# Physical therapy approach to spinocerebellar ataxia: a systematic review

*Abordagem fisioterapêutica da ataxia espinocerebelar: uma revisão sistemática*

*El abordaje fisioterapéutico de la ataxia espinocerebelosa: una revisión sistemática*

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**ABSTRACT |** The spinocerebellar ataxia (SCA) is an inherited disorder that leads to progressive degeneration of the cerebellum and its pathways with impairments of balance and other functions. Physical therapy studies for SCA treatment and their methodological quality were examined. We also investigated if the benefits achieved with treatment can be retained. The interventions identified included balance, gait and coordination training; strengthening; weights around the limbs during exercise and transcranial magnetic stimulation. The long-term improvements were related to the degree of SCA evolution and the continuity of exercise practice. Nevertheless, further studies with higher scientific accuracy are necessary to elect the best physical therapy approaches for SCA patients.

**Keywords |** spinocerebellar ataxia; physiotherapy; hereditary ataxia.

**RESUMO |** A ataxia espinocerebelar (SCA) é uma afecção hereditária que cursa com a degeneração progressiva do cerebelo e suas vias, causando alterações do equilíbrio e de outras funções. O efeito das abordagens da fisioterapia no tratamento da SCA e a qualidade metodológica desses estudos foram analisados. Foi investigado ainda se os benefícios alcançados com o tratamento são retidos. As intervenções encontradas incluem treino do equilíbrio, marcha e coordenação; fortalecimento; caneleiras nos membros durante exercícios e aplicação de estimulação magnética transcraniana. A retenção das melhoras obtidas com o tratamento foi relacionada ao grau de evolução da SCA e à continuidade da prática de exercícios. Porém, novos estudos com maior rigor científico são necessários para eleger as abordagens mais adequadas para o tratamento de portadores de SCA.

**Descritores |** ataxias espinocerebelares; fisioterapia; ataxias hereditárias.

**RESUMEN |** La ataxia espinocerebelosa (SCA) es una afección hereditaria que cursa con la degeneración progresiva del cerebelo y de sus vías, lo que causa alteraciones del equilibrio y de otras funciones. El resultado de los abordajes de la fisioterapia en el tratamiento de la SCA y la cualidad metodológica de estos estudios fueron analizados. Se investigó si los beneficios alcanzados con el tratamiento fueron retenidos. Las intervenciones encontradas incluyen entrenamiento del equilibrio, marcha y coordinación; fortalecimiento; canilleras en los miembros durante ejercicios y aplicación de la estimulación magnética transcraneana. La retención de las mejorías obtenidas con el tratamiento fue relacionada al grado de evolución de la SCA y a la continuidad de la práctica de ejercicios. Aunque nuevos estudios con mayor carácter científico son necesarios para elegir los abordajes más adecuados para el tratamiento de los portadores de la SCA.

**Palabras clave |** ataxias espinocerebelosas; fisioterapia; ataxias hereditarias.

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## INTRODUCTION

The first report of autosomal dominant cerebellar ataxia (ADCA) was made by Pierre Marie in 1893[1]. Since then, several classifications for these ataxias have been proposed by different authors. Harding's classification[2] became widely accepted, grouping the ADCAs for their clinical characteristics (ADCA I to III). The evolution of genetic research enabled the connection between the clinical disorders of ataxia and the affected chromosomal locus. After the description of the first defective gene to cause ADCA, each new defective gene was numbered[3], and ADCA began to be known as spinocerebellar ataxia (SCA). The current classification based on genetic changes comprehends 31 types of SCA[4]. Its prevalence is of 0.9 to 3:100,000, ranging according to type and continent[5]. SCA 1, 3 and 6 are the most common ones throughout the world[1]. In Brazil, SCA3, also known as Machado-Joseph disease, is the most prevalent type[6-8].

All kinds of SCA cause progressive cerebellar degeneration, but each type has additional clinical characteristics related to the other affected regions, especially by type (brain stem, cortex, bone marrow, cranial nerves and diencephalon)[3]. For instance, SCA7 is followed by visual loss, and SCA4, of sensory ataxia[9]. The degeneration caused by SCA, among other problems, leads to balance and gait changes, which increase the risk of falls. In fact, Van de Warrenburg et al.[10], in a study with 42 patients with SCA, demonstrated that 93% of them reported one or more falls in the period of one year. These falls can induce a vicious circle of immobility and the fear of falling[10]. The progressive character of SCA, added to this vicious circle, leads to the accumulation of motor loss, thus producing an inexorable scenario of immobility and wheelchair dependency around the 15$^{th}$ year of disease evolution[9].

