

Exercise programme intervention for persons with motor ataxia Programa de ejercicios de intervención para personas con ataxia motora

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Abstract. Introduction: Ataxia affects the nervous system by decreasing balance and coordination most mostly in the trunk, arms and legs. Physical activity is used to help lowering the symptoms of this disease. Objective: The study objective aims to determine the effects of an exercise program intervention focused on quality of life of this person in terms of body composition, hemodynamic parameters and functional capacity. Methods: A Longitudinal study-case with the exercise sessions performed in a gymnasium with a specialized Personal Trainer. A 43 years old male individual with motor ataxia as participated with a pre and post-test assessments with an exercise program comprised sessions twice a week of 30 minutes each, for 6 months, focused on cardio-fitness, strength and body stabilization. The Scale for the Assessment and Rating of Ataxia was applied to evaluate Gait, Stance, Sitting, Finger Chase, Nose-finger Test, Fast alternating hand movements and Heel-chin slide, and the program SPSS (v20) to present descriptive statistics to express the participant's improvements. Results: This study indicates that rehabilitation can improve health, well-being and life quality improvements in individuals with ataxia, however, a larger study is required to have analyse if these variables would suffer significant changes in all participants. Besides the improvements in body composition and blood pressure, benefits in all variables of SARA scale were observed, except in «Sitting» were the value remained unchanged. Conclusions: This type of exercise programme intervention can promote capacity and health-related quality of life. This study provides evidence for maintaining physical activity programs in patients with ataxia.

Keywords: ataxia, motor development, health, exercise programme.

Resumen. Introducción: La ataxia afecta al sistema nervioso al disminuir el equilibrio y la coordinación, sobre todo en el tronco, los brazos y las piernas. La actividad física se utiliza para ayudar a disminuir los síntomas de esta enfermedad. Objetivo: El objetivo del estudio es determinar los efectos de una intervención de un programa de ejercicio enfocado a la calidad de vida de esta persona en términos de composición corporal, parámetros hemodinámicos y capacidad funcional. Métodos: Un estudio-caso longitudinal con las sesiones de ejercicio realizadas en un gimnasio con un Personal Trainer especializado. Un varón de 43 años con ataxia motora que participó en una evaluación pre y post prueba con un programa de ejercicios compuesto por sesiones dos veces por semana de 30 minutos cada una, durante 6 meses, centradas en el cardio-fitness, la fuerza y la estabilización corporal. Se aplicó la Escala para la Valoración y Clasificación de la Ataxia para evaluar la Marcha, la Postura, la Sentada, la Persecución con los Dedos, la Prueba de los Dedos de la Nariz, los Movimientos Alternados Rápidos de la Mano y el Deslizamiento del Talón-Mentón, y el programa SPSS (v20) para presentar estadísticas descriptivas para expresar las mejoras del participante. Resultados: Este estudio es indicativo que la rehabilitación puede mejorar la salud, el bienestar y la calidad de vida de los individuos con ataxia, sin embargo, se requiere un estudio más amplio para analizar si estas variables sufrirían cambios significativos en todos los participantes. Además de las mejoras en la composición corporal y la presión sanguínea, se observaron beneficios en todas las variables de la escala de SARA, excepto en «Sentado», donde el valor permaneció sin cambios. Conclusiones: Esta intervención realizada con estos ejercicios puede promover la capacidad y la calidad de vida relacionada con la salud. Este estudio proporciona evidencia para mantener programas de actividad física en pacientes con ataxia.

Palabras clave: ataxia, desarrollo motor, salud, programa de ejercicio.

