CASE REPORT

Acute Motor Axonal Neuropathy: A Challenging Rehabilitation Case of a 6-Year-Old Child with Respiratory Failure

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ABSTRACT

Background: Acute motor axonal neuropathy (AMAN) is an autoimmune disorder that targets the axons of motor neurons without significant demyelination or sensory involvement. It primarily affects the lower motor neurons, leading to muscle weakness and diminished reflexes. It is a subtype of Guillain-Barre syndrome. Acute Motor Axonal Neuropathy (AMAN) in children shares similarities with adult presentations but also shows unique characteristics due to developmental and physiological differences. It typically appears after the third year of life.

Case Summary: A 6-year-old male child was reported to the emergency room with ascending weakness, drooling of saliva, and difficulty in breathing, along with flaccid tetraplegia and areflexia. An electrophysiological examination showed severe motor axonal neuropathy.

Interventions: A progressive program is designed using functional exercises.

These are as follows: Functional Training, Cardio-respiratory Training, Balance Training, and Coordination Training. Exercises progressed from passive ROM through gravity-eliminated AROM and antigravity-AROM to resisted functional exercises.

Outcome measures: The outcome measures are, Muscle Strength using Manual Muscle Testing (MMT); Disability and Progression of the disease using the Overall Neuropathy Limitation Scale (ONLS) and Hughes Severity Scale; Cardio-respiratory fitness using Borg Rating of Perceived Exertion (Original); (BBS); Mobility using Dynamic Gait Index (DGI).

Conclusion: Hence, structured and supervised exercises, including supervised cycling or prescribed unsupervised exercises and aerobic activities, have significantly improved strength, balance, gait, and cardiorespiratory functions.

Keywords: Guillain-Barre Syndrome; Child; AMAN; Respiratory Insufficiency, Functional Capacities, Muscle strength.

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INTRODUCTION

Acute motor axonal neuropathy (AMAN) is a subtype of GBS characterized by decreased compound muscle action potentials (CMAP). It is an autoimmune disorder, ascending polyneuropathy, which manifests itself as a lower motor neuron lesion. It occurs mainly after a prior infection [1]. GBS has a rare worldwide prevalence of 1.2–2.3 per 100,000 persons per year [2]. AMAN is rarely reported in Western countries (3-5% of all reported GBS cases) and has the most severe respiratory symptoms of all GBS variants [3,5]. However, a higher frequency of AMAN is reported in Japan, Pakistan, India, China, and Mexico, ranging from 28% to 67% [2]. Typically, it begins with distal, symmetrical paraesthesia, followed by increasing limb weakening. A common symptom is pain, which is generally reported as a deep ache or cramp in the buttocks, thighs, or between the shoulders. Most patients eventually recover completely or nearly completely, with the ability to walk alone after 3 months and with only minor remaining complaints by the end of the first year [3,4]. A case of a 6-year-old male with significant respiratory compromise caused by AMAN was treated with IVIG. The Patient was on positive pressure ventilatory support and under neurorehabilitation training.

Picture 1: Child on SIMV support

Patient information: On 25 September 2021, a 6-year-old male patient presented to the Emergency Department of Rainbow Children`s Hospitals, Malviya Nagar, New Delhi, with ascending weakness, drooling saliva, and difficulty breathing. He had no significant past medical history. He is a student and enjoys running and playing with his elder sister. He had no history of traveling or infection.

