WE NEED SLEEP!
Sleep issues in Batten Disease - A Parent Led Guide

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

At BDFA we are aware how, for many families, sleep disruption and loss of sleep on a regular basis becomes quite debilitating for the whole family. Listening to what parents told us about their child's sleep patterns in an interactive session has been invaluable, some children do sleep well, others have never slept well, we want to know why that is and how parents may be able to influence better sleep patterns.

There are 2 complex issues for children with Batten disease, both can cause problems with sleep-

- **Loss of vision** – For children with little or no sight they may be unable to produce enough Melatonin to help the sleep wake cycle, they may wake and be disorientated.

- **Childhood dementia** – This causes changes in how the children perceive their situation, added to it is that dementia can disrupt how the brain produces Melatonin to help the sleep wake cycle, they may wake and be disorientated. Children with Batten disease will compensate when they are disorientated, their brains want to fill in the missing pieces or make sense of the bits of memory they can retrieve.

Wakefulness at night after a few hours sleep, an inability to switch off and an active mind requiring stimulation is more apparent in children/young adults in the early to mid-stages of the disease process. Toward the later stages sleep issues tend to be more physically related to comfort, pain, seizures or incontinence.

IN OUR INTERACTIVE SESSION WE ASKED A RANGE OF QUESTIONS, THE SESSION WAS ATTENDED BY PARENTS FROM THE CLN2 AND CLN3 COMMUNITIES.

When did the sleep problems start for your child/children? Their age, Batten type and before or after diagnosis?

- CLN3 Around the age of 6yrs when they began losing their vision.
- CLN3 Before diagnosis. He has always been a bad sleeper from a baby until now. It got worse and worse over time, once we got the diagnosis it all made sense.
- CLN2 Just over 2, diagnosed when nearly 3
- CLN3 Sleep issues started around 12, after diagnosis.
- CLN3 No real issues yet
- CLN2 Never slept well. Got worse leading up to the few months before diagnosis age 4, however seizures had already started. She has slept through for 10hrs only 3 times in her life. She is 9 now.

What is the longest night-time sleep they might have now?
How many nights of 'good' sleep do you see in a month - Is there a pattern?

- CLN2 Best sleep EVER is 10hrs. Our "Good night" is about 7hrs. Happens maybe max about 8 times a month. It's SO erratic and there seems to be ZERO rhythm. The Paediatric Consultants haven't a clue what to do.
- CLN3 Longest sleep 8pm to 7am (although unusual). Normally would be 8pm to 4/5am (toilet), then back to sleep until 7ish (75% of the time) - she really needs her sleep! Tiredness can trigger seizures
- CLN3 He is normally asleep around 8.30-9. wakes around 12.30 and gets into my bed. Generally, up from 4/5am onwards. I think the longest we have is 6 hours.
- CLN3 When she does have issues, she may not sleep more than a couple of hours per night and in small snatches
- CLN3 My eldest sleeps from 8-5/6 and my youngest 8-7 and sleep like this every night. They fall asleep to audio books and will put these back on if Alexa says it's before 7am
- CLN3 My 14 year old daughter is now sleeping nearly all night 8.30pm to 7am. She sometimes needs to go to the toilet during the night but settles quickly. My 17 year old daughter sleeps from 8pm but wakes up 5 times usually. If she has seizures and has been given Buccolam that day, she will sleep all night. She very rarely sleeps all night. She wakes up from 1am onwards.
- CLN2 My 3 year old daughter has been sleeping better now. We have had over a full month of sleep every night. The longest night being 10/11 hours. Think this is due to new medication, before this we were going months without a full nights sleep. Constantly wakening.

How do the child's sleep problems present?

- CLN3 He wakes up sweating and gets hot. He has to sleep in bed with one of us.
- CLN2 She doesn't sleep for very long, very disturbed, can wake up and not go back to sleep. Tries to fight us and scratch. Wants to go downstairs.
- It started with night terrors.
- CLN3 They used to have great difficulty going over to sleep, I had to lie beside them until they were asleep. Now at 14yrs and 17yrs they fall asleep quickly, but they can't seem to stay asleep.
- CLN3 It takes ages for him to actually settle down and finally get into bed. We get I'm not tired (even when rubbing his eyes and yawning). I'm hungry, I'm hot.. once he's asleep he fidgets a lot. He will sleep for a good few hours but generally is awake from 4-5 am.
- CLN3 Sometimes we think Melatonin can make things worse - she goes to sleep too quickly and then wakes up and won't go back to sleep
- CLN3 Both of our boys sleep really well (luckily) and the only blip we had was a couple of years ago but we feel the addition of Reflexology/Reiki and an aromatherapy diffuser in the bedroom every night and a solid routine has worked. Both sleep from 8pm and awake around 6-7am . Aged 14 and 9
- CLN3 - when she has issues, she is very manic, just can't switch off and constantly repeats the same few words - like a stuck record on repeat.
- CLN3 Generally no sleep issues still, only maybe 2-3x/year (she will wake up every night - but only for going to the loo generally).

