

Batten Disease

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Batten disease is the most common type of a group of 4 disorders called neuronal ceroid lipofuscinosis or NCL's. It is rare occurring in 2-4 of every 100 000 births. The disorder has been found world wide however it appears to be more common in Finland, Sweden, Northern Europe, Newfoundland and Canada. Batten disease is usually regarded as the juvenile form of NCL, however this is the name now used to describe all forms of NCL. Two of the four disorders present themselves quite early in childhood and these are known as Infantile NCL (Santavuori-Haltia Disease) and Late Infantile NCL (Jansky-Bielschowsky Disease). Juvenile NCL and Adult NCL present later in life, have the same symptoms and are similar, however all four present at different ages and progress at different rates.

The symptoms of Batten disease are caused by a build-up of substances called lipopigments. These are made up of fats and proteins. In people who have Batten disease the build up of these fats and proteins can be found in the cells of the brain, eyes, skin, muscle and other tissues. These lipopigment build-ups are thought to be caused from a lack of enzymes that usually breakdown the lipopigments.

The Four types of Batten Disease

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| Infantile - | Early signs show at between 6 months and 2 years of age and children may show signs of microcephaly (small head), failure to thrive, delayed development with progressive deterioration, myoclonic jerks (short, sharp muscle contractions) seizures and other motor disorders. This is the most rapidly progressive of the four types. |
| Late Infantile - | Symptoms will become apparent at between 2 years and 4 years of age and include loss of muscle co-ordination (ataxia), seizures and progressive mental deterioration. |
| Juvenile - | Between 5 and 8 is when the typical symptoms of this type appear and present as progressive vision loss, seizures & ataxia (loss of muscular co-ordination). This form progresses less rapidly. |
| Adult - | Usually occurs before the age of 40. The symptoms progress more slowly and are milder. |

There is no known treatment to stop or reverse the effects of Batten disease, however some children who experience seizures may take medication/anticonvulsant drugs to reduce or control these.

Adapted from Batten Disease Support and Research Association: 2003

Implications for Inclusion

There may be some health implications for children with Batten Disease such as the seizures they may experience. It would be appropriate for the service to develop a policy and health management plan.

Please refer to:

Community Link Australia Pty Ltd (2003)

Risk Management Guidelines. CARATA: Brisbane.

To support the inclusion of a child with Batten disease, services can offer an inclusive program that is aimed at meeting all the children's needs within it. By observing the children within the context in different situations teachers can provide appropriate activities and games to include all children and to enhance all developmental areas.

Reference:

www.bdsra.org/batten.htm ***Batten Disease Support and Research Association:2003, accessed 12/12/2003.***

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