Rehabilitation in Guillian Barre syndrome

Fary Khan, MBBS, FAFRM (RACP), is Lecturer, Rehabilitation Studies, Department of Medicine, University of Melbourne, neuro-rehabilitation physician, the Melbourne Extended Care and Rehabilitation Centre, the Royal Melbourne Hospital, and Head, Orthopaedic and Musculoskeletal Unit, Caufield General Medical Centre, Victoria.

BACKGROUND

Guillian Barre syndrome (GBS) is the most common form of neuromuscular paralysis. It mostly affects young people and can cause long term residual disability.

OBJECTIVE

This article outlines the rehabilitation treatment for patients recovering from GBS.

DISCUSSION

Recovery from GBS can be prolonged. Early rehabilitation intervention ensures medical stability, appropriate treatment and preventive measures to minimise long term complications. Specific problems include deep venous thrombosis prevention, complications of immobility, dysautonomia, de-afferent pain syndromes, muscle pain and fatigue. Longer term issues include psychosocial adjustment, return to work and driving, and resumption of the role within the family and community. Effective communication between the GP and rehabilitation physicians is imperative for improved functional outcomes and successful social reintegration.

Guillain Barre syndrome (GBS) is the most common form of neuromuscular paralysis in developed countries. The incidence in Australia is similar to that of the United States of 1–2 cases per 100 000 annually, with a male-female ratio of 2:1.[1] The mortality of the condition in Australia is less than 1%.[2]

Guillain Barre syndrome often affects young people – with a relatively long life expectancy – and is therefore an important cause of long term disability for those patients with residual deficits. Most patients with GBS are discharged home with outpatient and home rehabilitation programs. However, 40% of all GBS patients require inpatient rehabilitation (especially those requiring ventilatory support).[1] Although general practitioners may see GBS sufferers infrequently in their practice, the needs and issues of these patients and their families are significant.

The most common type of GBS is acute inflammatory demyelinating polyradiculoneuropathy (AIDP). Other axonal subtypes include: acute motor axonal neuropathy and acute motor and sensory axonal neuropathy. Variants of GBS include Miller Fisher syndrome (cranial nerve involvement, ataxia) and acute pan-dysautonomia. The diagnostic features of GBS are shown in *Table 1*, the frequency of specific symptoms in *Table 2*, and differential diagnosis in *Table 3*.

Aetiology and pathogenesis

Guillain Barre syndrome is immune based – often acute fulminant demyelinating inflammatory poly neuropathy. Sensitisation of T lymphocytes to protein in the myelin sheath is necessary for disease induction.[3] Patchy areas of demyelination occur along peripheral nerves, nerve roots and myelin sheaths as a result of lymphocytic infiltration, causing impaired conduction of action potential leading to slow conduction velocity and conduction blocks. In axonal neuropathies, the conduction velocity is normal, but the number of functional motor units is decreased.[4] Cerebrospinal fluid (CSF) protein levels are elevated in the second week of illness. Within 2–3 weeks of the demyelination process, the inflammation resolves and re-myelination commences.[4,5]

Up to 60% of patients have had a preceding upper respiratory illness.[6] However, about 27% of patients with GBS have no identified preceding illness. Infection with cytomegalic virus (CMV) and *Campylobacter jejuni* has been implicated in the axonal form of GBS.

Guillain Barre syndrome has also been associated with diabetes, alcohol abuse, exposure to heavy metals or industrial toxins,[7] epidural aesthetic, and drugs (thrombolytic agents, heroin). Underlying systemic diseases such as systemic lupus erythematosus, sarcoidosis, Hodgkin disease, and other neoplasms have been known to cause a small number of GBS cases.[5]

Prognosis

The disease progression varies with severity of disease. Fulminant cases develop maximum paralysis within a couple of days, however, 50% of patients reach their peak severity of disease at about 2 weeks, and 80% by 3 weeks.[8] Recovery usually starts within 2–4 weeks after disease onset. Guillain Barre syndrome lasts for about 12 weeks in most patients and has a favourable outcome in the majority of cases. Factors associated with a poorer prognosis are listed in *Table 4*. Approximately 50% of patients have minor residual neurological deficits, 15% persistent residual deficits in function, and about 80% are ambulatory within 6 months of disease onset.[5,9]

Table 1. Diagnostic features based on clinical criteria supported by CSF and electrophysiological abnormalities[4,5,25]

