J. Neurol (1981) 225: 207–217

Journal of
Neurology

Treatment of Guillain-Barré Syndrome by Plasma Exchange*

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Summary. Plasma exchange has been used for therapy in eight patients with the Guillain-Barré syndrome. All patients were severely ill. They became tetraplegic and showed cranial nerve involvement. Five patients received assisted respiration, but the others were also at risk of ventilatory insufficiency. Recovery was abrupt in all cases after the first plasma exchanges. Improvement was more marked when plasmapheresis was done on three successive days with plasma exchanges of 2.0–3.0 l each in the initial progressive stage of the disease. A considerable advantage of this therapy is the avoidance of continued artificial respiration and nutrition, which both carry the risk of further complications.

Key words: Plasma exchange – Guillain-Barré syndrome – Artificial respiration – Recovery

Zusammenfassung. Die Wirksamkeit einer Plasmaaustausch-Behandlung wurde bei 8 Patienten mit einem Guillain-Barré-Syndrom geprüft. Alle Patienten hatten einen schweren Krankheitsverlauf gezeigt, mit schlaffen Tetraparesen und multiplem Hirnnervenbefall. Fünf Patienten mußten assistiert beatmet werden. Auch bei den anderen Patienten zeigten sich Hinweise für eine drohende respiratorische Insuffizienz. Nach den ersten Plasmapheresebehandlungen konnte eine abrupt einsetzende und deutliche Besserung der Ausfälle beobachtet werden. Die klinische Besserung verlief besonders eindrücklich, wenn der Plasmaaustausch an drei aufeinanderfolgenden Tagen mit einer Austauschmenge von jeweils 2,0–3,0 l und in der initialen progressiven Phase der Erkrankung erfolgte. Ein großer Vorteil dieser Therapie dürfte in der Vermeidung langzeitiger künstlicher Beatmung und Ernährung und den damit verbundenen Komplikationen liegen.

* Parts of the paper were presented at the International Symposion on Plasma Exchange Therapy, Wiesbaden, April 15/16, 1980 [23]

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0340-5354/81/0225/0207/$ 02.20

Introduction

The Guillain-Barré syndrome (GBS) is an acute polyradiculoneuropathy of unknown etiology [16]. The occurence of GBS after injection of rabies vaccine containing nervous tissue has supported the view that the disorder might be mediated through a common immunologic mechanism [25]. Experimental allergic neuritis has shown striking similarity with the disease in humans [5, 9]. The immune pathogenesis of GBS was further supported by the finding of complement fixing antibodies [24], of precipitating antibodies against trypsinized white matter extracts [29] and of myelinotoxic serum antibodies of the IgM class in patients with GBS [10]. Convincing evidence is still lacking that serum antibodies play the primary role in GBS [4, 31]. Cellular hypersensitization to peripheral nervous antigens presented by circulating immunoblasts and lymphocytes supported the role of cellular mechanisms in pathogenesis [2, 6, 21].

The role of treatment with prednisone or ACTH remains controversial [14, 15, 18, 19, 32]. There is also no striking success in therapy by the use of azathioprine [12]. In view of this lack of effective treatment and of the pathogenetic role of myelinotoxic antibodies Brettle et al. [7] tried plasma exchange in one patient with GBS. The good recovery of this patient suggested the use of plasma exchange for treatment of GBS in other patients.

Methods

Plasma exchanges were done using a Haemonetics 30 cell separator. The cells were reinfused into the patient. One third to one half of the plasma was replaced by prewarmed deep frozen fresh plasma, the rest was substituted for 5% human albumin solution with proper electrolyte adjustment. Up to 15 plasma exchanges varying from 0.5 to 3.0 l each in varying one day to one week intervals were undertaken. Cardiovascular problems, but also problems in coagulation and allergic reactions made it necessary to interrupt plasma exchange and therefore influenced the amount of exchanged plasma.

