Batten Disease and Childhood Dementia

BATTEN DISEASE FAMILY ASSOCIATION
Together we WILL make a difference
Registered Charity in England and Wales 1205650

This guide has been informed and shaped by parents of children with Batten disease during interactive sessions. Our hope is that other parents who may be navigating changes with their children will be able to make use of this information and understand that they are not alone. There are times when the disease can 'take over', we hope that the information helps parents, siblings and, indirectly, children and young adults with Batten disease to feel less isolated and more in control.

Childhood dementia results from progressive brain damage and is caused by over 100 rare genetic disorders, including the group of 13 types of CLN Batten disease. Each child's experience with dementia is unique.

In some instances, dementia symptoms can appear in the youngest of children and babies and progress quickly. While for other children, symptoms may not start until they are teenagers or young adults. Childhood dementia is progressive.
**For ease of reading throughout the document we refer to children. The information is relevant to children and young adults who have Batten disease.*

Symptoms of Childhood Dementia in Batten Disease

- Changes to speech pattern, range of language and word retrieval
- Communication difficulties
- Memory loss
- Fatigue
- Trouble concentrating and understanding
- Difficulty learning
- Personality changes
- Severely disturbed sleep
- Behavioural issues such as hyperactivity
- Emotional issues, anxiety, fear, anger
- Difficulty coping with change
- A need for routine

We asked parents 3 questions

1. **How do you see your child's dementia?**

Speech and word finding

Parents identified that changes in speech and word finding are often what they see first.
They told us their children find it hard to find the right word or words, they will become repetitive because of the word retrieval difficulties.
They can have problems with the fluidity of speech, so intent on getting their news across that they can blur or slur words, they need help to slow down and regulate their speech.
They can get stuck on a word, phrase or question, repeating over and over again, they don't seem to be able to move on or 'switch off' once they are in this pattern.
If they can't get the right word out, they may give up. This can, understandably, lead to frustration.
Over time the ability to speak in sentences diminishes, it becomes more common for the vocabulary range to shorten.
Parents see the way their children's communication changes.
Often children have such a desire to communicate they will speak non-stop. They often find it hard to have a 2-way conversation and focus on their story their way, Parents see stuttering develop to varying degrees.

Children with Batten disease can also flit between one topic and the next without staying on track with one so the whole narrative can be told and explored. They find it hard to keep a grasp of the topic beyond a few words. Saying Um or err repeatedly when they have lost their words. They develop a need for the person they are speaking to, to repeat what has been said rather than saying yes or no, they feel reassurance in being 'heard'.

Children find it harder to communicate in noisy environments or even when there is one other source of noise such as the TV. Lots of active communicating can cause tiredness and this is when the quality of speech and recall is most affected.

Obsessions or fixations

Parents told us about children's obsessions, children can become quite fixated on a past event or a slight/ disagreement with another child that happened in the past, they can use these memories to 'control the narrative' especially if nervous about going somewhere new.

There is a level of obsessiveness for all children, whether a tv programme, range of toys or games, cars, ponies, pets, people, you name it. For some the obsessions can be overwhelming and seem to be the safety net for children, they can keep the familiar and feel grounded and safe with it, they also feel validated for having 'their' knowledge heard.

Boredom

Parent told us that boredom is common, but when children say they are bored they do not always want to know what else they might be able to do, so everything is boring. Often, they become easily distracted, which can increase the boredom. These may be coping mechanisms as the child goes through stages of sight-loss*

Parents told us that progressively they see that children can't hold on to short term memories, and the ability to stay 'in the moment' becomes harder, the child struggles to remember something that happened earlier the same day especially if they are no longer in the place that something happened, so they get home from school and can't remember what they have done. For children who have good functional vision, parents say that they get to a point where they can't remember a face without some prompting.

Anxiety

Anxiety in new environments can come along suddenly, even in familiar schools as children move class each year, may have different Teaching Assistants and their peers may move to different classes. The increasing communication difficulties can lead to isolation as there will be difficulties engaging with peers, especially in mainstream school.
Anxiety can shift to becoming an angry outburst bought on by the flight or fight response when a child feels out of control with behavioural and emotional changes at the same time, along with the repetition and forgetfulness due to short term memory issues, children appear frustrated with changes and unsure how to cope. Some have described this as outbursts over nothing, or like being like watching a train whizz by on a station, or watching a fly in a jar, constant activity in all directions which are disorientating for everyone.
Children can become tired or fatigued as 'thinking and doing' require more energy as childhood dementia progresses.