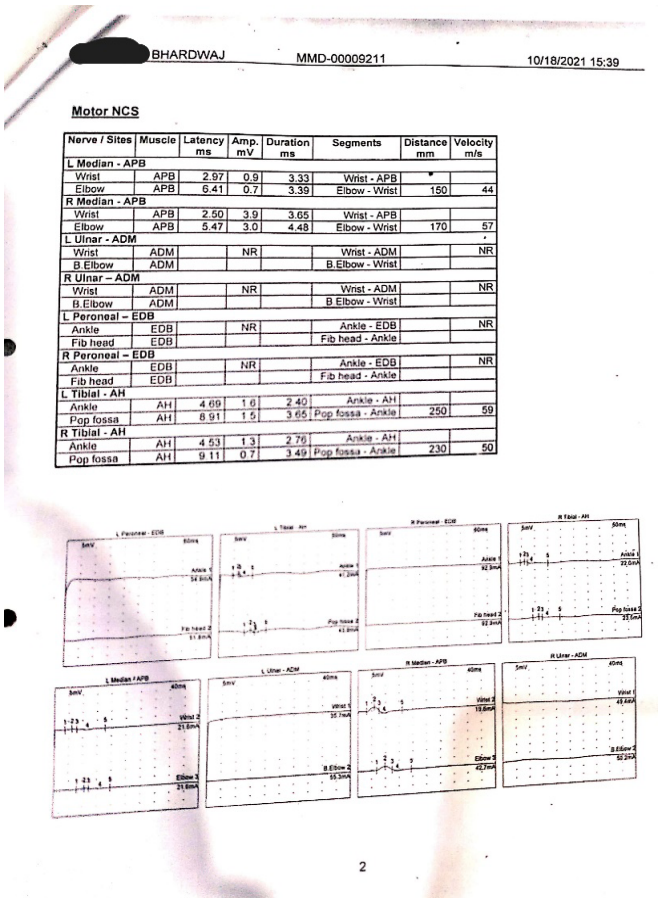
Physical Examination: Dysphonia, flaccid tetraplegia, and areflexia were found in the neurological examination.

Picture 2: Child after some weeks of hospital stay, improving with respiratory status

Diagnosis and Assessment: In electrophysiological studies, motor conduction blocks in all peripheral nerves suggest severe motor axonal neuropathy. The child started with IVIG. While re-examining the patient, it was found that the child was tachypneic and not maintaining saturation on room air; his Gag was also absent. The power in his limb was 1/5. As seen in Picture 1, the child was on ventilatory support for 28 days and gradually weaned off to BIPAP from the ventilator with oxygen support after some time. After some days, respiratory exercise and pulmonary conditioning were merged with the protocol, as depicted in Picture 2. Physiotherapy was started after the stabilization of vitals. The child's initial assessment at the first session was done by reviewing various systems, as shown in Table 1.

System Review	Result
Cardiopulmonary	Intact: RHR 90 beats per minute; BP 100/80 mm of Hg; Respiratory rate 20 breaths per minute
Musculoskeletal	Impaired: forward head posture, weakness in all extremities
Neuromuscular	Impaired: slow, uncoordinated gross movement, poor balance, decreased locomotion.
Communication, cognition, and Learning style	Intact: Able to understand and communicate in English and Hindi very well. Cognitively sound, visual, and verbal learner.

Table 1: Review of Systems at the Initial Assessment



Picture 3: NCV report of the patient

Furthermore, Tables 2 and 3 show the initial and discharge assessment of the case for various outcome measures and muscle strength in detail. A Nerve Conduction Velocity (NCV) test was also conducted, as seen in Picture 3, which showed severe motor axonal neuropathy in all tested nerves of the bilateral upper and lower limbs. The ICF model for Disability was considered for designing the child's rehabilitation intervention (Figure 1). The model shows physical difficulties and the child's aim of participation. Also, the family is supportive, and the house is located on the ground floor, which are supportive elements for rehabilitation.

