Guillain-Barré syndrome

Guillain-Barré syndrome is named after the French neurologists Georges Guillain and Jean Alexandre Barré, who described it with André Strohl in 1916.

The number of cases of GBS associated with COVID-19 is continuously increasing since the date of a review (May 17, 2020) [1)]

Key concepts

● acute onset of peripheral neuropathy with progressive muscle weakness (more severe proximally) with areflexia, reaches maximum over 3 days to 3 weeks

● cranial neuropathy: also common, may include facial diplegia, ophthalmoplegia

● little or no sensory involvement (paresthesias are not uncommon)

● onset often 3 days-5 weeks following viral URI, immunization, Campylobacter jejuni enteritis, or surgery

● pathology: focal segmental demyelination with endoneurial monocytic infiltrate

● elevated CSF protein without pleocytosis (albuminocytologic dissociation)

Guillain–Barré syndrome (GBS) is a rapid-onset muscle weakness caused by the immune system damaging the peripheral nervous system.

Clinical features

Many experiences changes in sensation or develops pain, followed by muscle weakness beginning in the feet and hands. The symptoms develop over half a day to two weeks. During the acute phase, the disorder can be life-threatening with about a quarter developing weakness of the breathing muscles and requiring mechanical ventilation. Some are affected by changes in the function of the autonomic nervous system, which can lead to dangerous abnormalities in heart rate and blood pressure.

This autoimmune disease is caused by the body's immune system mistakenly attacking the peripheral nerves and damaging their myelin insulation. Sometimes this immune dysfunction is triggered by an infection. The diagnosis is usually made based on the signs and symptoms, through the exclusion of alternative causes, and supported by tests such as nerve conduction studies and examination of the cerebrospinal fluid. Various classifications exist, depending on the areas of weakness, results of nerve conduction studies, and the presence of antiganglioside antibodies. It is classified as an acute polyneuropathy.

In those with severe weakness, prompt treatment with intravenous immunoglobulins or plasmapheresis, together with supportive care, will lead to good recovery in the majority. Some may experience ongoing difficulty with walking, painful symptoms, and some require long-term breathing support. Guillain–Barré syndrome is rare, at one to two cases per 100,000 people every year.

In Guillain-Barré syndrome (GBS) with antiganglioside antibodies, isolated absence of F waves is a frequent conduction abnormality especially in the early phase of the disease, and may be caused by axonal dysfunction, such as physiological conduction block or axonal degeneration at the nerve roots [2)].

Guillain-Barré variants

General information

A number of variants have been described (some may simply be incomplete forms of typical Guillain-Barré). Autonomic dysfunction may occur in some.

Miller-Fisher variant of GBS Ataxia, areflexia and ophthalmoplegia. May also have ptosis. 5% of cases of GBS. Serum marker: anti- GQ1b antibodies.

Acute motor axonal neuropathy (AMAN) This variant and AIDP are the most common to follow Campylobacter jejuni enteritis.

Pharyngeal-cervical-brachial variant Facial, oropharyngeal, cervical, and UE weakness, sparing the LEs.

Pure sensory variant Sensory loss accompanied by areflexia.

Atypical GBS May be accompanied by rhabdomyolysis

Treatment

Immunoglobulins may be helpful. In severe cases, early plasmapheresis hastens the recovery and reduces the residual deficit. Its role in mild cases is uncertain. Steroids are not helpful.

Mechanical ventilation and measures to prevent aspiration are used as appropriate. In cases of facial diplegia, the eyes must be protected from exposure (neuroparalytic) keratitis.

Outcome

Recovery may not be complete for several months. 35% of untreated patients have residual weakness and atrophy. Recurrence of GBS after achieving maximal recovery occurs in ≈ 2%.

Almost a third of patients with Guillain-Barré Syndrome (GBS) require mechanical ventilation, increasing mortality by 15-30% and proving poor functional outcomes. The Erasmus GBS Respiratory Insufficiency Score (EGRIS) is the most frequently used scale to assess the probability of respiratory insufficiency within the first week of admission. provided new specific clinical (deltoid muscle strength and autonomic dysfunction) and electrophysiological variables to discriminate GBS patients that will require IMV [3)]

Case series

From the International GBS Outcome Study, we selected patients whose modified Erasmus GBS Outcome Score at week 1 predicted a poor prognosis. We compared those treated with one IVIg course to those treated with two IVIg courses. The primary endpoint, the GBS disability scale at 4 weeks, was assessed with multivariable ordinal regression.

