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Case Report

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Adapted vestibular rehabilitation in Spinocerebellar Ataxia type 3: a case report

Abstract

Spinocerebellar ataxias (SCA) are a group of progressive hereditary neurodegenerative disorders, with type 3 (SCA3) being the most prevalent. SCA3 is characterized by cerebellar ataxia, ocular movement abnormalities, dysarthria, and vestibular symptoms, such as dizziness and imbalance. Vestibular rehabilitation (VR), although classically recommended for peripheral vestibular disorders, has been investigated as a therapeutic possibility for central vestibulopathies. This study aimed to describe the effects of adapted vestibular rehabilitation, based on the Cawthorne and Cooksey protocol, in a patient diagnosed with SCA3. A 44-year-old female patient underwent an audiological assessment, anamnesis, and application of the Dizziness Handicap Inventory (DHI) and Tinnitus Handicap Inventory (THI) questionnaires before and after 12 sessions of VR. The therapeutic program included habituation and adaptation exercises and postural control strategies. Results showed an improvement in balance and gait performance, as well as a reduction in tinnitus and dizziness-related symptoms. The findings reinforce the effectiveness of adapted vestibular rehabilitation in patients with central vestibular disorders such as SCA3.

Keywords: vestibular rehabilitation, spinocerebellar ataxia type 3, dizziness, cawthorne-cooksey protocol

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Introduction

One of the most complex sensorimotor functions of the human body is postural control, as it requires optimal balance and seamless integration of information exchanged among the vestibular, visual, proprioceptive systems and the central nervous system (CNS). Any breakdown in communication among these systems leads to motor, sensory and cognitive deficits that impair an individual's bodily movement. Certain neurological disorders, such as ataxias, create systemic disharmony by disrupting motor control and exposing the patient to vertigo symptoms. It is clinically characterized by an insidious and progressive onset of motor symptoms, especially cerebellar ataxia, which manifests as imbalance, dysmetria, dysarthria, and gait instability. In addition to cerebellar signs, extrapyramidal manifestations (such as rigidity, dystonia, and parkinsonism), pyramidal signs (hyperreflexia, spasticity, and Babinski sign), and peripheral neuropathy with reduced reflexes and sensory loss in the lower limbs may also occur. Oculomotor abnormalities, such as nystagmus and ophthalmoplegia, are also common.[1] The age of onset is variable, typically between 20 and 50 years, but it can occur during adolescence or in old age. The disease progresses slowly, gradually leading to a loss of functional independence.[2]

Diagnostic confirmation is achieved through molecular genetic testing, which identifies an abnormal expansion of the CAG trinucleotide repeat in the ATXN3 gene, located on chromosome 14q32.1. Healthy individuals have up to 44 CAG repeats, whereas patients with SCA3 have 55 or more. There is an inverse correlation between the number of repeats and the age of symptom onset that is, the greater the number of repeats, the earlier the disease begins. The diagnosis of SCA3 is confirmed when there are compatible clinical symptoms, a positive family history, and identification of the genetic mutation. This is an incurable condition, and management is focused on motor rehabilitation and a multidisciplinary approach to improving patients' quality of life.[3] According to the National Organization for Rare Disorders, spinocerebellar ataxias (SCAs) comprise a diverse group of progressive, hereditary, autosomal-dominant pathologies, with more than twenty distinct types identified worldwide.[4] Although rare, spinocerebellar ataxia type 3 (SCA3), also known as Machado Joseph Disease, is the most prevalent SCA, accounting for 20–50 percent of familiais ataxias and affecting approximately two in 100,000 people. The highest frequencies of SCA3 have been reported in Portugal (58–74 percent), Brazil (69–92 percent), China (48–49 percent), the Netherlands (44 percent), Germany (42 percent), and Japan (28–63 percent).[5] SCA3 arises from an expanded CAG trinucleotide repeat in the genome, which disrupts production of the ataxin-3 protein and triggers a degenerative process in the CNS. Its hallmark symptom is loss of balance and motor coordination. As the disease progresses, patients may also develop speech disturbances, swallowing difficulties and diplopia, typically with adult onset.[4] Dizziness and imbalance commonly accompany ataxia. Such symptoms may stem from primary (central) causes or secondary involvement of the labyrinth-the innerear organ responsible for spatial orientation and equilibrium. When communication between vestibular, visual and proprioceptive inputs is compromised, conflicting sensory-motor signals generate body imbalance and vertigo.[6] In cases of vestibular dysfunction, the CNS struggles to satisfactorily integrate visual and proprioceptive cues.[7] Vestibular rehabilitation (VR) has been advanced as a therapeutic resource because it acts directly on the labyrinth and engages central neuroplastic mechanisms namely adaptation, habituation and substitution to facilitate vestibular compensation.[8]

