

Dejerine-Sottas Disease

(CMT Type 3 or Chromosome 19 recessive form can be called CMT4F)

Last updated June 2010

Definition

A neurological disorder that causes damage to the peripheral nerves, which carry signals from the brain and spinal cord to muscles, and relay sensations, such as pain and touch, to the brain and spinal cord from the rest of the body ; generally, any peripheral nerve disorder that resembles Charcot-Marie-Tooth (CMT) disease but is more severe is called Dejerine-Sottas disease

Cause

Defects in the genes for proteins found in axons, fibers that carry electrical signals between the brain and spinal cord and the rest of the body, or in the genes for proteins found in myelin, a coating on axons that insulates and nourishes them.

Onset

Infancy or early childhood

Symptoms

Weakness, especially in the lower legs, forearms, feet and hands, with muscle wasting as disease progresses; reduced muscle tone; loss of sensation, particularly in the lower legs, forearms, feet and hands; curvature of the spine; foot deformities; slow acquisition of motor skills in childhood; occasionally, limitation of eye movements or other eye problems; occasionally, mild hearing loss.

Progression

Slow until teens; severe disability may eventually occur

Inheritance

Autosomal dominant, meaning can be inherited through a faulty gene contributed by either parent; or autosomal recessive, meaning can be inherited through a faulty gene contributed