For some children in main stream school, there may come a point where the complexity of school life, size of campus, number of other students and noise levels become overwhelming even with 1:1 Teaching Assistants and other structured support.

Memory

Parents all said their children's long term memories were strongest, for children who lose their vision these tend to be memories formed before all vision is lost. In CLN3 Batten disease parents say the memories from pre 10 -12 years are strongest.
Children prefer to use reminiscence, often finding it easier talking about memories as they may feel more in control and 'ordinary' with this, less exposed. There is a cognitive decline, depending on long term memories, watching old programmes they enjoyed earlier in their childhood. Along with this, parents told us that they see a regression in their child's interests.
As children move into mid teens, they can have lapses of where they are, who is with them and can quickly be disorientated to space and time so simple functions they can do when rested become almost impossible.

Sleep Disturbances

Parents told us of the progression of sleep disturbance from a young age and the inability to switch off at night. This is a huge topic that is covered in a further document.
Sight-loss in Batten disease, we have seen how changes to a child's functional vision can cause periods of instability and uncertainty as the child adapts to less visual perception, sometimes this is imperceptible to others but where sight-loss is progressive it needs to be considered as childhood dementia can appear to rapidly develop, then settle due to the fluctuating progressive nature of Batten disease.

2. How does it affect you and your family?

Parents told us: -

- We struggle with distraction at times as he always wants me (Mum).
- The boys like what they like and even though this is great in one way it makes it quite isolating for their sibling.
- Siblings feel sidelined.
- I don't think a lot of my family really understand what we are going through but also what he is going through.
- As a couple we work really hard to get our time as the boys are so full on and this could easily lead to us just manning a boy each and forgetting each other.
- Frustrating to repeat things and trying to stay calm
- Difficult to treat siblings the same - and explain that his brother can't say sorry or didn't mean it.
- They (affected children) have both stayed as young boys emotionally and intellectually and therefore their brother has now outgrown them both in that way, so he finds it hard that he doesn't have an older brother to chat with or share things with on an age appropriate level.
- Being 2 boys out of 3, it can be quite exhausting when they are both on full speed talking non stop especially for their middle sibling.
- Very hard for all of us to hear the same thing over and over again, for days and weeks, we try not to get frustrated.
- His brother finds it embarrassing
- We argue lots. Days out become a dread as we don't know what to expect. People like to stare as if you're a bad parent!
- Relationship changes. I feel alone at times.
- It's like torture sometimes, especially at 4am when you've had no sleep!
- Siblings are reluctant to help, especially the younger ones. They can be scared of how she (affected child) will react.
- All of us are very strained, tired and exhausted with the constant battles.
- It makes me feel stressed about leaving the house with her, especially if it's just me and the kids.
- The lack of sleep is a very difficult, makes me tired and grumpy.

3. What have you tried/what do you do that helps?

Parents told us: -

- A dog. He is our 4th boy and gives our affected children responsibility and therapy when they are stressed or upset and an outlet for their sibling to get out of the pressure cooker sometimes and take him for a walk
- MUSIC - incredibly important, every day
- Water. Swimming, baths, foot spas, hydro, water play, it's all so calming and the boys love it
- How did I forget Music!! 'Music isn't what I do its who I am' - a phrase our son used years ago and is up above his bed - music is on 24/7 in our home, car, garden it's a lifesaver
- One son listens to acoustic pop to go to sleep to, the other listens to audio books
- Support from school
- High expectations and keeping the brain busy has helped with everything from sleep to behaviour
- Letting her feel heard and not rushed
- A dog walk - getting outside
- A safe place/safe familiar thing to do - like a Disney film
- Distraction helps
- Just answer their questions calmly
- Singing
- Humour
- Reaching out to our extended community and friends and family - they want to help and be part of everything just sometimes need to be told how and be told when you need them.
- Holistic approaches are our saviour!!
- Being outside, distraction, water (bath time)
- They can't be rushed
- Respite care away from home – gives them a different focus
- Reflexology
- Reiki therapy
- Aromatherapy diffusers at bedtime
- Stephen Fry on Audible (other authors are available!)
- Rebound Therapy
- Horse riding
- Swimming
- Keeping adventurous as a family
- What helps- letting her calm down in her own time. Leaving the room. Getting in bed with her at night.
- Sleep- for me!
- Music as a distraction when she constantly talks about the same thing and trying to change the subject, ask Alexa to play particular songs, go for a walk, meet people.
- Support from school.