- Ascending motor symmetrical, flaccid areflexia paralysis (may be limited to the distal limbs or progress to full quadriplegia with respiratory and cranial nerve involvement)
- Paresthaesias and hypesthaesias (mild sensory involvement)
- Cranial nerve involvement (LMN VII)
- Autonomic dysfunction: sinus tachycardia or less often bradycardia, fluctuating hypertension and hypotension, loss of sweating or episodic profuse diaphoresis
- History of flu-like illness (common), afebrile
- Elevated CSF protein, fewer than 10 cells/3 mm
- Abnormal nerve conduction studies
- Recovery after 2–4 weeks of plateau of the disease process

Table 2. Frequency of features and clinical variants of acute GBS[1,5]

Features of syndrome	Frequency in fully developed cases
Weakness in the legs	95%
Weakness in the arms	90%
Areflexia	90%
Paresthaesia	85%
Sensory loss	75%
Oropharyngeal weakness	50%
Pain	30%
Respiratory failure	30%
Ophthalmoparesis	15%
Ataxia	15%
Sphincter involvement	5%
Clinical variants	
Fisher's syndrome	5%
Weakness without paresthaesia	3%
Pharyngeal, brachial cervical weakness	3%
Paraparesis	3%
Facial paresis with paresthaesia	1%
Pure ataxia	1%

Treatment

The acute setting

A detailed discussion of management of GBS in the acute stage is beyond the scope of this article. However, the following points are important in that they affect prognosis.

- Plasma exchange is the first treatment for GBS and is most beneficial when started within 7 days of disease onset, and of some benefit if started within 30 days of the onset.[10] Plasmapheresis shortens the time a patient stays on respiratory support, the time required to achieve independent walking, and is associated with greater functional mobility at 6 months[11,12]
- Immunoglobulin infusion (IVIg) hastens recovery in GBS as much as plasmaphoresis.[13] Administration of IVIg after plasma exchange has no added advantage over plasma exchange alone,[14] and there are reports of a high incidence of relapse following IVIg treatment[15]
- Treatment of GBS with steroids was ineffective in a large prospective randomised study using 500 mg methylprednisolone,[16] despite earlier suggestion of their usefulness in decreasing severity of illness.[17] Therefore, corticosteroids should not be used in the treatment of GBS.

Rehabilitation

About 40% of all cases require inpatient rehabilitation as most patients are very disabled and will have required ventilator support during the acute stage. Patients are initially closely monitored in the rehabilitation setting for signs of respiratory distress. They require intubation when the vital capacity decreases to <18 mL/kg and are transferred back to hospital for medical stabilisation.

A common scenario for a patient with severe GBS would be inpatient rehabilitation for 3–6 weeks followed by a community and home based rehabilitation program for 3–4 months.

The aim of rehabilitation is to restore and

Table 3. Differential diagnosis

- Spinal cord compression
- Transverse myelitis
- Myasthenia gravis
- Neoplastic meningitis
- Vasculitis neuropathy
- Paraneoplastic neuropathy

maintain a person's functional independence as soon as the patient is medically stable. Rehabilitation utilises an interdisciplinary team approach (eg. physiotherapist, occupational therapist, nurse, social worker) encouraging active patient and family education and participation using a time based, goal focussed, functional approach to minimise disability and maximise function and community participation.

Respiratory complications

Respiratory complications from GBS encountered in the rehabilitation setting and community are: incomplete respiratory recovery including chronic obstructive pulmonary disease, restrictive respiratory disease from pulmonary scarring and pneumonia, and tracheitis from chronic intubation and respiratory muscle insufficiency.[18]

Restrictive pulmonary function is associated with sleep hypercapnia and hypoxia during REM sleep.[19] Night time saturation records with pulse oximeter and BiPAP may be indicated for patients with hypoxia or hypercapnia. Physical therapy measures (chest percussion, breathing exercises, resistive inspiratory training) are used to clear respiratory secretions to reduce the work of breathing. Special weaning protocol in tracheostomy patients is suggested to prevent over fatigue of respiratory muscles. Patients with cranial nerve involvement are more prone to aspiration and respiratory complications and need extra monitoring. Patients are encouraged not to recommence smoking if previously a smoker.

