

Prognosis in severe Guillain-Barré syndrome

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SUMMARY Recent studies of Guillain-Barré syndrome in adults have shown that the requirement for assisted ventilation correlates closely with a poor outcome, and the need for ventilation is now being used as an indication for plasmapheresis. As comparable studies in children have not been performed we reviewed our experience of patients who had Guillain-Barré syndrome severe enough to require assisted ventilation. In a group of 11 children two died in the acute stage of the illness and nine made an excellent recovery. It would seem that the need for mechanical ventilation is not necessarily a bad prognostic factor for neurological recovery in children.

Guillain-Barré syndrome affects at least 1.7 per 100 000 of the population every year.¹ This inflammatory demyelinating disease of peripheral nerves is often thought to have a benign prognosis, but roughly 7% of adult patients die and a further 7–22% suffer residual disability.^{2–4} No comparable figures are available for Guillain-Barré syndrome in children, although there is some evidence that severe residual deficits are rare and that the natural history of the disease in childhood may be different.⁵

There have been several recent multicentre studies on the treatment of Guillain-Barré syndrome in adults,^{2 6 7} and these have shown that steroids are ineffective^{6 7} but that plasmapheresis is beneficial both in shortening the duration of acute illness and improving neurological outcome.^{2 8} Reservations have been expressed, however, about using plasmapheresis indiscriminately in a disease where most patients will recover spontaneously and where the technique itself is not without hazard.^{9 10} Selection for treatment of those patients who would otherwise do badly has thus been advocated.^{9 10} Winer *et al* in a recent retrospective study of 71 adults have attempted to identify such patients.¹¹ In their study those patients who required assisted ventilation were shown to be the most likely to have an incomplete neurological recovery. As a result of these recent findings we have reviewed our experience, with particular reference to outcome, of those children with severe demyelinating polyneuropathy who required ventilation.

Patients and methods

During the seven years 1977–84, 11 children with

demyelinating polyneuropathy required assisted ventilation at our hospital. There were six boys and five girls in the series, aged from 3 to 13 years. Details of the acute illness were obtained from case notes. The survivors were reviewed between 15 months and six years after the onset of illness. At review, a questionnaire was completed in which details of school performance, duration of rehabilitation, functional neurological deficit, and subsequent respiratory problems were sought. The children were weighed, measured, examined, and given a detailed physiotherapy assessment, which included charting of muscle strength. Pulmonary function tests were performed. Lung volumes were assessed by helium dilution technique and full body plethysmography. Maximum expiratory flow volume curves were recorded by Fleisch pneumotachograph and from these the forced vital capacity, forced expiratory volume in one second, and volume maximum 25% vital capacity were calculated. Peak expiratory and inspiratory mouth pressures (an indication of respiratory muscle power) were measured. All children had diaphragmatic movement assessed by radiological screening.

Results

Acute illness. Nine of the children presented with a history and clinical findings of classical Guillain-Barré syndrome and the other two children (cases 10 and 11) showed the more protracted natural history of chronic, progressive/relapsing polyneuropathy (Table 1).

At presentation all the children had cerebrospinal fluid findings compatible with a diagnosis of demyelinating polyneuropathy, and in those five chil-

Table 1 Clinical features of acute Guillain-Barré syndrome in 11 children. Cases 10 and 11 had chronic relapsing polyneuropathy

	Case No										
	1	2	3	4	5	6	7	8	9	10	11
Cranial nerve involvement	+										+
Encephalopathy	+			+		+		+			
Hypertension		+			+						
Cardiac arrest		x2	+								
Treatment					1 Steroids			Steroids	Steroids	1 Steroids, immunosuppressants, plasmapheresis	1 Steroids, immunosuppressants, plasmapheresis
Outcome									Death		Death

dren in whom nerve conduction studies were performed there was evidence of slowing of conduction. Seven of the 11 children had abnormal chest x ray films at admission (Table 2).

Five children were being ventilated on admission to our unit and the others were referred because respiratory failure was thought to be imminent. Eight of the children required ventilation within two weeks of onset of illness, and the duration of ventilation ranged from three days to five weeks. Five children were treated with steroids for at least part of their illness, although no standardised regimen was employed, and the two children with the relapsing form of the illness were treated with immunosuppressants and plasmapheresis as well. Two children died, one with the acute illness and the other with the progressive relapsing form of the disease.

Follow up assessment. The nine survivors all had normal pulmonary function tests at review, apart from one boy who had developed asthma and had a mild obstructive defect. Respiratory muscle function was unimpaired in all the children.

All the children had returned to normal school after their illness. On examination, seven of the nine children had made a complete neurological recovery. The parents of these seven thought that complete recovery had been achieved within a few months of the onset of illness. One of the two children who had not made a complete neurological recovery (case 10) had minimal weakness of the hip extensors and the ankle dorsiflexors and plantar flexors but negligible functional disability. The other child who had not made a complete neurological recovery (case 3) had minimal wasting of the small muscles of the hands, feet, and calf muscles and there was weakness of the quadriceps, hamstrings, calf muscles, and intrinsic muscles of the feet. In this boy charting of muscle strength in the lower limbs ranged between 1 and 4 on the Medical Research Council scale. The boy was unable to walk on his heels and had a waddling gait but nevertheless could walk a mile without tiring and ride a bike for several miles (Table 3). There was no sensory deficit or autonomic dysfunction in any of the nine children.

