this report summarises progress and key findings to date. The project aims to ascertain the extent to which music can have a positive impact on children and young people in varying stages and different types of Batten Disease, with a specific focus on how music interventions can help with:
  - (a) the maintenance of expressive communication 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 memories;
  - (d) social inclusion through musical participation, and importantly
  - (e) a sense of emotional well-being.
- 1.2 Twelve participants were recruited in Year 1, and they have received 11 visits from the Research Officers (RO), Rebecca Atkinson and Katya Herman. (Rebecca went on maternity leave in February 2018, for a period of 12 months. Katya is an experienced music therapist and researcher who will cover the fieldwork work.) The remaining visits of this calendar year will take place in the Autumn, 2018. Information regarding participants' musical and wider development and functioning continues to be collected.
- 1.3 Six music therapists, a musician and two music teachers have this year delivered weekly sessions with the children and young people at school ( $n=9$ ), at home ( $n=2$ ) and in a residential setting ( $n=1$ ). The music practitioners are videoing their sessions at additional points in the project and submitting these using EthOS (the online video App) to the RO for analysis. This is 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 the RO is not present.
- 1.4 The new resources that were devised in Year 1 of the project, in particular the 'micro-songs', continue to be used, and additional specialist technologies have been trialled, with the aim of enabling those children and young people with very limited movement to take part actively in music-making. The relationships with the Batten

Disease Family Association (BDFA) ([www.bdfa.com](http://www.bdfa.com)) and The Amber Trust (a charity that, among other things, funds music therapy for children and young people with Batten Disease, [www.ambertrust.org](http://www.ambertrust.org)) have been sustained, and we continue to liaise closely with the Batten Disease Support and Research Association (BDSRA) in the US.

1.5 Four meetings of the steering group of parents and professionals have now been held. The research team has hosted two training sessions for the music therapy professionals to improve their understanding of the impact of Batten’s, and to introduce them to new resources as these are developed. The Erasmus-funded European project investigating Juvenile Neuronal Ceroid Lipofuscinosis (JNCL) and education, which involves seven countries (England, Scotland, Norway, Denmark, Finland, Germany and the US), has now completed, and the Ockelford and Atkinson have contributed a chapter on music to the book that is being published about Batten Disease and education. The research team has given presentations at a UCL Institute of Child Health conference, the British Association of Music Therapists conference, a BDFA conference in the UK, and a second presentation is scheduled for the BDRSA annual conference in the US.

1.4 Project activities undertaken from June 2016–June 2019 are shown in the following Gantt Chart; those completed are shaded in grey.

Task	2016 (terms)		2017 (terms)			2018 (terms)			2019
	summ er	winter	spring	summ er	winter	spring	summ er	winter	spring
Design and distribute questionnaire	█								
Set up advisory group with BDFA; meet	█			█			█		█
Devise materials	█	█							
Purchase technology; set up EthOS app	█								
Sol assessment training for RO	█								
Analyse questionnaire data	█	█							
Identify families	█	█							
Initial family visits; baseline Sol data	█	█							
Train families in EthOS app and music tech	█	█							
Gather developmental data on children	█	█		█		█		█	
Data analysis (including,	█	█	█	█	█	█	█	█	█
Continuing visits to families and schools	█	█	█	█	█	█	█	█	█
Gathering and analysing EthOS data	█	█	█	█	█	█	█	█	█
Final visits; exit strategies implemented	█	█							█
Evaluating materials				█		█		█	
Refining materials				█		█		█	
Making materials available online									█



## 2. Participants

### 2.1 Batten Disease

The Neuronal Ceroid Lipofuscinoses (NCLs) – collectively known as Batten Disease – comprise several different genetic life-limiting neurodegenerative conditions that share similar features. Although the presentation and trajectory of the disease can vary significantly with each form, there are a number of common symptoms, including blindness, epilepsy with complex seizures, dementia, myoclonic jerks of limbs, 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. There is no known cure, so specialist symptom management and therapy are essential to maintain as good a quality of life for the children and their families as possible.

The BDFA estimates that there are between 100–150 affected individuals currently living with Batten Disease in the UK, with 11–17 new diagnoses each year. There are currently 14 known forms of Batten Disease, five of which are represented in this study. Of the 12 participants in this study, five have CLN2, three have CLN3, two have CLN5, one has CLN6 and one has CLN8. (Each variant is classified by the gene that causes the disorder.)

<b>Participant number</b>	<b>Age at onset of study</b>	<b>Male/Female</b>	<b>Genetic variant</b>	<b>Level of functioning</b>	<b>Engagement with music</b>
1	3	Female	CLN2 Late-infantile	Vision still functional in most everyday contexts	Receives bi-weekly music sessions at school. Participates actively in singing and instrument play.
				No issues with motor function.	
				Minor language difficulties.	
				No seizures.	
2	5	Male	CLN2 Late-infantile	Blind.	Receives bi-weekly music sessions at home. Engages with some music technology, otherwise primarily engages receptively.
				Minimal mobility.	
				No expressive language.	
				Seizures occurring frequently.	
3	5	Female	CLN2 Late-infantile	Poor vision.	Receives weekly music therapy sessions at school. Participates actively in instrument play.
				Uses a wheelchair, some mobility preserved.	
				Minimal expressive language.	
				Seizures occurring infrequently	
4	6	Male	CLN2 Late-infantile	Poor vision.	Receives weekly music therapy sessions at school. Participates actively in singing and instrument play.
				Walks with support.	
				Moderate language difficulties.	
				Seizures occurring infrequently.	

