CASE REPORT

Use of the Modified Functional Reach Test in a Patient with Friedreich's Ataxia

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ABSTRACT

Background: Friedreich's Ataxia (FRDA) is a progressive, multi-system degenerative disorder of the spinal cord, peripheral nerves, and cerebellum. A progressive loss of lower extremity strength and coordination leads to a loss of ambulation and an increased reliance on postural stability for wheelchair positioning and transfers. Quantifying seated balance has added importance for non-ambulatory patients. The Friedreich's Ataxia Rating Scale (FARS) is the gold-standard to measure functional disability, activities of daily living, and disease progression; however, patients in the advanced stage of the disease are unable to complete many test items. The Modified Functional Reach Test (mFRT) may provide a clinically relevant alternative. The purpose of this case report is to demonstrate the use of the mFRT to document changes in seated balance for a non-ambulatory patient with FRDA.
Case Description: The subject was a non-ambulatory, twenty-four year old male with FRDA. His primary physical therapy goal was to improve sitting balance. The FARS was administered initially and after six weeks of physical therapy service and the Modified Functional Reach Test (mFRT) was administered weekly. ***Outcomes:*** The patient required increased assistance to safely complete transfers and bed mobility. The FARS scores remained unchanged. Scores on the mFRT decreased from 30.5 cm to 5.0 cm (forward direction) and from 7.5 cm to 4.0 cm (right direction), demonstrating a quantitative change in seated balance for this non-ambulatory patient. ***Conclusion:*** Physical therapists should consider using the mFRT as a quantitative measure of seated balance in this patient population.

Background

Friedreich's Ataxia (FRDA) is the most common autosomal recessive ataxia in the United States with an estimated prevalence of 1:50,000 individuals.[10] Typical onset for this multi-system degenerative disease is as early as five years old. A loss of frataxin, a protein critical for mitochondrial homeostasis, results in cell death in the dorsal root ganglia, posterior columns, and spinocerebellar tracts. As the disease progresses, the corticospinal tracts are also affected. Patients first experience gait ataxia, followed by progressive symmetrical muscle weakness in the proximal extremities, tremors, and impaired coordination. As the disease progresses, affected individuals loose the ability to independently ambulate. At this advanced stage of the disease, weakness spreads to the trunk and affects seated balance.

Seated balance is defined as "the ability of a person to maintain control over upright posture during forward reach without stabilization.[8]" For individuals who are non-ambulatory, seated balance is critical in maintaining independence during functional activities.[5,6] This is particularly true in the advanced stages of FRDA, as an increased reliance on seated postural stability is necessary for proper wheelchair positioning, transfers, and bed mobility.[9] In the absence

of functional seated balance, an individual's independence is severely limited.

At this time, clinically relevant outcome measures of seated balance designed for individuals with advanced FRDA are lacking. Balance measures for individuals that can stand are not valid for individuals that are non-ambulatory and drive power wheelchairs from the seated position.[4,5,11,16] This information is vital for therapists to document this impairment that has the potential to result in significant functional changes and to develop appropriate interventions. Without such documentation, clinicians struggle to not only advocate for the necessity of services, but also to determine the effectiveness of their treatment plan.[3]

The two most commonly selected scales currently utilized to measure progression and severity of FRDA include the Friedreich's Ataxia Rating Scale (FARS) and the International Cooperative Ataxia Rating Scale (ICARS). When compared in a recent study by Fahey et al[3], the authors concluded that the FARS was the superior measure, as it had a higher effect size. Although the Friedreich's Ataxia Rating Scale (FARS) is considered the gold-standard to measure functional disability, activities of daily living scores, and disease progression in patients with FRDA, the test itself is problematic.[3,7,9,15] In addition to being lengthy to administer, the FARS is difficult for patients in the advanced stage of the disease who are non-ambulatory to complete.[1] This is particularly true in regards to the "Upright Stability" section, as six of the seven tasks are to be completed in standing. With significant strength and coordination deficits, these patients are often unable to complete many items that comprise the test and are automatically scored at the basal level. Please refer to Table 1 for more information about the tasks that comprise the "Upright Stability" section.