Case Reports

Case 1. A 66-year-old male patient with no history of previous illnesses became ill with weakness of both his legs 10 days after a mild afebrile upper respiratory tract infection and four days after influenza vaccination. The weakness progressed rapidly to involve the arms. On admission, on the second day of disease, the patient was already tetraplegic including mild bilateral facial nerve palsy with complete abolition of the tendon reflexes. The pupils and ocular movements were normal. Sensory examination revealed no abnormality. Two days later, the patient had to be tracheotomized and needed assisted ventilation for the next three weeks. After this period spontaneous respiration was regained. At this time the first 1.5 l plasma exchange was tried. One day later, the patient was able to swallow. A second 1.5 l plasma exchange was performed in the following week. A further slight improvement appeared with return of some finger movements. One week later, the patient started to move both his arms and legs. The patient underwent a total of ten 1.5 l plasma exchanges. Four months later, the patient was able to walk with support. Eight months later, there only was slight weakness of the extensors of the left toes.

The CSF on admission contained 36 mg/dl of protein and 1/mm^3 lymphocytes. A repeat lumbar puncture revealed 1/mm^3 lymphocytes and a total protein of 112 mg/dl two months later.

General examination, chest-X-ray, ESR, urine composition, full blood count and serum electrophoresis revealed normal findings. At no time did the patient receive corticosteroids or ACTH.

Case 2. A 28-year-old male patient suffered from rather frequent upper respiratory tract infections. He noticed weakness of both legs followed by a feeling of numbness in his arms within 24 h. On the third day of his illness he found it hard to speak and to swallow. On admission there was generalized weakness including bilateral facial nerve palsy with complete abolition of the tendon-reflexes. The patient was still able to stand and walk. Speech was slightly slurred. The sensory examination showed a slight distal impairment of the sense of joint position and of vibration in his legs. Over the next three days weakness progressed and the patient needed artificial respiration over a period of three weeks. Flaccid paresis of all limbs and multiple cranial nerve dysfunction including the ocular movements persisted. After the first of two 1.5 l plasma exchanges at a two days interval, spontaneous respiration was regained. The patient underwent a total of fifteen 1.5 l plasma exchanges. Further treatment was complicated by infective hepatitis with full recovery within three weeks. Three weeks after the first plasma exchange the patient could raise his arms against gravity and make fists on both sides. He was still unable to elevate his legs. Two months later the patients was ambulant, although general weakness persisted. Corticosteroids or ACTH were not given.

The CSF on admission contained 34 mg/dl of protein and 1/mm^3 lymphocytes. Nine days after the initial symptoms lumbar puncture was repeated. The CSF now contained 96 mg/dl of protein and 1/mm^3 lymphocytes.

Apart from signs related to his infective hepatitis general examination was normal. A chest-X-ray, ESR, full blood count, urinalysis and serum electrophoresis revealed normal findings. Immunoglobulins showed a small IgM-gradient.

Case 3. A 53-year-old male patient was known to suffer from hypertension due to polycystic kidneys with clinical signs of uremia. There was no antecedent febrile illness. Ten days before admission the patient developed bilateral deafness accompanied by tinnitus and vertigo. On admission bilateral hearing loss and bilateral facial nerve palsy were noted, together with an incomplete oculomotor palsy on both sides and bilateral trigeminal nerve involvement. There was no alteration of consciousness, but speech was slurred and the patient was unable to swallow. There was no weakness in the extremities, but fine movements were poorly performed. Sensory examination revealed a slight sensory disturbance distally in the lower limbs with absent ankle jerks. The other tendon reflexes were normal. Pyramidal signs were absent. During the next few days a transient paralysis of conjugate vertical ocular movements occurred and was accompanied by vertical nystagmus. Within 14 days after the initial symptoms the patient developed generalized weakness of all four extremities, starting in the distal muscles, and finally became tetraplegic. At the time all tendon reflexes were abolished and no pyramidal signs could be elicited. In anticipation of respiratory insufficiency the patient underwent three successive 3.0 l plasma exchanges. Rapid improvement occurred after the first exchange. All cranial nerves showed good recovery with the exception of the eighth. One month later, the patient was fully ambulant, but extreme hypacusis remained.

