Pathological entities most likely have their own pathogenesis. In about two thirds of all patients, Guillain-Barré syndrome is preceded by infections. We pointed out that motor Guillain-Barré syndrome without sensory loss is characterized by rapid onset of weakness, early nadir, distal-dominant weakness, sparing of the cranial nerves, and a preceding gastrointestinal infection, often subsequent to *Campylobacter jejuni* infection (Visser et al., 1995). The electrodiagnostic findings of the axonal types of Guillain-Barré syndrome show little evidence of demyelination; low distal CMAP amplitudes usually characterize signs of axonal damage. The presence of spontaneous activity in the renfainder of the muscle fibers on redle electromyography also confirms this. The clinical picture may show many similarities with CIP. Many patients with acute motor neuropathy have high IgG anti-GM1 titers or IgG antibodies to anti-Gal IgG antibodies are important in cytomegalovirus-associated Guillain-Barré syndrome.

**Guillain-Barré Syndrome**

Guillain-Barré syndrome is a subacute, immune-mediated polyneuropathy. Clinical features that are required for diagnosis are (1) progressive motor weakness of more than one limb (degree ranges from minimal ataxia, to total paralysis of the legs, to total paralysis of the muscles of all four extremities and the trunk, to bulbar *and facial paralysis*, and to external ophthalmoplegia) and (2) areflexia (universal areflexia is the rule, although distal areflexia with definite hyporeflexia of the biceps and knee jerks vii suffice if other features are consistent).

The diagnostic criteria are based on clinical, laboratory, and electrodiagnostic criteria and are defined by Asbury and Cornblath (1990).

The clinical spectrum of Guillain-Barré syndrome consists of acute inflammatory demyelinating polyradiculoneuropathy, acute motor axonal neuropathy, acute motor sensory axonal neuropathy, and Miller-Fisher syndrome. These heterogeneous groups of pathological entities most likely have their own
nificantly lowered after about 4 to 6 weeks. The sensory nerve action potential amplitude drops to 20% or less of normal values and commonly disappears between 3 to 4 weeks after onset of the disease. The conduction velocity and latency do not drop below 80% to 90% of the normal mean value. Conduction block or axonal loss is suspected when the alteration in sensory nerve action potential amplitude is larger than in conduction velocity.

Somatosensory Evoked Potential Conduction Studies

To study the proximal aspects of the sensory conduction system in Guillain-Barré syndrome, somatosensory evoked potential techniques are useful. Segmental conduction times, using Erb's point (N9) and cervical (N1) potentials (upper extremity), lumbar (N20) potential, and the central conduction, can be calculated. Central conduction times are essentially normal.

In Guillain-Barré syndrome, there seems to be a predisposition toward the proximal or nerve root regions. These features explain why patients have complaints of sensation and clinical abnormalities and only a few distal electrophysiological sensory nerve action potential abnormalities. Somatosensory evoked potential studies of both the upper and lower limbs should be performed, but because the neural pathway of the lower limbs is considerably longer, it is more beneficial to study them first.

Motor Conduction Studies

To establish the diagnosis and monitoring of Guillain-Barré syndrome, distal motor latencies, conduction velocities, F waves, H reflexes, and CMAP amplitudes; duration, and morphology are used. In 80% to 90% of the patients, at least one of these motor nerve parameters is disturbed. The distal motor latency and CMAP conduction velocity measurements reach a peak reduction of 60% to 80% of the normal mean values about 3 weeks after the onset of the clinical symptoms. After 4 weeks, the values begin to increase to normal over several weeks to months, although 1 year or longer can be required. In general, there is little correlation between the clinical presentation and nerve conduction velocity or distal motor latency.

To perform an examination of the F waves in Guillain-Barré syndrome, high amplifier gains (100 to 200 μV/cm), prolonged pulse duration, and increased current intensities should be used to conclude that F waves are reduced in number. Some type of abnormality can be expected in 80% to 90% of the patients, and the absence of F waves should be considered a definite abnormality. H reflexes should also be tested in the lower limbs to assess possible disturbed proximal neural conduction.

