

Supporting anxiety, panic attacks, hallucinations and night terrors in juvenile CLN3 (Batten) disease

Please note: the information contained in this document is intended to provide supportive guidance to families, carers and associated professionals. It is not intended to be, nor is it, medical advice for individual children or adults with juvenile CLN3 (Batten) disease. Parents and carers should consult the person's GP prior to changing medication, medical treatment or prescribed activities. If you are a professional and you require more information, in the first instance please contact Sarah Kenrick (contact details at end), who can provide you with a link to the Adult Neurology Consultant for Heather House and/or the GP for Heather House.

My name is Sarah Kenrick. I have worked with children and adults with juvenile CLN3 (Batten) disease for nearly 30 years, for the past 20 years with SeeAbility at Heather House. I am a registered nurse for people with a learning disability. Aside from epilepsy, the issues around anxiety, behaviour, hallucinations, panic attacks, night terrors and insomnia are of huge concern to nearly all parents I have met. There are some great research articles that look at these issues and some great clinicians in the UK and other countries that have extensive knowledge and skills in assessment and treatment of children in these areas. But the questions I am most frequently asked are around why is my child doing this and what can I do to help them cope or manage.

Unsurprisingly, I don't have all the answers, but over the years I have seen some similar themes for people living with juvenile CLN3 (Batten) disease and have learnt how I can adapt my behaviour to better understand and work with them to help them be in control and manage their own behaviour. The one thing I am certain that I don't know, is what it is actually like to have this neurodegenerative disease and live with its effects in my head and body every day. That is not to say every day is awful or dreadful – most are quite ok and ordinary, some are great!

Each child is unique in how they exhibit their feelings and will have their own thresholds or tolerances with regard to triggers for panic, anxiety and hallucinations, although there are some common signs that can be seen, and each family member or carer has their own coping strategies. So the best advice I can start with is to have a solid and consistent method of recording or describing your child's particular issue(s) and ask everyone around your child to use the same method. Something like an ABC chart is often the easiest thing to use – this has four columns:

1. **Antecedent** (what happened immediately prior to the behaviour)
2. **Behaviour** (what is it that you have seen)
3. **Consequence** (what happened as a result of the behaviour)
4. **The time, duration and who was involved** with the person during the episode

Gathering this information does take time, but this type of detail is what I and other professionals will need to know in order to help, and once you start doing it you can often see common trends for your child.

I would also advise a video of the person's behaviour be taken on more than one occasion, as this is invaluable in enabling professionals involved have a clearer understanding of what's happening. Any trends or repetition in words or movements can help to shape a diagnosis. Please be mindful for adults that a capacity assessment and, if necessary, a best interest process may need to be

completed. The seizure activity that children and adults develop can be very misleading and subtle in its presentation so the potential for seizures to contribute to the behaviour must always be ruled out or in.

Another area to consider is what the visual perception status is for the person. Having worked with some inspiring rehabilitation practitioners for visual impairment and with information from the annual Ophthalmology Consultant eye clinics held at Heather House each year, we have seen that individuals tend to lose their sight in the same or similar pattern, but over different lengths of time. So some people who are in their late teens or early twenties can still have light and dark perception, and occasionally some degree of functional vision. Understanding whether the person has light perception or can identify detail and how they need to hold their head to 'see' well is important. Sometimes the behaviour the person exhibits is influenced by external factors, including the quality of the light and where it is coming from, as well as frustration at not being able to interpret well what they can see.

Heather House

At Heather House our team consists of an Adult Neurology Consultant, a GP, Nurses (both general and Learning Disability fields), Speech and Language Therapy, Physiotherapy, Rehabilitation Practitioners for Visual Impairment, our team of carers and day service staff and the families of the people we support. We also have internal access to Personal Behavioural Support Practitioners and through our GP to Psychiatry. We have also learnt much from clinicians and specialist practitioners in other countries across the world.

To better understand how to support a person with juvenile CLN3 (Batten) disease who experiences anxiety, panic attacks, hallucinations or night terrors, over the years we have identified general descriptors for each category, what we know about why they happen and what we can do to help alleviate these episodes.

