Editorial

iMedPub Journals
www.imedpub.com

Insights in Biomedicine
ISSN 2572-5610

2021
Vol.6 No.12:57

Editorial on Gerstmann Straussler Scheinker Syndrome

Chih-Chang Chu*

Department of Medicine, Florida State University, USA

Received: December 20, 2021; **Accepted:** December 25, 2021; **Published:** December 30, 2021

***Corresponding author:**
Dr. Chih-Chang Chu

cc62@cornell.edu

Department of Medicine, Florida State University, USA.

Citation: Chu CC (2021) Editorial on Gerstmann Straussler Scheinker Syndrome. Insights Biomed Vol.6 No.12: 57

Editorial

Ataxia and dementia are symptoms of Gerstmann-Straussler-Scheinker disease, a neurodegenerative brain condition. It is virtually always inherited and just a few families worldwide have it. Between the ages of 35 and 55, the condition frequently manifests itself. The transmissible spongiform encephalopathies (TSEs) or prion diseases are a group of human and animal diseases known as Gerstmann-Straussler-Scheinker disease. Creutzfeldt-Jakob disease, kuru, and fatal familial insomnia are all TSEs.

GSS (Gerstmann–Sträussler–Scheinker syndrome) is an extremely rare, generally familial, deadly neurodegenerative disease that affects people aged 20 to 60. It is only passed down via families and is present in only a few families around the world. Due to the causal involvement of PRNP, the human prion protein, it is categorised with the transmissible spongiform encephalopathies (TSE). The Austrian physicians Josef Gerstmann, Ernst Sträussler, and Ilya Scheinker were the first to disclose GSS in 1936. Autosomal-dominant inheritance is linked to familial cases. Progressive ataxia, pyramidal indications, and even adult-onset dementia are all hallmarks of GSS, and they get worse as the disease advances.

Dysarthria (difficulty speaking) and cerebellar truncal ataxia (unsteadiness) appear first, followed by progressive dementia. The initial indication of GSS can be memory loss. Extrapyramidal and pyramidal symptoms and signs may appear, and the condition may first resemble spinocerebellar ataxias. In contrast to Creutzfeldt–Jakob disease, myoclonus (spasmodic muscular contraction) is less common. Nystagmus (involuntary eye movement), visual abnormalities, and even blindness or deafness are common in sufferers. GSS causes widespread deposition of amyloid plaques made up of improperly folded prion protein, according to neuropathological results.

GSS illness, also known as Gerstmann-Sträussler-Scheinker disease, is a rare inherited degenerative brain ailment. Symptoms, condition progression, and overall severity might vary significantly among affected families and individuals. Even within the same family, this is the case. A progressive loss of coordination, which can manifest as unsteadiness of gait, difficulty walking, and clumsiness, is a common sign. Other symptoms emerge as the disease advances, such as dementia, which is characterised by growing issues with thought, cognition, memory, language, and behaviour. GSS is caused by an aberrant version of the prion protein (PRPN) gene in all cases.

The human prion protein is encoded by the PRNP gene (PrPc). Changes in this gene cause improperly shaped (misfolded) prion protein (PrPSc), also known as a prion, to be produced, which is damaging to the body. The aberrant prions accumulate largely in the brain in GSS. These results in the loss of nerve cells (neurons) over time, as well as the many symptoms linked with this condition. People in their 40s and 50s are the most commonly affected. Although there is no cure, researchers are looking for the best strategies to treat and control GSS.

