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Guillain-Barre syndrome

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Abstract
The term Guillain-Barré syndrome (GBS), the most frequent cause of acute paralytic neuropathy, covers a number of recognisably distinct variants. The exact cause of GBS is unknown, but 50-70% of cases appear 1-2weeks after a respiratory or gastrointestinal infection, or another immune stimulus that induces an aberrant autoimmune response targeting peripheral nerves and their spinal roots. The interplay between the microbial and host factors that dictate whether and how the immune response shifts towards autoreactivity is still unclear, and nothing is known about the genetic and environmental factors that affect an individual's susceptibility to the disease. All patients with GBS need meticulous monitoring, and can benefit from supportive care and the early start of specific treatment. This review summarises the clinical features and diagnostic criteria of GBS and talks about different symptomatic approach for its management. An analysis of the literature showed that, about one century after it was first described, new information concerning its etiopathogenesis has allowed the development of new treatment strategies that should be started immediately after diagnosis; however, the available therapies are not sufficient in many clients, especially in the presence of the acute inflammatory demyelinating polyneuropathy.

Keywords: Guillain-Barré syndrome, autoimmune, demyelinating polyneuropathy

Introduction

Definition of Guillain-Barré syndrome

Guillain-Barré syndrome (GBS) is an acquired disorder of the peripheral nerves, described best as a polyradiculoneuropathy. With the widespread eradication of poliomyelitis, GBS is the most common cause of acute and subacute flaccid paralysis in infants and children.

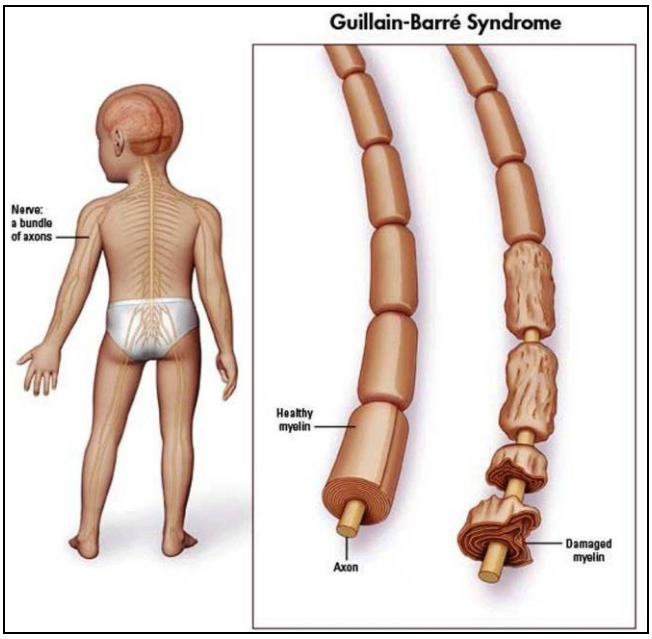

Fig 1

Etiology of Guillain-Barré syndrome

The Guillain-Barre syndrome (GBS) and its variants are considered post-infectious, immune-mediated neuropathies. Evidence from animal models suggests a key role of molecular mimicry. In Campylobacter jejuni gastrointestinal infections, a lipooligosaccharide present in the outer membrane of the bacteria is similar to gangliosides that are components of the peripheral nerves. Therefore, an immune response triggered to fight infection can lead to a cross-reaction on host nerves.

Many infections have been linked with GBS. The most common are gastrointestinal or respiratory illnesses. Up to 70% of patients have reported an antecedent illness in the 1 to 6 weeks before the presentation of GBS. During the Zika virus outbreak, many GBS cases were described [4] and the results of this study indicate GBS pathophysiologic mechanisms that may be more common after Zika infection. Case reports detail many other possible etiologies linked to GBS including medications and surgeries [5].

In 1976, flu vaccination against the influenza A/H1N1 antigen led to a well-documented, increased incidence of cases of GBS; however, further surveillance data of flu vaccinations in subsequent years have described only one additional case of GBS for every 1 million vaccines. Subsequent studies estimate that developing GBS after a flu infection is up to 7 times more likely than developing GBS after a vaccination [5].

Causes of Guillain-Barré syndrome

Most children who develop Guillain-Barré syndrome have a bacterial or viral infection prior to developing the signs and symptoms of the condition. However, only a very small percentage of children who have an infection develop Guillain-Barré syndrome. In order to fight the infection, specialized immune cells produce proteins called antibodies that recognize specific proteins or molecules on the bacteria or virus (pathogen). Some

research shows that antibodies that recognize molecules on some pathogens may also recognize proteins on the body's own nerves. As a result, the immune system attacks the nerves, causing inflammation and damaging the axons and myelin, which can lead to the signs and symptoms of Guillain-Barré syndrome.