5	9	Female	CLN2 Late- infantile	Blind.	Receives family music therapy sessions every week at school. Engages primarily on a receptive level.
				Uses a wheelchair. Minimal independent motor function.	
				No expressive language.	
				Seizures occurring frequently.	
6	12	Male	CLN3 Juvenile	Blind.	Receives weekly music therapy sessions, music lessons at school, private piano instruction, and weekly music support sessions at home. Plays the piano, and is working on recording an album. Highly motivated and engaged by music.
				Normal motor function.	
				Minor expressive language difficulties.	
				No seizures.	
7	18	Female	CLN3 Juvenile	Blind.	Receives weekly music therapy sessions at her residential home. Participates actively in singing and instrument play. Highly motivated by and engaged in music.
				Uses a wheelchair, some mobility preserved.	
				Significant language difficulties.	
				Seizures occurring frequently.	
8	15	Female	CLN3 Juvenile	Blind.	Receives weekly music therapy sessions at school. Participates actively in singing and instrument play. Highly motivated by and engaged in music.
				Uses a wheelchair, some mobility preserved.	
				Significant language difficulties.	
				Seizures occurring frequently.	
9	10	Male	CLN5 Late- infantile variant	Blind.	Receives weekly music therapy sessions at school. Participates actively in instrument play. Readily engaged in music.
				Uses a wheelchair, some mobility preserved.	
				No expressive language.	
				Seizures occurring infrequently.	
10	10	Male	CLN5 Late- infantile variant	Blind.	Receives weekly music therapy sessions at school. Participates actively in instrument play. Highly engaged in music.
				Uses a wheelchair, some mobility preserved.	
				No expressive language.	
				Seizures occurring infrequently.	
11	5	Male	CLN6 Late- infantile	Blind.	Receives weekly music therapy sessions at school. Participates actively in instrument
				Uses a wheelchair, some mobility preserved.	

			variant	Significant language difficulties.	play. Readily engaged in music.
				No seizures.	
12	8	Male	CLN8 Late-infantile variant	Blind.	Receives weekly music therapy sessions at school. Engages with some music technology, otherwise primarily engages receptively.
				Total loss of mobility.	
				No expressive language.	
				Seizures occurring frequently.	

### 3. Progress in relation to the research questions.

- ) *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?*

*Data will be gathered in a number of ways: initially through a survey of parents in the UK (contacted via the BDFA; N ≈ 60), using questions drawn from the Focus on Music studies (Ockelford, et al., 2006; Ockelford and Matawa, 2009), which examined the impact of different eye conditions associated with particular syndromes on developing musicality.*

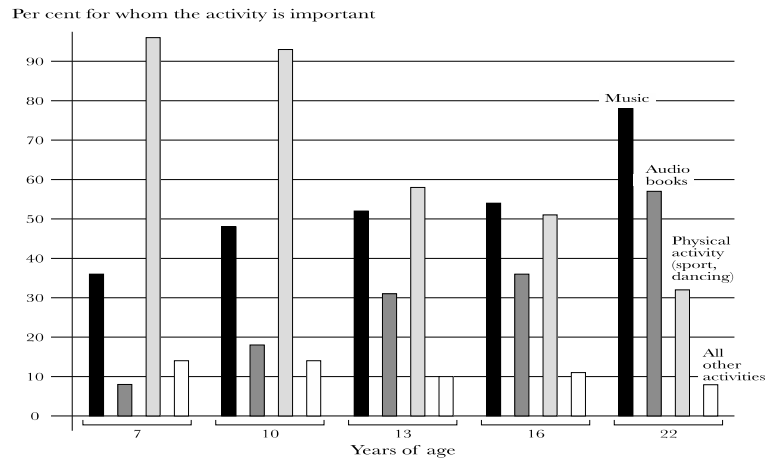
The opportunity arose to be part of a Europe-wide Erasmus-funded study, which gathered questionnaire data from parents and professionals from countries across Europe (including the UK) and the US. The questions from the *Focus on Music* studies questions were incorporated into the questionnaire. The results are to be published in 'Music in Lives of those with JNCL',<sup>1</sup> *Juvenile Neuronal Ceroid Lipofuscinosis (JNCL) and Education: A European Study*, edited by Bengt Elmerskog and contributing authors; publication is expected late in 2018.

The respondents (N=183) came from six countries across Europe and comprised parents (n= 110) and professionals (n=73). Key findings were that:

- Nine out of 10 parents reported that music had (or once had) a high or very high impact on the lives of their children.
- Moreover, the importance of music was perceived to increase markedly as the children grew up, when other activities, such as sport and dancing, were no longer accessible – see Figure 1.

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<sup>1</sup> 'JNCL' is an alternative term for 'CLN3'.



**Figure 1** The importance of music for children with JNCL, in relation to other activities.

- Around a third of children and young people with JNCL were having or had had music therapy at some point in their lives: the median age for starting was 14, and the mean duration of the therapy was eight years.
- Nine out of 10 parents, and eight out of 10 professionals, reported that music therapy had (or once had) a high or very high impact – primarily as a source of stimulation and comfort.
- Around half of the children and young people with JNCL 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. Most children started playing around the age of eight and continued for around five years, never getting beyond the level of beginners.
- It seems that children started to engage more with music 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).
- Children’s singing was said to be at its best (and on a par with fully-sighted peers) around the age of 12.
- Around one in five of children and young people with JNCL were reported to make up (or to have made up) their own music, which occurred largely between the ages of 11 and 15.

