

Outcome of Guillain-Barré Syndrome after Five Sessions of Plasmapheresis

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ABSTRACT

OBJECTIVE: To find out the outcome of Guillain-Barré Syndrome after five sessions of Plasmapheresis.

METHODOLOGY: This was prospective, descriptive observational study conducted from January 2018 - July 2019 in neurology ward 28, JPMC Karachi. Patients above 12 years of age diagnosed as Guillain-Barré Syndrome admitted through OPD and emergency were included. Patients diagnosed on the basis of clinical findings, cerebrospinal fluid detail report and nerve conduction studies. In Cerebrospinal fluid detail report, albuminocytologic dissociation was observed. Double lumen catheter was passed and plasmapheresis was done on every alternate day, 1.5-2.0 liters of plasma was removed and replaced by 4FFPS, 1000ml of normal saline and 500 ml of haemocoel for patients of diabetic neuropathy. Outcome of the patients was recorded as complete recovery, residual deficit and mortality. Patients were followed for six months. Data was analyzed by SPSS version 24.

RESULTS: Eighty nine patients of Guillain-Barré Syndrome were enrolled. Male patients were 60 (67.42%) whereas female patients were 29(32.58%). There were 15 patients (16.85%) within 12-20 years of age, 46 patients (51.68%) in 21-40 years of age, 19 patients (21.34%) from 41-60 years of age, 9 patients (10.11%) were above 60 years of age, 5 patients (5.62%) died while 64(71.91%) patients were fully recovered after plasma exchange. Twenty patients (22.47%) were partially recovered with residual motor weakness.

CONCLUSION: Plasma exchange is a good treatment modality for the Guillain-Barré Syndrome patients.

KEYWORDS: Guillain-Barré Syndrome, Plasmapheresis, Neuropathy.

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INTRODUCTION

Guillain-Barré Syndrome (GBS) an autoimmune disease is a common cause of generalized progressive paralysis. Guillain-Barré Syndrome is actually a poly radiculoneuropathy with available clinical course. Guillain-Barré Syndrome is mainly a post infectious disease manifested by rapid onset progressive, symmetrical weakness of the extremities. About 25% of Guillain-Barré Syndrome patients may develop respiratory failure and sometimes autonomic dysfunction¹.

Diagnosis is usually made clinically and once cerebrospinal fluid detail report, albuminocytologic dissociation and electrophysiological studies are done to make diagnosis². Plasma exchange in Guillain-Barré Syndrome shortens the duration and severity of illness. Plasma exchange is a procedure in which patients' blood is passed through machine, which separates the different type soft blood cells and returns all the cellular components like RBC, WBC and platelets along with either donor plasma or a replacement solution. This procedure removes substances which attacks myelin. Plasma exchange is very beneficial when started within a week after illness

but it was also beneficial in patients treated up to 30 days after disease onset³.

Plasma exchange is a better option for patients of Guillain-Barré syndrome because of negligible complications and highly cost effective³. It reduces the economic burden as compared to Intravenous globulin. Guillain-Barré Syndrome has no cure but plasma exchange can reduce the duration of illness and aids patient's recovery⁴. Spontaneous recovery may occur, however up to 75% of patients may develop sequelae⁴. Mortality in Guillain-Barré Syndrome patients is 5%. Plasma exchange removes antibodies from the blood stream. In Guillain-Barré Syndrome 5-6 therapeutic plasma exchange over 14 days are recommended with 5% albumin replacement. Patients of Guillain-Barré Syndrome benefit from plasmapheresis irrespective of initial severity so it should be started early as more benefit will occur. Plasma exchange reduces the risk of developing respiratory failure.

The rationale of study is that plasmapheresis treatment of Guillain-Barré Syndrome is available only in few centers and rarely the studies are done on this procedure in Pakistan. We are doing plasmapheresis

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in patients of Guillain-Barr'e Syndrome in J.P.M.C. Karachi to look for prognosis of Guillain-Barr'e Syndrome patients in our setup.

