Medica Hospitalia
Journal of Clinical Medicine
Med Hosp 2024; vol 11 (1) : 113–118

OPEN ACCESS

Case Report

Rehabilitation of A Child with Low Endurance in the Recovery Phase of Guillain-Barré Syndrome

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p-ISSN: 2301-4369 e-ISSN: 2685-7898
https://doi.org/10.36408/mhjcm.v11i1.1020

Accepted: August 25th, 2023
Approved: December 28th, 2023

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Abstract

Background : Guillain-Barré Syndrome (GBS) usually has a good prognosis of recovery. However, some patients can have remaining disabilities due to low cardiorespiratory fitness or endurance and needs to be managed. This study was aimed to reports the rehabilitation assessment and management of a child with low cardiorespiratory endurance in the recovery phase of GBS.
Case : A 12-year-old girl with a history of hospitalization due to Acute Motor and Sensory Axonal Neuropathy (AMSAN)-type of GBS was referred to the Physical Medicine and Rehabilitation (PMR) outpatient clinic with tiredness that restricted her school participation. She had low cardiorespiratory endurance, which was confirmed by a six-minute walk test (6-MWT). After the rehabilitation program, her endurance level was increased, and she can return to school.
Discussion : A comprehensive assessment showed that the muscle weakness, accompanied by obesity, anemia, and inactivity, led to low cardiorespiratory endurance that restricted the activity and participation. A rehabilitation program that consisted of aerobic and strengthening exercises improved cardiorespiratory endurance, walking ability, and school participation.
Conclusion : Rehabilitation management in children with low cardiorespiratory endurance due to the sub-acute phase of GBS could help them regain their activity and participation during the recovery phase.

Keywords : Guillain-Barré Syndrome, endurance, rehabilitation

INTRODUCTION

Guillain-Barré Syndrome (GBS) is an autoimmune disorder that targets the myelin sheaths of the nerve roots and peripheral nerves, resulting in myelopathy and neuropraxia.[1] It is considered a rare case, especially in the pediatric population.[2] The prognosis is typically good, with 70% of patients regaining functional status similar to the premorbid condition. However, about 20% of children have remaining disabilities after GBS.[1–3] Several studies reported that physical training in GBS patients also aimed to increase cardiorespiratory fitness.[4] Low cardiorespiratory fitness or endurance can lead to symptom of fatigue or tiredness that can interfere with activity and participation.[5] Therefore, it needs to be identified and managed appropriately for a better clinical outcome.

The rehabilitation management of children in the recovery phase of GBS, particularly for the cardiorespiratory endurance problem, has not been reported before. This study presents the rehabilitation assessment and treatment of a child with low cardiorespiratory endurance in the subacute phase of GBS.

CASE PRESENTATION

A 12-year-old girl was referred to the PMR outpatient clinic with a history of hospitalization due to GBS. Two months before, the patient began to feel weakness, as well as tingling sensations and numbness on both lower limbs. The weakness was slowly spreading to the upper limbs and getting worse day by day, until she was no longer able to move her legs. She also felt discomfort when breathing. At first, she got treatment at a remote hospital, but due to limited facilities, she was referred to a National Referral Hospital. The results of the cerebrospinal fluid examination correspond to the diagnosis of GBS (Table 1). Nerve conduction studies (NCS) showed signs of motor and sensory demyelination and axonal polyneuropathy, which are consistent with the acute motor sensory axonal neuropathy (AMSAN) type GBS. She received five regimens of plasmapheresis treatment, after which she felt the strength and sensibility of the extremities gradually improved for the next several days. The patient was admitted for two weeks. When she was discharged, she was able to stand up while holding on to something for less than five minutes.

Three weeks after discharge, the patient visited PMR outpatient clinic. The chief complaint at that time was easily getting tired when walking more than 100 meters. She had not returned to school yet because of the difficulty of walking for about 1 km to reach her school and climbing the stairs to reach her class room on the second floor. She also could not do her hobbies, such as playing badminton and cycling. However, she is still able to do all the self-care by herself. The patient was able to walk at a slower pace but had to stop for several minutes because of tiredness after walking 100 meters. The fatigue complaint also made it difficult for her to climb more than five stairs.

