TABLE 460-3 BRIGHTON CRITERIA FOR DIAGNOSIS OF GUILLAIN-BARRÉ SYNDROME (GBS) AND MILLER FISHER SYNDROME

Clinical case definitions for diagnosis of GBS

Level 1 of diagnostic certainty

Bilateral AND flaccid weakness of the limbs

AND

Decreased or absent deep tendon reflexes in weak limbs

AND

Monophasic illness pattern and interval between onset and nadir of weakness between 12 h and 28 days and subsequent clinical plateau

AND

Electrophysiologic findings consistent with GBS

AND

Cytoalbuminologic dissociation (i.e., elevation of CSF protein level above laboratory normal value AND CSF total white cell count <50 cells/μL)

AND

Absence of an identified alternative diagnosis for weakness

Level 2 of diagnostic certainty

Bilateral AND flaccid weakness of the limbs

AND

Decreased or absent deep tendon reflexes in weak limbs

AND

Monophasic illness pattern and interval between onset and nadir of weakness between 12 h and 28 days and subsequent clinical plateau

AND

CSF total white cell count <50 cells/μL (with or without CSF protein elevation above laboratory normal value)

OR

If CSF not collected or results not available, electrophysiologic studies consistent with GBS

AND

Absence of identified alternative diagnosis for weakness

Level 3 of diagnostic certainty

Bilateral and flaccid weakness of the limbs

AND

Decreased or absent deep tendon reflexes in weak limbs

AND

Monophasic illness pattern and interval between onset and nadir of weakness between 12 h and 28 days and subsequent clinical plateau

AND

Absence of identified alternative diagnosis for weakness

Clinical case definitions for diagnosis of Miller Fisher syndrome

Level 1 of diagnostic certainty

Bilateral ophthalmoparesis and bilateral reduced or absent tendon reflexes, and ataxia

AND

Absence of limb weakness

AND

Monophasic illness pattern and interval between onset and nadir of weakness between 12 h and 28 days and subsequent clinical plateau

AND

Cytoalbuminologic dissociation (i.e., elevation of cerebrospinal protein above the laboratory normal and total CSF white cell count <50 cells/μL)

AND

Nerve conduction studies are normal, OR indicate involvement of sensory nerves only

AND

No alterations in consciousness or corticospinal tract signs

AND

Absence of identified alternative diagnosis

Level 2 of diagnostic certainty

Bilateral ophthalmoparesis and bilateral reduced or absent tendon reflexes and ataxia

AND

Absence of limb weakness

AND

Monophasic illness pattern and interval between onset and nadir of weakness between 12 h and 28 days and subsequent clinical plateau

AND

CSF with a total white cell count <50 cells/μL) (with or without CSF protein elevation above laboratory normal value)

OR

Nerve conduction studies are normal, OR indicate involvement of sensory nerves only

AND

No alterations in consciousness or corticospinal tract signs

AND

Absence of identified alternative diagnosis

Level 3 of diagnostic certainty

Bilateral ophthalmoparesis and bilateral reduced or absent tendon reflexes and ataxia

AND

Absence of limb weakness

AND

Monophasic illness pattern and interval between onset and nadir of weakness between 12 h and 28 days and subsequent clinical plateau

AND

No alterations in consciousness or corticospinal tract signs

AND

Absence of identified alternative diagnosis

Abbreviation: CSF, cerebrospinal fluid.

Source: From JJ Sejvar et al: Guillain-Barré syndrome and Fisher syndrome: case definitions and guidelines for collection, analysis, and presentation of immunization safety data. Vaccine 29:599, 2011. Validation study published by C Fokke et al: Diagnosis of Guillain-Barré syndrome and validation of Brighton criteria. Brain 137:33, 2014.

of intubation) of tracheotomy, and chest physiotherapy. As noted, ~30% of patients with GBS require ventilatory assistance, sometimes for prolonged periods of time (several weeks or longer). Frequent turning and assiduous skin care are important, as are daily range-of-motion exercises to avoid joint contractures and daily reassurance as to the generally good outlook for recovery.

Prognosis and Recovery Approximately 85% of patients with GBS achieve a full functional recovery within several months to a year, although minor findings on examination (such as areflexia) may persist and patients often complain of continued symptoms, including fatigue. The mortality rate is <5% in optimal settings; death usually results from secondary pulmonary complications. The outlook is worst in patients with severe proximal motor and sensory axonal damage. Such axonal damage may be either primary or secondary in nature (see "Pathophysiology," above), but in either case successful regeneration cannot occur. Other factors that worsen the outlook for recovery are advanced age, a fulminant or severe attack, and a delay in the onset of treatment. Between 5 and 10% of patients with typical GBS have one or more late relapses; such cases are then classified as chronic inflammatory demyelinating polyneuropathy (CIDP).

CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY

CIDP is distinguished from GBS by its chronic course. In other respects, this neuropathy shares many features with the common demyelinating form of GBS, including elevated CSF protein levels