The most frequently encountered abnormality early in Guillain-Barré syndrome is conduction block. Such a block is present if there is a reduction in the peak-to-peak CMAP amplitude of more than 20% (a drop in proximal compared with distal CMAP), as defined by Asbury and Cornblath (1990), in the following nerves: (1) median (proximal arm compared with wrist, recording from thenar muscles), (2) ulnar (Erb’s point compared with wrist, recording from hypothenar muscles), and (3) peroneal (popliteal fossa to ankle, recording from extensor digitorum brevis). Due to the lack of unanimity on the percentage of amplitude reduction; a range of 20% to 40% is used. In pseudo-conduction block, a reduction in amplitude is a result of excessive temporal dispersion, which may increase the duration of the potential, with a concomitant and con- peratortic reduction in amplitude. To distinguish between conduction block and temporal dispersive effects, small-segment stimulation can be used to localize focal reduction in amplitude. The conduction block is pathophysiologically caused by the loss of myelin, leading to conduction failure and symptoms of weakness and sensory loss. Permanent reduction of function is secondary to axonal loss.

Abnormalities with regard to the phrenic nerve are frequently noted, although reduced ventilatory capacity is not due to reduced conduction velocity in the phrenic nerve. If present, axonal damage of this nerve can be diagnosed with needle electromyography of the diaphragm.

In addition, abnormalities of the facial nerve can be tested in Guillain-Barré syndrome, as well as those of the supraorbital nerve. Direct facial nerve stimulation and the blink reflex reveal abnormalities in either or both pathways.

Needle Electromyography Examination

Positive sharp waves and fibrillation potentials at rest appear between 2 to 4 weeks, peaking about 6 to 15 weeks (earlier in the proximal than in the distal muscles). Within the first 3 weeks, myokymia (complex bursts of repetitive discharges that cause vermicular movements of the skin) are detected, especially in the facial muscles.

This examination is mainly adjunctive to explore other disease entities.

In Guillain-Barré syndrome, a reduced recruitment for motor unit potentials is one of the earliest findings. After about 6 to 16 weeks, voluntary motor unit potential amplitude, duration, and number of phases increase. These findings imply (1) axonal loss with motor unit remodeling and (2) reverse motor unit remodeling during axonal regrowth.

Single-fiber electromyography shows mild-to-moderate increase in fiber density later in the course of Guillain-Barré syndrome, substantiating the initial phases of motor unit remodeling in patients with axonal loss.

ELECTRODIAGNOSTIC CRITERIA of GUILLAIN-BARRE SYNDROME AND THEIR VALIDATION

Most electrodiagnostic criteria have been defined on the basis of demyelination. Alam (1998) studied
e six different sets of criteria that have been used previous studies and applied them to 43 patients with the clinical diagnosis of Guillain-Barré syndrome. This resulted in 21% to 72% of the patients ving the diagnosis of acute inflammatory demye- lating polyradiculoneuropathy. The sets were de- led by Albers and colleagues (1985), Albers and Ally (1989), Cornblath (1990), Ho and associates (1997), and Meulstee and van der Meché (1995). though the criteria of Albers (1985; Albers and Kelly, 1989) identified most cases as ute inflammatory demyelinating polyradiculo- neuropathy, the importance of performing analyses on the variability in diagnosing the acute inflammatory emyelinating polyradiculoneuropathy variant of villain-Barré syndrome was emphasized.

In acute inflammatory demyelinating polyradiculo- neuropathy, most distal sites, roots, and physiologi- cal entrapment sites are fragile, and early demyelin- ation and secondary axonal degeneration occur there. Axonal degeneration easily masks demyelinat- ing conduction changes. However, with careful fol- low-up, the presence of delayed F waves or in- creased distal motor latencies definitely mitigates against primary axonal pathology, as in acute motor ional neuropathy or acute motor sensory axonal europathy.

