it has also been found in some cases of CMT1B (P0). The pathology in autosomal recessive CMT2 is more severe than in autosomal dominant CMT2. The genetic defect in some autosomal dominant CMT2 families is linked to chromosome 1p36.

HETEROZYGOUS MUTATION IN SEVERE, INFANTILE, HYPERTROPHIC HEREDITARY MOTOR AND SENSORY NEUROPATHY

Oral Presentation

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Severe, early-onset cases of demyelinating hereditary motor and sensory neuropathy (HMSN) and hypomyelination in nerve biopsy are often designated as Dejerine–Sottas syndrome or HMSN type III; this entity was defined as an autosomal recessive disorder. Formerly, the authors have argued that the cases fulfilling this definition, but with mainly classical onion bulbs in nerve biopsy, are not autosomal recessive cases. Indeed, a few cases classified as Dejerine–Sottas syndrome or HMSN type III, but with classical onion bulbs, resulted from heterozygous mutations in the PMP-22 gene or the P_0 gene. The authors present the genetic investigation of one of our patients with a severe, early-onset demyelinating neuropathy, high cerebrospinal fluid protein and hypomyelination with classical onion bulbs. DNA analysis revealed a heterozygous *de novo* mutation in the PMP-22 gene. This implies the existence of an autosomal dominant mutation in this patient.

SELF-MUTILATION IN HEREDITARY SENSORY NEUROPATHIES

Poster

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Self-mutilation (SM) in childhood is a complicated symptom with numerous aetiologies. Traditionally, severe mental retardation and/or profound psychiatric disorders are the main underlying conditions. The existence of SM in Tourette and Rett syndromes has only recently been recognised, while its mechanism in Lesch–Nyhan syndrome is still unclear. The authors have documented severe and disablng SM in familial dysautonomia and in hereditary insensitivity to pain with anhidrosis. (HASN types III and IV, respectively). Both conditions share a similar neuropathological picture consisting of selective loss of small myelinated fibres in peripheral nerves. This selective loss of pain-conducting fibres with preservation of larger fibres may be responsible for the hedonistic SM.

A FAMILY APPROACH TO THE APPLICATION OF COMPUTER GAMES FOR THE REHABILITATION OF CHILDREN WITH CEREBRAL PALSY

Poster

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The aim of this presentation is to provide an analysis of the application of computer games in the process of rehabilitation for disabled children. 150 children were examined aged between three and 15 years (78 boys, 72 girls) from families of differing social status and mostly with the spastic forms of cerebral palsy (two cases of familial cerebral palsy). 120 children have intellectual impairment. An IBM personal computer AT with different devices (joystick, mouse, keyboard, spatial hand manipulator created to order by the authors' centre which enables children with finger paresis to play computer games, and different software) was used. In two to three days the authors managed to teach a child (for whom the computer was a new experience) to correlate the actions