

Postural support in the later stages of juvenile CLN3 (Batten) disease – Supportive positioning during sleep and when resting on the bed

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. The pictures contained in this document are posed by staff working at Heather House.

SeeAbility's Heather House opened in July 1999 specifically to provide long term and palliative care and support for people with juvenile CLN3 (Batten) disease. For ease of reading, in this article we will refer to 'Batten' rather than juvenile CLN3 (Batten) disease. Since opening, we have supported 30 people with Batten aged between 16 and 32.

Our multidisciplinary team work together with each person to ensure the symptoms that Batten causes are supported with knowledge of the condition and with empathy, to enable the person to enjoy the best quality of life possible according to the progressive nature of the disease. One area that our Physiotherapy and Nursing Team have developed skills in is postural support, focusing on the seated position (which will be explained separately) and on the position in bed.

Why is it important to support the resting or sleeping position?

In the later stages of the disease, we have seen how people with Batten gradually spend more time on the bed. This happens because the person becomes more quickly fatigued with day-to-day activities. They experience a weakening of muscles, which means that sitting and being engaged with other people seems to be tolerated for shorter periods of around 1-3 hours, depending on the person, the time of day and the activity.

Not having the ability to move and change position to find comfort whilst sleeping or resting on the bed can be very distressing for any person. The descriptors for the disease will often refer to the person becoming bed-bound or bed-ridden in the later stages, however, in working with people with Batten for many years, we have seen how building in regular periods of rest on the bed, along with ensuring wheelchairs are commissioned with the person's specific needs in mind, and being focused on safe and effective postural support, the person can be enabled to continue doing the things that they enjoy every day.

At Heather House we are always mindful that we are working with adults or older children with Batten, who often have very clear opinions of how they want us to support them. So we always work with the person, listening to their voice and their body language in order to understand their fears. We work slowly to gain trust and support them to find out how they can control what they are doing and show us their preferred posture and position in bed. We would always encourage a person to lie on their side (side lying) once they are no longer able to change position themselves, as this position can help minimise risks, discomfort and pain associated with increasing immobility.

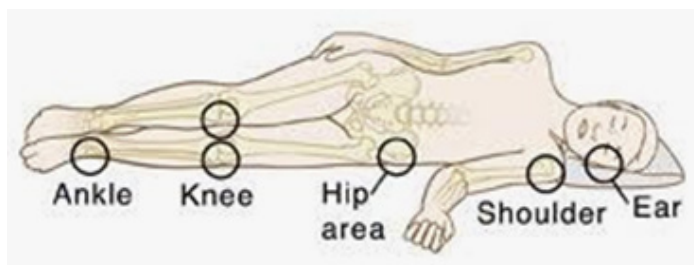
In the main, people in the later stages of Batten appear to prefer side lying, but we have worked with a small number of people who were only comfortable lying on their backs. We supported those people to find comfort and a safer position by using 4 way profiling beds.

Led by our Physiotherapist, we have put together guidance around side lying and postural support for families, particularly parents, care givers and professionals living and working with Batten.

Side lying explained

First we look at the areas of the body that are at risk of increased pressure when laying on the side:

Other important things to consider are: the type of mattress, pillow density, room temperature, bed linen, time spent on the same position, tilt in space bed mechanism and the need for specialised bed support to help maintain the body position.



Getting ready

The bed must be flat, clean and with no creases on the sheets. At Heather House we always follow safe moving and handling practice. We always make sure that pillows and other equipment needed are close to hand and that we don't move away from the bedside during the process.

We look at how to support a person with one care giver and when it is safest to have two.

For the purposes of the pictures (all posed by staff working at Heather House), we do not show bed rails raised. However whenever we have one carer supporting a person with high levels of need we always ensure the bed rail furthest from the carer is locked in the up position to prevent falls.

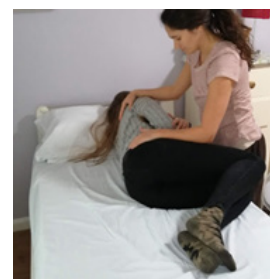
1. For a person who has some limited ability to help move themselves but needs another person to help them turn

- This is often a pivotal point for the person to be able to feel safe with their care giver or to feel out of control. So starting in the back-lying or prone position, encourage the person to shuffle over toward one side of the bed if they can. If they can't, then follow the support guidance for two care givers below.

Once on one side of the bed, ensure both legs are bent (feels safer than just one leg bent). This ensures that when they are supported to turn to their side they will then be in the middle of the bed.



Then the carer has to put one hand behind the person's shoulder and the other hand on the person's hip; then turn. Communication is crucial at this point, as turning to the side can be very scary for the person.



