# Vestibular rehabilitation with virtual reality in spinocerebellar ataxia

## Reabilitação vestibular com realidade virtual na ataxia espinocerebelar

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## ABSTRACT

The purpose of this study was to verify the benefits of the vestibular rehabilitation (VR) with virtual reality through the assessment before and after the application of the Berg Balance Scale (BBS) in four cases of spinocerebellar ataxia (SCA). The cases were underwent the following procedures: anamnesis, ear inspection, vestibular assessment and application of the BBS before and after VR with virtual reality using games from Wii Fit device. The cases describe four patients that were diagnosed with genetically inherited SCA (two type 2, one type 3 and one still under investigation), three of them were female and one was male, with ages ranging from 30 to 62 years. The patients presented otoneurological symptoms and the vestibular test showed the presence of semi-spontaneous nystagmus, absence of post-rotational nystagmus, hyporeflexia, and asymmetric directional preponderance of the nystagmus in the caloric test. Patients from cases 1 and 2 have showed an improvement in motor coordination and in body balance, even though the score presented by the BBS had demonstrated medium risk for falling before and after the exercises. In case 3, the patient's loss of balance and BBS score have improved, presenting low risk of falling; whereas the patient in case 4 did not show any improvement in the assessment after the exercises. This case study shows the applicability of VR exercises with virtual stimuli in SCA with improved motor coordination and postural balance.


**Keywords:** Spinocerebellar degenerations; Ataxia; Vestibular diseases; Electronystagmography; Rehabilitation


## RESUMO

O objetivo do estudo foi verificar os benefícios da reabilitação vestibular (RV) com realidade virtual, por meio de avaliação pré e pós-aplicação da Escala de Equilíbrio de Berg (EEB), em quatro casos de ataxia espinocerebelar (AEC). Os casos foram submetidos aos seguintes procedimentos: anamnese, inspeção otológica, avaliação vestibular e aplicação da EEB pré e pós-RV, com a realidade virtual representada por meio da utilização de jogos do equipamento Wii Fit. Os casos retratam quatro pacientes com diagnóstico genético de AEC (dois tipo 2, um tipo 3 e um em investigação), sendo três do gênero feminino e um do gênero masculino, na faixa etária de 30 a 62 anos. Os pacientes referiram sintomas otoneurológicos e, no exame vestibular, observou-se a presença de nistagmo semiespontâneo com características centrais, ausência de nistagmo pós-rotatório, hiporreflexia e preponderância direcional do nistagmo assimétrica à prova calórica. Nos casos 1 e 2, os pacientes referiram melhora na coordenação dos movimentos e do equilíbrio corporal, independente do escore na EEB ter demonstrado médio risco para queda, antes e após a realização dos exercícios. No caso 3, o paciente apresentou melhora do escore na EEB, bem como do equilíbrio, apresentando baixo risco para queda. O caso 4 não evidenciou melhora na avaliação após a execução dos exercícios. Este estudo de caso demonstrou a possibilidade da aplicação dos exercícios de RV com estímulos virtuais na AEC, com melhora da coordenação motora e do equilíbrio postural.


**Descritores:** Degeneração espinocerebelar; Ataxia; Doenças vestibulares; Eletronistagmografia; Reabilitação

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## INTRODUCTION

Spinocerebellar ataxias (SCA) are a heterogeneous group of neurodegenerative diseases that are characterized by the presence of progressive ataxia and have as initial clinical manifestations deterioration in balance and coordination, and eye disorders[1]. The ataxias are classified as: sensory, frontal, vestibular and/ or labyrinth, and cerebellar[2].

The etiology in most cases is caused by mutations characterized by the presence of an expansive and unstable trinucleotide CAG repeat in the coding region of the gene reported. In Brazil, more specifically in the South, we have evaluated a large number of families suffering from SCAs[1,2]. Machado-Joseph disease (MJD), known as SCA type 3, initially described in the Azores, is the most common form of hereditary ataxia with autosomal dominant inheritance. Found in prominent worldwide epidemiological studies, it is also the most common form of SCA in Brazil[2,3].

