

adherence to recommended titration procedures was followed by improvements in total and inattentive ADHD symptoms on the ADHD Rating Scales and SNAP-IV parent and teacher scales. Computer-assisted medication management may contribute to better treatment outcomes in pediatric primary care management of ADHD. (Lavigne JV, Dulcan MK, LeBailly SA, Binns HJ, Cummins TK, Jha P. Computer-assisted management of attention-deficit/hyperactivity disorder. *Pediatrics* July 2011;128(1):e46-e53). (Respond: John V Lavigne PhD, Department of Child and Adolescent Psychiatry (#10), Children's Memorial Hospital, 2300 Children's Plaza, Chicago, IL 60614. E-mail: jlavigne@childrensmemorial.org).

COMMENT. Successful management of the child with ADHD is benefitted by computer-assisted titration of medications. Brief training periods for physicians are not sufficient to obtain a better outcome. Computer-assisted dose-response decision-making increases adherence to guidelines. The optimal dose is the lowest dose producing a clinically significant change and symptom rating in the average range. This is associated with significant improvements in classroom behavior. Practices that stress to the parents the importance of obtaining school reports are more likely to obtain optimal results.

NEUROMUSCULAR DISORDERS

COURSE OF SELENOPROTEIN-RELATED MYOPATHIES

The clinical course and genotype-phenotype correlations in 41 patients aged 1-60 years with selenoprotein-related myopathy (SEPNRM) due to *SEPN1* gene mutations were evaluated retrospectively in a study at The Dubowitz Neuromuscular Center, London, and other centers in the UK. Mean age at onset was 2.7 years, ranging from birth to second decade. One third were congenital in onset with hypotonia. In 46%, onset was at 6 months to 5 years of age with delayed motor milestones. Creatine kinase was minimally elevated in 16% and markedly elevated only in 1. All but 2 patients remained independently ambulant. Respiratory insufficiency generally developed and nocturnal noninvasive ventilation was started at a mean age of 13.9 years. Scoliosis preceded by rigid spine developed at a mean age of 10 years and was treated surgically at 13.9 years. Joint contractures were present in 26 (63%) at a mean age of 10.4 years; 2 patients had finger contractures at birth. Motor abilities remained static over time. Twenty of 35 (57%) were underweight. Two patients died from respiratory failure at age 10 and 22 years. Muscle biopsy performed in 34 patients showed multimimicres and nonspecific myopathic changes. Genetic sequencing showed 14 new *SEPN1* mutations, 48% missense. (Scoto M, Cirak S, Mein R, et al. *SEPN1*-related myopathies. Clinical course in a large cohort of patients. *Neurology* June 14, 2011;76(24):2073-2078). (Respond and reprints: Dr Francesco Muntoni, The Dubowitz Neuromuscular Centre, UCL Institute of Child Health, 30 Guilford Street, London WC1N 1EH, UK. E-mail: f.muntoni@ich.ucl.ac.uk).

COMMENT. This study expands the spectrum of myopathy with *SEPN1* mutations, with respect to severity of the disease, age at onset, and long-term outcome.