2. For a person who has no functional ability to help move themselves it may be safer to have two people to help them turn – We always look at how calm the person is. Supporting someone to turn when they have no control can be very disorientating and sometimes this can lead to the person grabbing tight to the bed or the carer, so with one carer taking the lead and the other following, the person can feel safer and calmer.

Firstly we look at the bed height – here the bed is too low for the care givers: the lower back is at high risk of injury



Now the bed is too high for the care givers: the shoulders of the person supporting will suffer when the bed is too high.



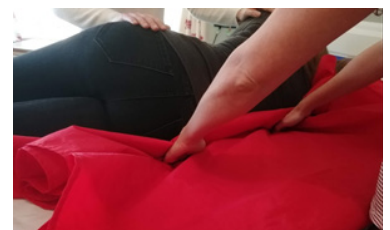
How to support someone to slide sideways using a slide sheet

Here the bed is the right height for the care givers

1. Help the person to turn to the side



2. Place the slide sheet behind the person and tuck it underneath them, always explaining what you are doing



3. Then support the person to roll onto their back and slightly to the opposite side



4. Now pull the slide sheet so it is flat on the bed



5. Coordinate with the other carer, communicate with the person and at the count of three: one carer has to pull the top of the sheet and the other carer helps this movement by pushing the person lying on the bed towards the other carer



Side-lying position

Once the person is side-lying, check the following:

1. Head position

The density of the pillow will determine the position of the head. A pillow too thick or too slim can make the head be tilted:



Correct position of the head



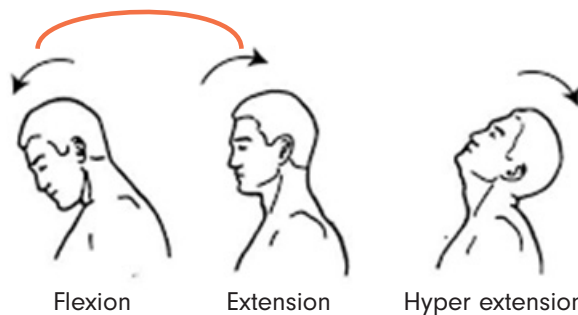
Pillow too thin. The head slightly drops down – can cause the neck to ache and pressure on the ear.



Pillow too thick. The head is pushed up - can cause the neck to ache and pressure on the ear.

The person's airway and breathing can be compromised and the risk of aspiration (saliva or stomach contents going into the lungs when the swallow mechanism is reduced or weakened) is increased depending on the level of extension of the head:

It is easier for the body to breathe and swallow in these two positions.



More difficult to breathe and higher risk of aspiration.

For this reason, it is very important to slightly flex the head and, most importantly, make sure that it does not go to extension throughout the night.



Correct

Too flexed

Too extended

2. Spine

It is very difficult to position someone so their spine is perfectly aligned. The most important thing to avoid is a rotation in the spine (twist in the upper body – see pictures below).

When the spine is twisted, the chest area and all the organs inside are slightly stretched, which could increase the reflux symptoms and affect the ability to breathe (the ribs find it more difficult to expand in this position). Therefore, to minimise these risks, we recommend positioning the person with no rotation in the spine.



It is easy to check if the body is rotated (twisted), as the upper shoulder will look out of line with the rest of the body.

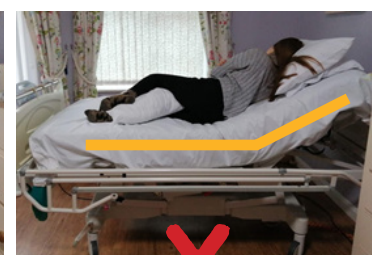
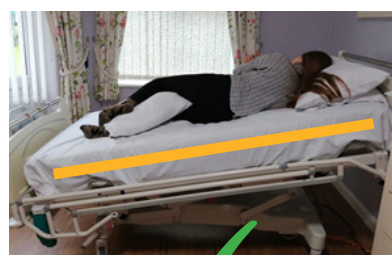


In these pictures, the left shoulder is out of alignment due to the spine rotation. This position would be unsafe for people who are at risk of aspiration or people who suffer from reflux.



This can be easily modified by supporting the left arm in front of the body, so the one shoulder is above the other. Like in the recovery position, but with supportive pillows to the upper leg and the upper arm in front of the person's body.

When lying on the side, is preferable to sleep flat and tilt the bed up (so feet are lower and head is higher) rather than profiling the bed as this causes a sideways bend.



