

PLASMAPHERESIS IS THE TREATMENT OF CHOICE IN MYASTHENIC CRISIS

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Myasthenic crisis is a life threatening medical emergency requiring early diagnosis and respiratory assistance. It can affect between one fifth and one third of all patients with generalised autoimmune myasthenia gravis caused by circulating autoantibodies to acetylcholine receptors (AChR) or muscle-specific tyrosine kinase (MuSK) (1). High dose corticosteroids in combination with therapeutic plasma exchange (TPE) or human intravenous immunoglobulin (IVIg) are the cornerstone of treatment for this fully reversible cause of neuromuscular paralysis. Establishing relative superiority of one intervention over the other has been difficult because of the design of some of the studies. Response to TPE is considered to be more predictable and TPE may be a superior choice because of faster onset of treatment effect. In one retrospective multi-centre review of 54 episodes of myasthenic crisis (2), plasma exchange had a superior outcome for ventilatory status at 2 weeks ($p=0.02$) and functional outcome at 1 month ($p=0.04$). A double blind, placebo-controlled randomised trial of IVIg confirmed efficacy of IVIg in patients with worsening symptoms of myasthenia gravis with greater response in patients with more severe disease (3). The recommended therapeutic dose of IVIg is 2g/kg but a randomised double-blind clinical trial found no significant superiority of 2g/kg dose over 1g/kg dose in patients with exacerbation of myasthenia gravis compared to limited TPE over 3 days (4). More recently, a head-to-head trial of IVIg (1g/kg for 2 days) and TPE (1.0 plasma volume exchanges for 5 days) showed no significant difference in the outcome of patients between two groups (5); however, the majority of patients in this trial had worsening neuromuscular weakness of moderate severity (MGFA Grade 2) only.

Whilst the efficacy of TPE and IVIg seems comparable in worsening myasthenic weakness of moderate severity, there is no trial evidence that IVIg response is same as TPE in myasthenic patients with impending or established respiratory weakness requiring ventilatory assistance (myasthenic crisis). There is some uncertainty about the most effective IVIg dose (2g/kg or 1g/kg), duration of therapy and the time of onset of therapeutic response. In contrast, TPE offers the option of more predictable treatment response in myasthenic patients in crisis. The clearance of serum MuSK-antibody during serial sessions of TPE is similar to AChR-antibody, and TPE also aids the proof of concept of diagnosis in antibody-negative patients. Patients usually respond with rapid improvement after TPE that may be critical in ventilated and post-operative patients. The predictability of treatment effect makes TPE a superior choice over IVIg in myasthenic crisis, provided the facility is available.

Reference

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