(42%) patients. Nerve conduction studies performed in 15 GD1 patients showed abnormalities in 12 (2 children); 26 – 40% showed reduced amplitude or abnormal wave forms in one to 3 nerves. Motor nerve conduction was normal, but sensory nerve conduction amplitude was reduced in 26% patients. Twenty-two patients receiving enzyme replacement therapy for 5-12 years showed neurological manifestations in 9. In adults, these were sensory neuropathy, parkinsonism, dementia, cognitive impairment and stroke; and in children, hypoacusia, myoclonus, and psychomotor delay. (Capablo JL, de Cabezon AS, Fraile J et al. Neurological evaluation of patients with Gaucher disease diagnosed as type 1. **J Neurol Neurosurg Psychiatry** February 2008;79:219-222). (Respond: Dr P Giraldo, Servicio de Hematolgia, Hospital Universitario Miguel Servet, Paseo Isabel La Catolica 1-3, 50006 Zaragoza, Spain).

COMMENT. Gaucher's disease, characterized by cerebroside storage in the reticuloendothelial system, occurs in 3 forms: Type 1) chronic GD, slowly progressive with visceral but rarely nervous system involvement; type 2) infantile GD with rapidly progressive CNS disease; and type 3) juvenile GD, with slowly progressive hepatosplenomegaly, intellectual deterioration, cerebellar ataxia, myoclonic seizures, and spasticity. An adult chronic GD is more common and may present in infancy, with rare CNS involvement. The above study shows that neurological abnormalities, including subclinical neuropathy, occasionally present in childhood GD1, and the classification of GD should be considered as a phenotype continuum.

DEMYELINATING DISEASES

SEROPREVALENCE OF NEUROMYELITIS OPTICA-IgG OF CHILDHOOD COMPARED TO ADULTS

The clinical and radiological characteristics and serostatus of neuromyelitis optica (NMO)-IgG in 87 children with inflammatory demyelinating CNS disorders were analyzed in a study at the Mayo Clinic, Rochester, MN, and other centers in the US, Canada, and Argentina. Seventeen patients had NMO and of these, 8 (47%) were seropositive. The prevalence of seropositivity was higher with relapsing NMO (7 of 9, 78%) than monophasic NMO (1 of 8, 12.5%, p=0.01). The majority of children with NMO (14 of 17) were enrolled from the program in Argentina, and few came from Canada. None showed oligoclonal bands in the CSF. MRI abnormalities were revealed in 9 (53%). After a follow-up of 36 months (range 1.2-126 months), 6.3% children with NMO were wheelchair-bound and 23% had severe visual impairment. One of 5 children with relapsing optic neuritis and none of 8 with monophasic optic neuritis was seropositive. Among 41 with relapsing-remitting multiple sclerosis, 9 with transverse myelitis, and 3 with ADEM, none was seropositive. The frequency of NMO-IgG in children is similar to that in adults. Longitudinally extensive

spinal lesions on MRI are not as predictive of NMO in children as in adults. (Banwell B, Tenembaum S, Lennon VA et al. Neuromyelitis optica-IgG in childhood inflammatory demyelinating CNS disorders. **Neurology** January 29, 2008;70:344-352). (Reprints: Dr Sean Pittock, Department of Neurology, Mayo Clinic, Rochester, MN 55905).

COMMENT. Neuromyelitis optica is characterized by monophasic or recurrent episodes of optic neuritis and longitudinally extensive transverse myelitis, either monophasic or recurrent. The autoantibody NMO-IgG is present in 73% of adults with NMO and is 92% specific for NMO and related disorders, recurrent optic neuritis or transverse myelitis. In children with NMO in the above study, 47% were seropositive. The role of NMO-IgG autoantibody in NMO is unknown, but it may be important in recurrent disease. An overview of NMO in children is provided in an editorial (Levy M et al. **Neurology** Jan 2008;70:334-335).

The most useful diagnostic feature of NMO or NMO-spectrum disorders in adults is a longitudinally extensive spinal cord lesion. This contrasts with the well-circumscribed foci of increased T2-weighted signal in multiple sclerosis, typically seen in adults. In children with MS, the discrete spinal lesions are common, but 14% also show the longitudinally extensive spinal lesions, rare in adults with MS. A longitudinally extensive spinal lesion in a child with demyelinating disease does not exclude a diagnosis of MS and is less predictive of an NMO-spectrum disorder than in adult patients.

Relationship between NMO and autoimmune disease. The association of NMO-IgG and non-organ-specific autoantibodies in patients with systemic lupus erythematosis (SLE) and in those with NMO spectrum disorder was evaluated at the Mayo Clinic and University of Lille, France (Pittock SJ et al. **Arch Neurol** 65:78-83). Patients with NMO were seropositive for NMO-IgG, and those with SLE without NMO were seronegative for this autoantibody. NMO-IgG is specific for distinguishing NMO spectrum disorder from multisystem autoimmune disorders. NMO may coexist with SLE and other autoimmune disease and is not a complication of SLE.

CORTICAL DEMYELINATION IN MULTIPLE SCLEROSIS

Cortical demyelination in CNS inflammatory demyelinating diseases is reviewed in an editorial and in a study by Moll NM et al (**Neurology** 2008;70:336-343). Cortical demyelination occurs in 3 different patterns: 1) leukocortical lesions, affecting both gray and white matter; 2) small perivenous intracortical lesions; and 3) widespread subpial demyelination. Type 3 is most abundant in multiple sclerosis (MS) and is related to chronic inflammation of the meninges. Cortical lesions in progressive multifocal leukoencephalopathy (PML) are similar to those in MS but are absent in HIV-encephalitis and adrenoleukodystrophy. T-cell inflammation is sparse in MS and PML cortical lesions in contrast to white matter lesions. Widespread subpial demyelination may be pathognomonic for MS and related inflammatory demyelinating diseases. (Lassmann H, Lucchinetti CF. Cortical demyelination in CNS inflammatory demyelinating diseases. **Neurology** Jan 29, 2008;70:332-336). (Reprints: Prof Dr Hans Lassmann, Div Neuroimmunology, Centre for Brain Research, Medical University of Vienna, Spitalgasse 4, A-1090 Wien, Austria).