home (97%). Here the mothers did not see the explanations in the Baby Check booklet, so they probably used less stringent criteria for each symptom/sign, and overscored. All three controls scoring 13–19 (table 2) had either green vomit (13 points) or recession (15 points), which are very rarely seen in the community. These would probably have been reported absent with Baby Check, where parents are told not to score items if in doubt.

The results suggest that Baby Check could help parents recognise a small proportion of babies at risk of sudden unexpected death. Babies who die suddenly and unexpectedly are more likely to be taken to the general practitioner in the week before death than control babies. This difference is unrelated to reported symptoms, but parents clearly react to their perception of the babies’ well being. If such families used Baby Check they might be empowered to obtain timely medical help, so that some of the babies would not die.


Guillain-Barré syndrome in three siblings less than 2 years old

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Abstract

Three of five children who were born to consanguineous parents developed Guillain-Barré syndrome before they were 3 years old. The syndrome is rare in early childhood and we suggest that there may be a genetic element in the pathogenesis.

Polyradiculoneuritis (Guillain-Barré syndrome) has become the most common form of acute paralysis since the decrease in the incidence of poliomyelitis. Though the incidence in children seems to be increasing, it is still rare below the age of 2 years. 2-3 There have been only three reports of familial Guillain-Barré syndrome in adults, and none in children, to our knowledge. 4-6 We describe three siblings from a consanguineous marriage who developed Guillain-Barré syndrome when they were less than 2 years old. The parents were young, healthy Arab Moslems who were second cousins and had five children (three boys and two girls). Their second, fourth, and fifth children are described in this report; the other two have always been healthy.

Case reports

CASE 1

A 2 year old boy was admitted to another hospital in 1980 with progressive weakness and difficulty in swallowing. Two weeks before admission he had a short febrile illness and a cold. On admission he was pale and in a 'poor general condition', with drooling, generalised hypotonia and hyporeflexia. Results of laboratory tests did not indicate a diagnosis.

The child was treated with corticosteroids and intravenous fluids replacement. His condition deteriorated and he died on the sixth day in hospital with the presumptive diagnosis of encephalitis.

CASE 2

The younger sister of case 1 was admitted to this hospital in 1984 at the age of 22 months because of weakness in the lower limbs. One week before admission she had had pharyngitis, and subsequently she became sleepy and developed unstable gait. Neurological abnormalities included ataxic gait, inability to stand, and areflexia. Examination of the cerebrospinal fluid showed no cells, glucose 3.1 mmol/l, and a protein concentration of 1.6 g/l. The electroencephalogram was interpreted as normal. Electromyography showed reduced nerve conduction velocity with prolongation of distal motor latency. Poliomyelitis was ruled out. HLA typing failed to show the DR3, A3, and B8 antigens.

Within a week she started to improve spontaneously, and at follow up examination two years later she had no neurological deficit.

CASE 3

The younger brother of case 2 was admitted to the paediatric intensive care unit in 1987 at the age of 9 months with respiratory failure follow-
Guillain-Barré syndrome in three siblings less than 2 years old

Guillain-Barre syndrome in three siblings less than 2 years old

ing progressive generalised weakness. Two weeks before admission he had had an episode of fever and rhinorrhea. On admission he was fully awake and had pronounced hypotonia with flaccid, almost complete, paralysis of all extremities. Deep tendon reflexes were absent. Examination of cerebrospinal fluid showed no cells. The concentration of protein was 0.6 g/l, glucose 2.9 mmol/l, and myelin basic protein 0.16 g/l (normal <0.07). He had a strong positive reaction to oligoclonal antibodies to central nervous system proteins. Red cell cholinesterase was 12.8 IU/ml (within the reference range). Electromyography showed severe neuropathy and a peripheral nerve demyelination pattern. Culture and antibody screens of cerebrospinal fluid were negative for all viruses, and tissue typing did not show the DR3, A3, or B8 antigens.

The child's general and neurological condition improved gradually, and he was weaned from mechanical ventilation after 40 days. Subsequently he showed slight but slow improvement, but at 3-5 years of age he had pronounced hypotonia, areflexia in all limbs, and was unable to stand or walk unsupported.

Discussion

The clinical features and laboratory findings in cases 2 and 3 fitted the criteria for Guillain-Barré syndrome.1 They had the typical cytoalbumin dissociation and peripheral nerve demyelination pattern on nerve conduction studies. Case 1 presented with a clinical picture identical to that of case 3, and we suspect that he died of respiratory failure caused by respiratory muscle paralysis and that the diagnosis of encephalitis was wrong.

Other possible causes of acute onset muscle weakness, such as poliomyelitis, diphtheria, botulism, and poisoning by organophosphates, lead, or hexacarbons were ruled out by the history, results of laboratory tests, and clinical course.

Guillain-Barré syndrome occurs rarely in infancy and early childhood.2-3 In a series of 30 children with Guillain-Barré syndrome only three were less than 2 years of age and in the series of Peterman et al only three of 26 were less than 2 years of age.4 The youngest patient in a recent series of 157 cases from Massachusetts General Hospital was 8 years old.2 Patients younger than 1 year of age (as case 3) were not reported.

Familial Guillain-Barré syndrome has not to our knowledge been reported in children, and we could find only three instances in adults.4-6 Two elderly siblings with painful, slow onset paralysis of all four limbs and raised concentrations of protein in the cerebrospinal fluid have been described.4 However, symptoms were initially sensory (severe pain in the lumbar region and buttocks), progressed over months, and were not associated with autonomic dysfunction. Such findings do not fit well with the diagnostic features established by the Guillain-Barré syndrome study group.1 Another report described a father and a 24 year old daughter whose presentations were compatible with Guillain-Barré syndrome.5 A third report described two brothers who acquired a paralytic disease while working in Gabon and taking chloroquine;6 both had raised concentrations of protein in the cerebrospinal fluid.

The occurrence of such a rare disorder in three siblings born to healthy, consanguineous parents suggests the presence of a genetic defect that is transmitted in an autosomal recessive way. An abnormal immune response against the peripheral nervous system has been implicated in the pathogenesis of the Guillain-Barré syndrome. The syndrome is often triggered by a previous viral infection or by certain immunisations. It has clinical and pathological features similar to those found in experimental allergic neuritis, which is an autoimmune disease that can be produced in animals by injecting peripheral nerve antigens. The considerably increased concentrations of immunoglobulin in the cerebrospinal fluid, sometimes with raised oligoclonal bands (as in case 3) also imply an alteration in local immunoregulation. Lymphocyte reactivity to myelin components of peripheral nerves has been reported in patients with Guillain-Barré syndrome, but the evidence that the pathogenesis has an immunological basis is indirect and circumstantial.

Disorders associated with immune aberrations are sometimes related to the HL A system. A few studies have reported an increased incidence of class I A3 and B8 and class II DR3 antigens in Guillain-Barré syndrome.7 As these specific antigens were not isolated in this family, the HL A system cannot be implicated.

This evidence suggests that genetic factors may have a role in the still uncertain aetiology of this disorder.

We thank Professor Oded Abramski and Dr Talma Brenner (laboratory of neuroimmunology, department of neurology, Hadassah University Hospital, Jerusalem) for performing the immunological tests on the central nervous system. We thank Mrs Onat Gideon (department of immunology, Rambam Medical Center and the Technion Faculty of Medicine, Haifa) for doing the tissue typing studies.

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Arch Dis Child 1991 66: 1078-1079
doi: 10.1136/adc.66.9.1078

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