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Research Article

Dynamic Balance in Spinal and Bulbar Muscular Atrophy: Relationship between Strength and Performance of Forward Lunge Step Up and Over and Step Quick Turn

Joseph A. Shrader [1] Ashwini Sansare [1] Vincent Shieh [1] Joshua G. Woolstenhulme [1] Julie Rekant [1] Rafael Jiménez-Silva [1] Galen O. Joe [1] Angela Kokkinis [2] Kenneth H. Fischbeck [2] Christopher Grunseich [2] and Cristiane Zampieri[1]

[1]*Rehabilitation Medicine Department, Clinical Center, National Institutes of Health, Bethesda, MD, USA*
[2]*Neurogenetics Branch, National Institute of Neurological Disorders and Stroke, NIH, Bethesda, MD, USA*

Correspondence should be addressed to Joseph A. Shrader; jshrader@cc.nih.gov

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Introduction. Spinal and bulbar muscular atrophy (SBMA) is a neuromuscular disorder that leads to progressive weakness of bulbar and extremity muscles. Dynamic balance during functional tasks has not been reported in people with SBMA. *Objectives*. (1) To evaluate the ability to safely complete a forward lunge (FL), step quick turn (SQT), and step up and over (SUO), (2) to determine the presence and severity of dynamic balance impairments by comparing performance to normative data, and (3) to investigate the relationship between lower extremity strength and ability to complete each task. *Design*. Cross-sectional analysis. *Participants*. Fifty-three people with SBMA were included in a cross-sectional analysis. Normative datasets provided by the NeuroCom manufacturer and isometric strength literature facilitated patient comparisons. *Outcome Measures*. Force plate-based dynamic balance measures included FL (distance, impact index, contact time, and force impulse), SQT (turn time and turn sway), and SUO (lift up index, movement time, and impact index). Maximal isometric contractions of knee extensors, ankle dorsi exors, ankle plantar exors, and hip extensors were measured with fixed frame dynamometry. *Results*. The most difficult test, per completion rate, was SUO (52), followed by FL (57) and SQT (65). *t*-tests revealed significant abnormalities in eight of nine balance variables ($p < 0\ 05$) accompanied by large Cohen s *D* effect sizes 0 8. Receiver operating characteristics analysis showed knee extensor (SUO 95 CI =0.78–1.00, SQT 95 CI =0.64-0.92) and ankle plantar exor strength (SUO 95 CI = 0 75 – 0 99, SQT 95 CI = 0 64 – 0 92) significantly discriminated the ability to perform SUO and SQT tests with acceptable to excellent areas under the curve. *Conclusions*. Considerable dynamic balance abnormalities were observed. Lower extremity strength helps explain low test completion rates. Patients modified task movement patterns, enabling safe task performance. Study results can help direct patient care and future protocol design for people with SBMA.

1. Introduction

Spinal and bulbar muscular atrophy (SBMA), also known as Kennedy s disease (KD) [1], is an X-linked neuromuscular disorder, resulting from a mutation in the androgen receptor gene [2]. Recent research indicates the toxic effects of the mutant gene on skeletal muscle as well as motor neurons, contributing to a pathophysiology characteristic of both a motor neuron disorder and myopathy [3]. SBMA primarily affects males, while female carriers of the mutation are usually unaffected clinically [4]. The disease causes slowly progressive weakness of bulbar and extremity muscles with onset in adulthood. Musculoskeletal presentations include muscle cramps, fasciculation, tremors, weakness, and decreased or absent deep tendon re exes [5].

Major muscle group weakness leads to considerable limitations of function and endurance in people with SBMA [6]. Falls were reported in 64 of individuals from the placebo

group in a recent randomized clinical trial [7]. A study including 223 people with SBMA reported the median age of onset for handrail use on stairs to be forty-nine years, for the use of a cane to be fifty years, and for requiring a wheelchair for primary mobility to be sixty-one years [8]. Common endurance limitations in people with SBMA include difficulty with sustained and repeated movements such as sit-ups, step-ups, and heel raises [6], and walking endurance limitations have also been reported [5, 9].

Characterizing and quantifying functional limitations associated with SBMA are critical in guiding patient care and research. Some performance-based clinical tools have been studied, such as the Adult Myopathy Assessment Tool (AMAT) [6] and the six-minute walking test [9]. Recently, posturography was used to quantify static postural sway in people with SBMA [10]. However, no SBMA research has explored objective dynamic balance assessment during important tasks such as forward lunging, turning while walking, and stepping.