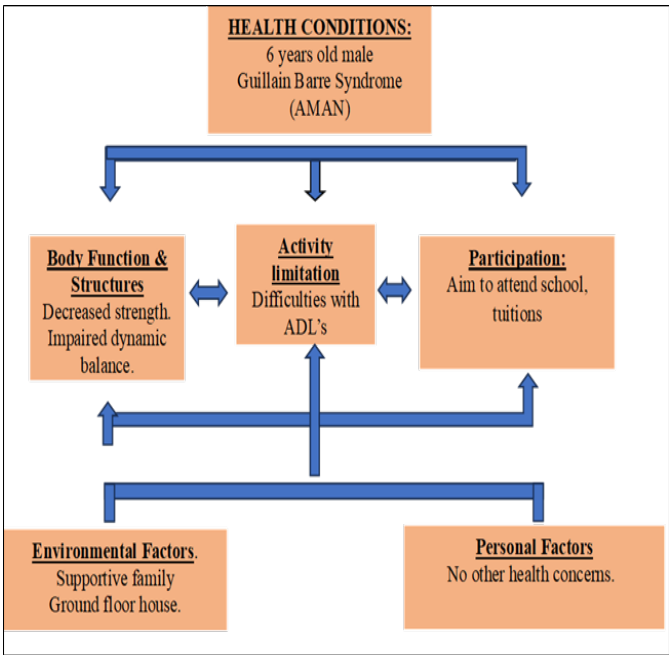


Figure 1: ICF model with factors specific to the case

Test and Measures	Initial (November 2nd, 2021)	Discharge (October 31st, 2022)
Active Range of motion (AROM)	All AROMs were complete (except dorsiflexion 5^0 B/L	All AROMs were complete (including ROM of dorsiflexion 15^0 B/L)
Posture	The forward head is scapular protected, the right shoulder is elevated, and there is increased lumbar lordosis.	No deviation seen
Locomotion/gait	Contact guarding of 1 person i s required. Not able to maintain sitting balance. No independent gait.	Ambulates independently and can run. There is a slight difference in the spatial gait parameter between the left and right foot.
Manual muscle testing (MMT)	Increased weakness of distal extremities > proximal extremities.	Predominantly weakness in B/L G. Medius
Gross motor coordination	Impaired heel-to-shin and rapid alternating movements.	Intact with accuracy, precision, and speed.
Single leg stance	Unable to perform	30-second bilaterally
Overall neuropathy limitation scale (ONLS)	12/12 (maximum disability)	0/12 (No disability)
Hughes Severity Scale	Hughes score 4 (Bedridden or chair bound)	Hughes score 1 (slight clinical symptoms and signs)
Pediatric Berg Balance Scale	16/56 (Wheel-chair-bound)	47/56 (Independent)
Modified Functional Independence Measure (FIM)	52/126 (Level 2- Maximum assistance)	124/126 (Level 7 – Complete independence)
Dynamic Gait Index (DGI) with modification for children	0/24 (Severe fall risk)	22/24 (safe ambulator)
Borg Rate of perceived exertion (RPE)	At rest: 9/20 On activity: 19/20	At rest: 6/20 On activity: 14/20

Table 2: Initial and Discharge assessment including various tests and outcome measures

Manual Muscle Testing (MMT)	Initial		On Discharge	
	Left	Right	Left	Right
Shoulder flexor	4	4	5	5
Shoulder extensor	3	3	5	5
Shoulder abductor	3	3	5	5
Shoulder IR	3	3	4	4
Shoulder ER	3	3	4	4
Elbow flexor	4	4	5	5
Elbow extensor	4	4	5	5
Forearm supinator	3	3	5	5
Wrist flexor	3	3	5	5
Wrist extensor	3	3	5	5
Hip flexor	3	3	5	5
Hip extensor	4	3	5	5
Hip abductor	4	3	3	3
Hip adductor	3	3	4	4
Hip IR	3	3	4	4
Hip ER	3	3	4	4
Knee flexor	4	4	5	5
Knee extensor	4	4	5	5
Ankle dorsiflexor	3	3	5	5
Ankle planter flexor	3	3	5	5
Trunk Flexor	2		4	
Trunk Extensor	2		4	

Table 3: MMT (Manual Muscle Testing) of major muscle groups

INTERVENTION

The intervention was designed to keep the short-term and long-term rehabilitation goals in mind. The goals are as follows. A progressive Program using Functional

exercises was used. It is further divided into four subcategories.

I. Functional Training
II. Cardio-Respiratory Training
III. Balance Training
IV. Coordination Training

Exercises progressed from Passive ROM through gravity eliminated AROM and antigravity, AROM to resisted functional exercises. Exercises were performed for the upper extremities, lower extremities, and trunk with 5-10 repetitions. The exercises were stopped before the patient reported fatigue. The session typically lasted 60 minutes. The detailed rehabilitation Program is shown in Table 5.

Intervention Goals	
Short-term goals to be achieved in 4-6 weeks.	**Long-term goals to be achieved in 8- 12 weeks.**
➢ Restoration of maximal independence. ➢ To improve strength and range of affected joints. ➢ Maintenance of airways. ➢ Patient and family counselling and motivation to decrease fear of fall and boost up level of confidence.	➢ Good alignment of posture. ➢ To maintain static balance and improve dynamic balance. ➢ Maintain proper gait pattern with coordinated movements. ➢ Maintain the strength of muscles and range of motion. ➢ Maintenance of airways. ➢ Lifestyle modifications. ➢ Regular follow-ups and reassessment.

