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Gerstmann-Straussler-Scheinker Disease

National Institute of Neurological Disorders and Stroke (NINDS)

Source

National Institute of Neurological Disorders and Stroke (NINDS). Gerstmann-Straussler-Scheinker Disease Information Page.

Gerstmann-Straussler-Scheinker disease (GSS) is an extremely rare, neurodegenerative brain disorder. It is almost always inherited and is found in only a few families around the world. Onset of the disease usually occurs between the ages of 35 and 55. In the early stages, patients may experience varying levels of ataxia (lack of muscle coordination), including clumsiness, unsteadiness, and difficulty walking. As the disease progresses, the ataxia becomes more pronounced and most patients develop dementia. Other symptoms may include dysarthria (slurring of speech), nystagmus (involuntary movements of the eyes), spasticity (rigid muscle tone), and visual disturbances, sometimes leading to blindness. Deafness also can occur. In some families, parkinsonian features are present. GSS belongs to a family of human and animal diseases known as the *transmissible spongiform encephalopathies* (TSEs). Other TSEs include Creutzfeldt-Jakob disease, kuru, and fatal familial insomnia.