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Case Report Open Access

# Guillain Barre Syndrome and its Variants: A Case Report on Acute Motor - Sensory Neuropathy

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## Abstract

Guillain Barre Syndrome is an autoimmune disease and acute idiopathic polyneuritis condition. This is considered as Monophasic Immune Mediated Disorder (MIMD) and as acute inflammatory demyelinating poly-radiculoneuropathy in peripheral nervous system. Around 1-3 people affecting in 1,00,000 population. AMSAN and AMAN are rare subtypes of Guillain Barre Syndrome. The accurate cause of this disease is unknown. Around 2-12% people die due to GBS fallowed respiratory paralysis. Generally, Immunoglobulins (IgG) and plasma exchange given as effective treatment. Here we find a case of 18 years male patient presented with complaints of generalized weakness since 10 days and ascending paralysis followed by descending paralysis for one day. Based on family history and nerve conduction studies, it was concluded with AMSAN type of GBS. In order to treat the condition nutrition therapy, physiotherapy and steroids were given to the patient, though steroids are considered as ineffective treatment.

**Keywords:** Immunoglobulins; Monophasic immune mediated disorder; Plasma exchange

**Abbreviations:** Acute Motor Axonal Neuropathy (AMAN); Acute Motor and Sensory Axonal Neuropathy (AMSAN)

## Introduction

Guillain Barre Syndrome, also known as Landry paralysis, it is the acute idiopathic polyneuritis; it is immunologically mediated disorder [1]. Guillain Barre Syndrome (GBS) is an acute onset and monophasic immune-mediated disorder (MIMD) in the peripheral nervous system. The term GBS is the synonymous of Acute Inflammatory Demyelinating Polyradiculoneuropathy (AIDP) [2]. It is a rare immune mediated polyradiculoneuropathy, which has an incidence of 1-3 per 1,00,000 population according to epidemiological studies in Europe, USA, Australia [3]. Females and Males are equally at risk [4-11]. Acute Motor Axonal Neuropathy (AMAN) and Acute Motor and Sensory Axonal Neuropathy (AMSAN) are subtypes of GBS mainly occur by acute inflammatory demyelinating nerves and Miller Fisher Syndrome (MFS) is also considered as an uncommon variant of Guillain-Barré syndrome (GBS). It is characterized by ataxia, areflexia and external ophthalmoplegia [12-15]. GBS remains as life threatening disease with mortality rate of 2-12% people [7] and 3-7% in USA and Europe. An American fatality study found that the most common complications that cause death in GBS were cardiovascular and respiratory paralysis [5]. The exact etiology and pathophysiology of GBS was not completely understood, but genetic and environmental factors that affect an individual's susceptibility to develop the disease [10]. The majority of GBS cases are affected by prior infection of Gastroenteritis and Respiratory tract infections and subsequence abnormal immune response towards the infectious agents, typically with the onset of symptoms 2-4 weeks after the primary infections [14]. It is thought to be an immune mediated process, results from the generation of auto-immune antibodies and inflammatory cells that cross- react with epitopes on roots of pheripheral nerves leading to axonal damage and demyelination or both [4]. The immune response towards many infections have been identified, including cytomegalovirus (CMV) *Campylobacter jejuni*, influenza virus, mycoplasma pneumonia, Epstein Barre virus, Japanese encephalitis virus (JEV) can trigger GBS [1,4,10]. Vaccines are other antigenic stimulus that potentially associates with GBS have been reported, they include formulations of simple rabies vaccine, tetanus toxoid vaccine and some formulation of influenza vaccine [4]. The AIDP, histological appearance resembles the experimental autoimmune neuritis that which commonly caused by T-cells directed P0,P2 and PMP22 [1,10]. In AIDP-the role of T-cell mediated immunity remains unclear and there is evidence that proves the involvement of antibodies and complement. Strong evidence that exist now are acute motor axonal neuropathy (AMAN) and acute motor and sensory neuropathy (AMSAN) are caused by antibodies towards gangliosides on the axolemma that target macrophages to invade the axon at Node of Ranvier [2]. In early detection and characterization of inflammatory demyelinating polyradiculopathies Electro diagnosis play an important role [11]. In order to treat GBS the Immunoglobulin therapy and plasma exchange are recommended as equally effective treatments [2], addition of nutrition therapy can show faster effect [3]. Hypokalaemia is a well-recognised common complication for therapeutic plasma exchange. There is a possibility of sudden death due to cardiac arrhythmias which is predisposing effect of hypokalaemia [6]. Here we report a case of AMSAN type of GBS. In this, the patient was treated with Corticosteroids and with additional nutritional therapy & physiotherapy as there is no availability of IgG and PE.

