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Physical Therapy for Cerebellar Ataxia

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Abstract

Ataxia, the incoordination and balance dysfunction in movements without muscle weakness, causes gait and postural disturbance in patients with stroke, multiple sclerosis, and degeneration in the cerebellum. The aim of this article was to provide a narrative review of the previous reports on physical therapy for mainly cerebellar ataxia offering various opinions. Some systematic reviews and randomized control trial studies, which were searched in the electronic databases using terms “ataxia” and “physical therapy,” enable a strategy for physical therapy for cerebellar ataxia. Intensive physical therapy more than 1 hour per day for at least 4 weeks, focused on balance, gait, and strength training in hospital and home for patients with degenerative cerebellar ataxia can improve ataxia, gait ability, and activity of daily living. Furthermore, the weighting on the torso, using treadmill, noninvasive brain stimulation over the cerebellum for neuromodulation to facilitate motor learning, and neurophysiological assessment have a potential to improve the effect of physical therapy on cerebellar ataxia. Previous findings indicated that physical therapy is time restricted; therefore, its long-term effect and the effect of new optional neurophysiological methods should be studied.

Keywords: ataxia, cerebellum, physical therapy, balance training, noninvasive brain stimulation, stroke, degenerative ataxia, multiple sclerosis

1. Introduction

Incoordination and balance dysfunction in movements without muscle weakness are the most accepted definition of ataxia, which has three subcategories: sensory, vestibular [1], and cerebellar ataxia [2, 3]. When the cerebellum is damaged, activity of daily living (ADL) is disturbed in patients with the ataxia owing to diseases, such as spinocerebellar degeneration [4–8], multiple sclerosis (MS) [2, 3, 9–13], and stroke [14–17]. In contrast, these can have several causes in children, such as infection and tumor [18].

The cerebellum contributes to sensory motor control [3, 19–25], gait [11, 21, 26–31], and balance [5] for maintenance of upright posture. Furthermore, the cerebellum has an important role in motor learning [32] and adaptation [33]. Therefore, if the cerebellum is damaged, then these functions are disturbed, and dysmetria, tremor, rebound phenomenon, dysdiadochokinesia, dyssynergia, and hypotonia [24, 34].

Intervention is provided to patients with cerebellar ataxia to recover motor function and ADL. The intervention for rehabilitation includes medication [35, 36], surgery, and physical therapy [6, 9]. The effect of medication and surgery depends on the cause of ataxia and extent of neuronal damage [37, 38]; however, there is no radical treatment for these diseases yet.

Patients with cerebellar damage have impaired motor learning [15, 39–46], but their ataxia, gait, and ADL can be improved. Studies with high methodological quality and scientific evidence regarding the intervention using physical therapy for cerebellar ataxia occur as systematic reviews, randomized control trials (RCT), and guidelines. Systematic reviews can particularly provide important information on physical therapy use for cerebellar ataxia. The systematic reviews offer various viewpoints to solve various clinical questions. The aim of this article was to review the important points for physical therapy to improve the motor function and ADL in patients with cerebellar ataxia and to search for the point which we should study in the future to establish the evidence of the effect of physical therapy for cerebellar ataxia. Based on these previous findings and opinions in this search, I will mention about recommended and possible physical therapy which includes the measure and intervention for cerebellar ataxia and ADL in the patients with cerebellar damage.

2. Method of search

A search for articles written in English, using the word “ataxia,” was performed by the author in electronic databases of the Physiotherapy Evidence Database (PEDro), which is provided by the George Institute for Global Health for Physical Therapy. The searched systematic reviews were scrutinized based on the aim of the articles, including RCTs for physical therapy, and the year of publication of the target articles. Subsequently, article search was conducted using the words “ataxia” and “physical therapy” or “physiotherapy” in PubMed [1970–2016]. The effective methods of measurement and intervention for ataxia were mainly assessed transversely in systematic reviews and RCT studies. Furthermore, other than these reports, we also considered the possible method of measure and intervention for cerebellar ataxia.