The CSF on admission contained 450 mg/dl of protein and 5/mm^3 lymphocytes. Twelve days later a repeat lumbar puncture showed 97 mg/dl of protein and 1/mm^3 lymphocytes. Motor conduction velocity in the right peroneal nerve was 40 m/s, distal motor latency 5.6 ms (7 cm); on the left the values were 35 m/s, distal motor latency 7.2 ms (7 cm).

General examination revealed hypertension due to chronic renal failure and hypertensive heart disease. The course of the disease was complicated in that the patient had lived for many months with severe uremia and had not been dialyzed nor had had a restricted intake of protein. The plasma urea was elevated to 216 mg/dl and creatinine to 9.1 mg/dl, with only slight variations during the course of the acute polyneuropathy. After recovery from the polyneuritis the patient was further treated by chronic hemodialysis. Serum electrophoresis and immunoglobulins were normal. ESR was elevated to 82/116 on admission and decreased to 13/35.

The patient was treated with 100 units ACTH daily for the first five days. There was no response to this therapy and the patient worsened. During the next three weeks methyl prednisolone 20 mg daily was given.

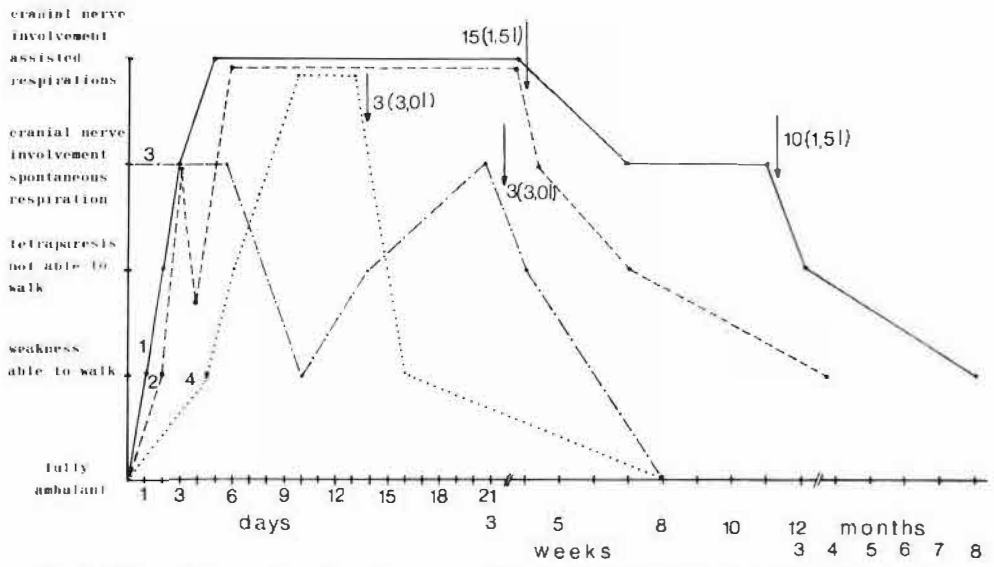

Fig. 1. Effect of plasmapheresis on the course of the Guillain-Barré syndrome in cases 1–4. First plasma exchange is marked by arrow, accompanied by number and amount of plasma exchange. The severity of the illness is classified in five stages. Note that the time scale is interrupted twice. In case 3 the disease started with cranial nerve involvement and then spread to the peripheral nerves

Case 4. A 28-year-old female patient had a history of a febrile infection three weeks before she became ill. The patient complained of pain, numbness and tingling in the limbs more pronounced on the left arm. During the following four days weakness developed in the left leg and then spread to the right. On admission she showed slight paresis of the upper and lower extremities, more severely proximally. The tendon reflexes were reduced. There was no sensory loss. Eight days after the onset of the illness the patient had developed a severe tetraparesis, bilateral facial weakness and an increasing bulbar palsy. Intensive care was needed because of ventilatory failure 2 days later. The patient needed assisted respiration for 3 days. During this time she underwent three successive 3.0 l plasma exchanges. Rapid recovery was seen, and the patient was able to walk and to eat within the following 3 days. One month later, the patient only showed slight weakness in the left hand and of the lower limbs, most pronounced in the distal muscles.