- He (CLN3) doesn't really have any sleep issues at present. He will sometimes wake in the night either nightmare or too hot, which can wake his older sister up in the night. He usually comes to wake us up to help settle him back down again.
- CLN2 Wakening frequently during the night. She wakens up and jumps out of bed before she is even fully awake and you can see she is confused but will try to walk about and act like she hasn't been asleep at all. It's as if she can't switch off, wants to go downstairs.
- CLN2 Her dystonia and epilepsy meds make her SO drowsy that need to be taken on a strict routine, however it makes her fall asleep. It's just STAYING asleep that's the issue.
- My 14 year old is always very warm and needs a wee fan on.

How are the rest of the family affected?

- We take it in turns to co-sleep with her. We only sleep together in the same bed once a month when carers come in. We really find it hard.
- Our son gets the rough end on the deal with our impatience and tiredness. It's affecting everything as our social workers don't get it when we want to use direct payment hours at night.
- It affects the child with batten disease as she falls asleep for an hour at school, but they can't keep her awake.
- I really struggle as I have sleep apnoea myself. He always wants me at bedtime, the constant waking up does take its toll on us both. Our relationship is changing.
- I'm a single parent so the lack of sleep greatly affects me. Physically and emotionally draining and exhausting. It greatly affects your health and well-being.
- We haven't really slept "properly" since diagnosis - even when she sleeps, we wake up with the slightest noise/movement. 95% of her seizures are at 4-5am
- The boys are in their routine and sleep well which allows their sibling to have his own time and gives him space and an opportunity to spend time with us or on his own. He also enjoys the aroma filling our upstairs!
- Having night carers and siblings seeing them in their home if they wake up during the night. They may have never met them before - scary seeing a stranger in their home.
- Even though both boys are seizure free still, we have a Sami Cam which gives us the most incredible sense of reassurance and ability to switch off and sleep which didn't happen before the introduction of these.
- Her brother tends to sleep through her sleep issues! (He's 17!)
- We find it really hard when issues happen – the lack of sleep is like torture
- Broken sleep and then having to work afterwards is a bad combination
- My 13yr old tends to sleep through it. But we try to get up with our daughter to keep her quiet, so it doesn't affect him for school etc
- With me and my husband it's been very hard trying to run on empty during the times where the sleep was so bad. My husband works, I don't. So I sleep with my daughter every night in her room. Then during the day I would have a nap at some point.

Does the lack of sleep have an effect on your child with Batten disease? if so, what does it look like?

- CLN3 When she has issues, she finds it difficult to break the cycle - she just seems to "power through" and then have naps (like when in the car) but not at night-time
- CLN3 He gets very grumpy but will still continue to carry on with his day like he's slept.
- CLN3 Yes, they will be very tired the following day. Sleeping more in school and in transport. Very difficult to keep them awake.
- CLN3 If our eldest 14 doesn't sleep well it affects his balance and mood and impacts his word finding, he becomes zombie like.
- CLN3 If our youngest 9 doesn't sleep well his is the opposite and becomes a full on, hyper, highly strung young man who can't sit still and talks a hundred miles an hour.
- CLN3 If he has a disturbed night, his mood is affected, and he is more likely to lash out with short temper.
- CLN3 He doesn't really have any sleep issues at present. He will sometimes wake in the night either nightmare or too hot, which can wake his older sister up in the night. He usually comes to wake us up to help settle him back down again.
- CLN2 Seizure activity seems to be worse with her when she isn't sleeping well
- CLN3 He gets very grumpy, but will still carry on with his day like he's slept.
- CLN2 It affects her as she falls asleep for an hour at school, but they can't keep her awake.

What are the routines you use or have tried to help the child sleep? Do they work?