Deep venous thrombosis

Pulmonary embolism has been reported in up to a third of patients with GBS.[20] Although prophylaxis for deep venous thrombosis is recommended, the optimal type and length of prophylaxis is unknown.[18] In the rehabilitation setting, patients are encouraged to be active and wear compression stockings. Progressive mobilisation protocols such as strategies to improve bed mobility, practising sitting up from bed, and safe transfer techniques (bed to chair) with or without adaptive equipment is a priority. Training carers and partners of more severely affected patients is important.

Dysautonomia

Dysautonomia is associated with severe forms of GBS, extending the duration of acute care. It can cause life threatening cardiac arrhythmia.[19,21] The morbidity and mortality associated with dysautonomia is significant. In one study of 100 patients, 11 developed cardiac arrhythmia of which seven patients died.[22]

Patients with excessive sympathetic outflow and hypertension are sensitive to vasoactive drugs and are prone to cardiac arrhythmia.[9] Approximately 50% of patients develop difficulty with blood pressure control especially postural hypotension.[21] Rehabilitation treatment includes: education and awareness of staff, patient/family, use of compression stockings, adequate hydration, 'profiling techniques' (patients undergo posture training to allow for baroreceptor control and stabilisation of BP) and the use of tilt tables.

Dysautonomia can cause early lower motor bladder and bowel involvement; although this usually resolves. Bladder and bowel programs are implemented in rehabilitation units to ensure social continence and to avoid complications such as bladder overdistension and urinary infections (which can occur in about 30% of patients with GBS).[23] These programs often continue as patients are discharged home. Other dysautomic features such as impotence settle over time.

Immobilisation

Prolonged immobilisation leads to reduction of blood volume, and this with concurrent postural hypotension can be difficult to manage. A tilt table for immobilised patients can be effectively used in rehabilitation units. Early mobilisation of patients will lower serum calcium levels and counter immobilisation hypercalcaemia.[1]

Physical therapy encompasses a graduated mobility program which includes: maintenance of the patient's posture and alignment, maintaining joint range of motion (passive, active, active assisted), providing ankle foot orthosis to prevent plantar contractures, improving endurance (repetitive exercises with low resistance), strengthening different muscle groups, and improving flexibility with a progressive ambulation program that commencing with bed mobility techniques and the use of a wheelchair – to patients walking using adaptive gait aids

Table 4. Features associated with poorer outcome in GBS[4]

- Older age
- Requirement for respiratory support
- Abnormal peripheral nerve function
- No plasmapheresis is performed
- Subgroup of GBS with primary axonal degeneration
- Patients with rapid onset
- Progression to quadriplegia
- Respiratory dependence
- Severe disease at presentation
- *Campylobacter jejuni* infection
- Patients showing no improvement at 3 weeks of plateau of disease

(frames, crutches). Other problems relating to immobilisation include:

- compression nerve palsies (ulnar, peroneal and cutaneous femoral nerve palsies occur commonly). Special attention is given to correct positioning of the patient
- pressure sores (loss of muscle mass, combined with sensory loss). Patient and carer/partner education in skin care is essential, and
- heterotopic ossification (periarticular bone formation in the muscle planes). This can be prevented by early aggressive joint range of motion exercise and mobilisation.

Fatigue and pain

De-afferent pain syndromes and muscle pain can be presenting symptoms in GBS.[24] In one study, 55% of patients reported pain and 72% reported pain during the entire course of their illness.[6] Symptoms of depression and mental fatigue are also common.[5] Treatment includes occupational therapy (OT), and supervised desensitisation therapy to enable patients to tolerate practising their daily living tasks (eg. grooming, dressing). The use of antidepressants (SSRIs, tricyclic antidepressants), membrane stabilising agents (tegretol), and gabamimetic agents (gabapentin) can be useful in neuropathic pain. Tramadol or narcotics are

Case study

Mr M, 46 years of age, presented to the emergency department 2 weeks earlier with sudden onset of ascending paralysis involving both lower limbs. He had had a preceding sore throat, chest infection and generalised aches and pains. He was diagnosed with GBS and treated with immunoglobulin for 5 days. He did not require ventilatory support. He was transferred to the medical ward initially, and then 2 weeks later to the rehabilitation unit.

