Dejerine-Sottas syndrome

INSERM

Source

INSERM. (1999). Orphanet: an online rare disease and orphan drug data base. Dejerine-Sottas syndrome. ORPHA:64748

Dejerine-Sottas syndrome is a clinical entity that represents a severe phenotype of Charcot-Marie-Tooth disease (see this term) characterized by onset occurring in infancy, severe motor weakness, delayed motor development, extremely slow nerve conduction (< 10-12 m/s), areflexia and foot deformity. Mutations in the genes PMP22 (17p12), MPZ (1q22), EGR2 (10q21.1) and PRX (19q13.2) have been implicated.