From interviews with parents and professionals, it also became apparent how music can help to sustain communication when words alone no longer function; how it can give young people a medium through which to articulate their feelings when other channels of emotional expression are occluded; how it can enable memories to be

accessed that would otherwise be lost; and, above all, how it can help to maintain a sense of wellbeing.

These results suggest that for children and young adults with JNCL, music plays an important part in everyday activities and well-being throughout the progression of their disease. Whilst this sample population only represents one of the thirteen variants of Batten Disease, JNCL has the largest incidence of Batten Disease in Europe and the US, and the RO's initial visits with families affected by Batten Disease, have shown that similar findings pertain to children with other variants of the condition, although the ages at which developments and challenges arise vary. One key difference is language: those with infantile or late infantile forms of the disease may develop little or no speech. Hence, rather than scaffolding and sustaining expressive language, music may necessarily be used as a substitute.

*ii) 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?*

*Data pertaining to this question will be obtained through longitudinal studies of the children, following their baseline assessments. Their levels of musical functioning will be re-assessed every term over three years using the Sounds of Intent scheme, enabling the researchers to ascertain the trajectories of the children's music development as the disease progresses. These will be analysed and compared to data from regular psychological and neurological testing that are undertaken as part of the children's specialist medical care.*

With the exception of the children involved in a Biomarin clinical trial at Great Ormond Street Hospital, we discovered that regular psychological and neurological testing was not being undertaken, and the data from those children in the trial are not available, even to the patients. The one scale available and universally accepted is the Hamburg Scale (devised in 1988 by the Department of Paediatrics at the University of Hamburg): a rating system that assesses disease progression in two major functional areas: motor function and language ability. Scores range from 3 (normal function) to 0 (total loss of function) with unit decrements representing milestone events in the loss of function (e.g. increasing difficulty with abstract reasoning, or losing the ability to walk or crawl). In addition to motor and language function assessments, there are also functional domains (each with a 3-point scale) to assess vision, intellect, and the frequency of grand mal seizures. These are rated as follows:

<b>Problem</b>	<b>Functional state</b>	<b>Coding score</b>
Vision	Normal	3
	Poor, but orientation good	2
	Poor, orientation difficult	1
	Blind	0
Intellect	Normal	3
	Abstract reasoning (mathematics) has become difficult	2
	Dementia clearly evident	1
	Apparent total loss	0



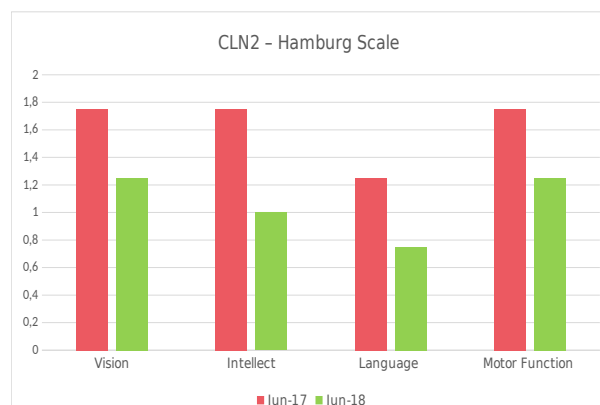
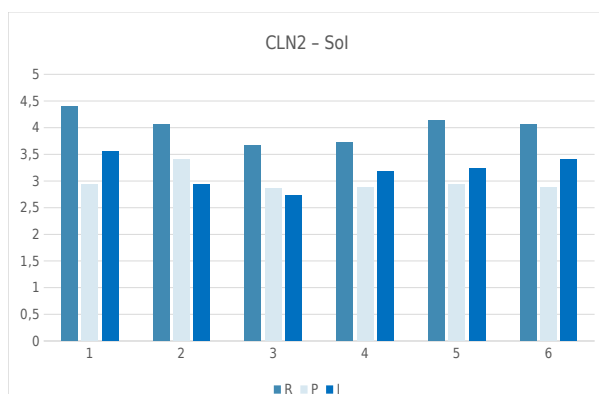
Language	Normal	3
	Minor difficulties recognised	2
	Hardly understood	1
	No verbal contact	0
Motor Function	Normal	3
	Minor difficulties recognised	2
	Mostly wheelchair, some mobility preserved	1
	Immobile, bedridden	0
Epilepsy (only Grand Mal)	No seizures	3
	1-2/year	2
	<1/month, 12/year	1
	>12/year	0