## Case Report

An 18-year old male, 3rd child & 1st son to his parents presented with complaints of generalised muscle weakness since 10 days and ascending paralysis followed by descending paralysis since day 1. On examination, the patient was conscious and coherent. Pallor, cyanosis, jaundice, clubbing, edema and lymphadenopathies were absent. His blood pressure was found to be 125/80 mmHg with no orthostatic hypotension. His pulse was 86/minutes and regular. Respiratory

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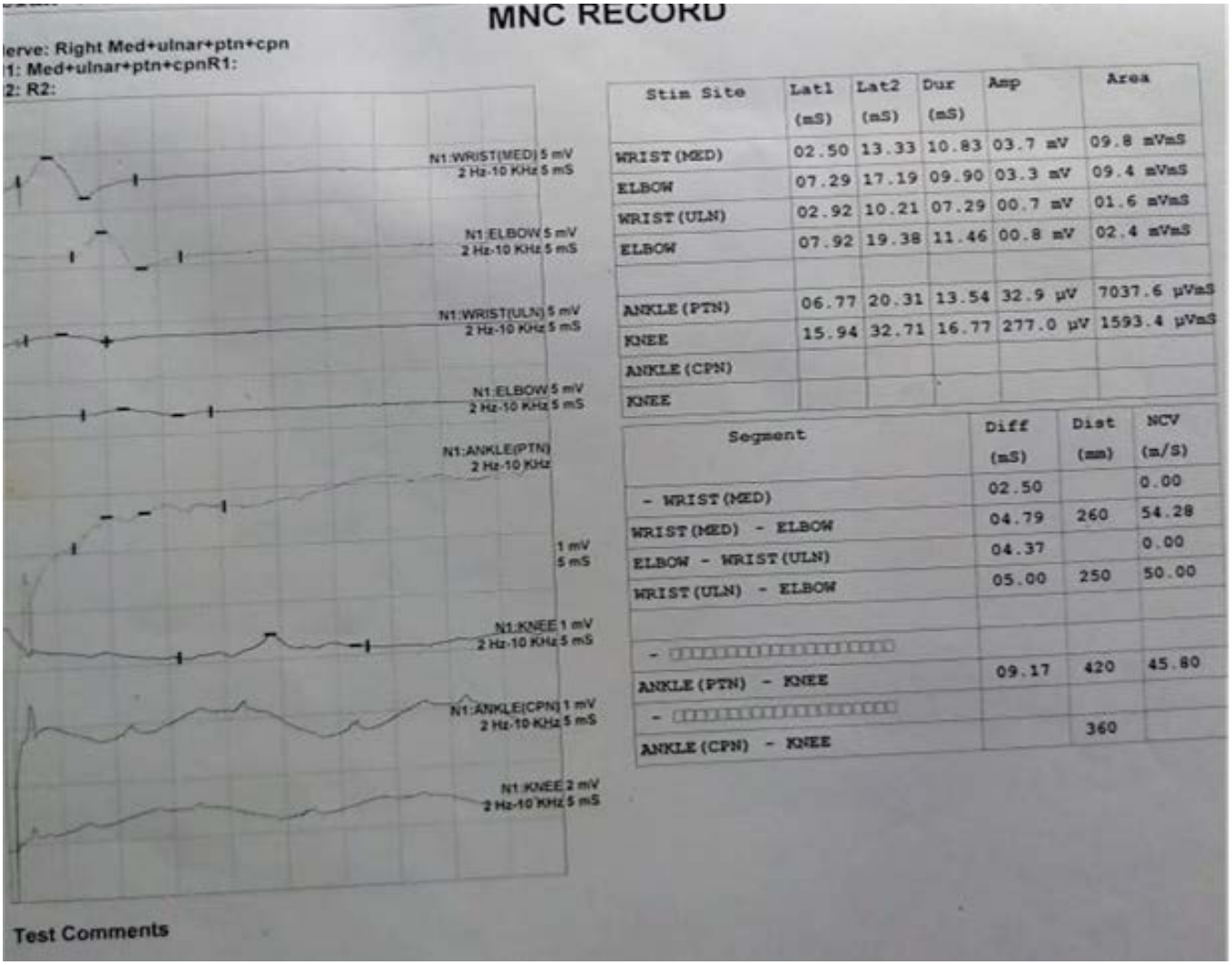
rate was 18/minute. There were no complaints of unconsciousness, convulsions, hearing defects, speech problems, vertigo, dysphagia, tinnitus. Bladder and bowel functions were normal. He had no history of upper respiratory tract infection and loose motion in last one month but affected with mild fever 2 weeks back. He had no recent history of any surgery, drug abuse, vaccination, alcohol addiction and extra marital sexual exposure. On examination of nerve conduction studies on sensory and motor nerves, the result was found to be decreased amplitude in motor nerve studies shown in Figure 1.

