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CASE REPORT

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DETERMINING THE OPTIMAL EXTERNAL WEIGHT FOR AN INDIVIDUAL TREATMENT PROGRAM IN AN ADOLESCENT WITH FRIEDREICH ATAXIA: A CASE REPORT

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ABSTRACT

Purpose: This study investigated the amount of external weight to optimize postural control, gait, and balance parameters in an adolescent with Friedreich ataxia (FRDA).
Method: The study included a 16-year-old adolescent (height: 1.57 m and weight: 60 kg) with FRDA. The adolescent's postural control, gait, and balance were evaluated by attaching different weights (unweight, 5%, 10%, 15%, and 20% of the body weight (BW)) to the trunk. The Bertec Balance Check Screener™, GAITRite and Timed up and Go (TUG) were used to assess postural control (postural sway, limits of stability), gait, and dynamic balance, consecutively.
Results: Regarding limits of stability, the adolescent with FRDA performed best at 10% of BW in the anterio-posterior directions and at 15% of BW in the lateral directions. In the antero-posterior directions, the postural sway range on the normal surface was the lowest in unweighted and the lowest at 15% of BW on the perturbed surface. In the lateral directions, she performed the minimum postural sway range at 10% of BW on both the normal and perturbed surfaces. She had a higher cycle time, stride length and narrower support surface at 20% of BW. The value of TUG was the lowest at 20% of BW (10.44 sec).
Conclusion: These findings may contribute to the literature by revealing the need for individual determination of the optimal amount in external weighting applications in FRDA.
Keywords: Friedreich Ataxia, Gait, Postural Balance, Body Weight

ÖZET

Amaç: Bu çalışma Friedreich Ataksi (FRDA)'li bir adölesanda postüral kontrol, yürüyüş ve denge parametrelerini optimize etmek için eksternal ağırlık miktarını araştırdı.
Yöntem: 16 yaşındaki bir FRDA'lı adölesan (boyu:1,57 m ve kilosu:60 kg) bu çalışmaya dahil edildi. Adölesanın postüral kontrol, yürüyüş ve dengesi gövdeye bağlanan farklı ağırlıklarla (ağırlıksız, vücut ağırlığının %5'i, %10'u, %15'i ve %20'si) değerlendirildi. The Bertec Balance Check Screener™, GAITRite ve Zamanlı Kalk Yürü (TUG) testleri sırasıyla postüral kontrolü, yürüyüşü ve dinamik dengeyi değerlendirmek için kullanıldı.
Bulgular: Stabilite limiti açısından FRDA'lı adölesan anterio-posterior yönlerde vücut ağırlığının %10'unda ve lateral yönde vücut ağırlığının %15'inde en iyi performansı gösterdi. Anterio-posterior yönlerde, normal yüzeyde postüral salınım aralığı ağırlıksızda ve köpük yüzeyde vücut ağırlığının %15'inde en düşük değerdeydi. Lateral yönlerde, normal ve köpük yüzeyde adölesan vücut ağırlığının %10'unda minimum postüral salınım aralığı gösterdi. Vücut ağırlığının %20'sinde daha yüksek döngü süresi, adım uzunluğu ve daha dar destek yüzeyine sahipti. TUG değeri vücut ağırlığının %20'sinde en düşüktü.
Sonuç: Bu bulgular, FRDA'da eksternal ağırlık uygulamalarında optimal miktarın bireysel olarak belirlenmesi ihtiyacını ortaya koyarak literatüre katkı sağlayabilir.
Anahtar Kelimeler: Friedreich Ataksisi, Yürüyüş, Postüral Denge, Vücut Ağırlığı

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INTRODUCTION

Low back pain (LBP) is a highly prevalent problem, with a prevalence of 37% of the general adult population and a lifetime prevalence between 60% and 85%. It has been reported that the lumbar disc herniation is among the top three causes of LBP. (1-3). It is also emphasized that the number of spine surgeries has recently tended to increase (4). However, it is noteworthy that the incidence of failed lumbar spinal surgery ranges from 10% to 46% (5). The International Association for Pain Research defines failed back surgery syndrome (FBSS) as lumbar spine pain of unknown cause that persists despite surgical intervention, or that occurs after surgery for spine pain in the same topographic location at baseline (6). The main reasons for failing lumbar surgeries have been explained as failure to clearly identify the etiology of the patient's pain, inadequate identification of high-risk parameters for surgery, and disregard of preoperative conservative measures. (7).