Discussion

Study of the acute illness in our patients revealed some interesting facts. Seven of the 11 children had evidence of pulmonary collapse or consolidation on chest radiograph at the time of admission, indicating that the dangers of oral feeding in the presence of bulbar involvement and poor respiratory function are still not sufficiently appreciated. The degree of

Table 2 Aspects of ventilation during acute Guillain-Barré syndrome in 11 children. All figures denote days

	Case No										
	1	2	3	4	5	6	7	8	9	10	11
Initial chest radiograph	A	A	A	N	A	A	N	N	A	N	A
Onset of ventilation	6	7	4	5	6	40	4	10	12	210	180
Duration of ventilation	3	42	56	11	18	21	3	10	21	34	20
Onset of improvement	9	22	24	8	10	54	5	15	—	240	198
Plateau time	3	15	20	3	4	14	1	5	—	30	18

A=Abnormal; N=normal.

Table 3 Clinical features in the two survivors with neurological impairment

	Muscle wasting	Weakness	Deep tendon reflexes	Functional disturbance
Case 3	Small muscles, hands, distal muscles of lower limbs	Interossei, distal muscles of lower limbs	Absent supinator, knee, ankle	Heel walking
Case 10	None	Hip extensors, ankle flexors, ankle extensors	Absent ankle	Heel walking

radiological abnormality was not gross in any child (the most severe being lobar collapse or consolidation) and not sufficient to account for respiratory failure per se. Weakness of the respiratory musculature led to hypoventilation and atelectasis, and this was aggravated in many cases by aspiration or overspill due to bulbar failure. Although details have not been provided in the adult studies, we believe that for patients of any age with Guillain-Barré syndrome, developing respiratory failure is often worsened by an episode of aspiration and may indeed precipitate the need for mechanical ventilation.

All the children had bulbar impairment, but in six there was additional cranial nerve involvement, most often unilateral or bilateral facial weakness. Five developed an encephalopathy, and although this has previously been reported, it is not a commonly recognised feature of the disease.¹² Autonomic neuropathy was prevalent, with hypertension, hypotension, and sphincter disturbance among the commonest symptoms. Cardiac dysrhythmias are the most hazardous form of autonomic dysfunction. These may occur spontaneously but more often are provoked by some manipulation, such as changing a tracheostomy tube or induction of anaesthesia.^{13 14} Six cardiac arrests occurred in the group (two in one child), and we think that there should be greater awareness of this common complication of the illness.

There were two children in our group with the rare chronic form of the disease (cases 10 and 11). This pursues a slowly progressive or relapsing course and its exact relation to acute demyelinating neuropathy is uncertain.^{15 16} Pathological findings in the two illnesses are identical; it is the tempo of clinical

evolution that is strikingly different. The peak of disability is usually reached slowly in the chronic form (after six to 12 months), with a plateau of disability that may last for many months. Recovery from second and later relapses is usually slow and incomplete.

There were two deaths. One girl had the syndrome with a concurrent endocarditis as a result of overwhelming coxsackie B infection. She developed intractable heart failure and postmortem findings confirmed the presence of an endocarditis and demyelination in the dorsal roots of the spinal cord. The second girl (case 11) had the chronic relapsing form of the disease and developed irreversible ventricular fibrillation during induction of anaesthesia for intubation.

At follow up of the nine survivors it became clear that there was little evidence of persisting functional impairment. All the children assessed had impeccable respiratory function (apart from one child with mild asthma) and none had recurrent chest infections. In the two children who had abnormal neurological signs one had minimal functional deficit and the other's ability to lead a normal life was only mildly affected.

In Winer's study on adults outcome was graded according to a simple objective of functional ability at 12 months after illness, as follows:¹¹

- (0) Healthy.
- (1) Minor symptoms or signs.
- (2) Able to walk five metres without help, walking frame, or stick but unable to do manual work, including shopping or gardening.
- (3) Able to walk five metres with help, walking frame, or stick.
- (4) Chair or bed bound.

(5) Requiring assisted ventilation (for at least part of day or night).

(6) Dead.

Patients who were still restricted in their activity (with a functional grade of 2 or greater at this time) were considered to have a poor outcome; a score of 0 or 1 denoted a good outcome. Twenty nine patients in his adult series were ventilated and 16 (55%) scored badly. By contrast, considering our nine children with the classical form of Guillain-Barré syndrome, allowing for the one death in this group, all our eight survivors scored very well (functional grade 0 or 1). Although our highly selected group is small, outcome in our children thus seems at variance with the adult experience. We acknowledge that there are inherent difficulties in making a direct comparison between our group of patients and the adult series, particularly in a retrospective study, but it does seem that children requiring ventilation for Guillain-Barré syndrome do well, and the need for ventilation need not necessarily be considered a bad prognostic factor.

Management of this condition remains controversial. Although five of our children received steroids at some time during their acute illness, several recent large trials in adults have shown such treatment to be of no benefit in this condition.^{6,7} There is some anecdotal evidence that chronic demyelinating neuropathy responds to plasmapheresis,^{15,17} and in our two patients with this disease we performed plasmapheresis and used immunosuppressants.¹⁸ The patient who survived seemed to respond appreciably to this treatment and has made an almost complete recovery (case 10). A recent American multicentre trial has shown plasmapheresis to be effective in acute Guillain-Barré syndrome in adults, both in reducing duration of the disease and improving neurological outcome.² As most adult patients recover spontaneously, however, it has been advocated that the potentially complicated, uncomfortable, and costly treatment of plasmapheresis should only be employed in those patients likely to have a poor outcome—that is, most of those requiring ventilation.^{9,10}

The findings in our group suggest that the outcome in severe Guillain-Barré syndrome in childhood is much more favourable than in adults. We believe that provided the child survives the acute illness the prognosis for good functional recovery with intensive supportive measures only is excellent.

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