In motor nerve studies, at right upper limbs amplitudes were found as wrist med-3.7 mV; Elbow-3.3 mV; Wrist ULN-0.7 mV; Elbow-3.7 mV; Ankle PTN-32.9 μV; Knee-277.0 μV; Inp upper left limbs the amplitudes was noted as wristmed-2.2 mV; Elbow-2.1 mV; Wrist ULN-98.6 μV; Elbow-nm 28.2 μV; Ankle PTN-50.2 μV; knee-173.7 μV; Ankle CPN-154.9 μV; Knee-14.1 μV. The 1st latency interval was increased in upper limb right AnklePTN-6.77 and in upper limb left Ankle PTN-6.98; Ankle CPN-7.19 (Figure 2).

On examination of family history, the patient's grandfather (maternal side) was affected with same condition and younger sister (2nd child to his mother) was affected minorly with similar symptoms but not as severe as this patient and she was managed with nutritional therapy. Based on family history and nerve conduction studies it was considered as severe axonal and demyelinating sensory motar polyneuropathy, with clinical correlation of progressive weakness for 10 days consistent it was concluded with AMSAN variant of GBS.

## Treatment

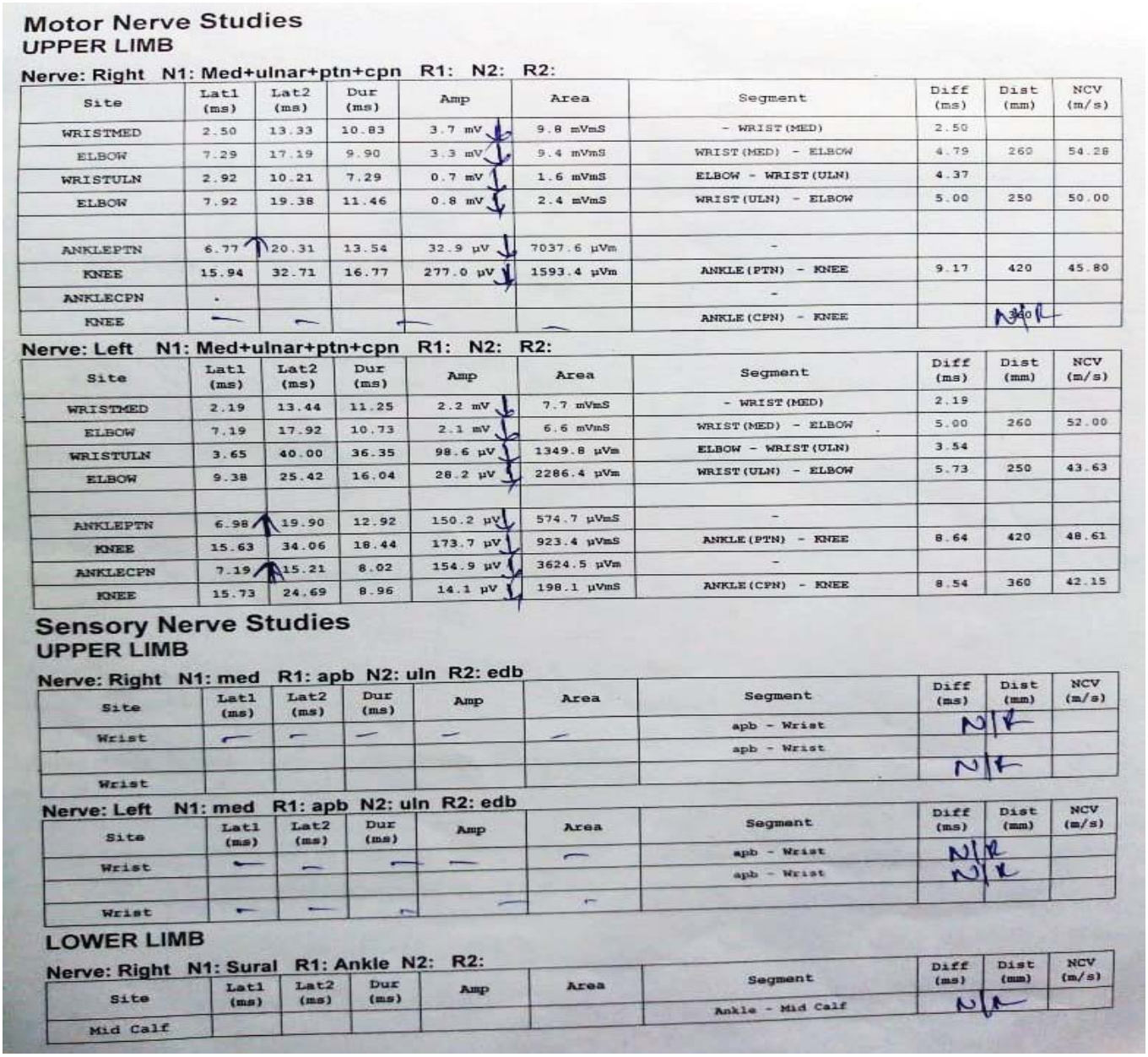
As there is no availability of Immunoglobulins and plasma exchange conditions in the hospital and at nearby locations of hospital, so, the patient was treated with Injection Methyl Prednisolone – 500 mg/IV/OD/5 days (Corticosteroid); Tablet Gabapentin- 100 mg/OD/5 days; Tablet Multivitamin/OD/1 month; and Tablet Calcium carbonate/OD/1 month along with Physiotherapy treatment