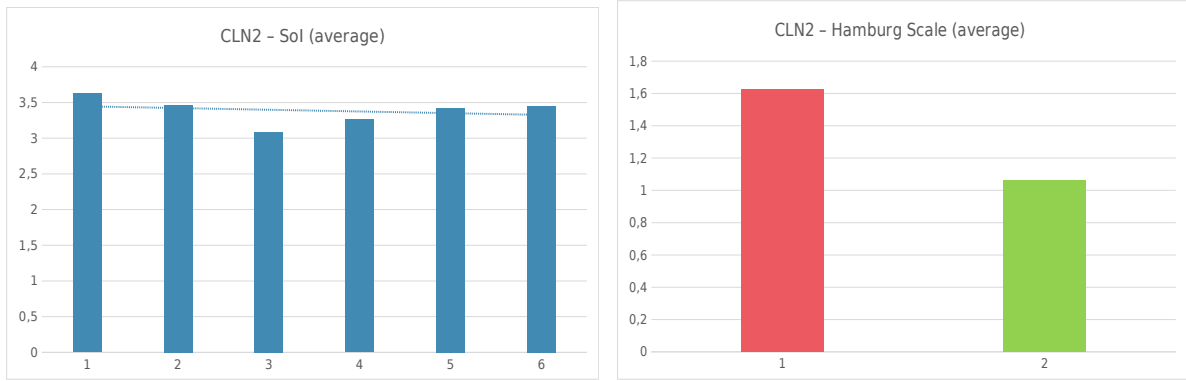
**Figure 2** ‘Single coding scores possible for each of 5 major clinical problems in JNCL patients.’ (from Kohlschütter *et al.*, 1988, p 868.)

Although this measure only offers a general sense of a child or young person’s place on their Batten’s journey, the scale allows quantitative description for each individual course of a disease that varies significantly from case to case. It also has practical value in that, although the scale was designed for use with children with JNCL (CLN3), it can be applied equally to all the variants, and doesn’t require special training to administer. The Hamburg Scale results (seen here as bar charts) are presented with the overall scores averaged by variant. Information regarding the participants’ seizure activity has been omitted, as these are often ameliorated by changing levels and types of medication.

The children’s level of musical engagement is measured using the Sounds of Intent (Sol) framework ([www.soundsofintent.org](http://www.soundsofintent.org)), and the Sol graphs depict the children’s receptive engagement, participation, and interactive engagement, followed by a consolidated score.

Over the 24 months of the study to date, we can see from the data pertaining to the Hamburg Scale data that most of the children have experienced a decline in at least two functional areas, with a 36% function loss averaged across the domains. However, from the Sounds of Intent analyses (‘R’ - ‘reactive’; ‘P’ - ‘proactive’ and ‘I’ - ‘interactive’), we can see that there has been no significant decline in their ability to engage in music.

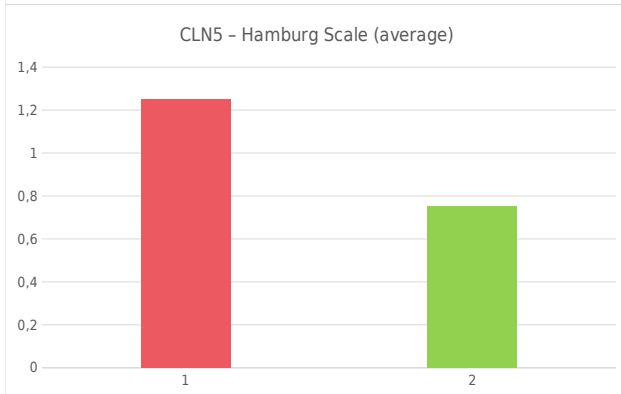
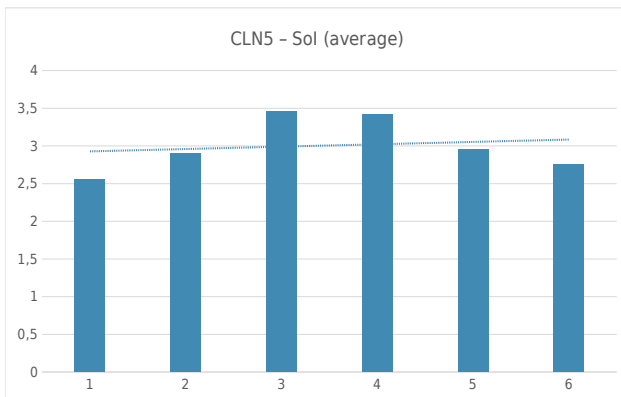
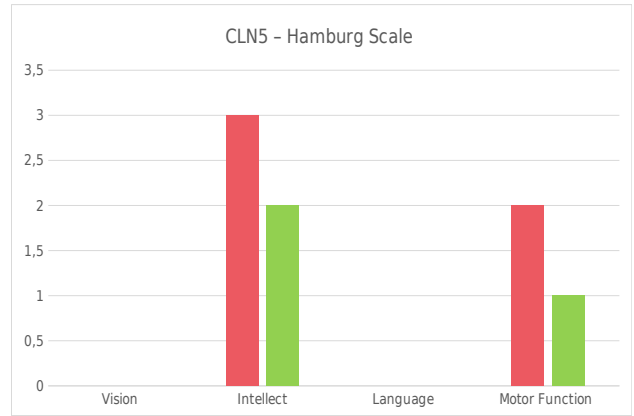
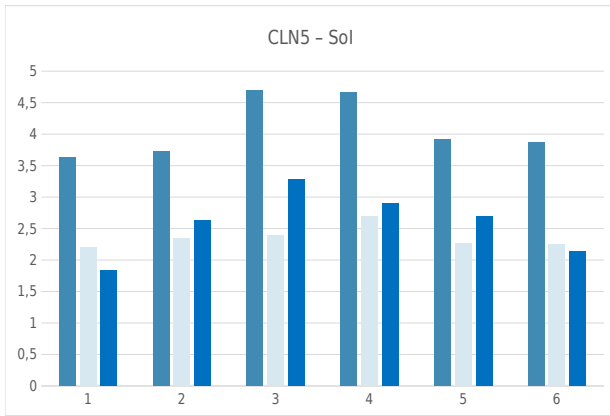