On the physical examination, there was obesity with Body Mass Index 25.7 kg/m^2. The resting heart rate was 110 times per minute, which is slightly higher than the normal value (60–100 beats per minute). The respiratory rate was 20 times per minute, which is normal for 12 year-old child. The proprioceptive and light touch functions were normal in all extremities. All the muscles strength was normal with Manual Muscle Testing (MMT) of 5, except for a slight decrease in several lower extremity muscles, such as the shoulder and hip flexor, extensor, abductor, adductor, internal rotator, and external rotator

TABLE 1
Cerebrospinal Fluid Analysis

Parameters	Results	Normal
Color	Yellowish*	No color
Turbidity	Cloudy*	Clear
Clarity	Positive*	Negative
Cells Count	127 cells/mL*	0–10 cells/mL
PMNs (segment)	8%*	0–6%
MNs (lymphocyte)	119%*	40–80%
Protein	1326 mg/dL*	15–45 mg/dL
Glucose level	136 mg/dL*	60–100 mg/dL
Chloride	109 mEq/L	115–130 mEq/L

PMNs = Polymorphonucleocytes, MNs = Mononucleocytes, *abnormal results muscles strength (MMT 4). Her power prehension and precision grips were normal for both hands. The physiological reflexes for biceps, patellar tendon, and Achilles tendon were decreased.

Chest expansion on the axillary, xiphoid, and umbilical levels were 3, 5, and 5 cm, respectively, which are considered normal for her age. There was no accessory respiratory muscle contraction. The single breath count test (SBCT) was performed 30 times (normal cut-off value is 25 times). Standing balance, both static and dynamic, were adequate. The Borg rating of perceived exertion (RPE) at rest was 9, or "very light exertion" (on a scale of 6 to 20). We performed a six-minute walking test (6-MWT), and the distance was 150 meters, which is equal to VO2 max of 2.9 cc/kg/minute or 0.9 METs (metabolic equivalents). From the laboratory results, there was microcytic hypochromic anemia with a hemoglobin level of 9.7 g/dl. From the spirometry, FEV1 and FVC were 101% and 93%, respectively (FEV1/FVC: 110%), and were still considered normal. The peak cough flow was 360 L/minute (normal range is 147–488 liters/min for female children ages 4–18 years old). Thorax plain radiography showed no abnormality in the heart or lungs.

We prescribed a rehabilitation program that consisted of therapeutic exercise, education for increasing physical activity, and dietary management based on clinical nutritionist guidance. The hospital-based therapeutic exercises that she received were aerobic and strengthening exercises. We prescribed the exercise intensity according to the target of heart rate (HR) that was calculated with Karvonen Formula below.

Target HR = {(Maximal HR Resting HR) x Intensity %}+ Resting HR

We get the maximal HR from subtraction of 220 with the age. The exercise intensity was started at a low intensity (30–39%) for exercise adaption. Based on the formula, we got the target of heart rate of 120-140 times per minute. Aerobic exercise was performed by supervised leg ergo-cycle training for 30 minutes per session, 3 times per week. A strengthening exercise with 1 kg dumbbells was performed for the shoulder flexor, extensor, abductor, and adductor muscles, each for 3x10 repetitions. We also encouraged the patient to perform physical activity at home, which included sit to stand exercise for 3x10 repetitions and walking exercise for 30 minutes, both of which were performed 5 times per week.

At the follow-up two months later, the patient said that the tiredness when doing activities had improved. She was able to walk to school but still felt easily get tired in both legs after walking. Our physical examination showed a lower resting heart rate (91 times per minute) and a Borg RPE scale of 6. She also had increased muscle strength for the shoulder and hip muscles with MMT 5 on both sides. The 6-MWT result was 350 meters, which was equal to 4.25 METs (VO2 max 14.9 cc/kg/minute). We asked her to continue the rehabilitation program with the same types of interventions as the previous regimen. The exercise intensity was increased into moderate level. At this time, the target of heart rate was calculated with the current resting heart rate and moderate training intensity (40–60%) using Karvonen Formula, which resulted the target heart rate of 140–160 times per minute. We also encouraged the patient to start doing her previous sports activities, such as badminton and bicycling.

Two months after the last follow-up, the patient said that she no longer had complaints when carrying out her daily activities and going to school. The patient had been participating in sports activities at the school once a week. Patients routinely play bicycle three times a week for a duration of 30–60 minutes without any complaint. We performed another 6-MWT with a result of 400 meters, which was equal to 5,1 METs (VO2 max 17.8 cc/kg/minute).

DISCUSSION

Guillain-Barré Syndrome (GBS) is an autoimmune disorder manifested as myelopathy and neuropraxia.[5,6] The estimated incidence in Indonesia is 1 to 2 cases per 100,000 children, which is considered a rare case.2 It is preceded by an infection in four to six weeks before neurological symptoms emerge. The clinical manifestation of GBS is heterogeneous, with the peculiarities of progressive bilateral weakness of the limb and accompanied by the deprivation of reflexes.[7] It has several subtypes such as Acute Inflammatory Demyelinating Polyneuropathy (AIDP), Acute Motor Axonal Neuropathy (AMAN), Acute Motor and Sensory Axonal Neuropathy (AMSAN), and Miller-Fisher Syndrome (MFS).[8,9]

The National Institute of Neurological Disorders and Stroke (NINDS) has developed diagnostic criteria for GBS.[10] In this case, the patient met the NINDS criteria for GBS. She experienced progressive bilateral sensory impairment and weakness, accompanied by decreased physiological reflexes. The CSF examination showed increased CSF protein. The NCS showed signs of motor and sensory polyneuropathy with demyelination and axonal features. Thus, she was diagnosed with AMSAN-type GBS.

