**ORIGINAL ARTICLE**

# Outcome of various treatments in Guillain–Barré Syndrome in Children

JAMAL ANWER, SAJIDA KHALID, RIAZ UL HAQ, RAGIB IQBAL, ZAHID MAHMOOD, HAFIZ TAYYAB
*Department of Pediatric Medicine, Sh. Zayed Medical College / Hospital, Rahim Yar Khan, Punjab.*
*Correspondence to:Professor Dr. Jamal Anwer, E-mail: drjamalanwer2000@yahoo.com, Cell: 03007829858*

## ABSTRACT

**Aim:** The outcome of various treatments of GBS in children.
**Study design:** Cross sectional study
**Study location:** Department of Paediatrics, SZMC and Hospital, Rahim Yar Khan
**Study duration:** From November 2014 to December 2015
**Methods**: All the consecutive children having Guillain–Barré syndrome.These children were selected according to inclusion and exclusion criteria.
Sample size: Thirty-fourchildren with GBS.
There were three types of treatment offered; group A Intravenous Immunoglobulin, group B Plasma exchange and group C, Inj. Dexamethasone and oral prednisolone. All groups were given supportive care.
**Results:** In this study Plasma Exchange has the best (100%) cure rate while IV Immunoglobulin G has moderate (64%) and steroid has worst (43%) cure rate.
**Conclusion:** In our study, we found that some treatments used in GBS are very effective, however some patients do not give response to the treatment and plasma exchange has best cure rate.
**Keywords:**Guillain–Barré syndrome (GBS), IVIG, Plasma exchange (Plasmapheresis), Dexamethasone/Prednisolone

## INTRODUCTION

Guillain–Barré Syndromeis a polyneuropathy which occurs after some infectious disease and involves mainly the motor nervous system. Sometimes,it may involve the sensory and autonomic nervous system as well. It affects all age groups[1]. However it is rare in children[2]. The incidence is 1.2–2.3 per 100,000 populations per year. Most commonly Campylobacter jejuniis the preceding infection. Others infections are Epstein–Barr virus, cytomegalovirus, hemophilus influenza and mycoplasma pneumoniae. There are some documented associations of GBS with vaccinations, operations or stressful conditions.

The commonesttype of GBS is acute inflammatory demyelinating polyneuropathy (AIDP) which constitutes about 75% of the syndrome. It may also involve the sensory neurons. However, acute motor axonal neuropathy (AMAN)is exclusively motor disorder and involve the axons only. In this disease, the antibodies are formed against gangliosides, especially GM1. A third variety of GBS is Acute Motor and Axonal Neuropathy. It is an acute axonal disorder which involves both motor and sensory nerves. Miller Fisher syndrome is a variant of GBS with no weakness. There is ophthalmoplegia, ataxia and areflexia. This disorder is characterized by antibodies to neuromuscular junction ganglioside GQ1b and interfere with transmitter release.

The GBS is an antigen antibody reaction against peripheral nerves after some respiratory or gastrointestinal infectionsspecially in AMAN and Miller Fisher syndrome[3]. However, in AIDP, cellular mechanism may be more important.Although antibodies have a role in the process of demyelination.

The most important clinical feature of GBS is rapid progression of weakness. The most serious complication of GBS is respiratory failure due to respiratory muscles weakness leading to fatigue. Ventilation is required in around 10% of cases[4]. Maximum weakness occurs within 4 weeks. The plateau phase ranges from days to weeks or even months. Then a slower and variable recovery phase starts. In spite of giving, intravenous immunoglobulin (IVIG) or plasma exchange (PE) about 20% of patients remain unable to walk after 6 months. Some patients remain disabled or severely fatigued and in these patients even after 3 to 6 years, there is a great hindrance in social life and daily activities.

The treatment of GBS is mainly supportive. Specific treatment is IVIG or plasmapheresis.[5]In plasma exchange (plasmapheresis), the plasma is removed from the blood of the patient. The blood cells are mixedwith normal saline and infused again intothe body. In this way plasma containing the autoantibodies is removed. In immunoglobulin therapy, high doses of intravenous immunoglobulin (IVIG)are given which can block the antibodies damaging the neurons. Steroids have controversial role. It can be given in oral or injectable form. Prednisolone, methylprednisolone and dexamethasone are having the same results.

