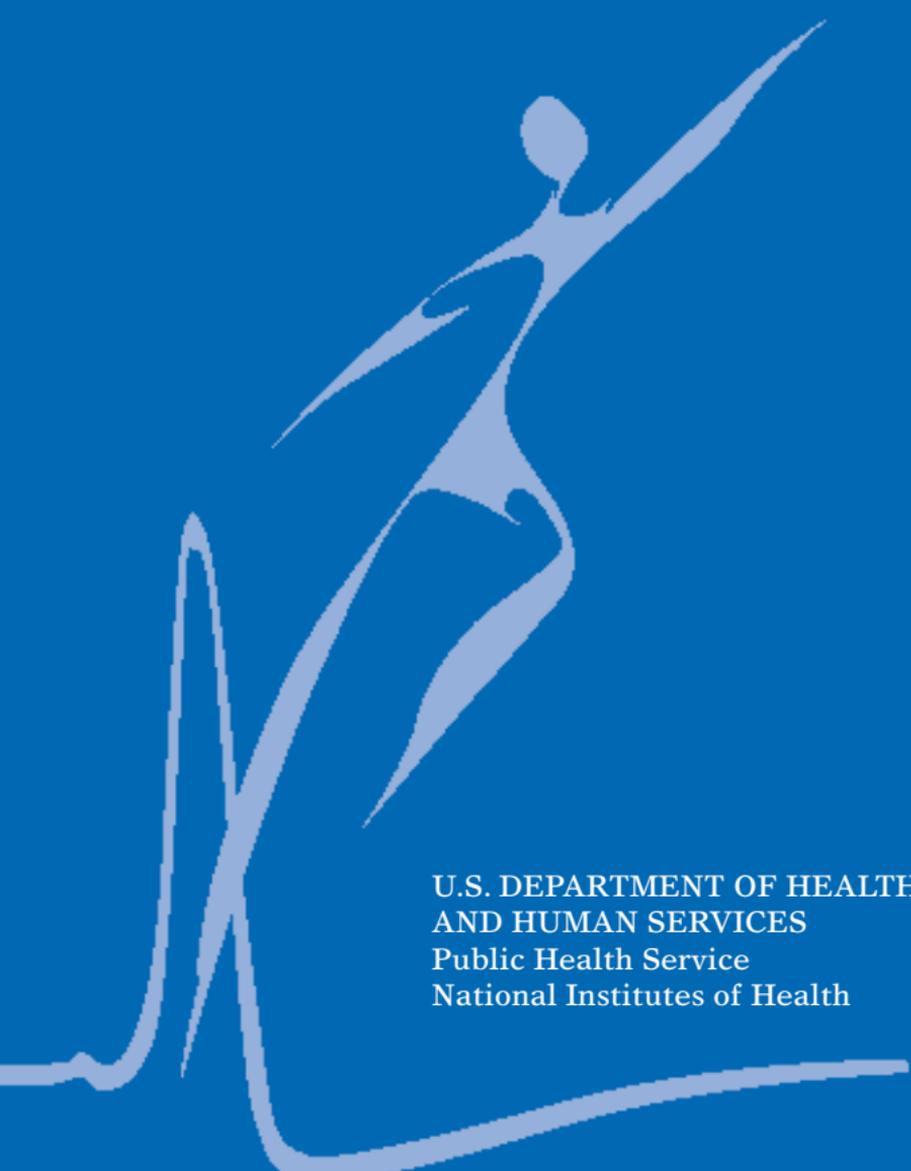


Guillain-Barré Syndrome



U.S. DEPARTMENT OF HEALTH
AND HUMAN SERVICES
Public Health Service
National Institutes of Health

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What is Guillain-Barré syndrome?

Guillain-Barré syndrome is a disorder in which the body's immune system attacks part of the peripheral nervous system. The first symptoms of this disorder include varying degrees of weakness or tingling sensations in the legs. In many instances the symmetrical weakness and abnormal sensations spread to the arms and upper body. These symptoms can increase in intensity until certain muscles cannot be used at all and, when severe, the patient is almost totally paralyzed. In these cases the disorder is life threatening—potentially interfering with breathing and, at times, with blood pressure or heart rate—and is considered a medical emergency. Such an individual is often put on a ventilator to assist with breathing and is watched closely for problems such as an abnormal heart beat, infections, blood clots, and high or low blood pressure. Most people, however, recover from even the most severe cases of Guillain-Barré syndrome, although some continue to have a certain degree of weakness.

