Prion Disease

A rare, rapidly progressive and always fatal neurological disease.

Sporadic

- Sporadic CJD (sCJD) has no known cause and occurs randomly with a lifetime risk of 1:6,000.
- sCJD accounts for approximately 85%-90% of all cases of prion disease.
- Atypical forms of sCJD account for less than 1% of all sporadic cases. These include Variably Protease Sensitive Prionopathy (VPSPr) and Sporadic Fatal Insomnia (sFI).

Genetic

- Genetic prion disease accounts for approximately 10-15% of all prion disease cases.
- Genetic prion disease includes Genetic CJD (gCJD), Gerstmann–Sträussler–Scheinker disease (GSS) and Fatal Familial Insomnia (FFI).
- DNA can be tested to identify if an individual carries a mutation for prion disease.
- The vast majority have an autosomal dominant pattern - if one parent carries a mutation for prion disease there is a 50-50 chance for each child to inherit the gene.

Acquired

- Acquired prion disease is the rarest cause representing less than 1% of all prion diseases.
- Iatrogenic or medically acquired CJD from contaminated surgical instruments, corneal transplant, dura mater graft, and human pituitary hormone treatment for infertility and short statue.
- Variant CJD (vCJD) acquired from exposure to BSE through consumption of contaminated beef or blood plasma transfusion.

CJDISA
INTERNATIONAL SUPPORT ALLIANCE

Providing support for patients and their families affected by prion disease globally.

Visit our website for more information: **www.cjdisa.com**