The Brazilian Society of Otorhinolaryngology's 2000 Consensus on Vertigo recommends a standardized protocol for dizzy patients, encompassing: (1) patient and family education on balance physiology, symptomatology and proposed interventions; (2) initiation of VR exercises repeated eye, head and trunk movements and gait training; and (3) guidance on the importance of continued follow-up and progress monitoring over a minimum three-month period.[9] Authors report that SCA3 not only impairs motor function but also provokes neuropsychological symptoms. Lin notes that patients complaining of dizziness often experience depression, anxiety, fatigue and poor sleep quality, all of which significantly impact quality of life.[10] Thus, beyond improving visual and postural stability and enhancing vestibulo-visual interaction during head movements, VR restores physical autonomy and confidence.[7] Spinocerebellar ataxias intersect multiple clinical fields, notably speech-language pathology and otoneurology. Given the topic's relevance to these disciplines, the therapeutic approaches employed and the patient's clinical evolution, this study aims to describe the contributions of vestibular rehabilitation using an adapted Cawthorne & Cooksey protocol in a patient with spinocerebellar ataxia type 3.

Case report

This prospective, observational case report was approved by the Research Ethics Committee (CEP) of the State University of Bahia (Uneb; protocol nº. 253.990) and began after the subject provided written informed consent via the Free and Informed Consent Form (TCLE). A 44-year-old female patient was referred by her otolaryngologist for vestibular rehabilitation with a diagnosis of spinocerebellar ataxia type 3 (SCA3). She attended sessions at the Otoneurology Division of the Speech-Language Pathology teaching clinic at Uneb from May 3 to July 21, 2022. During the otoneurological evaluation, the patient reported constant dizziness (objective vertigo), gait imbalance, and tinnitus. She also experienced associated symptoms of falls, nausea, emesis, and visual distortion. Her overall medical history included ataxia, migraine, dysphagia, and diplopia. She was taking a labyrinth suppressant and anxiolytics daily. For mobility, she used a fourpoint cane and, occasionally, a walker. Figure 1A illustrates a therapeutic activity performed during gait training with cane support.

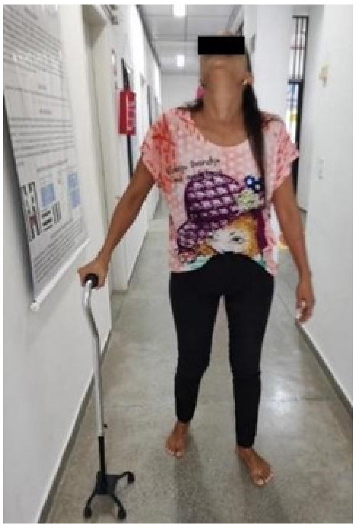

Figure 1A Patient in therapeutic activity.

Assessment and intervention instruments

To quantify the patient's progress, two measurement points were established-prior to the start of vestibular rehabilitation (VR) and after one month of therapy-using the pre- and post-treatment administrations of the Dizziness Handicap Inventory (DHI) and the Tinnitus Handicap Inventory (THI).

Dizziness handicap inventory (DHI)[11]

The DHI is a 25-item questionnaire that assesses how dizziness affects quality of life. Seven items address physical aspects, nine items cover emotional aspects, and nine items examine functional limitations. It was administered as an interview: each "yes" response scores 4 points, "sometimes" scores 2 points, and "no" scores 0 points. Total scores range from 0 to 100; the closer to 100, the greater the impact of dizziness on quality of life.

Tinnitus handicap inventory (THI)[12]

Developed by Newman et al. and adapted for Brazilian Portuguese, the THI comprises 25 items divided into three subscales:

i. Functional (11 items): gauges tinnitus-related limitations in mental, social, occupational, and physical domains.

ii. Emotional (9 items): addresses affective responses such as anger, frustration, irritability, and depression.

iii. Catastrophic (5 items): explores the most severe reactions, including despair, loss of control, inability to face problems or escape the tinnitus, and fear of serious illness.

Responses are scored "yes" = 4 points, "sometimes" = 2 points, and "no" = 0 points. Total scores range from 0 to 100; higher scores indicate greater disadvantage due to tinnitus.