- My daughter’s fixation is often on certain people, so we have found her seeing them and having a chat helps. She still talks about them but not so repetitive
- High expectations and keeping the brain busy has helped with everything from sleep to behaviour.
- Getting others outside the family to attend youth clubs and other groups with her.
- Getting her "grounded", a weighted blanket
- We have found out that he can’t regulate his body temperature, it’s called thermo-regulatory dysfunction (common in CLN3) so weight blankets didn’t help him.

What else can help?

Understanding how a Batten disease child’s brain works differently can help families to adapt to childhood dementia.

Children have to cope with many demands that others may take for granted, those that have vision loss have to re-learn how to compensate as their vision deteriorates, they want to use their visual memory but when faced with new situations or tasks they have to start to use a completely different ‘road map’ rather than learned and embedded memories.

Those that have functional vision are also coping with losses in memory function and cognitive skills.

Think of the brain like a high street, at different times the high street can be more or less busy, when it is more busy with lots of people, traffic, rubbish and noise, it can be more tiring to negotiate. In Batten disease, it struggles to filter what is needed and necessary, against what it needs to shut out, it tries to focus on everything and gets tired, frustrated, scared, anxious, angry and can’t communicate well.

When you see this happening, as early as possible imagine being able to give that brain some space, fresh air, peace, gentle sounds, a soothing place to calm, less people, less ‘noise’, then it can re energise ready to move forward again.

In many ways we all do this for ourselves, mindfulness can be simple if we have the tools to initiate it, for the children we need to provide it. Building frequent breaks into the day is helpful, it could be a story, walking the dog, a hand massage, whatever works for your children. Massage, breathing exercises and simple physical exercises are very helpful, especially if there is some instrumental mood music with a steady rhythm the background, as the parents have found in the list above of what they have tried and what works for them.

How can we influence brain function?

- Continuity of people and spaces, children need a degree of routine for school days and in a more relaxed way at weekends/holidays.

- How do we help the child remember? What is the best way to do this? Repetition is key, but it needs to be in the same way each time, for new people, wearing something identifiable (but safe) that the child can associate with the person, and with affirmative name dropping when someone new is involved, eg 'I like your new teacher Mrs Smith', 'Mrs Smith your new teacher thought you were great at reading', 'Mrs Smith your new teacher is going to help you make a birthday card tomorrow', etc.

- How can we help the child have time away from so much background noise, how often does the child need this? Each child is different, and the course of the disease process causes each child's resilience to 'noise' to fluctuate. Most children will benefit from a break of around 20 minutes after each 40 minutes of intense interaction, especially at school. However, if a child is intently focusing on something, then they can become distressed if it ends abruptly so the use of the 'now and next' process is helpful.

- Use key words and phrases when the child appears lost, confused or irritated, be consistent within your environments, if you are slow and thoughtful, it influences the child to slow. Slow your breathing, your voice, let rhythm of breath, speech and movement provide calm.

At times families can come across a roadblock with how their child copes with their changes, how the childhood dementia brings changes to thoughts and functional abilities, for the child, family and school there can be collective distress and worry over behaviours that can challenge others.

If a family is registered with BDFA please get in touch with Support and Advocacy, we can help by listening, providing support for family schools and the wider team around the child, we can provide education sessions bespoke to the child and their needs.

If you are resident in UK but not linked to BDFA, please consider getting in touch. BDFA can help by listening, supporting at your child's reviews, ensuring that sibling needs are included in Education, Health and Care Plans (or equivalent in devolved nations), we can refer you for counselling if you feel it would help, put you in touch with other families through Peer Befriending, and provide guidance and support for the whole family and for the professional team around the child.

If you are in a country outside the UK please reach out to your local professionals or charity networks.

This guide was led by parents, Sarah, Cheryl, James, Tracey, Naomi, Lynne and Lizzie, and compiled by Sarah Kenrick, BDFA Head of Support and Advocacy. April 2025