- CLN3 They have the same routine every night. Supper, meds, washed, in bed, story and then listen to a story online.
- CLN2 Nope nothing really works. we just try and keep to the same routine
- CLN3 Medication not too close to bedtime, in particular lamotrigine (does help)
- CLN3 Audio book (which she chooses) with noise cancelling headphones - which we remove when we go to bed. Weighted blanket, routine really helps
- CLN3 He always wants me to help him get to sleep
- CLN2 Reading in bed, no screens 30 mins before bed, the same tonie box character to fall asleep, hugs down stairs, just mum or just Dad. Some things make her calmer, but not always.
- CLN2 She seems to be very sensitive to routines. We try to keep every evening the same. Food. Medicine. Wash. Bed. So far that seems to be working. We notice her a bit distressed if anything's changed in a big way, she was also diagnosed with autism before the Batten diagnosis. We also use white noise
- CLN1 I used to play the same lullaby on the i-pad for him every night to let him know it was night time and a different song in the morning (more upbeat and active) to let him know it was the morning
- CLN2 She is always cold. Won't keep blankets on her, so sleeps with Pyjamas and a onesie on. If I didn't put the onesie on, she would waken during the night with cold hands and feet
- CLN3 My 14 year old is always very warm and needs a fan on.
- CLN3 My 17 year old uses a sleep system to keep her comfortable.
- CLN3 Both boys only like a thin quilt or just a blanket and the texture is very important to settle.
- 14 Year old, A warm but not hot bath, audible on his Alexa, aromatherapy diffuser in his room, a timer so that the story ends because Alexa has said time to finish. A thin quilt and the rule that if he wakes before 7am he is to put his Audible back on.

- Fortnightly reflexology/reiki session are the absolute key to good sleep and I do shorter fill in session in between these.
- 9 year old A warm but not hot bath, acoustic pop on Alexa or Tonie story and a snuggly thin blanket. Aromatherapy diffuser and monthly reflexology sessions with me doing smaller session in between.

How do you manage or support them when they wake during the night?

- CLN3 When my older daughter wakes up, she needs a lot of reassurance and listens to a wee story online. Sometimes this settles her and sometimes it doesn't. She also can be very confused and asks what's that e.g. her blanket or toy etc. Younger daughter just likes to listen to story online and falls back to sleep quickly.
- CLN3 Help her to get to the toilet. Keep calm and quiet.
- CLN3 When she wants to chat, don't engage and tell her that it is night time and to go back to sleep, an audio book as a distraction sometimes works.
- Rarely, we'll bring her into our bed if she's really unsettled
- CLN3 Melatonin (liquid) seems to work best when she wakes up in the middle of the night - not at normal bedtime
- CLN3 I struggle as I never know what kind of mood I will see in him at whatever time he wakes up. I try hard to get him back to sleep or we just get up and start our day.
- CLN3 Consultant suggested Valerian root (Kalms) but she won't easily take tablets so we've not tried this. Also heard from someone about magnesium cream on the soles of the feet - not tried either.
- CLN3 He usually gets in our bed for a cuddle for 5 minutes and then we take him back to his bed and tuck him in. He usually goes straight back off.
- CLN3 If the boys do wake in the night which happens extremely rarely it’s just very minimal conversation as not to ignite a lengthy repetitive dialogue and a few minutes of whale music on their Alexa.
- Again, it's just the waking up that's the issue. She tends to be frantic and wild, scratching, biting, pulling etc 15 mins before her falling asleep. We give her a fiddle toy which MASSIVELY helps the behaviour. If she doesn't have a fiddle toy, I come down bleeding.

Describe the bedroom, type of bed/mattress, thickness of duvet, colour scheme, light on or off, other light sources, access to toys/books/media?

- Toy story bedroom, so blue with a double bed, normal air sprung mattress.
- duvet not to thick but has a blanket at the bottom of his bed if needed. He has a lamp on but gets turned off when daddy gets into bed. He has toys, books, audio in room.
- He like the TV on to fall asleep with.
- Dark room, as quiet as possible (to avoid external distractions), she has an audio book.
- Single bed, normal mattress, weighted blanket. Thin duvet. Epilepsy pillow, no toys, no access to her Alexa (otherwise she'll get it to play music - like Coldplay at 3 bloomin’ a.m.!!)
- Both girls I think can only see light and dark. They both like a wee lamp on in their bedroom. They have their own rooms. Eldest sleeps on a special mattress (I forget the name). It's a hospital bed with sides up and we use a sleep system. Younger daughter sleep on a small double bed. Both bedrooms are calm colours. They don't look at any screens at bedtime.

- Small double bed. With memory foam mattress. No toys in a box at the bottom of the bed so she has no access to them during the night. We have a machine that plays white noise. No light. she needs all lights off to go to sleep.
- He shares a small room with big sister, bunk beds, he's on the bottom. Normal mattress, in winter a sheep's wool duvet (its good for temperature regulation as it breaths), in summer 3 tog. He has set teddies that have to be in bed with him. Main lights off, red fairy lights on one side of his bed. He likes it dark. There are books under his bed, but he doesn't access them at night. There is a small orange night light.
- CLN2 Double bed, with a firmer mattress, but foam topper. light off and as dark as possible. There is a TV, but it doesn't go on until she wakes up and is awake for over an hour. Book rack, White and pink colours. She has good vision, can see things 8-10 metres away

How well do they sleep when away from home? Do they go anywhere regularly overnight?

