Guillian Barre Syndrome

Guillain–Barré syndrome (**GBS**) (French pronunciation: [giˈlɛ̃ baˈʁe], English pronunciation: /ˈgiːlænˈbɑreɪ/), sometimes **Landry's paralysis** or **Guillain–Barré–Strohl syndrome**, is an acute polyneuropathy, a disorder affecting the peripheral nervous system. Ascending paralysis, weakness beginning in the feet and hands and migrating towards the trunk, is the most typical symptom, and some subtypes cause change in sensation or pain as well as dysfunction of the autonomic nervous system. It can cause life-threatening complications, in particular if the respiratory muscles are affected or if there is autonomic nervous system involvement. The disease is usually triggered by an infection.

The diagnosis is usually made by nerve conduction studies and with studies of the cerebrospinal fluid. With prompt treatment by intravenous immunoglobulins or plasmapheresis, together with supportive care, the majority will recover completely. Guillain–Barré syndrome is rare, at 1–2 cases per 100,000 people annually, but is the most common cause of acute non-trauma-related paralysis. The syndrome is named after the French physicians Georges Guillain and Jean Alexandre Barré, who described it in 1916.

- **Acute inflammatory demyelinating polyneuropathy** (AIDP) is the most common form of GBS, and the term is often used synonymously with GBS. It is caused by an auto-immune response directed against Schwann cell membranes.
- **Miller Fisher syndrome** (MFS) is a rare variant of GBS. Accounting for approximately 5% of GBS cases, it manifests as a descending paralysis, proceeding in the reverse order of the more common form of GBS.[1] It usually affects the eye muscles first and presents with the triad of ophthalmoplegia, ataxia, and areflexia.[2] The ataxia predominantly affects the gait and trunk, with the limbs relatively spared. Anti-GQ1b antibodies are present in 90% of cases.
- **Acute motor axonal neuropathy** (AMAN),[3] also known as **Chinese paralytic syndrome**, attacks motor nodes of Ranvier and is prevalent in China and Mexico. It is probably due to an auto-immune response directed against the axoplasm of peripheral nerves. The disease may be seasonal and recovery can be rapid. Anti-GD1a antibodies[4] are present. Anti-GD3 antibodies are found more frequently in AMAN.
- **Acute motor sensory axonal neuropathy** (AMSAN) is similar to AMAN but also affects sensory nerves with severe axonal damage. Like AMAN, it is probably due to an auto-immune response directed against the axoplasm of peripheral nerves. Recovery is slow and often incomplete.[5]
- **Acute panautonomic neuropathy** is the most rare variant of GBS, sometimes accompanied by encephalopathy. It is associated with a high mortality rate, owing to cardiovascular involvement, and associated dysrhythmias. Frequently occurring symptoms include impaired sweating, lack of tear formation, photophobia, dryness of nasal and oral mucosa, itching and peeling of skin, nausea, dysphagia, and constipation unrelieved by laxatives or alternating with diarrhea. Initial nonspecific symptoms of lethargy, fatigue, headache, and decreased initiative are followed by autonomic symptoms including orthostatic lightheadedness, blurring of vision, abdominal pain, diarrhea, dryness of eyes, and disturbed micturition. The most common symptoms at onset are related to orthostatic intolerance, as well as gastrointestinal and sudomotor dysfunction (Suarez et al. 1994). Parasympathetic impairment (abdominal pain, vomiting, constipation, ileus, urinary retention, dilated unreactive pupils; loss of accommodation) may also be observed.
- **Bickerstaff's brainstem encephalitis** (BBE) is a further variant of Guillain–Barré syndrome. It is characterized by acute onset of ophthalmoplegia, ataxia, disturbance of consciousness, hyperreflexia or Babinski's sign. The course of the disease can be monophasic or remitting-relapsing. Large, irregular hyperintense lesions located mainly in the brainstem, especially in the pons, midbrain and medulla, are described in the literature. Despite severe initial presentation, BBE usually has a good prognosis. Magnetic resonance imaging (MRI) plays a critical role in the diagnosis of BBE. A considerable number of BBE patients have associated axonal Guillain–Barré syndrome, indicative that the two disorders are closely related and form a continuous spectrum.