for 1month then discharged. After 10 days of discharge, the patient reconsulted doctor with complaints of shortness of breath since 2 days. Then X- ray of chest was performed and conclude as Bronchitis (prominent Broncho vascular markings in bilateral lung fields). To treat the condition with Capsule Doxycycline, Tablet Azithromycin and Tablet Digoxin were prescribed for 5 days. After 10 days, the patient was normalised but affected with generalised weakness after recovery from bronchitis condition. So, to treat the condition he was treated with 1 month course of Tablet Multivitamin and Tablet Calcium Carbonate.




| Stim Site | Lat1 (mS) | Lat2 (mS) | Dur (mS) | Amp | Area |
|---|---|---|---|---|---|
| WRIST (MED) | 02.50 | 13.33 | 10.83 | 03.7 mV | 09.8 mVmS |
| ELBOW | 07.29 | 17.19 | 09.90 | 03.3 mV | 09.4 mVmS |
| WRIST (ULN) | 02.92 | 10.21 | 07.29 | 00.7 mV | 01.6 mVmS |
| ELBOW | 07.92 | 19.38 | 11.46 | 00.8 mV | 02.4 mVmS |
| ANKLE (PTN) | 06.77 | 20.31 | 13.54 | 32.9 μV | 7037.6 μVmS |
| KNEE | 15.94 | 32.71 | 16.77 | 277.0 μV | 1593.4 μVmS |
| ANKLE (CPN) | | | | | |
| KNEE | | | | | |

| Segment | Diff (mS) | Dist (mm) | NCV (m/S) |
|---|---|---|---|
| - WRIST (MED) | 02.50 | | 0.00 |
| WRIST (MED) - ELBOW | 04.79 | 260 | 54.28 |
| ELBOW - WRIST (ULN) | 04.37 | | 0.00 |
| WRIST (ULN) - ELBOW | 05.00 | 250 | 50.00 |
| - | | | |
| ANKLE (PTN) - KNEE | 09.17 | 420 | 45.80 |
| - | | | |
| ANKLE (CPN) - KNEE | | 360 | |

