

weakness and/or wasting, ataxia, dysarthria, and sensory impairment. Impaired autonomic function in 11 patients was manifested by postural hypotension and anisocoria. Eleven were intellectually impaired. Ten had hyperkeratosis and fissures of their palms. Siblings were affected in 3 families, and 7 siblings had died in childhood. (Grant DB et al. Neurological and adrenal dysfunction in the adrenal insufficiency/achalasia/achalasia (3A) syndrome. Arch Dis Child 1993; **68**: 779-782). (Respond: Dr DB Grant, Hospital for Sick Children, Great Ormond Street, London WC1N 3JH).

**COMMENT.** The most frequent presenting symptom was hypoglycemia, often associated with Addisonian skin pigmentation. In 4 cases the diagnosis of achalasia preceded the recognition of cortisol deficiency. The authors recommend a test of cortisol secretion in children with achalasia, if associated with alacrima or with any of the above neurologic abnormalities.

### INFECTIOUS DISORDERS

#### **HEMOPHILUS INFLUENZAE VACCINE AND GUILLAIN-BARRE**

A four-year-old girl with Guillain-Barre syndrome that developed 10 days following *H influenzae* type b conjugate vaccine immunization is reported from Geneva, Switzerland. CSF contained 1 mononuclear cell/cmm and 0.89 g/l protein. Nerve conduction velocities were decreased in upper and lower extremities. Measurements of immunoglobulins against the polysaccharide (PRP) component of the vaccine showed a marked elevation (100 mcg/ml) of the anti-PRP IgM antibody level. Following treatment with two 5-day courses of intravenous immunoglobulins (0.4 g/kg/day) 2 weeks apart, cranial nerve function and muscle strength improved. Apart from absent deep tendon reflexes, the neurological exam was normal at one month follow-up. (Gervais A et al. Guillain-Barre syndrome following immunization with *Hemophilus influenzae* type b conjugate vaccine. Eur J Pediatr July 1993; **152**: 613-614). (Respond: Dr A Gervais, Children's Hospital, University Hospital of Geneva, 30 Blvd de la Cluse, CH-1211 Geneva 4, Switzerland).

**COMMENT.** A total of five cases of Guillain-Barre syndrome (GBS) following vaccination with the *H influenzae* type b diphtheria toxoid-conjugate vaccine have been reported. The authors speculate that an excessive anti-PRP IgM antibody response to the vaccine might explain this rare complication.

Treatment of acute GBS is controversial and not without risk, especially in patients with vascular disease. Two adults died from vascular thrombotic complications following gamma globulin therapy for GBS. (McFarland HR. Arch Neurol July 1993; **50**: 687).