Rehabilitation assessment showed a number of disabilities. He was wheelchair bound with decreased muscle strength in both lower limbs (2+/5) with bilateral foot drop, decreased muscle performance, fatigue and a propensity for falls. His sensory examination was normal. He reported a burning sensation in both the feet and shin areas. He still had the indwelling catheter as his previous residual urine volume was about 800 mL. He also had severe constipation. His mood was low and he reported difficulty sleeping. This 'episode' had made him aware of his mortality and changed how he perceived life in general. His wife was distressed and looking after their 2 young children. He worked as a computer support person with a local firm and had a number of financial concerns. Before his illness he enjoyed playing cricket.

The rehabilitation process

Mr M and his wife will initially have a session with the treating rehabilitation team regarding implications of GBS, prognosis, outcome and expectations from rehabilitation intervention. He is expected to make an excellent recovery.

A graduated mobilisation program (physiotherapy) will commence to attain independent bed mobility, safe transfer skills, improve sitting balance, progressive ambulation (parallel bars, coordination work, bilateral ankle foot orthosis) and gait training with increasing weight bearing on lower limbs. Complications resulting from immobility such as pressure sores and nerve palsies will be specifically targeted. He will have desensitisation therapy to allow weight bearing on his lower limbs accompanied by range of joint movement exercises for the prevention of contractures. His exercise program will address endurance and muscle strengthening. Meantime wheelchair skills will be taught. His recovery for ambulation will be gradual (over the next 6–12 weeks).

His mood symptoms need to be fully assessed and monitored. Counselling relating to adjustment issues will be undertaken and specific treatment with antidepressants if indicated. Tegretol will be an option (membrane stabilising affect) in pain modulation. Other modalities such as transcutaneous electrical stimulation may also be used.

Mr M will have his catheter removed (after a trial of void) and a bladder program will be initiated. It is unlikely that he will require an urodynamics study once the myopathic distension of his bladder settles. A bowel program will also commence. He will practise performing his daily personal care (showering, dressing) and domestic skills (making a cup of tea) with the OT. Appropriate equipment to facilitate care needs (long handled grabbers) will be provided using energy conservation strategies. The OT will do a home visit for environmental modification (safe access, steps, ramps). Any equipment required will be provided (eg. shower stool, long handled aids).

Mr M is expected to stay in the rehabilitation ward for about 4–6 weeks with 'trial leave' on weekends. He will continue with outpatient therapy (physical and OT) for a further 6–8 weeks. He will be discharged home under the care of his GP (with whom he has a good rapport). He may also be eligible for 'rehabilitation in the home program' if there are transport difficulties. Referral to the state GBS Society will be followed up.

There will be a number of long term care issues, which will be monitored by his GP and the rehabilitation team. These include: psychosocial adjustment, altered family role and self image, increased care needs for the family, inability to return to driving and work, financial constraints, marital stress and general limitation with participation. A referral for a driving assessment will be considered in 2–3 months (if appropriate). Return to work is important, the OT will do a worksite visit and contact his employer; there may be consideration for alternate or limited duties and a referral to a commonwealth rehabilitation service will be forwarded.

sometimes used for severe pain. Transcutaneous electric nerve simulation (TENS) can also be used as an adjuvant therapy.

The OT can advise specific energy conservation strategies to manage fatigue and facilitate patient functional independence and provide adaptive equipment (eg. grabbers, sock donners, plate guards) for the patient to facilitate personal care. This continues over time to incorporate domestic (eg. making tea) and community tasks (eg. banking, crossing roads). Home modifications are also undertaken to make the environment safer and accessible (eg. nonskid mats, proper lighting, grab rails). In severe cases, electric wheelchairs may be provided for community mobility. Other longer term issues such as return to driving and work are also coordinated by the OT, along with the GP and the rehabilitation physician.

Psychological and social issues

There are no studies in GBS that address the psychosocial and vocational outcomes. Mild depression and mental fatigue have already been mentioned. Education and counselling of the patient (and their family) is very important. On discharge home, many patients continue to have significant impairments that improve over 12 months. In this transition period, various adjustment issues such as the patient's perception of self worth and self image, and the altered role within the family surface. Families often struggle to cope with new demands associated with increased care needs, inability to return to driving and work, financial constraints, marital stress and the general limitation of the patient's participation. Carer support and respite care are important. Referral to a local GBS society such as the National Stroke Foundation and support groups such as GBS Survivors can be helpful in providing patients and their family with ongoing support, resources and equipment needs.

