

[Open Peer Review on Qeios](#)

# Gerstmann-Straussler-Scheinker Disease

National Cancer Institute

## Source

National Cancer Institute. *Gerstmann-Straussler-Scheinker Disease*. NCI Thesaurus. Code C84727.

A very rare and fatal disorder of spongiform encephalopathy usually caused by mutations of the prion protein (PRNP) gene. It is characterized by the accumulation of amyloid in the brain. Signs and symptoms include lack of motor coordination, unsteady gait, and difficulty walking. As the disease progresses, patients develop speech difficulties and dementia.