

Key educational issues
for children and young
people with Batten
disease including the
impact on memory and
learning.



BATTEN DISEASE FAMILY ASSOCIATION

Together we WILL make a difference

Dementia

Children and young people with Batten Disease:-

- ▶ Will develop childhood dementia, resulting in learning difficulties, unusual behaviours, poor concentration, anger, mood swings, confusion and memory loss. This is in addition to blindness, epilepsy, and increasing physical and communication difficulties and medical needs.
- ▶ Will lose their short term memory skills but their long term memory will stay largely intact
- ▶ Have difficulties in concentrating and are very 'distractable'
- ▶ Will have increasing difficulties in learning and socialisation
- ▶ Are resistant to change and like things to be in a very set routine
- ▶ May be perceived as 'naughty' or challenging because of the impact of the disease and the resulting dementia on their behaviour
- ▶ May not remember where they put their toys or books, or what they did yesterday or even a few minutes ago. This can make children incredibly distressed, frustrated and angry

Supporting children and young people with dementia

- ▶ A multisensory time table at school will help children and young people understand the pattern of their day and to 'orientate' themselves to where they are and what comes next and in transitions from one activity to another
- ▶ A home/school contact book will help parents and carers to know what has happened in school so they can't talk with the child about their day and reinforce memory
- ▶ A predictable learning environment with toys, books and equipment in the same place supports poor short term memory
- ▶ Having familiar adults and peers working with them in a consistent routine gives the child/young person a sense of security especially when they are feeling anxious
- ▶ Using simple language with frequent checks for understanding will support communication
- ▶ Simple activities involving lots of repetition, such as singing favourite songs, looking at family photographs, listening to videos of outings all help to reinforce long term memory
- ▶ Reinforcing the key points at the end of each session and repeating the same information/ideas in different ways helps to overcome short term memory difficulties and strengthen the long term memory

Supporting short term memory loss

- ▶ Children/young people with Batten Disease can still learn-but it may take them longer!
- ▶ Repeat tasks in a slightly different way each time to prevent boredom
- ▶ Play memory games or matching games
- ▶ Take lots of pictures and videos and use them to reinforce memory and to support communication
- ▶ Make talking 'books' on an Ipad or laptop so they can look back on their week in school with family and friends at weekends and during holidays
- ▶ Ensure parents know the topics you are covering at school
- ▶ Use a home/school diary
- ▶ Build up routines in in school and try to keep these routines as consistent as possible
- ▶ Prepare the child/young person for changes of staff or classroom

Communication

- ▶ Some children with infantile or late infantile disease may not develop spoken language and may be learning Makaton before the diagnosis of Batten Disease
- ▶ Difficulties in making themselves understood can cause frustration and anxiety for children and young people
- ▶ Support from the speech and Language Therapist is of vital importance in maintaining spoken language for as long as possible and in preparing for the time when speech becomes difficult to understand
- ▶ Augmented Communication (I pads, switches etc) can be used
- ▶ On body signing and Makaton should be used as soon as possible to support understanding and to encourage communication
- ▶ Objects of Reference can be used along side visual and tactile timetables
- ▶ Music cues can be helpful for transitions from one activity to the next



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Personalised learning

- ▶ Batten Disease is a neurodegenerative and life-limiting condition
- ▶ Children and young people with Batten Disease need challenge and stimulation together with very individualised support and learning programmes.
- ▶ Their skills are declining whilst their peers are progressing
- ▶ They are aware of their declining skills and are very sensitive to feeling 'different'
- ▶ They need a personalised programme that includes opportunities for both individual and group learning and focuses on activities that are enjoyable and achievable
- ▶ Routine and consistency are helpful but there also needs to be flexibility on those 'not so good' days
- ▶ The best possible quality of life is of the utmost importance

Equipment and access

- ▶ Having the right equipment at the right time is vital. Walkers, adapted trikes and standing frames all help to maintain physical skills and independence but need to be ordered in good time
- ▶ Staff need support and training in safe lifting and handling
- ▶ Maximising the accessibility of school buildings
- ▶ A classroom layout that maximises access to learning and social opportunities for the child/young person in a walker or wheelchair
- ▶ Opportunities for changing position-chair, bean bag, 'peapod', quiet area in classroom
- ▶ Regular assessment by physiotherapist/occupational therapist

Therapy support

- ▶ All children/young people with Batten disease need access to a range of therapies within their school curriculum.
- ▶ Hydrotherapy
- ▶ Physiotherapy
- ▶ Occupational Therapy
- ▶ Speech and Language Therapy
- ▶ All of these therapies are important as they can enable children to be as independent as possible for as long as possible.
- ▶ Sometimes a piece of equipment such as a walker or a standing frame is 'dismissed' if the child doesn't like it after the first few times of using it. We need to remember these children have never had to use equipment to support them to walk. They aren't going to like it but with time realise that the equipment helps them to get back some independence!

