Appendix A

Human Prion Diseases

FORM	CAUSE	DISTINGUISHING FEATURES
Sporadic CJD (sCJD)	Unknown	Affects mainly people over the age of 60. Common symptoms include ataxia and dementia. Short course. Upon tissue examination there is spongiform change, but plaques are rarely present.
Sporadic Fatal Insomnia (sFI)	Unknown	Has clinical and histopathologic features indistinguishable from those of FFI, but does not have the mutation on the prion gene that characterizes FFI.[2]
Inherited Prion Diseases - Familial CJD (fCJD) - Gerstmann-Straussler-Scheinker Disease (GSS) - Fatal Familial Insomnia (FFI)	Inherited mutation in PrP gene	Often younger onset than sCJD. Symptom pattern depends on type of mutation, but can be similar to sporadic. Course of illness is usually longer.
Acquired CJD -Iatrogenic CJD (iCJD)	Contamination through brain surgery, corneal transplant, dura mater graft or growth hormone	Age at onset depends on the age at exposure and on the incubation time. Clinical and pathological features are often indistinguishable from sCJD. Growth hormone cases show plaques.
-Variant CJD (vCJD)	Exposure to BSE through consumption of infected beef or blood or plasma transfusion	With exposure to BSE, younger onset and longer duration than sporadic CJD. Psychiatric symptoms often seen at disease presentation. Distinctive "daisy" plaques upon tissue examination. For blood or plasma transfusion, age at onset depends on the time of exposure.

Taken from CJD foundation CJD and other Prion Diseases 2009