Conclusion

General practitioners are primary carers for GBS patients and their families in the community. The prognosis from GBS is good, but recovery is prolonged. Close liaison between the GP and the rehabilitation team will assist the patient by minimising disability, improving functional outcomes and their quality of life.

Conflict of interest: none declared.

References

1. Meythaler JM. Rehabilitation of Guillain Barre syndrome. Arch Phys Med Rehabil 1997;78:872–879.
2. GBS society Australia. Available at: www.members.ozemail.com.au/guillain/.
3. Brosnan CF, Claudio L, Tansy FA, Martiney J. Mechanisms of autoimmune neuropathies. Ann Neurol 1990;27:S75–S79.
4. Hallum A. Neuromuscular diseases. In: Neurological Rehabilitation. Umphred DA, ed. 4th edn. St Louis: Mosby, 2001;363–415.
5. Ropper AH. The Guillain Barre syndrome. N Engl J Med 1992;326:1130–1136.
6. Roper AH. Severe acute Guillian Barre syndrome. Neurology 1986;36:429–432.
7. Ringel SP, Cooper WH. Classification of neuromuscular disorders. In: Maloney FP, ed. Interdisciplinary Rehabilitation of Multiple Sclerosis and Neuromuscular Disorders. Philadelphia: JB Lippincott, 1985.
8. Pascuzzi RM, Fleck JD. Acute peripheral neuropathy in adults. Neurol Clin 1997;15:529–547.
9. Winer JB, Hughes RAC, Greenwood RJ, et al. Prognosis in Guillain Barre syndrome. Lancet 1985;1:1202–1203.
10. Raphael JC, Chevret S, Hughes RAC, Annane D. Plasma exchange for Guillain Barre syndrome (Cochrane Review). In: The Cochrane Library, Issue 1, 2004. Chichester UK: John Wiley & Sons, Ltd.
11. French Cooperative Group. Plasma exchange in Guillain Barre syndrome: role of replacement fluids. Ann Neural 1987;22:753–761.
12. Guillain Barre Study Group. Plasmapheresis and acute Guillain Bare syndrome. Neurology 1985;35:1096–1104.
13. Van der Meche FGA, Schmitz PIM, the Dutch Guillain Barre Study Group. A randomised trial comparing intravenous immune globulin and plasma exchange in Guillain Barre syndrome. N Engl J Med 1992;326:1123–1129.
14. Hughes RAC, Raphael JC, Swan AV, van Doorn PA. Intravenous immunoglobulin for Guillain Barre syndrome (Cochrane Review). In: The Cochrane Library, Issue 1, 2004. Chichester, UK: John Wiley & Sons, Ltd.
15. Hughes RA, Rees JH, Guillain Barre syndrome. Curr Opin Neurol 1994;7:386–392.
16. Guillain Barre Syndrome Steroid Group. Double blind trial of intravenous methylprednisolone in Guillain Barre syndrome. Lancet 1993;341:586–590.
17. Feasby TE. Inflammatory demyelinating polyneuropathies. Neurologic Clin 1992;10:651–670.
18. Garth PJ. Guillain Barre syndrome. New York: Thieme Medical Publishers, 1993.
19. Bach JR. Rehabilitation of the patient with respiratory dysfunction. In: Delis JA, ed. Rehabilitation medicine: principles and practice. 2nd edn. Philadelphia: JB Lippincott, 1993;952–972.
20. Raman TK, Blake JA, Harris TM. Pulmonary embolism in Landry-Guillian-Strohl syndrome. Chest 1971;60:555–557.
21. Zochodne DW. Autonomic involvement in Guillain Barre syndrome: a review. Muscle Nerve 1994;17:1145–1155.
22. Sedan MJ, Calera J, Conga E, Berciano. Guillain Barre syndrome in Cantabria, Spain: an epidemiological and clinical study. Acta Neurologica Scand 1994;89:287–292.
23. Ropper AH. The Guillain Barre syndrome. N Engl J Med 1992;326:1130–1136.
24. Ravn H. The Landrey Guillain Barre syndrome: a survey and a clinical report on 127 cases. Acta Neurol Scand 1967;43:S30:8–64.
25. Young RR, Asbury AK, Corbett JL, Adams RD. Pure pan dysautonomia with recovery: description and discussion of diagnostic criteria. Brain 1975;98:613–636.

AFP

Correspondence
Email: fary.khan@mh.org.au