There are four additional diseases included in the Batten Disease/NCL group:

Finnish Late Infantile
Variant Late Infantile
Turkish Late Infantile
Northern Epilepsy/ERMP

How are NCLs inherited?

Childhood NCLs are autosomal recessive disorders; that is, they occur only when a child inherits two copies of the defective gene, one from each parent. When both parents carry one defective gene, each of their children faces one in four chance of developing NCL. At the same time, each child also faces a one in two chance of inheriting just one copy of the defective gene. Individuals who have only one defective gene are known as carriers, meaning they do not develop the disease, but they can pass the gene on to their own children.

Adult NCL may be inherited as an autosomal recessive (Kufs) or, less often, as an autosomal dominant (Parrys) disorder . In autosomal dominant inheritance, all people who inherit a single copy of the disease gene develop the disease. As a result, there are no unaffected carriers of the gene.

How many people have these disorders?

Batten Disease/NCL is relatively rare, occurring in an estimated 2 to 4 of every 100,000 births in the United States. The diseases have been identified worldwide. Although NCLs are classified as rare diseases, they often strike more than one person in families that carry the defective gene.

Batten Disease takes the lives of about 1000 children a year in the United States alone.

Batten Disease has many beautiful faces….

Bryce Butenhoff
Born 10/17/1999
Late Infantile NCL
Lived in Mora, Minnesota
Bryce lost his fight on,
Monday, April 7, 2008.

Sarah Inks
Born in July 7, 1994
Juvenile
lives in Bellevue, Nebraska

Nick's Battle Foundation, Inc

What is Batten Disease??

www.nicksbattle.org

To see more about Nicholas log on to

www.nicksbattle.com

Nicholas Heuchan
Born 12/18/1996
Juvenile
Lives in Baltimore, Maryland

Nick's Battle Foundation was formed in honor of Nicholas Heuchan. We formed to educate the public about Batten Disease and to raise funds for research and financial assistance for those who may need it.

Nicholas is a bright boy who loves to play video games, swim, and ride his bike. He likes fast cars and drag races. He loves being a part of his sisters pit crew and racing. He likes playing with his dog, Jake. Nick loves all the things every other boy his age likes to do. However, Nicholas is different, he has Juvenile Batten Disease. His life will change very fast.

What is Batten Disease?

Batten Disease is named after the British pediatrician who first described it in 1903. Also known as Spielmeyer-Vogt-Sjogren-Batten Disease, it is the most common form of a group of disorders called Neuronal Ceroid Lipofuscinoses (or NCLs). Although Batten Disease is usually regarded as the juvenile form of NCL. The forms of NCL are classified by age of onset have the same basic cause, progression and outcome but are all genetically different.

Over time, affected children suffer mental impairment, worsening seizures, and progressive loss of sight and motor skills. Eventually, children with Batten Disease/NCL become blind, bedridden, and unable to communicate and presently is always fatal.

Batten Disease is not contagious or, at this time, preventable.

What are the forms of NCL/Batten Disease? There are four main types of NCL, including two forms that begin earlier in childhood and a very rare form that strikes adults. The symptoms are similar but they become apparent at different ages and progress at different rates.

Infantile NCL, begins between about 6 months and 2 years of age and progresses rapidly. Affected children fail to thrive and have abnormally small heads (microcephaly). Also typical are short, sharp muscle contractions called myoclonic jerks. Initial signs of this disorder include delayed psychomotor development with progressive deterioration, other motor disorders, or seizures. The infantile form has the most rapid progression and children live into their mid childhood years.

Late Infantile NCL, begins between ages 2 and 4. The typical early signs are loss of muscle coordination (ataxia) and seizures along with progressive mental deterioration.. This form progresses rapidly and ends in death between ages 8 and 12.

Juvenile NCL (Batten Disease) begins between the ages of 5 and 8 years of age. The typical early signs are progressive vision loss, seizures, ataxia or clumsiness. This form progresses less rapidly and ends in death in the late teens or early 20s, although some may live into their 30s.

Adult NCL (Kufs Disease or Parry's Disease) generally begins before the age of 40, causes milder symptoms that progress slowly, and does not cause blindness. Although age of death is variable among affected individuals, this form does shorten life expectancy.