

whereas in MS they are less than two or three vertebral segments in length. In ADEM, cerebral MRI lesions may be massive, sometimes mistaken for tumors, Schilder's or Balo's disease, or strokes. Devic's syndrome or neuromyelitis optica, called 'Oriental form of MS' in Japan, has been classified as a form of MS in the past. Recent reports suggest that Devic's syndrome is different from MS and more closely resembles DEM. Treatment and prognostic considerations are important reasons to differentiate between a CIS of MS and acute DEM, and between relapsing-remitting MS and recurrent DEM. Lifelong immunomodulatory treatment is initiated immediately after an MS CIS, whereas IV methylprednisolone, IV immunoglobulin G, or plasma exchange are accepted treatments for ADEM. (Poser CM, Brinar VV. Disseminated encephalomyelitis and multiple sclerosis: two different diseases – a critical review. *Acta Neurol Scand* October 2007;116:201-206). (Respond: Charles M Poser MD, 11 Rutland Square, Boston, MA 02118).

**COMMENT.** The differentiation of ADEM and MS is made on clinical history, neurologic and MRI findings, and CSF analysis. Given the variety and remitting nature of clinical symptoms of demyelinating disease, a neurologist with expertise in neuroradiology may be best qualified to correctly diagnose the disorder. The above review regards the two diseases as distinct entities and not parts of a spectrum, as sometimes proposed.

**The spectrum of neuromyelitis optica, (NMO).** Researchers from the Mayo Clinic, Scottsdale, AZ, and Rochester, MN, review the role of the serum autoantibody marker (NMO-IgG) in the pathogenesis of NMO and in the definition of an extended spectrum of NMO-related disorders, distinct from MS. (Wingerchuk DM et al. *Lancet Neurol* September 2007;6:805-815). The potential pathogenicity of NMO-IgG, the role of aquaporin 4 as the inducer and target of the autoimmune attack, with resulting inflammatory demyelination and necrosis, and treatment are discussed. The revised Wingerchuk diagnostic criteria for NMO require 2 of the following: 1) Brain MRI is not diagnostic of MS; 2) spinal cord lesion must be 3 or more segments in length; and 3) patient is NMO-IgG antibody seropositive.

**NMO in a mother and daughter** is reported from University of Michigan, Ann Arbor. (Braley T, Mikol DD. *Arch Neurol* August 2007;64:1189-1192). The ages of onset were 62 and 29 years, respectively. Few familial cases of NMO are described.

## **SEIZURE DISORDERS**

### **MORTALITY OF STATUS EPILEPTICUS IN ALL AGE GROUPS**

The in-hospital mortality and potential predictors of outcome of generalized convulsive status epilepticus (GCSE) were evaluated at the Neurological Institute, Case School of Medicine, Cleveland, OH. The cohort was identified from the Nationwide Inpatient Sample (NIS) databases for the years 2000 through 2004. The definition of status epilepticus employed was continuous seizures or repetitive seizures without recovery of consciousness of 30 or more minutes' duration. The analysis included 11,580 patients (mean age 39 +/- 25.6 years, 53.4% male, 42.4% white) with a median hospital stay of 3 days. Mortality was 3.45% overall. Adjusted mortality rates by age were 0.67% for patients < or =10 years, 1.33% for ages 11-20, increasing to 10.15% for patients >80 years. Potential

predictors of death identified female sex, mild Charlson Comorbidity Index (chronic illness without cancer), and Northeast hospital location. Mortality tripled in those requiring mechanical ventilation (7.43% vs 2.22%;  $p < 0.0001$ ), mostly older patients. Potential etiologies and complications of GCSE that predicted mortality included cerebrovascular disease, hypoxic-ischemic brain injury, cardiac disease, respiratory failure, and higher comorbidity index. In the first decade of life, GCSE is the most common neurologic emergency. The number of cases  $< 10$  years old in this cohort (2,524, 21.8%) was comparable to all cases older than 60 years of age (2,627, 22.7%). (Koubeissi M, Alshekhlee A. In-hospital mortality of generalized convulsive status epilepticus: a large US sample. *Neurology* August 28, 2007;69:886-893). (Reprints: Dr Mohamad Z Koubeissi, Department of Neurology, University Hospitals Case Medical Center, Case Western Reserve University, 11100 Euclid Ave, Cleveland, OH 44106).

COMMENT. A bimodal age distribution of cases of GCSE, with highest frequencies in children  $< 10$  years and in the elderly is similar to other reports. The low mortality rate in young children in this study, lower than some previous series, is explained by the authors as a reflection of improved management and availability of rectal antiepileptic medications, or the relatively low frequency of acute symptomatic epilepsies in their cohort. A more detailed analysis of etiological factors related to GCSE outcome in children would be of interest, especially in children  $< 1 - 2$  years of age, when the incidence is highest and febrile CSE is the most common cause.

## EPIDEMIOLOGY OF STATUS EPILEPTICUS IN CHILDREN

The incidence, etiology, seizure characteristics, and outcome in childhood convulsive status epilepticus (CSE) are reviewed by researchers from Great Ormond Street Hospital for Children, and the Institute of Child Health, London, UK. Neonatal seizures require separate consideration and were excluded. Despite attempts to modify the definition of CSE, without a set temporal criterion (ILAE (2001), or a duration as low as 5 min (operational definition), there is no clear reason to modify the “traditional” 30-min-duration definition. By 30 min CSE has become self-sustaining, drug resistance has occurred, and neuronal injury has begun.

Etiology is an important determinant of CSE, and a significant proportion of cases are associated with fever. ILAE classification of SE according to etiologies (1993) lists 1) *acute symptomatic* - previously neurologically normal child, within 1 week of underlying etiology including CNS infection, prolonged febrile seizure, encephalopathy, head trauma, cerebrovascular disease, metabolic or toxic; 2) *remote symptomatic* - preexisting CNS disorder  $> 1$  week before; 3) *idiopathic epilepsy related*; 4) *cryptogenic epilepsy related*; and 5) *Unclassified*. This classification should be revised, with febrile CSE as a distinct category with an overall favorable prognosis, and separate from acute symptomatic CSE.

Incidence estimates of CSE in children are few, the most recent conducted in North London, the only study of a wholly pediatric population. The incidence of childhood CSE in North London is 18-20/100,000/year, higher than the 4-6/100,000/year reported in adults, excluding the elderly. The incidence in children less than 1 year is 51/100,000/year, compared to those aged 1-4 (29/100,000/year). The higher frequency of CSE in the very young is related to a high proportion of acute symptomatic causes, to brain immaturity, or