Therapeutic program

The primary goal of therapy was to alleviate dizziness. Upon entering the vestibular rehabilitation program, the patient underwent:

i. An overview of the program's structure

ii. Theoretical instruction on the anatomy and pathophysiology of the vestibular system

iii. A speech-language anamnesis focused on auditory and vestibular systems

iv. A scapular-girdle relaxation exercise protocol

v. Cawthorne-Cooksey vestibular exercise protocols

Cawthorne-Cooksey vestibular rehabilitation protocol[13,14]

Sessions were held once a week for three months at the Speech-Language Pathology teaching clinic of the State University of Bahia. These exercises aim to restore static and dynamic balance and spatial orientation by engaging eye, head, and body movements in both seated and standing positions:

Seated eye and head movements

i. Look up and down

ii. Look right and left

iii. Focus on a finger as it moves closer and farther (slowly, then rapidly)

iv. Flexion/extension of the head with eyes open (slowly, then rapidly)

v. Lateral head turns with eyes open (slowly, then rapidly)

vi. Repeat exercises 4 and 5 with eyes closed

Seated head and trunk movements

i. Pick up an object from the floor using trunk flexion/extension while fixating on it

ii. Flex the trunk while passing an object in front of and behind the knees

Standing exercises

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i. Sit to stand with eyes open

ii. Repeat with eyes closed

iii. Repeat exercise 1, adding a 360° turn to the right

iv. Repeat exercise 1, adding a 360° turn to the left

Balance-enhancing activities

i. Walk while rotating the head right and left

ii. Stand and execute sudden 90° turns with eyes open, then closed

iii. Ascend and descend stairs (using the handrail)

iv. Stand on right foot then left foot, eyes open, then eyes closed

v. Stand on a soft surface

vi. Walk on a soft surface

vii. Tandem gait (heel-to-toe) with eyes open, then closed

viii. Repeat exercise 4 on a soft surface

Scapular-girdle relaxation protocol[15]

This protocol employs neck and shoulder stretches to promote relaxation and sensory integration, which can aid vestibular symptom management:

i. Shoulders: Elevate and lower, then rotate each shoulder clockwise and counterclockwise

ii. “Yes” nod: Flex and extend the head as if nodding “yes”

iii. “No” shake: Rotate the head right, then left as if saying “no”

iv. Lateral side-bend (“Maybe”): Tilt the head to the right, then left; assist by gently pulling the opposite arm to deepen the stretch

v. Rotation: Rotate the head in full circles, 10 times clockwise and 10 times counterclockwise

Relaxation exercises in vestibular rehabilitation are widely recognized for improving quality of life in patients with vestibular dysfunctions by reducing muscle tension and enhancing sensorimotor integration.

Therapeutic intervention

Based on the clinical history and prior otoneurological assessment, an otoneurological diagnosis could not be established due to an exacerbation of the patient’s dizziness triggered by sensory conflicts during the evaluation.[16] Patients with vestibular disorders often struggle to integrate sensory information from different systems (visual, vestibular, and proprioceptive), which can lead to heightened symptoms such as dizziness and imbalance.[17] The proposed therapeutic intervention used the exercises from the Cawthorne Cooksey protocol in an adapted form (Figure 1B).[18] Before beginning the therapeutic program, the patient completed the pre-treatment Dizziness Handicap Inventory (DHI), scoring 82 points overall, and the pre-treatment Tinnitus Handicap Inventory (THI), scoring 40 points. She was then instructed to perform the adapted exercises at least three times a day. The patient reported occasional dizziness while doing the exercises particularly during the head-rotation tasks. Because of the protocol’s adaptations, any stages requiring the patient to lie supine were omitted, as those positions provoked both dizziness and emesis (Figure 1C). After seven sessions, the DHI and THI were readministered. The patient demonstrated improvements in dizziness, as reflected by the scores: 50 points on the DHI and 2 points on the THI (Table 1).

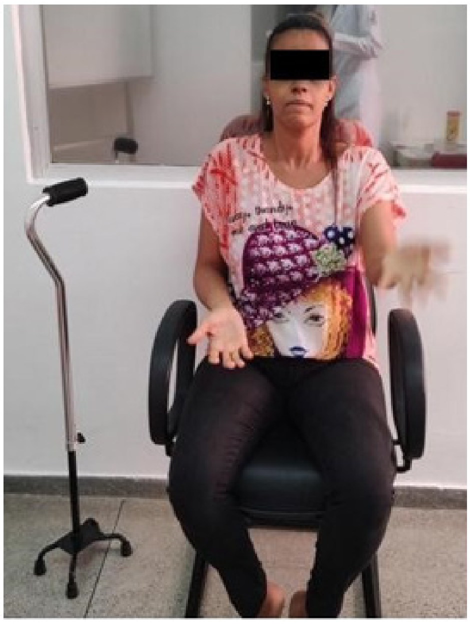

Figure 1B Patient in therapeutic activity.

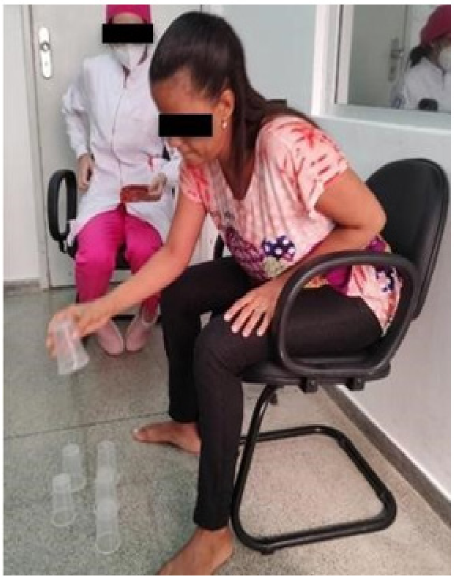

Figure 1C Patient in therapeutic activity.

