ANNOUNCEMENT

Criteria for Diagnosis of Guillain-Barre Syndrome

Diagnostic criteria for Guillain-Barre syndrome have been established by an ad hoc NINCDS committee. Because Guillain-Barré diagnosis is descriptive, the criteria are expected to help neurologists and nonneurologists recognize the syndrome's diagnostic boundaries.

Drafting of the guidelines was precipitated in part by the increased incidence of Guillain-Barré syndrome in 1977 associated with the swine flu vaccine and by the possibility of an increased incidence this year with the newly developed Russian flu vaccine. When asked for diagnostic criteria by the National Institute of Allergy and Infectious Diseases (planning Russian flu vaccine studies) and by the Center for Disease Control (planning vaccine-related epidemiological studies), Dr Tower requested that a committee chairman be appointed by the American Academy of Neurology and the American Neurological Association.

Consensus was achieved on the criteria that follow by committee members Arthur K. Asbury, MD, University of Pennsylvania (Committee Chairman); Barry G. W. Arnason, MD, University of Chicago; Herbert R. Karp, MD, Emory University; and Dale E. McFarlin, MD, NINCDS.

Definition of Guillain-Barré Syndrome and Criteria for Diagnosis

Guillain-Barré syndrome is a recognizable entity for which the basis for diagnosis is descriptive in our present state of knowledge. The features which allow a diagnosis include clinical, laboratory, and electrodiagnostic criteria. The problem is not with recognition of a typical case, but with knowing the boundaries by which the core disorder is delimited. The following criteria are established, in light of current knowledge and opinion, to define those limits.

The presence of preceding events is frequent, but they are not essential to the diagnosis. Most commonly, preceding events are viral infections, but the association of Guillain-Barré syndrome with preceding surgery, inoculations, and mycoplasma infections is also known. In addition, Guillain-Barré syndrome occurs more frequently than by chance in the setting of preexisting illnesses such as Hodgkin's disease, lymphoma, or lupus erythematosus. Many patients with Guillain-Barré syndrome will have no history of any of these events, and the diagnosis should be made independent of them.

I. Features Required for Diagnosis

A. Progressive motor weakness of more than one limb. The degree ranges from minimal weakness of the legs, with or without mild ataxia, to total paralysis of the muscles of all four extremities and the trunk, bulbar and facial paralysis, and external ophthalmoplegia.

B. Areflexia (loss of tendon jerks). Universal areflexia is the rule, though distal areflexia with definite hyporeflexia of the biceps and knee jerks will suffice if other features are consistent.

II. Features Strongly Supportive of the Diagnosis

A. Clinical features (ranked in order of importance)

1. Progression. Symptoms and signs of motor weakness develop rapidly but cease to progress by four weeks into the illness. Approximately 50% will reach the nadir by two weeks, 80% by three weeks, and more than 90% by four weeks.
2. Relative symmetry. Symmetry is seldom absolute, but usually, if one limb is affected, the opposite is as well.
3. Mild sensory symptoms or signs.
4. Cranial nerve involvement. Facial weakness occurs in approximately 50% and is frequently bilateral. Other cranial nerves may be involved, particularly those innervating the tongue and muscles of deglutition, and sometimes the extraocular motor nerves. On occasion (less than 5%), the neuropathy may begin in the nerves to the extraocular muscles or other cranial nerves.
5. Recovery. It usually begins two to four weeks after progression stops. Recovery may be delayed for months. Most patients recover functionally.
6. Autonomic dysfunction. Tachycardia and other arrhythmias, postural hypotension, hypertension, and vasomotor symptoms, when present, support the diagnosis. These findings may fluctuate. Care must be exercised to exclude other bases for these symptoms, such as pulmonary embolism.
7. Absence of fever at the onset of neuritic symptoms.

Variants (not ranked)

1. Fever at the onset of neuritic symptoms.
2. Severe sensory loss with pain.
3. Progression beyond four weeks. Occasionally, a patient's disease will continue to progress for many weeks longer than four or the patient will have a minor relapse.
4. Cessation of progression without recovery or with major permanent residual deficit remaining.
5. Sphincter function. Usually the sphincters are not affected, but transient bladder paralysis may occur during the evolution of symptoms.
6. Central nervous system involvement. Ordinarily, Guillain-Barré syndrome is thought of as a disease of the peripheral nervous system.

Evidence of central nervous system involvement is controversial. In occasional patients, such findings as severe ataxia interpretable as cerebellar in origin, dysarthria, extensor plantar responses, and ill-defined sensory levels are demonstrable, and these need not exclude the diagnosis if other features are typical.

B. Cerebrospinal fluid features strongly supportive of the diagnosis
   1. CSF protein. After the first week of symptoms, CSF protein is elevated or has been shown to rise on serial lumbar punctures.
   2. CSF cells. Counts of 10 or fewer mononuclear leukocytes/mm^3 in CSF.

   Variants
   1. No CSF protein rise in the period of one to ten weeks after the onset of symptoms (rare).
   2. Counts of 11 to 50 mononuclear leukocytes/mm^3 of CSF.

C. Electrodiagnostic features strongly supportive of the diagnosis

   Approximately 80% will have evidence of nerve conduction slowing or block at some point during the illness. Conduction velocity is usually less than 60% of normal, but the process is patchy and not all nerves are affected. Distal latencies may be increased to as much as three times normal. Use of F-wave responses often gives good indication of slowing over proximal portions of nerve trunks and roots. Up to 20% of patients will have normal conduction studies. Conduction studies may not become abnormal until several weeks into the illness.

III. Features Casting Doubt on the Diagnosis
1. Marked, persistent asymmetry of weakness.
2. Persistent bladder or bowel dysfunction.
3. Bladder or bowel dysfunction at onset.
4. More than 50 mononuclear leukocytes/mm^3 in CSF.
5. Presence of polymorphonuclear leukocytes in CSF.
6. Sharp sensory level.

IV. Features That Rule Out the Diagnosis
1. A current history of hexacarbon abuse (volatile solvents; *n*-hexane and methyl *n*-butyl ketone). This includes huffing of paint lacquer vapors or addictive glue sniffing.
2. Abnormal porphyrin metabolism indicating a diagnosis of acute intermittent porphyria. This would manifest as increased excretion of porphobilinogen and δ-aminolevulinic acid in the urine.
3. A history or finding of recent diphtheritic infection, either faucial or wound, with or without myocarditis.
4. Features clinically consistent with lead neuropathy (upper limb weakness with prominent wrist drop; may be asymmetrical) and evidence of lead intoxication.
5. The occurrence of a purely sensory syndrome.
6. A definite diagnosis of a condition such as poliomyelitis, botulism, paralysis, or toxic neuropathy (e.g., from nitrofurantoin, dapsone, or organophosphorus compounds), which occasionally may be confused with Guillain-Barre syndrome.