There is evidence concerning an important contribution of the cerebellum in the motor learning process[11-15]. Several studies have demonstrated that patients with cerebellar lesions may present impairment in the learning of motor skills[16-18]. Therefore, some authors question if the rehabilitation of patients with SCA could have any positive effect on their motor condition. The validity of rehabilitation in such cases becomes even more controversial, given the progressive aspect of the disease: it is questionable if the possible functional gain obtained with physical therapy intervention can be maintained in the long term.

In this context, the objectives of this review are: (1) to identify the current physical therapy approach for the treatment of SCA; (2) to analyze the methodological quality of the available studies concerning the treatment of motor sequels caused by SCA; (3) to assess if the benefits from the treatment can be retained in the long term.

## METHODOLOGY

A systematic literature review was conducted in the electronic data bases MEDLINE, SciELO, PEDro, Cochrane and Scopus, using the following MeSH descriptors, with their respective translations to Portuguese: "spinocerebellar ataxia", "spinocerebellar degenerations", "Machado-Joseph disease", "progressive ataxia", "rehabilitation" and "physiotherapy". Each search was performed with at least two combined descriptors so that one of them would be related to the pathology (ex.: "spinocerebellar ataxia"; "Machado-Joseph disease") and another one to the treatment (ex.: "rehabilitation"; "physiotherapy"). The search was complemented by related references cited by the found publications.

Clinical trials published in Portuguese, English and Spanish from 2000 on were included. There was no restriction as to the type of used physical therapy approach. Identification, selection and the posterior quality assessment of the article by means of the physiotherapy evidence database scale (PEDro)[19] was made by two researchers, independently. The articles were included when they received at least five out of the total of ten points in the PEDro scale.

## RESULTS

In the conducted search, 252 studies were found. Out of these, 246 were excluded for not meeting the inclusion criteria. Among the six eligible studies, one of them did not reach the minimum score for inclusion (Figure 1 and Table 1). Therefore, five articles were included in this review.

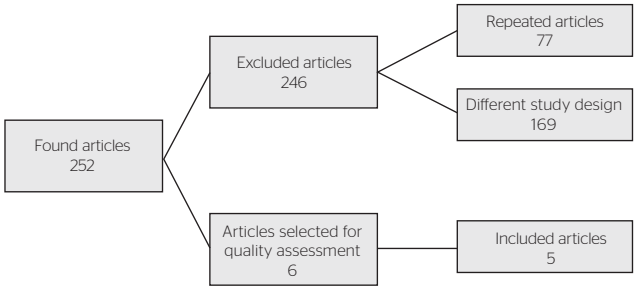
Ilg et al.[20] performed training of balance, coordination, amplitude of movement and falls prevention in six patients with sensitive ataxia and in ten patients with cerebellar ataxia (three SCA6 and six with other forms

Table 1. Quality assessment by the Physiotherapy Evidence Database Scale

| Item | Author | | | | | |
|---|---|---|---|---|---|---|
| | Ilg et al.[20] | Ilg et al.[21] | Shiga et al.[22] | Miyai et al.[23] | Dias et al.[24] | Pérez-Ávila et al.[25] |
| Eligibility criteria were specified | No | No | No | Yes | Yes | No |
| Subjects were randomly allocated to groups | No | No | No | Yes | Yes | No |
| Allocation was concealed | No | No | No | Yes | No | No |
| The groups were similar at baseline regarding the most important prognostic indicators | Yes | Yes | No | Yes | Yes | No |
| There was blinding of all subjects | No | No | Yes | No | No | No |
| There was blinding of all therapists who administered the therapy | No | No | No | No | No | No |
| There was blinding of all assessors who measured at least one key outcome | Yes | Yes | Yes | No | No | No |
| Measures of at least one key outcome were obtained from more than 85% of the subjects initially allocated to groups | Yes | Yes | Yes | Yes | Yes | Yes |
| All subjects for whom outcome measures were available received the treatment or control condition as allocated or, where this was not the case, data for at least one key outcome was analysed by "intention to treat" | Yes | Yes | Yes | Yes | Yes | Yes |
| The results of between-group statistical comparisons are reported for at least one key outcome | Yes | Yes | Yes | Yes | Yes | No |
| The study provides both point measures and measures of variability for at least one key outcome | Yes | Yes | Yes | Yes | Yes | Yes |
| Total | 6 | 6 | 6 | 7 | 6 | 3 |