Electrodiagnostic primary axonal Guillain-Barré syndrome was defined by Hadden and associates (1998) and Ho and colleagues (1997) as (1) no evidence of demyelination and (2) CMAP amplitude Ass than 80% of the lower limit of normal.

Treatment and Prognosis

The treatment of Guillain-Barré syndrome with in- ravenous immunoglobulin or plasma exchange results in earlier recovery, but morbidity rates remain nsiderable. Trials may lead to other methods of improving outcome.

The time of recovery depends on the extent of demyelination and axonal degeneration. Patients ith severe axonal loss may not regain motor func- tion for 1 to 2 years, implying a poor prognosis. Axonal regeneration takes considerably longer than emyelination. The Dutch multicenter study on prog- nostic factors that influence Guillain-Barré syndrome revealed that a preceding gastrointestinal illness, ider age (>50 years), severe weakness (a Medical research Council sum score of <40 at the start of ie treatment), and rapid progression of weakness within 4 days of onset of weakness were important independent, significant prognostic factors at 6 months of follow-up (Visser, 1999). Others found hat the electrodiagnostic finding of a low CMAP amplitude (<4 mV) is an important prognostic fac- JI.

To distinguish CIP from the acute motor axonal variant of Guillain-Barré syndrome, the following aiaeracteristics may be useful (De Letter et al., IUL).j}

1. Guillain-Barré syndrome is the primary neurologi- cal reason for admission to the intensive care unit.
2. On the other hand, CIP develops during a patient’s stay in the intensive care unit for another reason.
3. Infectious symptoms like fever and diarrhea usu- ally subside before the clinical features of Guillain- Barré syndrome appear.
4. The characteristic alterations in the cerebrospinal fluid of Guillain-Barré syndrome patients include a raised protein level and a normal to slightly elevat- ed cell count.
5. There is a possibility of detecting IgG antibodies against GM 1, GM 1 b, GD 1 a, arid GalNac-GD1a in the serum of axonal Guillain-Barré syndrome patients.
6. Electrodiagnostic changes in Guillain-Barré syn- drome occur in both sensory and motor nerves in about 80% of the patients in the Western world. In CIP, there is clinically a predominantly motor dysfunction. Both CIP and axonal-type Guillain- Barré syndrome show sensorimotor or pure motor axonal features. Critical illness polyneuropathy and myopathy can sometimes be distinguished from Guillain-Barré syndrome by the presence of myopathic motor unit potentials on voluntary activa-
7. During the progression of Guillain-Barré syndrome, the demyelinating features of the nerve conduc- tion study may change into a secondary axonal pattern. In the latter, slow nerve conduction velocity remains in some patients and the initial needle electromyography study lacks spontaneous activity (Chen, 1998). In CIP, spontaneous activity of the muscle fibers is an early feature. Further phrenic nerve conduction studies usually show no significantly prolonged latencies in CIP (Bolton et al., 1986).
8. Severe autonomic disturbances are more common in the patient with Guillain-Barré syndrome after the polyneuropathy has developed than in pa- tients with CIP (Bolton et al., 1986).
9. Septic encephalopathy may be present before the onset of CIP. Patients with Guillain-Barré syndrome do not have a disturbed consciousness.

Porphyrie Neuropathy

Porphyrie neuropathy is an acute or subacute, predominantly motor neuropathy. Disturbances of porphyrie metabolism are associated with acute attacks of neurological disease in cases of hepatic porphyrías. These porphyrías, consisting of acute intermittent porphyrie, hereditary coproporphyría, and variegate porphyrie, are caused by enzyme de- fects (uroporphyrinogen-1-synthetase, coproporphyrinogen oxidase, or protoporphyrinogen oxidase, re- spectively). Acute intermittent porphyrie (Stein and Tschudy, 1970: Kappas et al., 1993), hereditary coproporphyría (Magnussen et al., 1975), and varie- gate porphyrie (Eales et al., 1980) all occur on a genetic basis. The attacks of acute hepatic porphy- ries may be precipitated by drugs (must often bar bi-