Guillain–Barré syndrome

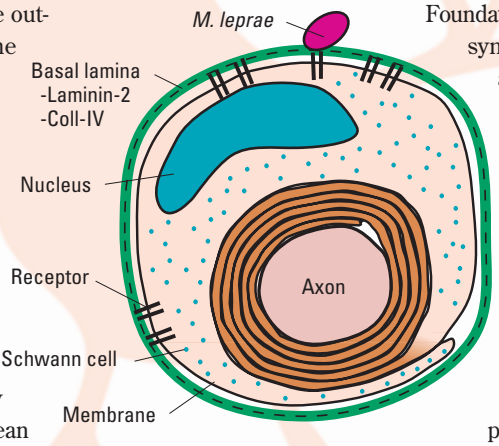
BY WILDER D. SMITH

Imagine that the most efficient fighting machine in the human body, the immune system, suddenly and inexplicably declares war on one of its closest allies, the peripheral nervous system. A biological version of civil war begins, and, if you're lucky, within two weeks you could come off a respirator to breathe on your own. This isn't the prelude to a new sci-fi movie but rather the real-life experience of victims who suffer from a type of acute idiopathic polyneuritis, or Guillain–Barré (ghee-yan bah-ray) syndrome.

Guillain–Barré syndrome is extremely rare. It affects only 2 people out of 100,000 per year. An attack begins when macrophages invade the basal lamina—the outermost layer surrounding the axon of the nerve—and eat through the insulation around the nerve (the myelin) down to the core or conducting part of the nerve (the axon). The symptoms—tingling or numbness in the limbs, slow nerve reflexes, general muscle weakness, and even respiratory or heart failure—can take a few weeks or months to notice.

Georges Guillain

The first case of Guillain–Barré was documented in 1859 by the French physician Jean B. O. Landry. The characteristics of the syndrome remained a mystery until 1916, when Georges Guillain, Jean Barré, and Andre Strohl found abnormal amounts of proteins in the spinal fluid of syndrome patients.



Schwann cell–axon unit

Many questions remain about Guillain–Barré syndrome. Perhaps the two biggest questions are whether it is a disease and, if so, whether it is hereditary. Bernard Ravina, program director for clinical trials at the National Institute of Neurological Disorders and Stroke in Bethesda, MD, explains that it is purposely called a syndrome because, unlike a disease, in which a specific gene can be linked or identified as a cause, Guillain–Barré syndrome is defined by its symptoms collectively.

Currently, there are no hereditary links, targeted age groups, or socioeconomic links among its victims. "Its strongest link has been to the bacterium *Campylobacter jejuni*," says Ravina. *C. jejuni*—a slender, curved, rod-shaped bacterium found in raw chicken, unchlorinated water, and unpasteurized milk—is often present in the diarrheal stools of patients suffering with Guillain–Barré syndrome. What *is* known is that the symptoms usually occur 2–4 weeks after a viral or bacterial infection such as a stomach or intestinal virus, infectious mononucleosis, viral hepatitis, or even the common cold. There are also cases of pregnant women in their third trimester developing the syndrome.

The only epidemic outbreak of the syndrome came in 1976 after swine flu vaccinations. In October of that year, the U.S. government launched the National Influenza Immunization Program to prevent another epidemic outbreak of influenza cases like that experienced in 1918. The vaccinations resulted in more than 500 cases of Guillain–Barré syndrome—25 of them fatal—by January of the following year.

Jean Barré

The syndrome can also strike without warning. "You can wake up one day and you're paralyzed," says Estelle Benson, executive director of the Guillain–Barré Syndrome Foundation International. The symptoms of an immediate attack often lead its victims to believe they are having a stroke. However, the two are not the same, says Ravina. "A stroke tends to affect one side of the body, whereas Guillain–Barré usually affects the entire body," he says.

An attack of Guillain–Barré is treatable and rarely fatal. The most common forms of treatment are plasmapheresis and high-dose immunoglobulin therapy. In plasmapheresis, some of the patient's blood is removed, the liquid component is separated and washed, and the clean blood cells are then returned to the body. In high-dose immunoglobulin therapy, small quantities of proteins that the immune system normally uses to attack invading organisms are injected intravenously into the patient at high doses. These injections reduce the ferocity of the immune system's attack on the nerves.

Reports show that 70% of the victims of Guillain–Barré syndrome return to a normal life; some reports put the number at 90%. Only a small percentage of patients (less than 15%) suffer permanent or long-term disabilities such as paralysis after an attack. Repeated attacks also are rare. "It's usually a one-time thing, and you're done," says Ravina.

With proper treatment, not only can Guillain–Barré syndrome be cured, but a truce can be called to the internal war. With civility restored, patients often return to their former lifestyles, in peace and harmony.

Sources

Guillain-Barré Syndrome Foundation International; www.Guillain-Barre.com.
Guillain-Barré Syndrome: From Healthy to Helpless; http:// medweb.pc.edu/students/med2003/miller1/guillain.html.
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PHOTOS: http://medweb.pc.edu/students/med2003/miller1/guillain.html

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