Anxiety and panic attacks

I think the best advice I can give is to look at this from the individual's point of view. The rationalisation of a situation, event or task is dependent on our own learnt perception of it, or of similar experiences we have had. The juvenile CLN3 (Batten) disease process causes a cognitive decline with difficulties in recall and processing, as well as, for some individuals, what we see as psychotic episodes. This all combines with the loss of vision and physical decline in posture, mobility, core strength and a gradual increase in fatigue. Sometimes the combination of the psychological, physical and emotional factors come together and could be described as a brainstorm, meltdown or temper tantrum.

At Heather House we work on the basis that we all need to have some control over what we do and how others interact with us. We have seen many times how people living with juvenile CLN3 (Batten) disease will struggle very quickly when they lose control over a situation that we may see as quite small, but must be so frightening for them. So when we see anxiety building we say 'Slow down..... take control..... breathe.' This not only gives the person time to process, but it also enables us to take a step back and think how we are affecting the person.

We have to remember that our good intentions do not always support the person's behaviour. An example of this is: I am scared of heights, so take me up a very tall building and tell me to walk over a glass see-through floor – it's not going to happen! Whatever your good intentions are in trying to help me overcome my fear, ('Don't be scared, I'll hold your hand, it's a really strong floor, now you're being silly!') it won't work. My anxiety will build and frustration and anger for both parties will be the outcome. Allowing me to take control ('I can see you're scared, what do you want to do?') is the only way for me to stay calm, then I will decide what I do.

This is particularly important for us to understand - our good intention to enable someone may well be the disabling factor, but seeing that is often painful, as we are all only trying to help. Little by little, this disease is taking control away from the person, so we need to be able to see if the disease is leading the behaviour or if the personality is leading the behaviour. So for parents, caregivers and professionals to understand how the disease affects thought processes, recall and coping is essential, and within this the knowledge of who this person is, their personality, interests, memories, must be at the centre of everything.

'We change other people's behaviour by changing our own' (anon)

Allowing the person to be able to retain control in whatever way, big or small, is so important, but there is a fine balancing act between encouraging them to be a good citizen, take responsibility and chances, but not becoming so dogmatic that they lose out on opportunities. The impact on other family members must at times be overwhelming, so for those of us who are professionals, we also need to be mindful of our good intentions when trying to find solutions. No, it's not easy, but being aware of our own behaviour and enabling others who need it to take control is essential when supporting anxiety issues.

The onset of mobility problems to the degree that the ability for smooth transitions between different postures becomes increasingly difficult can be very distressing for the person. At this time anxiety can be seen particularly around activities such as laying down, getting up, rising from sitting, and starting to walk. Equally, as they lose the ability to maintain full continence the levels of anxiety may rise. Often they may rationalise these events in a way that others find it hard to interpret, particularly as speech is more challenging when the person is distressed, and yet at the same time day-to-day life has to go on.

So as the individual loses their ability to mobilise and speak clearly and rely more on other people and carers for their daily needs, they may experience agitation. This may lead to panic attacks, if not supported consistently in a manner the person finds acceptable and affords them 'control' of the situation. They may hyperventilate, tremble, become flushed and perspire, have repetitive speech and may become unable to 'hear' what others are saying. This can appear similar to a hallucination, but the difference is often that one person is able to take control, have a calming influence, speak slowly and bring the person back into this calm space before moving on.

For example, getting in the car, going out with family or friends could be the number 1 on someone's list of favourite things to do. Then suddenly it's a huge thing with tears, screaming and refusal. Why? What has happened to change this? I think in this disease there are times where the person can cope well, but when the tipping point is reached, everything is too much to contemplate. How we react to that tipping point at the time can make a big difference to how the child copes with other tipping points. Of course this will normally happen when you really need to get to the shops, or are on the way to a doctor's appointment, or a wedding, or a birthday, theatre show, etc. It hardly ever happens at a time when everyone can say, "it's OK, let's go back inside, slow down, have a hug, calm down and then talk!"

