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Therapeutic Rehabilitation Program and Appreciating the Academic Performance of a Child with Marfan Syndrome at Inclusive School Settings

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ABSTRACT

Introduction: Marfan syndrome (MFS) is an unusual, severe, pervasive, and life-threatening condition that demands the best multidisciplinary interventions to enhance quality of life and normalize prognosis.

Objectives: The study aimed to concentrate the multidisciplinary rehabilitation program and appreciate the academic performance of a child with Marfan syndrome studying in an inclusive education system.

Case Description: The reported case for the study was a six-year-old boy recruited from an inclusive school of CRP.

Method and Material: A single subject with a repeated-measures design was utilized to measure the academic performance and physical status of the case. Purposive sampling technique was applied with medically diagnosed with MFS.

Intervention: The research was carried out between January 2023 and April 2023. The subject has participated in a multidisciplinary rehabilitation program of physiotherapy, speech therapy, occupational therapy, orthotics services, and special education. The Pediatric Functional Independence Measure (WeeFIM) and Individual Education Plan (IEP) were used in this study.

Results: After four months of evaluation, WeeFIM scores have found 33 where baseline assessment scores were 26. The cognition and self-care dimensions of WeeFIM have shown significant improvement. After schooling, surprisingly, his academic skills, communication, and participation have improved.

Conclusion: The study demonstrated that children with MFS had successfully improved communication skills, cognitive level, and academic performance. Early diagnosis of children with MFS is vital and crucial to executing a suitable early intervention. Multidisciplinary rehabilitation programs in inclusive school settings can be beneficial for dealing with rare hypermobile cases like MFS.

Keywords: Marfan Syndrome; Inclusive Education; Multidisciplinary Rehabilitation

Abbreviations: IEP: Individual Education Plan; CTEV: Congenital Talipes Equinovarus; PA: Physical Activity; IEP: Individual Education Plan; IDEA: Individuals with Disabilities Education Act

Introduction

Marfan syndrome is a hereditary condition that impacts connective tissue through the joints, bones, heart, and eyes [1]. The physical symptoms of this condition were first presented in 1896 by Marfan [2]. MFS appears in 1:5000 individuals [3]. It affects people of all racial and ethnic backgrounds and both sexes equally [4,5]. The fibrillin 1 (FBN1) gene has a pathogenic mutation that causes MFS, an autoimmune condition that is inherited in an autosomal dominant manner [5,6]. Joint hypermobility is one of the most prevalent musculoskeletal characteristics and affects 85% of kids [7]. Scoliosis is another prevalent condition that impacts roughly 60% of children with Marfan syndrome [8]. Hypermobility is driven by ligamentous laxity, which makes them more prone to pain, poor control of the joint, recurrent strains and sprains, and more likely to suffer from dislocation and subluxation. The Marfan populace is most typically affected in the finger, patellofemoral joints, and ankle joints [7]. Marfan syndrome primarily affects the circulatory, skeletal, and ocular systems [9]. The clinical identification of Marfan syndrome is determined mostly on the observation of particular physical symptoms and family history [10]. The Ghent criteria are determined by factors such as genetic history, organ system involvement (mainly skeletal, cardiovascular, and ocular), and the severity of the clinical symptom [11]. The study aimed at focusing the therapeutic rehabilitation program and appreciating the academic performance of a child with Marfan syndrome studying in an inclusive education system.

Case Presentation

The reported case for the study was a 6-year-old boy born with MFS. It was the mother's first pregnancy as well as her first delivery. The mother didn't have any significant illnesses or challenges during her pregnancy. The parents have consanguinity. The subject has another sibling, aged 1.6 years, who is also diagnosed with MFS. The baby was delivered at the hospital by cesarean section. After birth, the baby had to stay in the ICU for a few critical conditions, especially neonatal jaundice, iron deficiency anemia, protein energy malnutrition, a decrease in calcium level, an insufficient level of vitamin D, and weakness of both upper and lower extremities. At the age of 5 days, the baby was diagnosed with MFS. After birth, he was tested with an eco-color dopler, which identified a congenital heart defect. At the age of 15 months, another color Doppler report showed everything within the normal limit. USG of the brain was done at the age of 15 months, where both the lateral and 3rd ventricles were mildly dilated, showing triventriculo-megaly (mild). USG of the inguino-scrotal region showed undescended and smaller-sized right testis, whereas left testis are not seen in the inguino-scrotal region. On pelvic x-ray, we found upward and lateral dislocations of both femoral heads with increased joint spaces. Impression showed bilateral congenital hip joint dislocation (DDH), left > right. The cytogenetic report showed an apparently normal male karyotype.

