**Condition:** MERFF (myoclonic epilepsy with ragged-red fibers)

**Inheritance:**
Mitochondrial (maternal transmission).

**Genetic etiology:**
Mutation in *MT-TK* gene encoding mitochondrial tRNA lysine.

**Frequency:**
Approximately 1/400,000.

**Clinical features:**
MERFF is a highly variable disorder that affects energy metabolism in multiple tissues. Onset is most commonly in childhood, with myoclonus, generalized seizures, ataxia, weakness, and dementia.

**Management:**
Supportive care; administration of coenzyme Q10 and L-carnitine.

**Genetic counseling:**
Follows pattern of maternal transmission, but with widely variable expression due to heteroplasmy. Molecular genetic testing is available.