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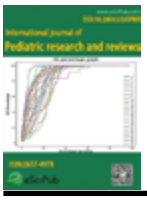

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Effect of Exercise on the Ability of Children with Cerebellar Ataxia to Perform Activities of Daily Living

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ABSTRACT

Background: People with ataxia cannot adjust the direction and intensity of movements due to poor voluntary muscle control and the lack of voluntary or reflexive muscle contraction necessary to improve position or posture problems. Ataxia is one of the main symptoms of cerebellar disorder. Most patients with cerebellar ataxia experience ataxia-related difficulties in performing activities of daily living. Exercise has positive effects on ataxia and the ability to perform activities of daily living. Balance exercises, walking exercises, muscle-strengthening exercises, activities of daily living exercises, torso weighting, etc., were reported to improve ataxia, gait capacity, and ability to perform activities of daily living in adults (including older people) with cerebellar ataxia. However, not much has been still done to clarify the effect of exercise on ataxia and the ability of children with cerebellar ataxia to perform activities of daily living. **Objective:** This review mainly aimed to add to existing knowledge and reveal the relationship between exercise and ataxia and the ability of children with cerebellum-related ataxia to perform activities of daily living. **Results:** Walking exercises, balance exercises, muscle-strengthening exercises, video games, tracking tasks, core stability exercises, and hippotherapy could improve ataxia, gross motor skills, and the ability of these children to perform activities of daily living. **Conclusion:** Given the lack of reports, studies are needed to clarify how exercise affects ataxia, gross motor skills, and the ability of children with cerebellar ataxia to perform activities of daily living.

Keywords: Cerebellum, Ataxia, Activities of daily living, Exercise, Children

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Introduction

People with ataxia cannot adjust the direction and intensity of movements due to poor voluntary muscle control and the lack of voluntary or reflexive muscle contraction necessary to improve position or posture problems [1]. Ataxia is one of the main symptoms of cerebellar disorders, and it is associated with the ability to stand, sit, and walk [1]. Most patients with cerebellar ataxia experience ataxia-related difficulties in performing activities of daily living (ADL) [2]. Children with cerebellar ataxia also experience declines in gait, transfer and stair capacity, gross motor skills, and motor developmental delays [3, 4, 5]. Therefore, treatment of symptoms including ataxia and ADL ability decline is important to maintain or improve the standing, sitting, and ADL abilities of these children.

Exercise is one of the methods that positively affect ataxia and ADL ability. Randomized controlled trials have reported that balance exercises, walking exercises, muscle-strengthening exercises, ADL exercises, torso weighting, etc., could improve ataxia, gait capacity, and ADL ability of adults (including older people) with cerebellar ataxia [6, 7, 8, 9]. The influence of exercise and physical activity on ataxia, gross motor skills, and ADL ability was also reported in children with ataxia [10]. However, not much has been still done to clarify the effect of exercise on the ataxia and ADL ability of children with cerebellar ataxia. Thus, this review mainly aimed to add to existing knowledge and reveal the relationship between exercise and ataxia and the ADL ability of children with cerebellum-related ataxia.

Functions of the cerebellum and symptoms of cerebellar disorders

The cerebellum is composed of the vermis and bilateral hemispheres [1]. The cerebellum facilitates the smooth and accurate execution of voluntary movements and adaptation to changes in motor tasks. Moreover, various combinations of complex movements have been stored accurately in the cerebellum via trial and error [11]. However, a study reported that prior experience but not the size of the error is associated with motor learning in young children [12].

Ataxia is the main symptom of cerebellar disorders, and it is often seen unilaterally on the lesion side. In patients with ataxia, they cannot adjust the direction and intensity of their movements because of their poor voluntary control of movements and the lack of voluntary or reflexive muscle contraction necessary for improving position or posture problems [1]. Impairments in the vermis and bilateral hemisphere of the cerebellum generally cause truncal and appendicular ataxia, respectively, and they affect equilibrium, standing, sitting, and gait [1]. Most patients with cerebellar ataxia experience ataxia-related difficulties in performing ADL [2]. Moreover, children with cerebellar ataxia were reported to experience dysmetria, which is one of the symptoms of appendicular ataxia and truncal ataxia, and

balance problems, decline in gross motor skills, ADL ability, and motor developmental delays [3, 4, 5, 13].

Assessment of ataxia, gross motor skills, and ADL ability

General assessments for appendicular ataxia include the nose–finger–nose test, finger–nose test, knee pat test, toe–finger test, heel–knee test, and shin-tapping test. In truncal ataxia, patients use their hands to support instability when sitting on the bed with their feet off the floor [1].

