## MUSCLE DISORDERS

### CONGENITAL MYASTHENIC SYNDROME AND AChR MUTATION

A 20-year-old woman from a consanguineous Moroccan marriage, with progressive muscle weakness noted from 2 years of age and evaluated at the University of Bonn, Germany, was found to have congenital myasthenic syndrome (CMS) due to homozygosity of the 1293insG e-acetylcholine receptor subunit mutation. Compared to the original case report of a CMS with end-plate acetylcholine receptor deficiency, heteroallelic for two e-AChR subunit mutations, and affected with mild muscle weakness, in this homozygous case, the weakness was profound and was associated with muscle wasting. (Sieb JP, Kraner S, Schrank B et al. Severe congenital myasthenic syndrome due to homozygosity of the 1293insG e-acetylcholine receptor subunit mutation. Ann Neurol September 2000;48:379-383). (Respond: Dr OK Steinlein, Institute for Human Genetics, University of Bonn, Wilhelmst 31, Bonn D-53105, Germany).

COMMENT. Differences in the acetylcholine receptor mutation haplotype can markedly influence the severity of congenital myasthenic syndrome. This profoundly affected patient is wheel-chair bound, and has almost complete external ophthalmoplegia and progressive kyphoscoliosis. Therapy with 60 to 120 mg of pyridostigmine daily was of limited benefit.

### ACETAZOLAMIDE IN HYPOKALEMIC PERIODIC PARALYSIS

The mechanism of action of acetazolamide in the K-deficient diet rat, an animal model of human hypokalemic periodic paralysis (hypoPP), was investigated at the University of Bari, Italy. In vivo administration of acetazolamide prevented paralysis and depolarization of the fibers induced by insulin. Intense sarcolemma Ca-activated K channel activity was recorded in the acetazolamide-treated animals. The serum K levels were also restored to normal by acetazolamide. (Tricarico D, Barbieri M, Camerino DC. Acetazolamide opens the muscular KCa channel: a novel mechanism of action that may explain the therapeutic effect of the drug in hypokalemic periodic paralysis. Ann Neurol September 2000;48:304-312). (Respond: Professor Conte Camerino, Unit of Pharmacology, Department of Pharmacobiology, Faculty of Pharmacy, University of Bari, 70126, via Orabona Number 4, Bari, Italy).

COMMENT. The observed therapeutic effect of acetazolamide in hypokalemic periodic paralysis may be mediated by the activation of the muscular KCa channel.

## ATTENTION DEFICIT AND BEHAVIOR DISORDERS

### MOTOR DYSFUNCTION IN CHILDREN WITH ADHD

The performance of children with attention deficit hyperactivity disorder (ADHD) in the Movement Assessment Battery for Children test was evaluated at the Motorik Lab, Department of Woman and Child Health, Karolinska Institute, Sweden. In tasks involving motor-memory representations, a special grip object recorded forces generated by the fingertips during a precision grip-lift task. Twenty-five boys, aged 11 years, with ADHD were grouped according to the presence (ADHD+) or absence (ADHD) of movement dysfunction, and compared to

a control group of 25 age-matched boys.

The ADHD+ group, with motor problems, showed deficits in grip-force control during the precision grip-lift task. They had difficulties in adapting motor output to target different weights, suggesting deficiencies in neuromotor control mechanisms, independent of ADHD symptoms. An overall high grip force output used by the ADHD+ group was a compensatory mechanism to increase grip stability, secondary to impaired tactile stimulation and sensory motor control. In the group with ADHD without motor problems, these deficits were less pronounced. (Pereira HS, Eliasson A-C, Forssberg H. Detrimental neural control of precision grip lifts in children with ADHD. Dev Med Child Neurol August 2000;42:545-553). (Respond: Heloisa S Pereira MD MSc, Motorik Lab, Astrid Lindgrens Children's Hospital, S 177 76 Stockholm, Sweden).

COMMENT. Discrete deficits in motor performance, independent of core symptoms of inattention and hyperactivity, occur in children with ADHD complicated by motor problems. Subtypes of ADHD, with and without motor dysfunction, argue against the hypothesis of impaired motor control related to a common neural mechanism for ADHD.

The descriptive term, DAMP (deficiency of attention, motor control, and perception) has been introduced in Scandinavian countries to characterize children with ADHD complicated by motor and perceptual disorders. In the USA, DAMP is regarded as a combination of ADHD and DCD (developmental coordination disorder). Before the American Psychiatric Association introduced the symptom complexes of ADHD and DCD, these patients were regarded as 'clumsy' or having "minimal brain dysfunction (MBD)," and having subtle neurologic abnormalities on examination. The reintroduction and grading of the neurologic findings in the diagnosis of children with ADHD would introduce some objectivity to the definition of the syndrome.

## STIMULANT TREATMENT OF ADHD IN PUBLIC SCHOOLS

A statewide school survey, supervised by school nurses, was performed to determine the prevalence of stimulant medication administered to Maryland public school students for the treatment of attention deficit hyperactivity disorder (ADHD), and reported by Johns Hopkins University Medical Institutions, and the Maryland State Department of Education, Baltimore, MD.

Of 816,465 students surveyed, 20,050 (2.46%) received methylphenidate and 3721 (0.46%) received other medications for ADHD. Methylphenidate was the most common stimulant prescribed, and represented 84% of all drug treatment for ADHD. Amphetamines were used in 11%, clonidine in 1.7%, pemoline in 0.4%, and tricyclic antidepressants in 0.4%. The male:female ratio was 3.5:1 for children receiving medication in elementary schools, and 4.3:1 in secondary schools. White children were medicated twice as often as black and Hispanic students.

Almost 50% of children receiving methylphenidate had special education accommodations, and 8.3% were eligible for Section 504 services, having an impairment that limited their major life activities. Children with Individual Education Programs (IEP), a marker for special education, received medication 5.6 times more often than students in regular education (8.7% of students compared to 1.55%, respectively). In high school, students with IEP were 10-fold more likely to be receiving methylphenidate in school than regular students. School-district rates of methylphenidate treatment varied 5-fold geographically, from a low of 1.18% to a high of 6.02%. Geographic variability was influenced by race/ethnicity demographics, and by large ADHD clinics in some localities.

By medical specialty, pediatricians were the prescribers of