were not associated. Also, first-degree family history of febrile seizure was not associated with behavioral problems in probands with uncomplicated or in those with complicated epilepsy. The familial clustering of these disorders suggests that behavioral disorders may be another manifestation of the underlying pathophysiology involved in or related to epilepsy. (Hesdorffer DC, Caplan R, Berg AT. Familial clustering of epilepsy and behavioral disorders: evidence for a shared genetic basis. **Epilepsia** Feb 2012;53(2):301-307). (Respond: Dr Dale C Hesdorffer, GH Sergievsky Center, Columbia University, 610 West 168th St, P & S Unit 16, New York, NY 10032. E-mail: dch5@columbia.edu).

COMMENT. Epilepsy and behavioral disorders appear to have a common underlying genetic predisposition, whereas in the above study febrile seizure had no significant familial association with behavioral disorders. Previous reports of behavior disorder in children with febrile seizure have varied findings. Friderichsen C and Melchior J (**Acta Paediatr** 1954;43:307-317) found behavior disorders in 12 (4%) of 282 febrile seizure patients, and Millichap JG et al (**Neurology** 1960;10:643-653) in a prospective study of 110 febrile seizure patients reported recurrent episodes of aggressive behavior, temper tantrums, and hyperactivity in 35% patients. Patients with a history of birth trauma and those with cryptogenic epilepsies were excluded from the Friderichsen and Melchior series of febrile seizures but not from the study by Millichap and colleagues.

Risk of behavioral, developmental, and physical comorbidities with epilepsy/seizure disorder in a nationally representative sample of US children. (Russ SA, Larson K, Halfon N. **Pediatrics** February 2012;129(2):256-264). Estimated lifetime prevalence of epilepsy/seizure disorder was 1%, and of current epilepsy/seizure disorder was 6.3/1000. Children with current epilepsy/seizure disorder were significantly more likely than those never affected to have ADHD (23% vs 6%), developmental delay (51% vs 3%), autism (16% vs 1%), and headache (14% vs 5%). Those with prior but not current seizures had lesser risks.

NEUROMUSCULAR DISORDERS

SPINAL MUSCULAR ATROPHY II/III AND FEEDING PROBLEMS

Researchers at Kaohsiung Medical University Hospital, Taiwan studied the prevalence and risk factors of feeding and swallowing problems in 108 genetically confirmed patients with types II and III spinal muscular atrophy (SMA), age range 3-45 years, 60 with type II and 48 with type III. A questionnaire survey showed the 3 most common feeding and swallowing difficulties were choking (30.6%), difficulty conveying food to the mouth (20.4%), and difficulty chewing (20.4%). Motor function status (sitters vs walkers) was an independent risk factor for feeding and swallowing difficulties; 28 were walkers, 76 sitters, and 4 nonsitters. All 4 SMA II nonsitters had feeding and swallowing difficulties. Poor head control when feeding was a factor in 13 (12%) patients. Age was not an independent risk factor in this study; 10 patients, all with type II SMA and age <20 years (range 3-19 years), had feeding and swallowing difficulties and required respiratory management. Respiratory assistance or suction was required in 17

patients (15.8%). Patients with feeding and swallowing difficulties had higher rates of underweight and aspiration pneumonia than those without these problems. Individual treatment plans for SMA II/III patients should depend on motor function status. (Chen Y-S, Shih H-H, Chen T-H, Kuo C-H, Jong Y-J. Prevalence and risk factors for feeding and swallowing difficulties in spinal muscular atrophy types II and III. **J Pediatr** March 2012;160:447-451). (Response and reprints: Yuh-Jyh Jong MD, Department of Pediatrics, Kaohsiung Medical University Hospital, 100, Shih-Chuan 1st Road, Kaohsiung 80708, Taiwan. E-mail: yjjong2@gap.kmu.edu.tw)

COMMENT. Classification of SMA types I, II, III, and IV is based on age at onset and the highest function achieved. (Lunn MR, Wang CH. **Lancet** 2008;371:2120-2133). In a Hong Kong, China study (Chung BH et al. **Pediatrics** 2004;114(5):e548-e553) survival probabilities for type I SMA (n=22) at 1, 2, 4, 10, and 20 years were 50%, 40%, 30%, 30%, and 30%, respectively. For type II SMA (n=26), survival probabilities at 1, 2, 4, 10, and 20 years were 100%, 100%, 100%, 92%, and 92%, respectively. Sixteen of the SMA I patients and 4 of the SMA type II patients died of cardiorespiratory failure. All SMA III patients were surviving. The probability of remaining ambulatory at 20 years after onset of type IIIa (age of onset <3 years) was 50%, and for type IIIb (age of onset 3-30 years) it was 68%. Interval between disease onset and inability to walk was 15 years for type IIIa and 21.2 years for type IIIb patients. In the Taiwan study, feeding and swallowing difficulties, especially choking, in SMA types II and III patients were correlated with current motor function status.

Double-trouble: SMA type II and seropositive myasthenia gravis in a 51 yr old male. (Jokela M, Udd B, Paivarinta M. **Neuromuscular Disorders** Feb 2012;22:129-130). A case report from Finland concerned a patient with SMA type II living to age 51 years and then developing worsening of dysphagia and chewing over a few weeks. A mild respiratory infection led to rapid deterioration in ventilatory function and need for tracheostomy and permanent night-time ventilator support. Ptosis of left eye, ophthalmoplegia, myopathic face, and left hand weakness followed. Serum acetylcholine receptor antibodies were elevated (44 nmol/L; normal 0.25-0.40 nmol/L), and edrophonium testing for myasthenia gravis was positive. CT chest was negative for thymoma or thymus hyperplasia. Myasthenia responded to a course of iv immunoglobulin, oral prednisone, and pyridostigmine. Azathioprine was substituted for the prednisone. Ocular findings are very atypical for SMA and a diagnosis of MG was suspected as a chance association of two rare diseases.

TREATMENT AND OUTCOME OF STIFF-MAN SYNDROME

Neurologists at the Mayo Clinic, Rochester, MN extended their reports of patients with stiff-man syndrome (SMS), first reported there by Drs Moersch and Woltman in 1956. They describe the characteristics of a large cohort of 99 patients (67 female), their treatment and outcome. Median age at symptom onset was 40 years (range 5-70 years); 5 presented before 18 years of age. Mean follow-up from symptom onset was 5 years (range 0-23 years). Phenotypic symptoms included low back stiffness and spasms in all of 59 classic cases, exaggerated lumbar lordosis in 52, lower extremity stiffness and