GBS has been linked to spontaneous recovery that happens soon after a plateau condition is established. After a plateau, the recovery phase may last many weeks, months, or even years.[1,4,8,11] The subtype of AMSAN has an advanced, rapid clinical course. Patients often have a poor prognosis, a high death rate, and a potentially lengthy recovery time.[12] In this case, the patient received

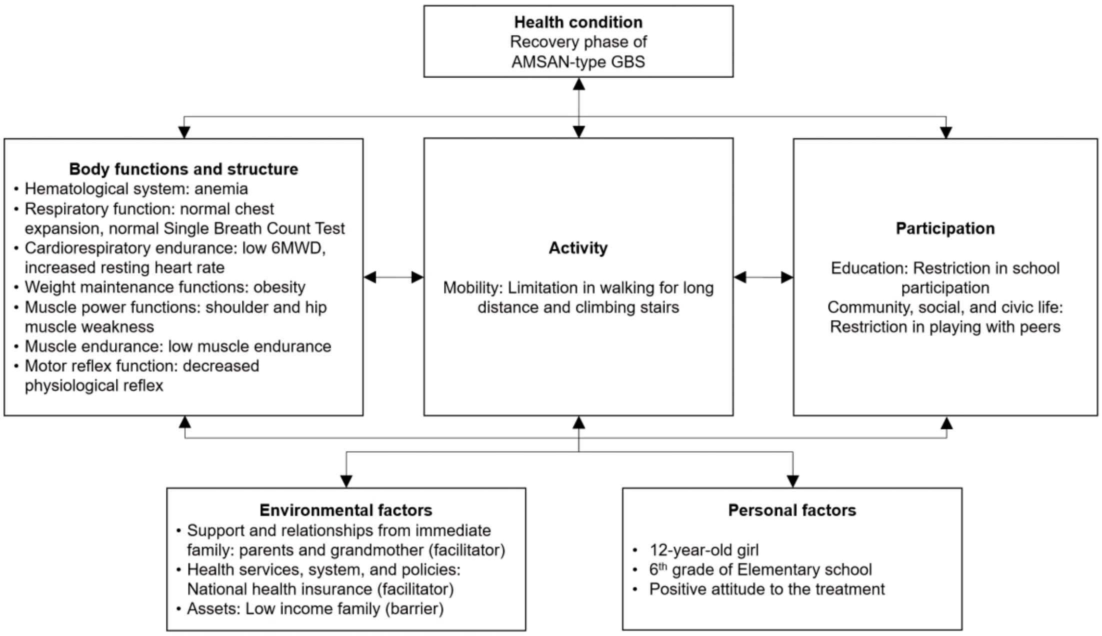

Figure 1. The International Classification of Functioning, Disability, and Health (ICF) (AMSAN = acute motor sensory axonal neuropathy, GBS = Guillain-Barré Syndrome)

five regimens of plasmapheresis treatment, which were followed by gradually improved muscle weakness. It has been demonstrated that intravenous immunoglobulin and plasma exchange are beneficial for improving illness outcomes by speeding up the recovery process. Four sessions of plasma exchange have been demonstrated to be successful; however, five sessions are more common in clinical practice.[13]

The prognosis of GBS is typically good, with 70% of patients regaining functional status, similar to the premorbid condition. However, about 20% of children have remaining disabilities after GBS.[1–3] Long-term residual complaints can include neuropathic pain, weakness, and fatigue.[1,7] The patient in this case was also complaining of tiredness at the first outpatient visit. It was the most disabling problem at that time because it limited the ability to walk more than 100 meters. It also restricted her from going to school because it was difficult for her to walk for about 1 km to reach the school and climb the stairs to her classroom.

Managing rehabilitation programs with a rehabilitation specialist and the team is a crucial step to improve the functional problems.[7,14] The goal is to optimize the activities of daily living.[7,14,15] A review study concluded that there is good evidence to support outpatient rehabilitation management in patients with GBS in the later stages of recovery to achieve long-term improvements in the levels of activity and participation.[15] The rehabilitation program in this case was arranged based on a comprehensive assessment of all the medical and non-medical factors, that can contribute to the residual disabilities.