Often just being close and making a quiet 'shh' noise whilst holding the persons hand or gently stroking their arm can be enough for them to reconnect with you, also gently, slowly and quietly saying the persons name until they say 'what?' so 'Jane' pause 'Jane' pause 'Jane' pause, will after a few repeats get a 'What' back. Once the person has said 'what', you can say 'I think this (whatever it is) is scaring you, is it?' Often then you can slowly move forward with gentle probing to find out what the problem is.

At Heather House, particularly in the later stages of the disease process, we have found this more prevalent when the person is unable to speak in simple sentences or full words and where they have multiple types of complex epilepsy. We found that with calm, controlled and constant approaches, particular with regard to moving and handling, and giving the person a feeling of being in control, the agitation and panic attacks diminished rapidly. Subsequently we were able to safely reduce some seizure medication with beneficial effects.

Where different or new types of epilepsy appear to be an issue, we would advise in the first instance a recorded detailed method is instigated, detailing how the person will be consistently supported. This should be followed in all environments prior to increasing medication. This is particularly important in the mid to later stages, where it is linked to some type of physical intervention or moving and handling, i.e. hosting, and/or incontinence using a commode or using shower chair/trolleys.

Factors that may contribute to an individual feeling disorientated – 'Golden Rules'

We are fortunate to be able to work with a group of people with juvenile CLN3 (Batten) disease at Heather House, and we are a large and diverse group of caregivers. So we have identified what we consider to be our golden rules. These may apply more in a school or care setting than at home, but are simple things that in our endeavour to be helpful and ease distress, we may not always see.

Too many people talking at once – is someone is easily confused, to help them make sure only one person is speaking at once. They can't cope with too many names, so our normal routine of saying who we are every time we walk in a room will raise the agitation level. Let the person supporting decide when to tell them who is in the room, and don't speak until you are invited to.

Too much noise – Be aware of the volume of your own speech and lower it, as hearing loud voices when they are already disorientated will make it worse. Turn down any music or TV.

Being too hot – People with juvenile CLN3 (Batten) disease find it hard to regulate their own temperature at times so open the windows and allow air to circulate. It is a natural response when someone's agitation levels rise that they need air. Being in a warm or stuffy environment can make a person feel closed in and unable to breathe.

Low fluid level – People with juvenile CLN3 (Batten) disease often won't want to drink when the weather is hot. Ideally aim for a fluid level of 1½-2 litres a day. This may not always be possible, but if fluid levels consistently drop below 1 litre daily, then this will cause dehydration, which will increase the possibility of agitation and confusion.

Constipation – If the person is not able to clear their bowels and feel they need to pass a stool, they will be agitated. In juvenile CLN3 (Batten) disease individuals progressively struggle to evacuate the bowels fully each day, which causes confusion as the individual is not able to fully

rationalise the need to go and may be embarrassed that they may be incontinent. Oral laxatives, more fibre in the diet, and encouraging a moist diet with not too much salt (particularly if they don't want to drink as much), may be helpful. Do also tell the person's GP, as there is so much support for this available.

Providing a positive rather than negative consequence for an action – People with juvenile CLN3 (Batten) disease may find it harder to rationalise negative comments as they age, and may start to believe they will come true. For instance, saying something like 'you need to put your shoes on or else you could cut your feet' can cause them to believe that their feet will indeed be cut and will instantly raise anxiety levels. Equally saying 'be careful, you might hurt yourself' can mean 'you will get hurt'. So we try to say 'shall I help you with your shoes, they look really smart and comfy and keep your feet safe'.

Lack of planning or too much planning – There comes a point when doing stuff outside of home or even outside the normal routine becomes 'A Big Thing' to the person, but maybe not to others who are on the periphery of the person's world. Planning is important to make sure things are accessible, planning an escape route if things get too much for the person, to think of the best time of day to do the thing and who is best to do it with. But too much planning with the person does not always allay fears. Sometimes it makes their anxiety worse to the point that on the day they may be poorly or have a seizure. We have learnt that we have to do the planning in detail and sometimes for days and maybe even weeks (depending on health and needs) before 'The Big Thing'. However, depending on the person we work with, we may limit how we plan with the person to:

- Where are we going?
- What we will do when we get there?
- Who do you want to go with you to help you stay safe?

