

Guillain-Barre and CIDP

Guillain-Barre syndrome (GBS) is an acute inflammatory disease of the peripheral nerves. An autoimmune attack on the myelin (insulation around individual nerve fibers, called axons) results in demyelination. Loss of myelin can occur in sensory, motor or autonomic nerves. Most patients recover spontaneously, but the recovery can be hastened by plasma exchange or intravenous immunoglobulins. In a small number of patients the inflammation in the nerve can be severe enough to cause degeneration of the whole nerve fibers. In those patients, the recovery is often slower and incomplete. A chronic form of this illness may present with progressive symptoms and result in CIDP (Chronic inflammatory demyelinating polyneuropathy).

Symptoms

GBS patients develop rapidly progressive sensory symptoms such as unusual sensations (paresthesias) and numbness, and motor symptoms such as weakness and cramping in their legs followed by their arms. Patients may also develop weakness of their breathing and difficulty chewing and swallowing. Difficulty breathing may create a neurological emergency as the patients can develop respiratory arrest. A sizeable number of patients also develop autonomic dysfunction where they experience fluctuations in their blood pressure and cardiac arrhythmias. GBS is one of the true neurological emergencies. Patients need to be monitored closely during the initial acute phase of the illness. In contrast, CIDP is a slowly progressive illness with diffuse sensory and motor symptoms.

Diagnosis

Diagnosis of GBS and CIDP is based on history, clinical examination and supporting laboratory investigations. These include electromyography with nerve conduction studies, blood tests and analysis of spinal fluid. In most instances CIDP requires nerve biopsy for histopathological evaluation.

Treatment

GBS patients with respiratory failure require full supportive care in the intensive care unit. Plasma exchange or intravenous immunoglobulins (IVIG) are administered to hasten the recovery from GBS. During the recovery, patients often need aggressive rehabilitation. CIDP as an autoimmune disease often responds to corticosteroids. However, long-term use of corticosteroids is associated with multiple complications and patients are often switched to "corticosteroid-sparing" agents. Acute exacerbations of symptoms can sometimes be treated with intravenous immune globulins (IVIG) or plasma exchange. Neuropathic pain due to CIDP can be treated with anti-seizure medications, antidepressants such as, or analgesics including opiate drugs. In severe painful conditions patients may be referred to the Blaustein Chronic Pain Clinic for a multidisciplinary approach to pain management.