3. Review

Six systematic reviews [6, 9, 10, 36, 47, 48] written in English regarding physical therapy were searched in these databases. Six RCTs [2, 49–54] were included in these systematic reviews that were published before October 2016. In addition, another RCT [55] was found in PubMed,

upon searching for all systematic reviews and RCT reports regarding physical therapy that were published from 1980 to 2016. An article regarding intensive physical therapy for spinocerebellar degeneration (SCD) written by Miyai et al. [50] was referred in three review articles [10, 47, 48], which were all systematic reviews published after 2012, as the most influential study with high methodological quality and scientific evidence on physical therapy for patients with degenerative cerebellar ataxia. These RCTs regarding MS or SCD were included in these reviews and that regarding stroke was not included. No RCT report on the long-term effect of physical therapy for cerebellar ataxia was found. In addition, no study regarding the meta-analysis on physical therapy for cerebellar ataxia was found.

3.1. Outcome measure for physical therapy

3.1.1. Evaluation for ataxia, balance, gait ability, and ADL

The definition of function and ADL should be measured before and after physical therapy [10, 34] because clients and therapists must judge the effectiveness of the intervention. Clinical scales are often used for investigating the effect of physical therapy. In cerebellar ataxia, the most often used specific scales for the severity of cerebellar ataxia in previous studies were the scale for the assessment and rating of ataxia (SARA) [5, 10, 16, 56–59] and international cooperative ataxia rating scale (ICARS) [55, 57, 60–64]. These scales include the measurement of not only incoordination movement of the upper and lower extremities but also balance in sitting, standing, and gait [10, 56, 62]. The Berg balance scale (BBS) was most often used in previous studies to measure balance ability comprehensively [5, 16, 26, 28, 51, 54, 62, 65–70]. Timed up and go test [54, 64, 69], 10-m walk test [5, 66], 2-min walking test [62], and 6-min walking test [10] were used to measure the gait ability in previous studies. Furthermore, the functional independence measure (FIM) was often used to measure the ADL in patients with cerebellar ataxia [4, 50]. These scales are recommended and should be used to measure for severity of cerebellar ataxia and ability of balance and gait before and after physical therapy for cerebellar ataxia.

In previous studies regarding stroke rehabilitation, the parameter, which measures severity of impairment and disability, is often used to predict for functional outcome and destination from hospital [71–83]. A report indicated that continued recovery is possible in cerebellar ataxia by trauma [84], but no report on the prediction of the rehabilitation outcome in patients with degenerative cerebellar ataxia was found. Therefore, the method of prediction of rehabilitation outcome using these measures in patients with cerebellar ataxia should be studied.

3.1.2. Possible scales for ataxia and postural disorder

Another possible test has been reported in a previous study regarding ataxia and postural disorder. Force control test was conducted to test the ability of force control by muscle contraction in the extremity by using a dynamometer in the isometric testing mode with Biodex (Medical Inc., Shirley, NY) [30]. The inadequacy of force control by muscle contraction can

cause posture and gait disturbances [30]. The repetitive and alternate movement with both legs is acquired in locomotion, such as gait and pedaling. Therefore, we may measure the amplitude and speed in these motions because the amplitude and speed of repetitive motion, which involve pedaling movement with both legs, are disturbed in patients with cerebellar ataxia [85]. The performance of leg placement task has a strong relationship with gait disturbance in patients with cerebellar ataxia [86]. Subjective visual vertical (SVV) is often measured in stroke patients [87, 88] because SVV deviated to one side laterally and the bias causes dysfunction of postural control in stroke patients not only with palsy [88] but also with cerebellar ataxia [89, 90]. The reason why SVV bias occurs in cerebellar patients is the estimated contamination of vestibular disorder, but it is unclear, so further study is needed.

3.1.3. Evaluation with neuroimaging

A functional magnetic resonance imaging (MRI) study revealed that cerebellar somatotopic maps are associated with voluntary limb movement in lobules and sublobules in the rostral and caudal spinocerebellar cortex [91]. Voluntary limb movement is disturbed in patients with the atrophy in intermediate and lateral cerebellum, which can be evaluated with MRI [92–94]. Furthermore, a voxel-based lesion-symptom mapping study using MRI revealed that limb ataxia was significantly correlated with lesions of the interposed and part of the dentate nuclei and ataxia of posture and gait was significantly correlated with lesions of the fastigial nuclei, including interposed nuclei [63]. Subsequently, MRI is useful to estimate location causing incoordination movement.