Issues around behaviour and social interaction

- ▶ Children and young people with Batten Disease:-
- ▶ Often have a high level of anxiety, cry uncontrollably,
- ▶ May have obsessive, repetitive or impulsive behaviours that isolate them from their peers
- ▶ Are easily disorientated, confused and frustrated and have 'meltdowns'
- ▶ May have episodes of aggressive behaviours, swearing, taking clothes off
- ▶ May be experiencing hallucinations or believe that they have heard or done things that never happened (confabulation)
- ▶ Have a limited range of interests and often repeat the same things over and over again



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Support for behavioural issues

- ▶ Ask parents and carers what works at home.
- ▶ Parents can be overwhelmed by daily reports of negative behaviours-balance this with positive reports.
- ▶ 'Meltdowns' can result when the child is overwhelmed and are distressing for the child/young person, those working with them and parents may be asked to take the child/young person home
- ▶ Careful recording and assessment can help to identify 'triggers' of negative behaviours.
- ▶ Careful planning of the days activities and the use of a tactile timetable and objects of reference can help to provide an easily understandable 'outline' of the school day and help with the often difficult transition from one activity to another.
- ▶ Having a 'safe' space, within the classroom or nearby, provides a familiar place in which to calm down, re-orientate themselves if they have become confused and to have a period of familiar, quiet activities before joining the class again.
- ▶ The dementia and 'brain disorganisation' can result in impulsive and aggressive behaviour that the child cannot control or remember afterwards. Distraction and refocusing on a favourite activity often works better than trying to reason with the child. Star charts and 'reward' systems may help but often the child cannot consistently control their behaviour. The child/young person can become even more anxious, frustrated and angry if those around them are not able to understand and allow for the impact of the disease on behaviour.

EHC Plans

- ▶ Importance-this is a legal document that sets out the child/young person's special education, health and social care needs and the provision needed to meet those needs. They should be 'specified' and 'quantified' with details of the support needed. They are 'needs led' and not 'resource led'. There should be a recognition that maintaining the best possible quality of life is a very important and relevant outcome.
- ▶ Speech and language therapy, physiotherapy and occupational therapy should be identified within the educational needs section-the disease results in loss of communication and fine and gross motor skills and there must be regular assessment and monitoring and provision of direct services as the condition deteriorates. These therapies are needed to enable the child/young person to continue to access educational and social interaction opportunities alongside their peers.
- ▶ The condition results in blindness and it is essential that support from a qualified teacher of the visually impaired is identified and provided.
- ▶ Hydrotherapy and music therapy are of great benefit to children/young people with Batten Disease but are often not mentioned in the EHCP
- ▶ Support at home, equipment, healthcare, medication, tube feeding, suctioning and any other need and the provision necessary to meet those needs must be specified and quantified in the EHCP.
- ▶ Careful thought should be given to EHCP meetings and Reviews-the child/young person is deteriorating, not progressing. Although the deterioration needs to be addressed and planned for, there also needs to be a strong focus on positive issues. Parents can be overwhelmed by the sheer number of people attending. It helps if there is someone there supporting parents and that only those professionals who need to be there are invited. EHCP reviews may need to be termly or 6 monthly as deterioration can be rapid and unpredictable.
- ▶ Parents may need support in preparing their parental contribution for review meetings. Reports need to be sent to them in advance so that they have time to read them and put together a list of questions that they may want to ask at the meeting.
- ▶ Reports from professionals are really important in updating information about the child/young person as the condition is a deteriorating one. Although progression through the P levels or National Curriculum levels are important measures of successful intervention, this is not appropriate for children/young people with a deteriorating condition. It is important that there are realistic and appropriate goals set and a real appreciation of the effort by the child and those working with him/her in maintaining skills for as long as possible.



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Supporting Parents

- ▶ Open and honest Communication with regular meetings.
- ▶ Sending home anything the child makes or does at school.
- ▶ Taking lots of photos and videos.
- ▶ Writing fully what the child did at school in the home school diary.
- ▶ Be factual but include positive information too.
- ▶ Do as much research about Batten disease as you can. The BDFFA can support you in this.
- ▶ Fundraise.
- ▶ Understand that parents are trying to make every day count and to make as many happy memories as possible. There may be special trips or holidays that may be in term time. Although regular attendance is very important, some flexibility is very important to children, siblings and parents.

Further sessions

- ▶ There will be a more detailed session on the impact of visual impairment on children and young people with Batten Disease.
- ▶ There will be a session on Epilepsy and motor disorders and Batten Disease
- ▶ There will be a 'Sharing Good Practice' session this afternoon

Thank you!