This research uses objective balance variables from tests of forward lunge (FL), step quick turn (SQT), and step up and over (SUO) to evaluate safe task performance and functional impairment severity in people with SBMA. While multiple different tasks would be needed to cover all the diverse aspects of dynamic balance, we selected these three tasks such that they are challenging yet doable by this clinical population; they are representative of activities of daily living that are most affected in patients with SBMA and are standard tests in the NeuroCom system previously used in multiple other clinical populations. Since one of the main characteristics of SBMA is muscle weakness, we investigated the relationship between lower extremity strength and the ability to perform the dynamic balance tasks.

2. Methods

2.1. Participants. Fifty-three males diagnosed with SBMA, with a mean age of 51 5 ± 3 5 years, BMI: 26 4 ± 3 4 kg m^2, and disease duration: 12 ± 4 2 years participated. The present investigation is a cross-sectional analysis of a larger randomized controlled trial conducted during 2011-2014 in a research hospital on the benefits of functional exercise for people with SBMA [11]. Healthy control subjects were not a part of this trial because the functional exercise group in this study was compared to a group who received stretching only. Inclusion criteria required participants to be ambulatory and able to travel to our hospital. All subjects were men over 18 years of age with genetically confirmed diagnosis of SBMA. The balance data were acquired at the baseline assessment. The protocol was approved by the Institutional Review Board, and informed consent was obtained from all participants. We compared balance scores to a normative dataset provided by the manufacturer, since healthy control participants were not included in the exercise trial. The normative dataset includes four age groups: 20-39, 40-59, 60-69, and 70-79 years old. All subjects in the normative dataset were reported to have no current or past diagnosis or injury affecting balance, be taking no medications affecting the central nervous system or known to affect balance or coordination, and have no symptoms of dizziness or lightheadedness, no symptoms suggestive of vestibular or neurological disorders, no psychological disorders including depression, no history of two or more unexplained falls within the past 6 months, and normal vision with or without glasses.

2.2. Outcome Measures. Participants performed the FL, SUO, and SQT bilaterally three times each without upper extremity support (NeuroCom Balance Master; previously Natus Medical Inc., Seattle, WA). Each subject received a detailed test description and a demonstration by the examiner. A physical therapist provided safe guarding against falls. Lower extremity strength was tested bilaterally, using a Quantitative Muscle Assessment (QMA) device consisting of a fixed-frame dynamometer (AEVERL Medical, LLC, Gainesville, GA) and load cells (Interface, Scottsdale, AZ) with computer-assisted data acquisition. Below is a description of each outcome measure.

2.2.1. Forward Lunge. Each subject was asked to step forward and lunge as far and as quickly as possible on one leg, while staying upright at the trunk, and then push backward and return to the starting position. Participants were required to bend their lunging knee to some degree for it to be considered a valid trial. For safety reasons, we could not predetermine a cut point for a minimal degree of lunging knee exion. The variables measured were (a) distance: length of the forward movement of the COG, expressed as a percentage of participants height; (b) impact index: the maximum vertical force exerted by the lunging leg as it contacts the force plate during landing, expressed as a percentage of body weight; (c) contact time: time lapsed, in seconds, between the lunging leg contacting the force plate and leaving the contact surface; and (d) force impulse: total vertical force generated by the lunging leg during the forward lunging and return to starting position phases of the movement, expressed as a percentage of body weight multiplied by the time, in seconds, that the force was exerted (total work). This test has been reported to have good to excellent reliability [12].

2.2.2. Step Quick Turn. Each subject was asked to take two steps on the force plate, make a quick pivot turn of 180 degrees, and walk back to the starting position as fast as possible. The variables measured were (a) turn time, the time to execute the turn portion of the test and (b) turn sway velocity, the distance travelled by the center of gravity (sway path), in degrees/second, during the turn. This test has been reported to have good to excellent reliability for both measures [13].

2.2.3. Step Up and Over. Each subject was asked to step up onto a four-inch curb/step (step up leg), lift the other leg (swing leg) up and over the curb without touching it, and safely lower the swing leg to land on the force plate. The variables of interest were (a) lift up index: maximum force exerted by the step up leg on the curb, expressed as a percentage of the body weight; (b) movement time: time elapsed between the initial weight shift to the nonstepping/swing leg and the impact of the swing leg down onto the force plate; and (c) impact index: the vertical force exerted by the swing