Hallucinations

The dictionary describes hallucination as '**an experience in which you see, hear, feel, or smell something that does not exist, usually because you are ill (or have taken a drug)**'.

Sometimes children and younger adults with juvenile CLN3 (Batten) disease will experience a hallucination where they believe they can see and feel and often hear something which is not there, such as spiders crawling towards or over them, falling from a great height, being dead, having bees in their ears, and so on. The important factor is that the person can describe it, and genuinely appears terrified. They will be wide eyed and have enlarged pupils, be shaking or rigid, be sweating and flushed, and depending on how someone else approaches, they can lash out and hit someone because that person becomes part of the hallucination. Hallucinations can last for minutes and sometimes for hours, sometimes come and go many times in a day, but will usually be debilitating for the person and frightening for others around them. During the hallucination the person will often be looking upwards and slightly to one side.

Why do hallucinations happen?

At times there is nothing we can think of as specific that triggers hallucinations, but when we can analyse information gathered over time we might be able to link this to pain, urine infection, constipation or an increase in seizures. We know that some people never experience hallucinations, others have recurring themes, and others may have a period of these for a year or two, then no

more. It is much harder to analyse this for people who find it hard to talk. So at Heather House we always use quiet, gentle reassurance, but focus less at the thing that is scaring the person (e.g. the spiders), because we know we can't necessarily talk them away, and we focus more at the physiological symptoms of flushing, sweating, shaking and pupil enlargement. It is this group of physiological symptoms that people with frightening hallucinations experience. The other common factor is that the person appears stuck, unable to move on or away from whatever is happening, often mumbling the same words or phrases, and if they can move they will often be making picking motions or have a one sided twitch or weakness.

We have worked with some young adults who have had periods where they are engaging with something we can't see, but appear very happy, chatting to something imaginary like a rabbit or cat, and being able to tell us where the thing is and what it looks like. The difference is in the behaviour exhibited, which is always calm and happy. Whether this is hallucination or not, we do not look to treat this with medication, as there appears to be no harm. To date we have not seen these happy episodes move on to seizure activity.

Is the physiological pattern of a frightening hallucination indicative of a form of epileptic activity?

At Heather House, with our team's experience, we believe the answer is yes. The patterns we see are most likely to be a type of aura leading into focal or simple partial or complex partial epilepsy. Juvenile CLN3 (Batten) disease is a disease that causes epilepsy to become more frequent and complex as the disease progresses, so we do not request repeat EEGs as we already know the seizure activity is there, and the stress of this procedure is not necessarily in the person's best interest.

What can we do to help?

When we see these episodes we give an 'as required' prescribed medicine, usually lorazepam if the activity does not stop after 15 minutes. Once we recognise the pattern, usually after the third one, we give this medicine as soon as is possible, depending on a variety of factors such as where the person is, who they are with and whether they have a gastrostomy device (sometimes the hallucination means the person is unable to swallow safely). We also ask our GP to liaise with the Consultant Neurologist as necessary to increase one of the regular anti-convulsants after we have ruled out urine infections or constipation, as we know that these can lead to increased epilepsy quite quickly in juvenile CLN3 (Batten) disease.

Over the years at Heather House, we have developed some simple rules for ourselves to ensure we do not inadvertently cause these 'hallucinations' or seizures to escalate further:

- Support the person to as quiet an environment as is possible and practical - if at home, support them to the room they feel safest in, e.g. their bedroom.*
- Try to make sure there is only one person talking to them. If other people are present they should try to remain quiet as extra voices will mean more confusion.
- Within your family, or team if at school, identify how you will approach the person during hallucinations and stick with this approach to maintain continuity.
- Talk in a quiet and calm voice, only a few words at a time, remembering to breath between short sentences, as this helps you stay calm. If you are calm this will help the person feel safe.