In light of these limitations, clinicians continue to search for alternative meaningful and objective outcome measures to document seated balance. Research suggests additional seated balance assessment tools are available; however, few have undergone validation for specific populations.[14] Lynch et al[7] demonstrated that in comparison to the FARS, single performance measures were valid assessment tools of FRDA progression and correlated significantly with functional disability and ADL scores. Another tool that measures seated balance, the Modified Functional Reach Test (mFRT), has been found to be a reliable measure of sitting balance in a variety of conditions including ataxia, sub-acute post-stroke and non-ambulatory patients with SCI's. [6,8,13] This tool may provide a clinically relevant alternative to quantify seated balance.

The Modified Functional Reach Test (mFRT) is adapted from the Functional Reach Test for individuals that are unable to stand. This tool assesses an individual's stability in the seated position by measuring the maximal distance he/she can reach forward. The individual's stability can also be assessed by reaching to the right and left. Normative data for scores on the mFRT has been established for patients with acute stroke.[6] This functional outcome measure is quick to administer in the clinical setting, taking less than five minutes, and cost effective, as the only material needed is either a tape measure or yardstick.[8] The mFRT is reported to be a reliable measure of seated balance in non-ambulatory patients with spinal cord injuries, ataxia, and in the sub-acute phase of stroke rehabilitation.[6,8,13] For this reason, the mFRT was selected in this study as a possible alternative to the FARS for a non-ambulatory patient with FRDA.

The purpose of this case report is to demonstrate the use of the mFRT to document changes in seated balance for a non-ambulatory patient with FRDA.

Case Description

Prior to initiation of the study, the patient signed a written document providing consent for inclusion in the study.

Patient Description

The patient is a twenty-four year old male diagnosed with FRDA at age twelve. The patient is independent in power wheelchair for mobility and wears bilateral ankle-foot orthotics to maintain lower extremity alignment. He resides in a fully accessible, one-bedroom apartment attached to his mother's home with twenty hours of aide support each week to assist in ADLs. Pertinent past medical history includes spinal fusion surgery at age seventeen secondary to scoliosis and Botox injections to bilateral hamstrings and ankle plantar flexors to improve lower extremity positioning. The patient takes Dantrium daily to reduce muscle spasticity. At the time of the case report, he was attending physical therapy two times per week to improve sitting balance, independence in all transfers, and maintain lower extremity range of motion for positioning.

Examination

A systems review revealed that the patient was alert and oriented with intact integumentary system. The patient reported episodes of dizziness, lightheadedness and palpitations and a cardiology evaluation was recommended. The extent of cardiovascular impairment was unknown due to inconsistent patient reports and poor attendance at cardiology appointments. The patient demonstrated bilateral upper and lower extremity ROM within functional limits with the exception of ankle dorsiflexion (L -30°, R -40°), knee extension (L -8°, R -9°) and hamstring length measured by the straight leg raise test (L 80°, R 75°). Upper extremity MMT scores were 4-/5 or greater at all joints. Bilateral lower extremity MMT scores were 2-/5 except for knee extension (4/5) and hip flexion (3/5).

The Friedreich's Ataxia Rating Scale (FARS) was selected due to prior research findings claiming it to be the superior functional rating scale for patients with FRDA.[1,3,7,15] It also included an "Upright Stability" subsection, which provided objective information on seated balance. Refer to Figure 1 for FARS scores at evaluation and after the six-week intervention program.

The Modified Functional Reach Test (mFRT) was selected as an alternative objective measure to document changes in this patient's seated balance. It allowed the examiner to gather information about the limits of seated balance in the forward and lateral directions. The patient was seated in his wheelchair against the back of the chair and his hips and knees were positioned at ninety degrees of flexion. A yardstick was mounted on the wall at shoulder height. His shoulder was flexed to ninety degrees with elbow extended and he was asked to lean forward as far as possible without rotating or touching the wall. After a practice trial, the distance of the forward lean was recorded at the distal end of the third metacarpal in centimeters. This was repeated two times and an average was calculated. The same procedure was repeated to record the distance of lateral reach to the right and left. Please refer to Table 2 for mFRT scores. Each measurement was obtained at the beginning of each session to prevent fatigue from therapeutic exercise affecting scores.

Evaluation

The patient presented with decreased seated balance and ataxia limiting his ability to

transfer and sit independently secondary to medical diagnosis of Friedreich's Ataxia. The American Physical Therapy Association (APTA) Guide to Physical Therapy practice pattern for this patient is 5E: Impaired Motor Function and Sensory Integrity Associated With Progressive Disorders of the Central Nervous System. The patient's prognosis was poor secondary to advanced disease progression and lack of motivation[2].