The International Cooperative Ataxia Eating Scale (ICARS) was developed to evaluate the severity of ataxia, including postural and stance disorders, limb ataxia, dysarthria, and oculomotor disorders as major categories, which are further divided into 19 minor categories. The scores range from 0 to 100 points (most severe) [14]. Furthermore, the scale for the assessment and rating of ataxia (SARA) is simple (eight items) and useful for assessing ataxia severity [15].

Regarding the assessment of motor skills and ADL ability, the gross motor function measure (GMFM), which was developed for cerebral palsy, can evaluate change over time and intervention outcomes of gross motor skills [16, 17, 18]. GMFM is scored using a total of 88 items (GMFM-88) in a 4-level Likert scale, and GMFM-66, with 66 simplified items, has been introduced [16, 17, 18]. The gross motor function classification system (GMFCS) is also a scale for the discriminative assessment of gross motor function and evaluates the severity of gross motor skills in five levels [18, 19, 20]. Moreover, the functional independence measure for children (WeeFIM) assesses the ADL ability of children, and it has a total of 18 items (motor domains, 13 items; cognitive domains, 5 items) scored from 18 (minimum) to 126 points (maximum) [18, 21, 22]. Besides, the pediatric evaluation of disability inventory (PEDI) is a tool that assesses the capacity and performance of ADL of children. PEDI evaluates functional skills (propriety of specific skills in daily living), degree of assistance from caregivers, and environmental adjustment [18, 23, 24].

Exercise for ataxia, balance, gross motor skills, and ADL ability

Exercise is a planned, structured, repetitive, and purposive physical activity to maintain or improve physical fitness, and physical activity is defined as any bodily movement produced by skeletal muscles that leads to energy consumption [25]. Randomized controlled trials have reported that balance exercises, walking exercises, muscle-strengthening exercises, ADL exercises, torso weighting, etc., improve balance and gait capacity, and SARA and FIM scores in adults (including older individuals) with cerebellar ataxia [6, 7, 8, 9]. Besides, exercise for balance and ambulation and ADL have been recommended to improve the ataxia, gait capacity, and ADL ability in people with spinocerebellar degeneration, which mainly manifests as cerebellar ataxia [26].

In children, the effect of exercise on ataxia and balance, which is related to ADL ability and gross motor skills, and ADL ability in children with cerebellum-related ataxia are mainly summarized in Table 1 (main outcomes, sample size including the number individuals in the control group, and some studies included individuals aged ≥18 years). Locomotor training with body weight support on a treadmill and overground improved the gait capacity and WeeFIM score [transfers: 3 (moderate assistance) to 6 (modified independence); walking: 2 (maximal assistance) to 5 (supervision); stairs: 1 (total assistance) to 4 (minimal assistance)] of children with severe cerebellar ataxia who had posterior fossa hemorrhage [3]. Yoo et al. (2021) also reported that robot-assisted gait training improved balance, walking capacity, and GMFM of children with ataxic cerebral palsy who had cerebellar atrophy [27]. On the other hand, task-oriented reaching activities, stretching, strengthening, and gait training resulted in a decrease in the rate of falls, while a decline in the gait speed of children with Friedreich ataxia [28].

Moreover, video games (Nintendo Wii and Xbox) were reported to improve the balance, gait parameters, and SARA scores of children with spinocerebellar ataxia [29, 30]. Multidimensional physical therapy (e.g., interventions for proximal stability, coordination of extremity movement, and balance) also had positive effects on the balance and gait parameters of children with cerebellar ataxia caused by traumatic brain injury [31]. Goal-oriented neuromuscular training (functional resistance and treadmill training, pool therapy, vibration-assisted therapy, etc.) was reported to improve gait parameters and GMFM in children with non-progressive and progressive ataxia, including cerebellum-related ataxia [32]. Besides, problem-based task training (10-step walk and return trial, walking while carrying an object, walking between parallel lines, and kicking a ball) was found to improve the balance, GMFM, ICARS, and PEDI score of children with cerebellar ataxia after brainstem glioma surgery [33]. Improvements in the SARA score, balance, GMFM, and Life Habits Questionnaire (LIFE-H) score, which is related to ADL, were also observed following balance and muscle-strengthening exercises and video games (Wii Fit Balance Board and Wii games) in children with ataxia–telangiectasia, which is characterized by symptoms such as cerebellar ataxia [34, 35].

