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

spasms in 59, neck stiffness and spasms in 10, upper extremity stiffness and spasms in 4, abdominal wall stiffness and spasms in 26, respiratory symptoms with spasms in 6, and falls in 30. Symptoms were exacerbated by emotional stress, startle, cold, and movement. Seventy-nine were GAD65 antibody seropositive, and 53 (67%) had at least one coexisting autoimmune disease; 3 (4%) had cancer. GAD65 antibody values were significantly higher in patients with classic SMA than in those with partial or variant SMA. Treatment with diazepam (40 mg/day) provided sustained improvements. Immunotherapy gave additional improvements. Sixteen (64%) of 23 patients with extended follow-up remained ambulatory. (McKeon A, Robinson MT, McEvoy KM et al. Stiff-man syndrome and variants. Clinical course, treatments, and outcomes. **Arch Neurol** Feb 2012;69(2):230-238). (Respond: Andrew McKeon MD, Department of Neurology, Mayo Clinic. E-mail: mckeon.andrew@mayo.edu).

COMMENT. Stiff-man syndrome occurs mainly in adults but can occur in children. Diagnosis may be confirmed with EMG documentation of hyperexcitability of spinal motor neurons, GAD65 antibodies, and response to diazepam, first described by Howard FM Jr (**Proc Staff Meet Mayo Clin** 1963;38:203-212).

Several reports of stiff-child syndrome are uncovered by a PubMed search. The disorder must be distinguished from hyperexplexia or hereditary stiff-baby syndrome, an autosomal dominant disorder. The EMG shows persistent hyperexcitability at rest, abolished by diazepam. The hypertonia lessens during sleep and increases with the slightest startle or tactile stimulus. Nose tapping will elicit the hyperexplexic startle response in affected newborns. (Tohier C et al. **Arch Dis Child** 1991;66:460-461) (**Ped Neur Briefs** May 1991).

MOVEMENT DISORDERS

BENIGN HEREDITARY CHOREA: RESPONSE TO LEVODOPA

A case of sporadic non-progressive chorea is reported in a 6 year-old girl from Hospital Sant Joan de Deu, Barcelona University and other centers in Spain and The Netherlands. At age 21 months she was diagnosed with severe motor delay and gait disorder. Birth and perinatal history including screening test for hypothyroidism were normal. A diagnosis of subclinical hypothyroidism was made at 2 years of age and she was treated with oral L-thyroxine. Language and learning skills have been age appropriate. At 3 years of age she was hypotonic, reflexes were normal, but her gait was unstable, clumsy, and wide-based, with frequent, sudden falls. Choreiform movements were generalized, affecting the mouth, limbs, and trunk, and were not progressive. A *TITF-1* de novo gene mutation test was positive. Levodopa therapy started at age 3 years 6 months controlled the chorea. When therapy was temporarily interrupted after 1 year, symptoms recurred with frequent falls and clumsy gait. Discontinuation of therapy slowly after 3 years of treatment was successful without relapse. (Fons C, Rizzu P, Garcia-Cazorla A, et al. TITF-1 gene mutation in a case of sporadic non-progressive chorea. Response to levodopa treatment. **Brain Dev** 2012;34:255-257). (Respond. Dr Carmen Fons, Department of Pediatric Neurology, Hospital Sant Joan de Deu, Barcelona, Spain. E-mail: cfons@hsjdbcn.org).