Description of Intervention

The patient was seen twice a week for six weeks in an outpatient physical therapy clinic. Due to the patient's poor cardiovascular and muscular endurance, sessions were limited to thirty minutes to prevent extreme fatigue. A variety of interventions were performed.[2,9,10,12] Emphasis was placed on seated balance exercises at the edge of the mat table to improve seated balance and core strength necessary for transfers and bed mobility. To ensure the patient's safety in the presence of poor seated balance, the physical therapist was positioned in tall kneeling behind the patient with her hands positioned on the anterior aspect of respective shoulders at all times. Examples of these seated balance exercises included: static sitting with and without the use of upper extremity support, reaching for cones outside of the patient's base of support, partner ball toss using balls of varying sizes and dynamic sitting on foam with upper extremity support. Each seated balance exercise was performed for an average of two to three minutes with thirty to sixty second rest breaks in between.

Additional exercises not specifically focusing on seated balance were also included in each session to assist in maintaining the patient's current functional mobility and comfort level. PROM was provided to bilateral hips, knees and ankles in order to maintain ROM for wheelchair positioning and comfort. Strengthening exercises including quadruped positioning, shoulder flexion with ten pounds, chest press with ten pounds and wheelchair push-ups were also included to maintain upper body strength for transfers. Lastly, the arm ergometer was used to work on improving cardiovascular endurance. All therapeutic exercises were progressed per patient tolerance.

Outcomes

The patient participated in ten therapy sessions over the course of the six-week intervention program. During this time, the patient required increased physical and verbal assistance during sessions to maintain proper form and encourage participation. A decline was also apparent during seated mat table exercises. During initial evaluation, the patient sat at the edge of the mat table with minimum/moderate assistance. After six weeks, he required maximal assistance to maintain the upright, seated position. This decline was also evident during wheelchair transfers, as an increase from minimal to moderate/maximal assistance was needed.

Despite the observable changes in the patient's functional mobility and seated balance, FARS scores remained unchanged over the six-week period of time (Figure 1). In contrast, scores on the mFRT decreased from 30.5 cm to 5.0 cm the in forward direction and from 7.5 cm to 4.0 cm to the right. The greatest change was noted in the forward direction. The patient's lateral reach to the left was inconsistent and demonstrated improvement from the initial 5.75 cm reach to 6.5 cm at the end of the six-week period. Please refer to Table 2 for more detailed scores. The mFRT provided quantitative documentation of the patient's decreased seated balance, something the FARS was unable to capture. This intensity and combination of specific therapeutic exercises appear be ineffective to maintain and/or improve seated balance for this patient.

Table 1: Items comprising the FARS Upright Stability Subsections

1. Sitting Posture
2. Stance-Feet Apart*
3. Stance-Feet Together*
4. Tandem Stance*
5. Stance on Dominant Foot*
6. Tandem Walk*
7. Gait*

**Denotes task completed in standing*

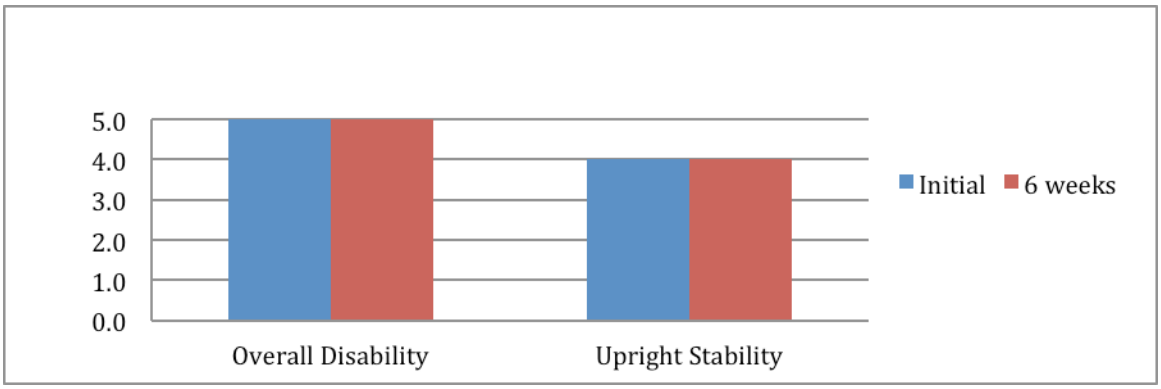
Table 2: Results of the Modified Functional Reach Test throughout the 6 week intervention program §