METHODOLOGY

This prospective, observational study was conducted from January 2018 till July 2019 at Jinnah post Graduate Medical Centre Karachi Ward-28. Eighty nine patients who were above 12 years of age both male and female were admitted emergency and OPD were diagnosed as Guillain-Barr'e Syndrome on clinical findings and lab investigations like Cerebrospinal fluid D/R and nerve conduction studies were included. Complete history and physical examination was done. Patients presented with quadriparesis, difficulty in breathing, paraparesis and autonomic dysfunction were also included. Their investigations like lumbar puncture were done. Albuminocytologic dissociation was observed in Cerebrospinal fluid D/R. Nerve Conduction Studies of all patients was done as well, before procedure of Blood Pressure, Pulse, Respiratory rate and temperature was recorded. Patients' consents were taken after explaining them the risks and benefits of the procedure. Double lumen catheter was passed in femoral vein or internal jugular vein. Plasma exchange was done in all patients. 4FFPs, in combination with normal saline 1 litre and haemocoel was given as a replacement fluid. 5 cycles of plasma exchange was done on alternate days and were completed. Post plasmapheresis, patient outcome was recorded as residual deficit or partial recovery and full recovery or mortality. After 2 weeks patients were discharged and were followed for 6 months in OPD to see the improvement in motor weakness. Results were analyzed on SPSS version 24 in frequency.

RESULTS

A total of 89 patients of Guillain-Barr'e Syndrome were enrolled in the study. Male patients were 60(67.42%) whereas female patients were 29(32.58%), so male patients were predominantly involved. There were 15 patients (16.85%) within 12-20 years of age range. 46 patients (51.68%) in 21-40 years of age range, 19 patients (21.34%) from 41-60 years of age range, 9 patients (10.11%) were above 60 years of age. Eighty four patients (94.38%) were recovered after 5 cycles of plasma exchange and five patients (5.62%) died due to respiratory failure and autonomic dysfunction. Fully recovered patients had Grade-5 power in all four limits while in partially recovered patients power improved from Grade-1 to Grade-3. In GBS, outcome of plasmapheresis is quick Figure I.

FIGURE I: NUMBER OF GUILLAIN-BARR'E SYNDROME PATIENTS

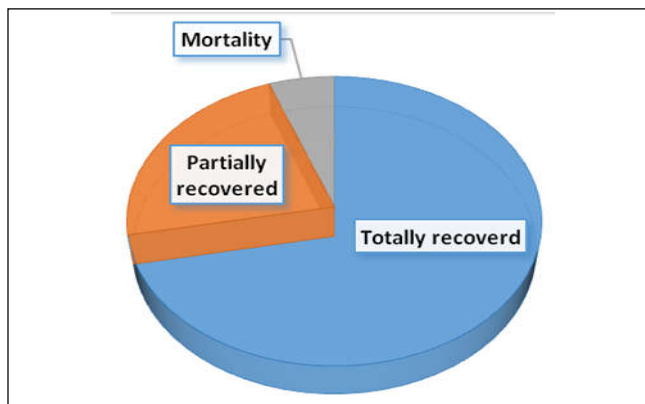


TABLE I: OUTCOME OF GBS PATIENTS AFTER FIVE CYCLES OF PLASMAPHERESIS

Outcome of GBS patients	No of Patients	Percentage
Totally recovered	64	71.91%
Partially recovered	20	22.47%
Mortality	5	5.62%
Total no of patients	89	100%

DISCUSSION

Guillain-Barr'e Syndrome is a serious disorder in which peripheral nerves become inflamed which leads to paralysis and sensory symptom. Many people who develop Guillain-Barr'e Syndrome have a history of preceding viral illness like upper respiratory tract infections and intestinal infections campylobacter jejuni-antibodies against the infection target the nerves myelin sheath and causes Guillain-Barr'e Syndrome. Plasma exchange removes soluble factors including anti bodies from the blood and it is the treatment for patients of Guillain-Barr'e syndrome. Plasma exchange replaces the patients' plasma with an artificial plasma substitute usually on albumin solution⁶.

Cause of Guillain-Barr'e Syndrome is not known yet. Men are 1.5 times more affected than women⁶. In this study males were affected more than females. Campylobacter jejuni gastroenteritis may be the preceding illness responsible for Guillain-Barr'e Syndrome,⁷ therefore immune response can lead to cross infection on host's nerves. 70% patients have been reported to have this infection⁷.

After the acute phase of disease Guillain-Barr'e patients do well. Most patients achieve independent ambulation after 6 months⁸. Mortality during acute phase of disease is less than 5%⁹, 20% patients may

have significant disability. Early identification of poor prognostic factors can lead to further treatment like plasma exchange. Early diagnostic findings in Guillain-Barré Syndrome include absent or prolonged H reflexes or frequency wave latencies¹⁰. The sural sparing pattern is considered specific for Guillain-Barré Syndrome as compared to other polyneuropathies¹¹. This pattern showed on intact sural sensory response with abnormal upper extremity sensory responses. Other findings depend on variants of Guillain-Barré Syndrome. Acute inflammatory demyelinating polyneuropathy more likely to have partial motor conduction block, temporal dispersion, slow conduction velocities, prolonged /absent F-wave latencies and prolonged distal latencies.

