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

with FC, 8 (13%) developed TLE within an average of 12 years after the first FC and 4 (7%) had other seizures. Of 213 family members without FC, only 1 had TLE. The mean duration of FC was 100+/-133 min in those with later TLE and 9+/-19 min in patients without TLE at prolonged follow-up (mean 32 years). The total number of FC, the number in one day, and age at onset did not differ significantly between groups. Of 27 patients with FC who had EEGs, 11 (41%) had epileptiform records and all but one had epilepsy. Neuropathological examination of resected temporal lobes from 5 of the patients with prolonged FC and TLE revealed mesial temporal sclerosis. (Maher J, McLachlan RS. Febrile convulsions. Is seizure duration the most important predictor of temporal lobe epilepsy? Brain 1995;118:1521-1528). (Respond: Dr RS McLachlan, University Hospital, 339 Windermere Road, London, Ontario, Canada N6A 5A5).

COMMENT. The duration of the febrile convulsion was the most important determinant of the later development of epilepsy and epileptiform EEGs. This finding echoes previous publications showing that prolonged febrile convulsions and seizure discharges in the EEG are the most significant criteria of a poor prognosis. (Millichap JG et al. Studies in febrile seizures. V. A clinical and electroencephalographic study in unselected patients. Neurology 1960;10:643-653). Millichap, JG. Febrile Convulsions. A monograph. New York, Macmillan, 1968). Epilepsy and recurrent afebrile seizures developed in 30% of patients with prolonged febrile seizures and in only 5% of patients with short convulsions of less than 20 min. The incidence of paroxysmal EEG tracings in children who developed epilepsy following FC was five times that observed in children with uncomplicated febrile convulsions. EEG abnormalities occurred in 36% of patients with pronged FC >20 min and in 10% of those having short FC <20 min duration.

Berg AT and Shinnar S, examining complex febrile seizures (Epilepsia Feb 1996;37:126-133), found a strong correlation between prolonged duration of the FC and focal features, both in first and recurrent FC. Also, complex features tended to repeat, especially the prolonged duration, suggesting genetic or constitutional factors. The authors recommend that such children may be candidates for diazepam given at the onset of fever to abort the occurrence of a prolonged seizure. The following authors report a conflicting viewpoint, a not uncommon happening among authorities on this subject.

Knudsen FU et al, examining the long term outcome of prophylaxis for febrile convulsions (Arch Dis Child 1996;74:13-18), found that the prevention of new febrile convulsions by intermittent diazepam at the onset of fever offered no advantages over treatment with diazepam administered at the time of onset of a seizure. The long term prognosis in terms of subsequent epilepsy, neurological, motor, intellectual, cognitive, and scholastic ability was not influenced by the type of treatment applied in early childhood.

ELECTROLYTE ABNORMALITIES IN FEBRILE SEIZURES

The role of serum sodium in susceptibility to complicated febrile convulsions was studied in 115 children admitted with simple or complicated febrile convulsions to the Kuopio University Hospital, Finland. Sodium levels were lower in children with complex FC in comparison with those having simple convulsions. The means were 136.07 (n= 42) and 137.62 mmol l^{-1}(n=71), respectively. Sodium levels were lowest in children with repeated seizures. Levels <135 occurred in 47% of children with repeated FC and only in 8% of those with simple FC, but 50% of these simple FC cases had later complicated, repeated seizures, status epilepticus, or they developed epilepsy within 3 years.