Inheritence Pattern

Almost all cases of Guillain-Barré syndrome are sporadic, which means they occur in people with no history of the condition in their family. A few families with more than one affected family member have been described; however, the condition does not have a clear pattern of inheritance. Multiple genetic and environmental factors likely play a part in determining the risk of developing this condition. As a result, inheriting a genetic variation linked with Guillain-Barré syndrome does not mean that a child will develop the condition.

Symptoms of Guillain-Barré syndrome

Symptoms can occur a bit differently in each child. They can include:

- Decreased feeling in fingers and toes
- Pain in fingers and toes
- Leg weakness
- Leg pain that moves to the arms
- Problems walking
- Irritability
- Breathing problems
- Trouble swallowing
- Facial weakness
- Vision changes

Clinical Presentation of Guillain-Barré syndrome

The typical patient with GBS presents 2-4 weeks following a relatively benign gastrointestinal or respiratory illness with complaints of finger dysesthesias and proximal muscle weakness of the lower limbs. The weakness may progress over hours to days to involve the arms, trunk, cranial nerves, and muscles of respiration. Variants of GBS may present as pure motor dysfunction or acute dysautonomia. a. "Typical" 'GBS is an acute, predominantly motor neuropathy involving distal limb paresthesias, relatively symmetric leg weakness, and frequent gait ataxia.

a) Most cases will have subsequent arm weakness, and possibly the weakness of facial, ocular, and oropharyngeal muscles.
b) Weakness is always bilateral, although some asymmetry in onset and severity is common.
    i) Proximal muscle weakness very frequent, especially initially, with subsequent distal arm and leg weakness.
    ii) GBS with a descending pattern of weakness seen in 14% cases; onset initially with cranial nerve or arm muscle weakness, followed by leg weakness.
    iii) In 1/3 of cases, the degree of weakness in the arms and legs is roughly equal.
c) Reduced or absent reflexes characterize GBS.
    i) Early loss of reflexes may be due to desynchronization of afferent impulses in reflex arc due to non-uniform demyelination.
    ii) About 70% of patients present with loss of reflexes; less than 5% retained all reflexes during the illness;
    iii) The presence of intact reflexes should suggest an alternative diagnosis other than GBS.
d) Sensory disturbance
    i) >50% will present with symmetric distal limb paresthesias, before clinically evident limb weakness. Early finger paresthesias suggest a patchy process, unlike the pattern seen with distal axonopathies.
    ii) Paresthesias of trunk or face unusual, but sensory loss over the trunk frequent and a psuedolevel may be evident
    iii) Beware if definite sensory level present as this may suggest structural cord disease.
e) Dysautonomia
    i) occurs in about 65% of cases
    ii) More frequent in patients with severe paralysis and ventilator difficulties but may develop in mild cases.
    iii) Most common manifestations include cardiac dysfunction such as sinus tachycardia, sinus bradycardia, sinus arrest and other supraventricular arrhythmias, paroxysmal hypertension, and hypotension (especially postural),
    iv) ICU monitoring necessary because of possible cardiac complications.
    v) Other features: ileus, urinary retention (1/4 cases), inappropriate ADH, altered sweating, mild orthostatic hypotension.
f) Cranial nerve involvement is observed in 45-75% of patients with GBS. Cranial nerves III-VII and IX-XII may be affected. Common complaints include:
    i) Facial Palsy
    ii) Diplopia
    iii) Dysarthria
    iv) Dysphagia
    v) Ophthalmoplegia
    vi) Pupillary disturbances.
    vii) Facial and oropharyngeal weakness usually appears after the trunk and limbs are affected. The Miller-Fisher variant of GBS is unique in that this subtype begins with cranial nerve deficits [12].

Differential Diagnosis of Guillain-Barré syndrome

a) Acute peripheral neuropathies

i) Systemic vasculitis
ii) Poliomyelitis
iii) Diphtheria
iv) Tick paralysis
v) Critical illness polyneuropathy

b) Disorders of Neuromuscular Transmission

i) Botulism
ii) Myasthenia gravis

c) Central Nervous system Disorders

i) Basilar artery occlusion
ii) Acute cervical transverse myelitis.

