Cone dystrophy with supernormal rod response

INSERM

Source

INSERM. (1999). Orphanet: an online rare disease and orphan drug data base. Cone dystrophy with supernormal rod response. ORPHA:209932

Cone dystrophy with supernormal rod response (CDSRR) is an inherited retinopathy, with an onset in the first or second decade of life, characterized by poor visual acuity (due to central scotoma), photophobia, severe dyschromatopsia, and occasionally, nystagmus. Night blindness usually develops later in the course of the disease, but it can also be apparent from childhood. A hallmark of CDSRR is the decreased and delayed dark-adapted response to dim flashes in electroretinographic recordings, which contrasts with the supernormal b-wave response at the highest levels of stimulation.