

# Who WE ARE



## OUR MISSION

Located in Central Ohio, BDSRA is determined to provide unwavering support, fund progressive research, and be a source of steadfast advocacy for all CLN types. Founded in 1987, Batten Disease Support, Research, & Advocacy Foundation (BDSRA) is the largest international nonprofit organization in North America for Batten disease. Our long-term vision is a world without Batten disease.

**BATTEN DISEASE** is a rare, fatal inherited disorder of the nervous system and has no known cure. Those affected with Batten disease often suffer progressive neurological impairment, seizures, blindness, and loss of speech and motor skills. Batten Disease, or Neuronal Ceroid Lipofuscinosis (NCL), is a family of lysosomal storage disorders which cause the build-up of fats and proteins (lipofuscins) in the body, causing cell death.

There are 13 identified forms of the disease (CLNs), which are most commonly diagnosed between infancy and school age. Kufs, Parry, and ANCL disease are known adult forms of Batten.

Though recent improvements in genetic testing have made diagnosing Batten disease much quicker and more reliable, many families experience long diagnostic journeys. Autism, seizure disorder, epilepsy, Pervasive Developmental Disorders, and others are common among roughly 30 early misdiagnoses. According to the Centers for Disease Control, 2-4 births per 100,000 in the U.S. are affected by Batten disease, though some researchers in the field suggest these numbers are low.



**Ohio families have lost two children this year, while many others cope with the Batten disease diagnosis, management of care, and a shared vision to find treatments and cures.**



Did you know?  
International Batten Disease Day is June 9 annually.



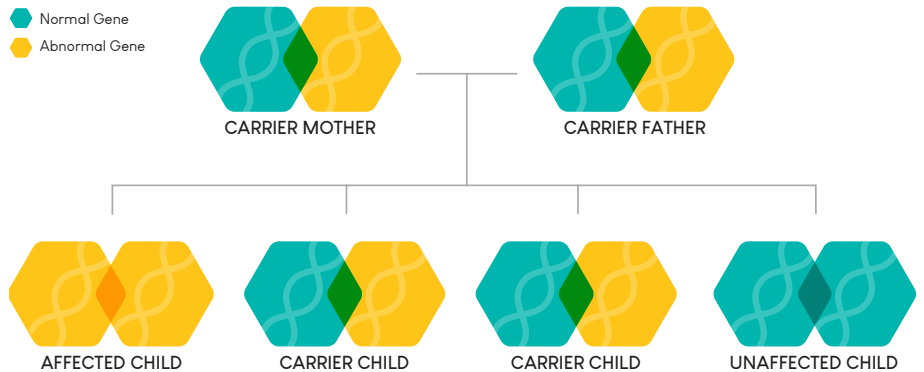
To learn more about Batten disease, visit:





# The Genetics of BATTEN DISEASE

Batten disease, for the most part, is an autosomal recessive disease. Autosomal recessive diseases require two copies of an abnormal gene to be passed down in order for the disease to develop. This means that both parents must be carriers in order to have the possibility of their child inheriting a recessive condition. The figure below illustrates the different inheritance possibilities.



When two carriers have children, **at each pregnancy** there is a one in four chance that the child will be born with autosomal recessive Batten disease.

If the child receives both abnormal genes

**25%**

chance this child is affected with the disease

If the child gets a normal gene from one parent and an abnormal gene from the other parent

**50%**

chance this child is not affected but is a carrier of the disease

If the child receives both normal genes

**25%**

Chance this child is not affected and is not a carrier

## What Can You DO TO HELP?

- Support H.B. 177, which would limit unexpected and unaffordable pharmaceutical costs for Batten families once their copay assistance is exhausted.
- Support H.B. 130, which would cut the red tape – ensuring that Batten patients get the care they need, when they need it.
- Support H.B. 24, which would require Ohio health plans and Medicaid to cover biomarker testing for the diagnosis, treatment, appropriate management, and ongoing monitoring of a disease or condition like Batten disease.
- Subscribe to our monthly newsletter, *The Illuminator*.

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