arrangements for hearing impaired children. They advocate early identification by screening, not only in the at risk children but universally. Testing only 'at risk' children leads to identification of less than half those affected. Regular school screening should help in diagnosis, and genetic counselling might lead to a reduction of numbers in the largest etiological group of cases. Hearing tests are important in the neurologic evaluation of children presenting with learning or attention deficit disorders, especially in those with a positive family history of deafness or a prior risk factor illness.

MUSCLE DISORDERS

ADHALIN DEFICIENCY IN MUSCULAR DYSTROPHY

A 13-year-old boy previously diagnosed with Becker's muscular dystrophy and dilated cardiomyopathy was studied at the University of Wisconsin, Madison, and the University of Iowa College of Medicine, Iowa City, and was found to have a deficiency of the dystrophin-associated glycoprotein, adhalin. He was asymptomatic until 9 years of age, when proximal weakness developed. He had flexion contractures at the ankles, hypertrophy of calf muscles, and Gower's sign. The serum creatine kinase level was 11,560 U/L. Both his sister and mother had normal CK. There was no consanguinity. Analysis of dystrophin from the biceps by Western blot was normal. Congestive heart failure required heart transplantation. Immunostaining in both skeletal and cardiac muscle showed normal dystrophin, whereas adhalin was reduced in skeletal muscle and absent in cardiac muscle. (Fadic R, Lotz BP et al. Brief report: Deficiency of a dystrophin-associated glycoprotein (adhalin) in a patient with muscular dystrophy and cardiomyopathy. N Engl J Med Feb 8, 1996;334:362-365). (Reprints: Dr Lotz, Department of Neurology, University of Wisconsin Hospital and Clinics, 600 Highland Ave, Madison, WI 53792).

COMMENT. Adhalin deficiency is an autosomal recessive disorder and is indistinguishable from the dystrinopathies by clinical presentation and muscle pathology. The authors propose that constituents of the dystrophin-glycoprotein complex (adhalin) and merosin should be analysed histochemically in all patients with histological findings suggestive of a dystrophinopathy and with normal muscle dystrophin. The dystrophin-associated glycoprotein was named "adhalin" from the Arabic *adhal* (muscle). It has been linked in North African populations to a gene in chromosome 13q, but the deficiency is genetically heterogeneous. The adhalin gene has been mapped to chromosome 17q. See Ped Neur Briefs Oct 1995, pp73-74, for reference to a further case report of primary adhalin deficiency in a 16-year-old African-American girl with childhood-onset limb-girdle muscular dystrophy.

FEBRILE CONVULSIONS

FEBRILE SEIZURE DURATION AND TEMPORAL LOBE EPILEPSY

Clinical features of febrile seizures and EEG findings were compared in patients who did and did not develop later afebrile seizures among six selected families and 59 family members with febrile convulsions examined at the Department of Clinical Neurological Sciences, University of Western Ontario, London, Ontario. All six probands developed epilepsy, 5 with temporal lobe epilepsy (TLE), after onset of febrile convulsions (FC). Of 59 family members