frequent side effect of MPH than OCD. The above report should prompt a greater awareness of the potential risk of OCD in addition to tics in ADHD children treated with MPH. As with MPH-induced seizures, the incidence of this side effect may be higher than the literature documents.

Psychiatric, neuropsychological, and psychosocial features of DSM-IV subtypes of ADHD were assessed in 413 children and adolescents referred to the Pediatric Psychopharmacology Unit, Massachusetts General Hospital, Boston, MA. Combined-type subjects showed the greatest psychiatric impairments compared to other subtypes, but no differences in cognitive or psychosocial functioning. Inattentive subjects were more likely to require extra help in school. Hyperactive-impulsive patients were not different from controls on measures of depression, social functioning, IQ, and academic achievement. (Faraone SV, Biederman J, Weber W, Russell RL. I Am Acad Child Adolesc Psychiatry Feb 1998;37:185-193).

MUSCLE DISORDERS

CONGENITAL MYASTHENIC SYNDROMES: END-PLATE AChR LACK

Two families with 5 affected members suffering from congenital myasthenic syndrome are reported from University Hospital, Bonn, Germany. The age at onset of ptosis, extraocular weakness, and exercise intolerance was in childhood or adolescence. The course was slowly progressive. The distribution of muscle weakness was more proximal than distal. All patients had abnormal decremental response on low-frequency 3-Hz nerve stimulation, and no repetitive responses to single nerve shock. All improved with anti-acetylcholinesterase drugs. Intercostal muscle biopsies from one patient in each family showed type 1 fiber predominance, and marked reduction in number of secondary synaptic clefts per neuromuscular junction and in the expansion of the postsynaptic area. Acetylcholine-receptor (AChR) density was reduced, and AChR-associated protein utrophin was deficient in the end-plate, on immunohistochemical analysis. Both patients showed a defect in the development or maintenance of the postsynaptic clefts. (Sieb JP, Dorfler P, Tzartos S et al. Congenital myasthenic syndromes in two kinships with end-plate acetylcholine receptor and utrophin deficiency. Neurology Jan 1998;50:54-61). (Reprints: Dr JP Sieb, Department of Neurology, University Hospital, Sigmund-Freud-St 25, D-53105 Bonn, Germany).

COMMENT. Since the clinical description of a *congenital* myasthenic syndrome (Millichap, JG, Dodge PR. Neurology 1960;10:1007), as distinguished from the *neonatal transient* form, several congenital myasthenic syndromes have been identified. Engel AG et al at the Mayo Clinic have reported patients and families with endplate acetylcholine and AChR deficencies, a slow-channel syndrome, defects in resynthesis of ACh and kinetics of AChR, and abnormal interaction of acethylcholine with its receptor. (see Progress in Pediatric Neurology III, 1997;pp346-347, for reviews of Mayo Clinic reports).

The Bonn families with congenital myasthenic syndrome had defects in the postsynaptic clefts with deficiencies in end-plate acetylcholine receptor and utrophin (dystrophin-related protein). The authors note that their patients resemble reports of a so-called congenital paucity of secondary synaptic clefts syndrome (CPSC) (Smit et al. 1988; Wokke et al. 1989). These familial syndromes have different clinical features and absent anti-AChR antibodies that distinguish them from autoimmune myasthenia gravis.