At the time of birth, the patient had limited joint ROM at the cervical spine, both upper and lower limbs, and bilateral Congenital Talipes Equinovarus (CTEV). Both wrist, hip, and knee joints were in a flexed position. The child's ankle joint was in an inverted and plantar-flexed position. The child was not able to sit properly except for modified cross-sitting, stand, or walk due to deformities. The patient had no history of operations or serial plaster casts for this deformed condition. His vision, hearing, and cognitive status were good. He was a non-residential student and attended an inclusive school where he participated in a regular special education program. He had weakness of the upper and lower limbs, hypotonicity, poor transitional and bed motilities, slurred speech, and ADL difficulties. During the course of the study, the child used a modified special wheelchair as an assistive device, though he needed total help to propel the wheelchair. He took routine-based individual physiotherapy, occupational therapy, and speech therapy. Physiotherapy treatment was directed towards maintaining range of motion in all joints and strengthening weak muscles. Speech and language therapy was given to improve oral muscle movement, tone and volume, breath support and phonation, and speech intelligibility. Occupational therapy was provided to help with the activities of daily living and further plan the vocational program according to the conditions. His delayed gross motor skills caused challenges with daily tasks and social interaction. At the age of 5 years, he was admitted to an inclusive school for improving interaction, communication, academic knowledge, and therapy interventions, as inclusive schools provide all supportive services for children with disabilities.

Ethical Approval

A subject for this case study was recruited from an inclusive school. Approval was obtained from the selected school and from the parent's willingness to participate in this study. The child and his mother agreed to participate in this observational case report study for 4 months. His mother also consented for his child and photograph to be published; his face is blocked out for anonymity.

Method and Material

The study was conducted in the inclusive school named "William & Marie Taylor School" at CRP. A single subject with a repeated-measures design was utilized to measure the academic performance and physical status of a student attending an inclusive school. Purposive sampling technique was applied to select the cases that matched the criteria, especially those that were medically diagnosed as MFS. The research was carried out between January 2023 and April 2023. The study was started after the acceptance of the protocol, following which data collection was started after getting consent, and the study period was over after the submission of the final report.

Physical Rehabilitation

Marfan syndrome (MFS) is an unusual, severe, pervasive, and life-threatening condition that demands the best multidisciplinary

interventions to enhance quality of life and normalize prognosis [12]. Exercises have been demonstrated to be treatment alternatives in MFS from a variety of perspectives, including both safety and efficacy [4]. Physical activity (PA) is still not generally advised for MFS patients. The study encourages extremely low intensities of PA for MFS [13]. The regimen of mild PA appears to have beneficial effects. Cardiorespiratory and muscular endurance are improved as a result of physical exercise [14]. The MFS must continue to be active in order to maintain their physical health. The purpose of exercise is to maintain muscle mass to enhance joint stability, improve cardiovascular fitness to lower blood pressure, and reduce pain. Every patient will require a unique strategy that takes into consideration their symptoms, especially those impacting the musculoskeletal, cardiovascular, and ophthalmic systems [15]. Individuals with MFS are encouraged to initiate regular physical exercise in order to preserve optimal wellness and health. Finding activities and sports that are enjoyable, safe, and improve fitness is challenging. Exercise should always be with little resistance and low impact and focus more on endurance than increasing muscle strength. Swimming, Tai Chi, dance, and Pilates are all recommended for water-based exercise [16].

Stretching is discouraged because of the awareness that one can overestimate and harm the joints. Stretching out gently has many advantages and is not always to be ignored. In general, splints and braces offer more efficient and effective support. Improvements in proprioception and injury prevention have been associated with the use of tape on skin surfaces to support joints while exercising [17]. Exercises that involve a closed kinetic chain are the most beneficial in this regard. Additionally, open-chain exercises can be included later to make them more practical [18]. Orthotics and functional foot insoles may be necessary for some people as supplementary support. The recovery of typical movement may benefit from gentle manual exercise. Programs for joint stability and motor control can improve proprioception and lead to efficient muscle contraction around joints [17] (Figures 1 & 2).