Guillain-Barré syndrome (GBS) can affect anybody. It can strike at any age and both sexes are equally prone to the disorder. The syndrome is rare, however, afflicting only about one person in 100,000. Usually GBS occurs a few days or weeks after the patient has had symptoms of a respiratory or gastrointestinal viral infection. Occasionally surgery will trigger the syndrome. In rare instances vaccinations may increase the risk of GBS.

After the first clinical manifestations of the disease, the symptoms can progress over the course of hours, days, or weeks. Most people reach the stage of greatest weakness within the first 2 weeks after symptoms appear, and by the third week of the illness 90 percent of all patients are at their weakest.

What causes Guillain-Barré syndrome?

No one yet knows why Guillain-Barré syndrome—which is not contagious—strikes some people and not others. Nor does anyone know exactly what sets the disease in motion.

What scientists do know is that the body's immune system begins to attack the body itself, causing what is known as an autoimmune disease. Usually the cells of the immune system attack only foreign material and invading organisms. In Guillain-Barré syndrome, however, the immune system starts to destroy the myelin sheath that surrounds the axons of many peripheral

nerves, or even the axons themselves (axons are long, thin extensions of the nerve cells; they carry nerve signals). The myelin sheath surrounding the axon speeds up the transmission of nerve signals and allows the transmission of signals over long distances.

In diseases in which the peripheral nerves' myelin sheaths are injured or degraded, the nerves cannot transmit signals efficiently. That is why the muscles begin to lose their ability to respond to the brain's commands, commands that must be carried through the nerve network. The brain also receives fewer sensory signals from the rest of the body, resulting in an inability to feel textures, heat, pain, and other sensations. Alternately, the brain may receive inappropriate signals that result in tingling, "crawling-skin," or painful sensations. The signals that travel to and from the arms and legs must travel the longest distances and are most vulnerable to interruption. Therefore, muscle weakness and tingling sensations usually first appear in the hands and feet and progress upwards.

When GBS is preceded by a viral or bacterial infection, it is possible that the virus has changed the nature of cells in the nervous system so that the immune system treats them as foreign cells. It is also possible that the virus makes the immune system itself less discriminating about what cells it recognizes as its own, allowing some of the immune cells, such as certain kinds of lymphocytes and macrophages, to attack the myelin.

Sensitized T lymphocytes cooperate with B lymphocytes to produce antibodies against components of the myelin sheath and may contribute to destruction of the myelin. In two forms of GBS, axons are attacked by antibodies against the bacteria *Campylobacter jejuni* which react with proteins of the peripheral nerves. Acute motor axonal neuropathy is particularly common in Chinese children. Scientists are investigating these and other possibilities to find why the immune system goes awry in Guillain-Barré syndrome and other autoimmune diseases. The cause and course of Guillain-Barré syndrome is an active area of neurological investigation, incorporating the cooperative efforts of neurological scientists, immunologists, and virologists.

How is Guillain-Barré syndrome diagnosed?

Guillain-Barré is called a syndrome rather than a disease because it is not clear that a specific disease-causing agent is involved. A syndrome is a medical condition characterized by a collection of symptoms (what the patient feels) and signs (what a doctor can observe or measure). The signs and symptoms of the syndrome can be quite varied, so doctors may, on rare occasions, find it difficult to diagnose GBS in its earliest stages.

Several disorders have symptoms similar to those found in GBS, so doctors examine and question individuals carefully before making a diagnosis. Collectively, the signs and symptoms form a certain pattern that helps doctors differentiate Guillain-Barré from other disorders. For example, physicians will note whether the symptoms appear on both sides of the body (most common in GBS) and the quickness with which the symptoms appear (in other disorders, muscle weakness may progress over months rather than days or weeks). In Guillain-Barré, reflexes such as knee jerks are usually lost. Because the signals traveling along the nerve are slower, a nerve conduction velocity (NCV) test can provide clues to aid the diagnosis. In individuals with GBS, the cerebrospinal fluid that bathes the spinal cord and brain contains more protein than usual. Therefore a physician may decide to perform a spinal tap, a procedure in which a needle is inserted into the person's lower back and a sample of cerebrospinal fluid is withdrawn from the spinal column for study.

How is Guillain-Barré treated?

There is no known cure for Guillain-Barré syndrome. However, there are therapies that lessen the severity of the illness and accelerate the recovery in most patients. There are also a number of ways to treat the complications of the syndrome.