Diagnostic Procedures of Guillain-Barré syndrome

- **Blood tests and urine tests.** These are done to check for infections and other problems.
- **Spinal tap (lumbar puncture).** This test uses a needle to help measure the pressure in the spinal canal and brain. The healthcare provider can also remove a small amount of cerebrospinal fluid (CSF) to send for testing.

CSF is the fluid that surrounds your child's brain and spinal cord. The fluid sample can help show if your child has an infection or other problems.

- **Electromyogram (EMG) and nerve conduction studies.** These tests measure the electrical activity of nerves and muscles. An EMG can find abnormal electrical muscle activity caused by diseases and conditions that affect the nerves and muscles. They are normal in the early stages but show typical changes after a week or so with conduction block and multifocal motor slowing, sometimes most evident proximally as delayed F-waves. The only way to classify a patient as having the axonal or nonaxonal type is electro diagnostically.
- Pulmonary function test. This is a breathing test done by a respiratory therapist. It shows your child's lung capacity and how strong his or her respiratory muscles are. This test is often used to decide if a child needs breathing support with a ventilator.
- Further investigative procedures can be undertaken to identify an underlying cause.
  For example
  i) Chest X-ray, stool culture and appropriate immunological tests to rule out the presence of cytomegalovirus or mycoplasma
  ii) Antibodies to the ganglioside GQ1b for Miller Fisher Variant.

Treatment of Guillain-Barré syndrome

Treatment will depend on your child's symptoms, age, and general health. It will also depend on how severe the condition is.

The goal of treatment is

- To prevent breathing problems and
- To ease symptoms.

Plasmapheresis

In plasmapheresis, blood is removed from the body, the red and white blood cells are separated from the plasma and only the blood cells are returned to the patient. It is thought that removing the plasma eliminates some of the immune factors that are responsible for the disease progression. Plasmapheresis helps in following ways:

a. Reducing the length of the illness
b. Shortened time on mechanical ventilation
c. Early ambulation

Intravenous Immune Globulin

IVIG has been shown to be safe and effective in the treatment of pediatric GBS. Although only one prospective, randomized treatment trial in childhood GBS has been published, multiple studies have shown that IVIG seems helpful in reducing the severity of the disease as well as the duration of symptoms. However, the long-term outcome may not be affected.

3. Futher medical management can be done according to medical symptoms.

A) Supportive Care

i) ICU monitoring
ii) Basic medical management often determines mortality and morbidity.

B) Ventilatory support

i) Atelectasis leads to hypoxia.
ii) Hypercarbia later finding; arterial blood gases may be misleading.
iii) Vidal capacity, tidal volume and negative inspiratory force are best indicators of diaphragmatic function.
iv) Progressive decline of these functions indicates an impending need or ventilatory assistance. Mechanical ventilation usually required if VC drops below about 14 ml/kg; ultimate risk depending on age, the presence of accompanying lung disease, aspiration risk, and assessment of respiratory muscle fatigue.
v) Atelectasis treated initially by incentive spirometry, frequent suctioning, and chest physiotherapy to mobilize secretions.
vi) Intubation may be necessary for patients with substantial oro-pharyngeal dysfunction to prevent aspiration.
vii) Tracheostomy may be needed in patients intubated for 2 weeks who do not show improvement.

C) Autonomic dysfunction

i) Autonomic dysfunction may be self-limited; do not over-treat.
ii) Sustained hypertension managed by angiotensin-converting enzyme inhibitor or beta-blocking agent. Use short-acting intravenous medication for labile hypertension requiring immediate therapy.
iii) Postural hypotension treated with fluid bolus or positioning.
iv) Urinary difficulties may require intermittent catheterization.

D) Nosocomial infections usually involve pulmonary and Urinary tracts

i) Occasionally central venous catheters become infected.
ii) Antibiotic therapy should be reserved for those patients showing clinical infection rather than colonization of fluid or sputum specimens.

E) Venous thrombosis due to immobilization poses a great risk of thromboembolism.

i) Prophylactic use of subcutaneous heparin and compression stockings.

Physiotherapy Management

Aims of physiotherapy management are:

1. Regain the patient's independence with everyday tasks.
2. Retrain the normal movement patterns.
3. Improve patients posture.
4. Improve the balance and coordination
5. Maintain clear airways
6. Prevent lung infection
7. Support joint in functional position to minimize damage or deformity
8. Prevention of pressure sores
9. Maintain peripheral circulation
10. Provide psychological support for the patient and relatives.

