Alpers' Disease

Alpers' disease is a rare, progressive neurodegenerative disease of the brain. It affects infants and children.

CAUSES

It is a disorder passed down from parents (*genetic*). It is sometimes seen in siblings. Some researchers believe that Alpers' disease may be caused by an underlying metabolic defect.

SIGNS

- The first signs of the disease usually happen in infancy. They include:
  - Seizures.
  - Failure to meet developmental milestones.

SYMPTOMS

Primary problems (*symptoms*) of the disease include:

- Developmental delay.
- Mental retardation.
- Poor muscle tone (*hypotonia*).
- Stiffness of the limbs (*spasticity*).
- Confusion (*dementia*).
- Seizures. These may include a type of seizure (*epilepsia partialis continua*) that consists of repeated muscle (*myoclonic*) jerks.
- Eye problems can lead to blindness.
- Many patients can have liver failure.

TREATMENT

There is no cure for Alpers' disease. Currently, there is no way to slow its progression. Treatment consists of supportive therapies that help with the symptoms of the disease.

- Anticonvulsants may be used to treat the seizures.
- Physical therapy may help to:
  - Relieve spasticity.
  - Maintain or increase muscle tone.

The outcome (*prognosis*) for individuals with Alpers' disease is poor. The lifespan for those diagnosed with Alper's disease is about 10 years. Liver failure is usually the cause of death. Cardiorespiratory failure may also occur.

RESEARCH BEING DONE

Research is continually being done to learn more about Alper's disease. The National Institute of Neurological Disorders and Stroke (NINDS) supports research on genetic *neurodegenerativ*e disorders such as Alpers' disease.

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