All forms of Guillain–Barré syndrome are autoimmune diseases, due to an immune response to foreign antigens (such as infectious agents) that is mistargeted at host nerve tissues instead, a phenomenon called molecular mimicry.[7] The targets of such immune attack are thought to be gangliosides, compounds naturally present in large quantities in human peripheral nerve tissues. The most common antecedent infection is the bacterium *Campylobacter jejuni*,[8][9] followed by cytomegalovirus (CMV).[10] However, 60% of cases do not have a known cause. Some cases may be triggered by the

influenza virus, or by an immune reaction to the influenza virus.[11] There was increased incidence of Guillain-Barré syndrome following influenza immunization during the 1976-1977 swine flu pandemic;[12] however, epidemiological studies since then have demonstrated either an extremely small increased risk following immunization (under 1 additional case per million vaccinations) or no increased risk.[13][14]

The end result of this autoimmune attack on the peripheral nerves is damage to the myelin, the fatty insulating layer of the nerve, and a nerve conduction block, leading to muscle paralysis that may be accompanied by sensory or autonomic disturbances. In mild cases, nerve axon (the long slender conducting portion of a nerve) function remains intact and recovery can be rapid if remyelination occurs. In severe cases, axonal damage occurs, and recovery depends on the regeneration of this important tissue. Approximately 80% of patients have myelin loss; in the remaining 20%, the pathological hallmark is axon loss. Guillain–Barré, unlike disorders such as multiple sclerosis (MS) and Lou Gehrig's disease (ALS), is a peripheral nerve disorder and does not in general cause nerve damage to the brain or spinal cord.

Hyperbaric Oxygen Therapy has been used to treat the following conditions:

Acute Acoustic Trauma
Acute & Chronic Anemia
Acute & Chronic Arterial Insufficiency
AIDS
ALS "Lou Gehrig's Disease"
Alzheimers
Autism Spectrum Disorder
Candida And Fungal Infections
Cerebral Edema
Cerebral Palsy
Chemical Poisoning
Chronic Fatigue Syndrome
Colitis
Cosmetic Surgery
Crohn's Disease
Compartmental Syndrome
Demyelination
Diabetes
Fibromyalgia
Flesh Eating Bacteria
Fracture Repair
Gastric And Duodenal Ulcers
Guillain-Barre Syndrom
Headaches, Cluster
Heart Attack
Hypoxic Birth Disorders
Hypoxic Induced Epilepsy
Inflammation
Inflammatory Arthritis
Lupus
Lyme Disease
Macular Degeneration
Migraines
Multiple Sclerosis
Musculoskeletal Injuries
Mycoplasma
Near Drowning
Near Hanging
Neurovascular Compression
Osteoporosis
Parkinson's Disease
Peripheral Nerve Injury And Neuropathies
Peripheral Vascular Disorders
Post Surgical Instability
Rehabilitative Care
Retintis Pigmentosa
RSD
Rheumatoid Arthritis
Sacroiliac Syndrome
Scelodrema
Silicone Induced Disorders
Spinal Cord Injury
Spider Bite
Stroke
Sudden Deafness
Surgery Healing, Pre And Post
Traumatic Brain Injury
Vegetative Coma
Wound Healing

Currently, there is no known cure for Guillain-Barré syndrome. The goal of treatment is to prevent breathing problems and provide supportive care (relief of symptoms). Medications are used to control pain and other conditions that may be present. In addition, treatments, such as plasmapheresis or immunoglobulin administration, may be used to suppress the immune system and/or reduce inflammation caused by the immune system's response to the disease. HBOT reduces inflammation and build a healthy immune system with an appropriate response.

Plasmapheresis is a procedure that removes the plasma (liquid part of the blood) and replaces it with other fluids. Antibodies are also removed with the plasma, which is thought to help reduce the symptoms of the disease. Another treatment is the administration of immunoglobulin, a blood product that helps to decrease the immune system's attack on the nervous system.

Other therapies include hormonal therapy and physical therapy (to increase muscle flexibility and strength). Through research, new treatments for Guillain-Barré syndrome are continually being identified.

