Factors Affecting Outcome and the Role of Plasmapheresis in Guillain-Barré Syndrome

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Guillain Barre syndrome (GBS) is a heterogeneous group of inflammatory polyradiculoneuropathies clinically exhibiting a monophasic course of acute flaccid quadriplegia and/or cranial neuropathies. Based on clinical and electrophysiological features, various forms of GBS have been designated. GBS can progress up to a period of 4 weeks and 25-30% of patients require artificial ventilation. Natural recovery occurs slowly over a period of months and incomplete recovery can leave behind residual deficits. Hence, treatment of GBS with immunomodulation such as plasma exchange (PE) or intravenous immunoglobulin (IVIg) becomes crucial in reducing mortality and morbidity, particularly in patients with rapidly progressive weakness.

PE and IVIg have been extensively studied in GBS. Class I evidence exists for the effectiveness of PE in treatment of GBS when the condition is severe enough to impair independent walking or require mechanical ventilation. Even in patients with mild weakness who are able to walk independently, rapid improvement has been noticed after initial 2 cycles of PE. Studies have shown further benefit of 4 and 6 cycles of PE over the initial 2 sessions but there is no added advantage of 6 PE sessions over 4. Five PE sessions (each exchange sessions of 2-3 litres of plasma volume or 1-1.5 times of colloid volume according to weight) over 2 weeks is generally considered to be a beneficial protocol. Currently, there are no studies to demonstrate the superiority of IVIg over PE or vice versa. Benefits of PE have been documented when it is used within 4 weeks from the onset of weakness. Some studies support that the benefit of PE is larger when used within 7 days after onset of weakness when compared to patients in whom PE was initiated between 8 and 28 days after onset. Thus, studies support the hypothesis of ‘time is nerve’ similar to ‘time is brain’ in acute ischaemic stroke. Efficacy of PE in patients with GBS who continue to progress after 4 weeks is unclear. Such patients with electrophysiological evidence of demyelination and progression more than 4 weeks are often termed as acute-onset chronic inflammatory demyelinating polyneuropathy (A-CIDP) or subacute idiopathic demyelinating polyneuropathy (SIDP). In absence of evidence supporting the efficacy of PE, use of PE should be individualised in such cases. Longer the duration of progression of illness beyond 4 weeks, more is the likelihood of CIDP. Such patients benefit from corticosteroids or alternative immunomodulation.

Electrophysiological tests help to confirm the diagnosis of GBS, detect its subtype (i.e. axonal or demyelinating) and hence, hint at the prognosis. Few important points need to be considered while interpreting nerve conduction study (NCS) findings in patients with GBS.

- NCS can be normal in early cases and it takes up to 2 weeks or longer, before characteristic changes become fully established. Hence, it is important for clinicians to understand that absence of abnormalities on NCS in early cases, particularly within 1 week does not rule out diagnosis of GBS. NCS can be repeated after few days to confirm the sub-type of GBS.
- Abnormality of the sural response is one of the earliest changes encountered in peripheral neuropathies. Sparing of sural responses with profound affection of upper limb sensory potentials is characteristic of non-length dependent neuropathy like GBS.
- Presence of low compound muscle action potentials (CMAP) < 20% amplitude of normal, suggests poor recovery pattern.
- Demyelinating variety of GBS is the most frequently reported sub-type from all around the world. But in clinical practice, axonal GBS is also encountered frequently in India and other Asian countries.

IVIg and PE have been found to be equally effective in all subtypes of GBS such as acute axonal motor neuropathy (AMAN), acute axonal sensory motor neuropathy...
Factors affecting outcome in patients with GBS have been well studied. Factors known to indicate poor recovery i.e. inability to walk independently at 6 months are mentioned below.

- A preceding gastrointestinal illness
- Age more than 50 years
- Medical research council sum score of less than 40
- Preceding cytomegalovirus (CMV) infection
- Initial rapid progression of illness within 4 days of onset
- CMAP amplitude less than 20% of normal
- Respiratory insufficiency

The timing of PE is always crucial as patients tend to fare well when PE is initiated within 7 days of onset of illness. However, impact of earlier use of PE within the 7 days window period on the recovery pattern has not been studied. The study by jignesh et al in this issue of JAPI aims to analyse and evaluate the impact of clinical in the first few days of the clinical symptoms. Physicians need to appreciate that GBS largely remains a clinical diagnosis in initial stages and other diseases presenting with acute flaccid quadriaparesis should be ruled out. From authors’ point of view, following features can help to rule out GBS mimics (Table 1).

Various variables can affect the outcome in patients with GBS. Such variables can be studied systematically by stratifying patients into different subgroups based on timing of PE after disease onset. While the present study by Jignesh et al makes a good beginning, a large, multicentric effort will be necessary in further assessing various clinical, electrophysiological and therapeutic factors that can affect the prognosis in patients suffering from GBS.

References