of ataxia). Training was conducted three times a week for four weeks (1h/day). After four weeks, patients were training at home for eight weeks. Scales assessing the staging of ataxia were used (impairment severity): SARA (Scale for the Assessment and Rating of Ataxia) and ICARS (International Cooperative Ataxia Rating Scale); for the treatment objectives of the patients, GAS (Goal Attainment Score) was used; for quantitative gait analysis (kinemetrics) and balance assessment (stabilometry, Berg scale and anticipatory reaction time). After practice, the staging of the disease improved for all of the patients. Only the group with cerebellar ataxia demonstrated significant balance improvement, increased velocity and step length, maintaining the benefits after eight weeks. This retention was not correlated with the staging of ataxia.

To assess the long term efficacy of this protocol, Ilg et al.[21] compared the results before and after 4 weeks of training with the results presented after 1 year, by 14 out of the 16 patients analyzed in the prior study. The patients performed the same exercises at home (1h/day), except for those that presented risk of falling, for 1 year. After this period, the staging of ataxia remained significantly better in relation to pre-intervention values. The improvement in GAS was also maintained. The benefits for balance and gait speed, however, did not remain significant. The authors concluded that the treatment was efficient in the long term.




Figue 1. Flowchart of article selection

Shiga et al.[22] summoned 74 patients with SCA and other ataxias and employed transcranial magnetic stimulation (TMS) over the cerebellum of 39 of them (stimulation group), while 35 of them received false stimulation (placebo group). Ten simple TMS pulses were applied once a day for 21 days. The stimulation group presented significant cadence improvement, as well as in relation to the time spent in the 10 m walking test, besides the increased number of possible steps in tandem and the ability to stay standing. After the 21 days of stimulation, 1 group of patients continued to receive TMS pulses once or twice a week. They retained the improvement obtained in the study for at least six months. Another group received TMS pulses only every two weeks. In this group, the scores of the used assessment methods returned to baseline two weeks after the end of applications.

Miyai et al.[23] assessed the effects of a short-term treatment in 16 people with idiopathic ataxia and 26 SCA, dividing them in 2 groups (immediate and control), with 21 patients each. Only the immediate group performed 1 hour daily trainings with conditioning exercises, amplitude of movement, muscle strengthening, stairs, static balance and gait for four weeks. In order to assess both groups, some tools were used: Sara, FIM (functional independence measure), functional ambulation categories, and gait speed/cadence, besides the number of falls per week. After practice, the immediate group presented improvements in all of the evaluations, except for cadence, when compared to the control group. After the four weeks, the control group was also trained and their data were analyzed together with the immediate group, accounting for 42 assessed patients before, right after and 4, 12 and 24 weeks after intervention. After two years, only the improvement in gait speed was maintained. However, it was observed that 22 out of the 42 patients had retained improvement in at least one of the assessed parameters during the period. By analyzing the correlation between staging and the assessed parameters, it was concluded that patients who retained some sort of improvement were those with less severe degrees of ataxia.

Dias et al.[24] conducted gait training with 10 patients (5 with SCA) wearing a 500 g shin pad on each lower limb (weight group), and with 11 patients (6 with SCA) without the shin pad (no weight group). The practice consisted of lateral gait, in a straight line, with obstacles and movements of the upper limbs. After 20 sessions of 30 min, there was significant balance improvement, as well as in the risk of falls, in FIM and in the staging of ataxia, only for the weight group. These results were maintained 30 days after the end of the intervention. The authors defend gait improvement even if there were no differences in Dynamic Gait Index scores.

## DISCUSSION

Out of the five studies analyzed in this review (Table 2), none of them investigated only patients with SCA. Individuals with ataxia of other etiologies were also included. Even so, the studies report the positive impact of physical therapy for patients with SCA. This suggests that the interventions used for ataxias of other etiologies can also be useful for the SCA treatment, even though this fact is not completely clear.

Only three studies[22-24] compared the performance of a trained group and a control group. This limitation does not make it possible to rule out the placebo effect, nor to assess if the benefits reported by them are only due to the proposed training.

Only Dias et al.[24] assessed if the protocols had an impact on the performance of activities and on the patient's independence. This is essential to observe the improvement in the quality of their daily lives.