In addition, tracking tasks (to track the movements of a target) displayed on the computer screen demonstrated a favorable influence on the dexterity of a child with ataxia after diagnosis with a cerebellar tumor [36]. Romano et al. (2022) also reported that upper body physical rehabilitation (motor activity program at home using the Niurion exergame) improved the hand dexterity, but not the SARA score and gait parameters, of children with ataxia, including cerebellum-related ataxia [37]. Furthermore, improvements in GMFM and

Table 1. Summary of the association of exercise with the ataxia, gross motor skills, and ADL ability

Study (years)	Sample size	Age (years)	Intervention	Outcomes
Harris-Love MO et al (2004) [28]	1	14	Task-oriented reaching activities, stretching, strengthening, and gait training	Decrease in the rate of falls Decline in gait speed
Cernak K et al (2008) [3]	1	13	Locomotor training using body-weight support on a treadmill and overground	Improvement of gait capacity Change of the wee-FIM score [transfers: 3 (moderate assistance) to 6 (modified independence); walking: 2(maximal assistance) to 5 (supervision); stairs: 1 (total assistance) to 4 (minimal assistance)]
Ada L et al (2009) [36]	1	5	Tracking tasks (to track movements of a target)	Improvement of dexterity
Frank A et al (2011) [4]	1	6	Hippotherapy	Improvement of the GMFM and PODCI scores
Ilg W et al (2012) [29]	10	11-20	Xbox coordinative training	Improvement of balance, gait parameters, and SARA score
Synofzik M et al(2013) [34]	1	10	Nintendo Wii games	Improvement of the SARA score
Sartor-Glittenberg C et al (2014) [31]	3	16-22	Multidimensional physical therapy	Improvement of balance and gait parameters
Schatton C et al (2017) [30]	10	6-29	Nintendo Wii and Microsoft Xbox Kinect	Improvement of the SARA score
Martakis K et al (2019) [32]	45	3-22	Goal-oriented neuromuscular training	Improvement of gait parameters and GMFM
Yoo M et al (2021) [27]	2	11-12	Robot-assisted gait training	Improvement of balance, walking capacity, and GMFM
Lee YS et al (2021) [33]	1	5	Problem-based task training	Improvement of balance, ICARS and PEDI score, and GMFM
Unes S et al (2021) [35]	1	9	Balance and strength exercises and Wii Fit balance-based video games training	Improvement of balance, GMFM, and the LIFE-H score
Elshafey MA et al (2022) [13]	40	5-9	Core stability exercises	Improvement of balance, coordination, and SARA score
Romano A et al (2022) [37]	18	5-17	Upper body physical rehabilitation	Improvement of the dexterity No significant change of SARA score (significant increment of SARA score in control group) No significant change of gait parameters (minor improvement)

ADL: Activities of Daily Living, GMFM: Gross Motor Function Measure, ICARS: International Cooperative Ataxia Rating Scale, LIFE-H: Life Habits Questionnaire, PEDI: Pediatric Evaluation of Disability Inventory, PODCI: Pediatric Outcomes Data Collection Instrument, SARA: Scale for the Assessment and Rating of Ataxia, Wee-FIM: Functional Independence Measure for Children

pediatric outcomes data collection instrument score, which includes upper extremity and physical functions, transfers, and basic mobility-related domains, were observed following hippotherapy in children with cerebral palsy and mild ataxia, which is caused by a congenital malformation of the cerebellum [4]. Elshafey et al. (2022) also reported that core stability exercises improved the SARA score, balance, and coordination of children with cerebral palsy and cerebellar ataxia [13]. Thus, walking exercises, balance exercises, muscle-strengthening exercises, video games, tracking task, core stability exercises, and hippotherapy could improve ataxia, balance, gross motor skills and ADL ability in children with cerebellum-related ataxia. However, relationships between exercise and ataxia and ADL ability should be further clarified in future studies given the scarcity of reports in children with cerebellar ataxia.

Conclusion

Walking exercises, balance exercises, muscle-strengthening exercises, video games, tracking tasks, core stability exercises, and hippotherapy could improve ataxia, balance, gross motor skills, and ADL ability of children with cerebellum-related ataxia. Given the lack of sufficient reports, studies are needed to clarify how exercise affects ataxia, gross motor skills, and ADL ability to maintain or improve the capacity of daily life in children with cerebellar ataxia.

Competing interests

The author declares that they have no competing interests

Abbreviations:

ADL, Activities of Daily Living

ICARS, International Cooperative Ataxia Rating Scale

FIM, Functional Independence Measure

GMFCS, Gross Motor Function Classification System

GMFM, Gross Motor Function Measure

LIFE-H, Life Habits Questionnaire

PEDI, Pediatric Evaluation of Disability Inventory

PODCI, Pediatric Outcomes Data Collection Instrument

SARA, Scale for the Assessment and Rating of Ataxia

SCD, Spinocerebellar Degeneration

Wee-FIM, Functional Independence Measure for Children

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