Autonomic Dysfunction is the main cause of morbidity and mortality attributable to Guillain-Barré Syndrome and involvement of respiratory muscles can lead to respiratory failures and death. In this study, mostly the cause of death was autonomic failure, like hypotension, hypertension, cardiac arrhythmia, and tachycardia.

Intravenous immunoglobulin's and plasma exchange are proven good treatment for Guillain-Barré Syndrome, but in spite of these treatments, the prognosis of Guillain-Barré syndrome is still not satisfactory. The zipper method of treatment seems to reduce the mortality. In this method plasma exchange is done by removing the plasma and replacement by albumin at the end of exchange. 4 gm/kg/Intravenous immunoglobulin should be given. This method seems to reduce the mortality, speed up weaning from mechanical ventilation and shortens the hospital stay in severe Guillain-Barré Syndrome. But in this study we have not given the albumin as a replacement fluid and also intravenous immunoglobulin.

Spontaneous recovery in Guillain-Barré Syndrome may occur. About 5% patients may develop ophthalmoplegia, ataxia and areflexia. Autoimmune pathogenesis is strongly suggested by the presence of antibodies against the four gangliosides-GM₁, GDA, GT1A and GQ1b which differ by number and position of sialic acid. The observation of preceding infection with campylobacter jejuni suggests cross reactive antibodies may be a component in disease pathogenesis. There are several scales to evaluate the severity and prognosis of Guillain-Barré syndrome disability. Medical research council sum score (MRCS. Score) ARASMUS score³. GBS Respiratory insufficiency score and ARASMUS GBS outcome score. Most of the treatment goal is to lessen the severity of Guillain-Barré Syndrome illness and to assess the patients' recovery.

High quality ICU care remains the most important aspect of management of Guillain-Barré syndrome.

Plasma exchange and high dose Intravenous globulin are equally effective modes of treatment¹². Physical therapy is advised to all patients when they are in the hospital. The mechanism, which intravenous immunoglobulin works in Guillain-Barré Syndrome is unclear. Intravenous globulin has minimal side effects like headache, skin reactions at infusion sites, flu-like symptoms, and aseptic meningitis, thromboembolic events such as pulmonary embolism due to increased viscosity of the blood are usually seen rarely¹³. The small volume plasma exchange has safety and feasibility with little side effects like sepsis, DVT and hypersensitivity reactions. Patients can be treated safely as compared to standard plasma exchange¹⁴. In this study we have not encountered any complications like pulmonary embolism and deep venous thrombosis.

Some people give the steroids alone in Guillain-Barré syndrome, but steroids are ineffective and can worsen the Guillain-Barré Syndrome, but steroids if given with Intravenous globulin has good effect on Guillain-Barré syndrome. Patients with Miller Fischer Variant also benefit from Intravenous globulin and 20% still unable to walk after ½ year.

Many patients have pain, fatigue or other residual complaints which persists for months for sometimes years. Prognostic model for Guillain-Barré Syndrome are now available. The ARASMUS GBS respiratory scale predicts the chance for artificial ventilation. Modified ARASMUS GBS outcome scale predicts the chance to be able to walk after 6 months.

Guillain-Barré Syndrome patients with poor prognosis potentially might benefit from more intensive management. A large increase in serum IgG levels after standard Intravenous globulin treatment 4gm/kg/B.W for 5 consecutive days seems to improve the Guillain-Barré syndrome. The aid GBS trial investigates second Intravenous globulin course improves outcome in GBS patients with poor outcome. Patients with Guillain-Barré Syndrome especially need excellent multidisciplinary care to prevent fatal complications, this shows Guillain-Barré Syndrome need careful monitoring of cardiac and respiratory function. 25% of severely involved patients require mechanical ventilation and regularly evaluation of vital capacity and timely transfer to an ICU. They need early prophylaxis for DVT, cardiac and hemodynamic monitoring, Pain management, possible management of bladder dysfunction. Complications like choking and airway obstruction may lead to death in ICU patients¹⁵. Physiotherapy during early course of disease should be started. Rehabilitation should be encouraged in recovery phase. For severe fatigue, pain killers and physical training is very beneficial. We started early physiotherapy.

The limitation in this study was that if we include the children below 12 years of age it can change the mortality and prognosis. Secondly, most patients presented earlier and plasmapheresis was done immediately so the results are better.

CONCLUSION

Plasmapheresis five cycles in Guillain-Barr'e Syndrome proved to be a good modality of treatment with good outcome of results.

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