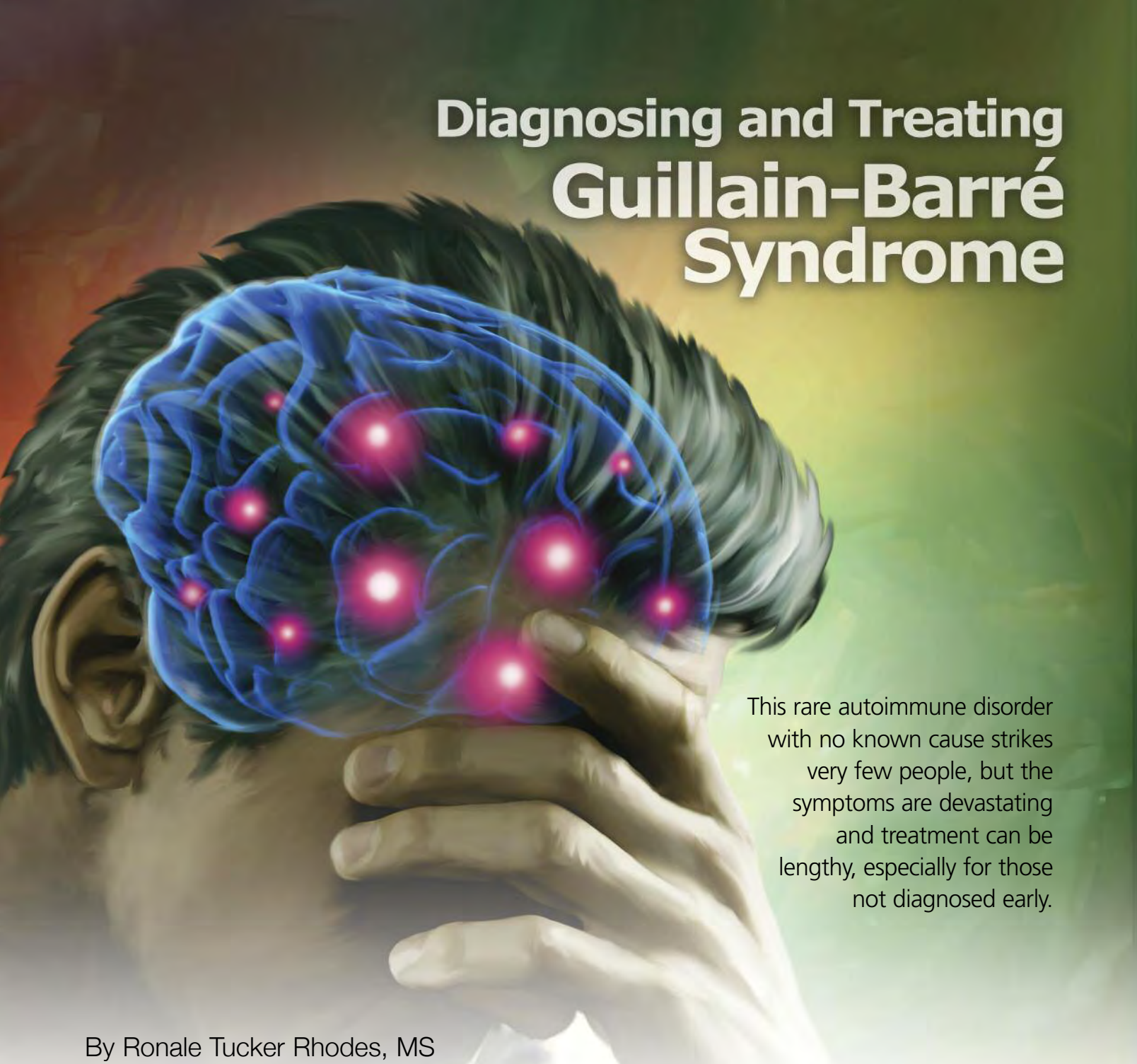
Diagnosing and Treating Guillain-Barré Syndrome



This rare autoimmune disorder with no known cause strikes very few people, but the symptoms are devastating and treatment can be lengthy, especially for those not diagnosed early.