Man with Facial Weakness Receives Hyperbaric Oxygen

Michael Vincent Anthony, MS, PA-C

Introduction

A 30-year-old man presented to the emergency department (ED) with limited mobility on the left side of his face and none at all on the right. The patient was also experiencing hyperacusis, a change in his sense of taste, and speech impairment.

This was his third visit to the ED. During his first visit two days earlier, he complained of facial weakness on the right side of his face subsequent to recent recovery from a viral upper respiratory infection. When a physical examination revealed no unusual findings, the patient was given a prescription for acyclovir and discharged with a diagnosis of Bell's palsy.

The patient returned to the ED the next day reporting that the facial weakness had progressed to the left side of his face. Again, physical examination was unremarkable. It was determined that his symptoms were caused by fatigue, and the patient was instructed to get some rest.

During the most recent visit, the patient was examined by the attending neurologist. The differential diagnosis included sarcoidosis and Guillain-Barré syndrome. Blood workups performed included complete blood cell count, HIV screening, and type-specific and antinuclear antibody testing; spinal fluid and urine were also tested. Both posteroanterior and lateral chest x-rays were performed, as was magnetic resonance imaging (MRI) of the head. All laboratory test results and chest x-rays were within normal limits. MRI revealed questionable swelling of cranial nerve VII on the left side of the face; cranial nerve VII on the right was unremarkable.

Shortly after the evaluation, the patient received a 30-minute infusion of methylprednisolone and an antacid (to combat potential gastric effects). Artificial tears were administered to prevent dehydration of the eyes. Three hours later, methylprednisolone (in conjunction with an antacid) was again administered.

The patient was sent home after his 10-hour stay in the ED. He received a prescription for prednisone to be taken for five days and then tapered for one week. He was advised to continue with the acyclovir regimen. A follow-up appointment was made with the consulting neurologist for three weeks following discharge. The diagnosis was bilateral facial paralysis of unknown etiology (ie, bilateral Bell's palsy).

Discussion

Known causes of facial paralysis include stroke, trauma (eg, laceration, fractures), neoplasm (metastatic lesions, parotid tumors), and congenital defects.[1,2] Bell's palsy, however, is an acute idiopathic condition involving damage to the seventh cranial facial nerve. It is the most prevalent form of facial paralysis[1] and usually presents unilaterally. Traditionally thought to be a diagnosis of exclusion, unilateral Bell's palsy can be positively identified based on clinical assessment, without performing expensive tests.[3]

Simultaneous bilateral facial palsies (SBFPs) are extremely uncommon and may indicate a more serious disease. In these cases, it is especially important to immediately rule out other causes.[4] See Table 1[1,5-7] (page 57) for the common differential diagnosis and diagnostic tools.

Some unusual presentations associated with SBFP include HIV[8] and intracranial hypertension.[9] In these cases, research has shown that the etiology was an unknown infectious process.

Classic Presentation

Typically, the onset of facial paralysis in patients with Bell's palsy is sudden. Paralysis may be preceded by pain in front of or behind the ears that can last until after paralysis becomes complete, usually between three and 72 hours. Patients may complain that the face feels stiff or pulled to one side but will have no demonstrable sensory loss. Hyperacusis (ie, abnormal acuteness of hearing due to increased irritability of the sensory neural mechanism) on the affected side, drooling, excessive tearing, and a change in the sense of taste can accompany facial paralysis.[2]

Bell's palsy affects approximately 25 persons per 100,000. Although incidence increases with age, peak incidence occurs from ages 10 to 40 years.[1,3] Men are affected at the same rate as women. At risk are those who have had influenza, a cold, or some other upper respiratory infection; persons with hypertension or diabetes; and pregnant women.[10-14] The right side of the face is affected 63% of the time.[2] Human herpesvirus 1 has been implicated as the likely cause of Bell's palsy.[15]

Between 60% and 80% of patients will experience a complete resolution of symptoms, while the remaining cases will have some residual -- or even permanent -- effects.[1,14] Indicators of a favorable prognosis include incomplete paralysis, younger age, and electrodiagnostic tests showing normal nerve excitability.[16]