3. Shoulders and arms

If the shoulder underneath the body is not well positioned, the person can end up having a numb and "sleepy" hand. We can accommodate this shoulder by moving the elbow in front of the chest area. Do not let the elbow underneath the body weight.

Some people will find it uncomfortable and painful to rest that arm on the mattress; in that case, you can support their hand to the opposite shoulder.



4. Pelvis, legs and feet

The main aim will be to use pillows to avoid pressure areas and to ensure alignment along the body. Some people will prefer to have:



Both legs bent



The top leg bent and bottom leg stretched

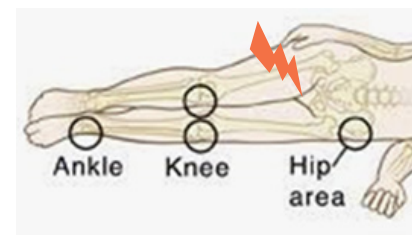


The top leg stretched and bottom leg flexed

A. Both legs bent:

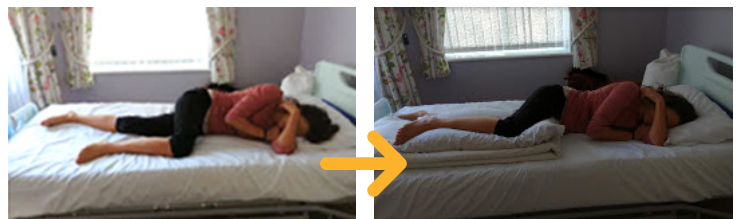
It is important to place a pillow in between both legs to prevent pressure areas in between the knees and ankles.

The pillow will also raise up the top leg. This is necessary, as if the top leg is not parallel to the hip, then it will naturally rest at a lower angle, potentially creating tension and pain in the hip joint.



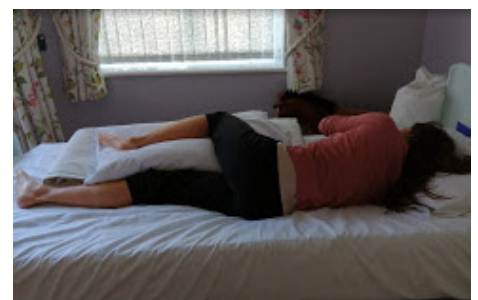
B. The top leg bent and bottom leg stretched:

In this situation, the top leg completely drops to the bed, turning the pelvis and the spine. The hip muscles of the same leg are also in constant stretch. To accommodate these two thick pillows will be necessary to use. These two pillows will be positioned in front of the bottom leg.



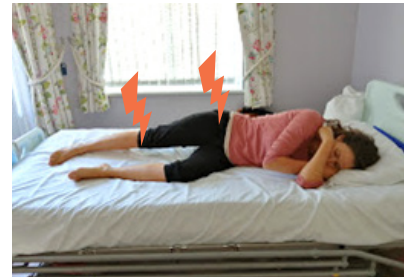
This is how the top leg should look like from behind:

In the picture you can see that both legs are parallel. There is no tension on the hip of the top leg and the pelvis is not rotated.



C. The top leg stretched and bottom leg bent:

Two joints are in a critical position if we do not accommodate the top leg. The hip muscles are constantly stretched; The knee joint is suffering. In addition to that, all the pressure would be held by the ankle.



Two pillows will be necessary to accommodate the top leg. The pillows must be positioned behind the leg resting on the bed. Once both legs are parallel, risks are minimised. Hip, knee and ankle are safe and free of tension and pressure.



Spatial awareness

This is the ability to be aware of oneself in space. It is an organised knowledge of objects in relation to oneself in that given space. It also involves understanding the relationship of these objects when there is a change of position. (Ref - Occupational Therapy for Children)

There is a correlation between having Batten and lack of spatial awareness. This does not mean that everybody living with the disease has a lack of spatial awareness to the same degree.

People with Batten who have a lack of spatial awareness can feel very anxious when they need to transfer or be supported to transfer from one position to another such as from sitting to standing or sitting to lying or from lying on their backs to side-lying. This anxiety is often seen when the person is still able to walk with assistance, showing heightened nervousness when walking on different surfaces or when there are people going past in the opposite direction such as in a corridor or on a pavement.

To enable the person to understand how they can be in control and feel safe, we recommend supporting everyone with Batten to spend time in side-lying from the very early stages, so that they are familiar with this position.

However, there may still be times where lying on the side, even when this is a familiar position, can still be very frightening and cause high levels of anxiety. In this case the first thing to focus on is how the people giving the support are doing it. Sometimes a different person or missing some parts of communicating the process or the way we use touch and physical assistance can cause much fear and anxiety - looking at ourselves and ensuring the same standards and attention from us is the simplest place to start.