Introduction

Ataxia can be defined as a change in the accurate movement coordination and can be a symptom of many diseases processes in general (Thakkar, Maricich & Alper, 2016). Ataxia can be described by different ways, such as, weakness, dizziness, stroke, falls or other non-specific major complaint. Classically, ataxia is associated with cerebellar dysfunction, but may occur due to different problems with a change in sensitivity that may cause sensorial ataxia. Huff (2016) reported that motor coordination performance is altered, even if motor systems and the cerebellum are intact. Sensory ataxias can be compensated by visual sensory information. However, loss of visual information leads to the observation that sensory ataxias often worsen in poor lighting conditions and may be exhibited during the examination. Motor ataxia is based on unstable and irregular steps, and

the lack of compensation for barriers in the environment may be insufficient. The gait of sensory ataxia resulting from the loss of proprioception is notable for the abnormal movement of the legs and the impact of the feet at each step (Bastian & Keller, 2014). Improvements were found in a study of Ilg, Synofzik, Brotz, Burkard & Giese (2009) where it was observed locomotor performance in people with cerebellar ataxia after a six-week home balance exercise program. A total of 14 patients with cerebellar ataxia participated in a 6-week individualized home-based balance exercise program and attended 5 testing sessions using the Scale for the Assessment and Rating of Ataxia (SARA). In the pre-training, post-training, and follow-up testing, assessments of balance and walking were included. Participants revelled in post-hoc comparisons significant rehabilitative improvements over the 6-week training period, but TUG (Timed-Up and Go) was retained 1 month later. There were no changes across other measures for the group. This study indicated that improvements in walking speed were affected by the level of balance challenge, but not by age, ataxia severity, proprioception, or duration of exercise.

Changes resulting from motor ataxia include problems of balance and motor coordination involving the trunk and limbs. Balance difficulties and inadequate posture generate difficulties in the execution of daily activities directly affect the functional capacity and quality of life of the patient (Armutlu & Karabudak, 2001; Castillo, Gómez-Carmona, Reche, Gil & Ortega, 2018).

According to these studies, an exercise program should include: 1) balance exercises to improve or maintain balance and stability during activities, such as, sitting, standing, walking and moving, and motor coordination exercises incorporated into functional activities of daily living (Abellán & Sáez-Gallego, 2015); 2) muscle strengthening carefully supervised to avoid fatigue. The number of repetitions, the amount of weight used and rest periods should be individually prescribed and adapted periodically. The patient's own weight should be used for global strengthening; strengthening the proximal musculature of the shoulders and hip is important to maintain functional use of upper and lower limbs; 3) physical resistance through cardiovascular exercises, such as walking on the treadmill and or bike exercises. Water exercises offer less limitation and can be beneficial; 4) a specific strategy for each individual, i.e., for patients with walking capacity and loss of proprioception, it is important to encourage compensation with visual feedback, such as looking at the feet while walking. The use of weight at the extremities may be indicated to increase sensory information; 5) Train independent functions, always considering ways to reduce energy effort and monitoring «sitting» and «standing» posture. Patients who do not wander in the community should be encouraged to maintain maximum mobility at home and to stand with support for weight bearing on lower limbs; 6) Evaluate the need, indicate and train the use of auxiliary means with the purpose of giving security to the transfers and locomotion. Assistance equipment with grab bars, crutches and wheelchairs may be important to compensate for the loss of coordination and strength required in different activities; 7) Maintain joint range of motion through postures and stretching exercises to prevent deformities (Robles-Rodríguez, Abad-Robles, Fuentes-Guerra & Benito-Peinado, 2017; Gomez-Marcos & Sanchez-Sanchez, 2019); 8) Monitor respiratory function by associating specific exercises as needed. Relaxation techniques may also be indicated. During balance training, the patient must be exposed to a movement or sequence of movements during which he is stimulated to maintain stability, (Gregorio, Pérez & Moro, 2019). This activity should be done slowly, in order to maintain a proper posture and to the maximum degree where the balance is required to «stop» and «balance» for a few seconds (Armutlu & Karabudak, 2001). Other study Ilg, Brotz, Murkard, Giese, Schols & Synofzik (2010) evaluated a rehabilitation programme with 26 patients with ataxic multiple sclerosis. It comprised sessions of coordination exercises, balance and walking over the ground three times a week for four weeks. In the end of this programme, balance, gait parameters and the Expanded Disability Status Scale (EDSS) scores were improved. Weyer, Abele, Schmitz-Hubsch, Schoch, Frings & Timmann (2007) evaluated a rehabilitation programme comprising static and dynamic balance exercises and coordination exercises (with three one-hour sessions a week

for four weeks) with 16 patients. Improvements in SARA scale was observed and the effect lasted for up to a year at least, Weyer et al. (2007). Therefore, this study aims to determine the effects of an exercise programme intervention focused on well-being, including body composition, hemodynamic parameters and functional capacity developed for an adult suffering from motor ataxia.