These patients should be managed in an intensive care unit (ICU). The vital signs monitoring should be done and input output of record should be maintained.Nutritional care and prevention from bed sore are important.Guillain-Barre syndrome is a progressive neuropathy leading to paralysis. The physiotherapy is the mainstay of supportive treatment in these patients which is requiredin every stage of recovery[6].

In most patients of Guillain-Barre syndrome following course of disease can be observed; after the first manifestation of disease, the condition will progressively increase in severity for about two weeks. Then thesymptoms reach a plateau in four weeks. Afterward recovery starts, usually lasting 6 - 12 months. Although in some patients the course may be as long as three years. In

children, GBS is usually benign, and spontaneous recovery begins within 2-3 weeks but in adults the clinical course of the disease is highly variable and difficult to predict. The objective of this study is to assess the outcome of different treatment used to treat Guillain–Barré syndrome among children.

## METHODOLOGY

This cross sectional study was conducted in the Department of Paediatrics, SZMC and Hospital, Rahim Yar Khan from November 2014 to December 2015. All the consecutive children having Guillain–Barré syndrome were selected according to inclusion and exclusion criteria. Sample size: Thirty-four children with GBS. There were three types of treatment offered; group A Intravenous Immunoglobulin, group B Plasma exchange and group C, Inj. Dexamethasone and or oral prednisolone. All groups were given supportive care.

**Inclusion criteria:**

- Children below 14 years of age suffering from symmetrical asending paralysis.
- CSF:

**Protein: raised**

WBC Count: not more then 50/mm$^3$

Nerve Conduction Studies: Polyneuropathy

**Exclusion criteria:** Patients not fulfilling the inclusion criteria. Ethical approval was sought from IRB of hospital? Data analysis was done by using SPSS version 20 and outcome variables were: Patients cured, Died, and left against medical advice.

## RESULTS

Thirty four patients were included in the study.Mean age was 9±3 years and 13(38%) were female patients.

Table I: Outcome of treatments used to treat Guillain Barre Syndrome patients

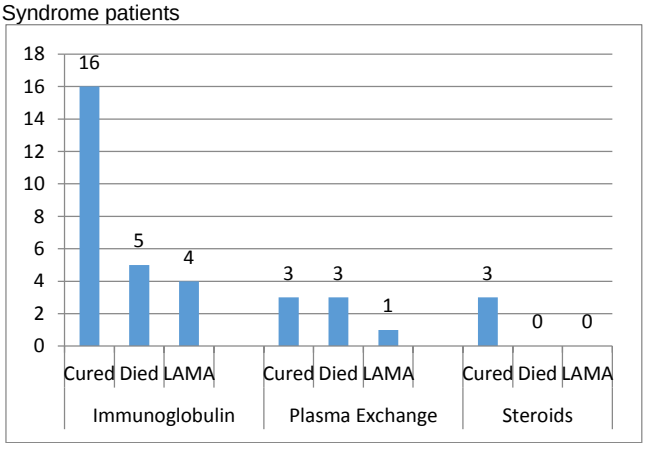


Table I shows treatment outcome among patients treated with three different modalities. In this study PE has the best (100%) while IVIG has moderate (64%) and steroid has worse (43%) result. In this study, 20% patients receiving IVIG died and 16% left against medical advice. 43% patients receiving steroids died and 14% left against medical advice.