Treatment

Although most Bell's palsy patients do not require specific treatment, a common regimen prescribed for facial paralysis is physical therapy, which can be used alone or in conjunction with surgery. The most serious complication associated with facial paralysis is ulceration of the cornea. This can be averted by using an artificial tear solution or applying a lubricating ointment.[2]

Antiviral agents (if infectious processes are suspected to be the cause of paralysis) and corticosteroids are usually prescribed to treat Bell's palsy. Research has shown that patients who receive such treatment within three days of symptom onset experience higher recovery rates than those who start therapy after four or more days.[17] Steroid treatments have been shown to be of little benefit when initiated more than four days after the onset of paralysis. While research has shown that patients given methylcobalamin with concomitant steroid therapy to treat Bell's palsy experience greater improvement than those treated with steroids alone,[18] use of this combination is not yet widespread.

Other nonsurgical treatments to combat the effects of facial paralysis include mime therapy, in which patients measure progress by the symmetry of their smile and judgment of their smile by others[19]; myofeedback[19,20]; Hyperbaric Oxygen Treatments, which have been shown to be more effective than treatment with prednisone[21]; and the use of high-voltage electrical muscle stimulation and chiropractic manipulation.[22]

Surgical intervention to reanimate the face is typically reserved for those cases in which the lesion in the facial nerve has been located.[23] Surgery types include cross facial nerve grafts,[24] rectus abdominis muscle transfers,[25] and nerve decompression.[26] Once general reanimation has been established, other therapies can be used to regain control of movement.

Psychosocial Implications

To choose the most appropriate treatment options, clinicians must take into account what their patients consider an acceptable level of functionality, as well as patients' values about their own health and social/moral structures.[27]
The psychosocial impact of facial paralysis can trigger depression, social withdrawal, or both. According to Neely and Neufeld,[28] the importance of the face as a symbol of personal identity and a tool for both verbal and nonverbal communication is often overlooked as part of the psychosocial impact of facial disfigurement. Results from their study show that the quality of patients' smiles influenced how they were perceived by others (see "One Patient's Experience With Bilateral Bell's Palsy," page 58).

Conclusion

Clinical assessment is sufficient to diagnose cases of unilateral Bell's palsy. However, when a patient presents with a complaint of SBFP, a complete and comprehensive examination must be performed. The distinctions among extracranial, intratemporal, infectious, traumatic, and idiopathic origin will dictate what type of treatment is necessary.
A conservative, all-inclusive approach is best. Treatment, whether with physical therapy, drugs, or surgery, should be aimed at what the patient feels is an appropriate recovery goal. Because the face is such an important component of nonverbal communication, the more fully patients can return to their original physical state, the better.

Tables

Table 1. [1,5-7] Differential Diagnosis for Bell's Palsy

Disease	Symptoms	Diagnostic tools
Melkersson-Rosenthal syndrome	Congenitally fissured tongue, orofacial edema, facial palsy [5]	
Möbius' syndrome	Congenital facial diplegia, ophthalmoplegia[6]	
Guillain-Barré syndrome	Ascending motor weakness, usually beginning in legs and possibly leading to respiratory muscle paralysis	Nerve conduction velocity; electromyography[1]
Facioscapulohumeral muscular dystrophy	Progressive weakness in the face, neck, upper torso, and upper arms	Serum creatine kinase; electromyography[1]
Myasthenia gravis	Weakness and fatigue, particularly of the extraocular, pharyngeal, facial, cervical, and respiratory musculature	Computed tomography; electrodiagnostic studies[1]
Sarcoidosis	Shortness of breath, cough, skin and ocular lesions; weakness, fatigue, fever, malaise[7]	Imaging studies; bronchoscopy; lymph node biopsy[1]