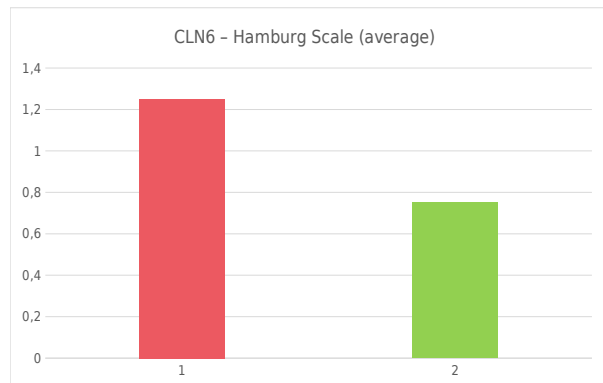
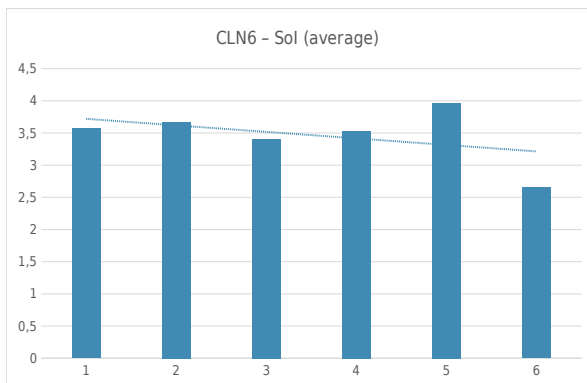
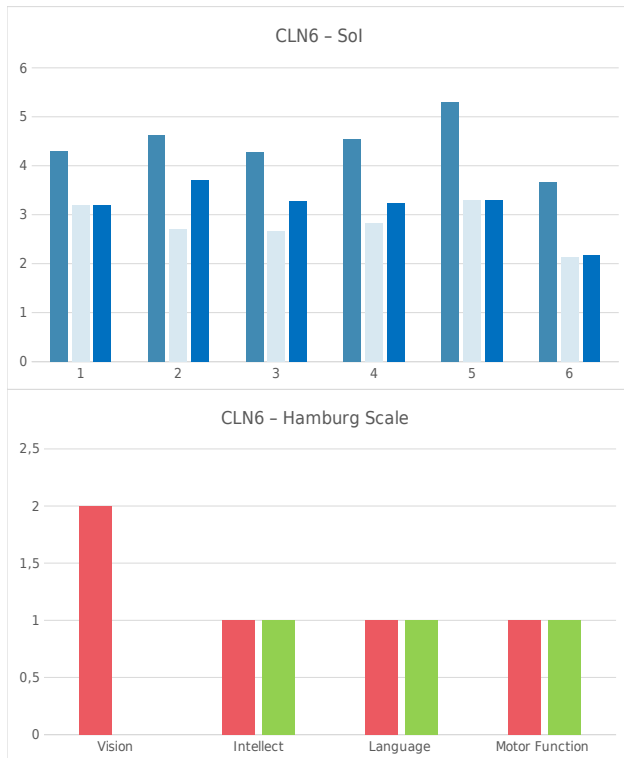
**Figure 3** Sounds of Intent and Hamburg Scale analyses, CLN2.



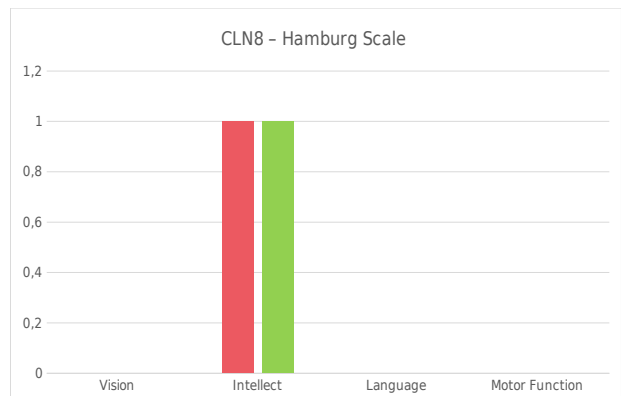
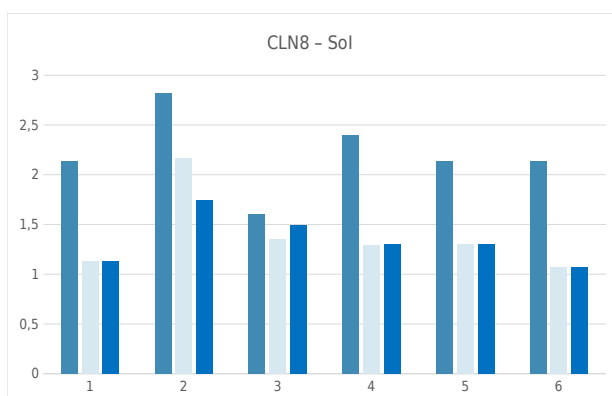
**Figure 4** Sounds of Intent and Hamburg Scale analyses, CLN3.

