Canavan Disease

Canavan disease is a rare, inherited, neurological disorder. It is characterized by spongy degeneration of the brain. This means the white matter of the brain is replaced by microscopic fluid-filled spaces.Canavan disease may occur in any ethnic group, but it tends to affect persons of Eastern European Jewish ancestry more often.

CAUSES

It is caused by a deficiency of an enzyme called aspartoacylase. Canavan disease is one of a group of genetic disorders called the leukodystrophies. They affect growth of the myelin sheath of the nerve fibers in the brain. The myelin sheath is the fatty covering surrounding nerve cells. It acts as an insulator.

SYMPTOMS

Symptoms of Canavan disease appear in early infancy and progress rapidly. They may include:

- Mental retardation.
- Loss of previously acquired movement (*motor*) skills.
- Feeding difficulties.
- Abnormal muscle tone (i.e., floppiness or stiffness).
- Poor head control.
- Abnormally enlarged head (*megalocephaly*).
- Paralysis.
- Blindness.
- Hearing loss.

TREATMENT

There is no cure for Canavan disease. There is no standard course of treatment either. Treatment is symptomatic and supportive.

The likely outcome (*prognosis*) for this disease is poor. Death usually happens before age 10.

RESEARCH BEING DONE

The NINDS supports research on genetic disorders including leukodystrophies such as Canavan disease. The goals of this research include finding ways to prevent, treat, and cure these disorders.

Research is continually being done to learn more about this problem.

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