	Forward*	**Right***	**Left***	**Comments**
Week 1	30.5	7.5	5.75	"Less tired" today
Week 2	16	5	9	Tired from boxing
Week 3	10.5	5.5	6.5	"Extremely tired" today
Week 4	8	5.5	9.5	Up late at graduation party night before
Week 5	NT	NT	NT	Session cancelled by patient
Week 6	5	4	6.5	Trip to Philadelphia, late night

§ *Test performed in wheelchair without armrests*

** Measurements recorded in centimeters.*

Figure 1: Comparison of change in FARS Scores (initial to final scores after six week intervention program)



Discussion

The purpose of this case report was to demonstrate the use of the mFRT to document changes in seated balance for a non-ambulatory patient with FRDA. Unlike scores of the FARS that remained unchanged, scores on the mFRT in the forward and right directions progressively decreased from week to week. This finding is consistent with the increased levels of assistance noted during functional tasks and seated balance exercises. Scores of the mFRT did increase to the left over the six-week time period, which could be attributed to the patient's participation in sled hockey. To date, no literature has been identified that has compared the results from the Upright Stability Subsection of FARS to the mFRT. The information from this case suggests that for this patient, the mFRT was a quantitative means of measuring changes in seated balance.

There are many ways in which a more sensitive measure of seated balance could assist clinicians in the treatment in patients with advanced FRDA, as small changes have a large impact on the functional abilities of a person who is non-ambulatory.[5] A recent study by Maring et al[10] found limited insurance coverage and a lack of access to services to be the most common external barriers to services in people with FRDA. An outcome measure that has the ability to detect small changes in seated balance would help provide clinicians a more accurate means to document changes in functional level. This more detailed and quantitative information may help advocate for the necessity of continued services to external payers. Furthermore, such measures could aid in treatment plan adjustments and durable medical equipment (DME) purchases and modifications. For this patient, the information served as an objective measure of daily documentation; however, results could support recommendations for continued therapy, prioritization of services in the plan of care, and the addition of a chest strap to the patient's wheelchair for increased safety.

Given that a case report lacks the control of a research study, possible alternatives may explain the outcomes in this case. In addition to the increased assistance required during seated exercises and transfers, additional factors may have negatively influenced mFRT scores. The patient's decline could be related to a lack of motivation to put forth his best effort, fatigue from previous daily activity, or the psychological effect associated with having a degenerative disorder. A valid measurement of a patient's level of fatigue could have been administered prior to the mFRT to rule out fatigue as a cause of the recorded scores. The addition of this information would have been an excellent way to validate his complaints; however, no measure of fatigue was administered with this patient. Therapists are encouraged to consider collecting this information to further understand the role of fatigue in this population.

Additionally, this intensity and combination of specific therapeutic exercises appear to be ineffective to maintain and/or improve seated balance for this individual. This case report was limited to six weeks in duration, which may not be enough time to see improvements in strength and motor control. Future research should investigate if different combinations of therapeutic activities as well as varying intensity of service are a more effective way to improve functional seated balance. These limitations provide an area in which future research is needed, as the psychology associated with having a degenerative disorder and its associated effects on physical performance were not explored in this case report.

The value in this case report lies in the fact that the mFRT is a clinically feasible outcome measure to administer to patients in the advanced stage of this disease who are non-ambulatory. The mFRT is cost effective, quick and easy to administer, and uses simple instructions to ensure patient understanding.

Conclusion

Friedreich's Ataxia is a condition that a physical therapist may encounter few times in a career. The pathophysiology of this condition is predictable; however, due to the small number of patients with this condition, randomized controlled trials are not available to offer guidance regarding best evaluation and treatment methods. This case report helps to fill this gap by providing a description of the use of one outcome measure to document changes in seated balance for this unique population.

Physical therapists should consider using the mFRT as an objective measure of seated balance. Despite the absence of a definitive cause-and-effect relationship, the mFRT demonstrated quantitative changes in seated balance for this non-ambulatory patient. Easy to administer and time efficient, the mFRT could serve as an effective method of documenting changes in seated balance for patients with advanced Friedreich's Ataxia. This information may help therapists in their decision-making regarding treatment planning and durable medical equipment needs for individuals with advanced Friedreich's Ataxia.

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