Rome, Italy (Mirabella M et al. Neurology April 1992; 42 Suppl 3:281). There was no muscle weakness and serum CK was only mildly increased.

HEREDITARY MOTOR AND SENSORY NEUROPATHY MUTATION

A duplication in chromosome 17 responsible for most cases of autosomal dominant HMSN 1 was present as a de-novo mutation in 9 out of 10 sporadic patients examined at the Department of Neurology, Academic Medical Center, Amsterdam, The Netherlands. (Hoogendijk JE et al. De-novo mutation in hereditary motor and sensory neuropathy type 1. Lancet May 2, 1992; 339:1081-82.) (Correspondence: Dr. JE Hoogendijk, Department of Neurology, Academic Medical Center, Meibergdreef 9, 1105 AZ Amsterdam, The Netherlands.)

COMMENT. The previous assumption that isolated cases of HMSN I are most frequently autosomal recessive appears to be incorrect and testing for the duplication in chromosome 17 is essential to establish the mode of inheritance for genetic counseling of isolated patients.

This duplication of part of chromosome 17 was found in affected individuals from 7 of 8 families with HMSN I. Patients with HMSN type II do not show the duplication (Hallam PJ et al. Ann Neurol May 1992; 31:570-572). (Correspondence: Dr. Malcolm, Molecular Genetics Unit, Institute of Child Health, 30 Guilford St., London WC1N 1EH UK.)

HMSN type I also known as Charcot-Marie-Tooth disease type I is known to be genetically heterogeneous. At least 5 genetic loci have been identified including 3 dominant genes and 2 X-linked recessive genes. Ionasescu VV et al. from the Department of Pediatrics, University of Iowa Hospitals, Iowa City and the Massachusetts General Hospital present a clinical and genetic linkage study of 8 families with X-linked dominant Charcot-Marie-Tooth neuropathy which supports a localization of the diseased gene between DXS14 and DXYS1 (Neurology April 1992; 42:903-908).

SEIZURE DISORDERS

TEMPORAL LOBECTOMY FOR INTRACTABLE SEIZURES

The results of temporal lobectomy in 16 children under the age of 12 years who had intractable seizures of temporal lobe origin are reported from the Miami Children's Hospital, Florida. All had seizures for at least 2 years despite multiple antiepileptic drugs. Structural lesions were demonstrated by neuroimaging in 11 patients. At follow-up from 1 to 4 years 11 children were seizure free, 3 were 90% improved, 1 was 50% improved and 1 was unchanged. Of 9 families contacted 7 reported substantial improvement in the child's psychosocial function. Neuropathological abnormalities identified in all children showed findings consistent with disordered neuronal migration in 7, tuberous sclerosis in 1, and mesial temporal sclerosis only in 2 children. (Duchowny M et al. Temporal lobectomy in early childhood. Epilepsia

March/April 1992; 33:298-303.) (Reprints: Dr. M. Duchowny Comprehensive Epilepsy Center, Miami Children's Hospital, 3200 S.W. 60 Court, Miami, FL 33155.)

COMMENT. Anterior temporal lobectomy tailored according to the extent of the lesion and epileptogenic field may be beneficial in preadolescent children with medically refractory seizures. Compared to adults, children undergoing temporal lobectomy have a higher incidence of abnormalities of neurogenesis and lesions that are poorly circumscribed and often widespread.

INTRACRANIAL ARACHNOIDAL CYSTS

Of 28 cases of intracranial arachnoidal cysts reported from the Clinica Las Condes, Santiago, Chile, 17 were located in the middle cranial fossa, 3 in the quadrigeminal cistern, 2 were parasagital, 3 suprasellar and 3 were in the posterior fossa. Of 23 (82%) treated surgically, 3 received craniotomy with fenestrations, 8 had shunts and 15 were treated by the insertion of cystoperitoneal drainage with excellent results. Twenty-one of the 22 treated cases are asymptomatic and leading a normal life. MRI or CT follow-up showed disappearance of cysts in 14 and marked reduction in size in 7. (Basauri L, Selman JM. Intracranial arachnoidal cysts. Child's Nerv Syst March 1992; 8:101-104.) (Reprints: Dr. L. Basauri, Neurosurgical Unit, Clinica Las Condes, Casilla 27014, Santiago, Chile.)

COMMENT. The diagnosis of intracranial arachnoidal cysts and reports in the literature have become more frequent since the routine use of neuroimaging. The danger of intracystic hemorrhage with a high possibility of secondary hydrocephalus following head injury has been quoted as a reason for surgical intervention, as a preventive measure in some cases. See Ped Neur Briefs Oct 1991, 5:80 for a report of 67 childhood arachnoidal cysts treated surgically and 1 report of spontaneous disappearance of a cyst. Small or medium sized cysts without clinical manifestations are not usually treated but are monitored carefully.

UNBOUND PHENYTOIN AND VALPROATE MONITORING

All antiepileptic drug blood level determinations at the University of Virginia Medical Center were analyzed over a 13 month period. Those values with unbound fractions outside the expected range and discordance between unbound and total levels were compared to seizure control and occurrence of side effects. Unbound phenytoin (PHT) levels were clinically significant (outside the expected range, a discordant ratio of unbound to total level, and seizures or side effects necessitating dosage change) in 61 of 254 determinations (24%) and 44 of 155 patients (28%). The clinical significance involved current seizures for 52 values and side effects for 24 (both for 15). The frequency of clinically significant values was similar with PHT monotherapy (20%) and polytherapy (27%). The rates of current seizures and side effects were also similar in these 2 subgroups. For valproate (VPA), 15% of unbound values from 18% of patients were clinically significant with regard to current seizures and side effects and the rate was similar for monotherapy and polytherapy. There were no clinically significant unbound