## DISCUSSION

Guillain–Barré Syndrome is a polyneuropathy which occurs after some infectious disease and involves mainly the motor nervous system. It may occur in any age. In 100,000 populations, the incidence of GBS is 1.2–2.3 per year. The treatment of Guillain-Barre syndrome is only supportive and there is no cure. Intravenous immunoglobulins and Plasma exchange (Plasmapheresis) are the most important ways of treatment which can hastethe recovery phase. These also reduce the severity of illness. However steroid may have a controversial role. Jaydip Ray Chaudhuri, Suvarna Alladi et al concluded that in developing countries, Plasmapheresis may be a better option in treatment of GBS.[7]According toPieter A. Van , Krista Kuitwaard et al most effective treatment of GBS is Intravenous immunoglobulin (IVIG)[8]. Matthew Harms concluded that different treatments of GBS can limit the severity but patients remained at risk of complications[9]. Diener HC, Haupt WF et al found significant improvement in disability after four weeks treatment withPlasmapheresis then the untreated control.[10]Hughes RA, Swan AV et al described that none of the treatments (plasma exchange, IVIG, corticosteroid) significantly reduced mortality[11]. R Korinthenberg and J S Mönting found that immunoglobulins were able to accelerate recovery in children who were not able to walk in the start of treatment. Corticosteroids were less potent. However, Plasmapheresis could not be evaluated because of itslimitation in the most severe cases[12]. Gürses N, Uysal S et al concluded that IVIG is a safe and effective treatment which also shortens the time to recovery[13]. Schessl J, Korinthenberg Ret al found that treatment with IVIG gives a faster recovery[14]. Hughes RA, Wijdicks EF et al described that the treatment with plasma pheresis or IV immunoglobulin (IVIG) hastensthe recovery from GBS but the combination of two treatment modalities in the same patient is not beneficial. Similarly, treatment with steroid alone is not useful[15]. Färkkilä M, Kinnunen E et al described that there is increasestrength of the muscles and decrease CSF protein. These findings are more significant in the plasma exchange group than in the control group, but this had no effect on duration of hospital stay or recovery periods[16]. Alshekhlee A, Hussain Z et al concluded that mortality rate in GBS is low, what so ever be the treatment[17]. Chiò A, Cocito D et al described the same thing that in GBS both mortality and morbidityare related to axonal involvement which ever be the treatment[18]. Bersano A, Carpo M et al described that 90% of GBS patients included in the study had more or less complete functional recovery with different modes of treatment[19]. Schröder A, Linker RA, Gold R successfully applied in chronic inflammatory demyelinating polyneuropathy[20]. Chevret S, Raphaël JC et al found that plasma exchange is superior to supportive treatment alone[21]. The controlled trial by Osterman PO, Fagius J et al in 38 patients with severe acute inflammatory polyradiculoneuropathy concluded that course of the disease was shortened in plasma exchange.[22] Christine Verboon, Pieter A Van described that intravenous immunoglobulin (IVIG) and plasma exchange are very useful in motor improvement of GBS[23]. Hughes RA, Pritchard J et al described that Chinese herbal medicine

tripterygiumpoly glycoside accelerated the recovery more than corticosteroids but this result needs more research[24]. Hugh J Willison, Bart C Jacobs et al described that the treatment of GBS withsupportive care and intravenous immunoglobulin or plasma exchange is the best management approach.[25]Porcher R, Orlikowski D et al suggest that viral replication may be associated with the neuropathological processes of CMV related GBS[26].

## CONCLUSION

In our study, we found that some treatments used in GBS are very effective, however some patients do not give response to the treatment and a long term follow up was required. Morbidity is more as compare to the mortality. In spite of poor resources, Plasmapheresis is better than other treatment options.

