Medical Hypotheses 117 (2018) 37–41

Contents lists available at ScienceDirect

Medical Hypotheses

journal homepage: www.elsevier.com/locate/mehy

Can pre-screening vestibulocerebellar involvement followed by targeted training improve the outcomes of balance in cerebellar ataxia?

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ARTICLE INFO

Keywords:
Spinocerebellar ataxia
Balance
Vestibulocerebellum
Subjective visual vertical

ABSTRACT

Balance problems and frequent falls are common among clients with Cerebellar Ataxia (CA). CA is not a disease by itself but a collection of symptoms due to the involvement of cerebellum or its pathways. Presently the treatment for balance problems for CA is not standardized. Interventions available to improve balance are not specific to symptoms presentation. Functionally the cerebellum is divided into the spinocerebellum, vestibulocerebellum and corticocerebellum. Each functional zone has a distinct role in maintaining balance. Therefore, the presentation of symptoms will vary according to the functional zone involved. Pre-screening clients with CA for identifying the part of cerebellum involved will facilitate clinicians to provide tailor-made interventions for targeting specific symptoms for better outcomes. Pre-screening clients with CA according to the part of cerebellum involved is not in practice and our study will introduce this concept. We hypothesize pre-screening participants with spinocerebellar ataxia (SCA) for the involvement vestibulocerebellum followed by prescribing vestibulocerebellum targeted exercises will have better outcomes when compared to conventional balance training. We plan to conduct two related studies. In study 1 we will screen participants with CA for the involvement of vestibulocerebellum. In study 2, the effects of vestibulocerebellum targeted balance exercises on balance will be studied. We will assess the Subjective Visual Vertical (SVV) deviation and postural sway pattern to screen participants into people with and without vestibulocerebellar involvement. SVV deviation will be estimated using a computerized Subjective Visual Vertical (cSVV) device and postural sway pattern will be assessed using the limits of stability program of the Bertec© Balance system. The obtained SVV deviation scores will be used to derive at cut-off scores to discriminate clients with and without vestibulocerebellar involvement. The second study will test the treatment effects of conventional exercises plus vestibulocerebellum targeted exercises to improve balance by correcting SVV deviation in SCA with vestibulocerebellar involvement. The intervention is planned as 12 one-to-one sessions over three months period. Participants will be reassessed after the intervention and 3 months post-intervention. The findings of this cutting-edge research are extremely important to the clinicians, researchers and clients with SCA.

Introduction

The concept of personalized medicine (categorizing patients in order to provide tailord interventions to enhance outcomes) is gaining popularity in healthcare. Better outcomes are achieved through early detection, improvement in diagnosis, and efficient intervention delivery. The possibility of personalized medicine for managing balance dysfunction in clients with cerebellar ataxia (CA) is worthwhile exploring. Cerebellar ataxia (CA) is an umbrella term that includes health conditions with genetic or non-genetic inheritance resulting in postural instability, in-coordination of gait, speech, limb, and eyeball movements. Though the incidence of CA is low [1], the burden of CA on quality of life [2] 2 and global economy [3] for those with CA is significant. Poor balance and walking difficulties are hallmarks of health conditions associated with CA [4]. Among the genetic disorders that result in CA, the spinocerebellar ataxias (SCA) is common having a prevalence of between 0.9 and 3.0 per 100 000 depending on the exact type. SCA is an autosomal dominant hereditary disease resulting in ataxia [5]. There are over 40 types of SCA and the most common type is the SCA 6 [5,6]. Frequent falls are common among clients with SCA [7].

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https://doi.org/10.1016/j.mehy.2018.06.001
Received 4 April 2018; Accepted 4 June 2018

Implementing a screening protocol to identify the target group and delivering tailored training is the most acceptable treatment strategy in modern medicine [8]. However, such screening protocols or standardized guidelines for intervention are not available for treating balance problems in SCA [9]. This is due primarily to the limited number of high-quality studies in this field as well as the heterogeneity of the health conditions resulting in CA [10]. In addition, the effects of physiotherapy and exercises for improving symptoms of CA including balance deficits are largely unknown [11]. This justifies the need for conducting a study to explore a pathway that improves the efficiency of interventions targeted for this population.

Importance of localizing the involvement of functional zones of the cerebellum

Functionally the cerebellum is divided into vestibulocerebellum, spinocerebellum, and corticocerebellum or neocerebellum [12]. Each functional zone has a distinct role in maintaining balance and equilibrium. The vestibulocerebellum has its primary connections with the vestibular nucleus. It has a significant role in the control of balance, gait, locomotion, and eye movements [6,13]. The vestibulocerebellum controls the axial muscles responsible for equilibrium and is also responsible for the compensatory postural adjustments to counteract falls during perturbations [14]. The exact role of the spinocerebellum on balance is not clearly understood [15]; it is thought to control and refine execution of on-going movement by comparing and correcting the actual movement happening against the intended movement [13]. The extensive inputs from the anterior and posterior spinocerebellar tracts enable spinocerebellum to control movement execution [15]. The corticocerebellum is involved in higher level functions such as planning or preparation of movement and evaluation of sensory information for action as a part of motor learning process [15,16]. Since the presentation of balance dysfunction is distinct between the functional zones involvement, we hypothesize that targeted tailored exercises based on the presentation will provide better outcomes for balance in clients with SCA. For instance, emphasis could be placed on training gait and equilibrium in clients with the vestibulocerebellar lesion [6], proprioceptive training could be administered for clients with spinocerebellar lesions and complex balance activities such as dual tasks training may be administered for clients with the corticocerebellar lesion [17]. This paper will focus on pre-screening participants for vestibulocerebellar involvement followed by prescribing vestibulocerebellum targeted exercises to improve balance in participants with SCA.