Material and Methods

Participant

We have studied one individual of male gender with 43 years old. This is a study-case with pre, post-test and follow-up assessments with an exercise program with sessions twice a week of 30 minutes each, for 6 months, focused on cardio-fitness, strength, body stabilization and neurologic evaluations in an indoor gymnasium. The follow-up test were made one month after this programme. The individual has this diagnostic confirmed by medical report, after neurologic evaluations and loss of coordination that lead to this conclusion.

Instruments and Procedures

We have used the Scale for Assessment and Rating Ataxia (SARA) to evaluate Gait, Stance, Sitting, Finger Chase, Nose-finger Test, Fast Alternating Hand Movements and Heel-Chin Slide. This scale is reliable and has been validated (Weyer, Abele, Schmitz-Hubsch, Schoch, Frings & Timmann, D. (2007). A specialized physical therapist trained the SARA instrument one month before the first assessment and was responsible for the measurements.

The training sessions included exercises, carried out with increasing load, such as, dorsal rowing (3x: 8,10,12 reps), chest press (3x: 8,10,12 reps), shoulder press (3x: 8,10,12 reps), spinning in bicycle (15m), assisted treadmill walking (15m), astatic body stabilization (2x: 1m), gluteus stabilization (2x: 15 reps), leg press (3x: 8,10,12 reps), leg extension (3x: 8,10,12 reps), butterfly (3x: 8,10,12 reps), inverted butterfly (3x: 8,10,12 reps), squat with and without load (3x: 8,10,12 reps to 12, 15, 20 reps depending on the load), elliptical (3x: 10,15,20 reps), trunk elevation in a inclined platform and with medical ball (2x: 15 reps), frontal swing with kettlebell (3x: 8,10,12 reps), small rope jumping (5x: 30s), mountain climbing elliptical (2x: 30s), jumping jacks (5x: 30 reps), deadlifts with bars (3x: 10,15,20 reps), TRX rowing (3x: 10,15,20 reps), Russian Twist with medical ball (2x: 30 reps), hack squats (3x: 10,15,20 reps), hand push-ups (3x: 10,15,20 reps), wall ball and lunges with discs on the head (3x: 20,25,30 reps). All these exercises were not applied in the same session, were developed during all the sessions practiced depending on the objective of each session. The effort intensity was kept approximately at 80% of the participant heart rate. Also, Borg Scale was used by the participant to manage his own effort, and in case of extreme fatigue or any signs of discomfort the session was interrupted. The participant's body composition was evaluated by bio-impedance (Tanita, Germany) and blood pressure was measured using a digital sphygmomanometer (HEM-Omron 907, Sweden). These evaluations were made in the morning before the training sessions and the blood pressure measurements were made on the left arm.

Table 1
Evaluation criteria of classification and methodology of the variables analysed.