SCAs have a geographic prevalence variable. Type 2, which is chromosome 12q24.1, ATXN2 gene, CAG mutation, Ataxin-2 protein, has a high incidence in Cuba, India, England, France, and the United States. Type 3, which is chromosome 14q32.1, ATXN3 gene, CAG mutation, Ataxin-3 protein, is highly prevalent in Portugal, Brazil, Germany, Japan, and China[1].

The disorders are characterized by progressive degeneration of spinocerebellar tracts. Among the neurological manifestations present, vision loss and nystagmus are common features of these diseases[4].

The cerebellum receives visual, auditory, vestibular, and somatosensory afferents from the brainstem and somatosensory receptor information from the limbs and from motor, premotor and prefrontal areas in the cortex of the brain. The cerebellum has three anatomical regions: a) medial zone (cerebellar vermis and core meridian), which is responsible for the control of posture, balance and movement; b) intermediate zone (intermediate and core interposite hemisphere), responsible for the control of movement and discrete ipsilateral limb reflexes; c) lateral zone (lateral hemisphere and dentate nucleus), responsible for motor planning and the complex movements of the limbs guided by vision[5].

Cerebellar lesions have classic symptoms such as ataxia, hypotonia, nystagmus, dysarthria and motion tremors[1].

The vestibulocerebellum is responsible for balance and body position in space. In the event of an injury at some of these systems, there is a conflict of information and there may be symptoms of body imbalance and dizziness. There are several types of dizziness, for example, spinning dizziness known as vertigo, and non-spinning, which can be characterized by instability, lightheadedness, and loss of balance, among others[6].

Vestibular rehabilitation therapy (VRT) has been shown to act physiologically on the vestibular system. It is considered a therapeutic option because of its proposed action, and is based on the central mechanisms of neuroplasticity, known as adaptation, habituation, and substitution in order to achieve vestibular compensation[6].

In adaptation, the vestibular system learns to receive and process information, even if inadequate or incomplete, with adaptation to stimuli. In habituation, the system processes to correct or reduce inappropriate answers when the vestibular system is stimulated and the body starts to respond appropriately. Substitution is focused on the prioritization of sensory perception, which aims to replace the missing or conflicting information, related to balance[6]. The central nervous system (CNS) processes this information and generates responses by reflexes, among which we highlight the vestibulo-ocular reflex (VOR) and the vestibulospinal reflex (VSR)[6].

Thus, neuroplasticity refers to the ability of the CNS has to modify some of its morphological and functional properties in response to environmental changes[6].

VRT using virtual reality consists of an interaction of graphical images, using an interface between the individual and a machine. The exploitation of composite applications by simulated scenes and situations makes the individual believe to be in another reality[7]. The benefits associated with this treatment, described in the literature, include correction of balance and posture, improved mobility, functionality of upper and lower limbs, and greater motivation for the patient in exercises[8].

SCA is part of a list of diseases that present major changes involving the fields of Speech, and more specifically, Otoneurology.

Thus, the aim of this study was to assess the benefits of VRT with virtual reality through the pre-application and post--application of the Berg Balance Scale in four cases of SCA.

## CLINICAL CASES PRESENTATION

We evaluated four patients, three female and one male, diagnosed with SCA (two cases of type 2, one case of type 3 and one case under investigation), between 30 and 62 years of age, referred from the Clinical Hospital to the Laboratory for Neurotology at Universidade Tuiuti do Parana (UTP). The study was approved by UTP's Ethics Committee, under n° 058/2008 and performed with authorization of patients after signing a consent form.

The diagnosis of ataxia was performed by genetic testing, using the Polymerase Chain Reaction (PCR) technique.

The study included patients with no middle ear conditions, without the use of devices for marching, and who had not performed any earlier rehabilitation therapy. Patients unable to meet and understand simple verbal commands, with otological alterations, severe visual impairment, or other abnormalities that prevented the realization of the proposed procedures were excluded.

We carried out the following procedures:

*Anamnesis*

A questionnaire focusing on otoneurological signs and symptoms.

*ENT evaluation*

Performed in order to rule out any alteration that could affect the test.

## Vestibular evaluation

The patients underwent the following tests that made up the vestibular examination: test for vertigo and for spontaneous, semi-spontaneous, and positional nystagmus.