**Figure 1:** Motor nerve conduction recordings in upper and lower limbs.



**Motor Nerve Studies**

**UPPER LIMB**

**Nerve: Right N1: Med+ulnar+ptn+cpn R1: N2: R2:**

| Site | Lat1 (ms) | Lat2 (ms) | Dur (ms) | Amp | Area | Segment | Diff (ms) | Dist (mm) | NCV (m/s) |
|---|---|---|---|---|---|---|---|---|---|
| WRISTMED | 2.50 | 13.33 | 10.83 | 3.7 mV | 9.8 mVmS | - WRIST (MED) | 2.50 | | |
| ELBOW | 7.29 | 17.19 | 9.90 | 3.3 mV | 9.4 mVmS | WRIST (MED) - ELBOW | 4.79 | 260 | 54.28 |
| WRISTULN | 2.92 | 10.21 | 7.29 | 0.7 mV | 1.6 mVmS | ELBOW - WRIST (ULN) | 4.37 | | |
| ELBOW | 7.92 | 19.38 | 11.46 | 0.8 mV | 2.4 mVmS | WRIST (ULN) - ELBOW | 5.00 | 250 | 50.00 |
| | | | | | | | | | |
| ANKLEPTN | 6.77 | 20.31 | 13.54 | 32.9 µV | 7037.6 µVm | - | | | |
| KNEE | 15.94 | 32.71 | 16.77 | 277.0 µV | 1593.4 µVm | ANKLE (PTN) - KNEE | 9.17 | 420 | 45.80 |
| ANKLECPN | - | | | | | - | | | |
| KNEE | | | | | | ANKLE (CPN) - KNEE | | 360 | |

**Nerve: Left N1: Med+ulnar+ptn+cpn R1: N2: R2:**

| Site | Lat1 (ms) | Lat2 (ms) | Dur (ms) | Amp | Area | Segment | Diff (ms) | Dist (mm) | NCV (m/s) |
|---|---|---|---|---|---|---|---|---|---|
| WRISTMED | 2.19 | 13.44 | 11.25 | 2.2 mV | 7.7 mVmS | - WRIST (MED) | 2.19 | | |
| ELBOW | 7.19 | 17.92 | 10.73 | 2.1 mV | 6.6 mVmS | WRIST (MED) - ELBOW | 5.00 | 260 | 52.00 |
| WRISTULN | 3.65 | 40.00 | 36.35 | 98.6 µV | 1349.8 µVm | ELBOW - WRIST (ULN) | 3.54 | | |
| ELBOW | 9.38 | 25.42 | 16.04 | 28.2 µV | 2286.4 µVm | WRIST (ULN) - ELBOW | 5.73 | 250 | 43.63 |
| | | | | | | | | | |
| ANKLEPTN | 6.98 | 19.90 | 12.92 | 150.2 µV | 574.7 µVmS | - | | | |
| KNEE | 15.63 | 34.06 | 18.44 | 173.7 µV | 923.4 µVmS | ANKLE (PTN) - KNEE | 8.64 | 420 | 48.61 |
| ANKLECPN | 7.19 | 15.21 | 8.02 | 154.9 µV | 3624.5 µVm | - | | | |
| KNEE | 15.73 | 24.69 | 8.96 | 14.1 µV | 198.1 µVmS | ANKLE (CPN) - KNEE | 8.54 | 360 | 42.15 |

**Sensory Nerve Studies**

**UPPER LIMB**

**Nerve: Right N1: med R1: apb N2: uln R2: edb**

| Site | Lat1 (ms) | Lat2 (ms) | Dur (ms) | Amp | Area | Segment | Diff (ms) | Dist (mm) | NCV (m/s) |
|---|---|---|---|---|---|---|---|---|---|
| Wrist | | | | | | apb - Wrist | | | |
| | | | | | | apb - Wrist | | | |
| Wrist | | | | | | | | | |

**Nerve: Left N1: med R1: apb N2: uln R2: edb**

| Site | Lat1 (ms) | Lat2 (ms) | Dur (ms) | Amp | Area | Segment | Diff (ms) | Dist (mm) | NCV (m/s) |
|---|---|---|---|---|---|---|---|---|---|
| Wrist | | | | | | apb - Wrist | | | |
| | | | | | | apb - Wrist | | | |
| Wrist | | | | | | | | | |