The International Classification of Functioning, Disability, and Health (ICF) was used as a framework to describe the relationship between those factors as we can see in the Figure 1. The functional problems of this patient in terms of activity level were limitations in long-distance walking and climbing stairs. This activity limitation restricted her participation in school and playing with peers. All the functional components, including impairments in body function and structures, environmental factors, and personal factors listed on the ICF chart, can contribute to the activity and participation level.

Based on the analysis of the clinical findings, the complaint of tiredness was mainly caused by low cardiorespiratory endurance. Cardiorespiratory endurance or fitness refers to the ability of the heart and lungs to deliver oxygen to working muscles during continuous physical activity, which is an important indicator of physical health.[16–18] Good cardiorespiratory fitness causes an increase in the ability to work with higher intensity with a longer time to achieve fatigue. When the fitness level is good and the recovery is fast, it is expected that the work does not cause excessive fatigue.[18]

The gold standard for CRF evaluation is the maximum oxygen uptake (VO2max) obtained from cardiorespiratory exercise testing (CPET). In low-resource environments, submaximal and field exercise tests could be implemented to estimate the VO2max.[1,2]

Dourado The six-minute walk test (6MWT) has been validated in several populations,[9,19,20] and the distance has been proven to adequately predict the VO2max obtained in the laboratory.[21–23] In addition, this test is more representative of activities of daily living than other walking tests.[21,24] Dourado Based on the reference value chart for the six-minutes walking distance (6-MWD) in Indonesian Children, the distance that can be achieved by the patient in this case was below the 5th percentile in the initial evaluation. After the rehabilitation programs, the 6-MWD increased to be between 5th and 50th percentile.[25] We also measured the VO2max prediction and the METs level based on the 6-MWD with the VO2max prediction equation from a study.[26] The level of the METs in the initial evaluation was very low; that was not enough to perform light activities such as walking with a slow speed.[27]

A study reports the presence of significant respiratory insufficiency both clinically and qualitatively during the subacute phase of GBS.[28,29] The weakness of inspiratory and expiratory muscles in GBS can lead to poor lung compliance. This can also be compounded by immobilization; intensive unit stays, and mechanical ventilation. A single breath count test (SCBT) is a simple clinical parameter to monitor lung function.[28–30] Chest expansion has been found to be an easily measurable variable that is statistically related to restrictive abnormalities on spirometry.[28,29] However, in this case, the SCBT[30] and chest expansion[31] results were still within the normal limit. It was supported by the normal spirometry test. Thus, the respiratory impairment in this patient may not be the main factor in the low cardiorespiratory endurance. It may be more related to muscle weakness, anemia, and immobilization.

Rehabilitation programs for GBS patients should aim to restore motor function and physical condition as optimally as possible to pre-disease conditions.[14] Early rehabilitation interventions for patients with GBS, including exercise programs, have been shown to decrease chronic fatigue symptoms, improve physical fitness, walking ability, and independence in activities of daily living. As the patient got improved in sitting and standing ability, the exercise prescription focused on regaining muscular strength through aerobic exercise, resistive exercise, and the practice of functional activities. However, the intensity of exercise must be closely monitored as overwork can cause fatigue and possibly slow progress.[1,14] In addition, heart rate response to exercise in anemic children was reported less than the healthy children, because of the decrease of cardiac reserve.[32] A graded, supervised exercise program has been shown to be useful in increasing endurance.[14] In physical conditioning patients, low-intensity aerobic activity can be secure, and evidence suggests that walking and cycling may have clinically significant advantages.[1,13,14] In this case, the patient received aerobic exercise, which was gradually increased from mild to moderate intensity according to the improvement of the patient's cardiorespiratory endurance level based on the 6MWT result. The patient also received upper and lower extremity strengthening exercises to increase muscle strength. Sit to stand training was also done as a daily exercise at home. It aims to establish coordination of movements between the trunk and lower limbs and improve lower extremity muscle strength.[8]

After the rehabilitation programs, the improvement of the patient's functional condition can be observed as the level of cardiorespiratory endurance increases. Increased cardiorespiratory endurance means the increase of the ability of the heart, lung, and blood vessels to more efficiently supply the metabolic demand both at rest and during activities.[18] This can be seen from the decline in resting heart rate and RPE scale on the follow-up evaluation. At the initial evaluation, the endurance level was 0,8 METs, which was increased to 4,25 METs after two months and then 5,1 METs after three months of rehabilitation programs. Based on the METs compendium in pediatric patients, the METs level between 3 and 6 was enough to perform walking either at normal or brisk walking speed and climbing up three staircases. At the last follow-up, the patient can return to school participation and leisure activities as pre-disease condition.

CONCLUSION

Rehabilitation management, consisting of comprehensive assessments and interventions, could help children in the recovery phase of GBS to increase the cardiorespiratory endurance and regain their activity and participation.

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