There are a limited number of studies examining the effectiveness of physiotherapy and rehabilitation after failed lumbar spine surgery. We could only find one trial that included physiotherapy and rehabilitation with exercise in this group of patients (8). In our case, reportedly, our patient had 3 lumbar spinal operations and had a more complex surgical history. In addition to severe pain, our patient had severe neurological symptom such as drop foot. We believe that the more complex surgical history of our case and the high severity of the clinical symptoms increase the originality of our report.

In this case report, we presented the medical and surgical history, physiotherapy and rehabilitation program and clinical results of a patient with lumbar disc herniation who had undergone 3 spinal surgeries for LBP and developed drop foot. The effectiveness of physiotherapy and rehabilitation applied to this patient was evaluated.

METHODS

Case Description

A 16-year-old adolescent with a height of 1.57 m and a weight of 60 kg was included in this study. She was taking idebenone, B12 and vitamin D as medication. She was able to climb stairs and walk without support. In 2013, following her sister's diagnosis of FRDA, she underwent genetic testing (homozygous for an 873 GAA expansion in the FXN gene) and received the same diagnosis. In 2020, she consulted her doctor due to complaints of knee buckling and falling. She did not receive physiotherapy during the Covid-19 pandemic. After the pandemic, her balance and gait problems became apparent. She has been attending physiotherapy sessions (3x/week) for the past year due to worsening symptoms. She who is a student in high school reported difficulty moving in social settings, on narrow paths, and with fine motor skills. She reported frequent ankle sprains. She did not have any muscle shortness. Her gross muscle strength score is 4 for the lower extremity and 4+ for the upper extremity according to the modified Medical Research Council (18). She did not have sensory complaints.

Outcome Measure

The adolescent's postural control, balance and spatio-temporal parameters of gait were assessed.

Postural control was assessed using the Bertec Balance Check Screener™ (BP5046 Bertec Co., USA). Limits of stability (LoS) and postural sway range were determined for postural control. During the test, the subject was asked to stand comfortably on a 50x46 cm surface with parallel feet. LoS was assessed by maximum center of pressure displacement in the anterior-posterior (AP) and lateral directions with the heels in contact with the platform. To assess the AP/lateral sway range of the center of pressure she was asked to stand relaxed with eyes open and then to stand on a 7.5 cm high foam as a perturbed surface on the platform in the same way for 10 seconds. Both tests were evaluated three times. Higher values for LoS and lower values for sway indicate better postural control. The highest LoS value and the lowest sway value of the three assessments were recorded as cm (19).

The spatio-temporal parameters of gait were assessed using GAITRite (CIR Systems Inc., Jersey, USA) electronic walkway system. The GAITRite uses a 4.88 m mat with sensors to objectively measure spatio-temporal gait parameters of individuals. Cycle time, stride length, heel to heel base of support (BoS), velocity and cadence were recorded. Three assessments were obtained and the mean gait values were recorded (20).

Dynamic balance was assessed with the Timed Up and Go test (TUG). The adolescent sat in a chair with hip and knee flexion at 90 degrees, facing a cone 3 meters away. She was instructed to stand up, walk around the cone, return to the chair, and sit down as quickly as possible. Three assessments were recorded as seconds (21).

The assessments were performed with unweight, 5%, 10%, 15% and 20% of the participant's body weight (BW), which was determined based on different studies that used different weighting procedures, weight application methods, amounts and placement on balance and gait (22-24). The weights were placed around the patient's waist with the help of a belt and secured from her shoulders with two straps. Assessments were conducted randomly with 5-minute rest intervals to avoid fatigue.

RESULTS

The adolescent's LoS and postural sway at different BWs are shown in Table 1. In the AP direction, LoS (16.73 cm) and postural sway on the perturbed surface (1.48 cm) were better at 15% of BW, whereas postural sway on the normal surface (0.87 cm) was better in unweighted conditions. In the lateral direction, LoS (25.13 cm) and postural sway (normal surface 0.88 cm; perturbated surface 0.68 cm) were better at 10% of BW.

