

# Major intra-familial variability in Unverricht-Lundborg disease

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## Progressive Myoclonic Ataxia (PMA)

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graph TD; PMA[Progressive Myoclonic Ataxia (PMA)] --> Acquired[→ Acquired causes]; PMA --> Inherited[→ Inherited causes];
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### → Acquired causes

- Autoimmune
- Infectious
- Paraneoplastic

### → Inherited causes

- Unverricht Lundborg Disease (ULD)
- Mitochondriopathies
  - Sialidosis
- Alzheimer's disease
- Down syndrome

**ULD**

```
graph TD; A[ULD] --> B["→ Intra-familial variability  
(Age at onset/Clinical features/Disease severity/  
Disease progression)"]; B --> C["→ No correlation between the number of  
dodecamers of the cystatin B (CSTB) gene and the  
severity of ULD phenotype"];
```

**→ Intra-familial variability**  
(Age at onset/Clinical features/Disease severity/  
Disease progression)

**→ No correlation between the number of  
dodecamers of the cystatin B (*CSTB*) gene and the  
severity of ULD phenotype**