Table 1 Pre- and Post-Treatment Scores on DHI and THI

Domain	Pre-DHI	Post-DHI	Pre-THI	Post-THI
Physical	28	18	–	–
Functional	30	22	–	–
Emotional	24	10	–	–
Catastrophic	–	–	24	0
Total	82	50	40	2

Discussion

Vestibular rehabilitation (VR) is grounded in specific mechanisms that leverage central nervous system plasticity to treat balance disorders. Its primary goals are to restore vestibulo-visual integration during head movements and to promote both static and dynamic visual and postural stability under conditions that produce conflicting sensory cues. Existing protocols are diverse, offering effective therapeutic options that can be tailored to each patient's individual profile. The success of VR may be influenced by factors such as patient age, emotional state, medication use, and the presence of central nervous system disorders that impair vestibular neuroplasticity. The patient's motivation to perform the exercises is also taken into account, since it directly affects rehabilitation outcomes. Recovery may be prolonged or limited if the patient restricts head movements or minimizes visual input. Patients with vertigo almost invariably limit their movements in an effort to avoid triggering symptoms of imbalance and nausea.[16,19,20] The exercises selected from the Cawthorne and Cooksey protocol, based on head movements, eye-head coordination tasks, global body movements, and balance tasks, have proven effective when adapted to meet the patient's specific needs, such as performing movements while seated, incorporating breathing exercises like inhaling and exhaling, and swallowing water. This fact is supported by the literature, demonstrating positive outcomes in patients with peripheral vestibulopathies. In the patient in question, an improvement in overall condition and quality of life was observed, despite the pathology being central. Cawthorne and Cooksey exercises have shown effectiveness in treating peripheral vestibulopathies, requiring regular execution. A specific number of sessions and frequency of exercises are recommended, as repetitive exposure to a provocative stimulus leads to a reduction in the pathological response. This technique is based on the phenomenon of vestibular habituation.[13,14,16]

According to the literature, AEC is characterized as a progressive and neurodegenerative disorder, whose manifestations include incoordination, body balance problems, and ocular disturbances. In the present study, an improvement in the symptom of dizziness was observed, leading to a decrease in the DHI score post-treatment. DHI is a tool used to assess the impact of dizziness on patients' quality of life. Studies show that vestibular rehabilitation not only improves physical symptoms but also has a positive impact on the emotional domain. The correlation between dizziness symptoms and quality of life suggests that improvement in vestibular symptoms can lead to a reduction in anxiety and depression, thus enhancing patients' emotional well-being.[5,17] Maria et al.[21] observed that their patients showed improvement in balance and functional independence after a vestibular rehabilitation program. These positive reports may be associated with adaptations of the central nervous system following repetitive sensory stimuli, evidencing suggestive signs of vestibular compensation through the phenomenon of habituation.[21] The effectiveness of vestibular rehabilitation (VR), assessed through the Dizziness Handicap Inventory (DHI), measures the individual's disability in three domains before and after treatment.[13]

Physical domain: Evaluates eye, head, and body movements, as well as worsening symptoms or dizziness triggers.

Emotional domain: Measures frustration, shame, depression, inability to perform tasks, concentration disturbances, and even changes in family dynamics.

Functional domain: Assesses the ability to perform tasks independently, such as walking or moving around the house unaided or in the dark.

The analysis is based on the difference in score pre- and post-treatment, with effectiveness determined by a minimum difference of 18 points.[13] Thus, VR's positive outcomes are justified by its activation of central systems linked to neuroplasticity, such as the equalization and adaptation of functional balance resulting from neural changes in the cerebellum and brainstem due to repeated sensory stimuli as well as consideration of the individual's psychosocial aspects.[13]

Conclusion

This case demonstrates that, despite spinocerebellar ataxia type 3 being a progressive central vestibulopathy with well-defined clinical and genetic diagnostic criteria, vestibular rehabilitation can yield meaningful symptomatic and functional benefits. While traditional diagnostic frameworks often emphasize the neurodegenerative and incurable nature of SCA3, the positive response to the adapted Cawthorne and Cooksey protocol in this case suggests that rehabilitation may play a more central role than typically acknowledged. These findings reinforce the need to complement diagnostic precision with individualized, interdisciplinary care strategies, highlighting the potential of tailored vestibular therapy to improve quality of life even within the constraints of a progressive disorder.

Acknowledgments

None.

Conflicts of interest

The authors declares that there are no conflicts of interest.

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