The adolescent's spatio-temporal gait parameters at different BWs are shown in Table 2. She had a higher cycle time (1.22 sec) and stride length (104.39 cm), narrower support surface (15.42 cm) and lower step length differential (0.68 cm) at 20% of BW.

At 20% of BW, the mean TUG duration was lowest at 10.44 seconds. The mean TUG results at different BWs are shown in Figure 1.

Table 1. The results related to postural control of case with Friedrich Ataxia

Postural Control Parameters		Body weight amount (%)				
		0	5	10	15	20
Antero-posterior	Limits of stability (cm)	16.15	13.65	15.91	**16.73**	15.19
	Postural Sway Range / NS (cm)	**0.87**	1.50	1.46	1.32	1.24
	Postural Sway Range / PS (cm)	1.76	2.00	1.68	**1.48**	2.41
Lateral	Limits of stability (cm)	25.04	19.73	**25.13**	23.89	21.84
	Postural Sway Range / NS (cm)	1.01	1.17	**0.88**	1.60	0.88
	Postural Sway Range / PS (cm)	1.07	1.25	**0.68**	1.36	1.65

NS: Normal Surface, PS: Perturbed Surface

Table 2. The results related to spatiotemporal parameters of gait of case with Friedrich Ataxia

Gait parameters	Body weight amount (%)				
	0	5	10	15	20
Cycle time (sec)	1.14	1.19	1.18	1.22	**1.22**
Stride length (cm)	90.31	91.12	101.38	93.16	**104.39**
Heel-Heel Base Support (cm)	19.85	18.55	17.58	17.95	**15.42**
Velocity (cm/sec)	79.00	75.40	85.80	76.00	84.00
Cadence (steps/min)	104.90	100.60	101.60	99.10	97.20
Step Length Differential (cm)	6.13	1.79	7.91	5.80	**0.68**

