Temporal pattern for patients worsening

- **Onset Neurologic Symptom (26.2)**
  - 10.4 years (0.34)
  - 100%

- **Diagnosis (36.6)**
  - 17.3 years (0.32)
  - 100%

- **Cataract (23.3)**
  - 13.4 years (0.29)
  - 100%

- **Childhood Onset Chronic Diarrhea (11.7)**
  - 11.4 years (0.26)
  - 64%

- **Onset Symptom (11.9)**
  - 24.7 years (13,34)

23% abnormality of the nervous system
- 15% abnormality of higher mental function
- 8% seizures

77% extra-neurological abnormality
- 62% abnormality of the abdomen
- 15% abnormality of the eye

100% pyramidal signs
81% ataxia, behavioural or psychiatric abnormality
78% xanthomatosis
63% dementia
37% peripheral neuropathy
36% psychomotor retardation
25% epilepsy
18% parkinsonism