By Ronale Tucker Rhodes, MS

No matter what age one is when Guillain Barré syndrome (GBS) strikes, it is debilitating and tragic. Like Amanda, an active 14-year-old who suddenly became weak and lost feeling in her feet and legs. After four days in the intensive care unit and a diagnosis of GBS, she was still sick months later, in so much pain she was not only unable to do sports, and she could barely make it to school. And, like Byron Comp, a 52-year-old computer scientist who writes in his book, *Guillain Barré Syndrome — My Worst Nightmare*: "For someone like myself who spent the first 52 years of his life as an independent spirit, doing *what* he wanted *when* he wanted most of the time, my worst nightmare would be losing that independence."

Not many people have heard of GBS. So, when struck with the sudden onset of painful symptoms that quickly worsen, individuals and very often their doctors are baffled about what is wrong. Left untreated for long, these individuals can develop permanent nerve damage and even risk death. Fortunately, there is a growing awareness of GBS, and once diagnosed, it can be treated and, in most cases, individuals do fully recover.

What Is GBS?

GBS is a rare autoimmune disorder in which the body's immune system mistakenly attacks part of its peripheral nervous system (PNS), believing it to be foreign material and invading organisms. The part of the PNS it attacks is either the myelin sheath, which surrounds the axons of many peripheral nerves, or even the axons themselves, which are long, thin extensions of the nerve cells that carry nerve signals. Once injured or degraded, the nerves cannot transmit signals efficiently, and the muscles begin to lose their ability to respond to the brain's commands. In addition, the brain receives fewer sensory signals from the rest of the body, resulting in an inability to feel textures, heat, pain and other sensations. Or, the brain may receive inappropriate signals, resulting in tingling, "crawling" skin or painful sensations. The arms and legs are most vulnerable, as the signals from the brain to those extremities must travel the longest distances.[1]

A grave disorder, GBS was first described in 1859 by Jean Landry, a French physician. In 1916, Georges Guillain, Jean Alexandre Barré and Andre Strohl diagnosed two soldiers with the illness.[2] GBS can develop over the course of hours or days, or it can take up to three to four weeks. The first symptoms include varying degrees of weakness or tingling sensations in the legs, and in many instances, the weakness and abnormal sensations spread to the arms and upper body. As these symptoms increase in intensity, certain muscles cannot be used at all, and when severe, the patient can be almost totally paralyzed and unable to breathe on their own.[2,3]

GBS afflicts approximately one person in 100,000. It is called a syndrome rather than a disease because it is not clear that a specific disease-causing agent is involved. In fact, it is unknown what causes GBS and why it strikes some people and not others. But, it can affect anybody at any age, and both sexes are equally prone to the syndrome. What is known is that GBS usually occurs a few days or weeks after the patient has had symptoms of a respiratory or gastrointestinal viral infection. And, occasionally, surgery or vaccinations will trigger the syndrome.[1,3]

The GBS-Vaccine Link

While experts at the Centers for Disease Control and Prevention (CDC) disagree that there is any evidence linking vaccinations with even a single case of GBS, other experts believe otherwise. However, because the associated link occurs in such small numbers, those experts don't support forgoing vaccination.[4]

The first link between GBS and vaccinations occurred in 1976 when many cases of GBS were reported after the swine flu vaccination was given.[2] That year, 43 million people were vaccinated against the swine flu, 500 of whom developed GBS and 25 of whom died.[4] Small increases in the incidence of GBS occurring after vaccinations were also noted in the 1992-1993 and 1993-1994 flu seasons.[5]

In 2009, there were 62 cases that had a high suspicion of being GBS from among 99 million people vaccinated against the H1N1 swine flu virus, all but two of which emerged within six weeks of getting the vaccine, which translated into a rate of about six per 10 million people.

