Alexander Disease

National Institute of Neurological Disorders and Stroke (NINDS)

Source

National Institute of Neurological Disorders and Stroke (NINDS). Alexander Disease Information Page.

Alexander disease is one of a group of neurological conditions known as the leukodystrophies. Leukodystrophies are disorders that result from abnormalities in myelin, the “white matter” that protects nerve fibers in the brain. In Alexander disease, the destruction of white matter is accompanied by the formation of Rosenthal fibers--abnormal clumps of protein that accumulate in non-nerve cells (astrocytes) in the brain. The most common type of Alexander disease is the infantile form that usually begins during the first two years of life. Symptoms include mental and physical developmental delays, followed by the loss of developmental milestones, an abnormal increase in head size, and seizures. The juvenile form of Alexander disease has an onset between the ages of two and thirteen years. These children may have excessive vomiting, difficulty swallowing and speaking, poor coordination, and loss of motor control. Adult-onset forms of Alexander disease are less common. The symptoms sometimes mimic those of Parkinson’s disease or multiple sclerosis, or may present primarily as a psychiatric disorder. The disease occurs in both males and females, and there are no ethnic, racial, geographic, or cultural/economic differences in its distribution. Alexander disease is a progressive and often fatal disease.