## **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).

## Acquired

- Acquired prion disease is the rarest cause representing less than 1% of all prion diseases.
- latrogenic 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.

## 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.



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

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