Figure 1: Providing rehabilitation services.

Figure 2: Physical structure and school performances.

Discussion

According to empirical findings, multidisciplinary rehabilitation care significantly enhances rehabilitation achievements for serious and rare cases like Marfan syndrome. The purpose of the study was to evaluate the academic performance and physical status of a child with MFS over four months after receiving inclusive education and multidisciplinary therapeutic rehabilitation. The Paediatric Functional Independence Measure (WeeFIM) tool was developed to assess children's functional abilities in the tasks of daily living. The tool assesses the child's level of functional autonomy, which is described as their regular and consistent performance, as well as the amount of support they require to successfully complete routine tasks [19]. It consists of 18 items that address the three key categories: eight are for self-care, five are for mobility, and five are for cognition. Each item is given a rating from 1, which requires complete reliance, to 7, which means full independence [20]. The WeeFIM tool is reliable for both children with and without disabilities [21].

After four months of evaluation, WeeFIM scores have reached 33, where baseline assessment scores were 26. The cognition and self-care dimensions of WeeFIM have found significant outcomes after four months of evaluation. In dimension cognition, we got scores of 13 at the initial assessment, and we got scores of 18 after four months of observation. In the dimension of self-care, we got scores of 8 at the initial assessment, and we got scores of 10 after four months of observation. In dimension transfer, we found no significant changes after the initial assessment (Table 1). The Individual Education Plan (IEP) refers to a written agreement specifically designed for pupils with disabilities. It includes the resources and facilities required to meet these objectives, as well as the educational goals for each student [22]. It serves as a management instrument for the method of learning and teaching and for assessing the progress of students [23]. It is crucial as a tool for managing and observing the entire learning and teaching process [22]. According to the Individuals with Disabilities Education Act (IDEA), every kid in the United States who must have an IEP receives special education programs [24]. The researcher applied IEP to measure the school performance of the children with MFS.

Table 1: Evaluation scores of the Pediatrics Functional Independence Measure (WeeFIM).

Dimension	Initial Assessment	After 4 Months
Self-care	05	07
Sphincter control	03	03
Self-care subtotal	08	10
Mobility-transfer	03	03
Locomotion	02	02
Mobility subtotal	05	05
Communication	06	09
Social cognitive	07	09
Cognition subtotal	13	18
WeeFIM Total	26	33

The kids with Marfan syndrome were assisted by the principal, general educators, special educators, counselors of the school, nutritionists, and rehabilitation specialists within the institution by offering academic and psychological counseling. The case demanded modifications in the classroom setting for the purpose of facilitating learning. Due to their close vision, teachers presented a comprehensive visual representation of the content. Hypermobility of the fingers and wrists was needed adjustments for children with poor writing skills. Based on the child's restricted abilities and the severity of the symptoms, the school therapist panelists developed and used a number of exercise cures. To prevent the youngster from falling behind academically, the special teachers interacted closely with the kid and the kid's parents. One study found that 17% of MFS patients had attention deficit issues and 13% had learning disabilities. The study predicted that hypermobility of the fingers and wrists may be the cause of the lower IQ ratings [25]. But after schooling here, surprisingly, his academic skills, communication, and participation have improved through multidisciplinary rehabilitation approaches, including special education, physical therapy, speech therapy, and occupational therapy services.

Conclusion

The study demonstrated that children with MFS had successfully improved communication skills, cognitive level, and academic performance. This study demonstrated that IEPs may be created and implemented successfully for CWD. Early diagnosis of children with MFS is vital and crucial to executing a suitable early intervention. It is possible to reduce significant morbidity and lengthen the life expectancies of individuals with MFS through regular follow-up checks. The results of this study might serve as a direction for a more successful application of IEPs in Bangladesh, especially in government primary schools. The research indicated that multidisciplinary rehabilitation programs in inclusive school settings can be beneficial for dealing with rare hypermobile cases like MFS.

Conflict of Interest

The authors disclose no conflict of interest relevant to this publication.

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Author Contribution

All authors contributed to preparing the write-up, revising the article, and giving final approval of the version to be published. The authors consent to accept responsibility for the contents of the tasks.

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