## REFERENCES


1. Harvey B. Sarnat “GuillainBarre Syndrome” Nelson text book of Pediatrics 19th Edition, P: 3010-3013.
2. Pervez Akbar Khan “Neuromuscular Disorders” Basis of Pediatrics, 8th Edition P 31-34.
3. Pervez Akbar Khan “Neuromuscular Disorders” Basis of Pediatrcs, 8th Edition P 31-34.
4. Mary Imelda Hughes "Neurology" Forfar and Arneil's, Textbook of Pediatrics, 7th edition. P 930.
5. Mary Imelda Hughes "Nurology" Forfar and Arneil's, Textbook of Pediatrics, 7th edition. P 930.
6. Harvey B. Sarnat “GuillainBarre Syndrome” Nelson text book of Pediatrics 19th Edition, P: 3010-3013.
7. Jaydip Ray Chaudhuri, SuvarnaAlladi, K RukminiMridula “Clinical outcome in-Barré syndrome with various treatment methods and cost effectiveness” A study from tertiary care center in South India, Neurology Asia 2014; 19(3): 263 – 270.
8. Pieter A. van Doorn, Krista Kuitwaard, Chrkista Walgaard, Rinske van Koningsveld,[3] Liselotte Ruts, and Bart C. Jacobs“IVIG Treatment and Prognosis in Guillain–Barré Syndrome” J ClinImmunol. 2010 May; 30(Suppl 1): 74–78.
9. Matthew Harms “In patient Management of Guillain-Barré Syndrome” Neurohospitalist. 2011 Apr; 1(2): 78–84.
10. Diener HC, Haupt WF, Kloss TM, “A preliminary, randomized, multicenter study comparing intravenous immunoglobulin, plasma exchange, and immune adsorption in Guillain-Barré syndrome” Eur Neurol. 2001;46(2):107-9.
11. Hughes RA, Swan AV, Raphaël JC, Annane D, van Koningsveld R, van Doorn PA Immunotherapy for Guillain-Barré syndrome: a systematic review Brain. 2007 Sep;130(Pt 9):2245-57. Epub 2007 Mar 2.
12. R Korinthenberg, J S Mönting “Natural history and treatment effects in Guillain-Barré syndrome: a multicentre study” Arch Dis Child. 1996 Apr; 74(4): 281–287.
13. Gürses N, Uysal S, Cetinkaya F, Işlek I,Kalayci AG. Intravenous immunoglobulin treatment in children with Guillain-Barre syndrome Scand J Infect Dis. 1995;27(3):241-3.
14. Korinthenberg R, Schessl J, Kirschner J, Mönting JS “Intravenously administered immunoglobulin in the treatment of childhood Guillain-Barré syndrome: a randomized trial” Pediatrics. 2005 Jul;116(1):8-14.
15. Hughes RA, Wijdicks EF, Barohn R, Benson E, Cornblath DR, Hahn AF, Meythaler JM, Miller RG, Sladky JT, Stevens JC “Practice parameter: immunotherapy for Guillain-Barré syndrome: report of the Quality Standards Subcommittee of the American Academy of Neurology” Neurology. 2003 Sep 23;61(6):736-40.
16. Färkkilä M, Kinnunen E, Haapanen E, Iivanainen MGuillain-Barré syndrome: quantitative measurement of plasma exchange therapy. Neurology. 1987 May;37(5):837-40.
17. Alshekhlee A, Hussain Z, Sultan B, KatirjiB.Guillain-Barré syndrome: incidence and mortality rates in US hospitals. Neurology. 2008 Apr 29;70(18):1608-13.
18. Chiò A, Cocito D, Leone M, Giordana MT, Mora G “Guillain-Barré syndrome: a prospective, population-based incidence and outcome survey” Neurology. 2003 Apr 8;60(7):1146-50.
19. Bersano A, Carpo M, Allaria S, Franciotta D, Citterio A, Nobile-Orazio E “Long term disability and social status change after Guillain-Barré syndrome” J Neurol. 2006 Feb;253(2):214-8. Epub 2005 Aug 17.
20. Schröder A, Linker RA, Gold R “Plasmapheresis for neurological disorders” Expert Rev Neurother. 2009 Sep;9(9):1331-9.
21. Raphaël JC, Chevret S, Hughes RA, Annane D “Plasma exchange for Guillain-Barré syndrome” Cochrane Database Syst Rev. 2001;(2):CD001798.
22. Osterman PO, Fagius J, Lundemo G, Pihlstedt P, Pirskanen R, Sidén A, Säfwenberg J “Beneficial effects of plasma exchange in acute inflammatory polyradiculoneuropathy” Lancet. 1984 Dec 8;2(8415):1296-9.
23. Christine Verboon, Pieter A van Doorn, Bart C Jacobs “Treatment dilemmas in Guillain-Barré syndrome” *J NeurolNeurosurg Psychiatry.*doi:10.1136/jnnp-2016-314862
24. Hughes RA, Pritchard J, Hadden RD “Pharmacological treatment other than corticosteroids, intravenous immunoglobulin and plasma exchange for Guillain-Barré syndrome” Cochrane Database Syst Rev. 2013.
25. Hugh J Willison, Bart C Jacobs, Pieter A van Doorn “Guillain-Barré syndrome” Lancet 2016; 388: 717–27.
26. Orlikowski D, Porcher R, Sivadon-Tardy V, Quincampoix JC, Raphaël JC, Durand MC, Sharshar T “Guillain-Barré syndrome following primary cytomegalovirus infection: a prospective cohort study” Clin Infect Dis. 2011 Apr 1;52(7):837-44.