Guillain-Barré syndrome

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Abstract

Guillain-Barré syndrome (GBS) is a rare immune-mediated neuropathy that occurs in previously healthy individuals. The incidence of GBS is 1.18 in 100,000 per year and increases with age. GBS clinical spectrum is heterogeneous and encompasses acute inflammatory demyelinating polyneuropathy (AIDP), acute motor axonal neuropathy (AMAN), acute motor and sensory axonal neuropathy (AMSAN) and Miller Fisher Syndrome (MFS). The disease is characterized by a rapid onset of symmetrical limb weakness, which progresses over days to 4 weeks, and occurs in patients of all ages. Most patients also have sensory disturbances (tingling or dull feelings). In the majority of cases, an infectious disease, mostly Campylobacter jejuni infection, precedes the onset of limb weakness. Due to progression of weakness, about 25% of patients need artificial ventilation for some time. Eventually muscle strength improves and sensory disturbances decrease. Many patients remain disabled for months or even years. Fatigue and endurance intolerance may persist for years. Optimal general hospital care providing intensive-care facilities is most essential. Treatment with intravenous immunoglobulin (IVIg) or plasma exchange (PE) was shown to be effective. Physiotherapy and rehabilitation are important. Immune responses against the triggering infectious agents are thought to be involved in the pathogenesis of GBS by cross reaction with neural tissues.

Keywords

Guillain-Barré syndrome – GBS – Miller Fisher Syndrome – Acute motor axonal neuropathy (AMAN) – Intravenous immunoglobulin (IVIg) – plasma exchange (PE) – ganglioside antibodies

Disease name and synonyms

Guillain-Barré syndrome (GBS) is the covering name of the syndrome. GBS is characterized by a heterogeneous clinical spectrum. The most frequent form in the Western World is the acute inflammatory demyelinating polyneuropathy (AIDP). The less frequent forms are: Acute Motor Axonal Neuropathy (AMAN), Acute Motor and Sensory Axonal Neuropathy (AMSAN) and the cranial nerve variant Miller Fisher Syndrome (MFS).
Excluded diseases
Other diseases that may give rise to rapid progressive weakness have to be excluded or made unlikely. The main disorders that may give rapid progressive weakness are electrolyte disturbances (hypophosphatemia, hyperkalemia), porphyria, polymyositis or necrotising myopathies, myasthenia gravis, poliomyelitis and Lyme borreliosis.

Diagnosis criteria / definition
GBS is a neuropathy characterized by a rapidly progressive weakness (and mostly also sensory disturbances) in at least 2 legs, and low or absent tendon reflexes. Cranial nerves may be affected. The maximal level of weakness is reached within 4 weeks, meaning that the duration of progression is at most 4 weeks. Cerebrospinal fluid (CSF) examination mostly shows an increased protein level, but no increased number of cells. Other causes of rapid progressive weakness need to be excluded or must be unlikely.

Differential diagnosis
History taking, standard blood tests and cerebrospinal fluid examination – in general – are sufficient to exclude other diseases. An electromyography (EMG) examination may be helpful to establish the diagnosis. It is necessary to make the distinction between AIDP and AMAN or AMSAN.

Frequency
The incidence of GBS is 1.18 in 100,000 per year and increases with age.

Clinical description
GBS onset is sudden and occurs at all ages in a previously healthy person. Most patients with GBS have had an infection within the 3 weeks prior to onset of weakness, most commonly consisting of diarrhoea (due to Campylobacter jejuni) or an upper respiratory infection. Patients may experience sensory disturbances (tingling or dull feelings in the hands and feet) a few days before the onset of weakness. Weakness most frequently starts in the distal legs, but may also first appear in the upper legs or in the arms. Most patients reach their maximal level of weakness within the first 2 weeks after onset. By definition, progression of weakness does not exceed 4 weeks. About one third of GBS patients remains able to walk during the course of disease. After a plateau phase of variable duration (weeks to months), muscle strength improves. Despite treatment, about 20% of patients are unable to walk unaided 6 months after onset of disease.

Many patients complain about fatigue and endurance intolerance during months or even years after they have had GBS. The probability to develop the chronic variety of GBS (chronic inflammatory demyelinating polyneuropathy/CIDP) or to have a second bout of GBS is very low.

Management including treatment
Optimal general hospital care providing intensive-care facilities is most essential. Besides prevention and treatment of general complications, active treatment has become available. A Cochrane Database review of selected trials showed that plasma exchange (PE) or treatment with intravenous immunoglobulin (IVIg) have equivalent efficacy in hastening recovery from GBS in patients when started within the first 2 weeks after onset of weakness. About 10% of patients need to be retreated because they have secondary progression of weakness after initial improvement following PE or IVIg. Mostly due to practical reasons (e.g. advantage of low risk and easy application), treatment with IVIg is presently the first-line treatment for patients with GBS. It is important to start physiotherapy in an early phase to prevent secondary problems like joint stiffness. Rehabilitation is very important and should be carried out once the patient is in a stable condition and able to do some exercise and to follow a training programme.

The subcommittee of the American Academy of Neurology has recently given the following recommendations for the treatment of GBS:
1) PE is recommended for nonambulant adult patients with GBS who seek treatment within 4 weeks of the onset of neuropathic symptoms. PE should also be considered for ambulant patients examined within 2 weeks of the onset of neuropathic symptoms; 2) IVIg is recommended for nonambulant adult patients with GBS within 2 or possibly 4 weeks of the onset of neuropathic symptoms; 3) Corticosteroids are not recommended for the management of GBS; 4) Sequential treatment with PE followed by IVIg, or immunoabsorption followed by IVIg is not recommended for patients with GBS; and 5) PE and IVIg are treatment options for children with severe GBS.
A recent trial compared the effect of IVIg, methylprednisolone and placebo. It showed that the combination of IVIg and steroids might be somewhat better when adjustment was made for well-known prognostic factors. The exact role of steroids in GBS remains to be established.

Pathogenesis
In AIDP, the peripheral nerve and spinal roots are initially infiltrated by T lymphocytes and...
GBS in general including general aspects of treatment

References

GBS in general including general aspects of treatment


Intravenous immunoglobulin (IVIg)


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**Plasma exchange (PE)**


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**Mechanisms of disease**