Table 4. Goals for Rehabilitation Protocol

TYPE/ EXERCISE	FREQUENCY	INTENSITY	DURATION/ TIME	DESCRIPTION
FUNCTIONAL TRAINING				
Hurdle crossing	4 Rounds of 6 hurdles / per session 6 days a week	½ kg weight cuffs, six hurdles	Within half an hour for FT	Forward and Sideways crossing of hurdles with a height of 1 foot
Stair climbing	1 set/ session 5 times a day	½ kg weight cuffs ~100 (50+50) stairs		For promoting hip and knee flexion
Bird Dog Exercise	2 sets/ session 5 days a week	½ kg weight cuff 12 reps with 5 sec hold time	5 minutes	For the core strengthening
Pushups against the wall	2 sets/ session 5 times a week	12 reps		For strengthening of shoulder, chest, and back musculature
Ball toss with therapist	2 sets/ session 5 times a week	12 reps		
Jumping jacks	2 sets/session 5 times a week	12 reps		
Step up and step down on stairs	2 sets/ session 5 times a week	12 reps		
Stepping over 19" foam pad	3 sets/ session 5 times a week	20 reps.		
Marching on plane	2 sets/ session 5 times a week		2 minutes for each set	
Jumping the hurdle of 4"	2 sets/ session 5 times a week	20 reps		
Hanging on ladder	3 sets/ session 5 times a week		For 1 minute for each set	
Squats	3 sets/session 5 times a week	20 reps		
Walking on a ladder	3 sets/ session 5 times a week			Quick step forward, backward and sideways
Kicking the ball	3 sets/ session 5 times a week	20 reps		

CARDIO-RESPIRATORY TRAINING				
Bicycle ergometry	5 times/ week	45% HRmax	15 minutes	
Incentive spirometry	10 inhalations/session Thrice a week	Breaths should be held for at least 2 or 3 seconds at a minimum at full inspiration. Expiration is performed slowly and calmly with the lips.		
Manual chest wall compression	1 cycle/ session Thrice a week		5 minutes	Done by pressing the bilateral lower rib cage during expiration.
BALANCE TRAINING				
One leg standing.	2 sets/ session 5 times a day	15 reps.	30 sec holds with eye open; 5 sec with eye closed	To improve stability and reduce falling risk
Toe and heel raise.	2 sets/ session 5 times a day	15 reps		
Marching on plane	2 sets/ session 5 times a day	15 reps		
Walking on a ladder	3 sets/ session 5 times a week			Quick step forward, backward and sideways
Mild perturbations (backward, forward, sideways) in standing position	2 sets/ session 5 times a day	15 reps		
COORDINATION TRAINING				
Drawing a circle	2 sets/ session 5 times a day	15 reps		
Touch therapist's finger with toe.	2 sets/ session 5 times a day	15 reps		
Heel on shin.	2 sets/ session 5 times a day	15 reps		
Alternate heel to knee and heel to toe	2 sets/ session 5 times a day	15 reps		
Drawing a circle	2 sets/ session 5 times a day	15 reps		

Table 5. Detailed Protocol Followed for Rehabilitation

FOLLOW-UP AND OUTCOME MEASURES:

The child showed improvement after 7-8 weeks of regular and supervised home exercises. The drastic improvement seen in Picture 4, where the child crosses hurdles. The progress was observed and documented regularly; the difference was seen between the various outcome measures from the initial assessment (November 2nd, 2021) to the day of discharge (October 31st, 2022).

Primary outcome: Muscle Strength using Manual Muscle Testing (MMT); Disability and Progression of the disease using the Overall Neuropathy Limitation Scale (ONLS) and Hughes Severity Scale; Cardio-respiratory fitness using Borg Rating of Perceived Exertion (Original). *Secondary outcome:* Balance using the Pediatric Berg Balance Scale (BBS); Mobility and Gait parameters using Dynamic Gait Index (DGI). Table 6 shows the difference in pre- and post-rehabilitation scores of outcome measures.