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However, research shows that the rate of GBS in the general population is estimated to be 34 to 400 per 10 million people. According to Dr. Nizar Souayah, a neuromuscular specialist and assistant professor of neurology at New Jersey Medical School, although this research "suggests Guillain-Barré syndrome may be triggered in some cases by H1N1 influenza vaccination, the very low incidence of H1N1 influenza vaccine-associated Guillain-Barré syndrome makes vaccination the first-line strategy for infection prevention and supports the current guidelines for vaccination." Dr. Souayah added, "There is more risk for not vaccinating than for vaccinating."[4]

Case reports of GBS after administration of several other vaccines also have been published. This includes vaccines against anthrax, haemophilus influenza type b, measles, rabies, rubella, tetanus-diphtheria and polio.[6] Most recently, there are reports of GBS after the new HPV vaccines.[7] However, once again, the incidence of cases appears to be no higher than the background rate of GBS incidence expected in an unvaccinated population.[6]

Diagnosing GBS

Because the signs and symptoms of GBS are so varied, doctors often find it difficult to diagnose it in its earliest stages. Several disorders have similar symptoms to GBS, such as chronic inflammatory demyelinating polyneuropathy, French polio, Landry's ascending paralysis and muscular sclerosis.[1,2]

To make a diagnosis, physicians need to establish a collective pattern of the signs and symptoms. For example, GBS

symptoms appear on both sides of the body, versus one or another, and symptoms appear more quickly (days or weeks) than other disorders, which often progress over months.[1]

Specific tests can help physicians to make a diagnosis. Since knee jerks are usually lost in GBS patients, a reflex test can be revealing. In addition, because signals traveling along the nerves are slower in GBS patients, a nerve conduction velocity test or electromyogram (EMG) can aid in diagnosis. And, GBS patients usually retain more protein than usual in their cerebrospinal fluid that bathes the spinal cord and brain, so a spinal tap, a procedure in which the doctor inserts a needle into the patient's lower back to draw cerebrospinal fluid from the spinal column, can be performed.[1,2]

Treating GBS

There is no known cure for GBS, but it can be treated to lessen the severity of the illness and to accelerate recovery. The two equally effective treatments currently used for GBS include plasmapheresis (plasma exchange) and high-dose immunoglobulin (IG). With plasmapheresis, whole blood is removed from the body and processed to separate red and white blood cells from the plasma (the liquid portion of the blood). Those blood cells are then returned to the patient without the plasma, which is quickly replaced by the body. Scientists don't yet understand why plasma-

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pheresis works, but it does reduce the severity and duration of GBS. They speculate that the plasma portion of the blood contains elements of the immune system that may be toxic to the myelin sheath. Patients given IG therapy receive injections of these proteins in high doses, which lessens the immune attack on the nervous system. Again, scientists still don't know how or why this works.[1]

Keeping the patient's body functioning during nervous system recovery is the most critical part of treatment. In some cases, the patient may need a respirator, heart monitor or other machines to assist with breathing and other body functions. Before recovery begins, caregivers can manually move the patient's limbs. Then, as the patient begins to recover, physical therapy is needed. Psychological counseling also may be necessary to help the patient adjust to sudden paralysis and dependence on others for help with routine daily activities.[1]

Depending upon how early GBS is diagnosed, recovery can be in as little as a few weeks or as long as a few years. Eighty percent of people diagnosed with GBS will recover completely with only slight residual weakness. Five to 10 percent may experience more serious permanent problems, and one in 10 may experience a relapse at some later time.[4]