- Try to avoid asking questions as we have found this can lead to increased agitation. Once the person is beginning to calm then it is safe to ask questions with a yes/no answer, such as 'Are you feeling better?' 'Are you ok?'
- If the person has to be physically assisted during the hallucinations, e.g. from one room to another or from chair to bed, have a pre agreed plan as to how this is supported, act quietly and apply team strategies, as above. Physical manoeuvres such as using a hoist for people who are not able to weight bear are, in our experience, best done quickly, with the person being quietly informed as to what is going on around them and gentle praise once the manoeuvre is complete.
- During any type of anxiety, panic or hallucination/seizure, the person may not be able to identify the potential dangers around them, so guarding or shielding the person from harm is important to prevent injury. However, we would never advocate any form of physical restraint, as this will reinforce their fears and cause the person to become more agitated.
- If the person's safety is in doubt they should be supported to sit or lie on the floor where they would be least at risk. Physical contact and reassurance should again be restricted to the main carer.
- Record each hallucination/seizure on the person's seizure chart. State clearly where they are and what they are doing immediately prior to the hallucination, what the person is doing and or saying, how you have interacted with the person, how long it lasts and whether any medication was required.

*In a school environment, it is helpful to think ahead and support the child to establish a space that they consider safe when they are able to. Environmental factors may well antagonise hallucinations (e.g. noise, crowds and prolonged periods of an activity, as some children with juvenile CLN3 (Batten) disease may struggle to focus for more than 20 minutes on a task). Have somewhere away from other children to go to, so that school staff can automatically have somewhere to support the child to when they are struggling to cope with the disease process. Use this space at least once a day for a short period of what the child sees as high value one to one activity, so that the space will be more meaningful more quickly.

Night terrors and insomnia

Many of the children with juvenile CLN3 (Batten) disease that I have supported over the years have had times when they have been very happy to stay awake for some or most of the night, often listening to stories on their tablets or media players. They do not appear unduly tired from this and would maybe sleep for longer for one or two nights, then be awake again. However, sometimes this affects other family members – sometimes the child will be bored and want to have others to play with or sometimes their behaviour could be dangerous. For some there are episodes of night terror, when the child wakes and is clearly terrified of something, usually appearing flushed, hot and wide-eyed.

It is helpful to be able to understand the degree of visual impairment the child has. The term 'blind' covers a wide range of visual impairments – sometimes the child may have peripheral vision or see light/dark, shadows and shapes. It may be that as vision decreases the child can see shapes and shadows at night depending on light outside the bedroom, whether there is a nightlight in the room and so on. It may be helpful to have either a very dark room with blackout blinds at the windows and no light on in the room, or a good amount of light in the room to prevent shadows. Again this is trial and error. Equally, a consistent bedtime routine can be helpful as it is for other non-affected children.

Why do some children have night terrors and others may never have them? This may be due to the way the disease progresses for each child. Night terrors appear similar to hallucinations, which may well be a form of focal epilepsy. Where night terrors become a more consistent theme and night time routines have previously aided sleep, then this may be more likely. Night terrors are most frequently seen in childhood. It is unusual for them to occur only in adulthood. As the person ages, the disease process sadly leads to more times when the person is asleep, so is much more a symptom of childhood, adolescence and late teens.

Some children are prescribed melatonin to help them sleep at night, as the onset of blindness can affect the production of melatonin in the brain. Some families report that melatonin works well, others not so well. At Heather House we have also seen this, so for each person it is trial and error as to whether melatonin is supportive to a good night's sleep. For night terrors that are not 'one offs', speak to your GP or Consultant. A small amount of clonazepam, clobazam or lorazepam at bedtime can be very helpful in controlling these, as they have some appearance of epilepsy. In our experience with teenagers and younger adults, these types of medicine have not only aided sleep, but has a beneficial knock on effect to the alertness and functional ability the next day.

And finally...

Sometimes we can find the cause or triggers for all or any of the above reasonably quickly depending on the information available, but at other rarer times, it is much more difficult. The most important thing is for everyone involved to be working together and if there are person centred plans in place to make sure they enhance the quality of life for the family and are not so complex that different people can put different interpretations on them, the term singing from the same hymn sheet is most apt.

For more information, advice or support please contact

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