The CSF on admission contained 16 mg/dl of protein and 4/mm^3 lymphocytes. Four days later, the CSF contained 67 mg/dl of protein and 1/mm^3 lymphocytes. After one month CSF revealed 115 mg/dl of protein and 3/mm^3 lymphocytes.

General examination, chest-X-ray, ESR, full blood count, serum electrophoresis and immunglobulins were normal. During the period or her deterioration the patient received no ACTH or corticosteroids. However, 28 days after the plasma exchanges ACTH 100 daily units, followed by 50 units daily, were given over a period of two weeks (Fig. 1).

Case 5. A 29-year-old female patient complained of burning sensations in her left arm 3 weeks after a febrile upper respiratory tract infection. Two days later she noticed the same symptoms in her right arm accompanied by a slight weakness. Four days after the initial symptoms the weakness spread to the left arm and the legs, which felt stiff. The weakness in the legs further increased and the patient was not able to walk at the time of admission 6 days after her first symptoms. She was then tetraplegic, with mild bilateral facial weakness. The tendon reflexes were abolished. Two days later, the patient had to be tracheotomized and needed assisted ventilation. At this time, she underwent the first of 3 successive 2.0 l plasma exchanges. After the second plasma exchange spontaneous respiration was regained. Two days after the third exchange the patient was able to walk, and 4 days later she was fully ambulant, with only some slight weakness in her arms. Four months later neurological examination revealed only slight weakness of distal

upper limb muscles and of the perioral muscles. Tendon reflexes were elicitable with the exception of the ankle jerks.

The CSF on admission contained 16 mg/dl of protein and 4/mm^3 lymphocytes. Four days later, a repeat lumbar puncture revealed 69 mg/dl of protein and 1/mm^3 lymphocytes, and a further examination one month later, 110 mg/dl of protein and 2/mm^3 lymphocytes.

General examination, chest-X-ray, ESR, urine composition, full blood count and serum electrophoresis revealed normal findings. The patient received 150 units ACTH daily during the first 3 days, but no ACTH or corticosteroids were given subsequently.

Case 6. A 20-year-old female patient had the history of a febrile infection one week before she complained of pain and weakness primarily involving her legs but rapidly spreading to her arms within 2 days. Three days after her initial symptoms she was unable to walk. On admission, neurological examination revealed a severe tetraparesis, more pronounced on the right, as well as bulbar palsy. The tendon jerks were absent. Sensory disturbances were present distally in the lower extremities. The patient was transferred to the intensive care unit, but assisted ventilation was not needed. Five days after the first symptoms the patient had the first of 3 successive 3.0 l plasma exchanges. There was no significant change after the first treatment, but after the second paresis of the arms decreased and the patient became able to swallow. Five days after the third plasma exchange she could elevate her limbs against gravity, after another 2 days she was able to walk; 22 days after the third plasma exchange she was fully ambulant, only showing some slight weakness in the proximal muscles of all extremities.

The CSF on admission contained 23 mg/dl of protein and 1/mm^3 lymphocytes. A repeat lumbar puncture 10 days later revealed 33 mg/dl of protein and no cells. Six days after the third plasma exchange, the right peroneal nerve motor conduction velocity was 29 m/s, distal motor latency 8.3 ms (7 cm), and 28 m/s, distal motor latency 8.3 (7 cm), on the left. One month later, the corresponding values were 39 m/s, distal motor latency 6.9 ms (7 cm), on the right, and 46 m/s, distal motor latency 6.1 (7 cm), on the left.

General examination including chest-X-ray, ESR, urine composition, full blood count, serum electrophoresis and immunglobulins, was normal. The patient received no corticosteroids or ACTH.