References

1. Medline Plus Health Information. Medical encyclopedia. Available at: www.nlm.nih.gov/medlineplus/encyclopedia.html. Accessed February 28, 2002.
2. Facial nerve (VII) paralysis. Neuromuscular. Available at: www.neuro.wustl.edu/neuromuscular/nanatomy/vii.htm#anatomy7. Accessed February 28, 2002.
3. Ruckenstein MJ. Evaluating facial paralysis: expensive diagnostic tests are often unnecessary. Postgrad Med. 1998;103:187-188, 191-192.
4. Keane JR. Bilateral seventh nerve palsy: analysis of 43 cases and review of the literature. Neurology. 1994;44:1198-1202.
5. Balevi B. Melkersson-Rosenthal syndrome; review of the literature and case report of a 10-year misdiagnosis. Quintessence Int. 1997;28:265-269.
6. Abramson DL, Cohen MM Jr, Mulliken JB. Möbius syndrome: classification and grading system. Plast Reconstr Surg. 1998;102:961-967.
7. Belfer MH, Stevens RW. Sarcoidosis: a primary care review. Am Fam Physician. 1998;58:2041-2056.
8. Krasner CG, Cohen SH. Bilateral Bell's palsy and aseptic meningitis in a patient with acute human immunodeficiency virus seroconversion. West J Med. 1993;159:604-605.
9. Capobianco DJ, Brazis PW, Cheshire WP. Idiopathic intracranial hypertension and seventh nerve palsy. Headache. 1997;37:286-288.
10. Bleicher JN, Hamiel S, Gengler JS, Antimarino J. A survey of facial paralysis: etiology and incidence. Ear Nose Throat J. 1996;75:355-358.
11. Marson AG, Salinas R. Bell's palsy. West J Med. 2000;173:266-268.
12. Savettieri G, Salemi G, Rocca WA, et al. Incidence and lifetime prevalence of Bell's palsy in two Sicilian municipalities. Acta Neurol Scand. 1996;94:71-75.
13. Noone J, Longe S. Bell's palsy in the primary care setting: a case study. Clin Excell Nurse Pract. 1998;2:206-211.
14. National Institute of Neurological Disorders and Stroke. Bell's palsy information page. Available at: www.ninds.nih.gov/health_and_medical/disorders/bells_doc.htm. Accessed February 28, 2002.
15. Billue JS. Bell's palsy: an update on idiopathic facial paralysis. Nurse Pract. 1997;22:88, 97-100, 102-105.
16. Jabor MA, Gianoli G. Management of Bell's palsy. J La State Med Soc. 1996;148:279-283.
17. Hato N, Honda N, Gyo K, et al. Treatment of Bell's palsy with acyclovir and prednisolone [in Japanese]. Nippon Jibiinkoka Gakkai Kaiho. 2000;103:133-138.
18. Jalaludin MA. Methylcobalamin treatment of Bell's palsy. Methods Find Exp Clin Pharmacol. 1995;17:539-544.
19. Van Gelder RS, Philippart SM, Bernard BG, et al. Effects of myofeedback and mime-therapy on peripheral facial paralysis. Int J Psychol. 1990;25:191-211.
20. Visscher CM, Lobbezoo F, Naeije M. Treatment of bruxism: physiotherapeutic approach [in Dutch]. Ned Tijdschr Tandheelkd. 2000:107;293-296.

21. Racic G, Denoble PJ, Sprem N, et al. Hyperbaric oxygen as a therapy of Bell's palsy. Undersea Hyperb Med. 1997;24:35-38.
22. Shrode LW. Treatment of facial muscles affected by Bell's palsy with high-voltage electrical muscle stimulation. J Manipulative Physiol Ther. 1993;16:347-352.
23. House JW. Iatrogenic facial paralysis. Ear Nose Throat J. 1996;75:720-723.
24. Tos T, Krag C. Surgical treatment of sequelae after facial paralysis [in Danish]. Ugeskr Laeger. 2001;163:5647-5651.
25. Koshima I, Tsuda K, Hamanaka T, Moriguchi T. One-stage reconstruction of established facial paralysis using a rectus abdominis muscle transfer. Plast Reconstr Surg. 1997;99:234-238.
26. Gantz BJ, Rubinstein JT, Gidley P, Woodworth GG. Surgical management of Bell's palsy. Laryngoscope. 1999;109:1177-1188.
27. Erde EL, Eden AR. Ethics and the care of persons with Bell's palsy. Soc Sci Med. 1991;32:559-563.
28. Neely JG, Neufeld PS. Defining functional limitation, disability, and societal limitations in patients with facial paresis: initial pilot questionnaire. Am J Otol. 1996;17:340-342.