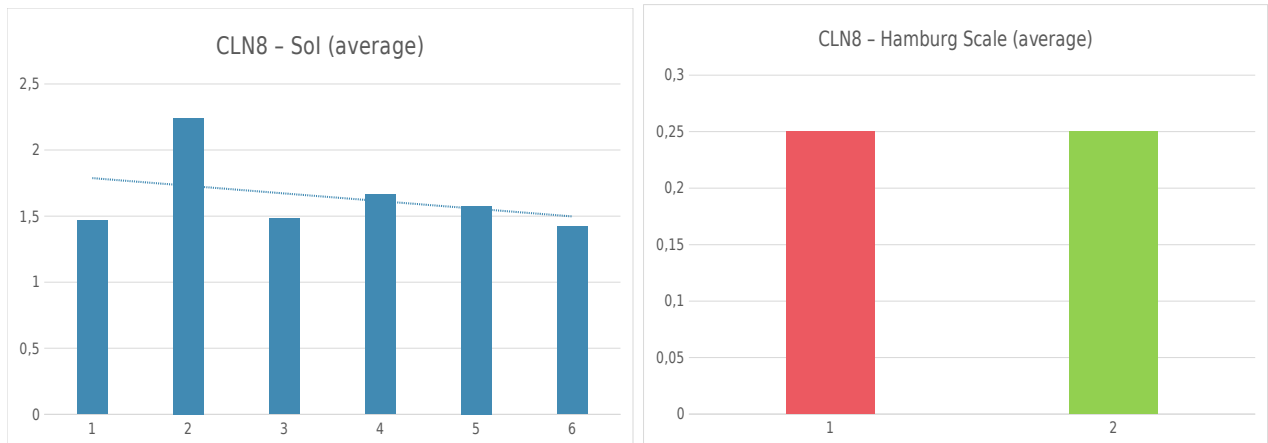


**Figure 5** Sounds of Intent and Hamburg Scale analyses, CLN5.

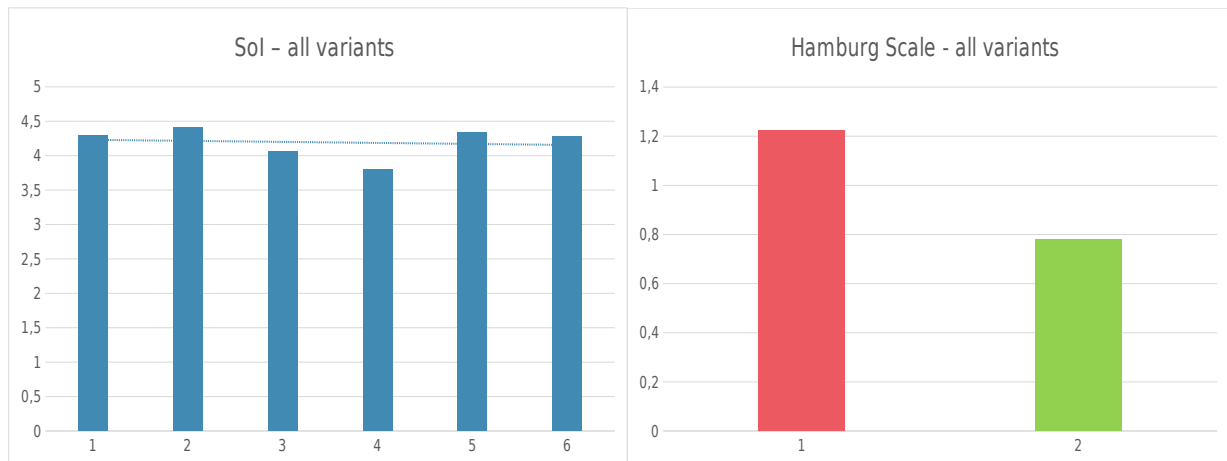


**Figure 6** Sounds of Intent and Hamburg Scale analyses, CLN6.





**Figure 7** Sounds of Intent and Hamburg Scale analyses, CLN8.



**Figure 8** Sounds of Intent and Hamburg Scale analyses, averaged across all variants.

- iii) *To what extent and in what ways can music be used an intervention to promote: (a) the continued use 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 memories; (d) emotional regulation and well-being; and (e) social inclusion through active musical participation?*

A series of musical interventions are being devised by the Principal Investigator (PI) – Adam Ockelford – as a development of previous work in this area. Specific examples can be found in the following areas.

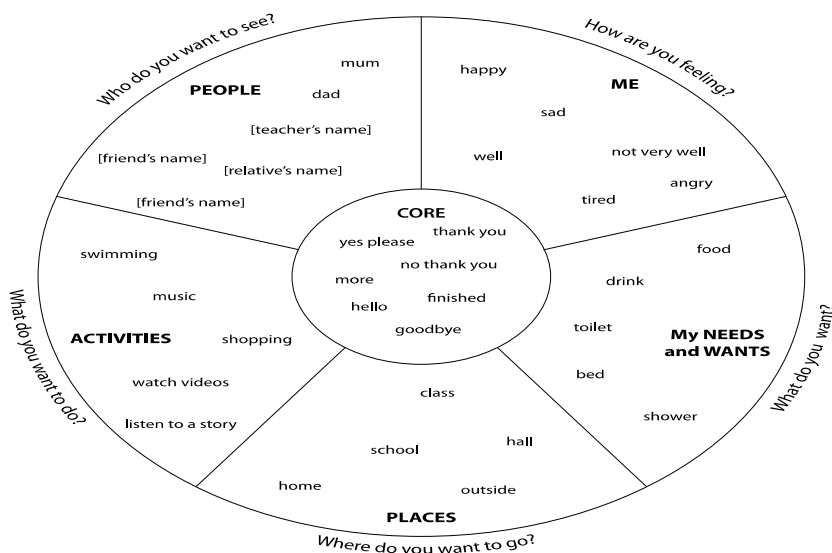
- (a) *Can music be used to promote or replace expressive language?*

*‘Materials will be devised that extend the thinking in the All Join In! project (Ockelford, 1996) a set of songs and activities for visually impaired children with learning difficulties, in which everyday language and simple melodies are coherently and consistently connected, the latter intended to scaffold the former, or even substitute for it, whereby music comes to serve as a proxy language (Ockelford, 2013).’*

(b) Can music be used as a symbol to communicate 'what, where, with whom and when'?