Picture 4: Phase III Rehab session

Outcome Measures	Range of Score	Pre	Post	Diff.
Hughes Severity Scale	0-6	4	1	-2
Overall Neuropathy Limitation Scale	0-12	12	0	-12
Pediatric BBS	0-56	16	47	31
DGI	0-24	0	22	22
RPE- Rest	6-20	9	6	-3
RPE- Activity	6-20	19	14	-5

Table 6: Pre and post-intervention values of outcome measures and differences among them

RESULT

The child showed improvement after 7-8 weeks of regular and supervised home exercises. The drastic improvement is shown in Picture 4, where the child can cross hurdles.

The progress was observed and documented regularly; there was a difference between the various outcome measures from the initial assessment (November 2nd, 2021) to the day of discharge (October 31st, 2022). Table 6 shows the pre and post-intervention scores and the Differences between them. The bar graphs are used to depict every outcome measure in detail.

Hughes Severity Index and Overall Neuropathy Limitation Scale

- Figure 2 shows that higher scores in Pre-Intervention for both tests indicate maximal disability.
- Hughes Severity Scale improved from moderate disability to mild disability.
- Overall neuropathy limitation scale (ONLS) depicts an initial maximal disability (Score 12) to no disability (Score 0) at discharge.

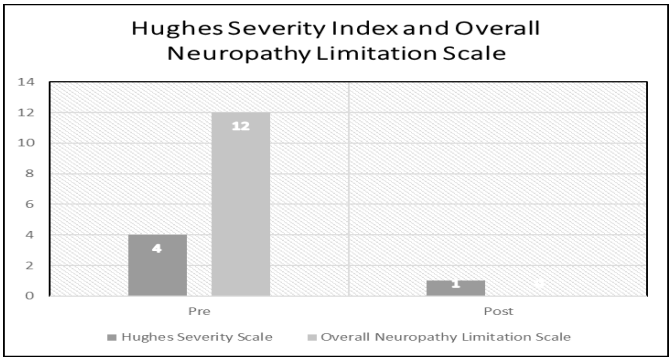


Figure 2: Hughes Severity Index and Overall Neuropathy Limitation Scale

Pediatric BBS and Dynamic Gait Index

- Figure 3 shows pre-intervention lower scores for both tests, indicating severe fall risk.
- Pediatric BBS improved from severe fall risk (16) to independent ambulator (47).
- Dynamic gait index (DGI) shows drastic improvement from severe fall risk (0) to safe ambulator (22).

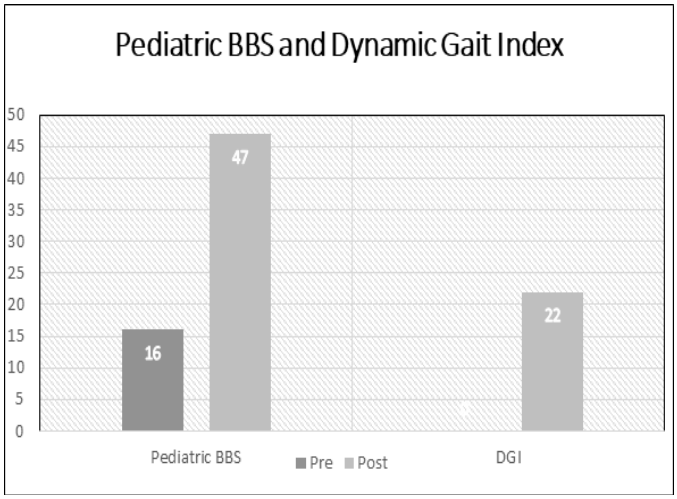


Figure 3: Pediatric BBS and Dynamic Gait Index

Borg Rate of Perceived Exertion (At Rest and Activity)

- The original version of borg was used (6-20 rating scale).
- Figure 4 shows that Borg rate of perceived exertion (RPE) showed drastic improvement at rest and activity between pre and post-rehabilitation.
- The B-RPE at rest improved from very light (9) to very-very light (6).
- The B-RPE at activity improved from very-very hard (19) to somewhat challenging (14).

