Gerstmann-Straussler-Scheinker Disease

Gerstmann-Straussler-Scheinker disease (GSS) is a rare, neurodegenerative brain disorder. It is almost always inherited. It is found in only a few families around the world. GSS belongs to a family of human and animal diseases known as the *transmissible spongiform encephalopathies* (TSEs). Other TSEs include:

- Creutzfeldt-Jakob disease.
- Kuru.
- Fatal familial insomnia.

SYMPTOMS

Onset of the disease usually happens between the ages of 35 and 55.

- In the early stages, patients may experience varying levels of ataxia. This is a lack of muscle coordination. It includes:
  - Clumsiness.
  - Unsteadiness.
  - Difficulty walking.

As the disease progresses:

- The ataxia becomes more pronounced.
- Most patients develop dementia.

Other problems (*symptoms*) may include:

- Slurred speech (*dysarthria*).
- Involuntary movements of the eyes (*nystagmus*).
- Rigid muscle tone (*spasticity*).
- Visual disturbances. These sometimes lead to blindness.
- Deafness.
- In some families, features of Parkinson`s are present.

TREATMENT

There is no cure for GSS. There are no known treatments to slow progression of the disease. Current therapies are aimed at:

- Reducing symptoms.
- Making the patient as comfortable as possible.

GSS is a slowly progressive condition. Usually it lasts from 2 to 10 years. The disease ultimately causes severe disability and finally death. This is often after the patient goes into a coma or has a secondary infection such as aspiration pneumonia due to an impaired ability to swallow.

RESEARCH BEING DONE

The NINDS supports and conducts research on TSEs, including GSS. Much of this research is aimed at:

- Locating agents that cause these disorders.
- Clarifying the mechanisms underlying them.
- Finding ways to prevent, treat, and cure them.

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

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