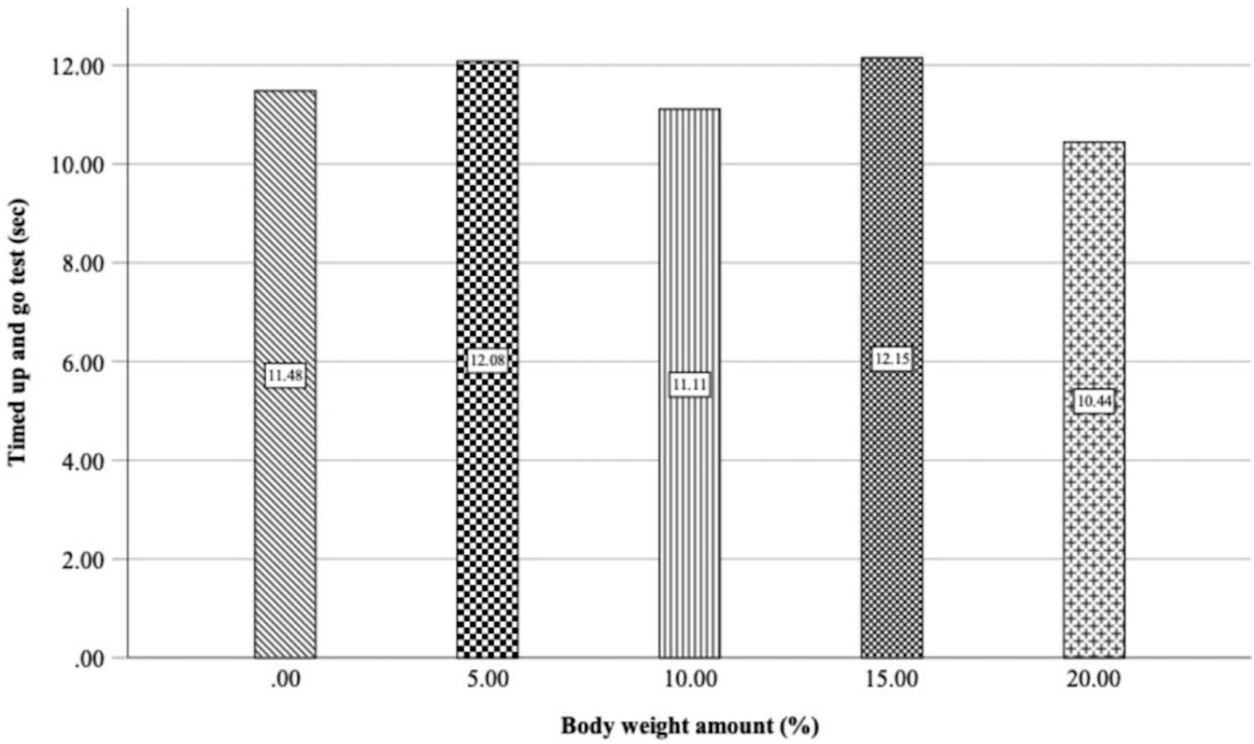

Figure 1. The mean duration of timed up and go test of adolescent with Friedrich Ataxia

DISCUSSION

In this case study to determine the optimum external weight amount to improve balance and gait parameters in FRDA, it was found that the optimal weight amount was 15% for AP balance parameters, 10% for lateral balance parameters and 20% for most of the gait parameters.

Progressive gait ataxia, characterized by impaired balance during walking, is the initial symptom in most patients, resulting from a combination of cerebellar and spinocerebellar degeneration, sensory neuropathy, and vestibular nerve involvement (7, 25). This case had decreased velocity, cadence, stride length and increased BoS compared to mean values of healthy girls aged 10-19 years at an unweighted condition (26). Moreover, her TUG time was twice as long in the unweighted condition compared to the mean values of healthy children aged 14-18 years (27). These results indicate gait and balance problems in this case.

The effects of external weights on gait have been studied in a variety of conditions (13, 14, 22). Morgan (28) observed an improvement in the gait of 11 out of 14 ataxia patients, including those with FRDA, with the use of external weights. In a study conducted in cerebellar ataxia (CA), 3 out of 5 patients showed improvement in certain gait parameters with weights applied at 10% of BW. However, throughout the study they found that changes in gait tended to worsen rather than improve (10). For most gait parameters in this case, optimal values were obtained with a 20% of BW. With external weights, an increase in velocity and stride length, along with a decrease in BoS may indicate that gait has become more stable. With a significant decrease in step length differential with 20% of BW, it can be interpreted that gait symmetry may be increasing.

Studies using strategically placed weights have demonstrated beneficial effects of external weight on balance in MS and Ataxia (12, 14-17). In this case, weight application at 15% of BW maximized AP LoS and minimized postural sway on the perturbed surface, but increased sway on the normal surface. The weight application significantly reduced the area of sway when one sensory modality was restricted, but showed no improvement without restriction in CA (12). In this case, weights applied at 10% of BW maximized lateral LoS and minimized postural sway on both surfaces. Two studies reported that external weights reduced lateral sway but had no effect on AP sway (29, 30). It can be concluded that more weight may be required to increase AP balance in patients than the weight effective for lateral sway.

The results for dynamic balance measured with TUG in this case showed that the optimal weight is the weight applied at 20% of BW. The improvement in TUG time with weight is consistent with the results of the studies conducted in MS (14, 16, 17). However, it contradicts the findings of Widener et al. that TUG times did not change with weight in ataxia patients (12). The authors suggested that different pathological processes and locations in these diseases might explain the conflicting findings. They hypothesized that increased sensory input from weights might improve coordination in ataxia, but not gait speed. Perhaps, in line with these findings, heavier weights may be needed to improve dynamic balance in individuals with CA.

The main limitation of this study is its single-case design. Another is the lack of weight application with Balance Based Torso Weighting.

CONCLUSION

In this case study, weight application at 10% of BW improved lateral stability, 15% improved AP stability and 20% improved gait stability, symmetry, and dynamic balance in an adolescent with FRDA. These findings may contribute to the literature by suggesting the need for individualized determination of the optimal amount of weighting application. There is a need for follow-up studies with more participants, different weights, and different methods.

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