Out of the five studies in this review, four of them[20,21,23,24] described the staging of SCA. This is necessary to investigate if, after treatment, the clinical condition of the patient, in short and long terms, became worsen, better or stable. Such description can also help identify the adequate treatments for the different stages of the disease. Ilg et al.[20], Miyai et al.[23] and Dias et al.[24] reported improved staging of ataxia after the treatment. Ilg et al.[20] stated that a short period after the end of the intervention proposed in their study, the improvement obtained in the assessments had no correlation with the staging of SCA. In the long term, Miyai et al.[23] observed that patients who mostly retained the improvements were those with less severe ataxia. Ilg et al.[21] demonstrated that one year after the end of their protocol, patients still had significantly better scores at SARA than in the first evaluation.

Apparently, maintaining the improvement obtained with interventions depends on continuing to practice exercises. Ilg et al.[20,21] and Miyai[23] recommended the daily practice of their protocols to patients after evaluation if there was long term retention, which happened in all of the cases. Shiga[22] also maintained the application of TMS pulses with better results in the group that continued to receive frequent applications. On the other hand, Dias et al.24 reported the maintenance of improvement 30 days after the end of the intervention, even without continuing to practice. The gain retention described in some studies may indicate that patients with cerebellar lesions are able to retain learning, even though their lesions are located in the cerebellum, which is such an important area for motor learning. Still, it is hard to distinguish if the lack of benefit retention after an exercise program is owed to the progressive degeneration, characteristic of the disease, to the inability to retain the learned movement patterns or to the need to elect more appropriate exercises for these patients.

Most of the found studies employed balance, gait, muscle strengthening, stretching and coordination exercises. However, none determined which the most efficient one was. Therefore, the good results found

Table 2. Summary of the analyzed articles

| Author, Year | Etiology | Control | Assessment | Staging | Long term assessment | Intervention |
|---|---|---|---|---|---|---|
| Ilg et al.,[20] 2009 | Idiophatic and sensitive SCA | – | SARA, ICARS, GAS, stabilometry Berg and kinemetrics | SARA and ICARS | 8 weeks SARA, balance, speed and step length | Training balance, coordination, amplitude of movement and against falls |
| Ilg et al., [21] 2010 | Idiophatic and sensitive SCA | – | SARA, ICARS, GAS, stabilometry Berg and kinemetrics | SARA and ICARS | 1 year SARA, GAS | Training balance, coordination, amplitude of movement and against falls |
| Shiga et al., [22] 2002 | SCA and acquired | Placebo stimluation | 10 m test, tandem steps, ability to stand up and walk | – | 6 months 10 m test, tandem steps, ability to stand up and walk | TMS |
| Miyai et al., [23] 2012 | SCA and idiophatic | Did not receive treatment | SARA, FIM, gait cadence/velocity and number of falls | SARA | 24 weeks Gait velocity | Training balance, conditioniong, gait, amplitude of movement, strenghetning, stairs |
| Dias et al., [24] 2009 | SCA and acquired | No shin pads | ICARS, FIM, DGI, *equiscale* BERG | ICARS | 30 days ICARS, BERG and FIM | Training lateral gait with obstacles, limb association, shin pads in lower limbs |

in this study indicate that different types of exercise could be equally beneficial for these patients. Indeed, Fryer et al.[26] noticed a beneficial effect of non-specific exercises for the progression of cerebellar degeneration in rats with SCA1. These findings may corroborate the idea that immobility leads to motor loss, which goes beyond those promoted by the characteristic degeneration of SCA itself. Therefore, it seems that keeping patients active alone can already being some sort of benefit.

Additionally, Dias et al.[24] showed that, regardless of the type of employed exercise, the use of shin pads on the lower limbs during practice can increment its beneficial effect.

Most studies, except for the one of Ilg et al.[20], do not describe the intervention protocol in detail. Such details are important to ensure replicability in new studies, including multicenter clinical trials, and to enable the reproduction of the protocol in clinical practice.

## CONCLUSION

In this review, studies demonstrating the positive effects of interventions to treat the sequels of SCA were identified: balance, gait and coordination training, muscle strengthening and stretching, use of shin pads in lower limbs and the application of TMS pulses on the cerebellum. Treatment seems to be more efficient less impaired patients. Besides, retaining the improvements obtained in the long term was related to the degree of disease evolution and to the continuous practice of exercises, even if at home. Future studies including only patients with SCA, which would describe the staging of the disease, including a proper control group, blind evaluators, description of the impact of treatment on activities and participation and containing a detailed description of the intervention protocol are necessary to select the most adequate approaches for the treatment of patients with SCA. These studies may contribute with the elaboration of protocols based on important evidence to face the problems caused by SCA properly.

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