If necessary, and ruling out the influence of other people, different positions must be taken into consideration. The supine position (lying on the back) can be an option.

If the bed can be tilted head up this will minimise the risk of reflux and aspiration.



If lying on the back is unsafe, and the person still feels frightened when side-lying, specialised equipment can be used.

We have found that the more the body is in contact with a surface, the less frightened they feel. Therefore, placing cushions, bean bags, or sleep systems against the body (mainly the back), can help the person feel safer and relaxed whilst on this position. Cushions can also be positioned on the abdominal area to prevent the person from lying on their front. This will also give a reassurance and reduce the stress of being in an unsteady position.

These black wedged cushions have Velcro hooks on one side, so when the person is positioned, the cushions do not move.

To be able to use these cushions, a special Velcro sheet is necessary.



Behind the back, there is a bean bag, robust enough to support the person's back even when the person is trying to push against it (something very common in people living with Batten).



Range of movement

People living with Batten do not necessarily suffer from rigid muscles or loss of range of movement. However, over time they do become fully dependent on other people to help them stretch and flex arms and legs, hands and feet every day. There is a tendency for a loss of range in the knee, and often younger children will show a preference for having knees flexed when lying, but exercise and encouraging a full range of movement enables a greater degree of comfort in the longer term. Once the person is unable to purposefully move themselves, our aim is to support the person in a "neutral" position, so not too flexed, not too extended /stretched.

During the night and rest times, it is very important that we support the person in allowing their body system to relax. The body will find it very difficult to relax if it is supported in a stretched position, as it increases the feeling of loss of balance and instability. We need to provide and allow a slight natural flexion, mainly in arms and legs. This slight flexion will provide greater comfort when side-lying. Now, if the body and all the joints are too flexed and the person spends great amounts of time in this flexed position, we are increasing the risk of shortened muscles and loss of range of movement. That is why having regular changes of position is so important.

Tips and advice on how to touch and move someone's body:

- Whenever possible, try to always use both of your hands. It is more comfortable and it helps the person feel safer.
- The carer's hands should hold the joints of the limb that is being moved.
- Avoid grabbing wrists and ankles. It feels better to be held.



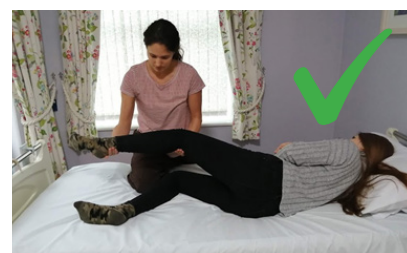
In the picture above, we can see that the person's left arm is stuck under the body. This could cause general discomfort and numbness on the hand. To accommodate this arm, use both hands to hold the elbow and the wrist of the person, then pull the arm away slowly.



Avoid grabbing the wrist and pulling as this will scare the person and may cause pain.



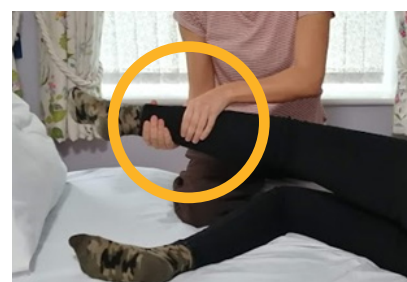
Use flat hands to hold the leg. Both hands should be positioned under the joints: knee and ankle. Using both hands, will spread the limbs weight, making it easier to the carer to move the person's body.



In the picture to the right, the carer is holding the leg allowing the joints to take strain, this can cause the person to feel scared, also the carer's left hand is too close to the person's intimate area.



In the picture to the right, the carer is not supporting the length of the leg, so is placing strain on the upper knee and hip.



This is another comfortable way to hold the leg when we can only use one hand. The knee should be resting on the carer's arm. We normally hold the leg like this when we are placing the pillow in between the legs with the other hand.



Regarding how often to help the person move position (especially at night), every person is different and needs different levels and intensity of support. For instance, there will be people who it might be better to let sleep through the whole night in the same position, because if we support them with personal care or to change position, they will not be able to fall sleep again. On the other hand, other people might feel uncomfortable after 2-3 hours lying in the same position and this might disrupt their sleep, not being able to rest until they change position.

To the person with Batten who is blind and unable to move autonomously, the impact of others can be supportive or frightening depending on the person. Everything we do on a physical level requires an amount of trust and partnership. Demonstrating to the person that we can help them find comfort in their position helps the feeling of being safe and belonging.

For more information, advice or support please contact

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