FAMILIAL FATAL INSOMNIA

1. **Description**
   first described in 1986

   “FFI is a progressive disorder that **begins with difficulty in initiating sleep and leads to total lack of sleep** within a few months and, later, to spontaneous lapses from quiet wakefulness into a sleep state with enacted dreams.” (ICSD, 2001)

   Course is one of a **relentless worsening of Sxs**, especially of **autonomic dysfunctions** (e.g. normal circadian rhythms of body temp, HR, respiration, hormone cycles are lost; elevated levels of cortisol and catecholamines; “zero” ACTH)

   **Is always fatal**, usually within 7-13 months after onset of Sxs

   Usually Sxs begin in 50s or 60s, or at least middle age on, M = F

   Associated with degeneration of **anterior & dorsomedial thalamus**
   Cerebral cortex (esp. frontal lobes) & **cerebellum** may also be affected

2. **Causes**
   a. suspected genetic origin, **autosomal dominant**!
      family pedigrees show up to 3 generations of affected Ss

   b. many involve an inherited or “spontaneous/sporadic” form of a brain-wasting disease based on a **prion disease** (deformed, defective protein)

   c. at this point prions causing FFI are **not** thought to be from contaminated meat/meat products