| Variable | Methodology | Classification |
|---------------------------------|--|--|
| Gait | (1) to walk at a safe distance parallel to a wall including a half-turn (turn around to face the opposite direction of gait) and; (2) to walk in tandem (heels to toes) without support. | From 0 ("Normal", no difficulties in walking) to 8 ("Unable to walk", even supported). |
| Stance | to stand (1) in natural position, (2) with feet together in parallel (big toes touching each other) and (3) in tandem (both feet on one line, no space between heel and toe). | For each condition, three trials were allowed. Best trial was rated, varying between 0 ("Normal", able to stand in tandem for > 10 s) and 6 ("Unable to stand" for >10 s even with constant support of one arm). |
| Sitting | the proband had to sit on an examination bed without support of feet, eyes open and arms outstretched to the front. | 0 ("Normal", no difficulties sitting >10 sec) and 4 ("Unable to sit" for >10 s without continuous support). |
| Finger Chase | Rated separately for each side, the proband had to seat comfortably. If necessary, the support of feet and trunk was allowed. The examiner seated in front of the proband and performed 5 consecutive sudden and fast pointing movements in unpredictable directions in a frontal plane at about 50 % of the proband's reach. Movements had an amplitude of 30 cm and a frequency of 1 movement every 2 seconds. Proband was asked to follow the movements with his index finger, as fast and precisely as possible. | Average performance of last 3 movements was rated varying between 0 (No dysmetria) and 4 (Unable to perform 5 pointing movements) |
| Nose-Finger Test | (rated separately for each side), the proband had to seat comfortably. If necessary, the support of feet and trunk was allowed. The proband was then asked to point repeatedly with his index finger from his nose to the examiner's finger, which was in front of the proband at about 90 % of proband's reach. Movements were performed at moderate speed. | Average performance of movements were rated according to the amplitude of the kinetic tremor. This item varied between 0 (No tremor) to 4 (Unable to perform 5 pointing movements). |
| Fast Alternating Hand Movements | (Rated separately for each side), the proband seated comfortably. If necessary, the support of feet and trunk is allowed. The proband was asked to perform 10 cycles of repetitive alternation of pro- and supinations of the hand on his/her thigh as fast and as precise as possible. This movement was demonstrated by the examiner at a speed of approximately 10 cycles within 7seconds. Exact times for movement execution had to be taken. | between 0 (Normal, no irregularities, performs <10s) and 4 (Unable to complete 10 cycles). |
| Heel-Chin Slide | (Rated separately for each side), the proband lied on the examination bed without sight of his legs. He was asked to lift one leg, point with the heel to the opposite knee, slide down along the shin to the ankle, and lay the leg back on the examination bed. The task was performed 3 times. Slide-down movements should be performed within 1s. | The task was performed 3 times. Slide-down movements should be performed within 1 s. If proband slides down without contact to shin in all three trials, rate 4. It varies from 0 (Normal) and 4 (Unable to perform the task). |

Ethical procedures

The individual was recruited by the investigators from a group of individuals with special needs, that perform physical activities in the gym. The study was approved by the Scientific Board of the Polytechnic Institute and was given full informed consent in writing by the individual.

Statistical Procedures

Program SPSS (v20) was applied to present descriptive statistics to express variables improvements.

Results

According to our findings, the participant made improvements in all variables under study, except for sitting and finger chase that remain equal. Improvements in gait and stance (two aspects directly related to body and posture control) were observed. The improvements in gait, stance and posture after the six-month exercise program are similar in magnitude to those seen after an intensive program with direct intervention previously conducted, Weyer et al. (2007).

literature, rehabilitation will not prevent progression of cerebellar degeneration, and future longitudinal training studies should optimize functions throughout and monitor this disease. Additionally, our program, through SARA evaluation, included sitting and standing balance exercises. For other individuals who are no longer able to walk, the use of this type of exercises is recommendable such as the exercises used by other authors (Ilg et al., 2009). A larger mobility in later stages of cerebellar degeneration will improve patients' quality of life (Kim, Lim, Lee & Koh, 2010) Also, trunk weighting has been recommended (Cernak, Stevens, Price & Shumway-Cook, 2008) and was studied in a randomized trial in 36 patients with ataxia. Trunk weighting of 1.5% (less than ours) improves balance and gait and involves an increase in contributions to movement control (including postural control) and a mechanical reduction in coordination disorders (Freund & Stetts, 2010; Castillo et al. 2018). We provided in our study an exercise on a treadmill that can be an important tool as well. Twenty minutes of treadmill training three times a week improved gait parameter in two patients with cerebellar syndrome (Cernak et al., 2008; Freund & Stetts, 2010). After the exercise program, less assistance with gait was required and the walking distance had increased. These case studies can be confirmed and supported by ours due to the intensive and repeated training, has it was found in Milne, Corben, Georgiou-Karistianis, Delatycki & Yiu (2017) that exercise interventions included coordination and balance training, balance exercises, respiratory muscle training, and treadmill training improved the quality of life of the participants.