*'Short musical excerpts will be devised for each child, which will have the capacity to function symbolically, using Ockelford's principle of 'sounding Objects of Reference' (Ockelford, 2001). These sound symbols can potentially function expressively or receptively, enabling children without speech (or sight) to communicate preferences and, in the later stages of mental decline, to understand what is happening, where, with whom and when. Sound symbols will be introduced as verbal language disappears.*

The development of the project resources and activities comes in the form of 'micro-songs'. Thirty-nine 'micro-songs' for the children in the study, are organised as follows:



**Figure 9** Map of 'micro-songs'

The scheme is being introduced to two of the young people with CLN3 in whom speech is all but lost, but for whom the ability to sing (with words) is maintained. The initial findings are very promising. Social greetings ('hello' and 'goodbye') are possible using the motifs from the songs. Choices using 'yes' and 'no' can be made. Feelings ('happy', 'sad', 'angry', 'tired') can be expressed. Short songs with 'open ended' answers have also been developed. For example, in response to the question 'What d'you want to eat, [name of child]?', answers such as 'crisps', 'pizza' and 'Weetabix' have been given. 'What d'you want to drink, [name of child]?' has produced responses such as 'squash', 'water' and 'Coke'. 'Who d'you want to see, [name of child]?' has options such as 'mum', 'dad' and other open-ended possibilities for family members and friends. The next step is to seek to embed these songs with

carers and parents beyond the music sessions. This aim will be supported at the end of the current project, when all the materials will be made freely available online (including audio files, videos and written materials) – a project for which funding has been obtained through The Amber Trust.

(c) *Can music be used systematically to support long-term memory?*

*'Music and sound associated with important events in a child's life will be recorded (for example, the sounds of the funfair on a family holiday, a busker encountered on a visit to Covent Garden, a song from the school's end-of-year concert, the music at a cousin's wedding) and subsequently used to evoke memories that can be shared and enjoyed with family and friends, who will additionally be able to recall in words what happened, and help the child re-live moments from the past.'*

In collaboration with colleagues in special schools in England and Scotland, the researchers are devising ways in which music can help facilitate the recall of events. For example, at the Royal Blind School in Edinburgh, 'memory books' are created for children and young people with Batten's, 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, and CDs (or similar materials associated with holidays or outings). In the case of a 16-year-old with CLN3, who struggles to find the words for day-to-day objects and activities, she can still remember the lyrics of entire songs. She is now working on constructing her own tactile books, which she revisits as a way of recalling favourite memories. From the start, she has played an active part in the process: first she will rehearse a song that she has chosen herself. This song will then be recorded and put onto a CD. She then selects the tactile materials that she deems relevant to each song (the 'Objects of Reference'), and attaches them to the page. For a Paloma Faith song, she chose a red flower as her Object of Reference, because Paloma Faith always wears flowers in her hair; for Sam Smith, a piece of leather, as he always wears leather trousers. Lastly, she writes out the song lyrics in uncontracted Braille, with support. This is a fully interdisciplinary approach involving music, English, Braille, and art, and 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.' As this young woman's disease progresses, 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.

It is intended that this type of activity will be implemented with participants in the project, focusing not only on music, but also other events such as holidays, outings, or special occasions. By encouraging this activity for individuals in the study who still have the necessary cognitive functioning, it is hoped that the maintenance of long-term memory will be facilitated, and potentially transferred from different schools or residential settings as individuals progress through different phases of education and care. It will also enable new support staff to become familiar with an individual's preferences and important life events, and to grasp their sense of identity.

(d) *Can music be used to support emotional regulation?*

*'One of the most distressing elements of Batten's is the emotional anguish that children suffer, and the available evidence suggests that self-selected music may promote self-regulation (Bills, et al., 1998), as it can in neurotypical contexts (Saarikallio and Erkkilä, 2007), and offer relief at times of psychological crisis. It is*

*anticipated that here most data gathering will be by parents, using the EthOS app, and, again, qualitative analysis will be undertaken by the researchers.'*

From the European survey feedback gathered from parents and staff (see Figure 1) it has been reported that music can help support stimulation, comfort and self-expression. For example, parents have 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 reported 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'.

To date, video evidence gathered from parents and staff in the EthOS app has demonstrated how children are affected by music in respect of relaxation, excitement, motivation for movement and for improving mood. The table below outlines main themes of the videos uploaded by parents

<b>Video content</b>	<b>Number of videos</b>
Listening to music	10
Positive behaviour, e.g., smiling, laughing	11
Singing to music	7
Playing instruments	9
Music initiating movement/ dance	5
Music to reminisce while others look at photos	5
Music lessons	4
Eating	2
Music and special occasions	1
Music concerts	1
Listening to music playing in the park	1
Music while hair is cut	1
Music during car journeys	1
Music technology	1

**Figure 10**

Categorisation of video content

Of the 12 families in the project, nine report that they regularly use music at home to support their child's emotional wellbeing. For example, when it comes 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 correlation between their children's eyesight deteriorating and their increased appreciation of music. In these cases, music has 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'; and '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.'