To perform the electronystagmography (ENG) a thermosensitive unit with three recording channels from Berger® model VN316 was used. We used also a Ferrante® brand rotary chair, a Neurograff® EV VEC visual stimulator, and an NGR model 5 air otocalorimeter, also by Neurograff®.

The following ENG eye and labyrinth tests were performed: an eye movement calibration, a test for semi-spontaneous and spontaneous nystagmus, a pendular tracking test, an optokinetic nystagmus test, as well as pre-rotatory and post-rotatory, pre--caloric and post-caloric tests. The caloric stimulation time for each ear with air at 42°C and 18°C was 80 seconds at each temperature. The responses were recorded with eyes closed and then with eyes open to observe the inhibitory effect of ocular fixation.

## Application of the Berg Balance Scale (BBS), the Brazilian version

This scale has been culturally adapted to the Brazilian population by the author[9] and has been widely used to determine the risk factors for loss of independence and for falls. It evaluates the performance of functional balance in 14 items common to everyday life. Each item has an ordinal scale of five alternatives ranging from zero to four points. Therefore, the maximum score is 56. The points are based on the time in which a position can be maintained, the distance at which the upper limb is able to reach the front of the body and the time required to complete the task. The higher the score, the better the balance. The analyses are carried out based on the results obtained for each score, or the time needed to develop individual skills to perform tasks in controlling body balance. Scores for low falling risk (56-64), medium falling risk (53-46), high falling risk or 100% falling risk (below 36) are measured[10].

## Vestibular rehabilitation with virtual reality games from Wii Fit Plus

We used a virtual reality videogame from the Nintendo® Wii gaming platform, which allows the user to have the feeling of movement in different sports. The game assigned to this protocol was Wii Fit Plus, which simulates a circular motion of the pelvis, as well as extension, rotation and weight transfer exercises, aimed at changes in balance and postural instability. The following games were used: Bird's-eye Bull's-eye, Big Top Juggling, and Hula Hoop, displayed on a 1.10 $m^2$ projection screen, and using, as accessories, two handsets of the same brand, one loudspeaker and a balance platform called Balance Board. This board functions as a surface that measures the force applied and senses small changes in equilibrium by means of pressure sensors. Sensors are responsible for the interface between the machine and the player. Initially, patients were individually familiarized with the game and instruction was given regarding the movements necessary for its execution. All games used were played for 30 minutes for a total of ten times, twice a week.

## Case 1

Female, 44 years old, three years with disease, with symptoms of moderate and sporadic dizziness (did not know how long these symptoms has persisted), headaches, imbalance when walking, difficulty or pain with neck movements, dysphonia, dysphagia, and tremors. Vestibular testing revealed a central bilateral deficit vestibular disorder. Analyzing the application of the BBS and performance during the execution of the exercises, we observed the following scores: pre-treatment – 48 points, medium risk for falling, difficulty and uncoordinated movements during the execution of the exercises; post-treatment – 51 points, medium risk for falling and improvement in the coordination of movements during the games.

## Case 2

Female, 30 years old, two years with disease, with symptoms of intensive and frequent occurrence of dizziness since childhood, gait deviation to the left, hearing loss, dysphagia, dysarthria, agitation during sleep and insomnia. Vestibular testing noticed a central bilateral deficit vestibular disorder. Analyzing the application of the BBS and performance during the execution of the exercises, we observed the following scores: pre-treatment – 47 points, medium risk of falling, initial dizziness during the exercises, good mobility for the movements of the pelvis and difficulty transferring weight on the balance board; post-treatment – 50 points, medium risk of falling, greater ease in performing exercises that simulate weight transfer and reported improvement in balance after performing the exercises.

## Case 3

Female, 30 years old, six months with disease, with symptoms of imbalance when walking, dysphagia, dysphonia, and dizziness. Vestibular examination showed a peripheral bilateral deficit vestibular disorder. Analyzing the application of the BBS and performance during the execution of the exercises, we observed the following scores: pre-treatment - 52 points, medium risk of falling, no complaints during the execution of the exercises, with difficulty keeping on the balance board

and tracking the movements of the games; post-treatment - 55 points, low risk for falling and no imbalance for movements executed on the board.