In addition to these studies, Marques, Ferreira, Silva, Bezerra & Rabay (2018) evaluated also a study case of an individual with 20 years of age with the main objective of analyse the benefits of strength training. The benefits can be seen in aspects such as walking that previously was not possible without the help of people, walkers and even from a wheelchair, today you do not need any assistance for short walks. Through the stabilization of the body segments, by the balance of muscular strength and dynamic postural balance, which can achieve a more functional gait. The sitting and standing which also needed help, was able to sit and stand up alone. Thus, proving the effectiveness of strength training to improve motor coordination of limbs and trunks,

Table 2
Characteristics of the participant at the 1st and 2nd assessments.

| Variables | N | 1 st | 2 nd |
|----------------------------------|---|-----------------|-----------------|
| Body fat (%) | 1 | 12,0 | 8,1 |
| Body muscle Mass (%) | 1 | 54,4 | 65,5 |
| Body water (%) | 1 | 58,6 | 63,0 |
| Body visceral Fat (%) | 1 | 6,0 | 3,0 |
| Systolic Blood Pressure (mm/Hg) | 1 | 138,0 | 116,0 |
| Diastolic Blood Pressure (mm/Hg) | 1 | 112,0 | 84,0 |
| Bone Mass (%) | 1 | 3,2 | 3,6 |
| Right Arm Fat Mass (%) | 1 | 14,0 | 11,0 |
| Left Arm Fat Mass (%) | 1 | 13,5 | 10,0 |
| Right Leg Fat Mass (%) | 1 | 13,7 | 10,1 |
| Left Leg Fat Mass (%) | 1 | 15,6 | 11,7 |
| Trunk Fat Mass (%) | 1 | 19,0 | 15,4 |
| Right Arm Muscle Mass (%) | 1 | 3,1 | 3,5 |
| Left Arm Muscle Mass (%) | 1 | 3,2 | 3,7 |
| Right Leg Muscle Mass (%) | 1 | 9,3 | 10,1 |
| Left Leg Muscle Mass (%) | 1 | 8,4 | 9,5 |
| Trunk Muscle Mass (%) | 1 | 30,4 | 35,2 |
| Gait | 1 | 8,0 | 5,0 |
| Stance | 1 | 3,0 | 2,0 |
| Sitting | 1 | 0,0 | 0,0 |
| Finger Chase | 1 | 1,0 | 1,0 |
| Nose Finger Test | 1 | 2,0 | 1,0 |
| FAHM | 1 | 1,0 | 0,0 |
| HSS | 1 | 1,0 | 0,0 |

Discussion

It is valuable that an assisted training program can be very effective. Determining the appropriate amount of intervention is an important step, because according to

giving them a better and better lifestyle. Also through strength training you can observe the regression of ataxia symptoms, such as limb tremors, imbalances that were constant and after training are more controlled.

Salci, Fil, Armutlu, Yildiz, Kurne, Aksoy, Nurlu & Karabudak (2019) investigated the effects of different exercise protocols for 42 ataxia patients with multiple sclerosis. Participants were divided into three different groups: a balance training (BT) group, a lumbar stabilization (LS) group and a task-oriented training (TT) group. All groups have received balance training, and additionally, the LS group also received specific lumbar stabilization exercises. The TT group received task-oriented training. All these groups had 18 training sessions. The results showed significant improvements, however balance training alone is not enough for the rehabilitation of these patients. A combination of lumbar stabilization exercises or task-oriented training favours the values of balance rehabilitation.

Conclusions

This study showed improvement in all measures of mobility and body composition in the patient studied after a six-month training program. Individualizing the training program was not critical, as our results suggest that were important for improving these characteristics with an increasing exercise frequency. In table 2 its observable that all body composition variables have improved in the second evaluation. Continued training and progression of the program may be necessary to see optimal retention of results. Although, techniques such as treadmill exercises and others mentioned, with supported body weight and trunk exercises, appear to be valuable. This literature is limited by the small sample sizes, however this will be the challenge for incoming studies and we sure made another contribution towards a better life quality of patients suffering from ataxia.

Study limitations

This investigation, being a case study, has some limitations, such as: evaluating only one participant, which does not allow extrapolating the results for a better comparison between more generic results. The follow-up period could be longer to check if the results obtained during the program are maintained for a longer period. In addition to assessments at the level of motor capacity and body composition, it would be interesting in future studies to evaluate the individual in terms of personal satisfaction, in addition to the improvements presented, to analyse whether in terms of his personal status combined with these improvements, what is his personal and psychological perception regarding these improvements.

