

NCL Disease and Visual Impairment

BDFA Education Advisor

Deteriorating vision and blindness is sadly an inevitable part of CLN disease. The disease progression results in the deterioration of the nerve tissue that transmits impulses from the retina of the eye to the brain. In addition there is an accumulation of pigmented material in the retina. Visual loss is often the first sign of CLN3 disease but occurs later in the disease progression in the infantile and late infantile CLN diseases. The rate of vision loss varies but is not always obvious – the child adapts to the loss of vision and develops different ways of doing things. Parents sometimes feel guilty that they haven't realised that their child's vision has deteriorated but this gradual adaptation helps the child to adjust. A small area of relatively good vision may remain for a long time and is often used very well – the child may tilt their head or come very close in order to make the best use of their remaining vision. However, as the disease progresses, even light perception is eventually lost.

About 80% of learning takes place through vision and the loss of vision affects every part of the child's life. However, children and young people with CLN disease do have normal vision before the onset of symptoms and can use their 'visual memory' to help them understand and continue to interact with the world around them.

In the UK, local authorities have sensory support services for children who have visual impairment or hearing impairment. The organisation of these services vary across the country with some large authorities having an extensive team supporting children with visual impairment whereas others may only have one or two teachers. Qualified teachers of the visually

impaired (QTVIs) are specialist teachers who have successfully undertaken the mandatory visual impairment training course. The QTVI can:

- Work with parents/carers in the home to develop early learning skills
- Offer support and advice and works collaboratively with parents/carers, schools and other professionals on learning resources, strategies, specialist equipment, multisensory toys and activities and much more
- Provide ongoing assessment of the child's visual functioning
- Provide specialist support for independence and life skills (for example mobility training)
- Provide specialist teaching to develop tactile and auditory skills (for example braille teaching, training in the use of computers and specialist technology such as Braille-notes)
- Provide ongoing support as, the vision deteriorates, helping the child to adapt to the loss of vision and to learn adaptive skills such as braille, touch typing and using their other senses such as touch and hearing
- Offer advice and support during the Early Years, at school entry and at school transfers (for example transferring to a special school or to a secondary school)

Young people with CLN3 disease are often already known to the service for visual impairment. Sometimes though, the need for QTVI involvement with children with Infantile or Late Infantile CLN disease can

be overlooked within the context of severe medical needs. The loss of vision can be seen as 'the least of the child's problems'. Blind children rely heavily on their memory skills and on asking questions but the short term memory loss and deteriorating communication skills in CLN disease make this much more difficult. QTVI support is even more important to children with CLN disease. Some authorities have only termly visits from the QTVI for children in special schools. Parents and carers should talk with the school to make sure that the additional needs arising from CLN disease are recognised and met.

To ensure that their child receives support for their visual impairment, parents/carers can:

- Ask their Health Visitor, Early Years professional, school SENCO or Headteacher to make a referral to the VI Service (the child can also be referred by the Ophthalmologist or Neurologist)
- Ask that their child's visual impairment is carefully and comprehensively described in the Statement or Education, Health and Care Plan together with details of the support needed (for example the amount of time that the QTVI should be working directly with the child, preparing resources, training school staff etc)
- Ask for support with independence and living skills
- Ask for advice in finding ways of stimulating their child in the later stages of the disease and in choosing multisensory toys and activities
- Ask for regular updates from the QTVI, especially when there is an Annual Review or a change of school is proposed

If a child or young person with CLN3 learns braille early on, braille skills often remain good and very useful to the young person well into the later stages of the condition. Sometimes the teaching of braille is not offered as it is felt too difficult for the child but it is important that the teaching of braille is carefully considered. The young person will need intensive support in developing braille skills to overcome concentration and memory difficulties and there are considerable resource implications. Parents and carers should explore the option of braille carefully with those supporting their child. Another tactile 'code', Moon can be used with young people who find braille too difficult. Moon may be easier to learn as it is largely based on letter shapes already known to the child when they had vision.

Children and young people with CLN disease in Early Years provision or school need:

- Individual support to enable the child to be included in all activities in school
- Staff who understand the impact of CLN disease on every aspect of the child's functioning
- Increasing support as skills and abilities decline

Parents and carers can get advice and support regarding visual impairment from:

The BDFA Education Advisor and the Advocacy and Support Partner:
barbaracole@bdfa-uk.org.uk
support@bdfa-uk.org.uk

BlindchildrenUK - www.blindchildrenuk.org
SENSE - www.sense.org.uk
RNIB - www.rnib.org.uk
IPSEA - www.ipsea.org.uk
SOS!SEN - www.sossen.org.uk