syndrome in children (table 2). Although we found no mortality, our survey did not significantly differ from those reported earlier. The frequency of the need for assisted ventilation of 5/27 is a common figure, indicating that our patients represent a typical series of cases of Guillain-Barré syndrome despite being based on a nationwide survey.

Although Guillain-Barré syndrome is a rare disease with a relatively good long term outcome, it causes a long period of hospitalisation and a prolonged need for rehabilitation. The Guillain-Barré syndrome is still a serious and sometimes life threatening condition.

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Commentary
Rantala and colleagues report an age specific annual incidence of 0.38/100 000. Similar figures have been reported from Denmark,1 Norway,2 and the USA.3 The last mentioned study reported a linear increase in incidence with increasing age throughout childhood and adult life. These reports, however, can only describe the course of the condition in cases in whom the correct diagnosis is made.

Hospital based reports have emphasised the difficulties of diagnosis, especially in children. Three of the last five personal cases have been referred for the first time after cardiac arrest and still bearing other diagnoses. Of the remaining two, one had been misdiagnosed as encephalitis and the other had prominent pain and cramps, preserved reflexes and reasonable preservation of power at the time when cerebrospinal fluid examination and neurophysiology confirmed the diagnosis.

The classical signs of limb weakness and hyporeflexia may be unimpressive early in the illness. Again, abnormalities of cerebrospinal fluid protein and nerve conduction velocity may not be striking early in the illness. The Guillain-Barré study group listed,4 in addition to those features tabulated by Rantala et al, the following other clinical features strongly supportive of the diagnosis: mild sensory signs or symptoms, autonomic dysfunction, initial absence of fever, and relative symmetry of impairment.

The diagnosis should be considered in any child with acute or subacute unexplained ataxia, cranial nerve palsies, weakness or pain in the trunk and/or limbs. Patients with Guillain-Barré syndrome may be misdiagnosed as having encephalitis (which may also accompany the Guillain-Barré syndrome), myositis, cachexia, arthralgia, arthritis, transverse myelitis, behaviour disorder, or hysteria. Prominent cranial nerve involvement may lead to an apparent absence of brainstem reflexes and be misinterpreted as coma, particularly if accompanied by limb paralysis. The Miller-Fisher variant may present with cranial nerve involvement ataxia and proprioceptive loss without detectable motor weakness.

The differential diagnosis of subacute polyneuropathy includes glue sniffing, diphtheria, botulism, heavy metal poisoning, polyomyelitis, organophosphate poisoning, myasthenia gravis (tick paralysis), and rabies.

The series of Rantala et al is similar to others in reporting the need for assisted ventilation
Occurrence, clinical manifestations, and prognosis of Guillain-Barré syndrome

in between 10–20% of cases. Such severely affected cases have an appreciable mortality. Meticulous intensive nursing and medical care are essential for proper management. Pneumonia, autonomic instability, pulmonary embolism, hyponatraemia, and encephalopathy are well recognised complications. Cardiac arrest, particularly precipitated by external stimuli, such as tracheal suction, was reported by Cole and Matthew in five of 11 patients (including two with a more chronic polyneuropathy) requiring ventilation. In the presence of substantial fluctuations in blood pressure, central venous pressure monitoring is essential, particularly as excessive sweating and disturbed gut function may also increase parenteral fluid requirements. Oral fluids should be avoided because of the risk of aspiration and consequent respiratory failure. Total paralysis without impairment of awareness can be a terrifying experience: reassurance and good communication with the patient are therefore especially important. Sedatives may be helpful, although cardiovascular and respiratory side effects may be potentiated by the neuropathy.

Although disability after recovery is exceptional in children (and in most adult series), plasmapheresis has been justified in non-ambulant adults on the grounds that it speeds recovery and shortens the duration of hospital stay substantially. Epstein and Sladky reported a mean time to independent ambulation of 25–4 days in nine children treated with plasmapheresis compared with 60–2 days in 14 historical controls. In that study, only one child in each group required ventilation. However, plasmapheresis has its own morbidity, particularly when applied to infants. Steroids are not helpful and may predispose to relapse. Recently, intravenous gammaglobulin has been reported to be helpful in Guillain-Barré syndrome by Kleyweg et al., and subsequently by Shahar et al., using doses ranging from 0.4–1 g/kg/day in a small number of children. Further controlled studies are needed before routine use of these treatments can be recommended for childhood Guillain-Barré syndrome.

In summary, Rantala and colleagues report that Guillain-Barré syndrome is uncommon and that complete recovery is the rule. However, there may be an unmeasured mortality in undiagnosed cases: a high index of suspicion is required.

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The Herbst triad
Gastro-oesophageal reflux may present in unusual ways. Perhaps the most unusual is Sandifer’s syndrome but asthma and recurrent apnoea in infancy are well known. A report in the Journal of Pediatric Surgery (P Sacker, U G Stanfler, 1990;25: 1238–9) describes two children with another unusual syndrome, previously unknown to me, the Herbst triad. This consists of finger clubbing, hypoproteinaemia due to protein-losing enteropathy, and gastro-oesophageal reflux but it could be expanded to a tetrad with the addition of severe iron deficiency anaemia.

The clubbing and the hypoproteinaemia resolved after fundoplication suggesting that the enteropathy is secondary to the reflux. The mechanism is not known but the authors suggest that gastro-oesophageal reflux may lead to an alteration in gut flora which in turn might cause the enteropathy. Some of the patients have the head and neck movements of Sandifer’s syndrome. Who will tell me the cause of the clubbing?

ARCHIVIST