Case 7. A 28-year-old male patient suffered from a febrile diarrhea one week before he complained about painful muscles in his legs and felt generally weak. During the next three days the weakness in his legs advanced. Five days after his first symptoms he became unable to walk. One day later, his arms were weak and he found it hard to swallow. On admission weakness was most marked in the proximal muscles, but also involved the distal muscles and was more pronounced on the left side. However the patient could elevate his right leg against gravity. Tendon reflexes were abolished. There was no sensory loss. The weakness progressed to tetraplegia in the next two days and bulbar palsy, accompanied by bilateral facial weakness, developed. At this time he underwent five successive 1.0–1.5 l plasma exchanges. Fourteen days after this treatment the bulbar palsy recovered, but slight facial weakness persisted. Five days later the patient was able to elevate his right leg against gravity again and maintain both his arms in an outstretched position. After a further 18 days he showed only a mild left facial paresis together with weakness and atrophy in the limbs, more pronounced in the distal muscles. The patient was not able to walk without help for four weeks. After a further two weeks the patient was fully ambulant, but weakness in the left leg persisted.

The CSF on admission contained 32 mg/dl of protein and 2/mm^3 lymphocytes. Twenty-two days later, a repeat lumbar puncture revealed 6/mm^3 lymphocytes and 87 mg/dl of protein. One month later, the CSF showed 69 mg/dl of protein and 8/mm^3 lymphocytes. Seven days after the initial symptoms conduction velocity in the right peroneal nerves was 52 m/s, distal motor latency 4.2 (7 cm), and 56 m/s, distal motor latency 4.1 ms (7 cm), on the left.

General examination, including chest-X-ray, urine composition, full blood count, serum electrophoresis and immunoglobulins, was normal. No corticosteroids or ACTH were given during the course of the disease.

Case 8. A 29-year-old male patient had the history of a febrile infection two weeks before he observed painful paraesthesias in his fingers and toes. Five days later increasing weakness of the distal muscles of the upper and lower limbs appeared. After a further 6 days the patient was unable to walk. One day later, he could not elevate his arms. On the next day he found it hard to

swallow and bilateral facial weakness was observed. On admission a bilateral facial and bulbar palsy was accompanied by a severe tetraparesis. Only some slight finger movements could be observed. Tendon reflexes were absent. A sensory deficit was found distally in all four extremities. At this time six successive plasma exchanges were performed varying from 0.5 to 2.0 l each. Cardiovascular problems necessitated minimal exchanges of 0.5 on days two and three, while on the first and last day 2.0 l plasma exchanges were carried out. The other two plasma exchanges were of 1.5 l each. There was no change during the first three days of therapy. On the fourth day the bulbar palsy decreased and two days after the last plasma exchange the patient could elevate his arms and his left leg against gravity. Ten days later, he could maintain both his arms in an outstretched position and shortly afterwards could elevate both his legs. Symptoms of facial and bulbar palsy showed a further significant decrease. The act of swallowing was normal. During the next seven days the patient's strengh steadily increased and he became able to walk. Sensory disturbances had completely recovered. Four weeks later, the patient was fully ambulant with no clinical signs of weakness, but showed decreased tendon reflexes and some slight ataxia of movement.

The CSF on admission contained 20 mg/dl of protein and 1/mm^3 lymphocytes. Fourteen days later, the CSF revealed 110 mg/dl of protein and 2/mm^3 lymphocytes. On admission motor conduction velocity in the right median nerve was 59 m/s, distal motor latency 6.2 ms (5 cm). Sensory conduction velocity of the right median nerve measured antidromically was 50 m/s (elbow–wrist) and 41 m/s (wrist–second finger). Motor conduction velocity in the right peroneal nerves was 37 m/s, distal motor latency 10.5 ms (7 cm), and 44 m/s, distal motor latency 7.8 ms (7 cm), on the left. Eight days after plasma exchange a repeat examination revealed a median nerve motor conduction velocity of 45 m/s, distal motor latency 7.8 ms (5 cm), and a motor conduction velocity of 34 m/s, distal motor latency 10.1 ms (7 cm) in the right peroneal nerve and 37 m/s, distal motor latency 9.4 ms (7 cm) in the left. With proximal stimulation a median nerve sensory action potential was not elicitable on antidromic recording. Distal stimulation revealed a sensory conduction velocity of 34 m/s (wrist–second finger). Two weeks later, motor conduction velocities further decreased despite clinical improvement. The right median nerve motor conduction velocity was 40 m/s, distal motor latency 11.5 ms (5 cm). The corresponding values were 33 m/s, distal motor latency 9.1 ms (7 cm), on the right, and 32 m/s, distal motor latency