- CLN3 He sleeps well at my sisters, and apparently good at nanny and grandads. He's not had any other overnight respite.
- CLN3 Both boys sleep at extended families/friends houses and when we stay away from home on holidays etc they both sleep the same as when at home. They don't take everything with them such as aromatherapy diffuser etc. but do all sleep in the same room when away from home and wouldn't agree to staying away otherwise. Poor middle sibling would prefer this not to be the case but for now he plays along.
- CLN2 No rhythm. She has 2 nights away at the hospice per year, sometimes 7hrs of sleep, sometimes she treats the nurses to the grade A treatment of a 1am wake up. She has 2 nights away per year currently.
- CLN3 He has been away at Cub camp in September last year and slept well apparently. When we camp, he sleeps well, and when he stays at my parents.
- CLN3 Very variable - normally depends on her underlying mood at the time.
- CLN3 She generally doesn't sleep as well as at home (for the first night at least)
- CLN3 Stays at Grandparents from time to time, Granny insists on sleeping in the same room (because of seizures and she's not good great hearing) which really doesn't help - as she knows someone is there and it keeps her awake.
- CLN3 Just started 1x night/month with her PA funded by direct payments - she slept well for the first one :)
- CLN2 She has never stayed away from home and wouldn't at all.

What can parents do in general to help?

- All Children with Batten disease are different, their personality, rate of progression through the disease pathway, and their family life in general all play a part when it comes to sleep. Some children learn to self-manage their wakefulness without disturbing their parents too much! Others have times where their internal 'off switch' is faulty. Many Batten children don't seem to need as much sleep as others, some have cycles of good sleep then restless sleep.

Can we teach self-management?

It depends on the type and onset of Batten disease and the child's developmental age along with their personality. Children with a juvenile form of Batten disease are potentially more able to learn techniques as they are developing their skills and understanding better than children with an earlier disease onset. Second to this it depends on the house rules and how they understand these.

Can we break down self-management to help with learning?

Yes, we know children with Batten disease have childhood dementia, they need to learn by repetition, and we need to understand that when they are tired, worried or anxious they find it hard to focus and hear what is being said.
It is important to recognise the good things and to observe and listen to behaviours that are difficult to cope with.

How do we do it?

- Think like the child – think what they can see, feel and hear when they are in bed, how much stuff they have and where it is. How much shadow there is and how it might affect them. How safe they feel, what could make them feel safer.
- If they have good sight, think from the dementia perspective, if they have changing, little or no sight think from the sight-loss and dementia perspectives.
- Wee time – They may have changes in full bladder perception, can you develop a safe route for them to use the bathroom alone or develop a rough schedule of using the loo before they are fully awake? Do you have the same words to describe going to pee?
- The bed has to be comfy, big is not always better, sometimes if children are anxious and have sight- loss they will think they are somewhere else, they will forget they are in bed and go into the flight or fight mode, anxiety can escalate to panic very quickly and how you react will promote different emotions for them around their confusion. Sometimes it is best to have a single bed, one side against the wall and on the other a soft rug on the floor so they can understand the floor texture near the bed and the orientation of the bed in the room.
- The mattress, pillows and coverings make a big difference, especially for children and young adults who may not be able to manage their own night-time comfort in bed, for anyone that can't move well there will be a degree of potential discomfort that can trigger discomfort, pain and anxiety.
- In CLN3 it is common that children and young people are unable to regulate their own body temperature, they often get overheated in bed, other children may feel the cold and want to be wrapped up in their blanket or onesie.
- Focus on daytime as well as bedtime, think about day routines that can help remove excess energy.
- After tea, slow the activities before bed, minimise the use of electronic devices and screen time.
- Try to train your child early on to accept headphones, especially children with CLN3, they usually are quite content listening to a story, and it can help get them back to sleep.
- Try to be calm

- When they wake have a plan of how to interact and stick to it – as much as possible!
- Behaviour and sleep issues often go together sometimes it helps to look at the whole thing.
- Speak to your child's Consultant or prescribing Doctor, ask them how the disease drives wakefulness, explain the effects on the whole family and take a sleep diary with you to demonstrate wakefulness. Ask for guidance regarding how best to give any prescribed medicines, and if you are feeling overwhelmed, ask the Dr to write instructions down for you or to send you an email so you can be clear and feel safe and in control.
- Extra support at night, there may well come a point for each family where there is a clear need for extra support staff in the home at night. Social care is the starting point for this and BDFA Support and Advocacy can help you access the right support and provide evidence of need based on how Batten disease affects your child.

Sleep, and the lack of it, can have a huge impact for families living with Batten disease, BDFA is here to help. If you are resident in UK but not linked to BDFA, please consider getting in touch if you feel you would like our support.

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, Lauren, June, Sarah, Dave, Cheryl, James, Caroline, Tracey, and Mimi, and compiled by Sarah Kenrick, BDFA Head of Support and Advocacy . May 2025