

# 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