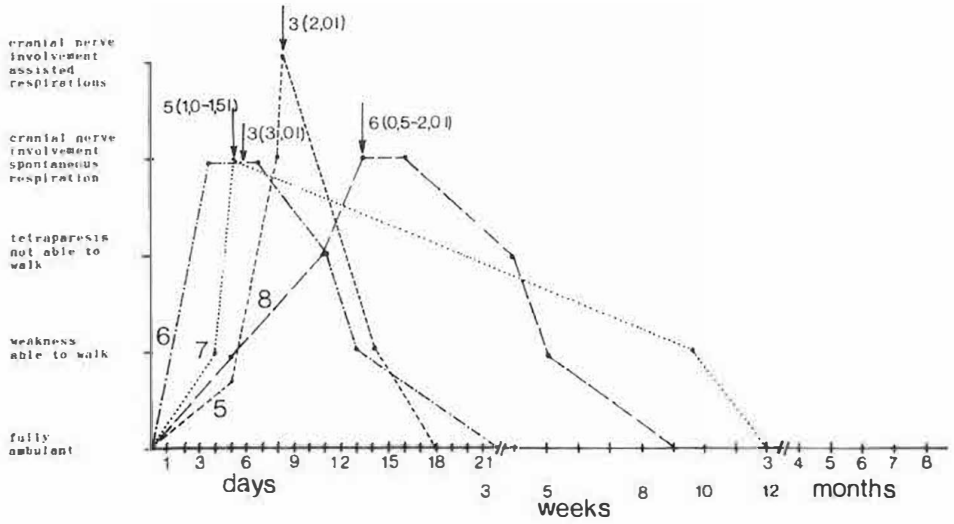

Fig. 2. Effect of plasmapheresis on the course of the Guillain-Barré syndrome in cases 4–8. First plasma exchange is marked by arrow, accompanied by number and amount of plasma exchanges. Same classification of severity and time scale as seen in Fig. 1

