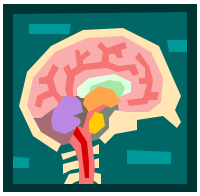

Disability Awareness Begins With You:

Alper's Disease

What is Alpers' Disease?
Alpers' disease is a rare, genetically determined disease of the brain that causes progressive degeneration of grey matter in the cerebrum. The first sign of the disease usually begins early in life with convulsions. Other symptoms are developmental delay, progressive mental retardation, hypotonia (low muscle tone), spasticity (stiffness of the limbs), dementia, and liver conditions such as jaundice and cirrhosis that can lead to liver failure. Optic atrophy may also occur, often causing blindness.

Researchers believe that Alpers' disease is caused by an underlying metabolic defect. Some patients have mutations in mitochondrial DNA. Researchers suspect that Alpers' disease is sometimes misdiagnosed as childhood jaundice or liver failure, since the only method of making a definitive diagnosis is by autopsy or brain biopsy after death.

Is there any treatment?
There is no cure for Alpers' disease and no way to slow its progression. Treatment is symptomatic and supportive. Anticonvulsants may be used to treat the seizures. Valproate should be used with caution since it can increase the risk of liver failure. Physical therapy may help to relieve spasticity and maintain or increase muscle tone.

What is the prognosis?
The prognosis for individuals with Alpers' disease is poor. Those with the disease usually die within their first decade of life. Continuous, unrelenting seizures often lead to death. Liver failure and cardiorespiratory failure may also occur.

What research is being done?
The NINDS supports research on gene-linked neurodegenerative disorders such as Alpers' disease. The goals of this research are to increase scientific understanding of these disorders, and to find ways to prevent, treat, and cure them.

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