Respiratory Care

The common respiratory complications in the rehabilitation setting include incomplete respiratory recovery including

chronic obstructive pulmonary disease, restrictive respiratory disease (pulmonary scarring, pneumonia), and trachitis from chronic intubation and respiratory muscle insufficiency. Sleep hypercapnia and hypoxia, which worsens during sleep can be the result of a restrictive pulmonary function.

Treatment methods are

- Night time saturation records with pulse oximeter and bilevel positive airway pressure (BiPAP) may be indicated for the patients.
- Physical therapy measures (chest percussion, breathing exercises, resistive inspiratory training) may be required to clear respiratory secretions to reduce the work of breathing.
- Special weaning protocol to prevent over fatigue of respiratory muscles can be recommended for more severe patients with tracheostomy. Patients with cranial nerve involvement need extra monitoring as they are more prone to respiratory dysfunction.
- Patients should be encouraged to cease smoking.
- Posturally drain areas of lung tissues, 2-hourly turning into supine or side lying positions.
- 2-4 litre anaesthetic bag can be used to enhance chest expansion. Therefore, 2 people are necessary for this technique, one to squeeze the bag and another to apply chest manipulation.
- Rib springing to stimulate cough
- After the removal of a ventilator and adequate expansion, effective coughing must be taught to the patient

Maintain Normal Range of Movement

Gentle passive movements through full ROM at least three times a day especially at hip, shoulder, wrist, ankle, feet.

Orthoses

Use of light splints (e.g. using PLASTAZOTE) may be required for the following purpose listed below:

a) Support the peripheral joints in comfortable and functional position during flaccid paralysis.
b) To prevent abnormal movements.
c) To stabilize patients using sandbags, pillows.

Prevention of Pressure Sores

2- Hourly change in patients position from supine to side lying. If the sores have developed then UVR or ice cube massage to enhance healing.

Maintenance of Circulation

a) Passive movements
b) Effleurage massage to lower limbs.

Relief of pain

a) Transcutaneous electrical nerve stimulation
b) Massage with passive ROM
c) Patient can demonstrate increased sensitivity to light touch, a cradle can be used to keep the bed sheet away from the skin. Low-pressure wrapping or snug fitting garments can provide a way to avoid light touch.
d) Reassurance and explanation of what to expect can help in alleviation of anxiety that could compound the pain.

According to Bensman (1970), the following four guidelines are to be followed for prescription of exercises:

a) Use short periods of non-fatiguing exercises matched to the patients strength.
b) Progression of the exercise should be done only if the patient improves or if there is no deterioration in status after a week.
c) Return the patient to bed rest if a decrease in muscle strength or function occurs.
d) The objective should be directed towards not only at improving function but also in improving strength.

References

1. Guillain-Barré Syndrome Fact Sheet http://www.ninds.nih.gov/disorders/gbs/detail_gbs.htm
2. Lo YL. Clinical and immunological spectrum of the Miller Fisher syndrome. Muscle & nerve. 2007; 36(5):615-27.
3. Dirlikov E, Major CG, Medina NA, Lugo-Robles R, Matos D, Muñoz-Jordan JL. *et al*. Clinical Features of Guillain-Barré Syndrome With vs Without Zika Virus Infection, Puerto Rico, 2016. JAMA neurology. 2018; 175(9):1089-97.
4. Nguyen TP, Taylor RS. Guillain Barre Syndrome. [Updated 2018 Oct 27]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2018. Available from: https://www.ncbi.nlm.nih.gov/books/NBK532254/
5. Skayhue Guillain-Barre Syndrome. wmv Available from: https://www.youtube.com/watch?time_continue=3&v=hnvw2zFd-Um.
6. Kuwabara S. Guillain-barré syndrome epidemiology, pathophysiology and management. Drugs. 2004; 1; 64(6):597-610.
7. http://www.ninds.nih.gov/disorders/gbs/detail_gbs.htm
8. Alshekhlee A, Hussain Z, Sultan B, Katirji B. Guillain-Barré syndrome incidence and mortality rates in US hospitals. Neurology. 2008; 70(18):1608-13.
9. Kushnir M, Klein C, Pollak L, Rabey JM. Evolving pattern of Guillain-Barre syndrome in a community hospital in Israel. Acta Neurol Scand. 2008; 117(5):347-50
10. Prevots DR, Sutter RW. Assessment of Guillain-Barré syndrome mortality and morbidity in the United States: implications for acute flaccid paralysis surveillance. J Infect Dis. 1997;175(1):S151-5
11. Lo YL. Clinical and immunological spectrum of the Miller Fisher syndrome. Muscle Nerve. 2007; 36(5):615-27.