Currently, plasma exchange (also called plasmapheresis) and high-dose immunoglobulin therapy are used. Both of them are equally effective, but immunoglobulin is easier to administer. Plasma exchange is a method by which whole blood is removed from the body and processed so that the red and white blood cells are separated from the plasma, or liquid portion of the blood. The blood cells are then returned to the patient without the plasma, which the body quickly replaces. Scientists still don't know exactly why plasma exchange works, but the technique seems to reduce the severity and duration of the Guillain-Barré episode. This may be because plasmapheresis can remove antibodies and other immune cell-derived factors that could contribute to nerve damage.

In high-dose immunoglobulin therapy, doctors give intravenous injections of the proteins that, in small quantities, the immune system uses naturally to attack invading organisms. Investigators have found that giving high doses of these immunoglobulins, derived from a pool of thousands of normal donors, to Guillain-Barré patients can lessen the immune attack on the nervous system. Investigators don't know why or how this works, although several hypotheses have been proposed.

The use of steroid hormones has also been tried as a way to reduce the severity of Guillain-Barré, but controlled clinical trials have demonstrated that this treatment not only is not effective but may even have a deleterious effect on the syndrome.

The most critical part of the treatment for this syndrome consists of keeping the patient's body functioning during recovery of the nervous system. This can sometimes require placing the individual on mechanical ventilator assistance, a heart monitor, or other machines that assist body function. The need for this sophisticated machinery is one reason why people with GBS are usually treated in hospitals, often in an intensive care ward. In the hospital, doctors can also look for and treat the many problems that can afflict any paralyzed patient—complications such as pneumonia or bed sores.

Often, even before recovery begins, caregivers may be instructed to manually move the patient's limbs to help keep the muscles flexible and strong and to avoid venous sludging (the buildup of red blood cells in veins) which could result in deep vein thrombosis. Later, as the individual begins to recover limb control, physical therapy begins. Carefully planned clinical trials of new and experimental therapies are the key to improving the treatment of patients with Guillain-Barré syndrome. Such clinical trials begin with the research of basic and clinical scientists who, working with clinicians, identify new approaches to treating patients.

What is the long-term outlook for those with Guillain-Barré syndrome?

Guillain-Barré syndrome can be a devastating disorder because of its sudden and unexpected onset. In addition, recovery is not necessarily quick. As noted above, individuals usually reach the point of greatest weakness or paralysis days or weeks after the first symptoms occur. Symptoms then stabilize at this level for a period of days, weeks, or, sometimes, months. The recovery period may be as little as a few weeks or as long as a few years. About 30 percent of those with Guillain-Barré still have a residual weakness after 3 years. About 3 percent may suffer a relapse of muscle weakness and tingling sensations many years after the initial attack.

Individuals with GBS face not only physical difficulties, but emotionally painful periods as well. It is often extremely difficult for people to adjust to sudden paralysis and dependence on others for help with routine daily activities. Some individuals may need psychological counseling to help them adapt.

What research is being done?

Scientists are concentrating on finding new treatments and refining existing ones. Scientists are also looking at the workings of the immune system to find which cells are responsible for beginning and carrying out the attack on the nervous system. The fact that so many cases of Guillain-Barré syndrome begin after a viral or bacterial infection suggests that certain characteristics of some viruses and bacteria may activate the immune system inappropriately. Investigators are searching for those characteristics. Certain proteins or peptides in viruses and bacteria may be the same as those found in myelin, and the generation of antibodies to neutralize the invading viruses or bacteria could trigger the attack on the myelin sheath. Neurological scientists, immunologists, virologists, and pharmacologists are all working collaboratively to learn how to prevent this disorder and to make better therapies available when it strikes.

Where can I get more information?

For more information on neurological disorders or research programs funded by the National Institute of Neurological Disorders and Stroke, contact the Institute's Brain Resources and Information Network (BRAIN) at:

BRAIN

P.O. Box 5801
Bethesda, MD 20824
800- 352-9424
www.ninds.nih.gov

Information also is available from the following organizations

American Autoimmune Related Diseases Association

22100 Gratiot Avenue
East Detroit, MI 48021
586-776-3900
800-598-4668
www.aarda.org

GBS/CIDP Foundation International

The Holly Building
104 ½ Forrest Avenue
Narberth, PA 19072
610-667-0131
866-224-3301
www.gbs-cidp.org

**National Organization for Rare
Disorders (NORD)**

P.O. Box 1968

55 Kenosia Avenue

Danbury, CT 06813-1968

203-744-0100

800-999-6673

www.rarediseases.org





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