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Case Report
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Treatment of Guillain-Barré Syndrome: Exploration of the Role of Plasma Exchange after Failure of Intravenous Immunoglobulin

Rajendram R[1,2]*

[1]Consultant, Department of Medicine, King Abdulaziz Medical City, King Abdulaziz International Medical Research Center, Ministry of National Guard - Health Affairs, Riyadh, Saudi Arabia

[2]Joint Appointment Assistant Professor, College of Medicine, King Saud bin Abdulaziz University of Health Sciences, Riyadh, Saudi Arabia

Abstract

Guillain-Barré syndrome is a life-threatening condition characterized by acute ascending areflexic paralysis. Treatment with plasma exchange reduces demyelination and results in faster recovery, than supportive therapy alone. However, plasma exchange is not widely available. Intravenous Immune Globulin (IVIG) is reported to be as effective as plasma exchange and has essentially replaced plasma exchange as the treatment of choice. It is more convenient and more widely available. However, managing patients who fail to respond to initial treatment is challenging. Administration of IVIG after plasma exchange is no better than either plasma exchange or IVIG alone. Conversely, anecdotal data suggests that patients have been improved by plasma exchange after IVIG failure. To increase the awareness of this phenomenon I outline an illustrative case in which a patient with Guillain–Barré syndrome was successfully treated with plasma exchange after failure of intravenous immunoglobulin.

Keywords: Guillain-Barré syndrome; Plasma exchange; Intravenous immunoglobulin

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*Correspondence:
Rajkumar Rajendram, Consultant, Department of Medicine, King Abdulaziz Medical City, King Abdulaziz International Medical Research Center, Ministry of National Guard - Health Affairs, Riyadh, Saudi Arabia.
E-mail: rajkumarrajendram@doctors.org.uk

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Introduction

Guillain-Barré syndrome is characterized by acute ascending areflexic paralysis. It is associated with high levels of Cerebrospinal Fluid (CSF) protein but normal CSF cell counts. Approximately 5% of patients with Guillain-Barré syndrome die. Plasma exchange and IVIG are non-specific immuno-modulatory therapies for GBS [1-3]. More specific treatments are required. However, although *in vitro* studies are underway there are currently no effective alternatives. Whilst IVIG has essentially replaced plasma exchange as the treatment of choice, the management of patients who fail to respond to IVIG is complex. The illustrative case described below highlights these issues.

Illustrative Case Report

A 43-year-old man was admitted to ICU 3 days after admission to his local district general hospital with a one week history of ascending weakness and numbness, dysarthria, dysphagia and facial weakness. Six weeks prior to presentation he had had a diarrhoeal illness which had lasted for a week. On lumbar puncture it was found that his CSF protein was raised (0.9 g/L) and the white cell count was only 1 cell/ml CSF. Findings on nerve conduction studies were also consistent with Guillain-Barre Syndrome (GBS) and so a 5-day course of intravenous immunoglobulin was given.

However, the patient continued to deteriorate. His cough was very weak and he developed dysphagia. Ten days after admission he developed a productive cough and fever. His chest x-ray showed right lower lobe collapse and consolidation. He was sedated, intubated and ventilated semi-electively.

Pain and paraesthesia in his feet were treated with pregabalin and a tracheostomy was formed on day 12 post-admission. Over the next week all attempts at weaning ventilatory support were unsuccessful. Twenty days after admission he was transferred to the Royal London Hospital for plasma exchange. He received 5 treatments over a period of 2 weeks. He was then transferred back to the ICU at his local district general hospital. Over the subsequent two weeks his muscle

power improved and he weaned from BIPAP. His tracheostomy was decannulated and he was moved to a medical ward 50 days after his initial presentation to hospital.

Discussion

Guillain-Barré syndrome is characterized by acute ascending areflexic paralysis with raised levels of protein in the Cerebrospinal Fluid (CSF) but normal CSF cell counts [1]. Approximately 5% of patients with Guillain-Barré syndrome die of complications such as sepsis, pulmonary emboli, or unexplained cardiac arrest, perhaps related to autonomic nervous system dysfunction [1].

Thus, close observation is required for the early detection of these complications [1]. Patients should remain in hospital for observation at least until there is evidence that disease progression has stopped [1].

Treatment with plasma exchange reduces demyelination and results in faster recovery, in comparison to supportive therapy alone [1]. The mechanism of action is unclear but plasma exchange non-specifically removes antibodies and complement so it is thought that plasma exchange removes the auto-antibodies which cause GBS. Plasma exchange seems to be most effective within 2 weeks of the onset of the disease [1]. Nerve conduction studies are not required prior to treatment. The usual regimen is five total plasma volumes exchanges over a period of 2 weeks [1]. However, plasma exchange is not widely available because it is expensive, labour-intensive and requires wide-bore venous access [1].

Treatment with Intravenous Immune Globulin (IVIG), within 2 weeks of the onset of the disease, is reported to be as effective as plasma exchange in patients with GBS who cannot walk independently [2,3].

IVIG has essentially replaced plasma exchange as the treatment of choice. It is more convenient and more widely available. The standard treatment is a total dose of 2 g/kg body weight in divided doses over 5 days [2,3]. The mechanism of action of IVIG is also unclear but immune globulin may neutralise pathogenic antibodies and inhibit autoantibody-mediated complement activation.

The pharmacokinetics of IVIG is variable and some patients have a smaller increment in serum IgG after IVIG administration [4]. These patients have a worse prognosis.

The management of patients who fail to respond to initial treatment is complex. Administration of IVIG after plasma exchange is no better than either plasma exchange or IVIG alone [2]. One study suggested that patients unresponsive to initial treatment with IVIG may benefit from a second course of IVIG [5].

The role of plasma exchange after administration of IVIG is unclear. It has been suggested that plasma exchange could remove any IVIG that was previously administered [6]. However a few case reports and case series have reported that patients have been improved by plasma exchange after failing IVIG treatment [7]. So, performing plasma exchange after administration of IVIG does not necessarily reduce the efficacy of IVIG.

Conclusion

There are several anecdotal reports of the successful use of plasma exchange to treat patients with GBS after failure of IVIG. Unfortunately, there are no randomised controlled trials of this management strategy for GBS. Indeed, such data will be extremely difficult to obtain as this is a relative rare situation and the availability of plasma exchange is limited. So, plasma exchange of patients with severe GBS should be considered if IVIG fails and their clinical condition deteriorates.

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