3.1.4. Neurophysiological evaluation

The motor-evoked potential induced by transcranial magnetic stimulation (TMS) over the primary motor cortex is inhibited by TMS over the contralateral cerebellum [95, 96]. This cerebellar brain inhibition (CBI) [95, 97–103] is modulated in patients with cerebellar ataxia [99, 103] and in healthy adults after motor learning [32], indicating that CBI reflects excitability of the cerebellum and the function of connectivity of the cerebellum and other tissues associated with motor control and learning [104]. Therefore, CBI may be useful for estimation whether ataxia may be due to the disturbance of the input, output, or cerebellum itself and the effect of physical therapy in cerebellar plastic change [61].

The cerebellum has output to the vestibular nuclei, red nuclei, reticular formation, and functional corroboration with the brainstem [105]. The vestibulospinal, rubrospinal, and reticulospinal tracts play an important role in postural control [106]. A previous study reported the possibility that cerebellar TMS facilitates spinal reflex [107] mediated with these extrapyramidal tracts [108]. This cerebellar spinal facilitation (CSpF) [107, 108] is modulated by motor tasks [107], which require cerebellar excitation [109]. In contrast, cerebellar TMS induces long latency electromyographic response (C-LER) in the hand muscles [110–112] particularly during visually guided manual tracking tasks and in the lower muscles [113–115], which can be mediated by the vestibulospinal and reticulospinal tracts because C-LER is affected by the task-modulated excitability of these tracts [114, 115]. Therefore, CSpF and C-LER may be useful to detect the function of cerebellar output and that of the cerebellum itself.

3.2. Intervention

3.2.1. Intensive physical therapy

Three systematic reviews [10, 47, 48] and two narrative review articles [37, 116] introduced and recommended intensive physical therapy for cerebellar ataxia in patient with SCD within 1 year from diagnosis [50]. Physical and occupational therapies, which were 2 hours $\times$ 5 days + 1 hour $\times$ 2 days per week for 4 weeks, were applied to patient in the hospital. This intensive physical therapy, which focused on balance, gait, and muscle strengthening, can improve SARA score and gait speed in the hospital, but the effect was carried over only until 12 weeks after the training, but not 24 weeks [50]. In another study including the patient with a diagnosis of SCD for more than 1 year, intensive physical therapy, which was 1 hour $\times$ 3 days per week for 4 weeks, including coordinative training, improves SARA and gait ability, and the period of effect was 8 weeks post-rehabilitation [52, 53]. Therefore, intensive physical therapy that is focused on balance, gait, and muscle strengthening can be recommended for patients with SCD with cerebellar ataxia to improve their severity of ataxia and gait ability. However, these effects can be restrictive in time; hence, continuing therapy intermittently and further studies acquiring more long-term effects are necessary.

In patients with cerebellar stroke, intensive training for the upper limb with cerebellar ataxia can improve the upper function [17]. In this study, the modified constraint-induced movement therapy was conducted for patients with subacute stroke, resulting in the improvement of the upper limb function. However, whether the effect harbors for more than 1 week is not known.

The study was conducted regarding the intensive physical therapy to chronic MS patient who does not have sufficient walking ability. The patients received physiotherapy for two sessions of 45-min each week on different days for 8 weeks. The result suggests that the intensive intervention can mildly improve the upper function and mobility in chronic MS patients [49].

3.2.2. Balance training

Balance training is an important intervention for patients with cerebellar ataxia [50, 52, 53, 117]. A previous RCT study revealed that balance training with placement on the torso with less than 1.5% of patient's body weight improves gait ability in patients with cerebellar ataxia with MS [10, 54]. Small weights were applied to the torso on a specially constructed vestlike garment that allowed Velcro application of weights to the front, back, or sides of the torso between the shoulders and waist to maintain the balance of the upright posture [54]. This balance-based torso weighting (BBTW) can be useful for treatment of cerebellar ataxic gait.

In another report that is not RCT, balance training without weighting can improve gait function in patients with SCD using the PhysioTools™ General Exercises First Edition software (PhysioTools; Tampere, Finland) in own home [64]. The home-based exercise program was modified by physical therapist based on severity of ataxia, and the tools, which were chair, exercise ball, or balance disk, were applied [64]. The improvement of gait function was maintained 4 weeks after the final training [64]. This balance training can be effective at home after rehabilitation in hospital.