RESULTS: Of 237 eligible patients, 199 patients received a single IVIg course. Twenty patients received an 'early' second IVIg course (1-2 weeks after start of the first IVIg course) and 18 patients a 'late' second IVIg course (2-4 weeks after start of IVIg). At baseline and 1 week, those receiving two IVIg courses were more disabled than those receiving one course. Compared with the one course group, the adjusted OR for a better GBS disability score at 4 weeks was 0.70 (95%CI 0.16 to 3.04) for the early group and 0.66 (95%CI 0.18 to 2.50) for the late group. The secondary endpoints were not in favour of a second IVIg course.

This observational study did not show better outcomes after a second IVIg course in GBS with poor prognosis. The study was limited by small numbers and baseline imbalances. Lack of improvement was likely an incentive to start a second IVIg course. A prospective randomised trial is needed to evaluate whether a second IVIg course improves outcome in GBS [4)].

Case reports

Agosti et al. reported a case of COVID-19 patient with acute monophasic Guillain-Barré syndrome (GBS), and a literature review on the SARS-CoV-2 and GBS etiological correlation.

A 68 years-old man presented to the emergency department with symptoms of acute progressive symmetric ascending flaccid tetraparesis. Oropharyngeal swab for SARS-CoV-2 tested positive. Neurological examination showed bifacial nerve palsy and distal muscular weakness of lower limbs. The cerebrospinal fluid assessment showed an albuminocytologic dissociation. Electrophysiological studies showed delayed distal latencies and absent F waves in early course. A diagnosis of Acute Inflammatory Demyelinating Polyradiculoneuropathy (AIDP) subtype of GBS was then made.

Neurological manifestations of COVID-19 are still under study. The case we described of GBS in COVID-19 patient adds to those already reported in the literature, in support of SARS-CoV-2 triggers GBS. COVID-19 associated neurological clinic should probably be seen not as a corollary of classic respiratory and gastrointestinal symptoms, but as SARS-CoV-2-related standalone clinical entities. To date, it is essential for all Specialists, clinicians, and surgeons, to direct attention towards the study of this virus, to better clarify the spectrum of its neurological manifestations [5)].

1)
Doneddu PE, De Sanctis P, Viganò L, Selmi C, Gentile F, Nobile-Orazio E. Response to: SARS-CoV-2 associated Guillain-Barré syndrome in 62 patients. Eur J Neurol. 2020 Sep 25. doi: 10.1111/ene.14543. Epub ahead of print. PMID: 32978868.
2)
Kuwabara S, Ogawara K, Mizobuchi K, Koga M, Mori M, Hattori T, Yuki N. Isolated absence of F waves and proximal axonal dysfunction in Guillain-Barré syndrome with antiganglioside antibodies. J Neurol Neurosurg Psychiatry. 2000 Feb;68(2):191-5. PubMed PMID: 10644786; PubMed Central PMCID: PMC1736791.
3)
López-Hernández JC, Jorge de Saráchaga A, Briseño-Godínez ME, May-Mas RN, Bazán-Rodríguez L, León-Manriquez E, Cruz-Pérez J, Becerra-Hernández I, Vargas-Cañas ES. Deltoid Muscle Strength and Autonomic Dysfunction as Independent Risk Factors for Invasive Mechanical Ventilation in Patients with Guillain-Barré Syndrome. Int J Neurosci. 2022 May 27:1-13. doi: 10.1080/00207454.2022.2082963. Epub ahead of print. PMID: 35633066.
4)
Verboon C, van den Berg B, Cornblath DR, Venema E, Gorson KC, Lunn MP, Lingsma H, Van den Bergh P, Harbo T, Bateman K, Pereon Y, Sindrup SH, Kusunoki S, Miller J, Islam Z, Hartung HP, Chavada G, Jacobs BC, Hughes RAC, van Doorn PA; IGOS Consortium. Second IVIg course in Guillain-Barré syndrome with poor prognosis: the non-randomised ISID study. J Neurol Neurosurg Psychiatry. 2019 Oct 5. pii: jnnp-2019-321496. doi: 10.1136/jnnp-2019-321496. [Epub ahead of print] PubMed PMID: 31586949.
5)
Agosti E, Giorgianni A, D'Amore F, Vinacci G, Balbi S, Locatelli D. Is Guillain-Barrè syndrome triggered by SARS-CoV-2? Case report and literature review [published online ahead of print, 2020 Jul 9]. Neurol Sci. 2020;10.1007/s10072-020-04553-9. doi:10.1007/s10072-020-04553-9