Familial Creutzfeldt-Jakob Disease

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Prions:

1. Normal prion (PrP-N) is not infectious, while variant prion (PrP-V) is.

2. A person can induce normal prion to change into variant prion, making it infectious.

3. PrPC and PrP-V are different proteins, and only prion diseases are transmissible.

Familial Creutzfeldt-Jakob Disease is a rare familial prion disease that attacks the nervous system. It is caused by a 

**Introduciton:**

**Prion Diseases:** Familial Creutzfeldt-Jakob Disease is a familial human prion disease that attacks the nervous system, and usually affects patients about 60 years old (Baron). It is caused by prions (proteinaceous infectious particles). Prions are infectious proteins with a normal structure that convert to a normal form of prions into the healthy brain (Heppner). Prions are unique in that they can be inherited, transmitted, or manifest sporadically. Other prion diseases include variant and sporadic CJD, kuru, fatal familial insomnia (FFI), and Gerstmann-Straussler-Scheinker syndrome (GSS). Prions can be transmitted through a blood transfusion, a transfusion, or by direct contact with infected tissue or fluid. Prion diseases are unique in that they can be inherited, transmitted, or manifest sporadically.

**Symptoms:**

1. Early symptoms:
   - Vision and sleep impairment
   - Difficulty thinking or remembering
   - Behavioral changes
   - Headache
   - Malaise

2. Later Symptoms:
   - These rapidly lead to more severe later symptoms, such as loss of muscular coordination (ataxia), uncontrollable movements (myoclonus), and dementia. At the later stages of disease, patients may experience hallucinations and memory loss. In the later stages of the disease, patients may not recognize family members or others. 

3. **Support:**

   - The following organizations provide answers and support for patients and families coping with CJD, and also information for the general public:
     - The CJD Foundation
     - The CJD Support Network
     - The Brain and Spine Foundation

   - **References:**