Acknowledgments

Nothing to report.

References

Abellán, J. & Sáez-Gallego, N. (2015). Rendimiento de los deportistas con discapacidad intelectual en pruebas motrices. Diferencias en función de la edad y el género. *Retos*, 27, 40-44.
 Armutlu, R. & Karabudak, G. (2001). Physiotherapy approaches

in the treatment of ataxic multiple sclerosis: a pilot study. *Neurorehabilitation Neural Repairment*, 15, 203-211
 Bastian, A. & Keller, J. (2014). A Home Balance Exercise Program Improves Walking in People with Cerebellar Ataxia. *Neurorehabilitation Neural Repairment*, 28(8):770-778.
 Castillo, A., Gómez-Carmona, C., Reche, P., Gil, P. & Ortega, J. (2018). Trunk stability assesment using an inercial device. *Retos*, 33, 199-20.
 Cernak, V., Stevens, R., Price, A. & Shumway-Cook, L. (2008). Locomotor training using body-weight support on a treadmill in conjunction with ongoing physical therapy in a child with severe cerebellar ataxia. *Physical Therapy*, 88: 88-97.
 Freund, J. & Stetts, D. (2010). Use of trunk stabilization and locomotor training in an adult with cerebellar ataxia: a single system design. *Physiotherapy Theory and Practice*, 26: 447-458
 Gomez-Marcos, G. & Sanchez-Sanchez, M. (2019). Description and differences in the psychological variables related to sports performance of triathletes and para-triathletes. *Retos*, 36, 22-25.
 Gregorio, M., María, P. & Moro, I. (2019). Analysis of the relationships between Developmental Coordination Disorder (DCD) and Attention Deficit and Hyperactivity Disorder (ADHD) in school age. *Retos*, 36, 625-632.
 Huff, J. (2016). *Ataxia and gait disturbances*. London: Emergency Medicine.
 Ilg, W., Synofzik, D., Brotz, S., Burkard, M. & Giese, L. (2009). Intensive coordinative training improves motor performance in degenerative cerebellar disease. *Neurology*, 73:1823-1830
 Ilg, W., Brotz, S., Burkard, A., Giese, L., Schols, M. & Synofzik, T. (2010). Long-term effects of coordinative training in degenerative cerebellar disease. *Movements Disorders*, 25: 2239-2246
 Kim, J., Lim, S., Lee, S. & Koh, L. (2010). Usefulness of the Scale for the Assessment and Rating of Ataxia (SARA) in ataxic stroke patients. *Annals of Rehabilitation Medicine*, 35: 772-780
 Marques, K., Ferreira, A., Silva, P., Bezerra, J., Rabay, J. (2018). A influência do treino de força para indivíduo atáxico: um estudo de caso. *Revista Diálogos em Saúde*, 2, 93-106
 Milne, S., Corben, L., Georgiou-Karistianis, N., Delatycki, M. & Yiu E. (2017). Rehabilitation for Individuals With Genetic Degenerative Ataxia: A Systematic Review. *Neurorehabilitation Neural Repair*, 31(7), 609-622
 Robles-Rodríguez, J., Abad-Robles, M., Fuentes-Guerra, J. & Benito-Peinado, J. (2017). Adapted sports as a contribution to education in values and the improving motor skills: the opinion of high school students. *Retos*, 31, 140-144.
 Salci, Y., Fil, A., Armutlu, K., Gökçem, Y., Kurne, A., Aksoy, S., Nurlu, G. & Karabudak, G. (2019). Effects of different exercise modalities on ataxia in multiple sclerosis patients: a randomized controlled study. *Journal of Disability and Rehabilitation*, 39(26), 2626-2632
 Thakkar, K., Maricich, M. & Alper, G. (2016). Acute Ataxia in Childhood: 11-Year Experience at a Major Pediatric Neurology Referral Centre. *Journal of Childhood Neurology*, 31:1156-1170.
 Weyer, A., Abele, M., Schmitz-Hubsch, T., Schoch, B., Frings, M. & Timmann, D. (2007). Reliability And validity of the scale for the assessment and rating of ataxia: a study in 64 Ataxia patients. *Movement Disorder*, 22:1633-7