A rehabilitation program including foot sensory stimulation, balance, and gait training with limited vision was adapted to patients with ataxic neuropathy. The results suggest that there is a possibility that the intervention improves the motor function in patient with ataxic neuropathies [69].

Trunk stabilization and locomotor training in cerebellar ataxia can improve the balance score [66]. Furthermore, core stability exercise increases trunk stability to facilitate skilled motor behavior of the upper extremities [118].

3.2.3. *Gait training*

The gait in patients with degenerative cerebellar ataxia is disturbed [28, 29]; however, no RCT report focused on especially gait training for patients with cerebellar ataxia. Human locomotor adaptive learning is proportional to the depression of cerebellar excitability [33], and patients with ataxia with cerebellar damage have partially impaired in motor learning [14, 41, 45, 46], but some previous studies reported that gait training can improve the function of gait. The locomotive training on the ground is applied [84], and the treadmill training is applied to cerebellar ataxic gait in adults [5, 66, 119–121] and children [120], resulting to the intervention that contributes gait function improvement in this report. Furthermore, body-weight support on a treadmill can improve balance and gait function with severe cerebellar ataxia [66].

Task-oriented (disability-focused) or facilitation (impairment-based) approach was conducted to cerebellar ataxia in MS to improve gait ability [51]. Both approaches improved gait function, but no significant difference was found on the effect in both approaches. Another possible gait function improvement on cerebellar ataxia was reported; auditory feedback control therapy can improve ataxic gait in MS [122], and orthopedic shoes were used to improve gait in Friedreich's ataxia in a single case report [123].

3.2.4. *Noninvasive brain stimulation (NIBS)*

NIBS, which is repetitive TMS (rTMS) [96, 98] and transcranial direct current stimulation (tDCS) [124], over the primary motor cortex can modulate the excitability of the motor cortex [125] and is often used as an effective tool for enhancing behavioral training after stroke with hemiplegia [125–128]. Furthermore, the NIBS to the cerebellum modulates cerebellar excitability [98, 129–131], motor function [132, 133], and motor learning [32, 134–138] in healthy population. In patients with cerebellar ataxia, some previous studies reported about the effect of tDCS to the cerebellum on motor function [131, 139–141]. A double-blind, randomized and sham-controlled study revealed that cerebellar tDCS improved SARA, ICARS, nine-hole peg test, and gait speed [141]. But it is not clear whether the effect remains for a long term. In another report, tDCS over the cerebellum and contralateral motor cortex reduced postural and action tremor due to the degenerative cerebellum in patients [140]. rTMS over the cerebellum in patients with cerebellar stroke and cerebellar ataxia improves ICARS subscore on gait and posture with improvement in neurophysiological parameters, such as CBI [61]. Therefore, some recent report revealed that NIBS over the cerebellum transiently improve ataxia and gait disturbance; however, there are no report on the long-term effect in any RCT.

4. Conclusion

Gait, balance, and ADL are disturbed in patients with cerebellar ataxia owing to diseases, such as stroke, SCD, and MS. Severity of cerebellar ataxia depends on the disease. The interventions for cerebellar ataxia are medication, surgery, and physical therapy to improve and maintain their function of gait, balance, and ADL due to cerebellar ataxia. However, very few reports, which are systematic reviews, RCT studies, and meta-analysis studies, on physical therapy for cerebellar ataxia with high methodological quality and scientific evidence, were found compared with rehabilitation for hemiplegia due to stroke.

Some systematic reviews introduced and recommended intensive physical therapy more than 1 hour per day for at least 4 weeks, which focused on balance, gait, and strengthening training for degenerative cerebellar ataxia in hospital and own home. However, these effects can be restrictive in time; therefore, the therapy should be applied intermittently. To facilitate the effect, the intervention program should be modified by physical therapist based on results of evaluation using SARA, ICARS, BBS, walking test, FIM, neuroimaging, and neurophysiological assessments. The weighting on the torso, treadmill, balance ball, balance disk, and the software to instruct the exercise can improve the effect of physical therapy in hospital and own home. NIBS over the cerebellum for neuromodulation to facilitate motor learning has potential to improve the effect of physical therapy for cerebellar ataxia. Previous findings indicated that physical therapy for cerebellar ataxia is time restricted; therefore, the long-term effect and the effect of new optional neurophysiological methods should be studied in patient with stroke, SCD, and MS.

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