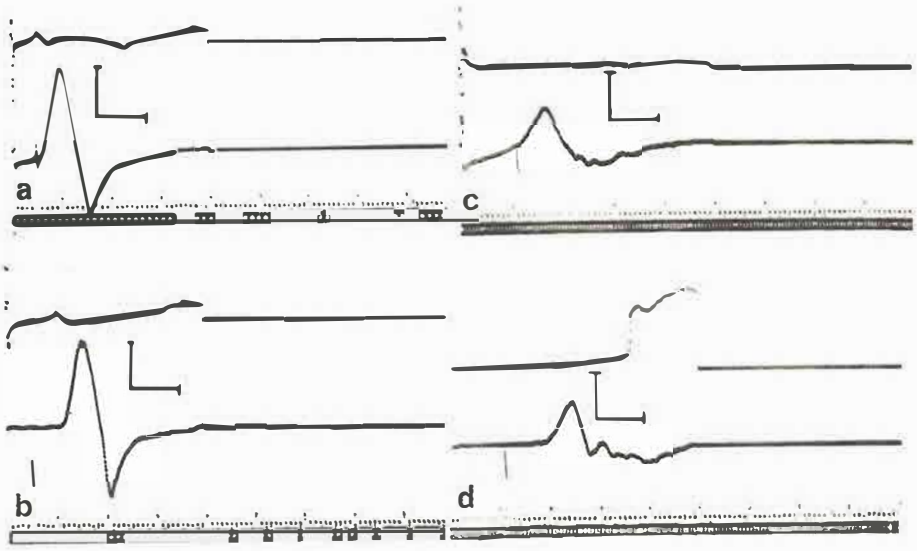

Fig. 3a–d. Conduction velocities in the right median nerve at different stages of the Guillain-Barré syndrome in case 8. a. On admission; Upper channel: antidromic sensory nerve action potential elicited by a maximal electrical stimulus at the wrist. Lower channel: response of the thenar muscles to the same stimulus. b. Upper channel: antidromic sensory nerve action potential elicited by a maximal electrical stimulus at the elbow. Lower channel: response of the thenar muscles to the same stimulus. Motor conduction velocity was 59 m/s, distal motor latency 6.2 ms (5 cm). Sensory conduction velocity was 50 m/s (elbow–wrist) and 42 m/s (wrist–second finger). c. Three weeks later; Upper channel: no antidromic sensory nerve action potential elicited by maximal electrical stimulation at wrist. Lower channel: response of the thenar muscles to the same stimulus. d. No antidromic sensory nerve action potential elicited by maximal electrical stimulation at the elbow. Lower channel: response of the thenar muscles to the same stimulus. Motor conduction velocity was 40 m/s, distal motor latency 11.5 ms (5 cm). Note the decrease of amplitude of motor action potentials, the slowing of motor conduction velocity and the loss of sensory nerve action potentials despite clinical improvement. Evoked muscle and sensory nerve action potentials were recorded by bipolar surface electrodes (50 μV/div.; 10 ms/div. upper channel; 2 mV/div.; 10 ms/div. lower channel; Medelec-EMG)

10.2 ms (7 cm) in the left peroneal nerve. An antidromic median nerve sensory action potential was no longer elicitable both with proximal and distal stimulation (Fig. 3). One week later, the patient was ambulant. Motor conduction velocity in the right median nerve was 31 m/s, distal motor latency 9.3 ms (5 cm), in the right peroneal nerve 30 m/s, distal motor latency 11.5 ms (7 cm), and in the left peroneal nerve 31 m/s, distal motor latency 13.8 ms (7 cm).

General examination, chest-X-ray, ESR, urine composition, full blood count serum electrophoresis and immunoglobulins revealed normal findings. The patient received no corticosteroids or ACTH (Fig. 2).

Discussion

The usefulness of plasma exchange in the treatment of acute polyneuropathy was recently demonstrated in one case [7]. Furthermore, recovery after plasma

exchange was also seen in a case of chronic progressive polyneuropathy [13]. In both cases the improvement was striking and abrupt, warranting the further assessment of plasma exchange in the treatment of acute or chronic progressive polyneuropathy. For this reason plasma exchange was used in eight patients with the Guillain-Barrè syndrome (GBS). In six cases the first symptoms were noticed in the lower limbs, rapidly followed by involvement of the upper limbs. In one case cranial nerve impairment was seen at the onset. The further spread of the illness was dramatic in all cases. All patients became tetraplegic and showed cranial nerve involvement. It is now generally accepted that the severity of the illness is highly significantly correlated with the presence of cranial nerve impairment, especially with weakness of the bulbar and respiratory muscles [14, 20, 22]. Five patients received assisted respiration both the others were so severely affected that is was thought the need might arise.

Intensive investigations were carried out to ascertain the cause of the polyneuropathy. In case 3, chronic uremia was found due to polycystic kidneys. However the albumino-cytologic dissociation in the CSF in two different stages of the illness [21] indicated that the acute cranial and generalized polyneuritis could be better explained by the GBS than by a polyneuropathy due to chronic uremia. The transient disturbanee of vertical ocular movements combined with vertical nystagmus was thought to be a central nervous complication of the GBS with involvement of the brain stem [20], although the patient's consciousness was never clouded. In case 2 infective hepatitis appeared during the course of the disease 30 days after the initial symptoms. Infective hepatitis was distinguished from serum hepatitis by sequential measurement of circulating HB_sAg, which was absent in this case. It therefore seemed unlikely that the hepatitis had been induced by plasma exchange. Because of the incubation time of infective hepatitis and the typical findings in the CSF, infective hepatitis was excluded as the cause of the acute polyneuropathy. In all of the other cases no explanation for the peripheral nerve disorder could be found other than the GBS.

Recovery was abrupt in all cases after the first plasma exchanges. Improvement was more marked, when plasmapheresis was performed on three successive days with plasma exchanges of 2.0–3.0 l each. This is supported by the successful therapy in a patient with the GBS with a 3.0 l plasma exchange on four successive days [7]. Recovery seemed to be delayed in cases when plasma exchanges were reduced to 0.5–1.5 l each and were spread over several days or weeks, even when the number of plasma exchanges was increased.

