Krabbe Disease

Krabbé disease is a rare, degenerative disorder of the central and peripheral nervous systems. It is one of a group of genetic disorders called the *leukodystrophies* that affect the growth of the myelin sheath, the fatty covering-which acts as an insulator-on nerve fibers in the brain. Symptoms vary in prevalence and severity among patients and may include loss of previously attained developmental skills, unexplained fevers, irritability, myoclonic seizures (sudden, shock-like contractions of the limbs), blindness, spasticity (stiffness of the limbs), and paralysis. Prolonged weight loss may occur also. Onset of the disorder generally occurs at 3 to 6 months of age, but juvenile and even adult-onset patients are known.

TREATMENT

Although there is no cure for Krabbé disease, bone marrow transplantation has been shown to have benefit for mild cases early in the course of the disease. Generally, treatment for the disorder is symptomatic and supportive. Physical therapy may help maintain or increase muscle tone and circulation.

WHAT WILL HAPPEN

The prognosis for individuals with Krabbé disease is poor. The disorder is generally fatal before age 2. Prognosis may be significantly better with early bone marrow transplantation in some patients.

RESEARCH BEING DONE

The NINDS supports research on genetic disorders including leukodystrophies such as Krabbé disease. The goals of this research are to increase scientific understanding of these disorders, and to find ways to prevent, treat, and cure them.

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

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