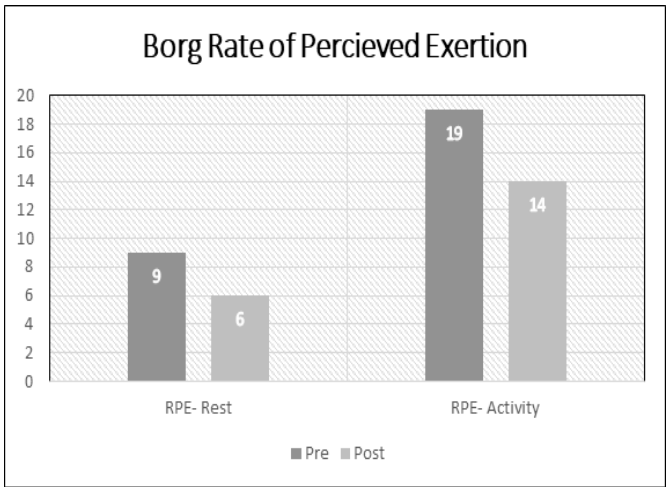


Figure 4: Borg Rate of Perceived Exertion (At Rest and Activity)

DISCUSSION

In this case report, we evaluated and rehabilitated a 6-year-old boy with the AMAN subtype of GBS. It was possible with the combined efforts of the patient, caretakers, and physiotherapists, which helped to develop the care and treatment plan. Although there were few clinical studies on managing GBS with physiotherapy,

the available evidence suggested that an exercise-based treatment plan that targeted the symptoms was necessary.

The Rehabilitation strategies were tailored to the patient's symptoms. Some symptoms were required to improve before other problems could be addressed. For example, respiratory control, weak muscles, and the trunk must be focused on before reeducating people to stand and walk. As a result, a structured treatment plan and procedure were developed for the patient.

Gradually, all the systems were targeted and showed improvement with continued exercise intervention. The symptoms were reduced gradually. A significant improvement was seen in the trunk muscle group, and the patient progressed to standing. At the time of discharge, he was able to run, jump, and play like other kids in school.

Evidence-based research, including longitudinal and randomised controlled trial studies, has yet to be conducted to make physiotherapy intervention effective.

The Physiotherapy treatment was effective because he enjoys spending time with his pals and has played football and other extracurricular activities. The protocol was designed to keep his interest in mind as a goal.

There is a dearth of case studies about the rehabilitation of GBS following such severe respiratory dependency. Our study structured various exercise types, tailored to patient needs, and intervened with supervision to the pediatric case.

The physical therapy sessions were carefully planned to focus on specific functional activities, considering the patient's presenting symptoms at each session. By adopting this approach, the physical therapy interventions successfully helped the patient achieve their goal of safe ambulation. Physical therapists must remain vigilant in assessing the patient's immediate and ongoing responses to treatment to ensure that the exercises prescribed are appropriate in type and intensity. The exercises should be challenging enough to elicit a physiological and therapeutic response but remain within the patient's irritability threshold to prevent undue fatigue and nervous stress [5].

Seven studies were included in a systematic review of the influence of exercise on adult patients with GBS. In most studies, aerobic training was performed, and cycling was found to improve the functional status of GBS patients. A study also gave progressive functional exercise intervention and found significant improvement in mobility and MMT. In the same review, supervised and structured exercises showed improvement in fatigue, isokinetic strength post 12 weeks, improvement in Physical domains of SF-36, and increased VO2 by 20% [6].

Although large-scale studies on this topic are limited, this case study demonstrates that physical therapy interventions can effectively aid Guillain-Barré Syndrome (GBS) patients in restoring function and reducing disability. The significance of this case lies in the rare opportunity to document the outcomes of physiotherapy interventions for GBS in a pediatric case. At discharge, the patient had achieved all goals and returned to their prior functional status, with only mild postural deviation and minimal weakness. This case shows the effectiveness of function-based interventions in managing Guillain-Barré syndrome (GBS) in childhood. Future research could explore and compare the outcomes of different physiotherapy approaches for GBS in pediatric populations.

CONCLUSION

Hence, structured and supervised exercises, including supervised cycling or prescribed unsupervised exercises and aerobic activities have shown significant improvement in strength, balance, gait, and cardiorespiratory functions.

This case report has been structured in accordance with the CARE (Case Report) guidelines to ensure the clarity, transparency, and completeness in reporting.

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