The observations suggest the use of plasma exchange in the initial progressive stage of the disease and to exchange 3.0 l daily in three or four successive sessions. The observations further indicate that plasmapheresis has to be advocated for all patients who develope a severe illness with respiratory insufficiency. In all of our patients with early treatment, respiratory distress was avoided or the time of artificial respiration was significantly shortened. Considering the high mortality in cases with respiratory complications [20], their successful management by plasma exchange seems to improve the prognosis for life. There was no death in our series of patients. The psychological benefit of plasma exchange as an effective treatment may be another aspect of this therapy. Most patients are anxious and severely emotionally affected when the illness spreads to various parts of the body and

respiratory insufficiency requires artificial ventilation. Similar benefits may arise by avoidance of artificial nutrition.

This advocacy of plasma exchange requires careful consideration, especially as there is little else to offer such patients. Some studies have shown that prednisolone is of no benefit in the GBS [14, 18]. Also a controlled trial of ACTH, 100 units daily for 10 days, did not to show any convincing effect of ACTH, and was also hampered by the fact that all severe cases requiring assisted ventilation were excluded [32]. Case 1, 2, 6, 7 and 8 received no corticosteroids or ACTH, case 3 had corticosteroids during the whole period of treatment, in case 4 ACTH was given 28 days after plasmapheresis and case 5 received 150 units of ACTH in the first three days of treatment. There was no obvious influence on the course of the disease from this therapy.

Unfortunately systemic nerve conduction studies were only carried out in case 8. Despite clinical improvement after plasma exchange and despite recovery from sensory disturbances, motor and sensory conduction velocities further decreased. Marked slowing of motor conduction velocity, impairment of sensory conduction and slowing of distal motor latencies indicate both segmental demyelination and axonal degeneration. A similar slowing of motor conduction was seen in case 6, while in cases 4 and 7 normal motor conduction velocities were found at a very early stage of the disease. As seen in the CSF, where both the cell count and the protein content depend considerably on the stage of the illness at which the CSF is examined [21], the same pattern may be displayed by nerve velocity studies. In case 3, prolonged nerve conduction times were perhaps better explained by the chronic uremia than by the cranial polyneuritis.

These electroneurographic findings are well supported by pathological examinations, which demonstrate segmental demyelination as the predominant type of lesion [3, 11, 35]. Axonal destruction was rare, but was seen in later lesions [8]. Also the appearance of lymphocytic and phagocytic infiltrates was a delayed effect of the disease [17]. It was further demonstrated that the lymphocytes or phagocytes of patients with the GBS are sensitised to peripheral nerve antigen [1, 28, 31]. It could be speculated that in the early stage of the GBS there is a factor in plasma which is active [10, 24], and that lymphocyte sensitization is secondary to damage of the peripheral nerves caused by this primary antibody [4, 14]. The convincing effect of early plasma exchange may support the hypothesis of circulating plasma factors [34]. A delayed cellular response may cause the delayed and progressive slowing of nerve conduction velocities. Moreover, the total recovery time seems to be shortened by plasmapheresis [22].

There are certainly striking similarities with the effect of plasmapheresis in patients with crises in myasthenia gravis. The benefit of plasmapheresis in patients with myasthenia gravis is well known [26, 27] and documented by the decrease of acetylcholine receptor antibody titers after plasma exchange, showing an inverse relationship between the clinical indices of muscle strengh and the acetylcholine receptor antibody titers [26, 33]. Antibody studies were not carried out in our patients, as well as in the patients reported earlier [7, 13]. The determination of immunoglobulins showed normal distributions with the exception of a small IgM-gradient in case 2.

Although the number of patients is small and spontaneous recovery is the rule in the GBS, plasmapheresis seems to be an available therapeutic tool in patients with the GBS whose illness develops dramatically and who require or seem likely to require artificial ventilation. The rapid elimination of the pathogenic antibodies seems to be important for the further course of the illness. The considerable risks and the high technical requirements may limit this therapy to the severer cases of the GBS. The use of a new hollow-fiber filter may facilitate plasma exchange and may also permit the use of this technique at less well equipped medical centers [30].

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Received December 9, 1980