Notably, of the three young people with CLN3, their parents all commented on their child'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.'

Another theme that emerged was the value of using music at school to support their child's 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.'

These qualitative findings align with those from the Erasmus survey, suggesting that music has a significant role in supporting the emotional regulation and wellbeing of children and young people with Batten Disease.

*(e) Can enabling active musical participation through technology promote social inclusion in the later stages of the disease?*

*'Each family will be given two interfaces, which the Research Officer will set up for them, with whatever computer technology is available at home (or at school) to enable the children to make music either through gestures (using the 'Leap' Motion Controller) or physical contact (using the 'Makey Makey' system, which enables any everyday item to be used as a switch). The RO will devise a set of simple musical activities in which family or friends can participate with the child concerned. Data gathering and analysis will follow the protocol set out above - with a particular focus on musical interaction, and the capacity of the child with Batten Disease (i) to*

*imitate the musical gestures of others and (ii) to lead the group [which may be a family group] musically.'*

There have been six computer programmes and music technology products trialled by children and young people in the project so far, with the aim being to identify devices that parents, relatives or school staff can utilise to engage in music activities with their children and young people.

#### LEAP Motion Controller

This device was trialled with one of the participants in the study, but was discarded as it was deemed inappropriate for the individuals in the study, since in setting the device up with a computer, it required sight to calibrate hand movements to specific functions or sounds in the programme.

#### iPad Music Technology

Programmes on the iPad such as 'Echostring', 'Drumbeats' and 'Sound Prism' have shown to be successful for the participants in the project who have some range of movement in their hands or arms, and for whom conventional instruments are too difficult to play. One parent commented on her son's enjoyment of 'Drumbeats': '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 really hold the real ocean drum as well anymore, but the iPad he can still grip and tilt.' These interactive programmes 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 the case where children and young people are able to engage in these more complex tasks, programmes such as 'Garageband' and 'Launchpad' are used regularly in sessions and at home, to encourage children to execute decision making, planning, and enjoyment in making their own music.

One young man is particularly engaged by music technology. He spends an hour every week recording his original music with his music support teacher, who visits him at home. He's hard at work on his second album, but 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 Garage Band/Pro Tools are too much for him.' He has stated a preference for physical equipment and instruments (in addition to playing the piano 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. ... A mixer that will talk to him - that's what we need!'

#### Soundbeam

Most schools and centres have a Soundbeam on site, which in most cases is largely unused. The device is relatively complex to use and can be time-consuming to set up. Work is ongoing to assess the value of the device with children and young people in the later stages of Batten Disease.

#### BEAMZ

The BEAMZ unit, an interactive laser-beam console, has been trialled with individuals in the project who have a good awareness of the cause and effect of movement, but who have limited gross motor skill in the upper or lower body. It was anticipated that, as a visually mediated device, this might be unsuccessful with visually impaired people. However, children and young people with a higher cognitive ability have enjoyed navigating their way around the console using their fingertips to determine where the lasers are situated and how sounds can be manipulated by movement. They have also enjoyed recording their pieces of music and hearing these played back to them. BEAMZ has not been as effective, though, in cases where younger children in the project enjoy the tactile nature of instruments they can pick up and hold. It has also not been a realistic option for those individuals who have extremely limited voluntary movement. The music on the device is pre-programmed and therefore specific music-movement calibration is not possible. Where appropriate, the children in the study have been purchased BEAMZ equipment that has been moderately successful in trial.

Further exploration of music technology products is continuing, to try to identify or create a device that is more sensitive to a wider range of movements.

#### 4. Conclusion

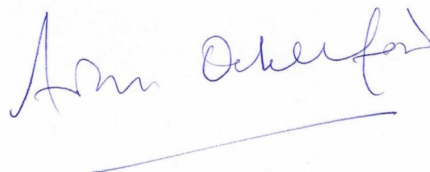
Two years into the research project, key findings are emerging:

- It appears that the capacity to engage with music remains intact while other abilities decline; indeed, music is reported to become relatively more important in the lives of children and young people as their capacity to participate in other activities wanes.
- However, increasing motor impairment presents an ever greater barrier to making music actively, and while movement-sensitive technology can ameliorate this difficulty to an extent, none of the equipment that is currently commercially available offers an ideal solution.
- For young people who are still able to 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.
- Music appears to have a unique capacity to stimulate more general memories.
- Music is valuable to help children and young people regulate their emotions, to produce a sense of wellbeing.
- Music offers a social activity in which other members of the family (or class, at school) can engage together.

The Amber Trust has secured funding to create a free online resource for all families of children and young people with Batten Disease (the Trust currently supports over 30 families across the UK, providing bursaries for music interventions, including therapy and instrumental tuition). The intention for the new resource is that it should build on our findings from this project, and support this small but extremely needy group of young people by offering specific, easy-to-use strategies to therapists, teachers and families to sustain communication when words alone no longer function, to give children 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 wellbeing in an increasingly confusing and frightening inner world.

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5<sup>th</sup> July 2018