Approximately 5 percent of people afflicted with GBS will die, despite treatment.[4] In a study conducted by the department of neurology at the Mayo Medical Center, Rochester, Minn., 14 of 320 patients (4 percent) admitted with GBS died as a direct result of the illness. Deaths most commonly resulted from ventilator-associated pneumonia. In comparison with 101 other patients with severe GBS admitted to the intensive care unit, the patients who died were older and more likely to have underlying pulmonary disease. In a specialized center, the primary event leading to death from GBS was ventilator-associated pulmonary infection, predominantly in elderly patients with significant comorbidity.[8]

Immunization for GBS Patients

Despite any proven connection between vaccines and GBS, it is known that vaccines may trigger the syndrome. Therefore, GBS patients wonder whether they should avoid vaccinations to prevent a relapse. According to Gareth J. Parry, professor of neurology at the University of Minnesota, the decision to avoid vaccinations "is a personal one that each patient must make, based on their best analysis of the risks and benefits."[9]

While recurrent attacks of GBS are rare, they do occur and they have been described following vaccines. But, recurrence of GBS also may occur following infections, such as influenza. "It is important to remember that the risk of developing new GBS [or] having a recurrence of GBS … are … considerably greater following an infectious illness such as influenza than it is after vaccination," explains Parry. "Thus, by preventing the infection, vaccinations may actually reduce the risk of one of these undesirable outcomes." Parry emphasizes that getting a disease such as polio, typhoid, tetanus, rabies and others is so devastating that it's easy to recommend the vaccine, even if it does cause GBS. But, he says, "It is with the less severe diseases such as influenza that the difficulties arise."[9]

Peter D. Donofrio, MD, who is on the medical advisory board for the GBS/CIDP Foundation International, agrees: "The patient must weigh the chances for relapses of GBS … after immunization to relapses from natural infections with the influenza virus and other pathogens, as well as the morbidity and mortality of influenza infection not affecting the peripheral nerves.… Some data exist that the relative risk of developing GBS is considerably higher after the natural flu than after vaccination."[5]

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Unfortunately, almost all studies that have looked at the incidence of GBS following vaccination in normal populations pertain to populations that have never had GBS. But there is one study in which the researchers distributed a questionnaire to members of the British Guillain-Barré Syndrome Support Group about their illness, immunizations given after their illness, and new symptoms developing within six weeks of these immunizations. Three hundred and eleven patients with prior GBS had received an immunization since recovering from GBS. Eleven patients reported new symptoms of fatigue, weakness, numbness and paresthesia, but in most instances, the symptoms were mild and did not require hospitalization. One patient could not work or drive for six weeks. The relapses were most associated with influenza, tetanus and typhoid immunization, but some relapses were observed after immunizations with polio, hepatitis A and B, BCG, yellow fever, meningococcal and diphtheria vaccines. From that study, the researchers determined that the chance for relapse in GBS requiring treatment is about 1 percent after receiving an immunization and only 0.3 percent of patients will experience significant disability.[5]

Parry suggests that GBS patients who are concerned about receiving an influenza vaccine answer the following questions: 1) Was the initial attack of GBS triggered by an influenza vaccine? 2) Was the initial attack of GBS triggered by influenza? 3) Is the individual at increased risk of significant complications of influenza? This includes individuals with chronic respiratory disease, such as asthma, chronic bronchitis or emphysema, people over the age of 70 years and people with other serious chronic diseases. If the answer to either of the first two questions is yes, while the answer to the third is no, then the risk of the vaccine may outweigh any benefit. If the answer to the third question is yes, while the first two are no, then the benefit of the vaccine clearly outweighs the slight risk that it will cause a recurrence of GBS. If the answer to all three questions is yes, "then I usually still recommend vaccination, but certainly emphasize to the patient the potential risk," says Parry.[9]

The Future of GBS?

Even though the incidence of GBS is rare, neurological scientists, immunologists, virologists and pharmacologists are all collaborating to learn how to prevent this disorder and to make better therapies available. Until then, those afflicted with GBS can only continue with the current treatment protocols and seek the insight of others who also suffer from the syndrome, such as Byron, who wrote his book to give support and hope to other victims. ◪

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