The vestibulocerebellum modulates balance through its connections with vestibular system [18]. The vestibular system is responsible for visual stabilization and postural stability during head movements and provides information on one's spatial orientation to the immediate environment. Visual stabilization is achieved through the vestibulo-ocular reflex [19]. Anatomically the vestibular system is classified into peripheral and central [20]. The semicircular canals and the paired otolith organs comprise the peripheral vestibular system. The peripheral vestibular organs communicate with the vestibular nuclei which have an extensive connection with the brainstem, reticular formation, thalamus and cerebellum. These connections are collectively called the central vestibular pathway. The cerebellum controls and modulates the vestibular system through the vestibulocerebellum [18,20].

Despite evidence that the clinical presentation of balance and postural control in clients with SCA may vary with the location of insult to the cerebellum [15], it is unclear how well current balance measures accurately identify the involved functional zone of the cerebellum. A previous Delphi survey and a systematic review conducted by our team identified a wide range of outcome measures for the assessment of balance in participants with CA [21,22]. Over half of these measures were either not standardized or were not psychometrically tested for eligibility in clients with CA. Subsequently, a psychometric analysis was conducted to identify a standardized set of balance outcome measures for people with CA. The psychometric analysis found Berg Balance Scale (BBS) and the balance sub-components of the Scale for the assessment and Rating of Ataxia (SARAbal) as most appropriate measures for balance assessment in CA [23,24]. However, these tools do not localize the functional zone involved. The recently developed cerebellar-specific measures such as Scale for the Assessment and Rating of Ataxia (SARA) and International Co-operative Ataxia Rating Scale (ICARS) [25,26] have not gained popularity among health-care practitioners for the assessment of balance in CA [9] The available cerebellar-specific measures are largely disease severity rating tools with balance assessment as a sub-component rather than a standalone balance quantifying tool [17,25]. It is therefore evident that current clinical practice utilizes inappropriate measures or use measures that do not have a focus on localizing the functional zone of cerebellum.

Assessing the integrity of vestibulocerebellum

Force platform

Presently there are no standardized methods available for identifying the involvement of functional zones of the cerebellum. The pattern of postural sway could be considered to differentiate functional zones involved using force plate [17,27]. Mauritz et al. (1979), demonstrated that studying the pattern of postural sway and sway amplitude using force plate enabled identification of lesions of different functional zones of the cerebellum [28]. Based on the findings they suggested that clients with spinocerebellar lesion may have a postural tremor in an anteroposterior direction with a specific frequency of 3 Hz [28]. Specifically, they found that lesions in the corticocerebellum showed limited postural instability without directional preference and lesions of the vestibulocerebellum may result in severely affected multidirectional postural sways [28]. The amplitude of postural sway along different directions in relation to the line of gravity ranged between 4 mm and 12 mm.

Sensory Organization Test (SOT)

A laboratory-based assessment for testing sensory interaction for maintaining balance [29] could be considered for determining the integrity of the vestibular system that might be related to vestibulocerebellar involvement. However, a previous study by our team found SOT unsuitable for this population [30] perhaps due to significant floor effects [31]. Among ten included participants, 4 (40%) were ineligible for SOT because they were unable to stand unsupported for 10 s. Although SOT might predict the integrity of vestibulocerebellum, it might not be suitable for many individuals with SCA. A tool that allows assessment of balance in seated position might be best for clients with SCA.

Subjective Visual Vertical (SVV) test

To study the integrity of central vestibular pathway in participants with CA, the current research team recently conducted an observation study on 20 participants with CA secondary to multiple sclerosis [32]. The integrity of central vestibular pathway was assessed using a computerized Subjective Visual Vertical (cSVV) device. This spatial orientation testing device studies the ability of the client to accurately perceive verticality and tests the function of utricle and superior vestibular nerve of the vestibular system [33,34]. The severity of vestibular dysfunction can be correlated to the magnitude of deviation of the SVV, which is a reflection of the difference between the actual vertical and perceived vertical of the client. Though this test has been extensively used in clients with peripheral vestibular disorders such as unilateral otholith dysfunction [35,36], studies have found that findings of cSVV could identify central vestibular pathway dysfunctions in clients with multiple sclerosis [37], stroke [38], Parkinson's disease [39] and CA [40]. In our study we found a statistically significant moderate correlation (Spearman's rho correlation) between SVV deviation and measures of balance [Berg Balance scale ($r = -0.59$), Timed Up and Go test ($r = -0.58$)], ataxia severity [ICARS ($r = 0.56$), SARA ($r = 0.62$) and