**LOWER LIMB**

**Nerve: Right N1: Sural R1: Ankle N2: R2:**

| Site | Lat1 (ms) | Lat2 (ms) | Dur (ms) | Amp | Area | Segment | Diff (ms) | Dist (mm) | NCV (m/s) |
|---|---|---|---|---|---|---|---|---|---|
| Mid Calf | | | | | | Ankle - Mid Calf | | | |

**Figure 2:** Readings of motor nerve studies and sensory nerve studies in upper and lower limbs.

After 6 months of normal condition, reconsulted doctor with complaints of generalized weakness and he was treated with Tablet Melalben fotel (Paracetamol + Aspirin + Caffeine) and Capsule Vit-E for 10 days, Tablet Multivitamin for 1 month along with physiotherapy for 1 month. He was also with complaint of severe muscle weakness during physically strained condition, so he was advised to have Capsule Felicita (benfotiamine, meobalamin, alpha lipoic acid, and chromium picolinate and inositol soft gelatin capsule). It can be used only at body weakened condition not as regular continuous medication.

## Discussion

Guillain Barre Syndrome is a rare case with incidence of 1-3 in 1,00,000 population. Here this case report is treated with the steroidal therapy, nutritional therapy and physiotherapy. The plasmapheresis and immunoglobulins were not used in this case though they are considered as effective and standard treatments.

About quarter of patients with GBS are affected with *C. jejuni* infection. The liposaccharide from *C. jejuni* infection bacterial wall contains gangliosides like structures and when injected into rabbits induce a neuropathy that resembles acute motor axonal neuropathy [2]. The Antibodies to GD1a, GM1, GM1b and GalNac-GD1 are in AMAN and GalNac-GD1 in AMSAN [2,10].

To diagnose the condition cerebro spinal fluid test and biochemical tests and nerve conduction studies of motor and sensory nerves are used. The cerebrospinal tests were performed but the parameters showed no signs of cerebral ad other infections and did not present with any fever. Nerve conduction studies are also performed and

amplitude in nerve conduction of motor and sensory nerves were decreased. Generally, in AMSAN type of GBS amplitudes of motor and sensory nerves decreases in nerve conduction studies.

In order to treat the condition, he was administered with methyl prednisolone 500 mg/I.V., for 5 days, once daily; Gabapentin 100 mg/p.o., once in a day; Multivitamin once daily and calcium carbonate once daily. But calcium carbonate should not be administered at the same time with methyl prednisolone and gabapentin, it leads to interaction. Calcium carbonate decreases the absorption of methyl prednisolone and with gabapentin it increases elimination. So, in a day they have administered in altered time to avoid interaction.

To treat the bronchitis, he was administered with antibiotics Azithromycin 500 mg and Doxycycline 100 mg; cardiac glycosides Tablet Digoxin. But Digoxin is not administered at same time in a day with antibiotics as it increases the level or effect of digoxin by altering intestinal flora. The further generalised weakness of the patient was treated with nutritional therapy (multivitamin) and with physiotherapy for 1 month. Thus, the AMSAN type of GBS was treated in this patient without use of immunoglobulins and plasmapheresis.

Intravenous immunoglobulin (IV Ig) is a beneficial treatment in other autoimmune diseases. This review was 1st published in 2001 and updated in 2003, 2005, 2007 ad 2010 [13-15]. The Cochrane systemic reviews shown that plasma exchange (PE) has significant recovery in GBS compared with other supportive treatment. The researchers concluded that "A short course of high dose Methyl prednisolone is ineffective in early stage of GBS is ineffective" [16]. In Cochrane systematic review of 6 trails with 587 patients it has shown that corticosteroids are ineffective for treating GBS. But in combination with immunoglobulins show effectiveness [12]. In this case as there is no availability of standard medication, steroids were used, and they show therapeutic and recovery effect on this patient with combination of nutritional therapy and physiotherapy. The patient was able to walk normally after 2nd physiotherapy (after onset of 3 months). About 20% of patients with Guillain-Barre syndrome cannot walk unaided 6 months after onset [10]. He was set up to be normal and with no fresh complaints for 12 months.

## Conclusion

The AMSAN type of GBS is a rare and leading type in GBS that cause death. To treat the GBS, Immunoglobulins and plasmapheresis are treated as effective treatment rather than steroids but as they were not available at area hospital and its surroundings the patient was treated with steroids and physiotherapy along with additional nutritional therapy and thus the patient was treated and recovered with GBS.

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