

GUILLAIN BARRÉ SYNDROME

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Guillain-Barré syndrome, or acute post-infectious polyneuropathy, is an acute, ascending, symmetric paralytic disorder diagnosed by consensus clinical criteria. Acute inflammatory demyelinating polyneuropathy (AIDP) is the classic form of this syndrome.

Epidemiology

Guillain-Barré syndrome has replaced poliomyelitis as the most common cause of acute flaccid paralysis worldwide. Its annual incidence in the U.S. increases with age, from 0.8 per 100,000 in individuals younger than 18 years to 3.25 per 100,000 in those over 60 years. There is an overall male to female preponderance of 1.5 to one for Guillain-Barré syndrome occurring in later life.

Guillain-Barré syndrome is associated with antigenic triggers, including minor respiratory infections in 50% of cases. *Campylobacter jejuni* infections are associated with 26% to 40% of cases in industrialized nations. Fewer than 5% of cases are vaccine-associated. Guillain-Barré syndrome has been reported with primary HIV infection and as part of the immune reconstitution syndrome during therapy.

Conditions associated with Guillain-Barré syndrome:

- Non-specific respiratory tract symptoms
- *Campylobacter jejuni*
- *Mycoplasma pneumoniae*
- Cytomegalovirus
- Epstein-Barr virus
- HIV-1
- Japanese encephalitis virus
- Hodgkin's disease
- Lymphoma
- Systemic lupus erythematosus
- Surgery
- Parturition
- Rabies
- Influenza A/New Jersey/76
- Vaccines
- Vaccinia

The 1976 swine influenza vaccine was associated with an increased frequency of Guillain-Barré syndrome. Among persons who received the swine influenza vaccine in 1976, the rate of Guillain-Barré syndrome was less than ten cases per one million persons vaccinated. Evidence for a causal relation of Guillain-Barré syndrome with subsequent vaccines prepared from other influenza viruses is unclear. Obtaining strong epidemiologic evidence for a possible limited increase in risk is difficult for such a rare condition as Guillain-Barré syndrome, which has an annual incidence of ten to 20 cases per one million

adults. During three of four influenza seasons studied during 1977 to 1991, the overall relative risk estimates for Guillain-Barré syndrome after influenza vaccination were slightly elevated, but were not statistically significant in any of these studies. However, in a study of the 1992 to 1993 and 1993 to 1994 influenza seasons, the overall relative risk for Guillain-Barré syndrome was 1.7 (95% CI = 1.0--2.8; $p = 0.04$) during the six weeks after vaccination, representing approximately one additional case of Guillain-Barré syndrome per one million persons vaccinated. The combined number of Guillain-Barré syndrome cases peaked two weeks after vaccination. Thus, investigations to date indicate that there is no substantial increase in Guillain-Barré syndrome associated with influenza vaccines (other than the swine influenza vaccine in 1976), and that, if influenza vaccine does pose a risk, it is probably slightly more than one additional case per one million persons vaccinated. Cases of Guillain-Barré syndrome after influenza infection have been reported, but no epidemiologic studies have documented such an association.

Clinical Description

Guillain-Barré syndrome is an acute monophasic demyelinating syndrome of peripheral nerves. It is multifocal, with a predilection for nerve roots. Demyelination is associated with deposits of antibody and complement as well as macrophage 'stripping' of outer myelin lamellae.

The acute afebrile paresis progresses over a few days to weeks. Initial symptoms are paresthesia in toes or fingers which occur before motor findings. Weakness is generally more profound in the legs but may predominantly affect the arms or cranial nerves in up to 10% of cases each. Weakness begins in the extremities and progresses centrally; progression may be quite rapid. As Guillain-Barré syndrome evolves, general symmetry of paresis is the rule. Dysfunction of the autonomic nervous system may be prominent but bowel or bladder incontinence is rare. Pain is common, involving the large muscles of the legs and back.

Examination shows relatively symmetric motor weakness, absent or greatly depressed tendon reflexes and minimal loss of sensation despite sensory complaints. In severe cases, respiration, airway control and autonomic function are affected.

Guillain-Barré syndrome is complicated by unstable blood pressure, cardiac arrhythmias or thrombosis. Mortality with optimal supportive care and treatment is less than 5%.

Guillain-Barré may resemble paralytic poliomyelitis but fever, headache, nausea, vomiting, and increased lymphocyte count in CSF are usually absent.

Laboratory Tests

There is no specific laboratory diagnosis.

Surveillance

Guillain-Barré syndrome is not a reportable condition except for outbreaks.

Case Definition

A case of Guillain-Barré syndrome is defined as an illness characterized by clinical symptoms beginning with progressive ascending weakness (usually beginning peripherally in the limbs), impairment of position and vibration sense, reduced or absent tendon reflexes and an elevated CSF protein with abnormal EMG results (indicating motor neuron involvement).

Investigation

The purpose of investigation is to identify cases, to elicit triggering infections and risk factors.

If an investigation is warranted:

- Upon receipt of a report of a case of Guillain-Barré syndrome, contact the physician and/or the hospital to confirm the diagnosis
- Determine whether the patient has had a recent respiratory, gastrointestinal or other illness or has recently received an immunization.
- If the history reveals a recent immunization, collect details about the vaccine: administration by private or public clinic, date, vaccine, lot number...
- All clinically significant adverse events after vaccination of children should be reported to Vaccine Adverse Events Reporting System (VAERS), even if it is not certain that the vaccine caused the event. It is specifically recommended to report demyelinating disorders such as Guillain-Barré syndrome, although no evidence exists of a causal relationship between influenza vaccine and neurologic disorders in children.

Treatment

Plasma exchange has yielded better results than supportive treatment alone. Intravenous immunoglobulin can speed the recovery from Guillain-Barré syndrome. Corticosteroids are not effective. The natural history of Guillain-Barré syndrome, with spontaneous full recovery in 80% of cases, makes it difficult to evaluate effective treatment. Guillain-Barré syndrome requires expert diagnosis and management available at tertiary referral centers.

Hospital precaution and isolation: Standard precautions