### Case 4

Male, 62 years old, four years with disease, type of SCA under study, with symptoms of imbalance when walking, double vision and dysphagia. Vestibular examination showed an irritative peripheral vestibular disorder. Analyzing the application of the BBS and performance during the execution of the exercises, we observed the following scores: pre-treatment – 46 points, medium risk of falling, difficulty with movements of pelvic mobility and lower limbs; post treatment – 45 points, high risk for falling, and the same initial difficulties and dizziness.

## DISCUSSION

It was found that, despite the difference in age, time since diagnosis, and classification of SCA, all cases reported initial difficulty in the use of virtual games, probably because of the innovation of the method used, which stresses the importance of familiarization with the protocol[11].

Chronic diseases affecting balance can generate higher inactivity and changes in physical conditions for these patients. Lack of physical fitness is potentially the most disabling consequence of vestibular dysfunction and therefore movement should be encouraged[11].

The use of virtual simulations and environments may cause the individual to believe he is truly in another reality, enabling the association of behaviors and responses with movements of virtual objects, allowing for integration with the user's virtual environment[7].

In the present study, we observed that in cases 1 and 2, the patients reported improvement of coordination of movements and body balance, regardless of the score in BBS showing an average risk of falling, before and after the exercises. In case 3, the patient showed improvement in the BBS score and in balance, with a low risk for falling. Thus, it was found that the virtual environment is favorable for movements for the treatment of imbalances and postural instability shown by patients. The authors[7] observed through the virtual game that the patients showed improvement in posture correction and balance, increasing the capacity of movement and range of motion of the upper and lower limbs, which confirmed the above results.

Case 4 showed no improvement in the evaluation after the execution of the exercises. Probably, his age group was responsible for the observed difficulties, as it is scientific knowledge that, with physiological aging, there is a decrease in mobility and ability of movement, as well as other factors, associated with senescence and senility which may justify an increase in the risk of falling[12].

Virtual reality presents itself as an important resource in helping in patients with balance disorders. By means of virtual games, the patient may interact with the proposed virtual environment and receive visual feedback regarding the changes in movements, thereby creating strategies to restore and/ or maintain body balance[13].

The authors[14] used the virtual games of golf, boxing, tennis, bowling and baseball for Wii Fit in patients with cerebral palsy and observed an improvement in the process of visual perception, postural control, and functional mobility, which highlights the VRT benefit of virtual reality in accordance with the research presented, differing only in the underlying disease.

Studies conducted by the authors[15] in a patient with ataxia, using Wii Fit to stimulate latero-lateral and antero-posterior balance revealed that the patient improved, i.e., an increase of 23.21% in BBS, suggesting that VRT with virtual reality was important for balance, providing greater independence to carry out daily tasks, a fact also noted by the authors of this study.

It is known that body balance depends on the integrity of the visual, vestibular and somatosensory systems, all controlled by the cerebellum. VRT exercises performed by conventional protocols or using virtual reality have the same goals, which are to restore body balance, accelerate and stimulate natural mechanisms of vestibular compensation and thus maximize neuroplasticity of the individual with peripheral and/ or central vestibular disorders. What distinguishes the two protocols is the increasingly improving approach that was used, making the quantitative methods more effective for assessment and rehabilitation of the systems responsible for body balance.

In Brazil, there is an interest, now insignificant but increasingly growing, in the use of virtual reality for VRT with various neurological disorders. The literature that deals with the subject in question is still scarce, which demonstrates not only the importance of this study, but also the possibility of a better quality of life to be provided to patients already suffering and often weakened by the consequences of cerebellar disorders.

## FINAL COMMENTS

In the cases studied, it was found that labyrinthine alterations indicate disorders of the vestibular system. Improved coordination of movements and postural balance when comparing pre- and post-VRT using virtual reality in patients with SCA was evident. This improvement can be attributed to central structures that act in neuroplasticity and thereby enhance the natural mechanisms of compensation for adaptation, habituation, and substitution in the nervous system, enabling individuals with SCA to improve performance of these functions.

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