



Literature review / Revue de la littérature

The assessment and treatment of postural disorders in cerebellar ataxia: A systematic review

Ataxie cérébelleuse posturale : évaluation et traitement

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Abstract

Gait and balance disorders are often major causes of handicap in patients with cerebellar ataxia. Although it was thought that postural and balance disorders in cerebellar ataxia were not treatable, recent studies have demonstrated the beneficial effects of rehabilitation programs. This article is the first systematic review on the treatment of postural disorders in cerebellar ataxia. Nineteen articles were selected, of which three were randomized, controlled trials. Various aetiologies of cerebellar ataxia were studied: five studies assessed patients with multiple sclerosis, four assessed patients with degenerative ataxia, two assessed stroke patients and eight assessed patients with various aetiologies. Accurate assessment of postural disorders in cerebellar ataxia is very important in both clinical trials and clinical practice. The Scale for the Assessment and Rating of Ataxia (SARA) is a simple, validated measurement tool, for which 18 of the 40 points are related to postural disorders. This scale is useful for monitoring ataxic patients with postural disorders. There is now moderate level evidence that rehabilitation is efficient to improve postural capacities of patients with cerebellar ataxia – particularly in patients with degenerative ataxia or multiple sclerosis. Intensive rehabilitation programs with balance and coordination exercises are necessary. Although techniques such as virtual reality, biofeedback, treadmill exercises with supported bodyweight and torso weighting appear to be of value, their specific efficacy has to be further investigated. Drugs have only been studied in degenerative ataxia, and the level of evidence is low. There is now a need for large, randomized, controlled trials testing rehabilitation programs suited to postural and gait disorders of patients with cerebellar ataxia.

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Keywords: Postural and balance disorders; Cerebellar ataxia; Rehabilitation; Posturography

Résumé

L'ataxie posturale cérébelleuse est source d'incapacité souvent majeure pour les patients. Elle a longtemps eu la réputation de ne pas être accessible au traitement. Plusieurs études récentes suggèrent au contraire que les patients ayant une ataxie posturale cérébelleuse peuvent être améliorés par la rééducation. Cet article est une des premières synthèses de la littérature sur le traitement de l'ataxie posturale cérébelleuse. Dix-neuf articles dont 3 essais randomisés portant sur la rééducation sont analysés. Ces études portent sur des pathologies variées (5 essais portent sur des patients porteurs de sclérose en plaques, 4 sur des patients ayant une ataxie dégénérative, 2 sur des patients après accident vasculaire cérébral et 8 portent sur des étiologies variées). Ceci est une limite pour la généralisation des résultats. Ces études pointent l'intérêt d'une évaluation performante de l'ataxie posturale cérébelleuse, à la fois pour la clinique et la recherche. La Scale for the Assessment and Rating of Ataxia (SARA), validée pour évaluer l'ataxie cérébelleuse, comporte 18 points sur 40 sur l'évaluation de l'ataxie posturale cérébelleuse. Simple d'utilisation et possédant de bonnes qualités métrologiques, elle est utile pour suivre l'évolution des patients. Les résultats des études apportent un niveau de preuve modéré sur l'efficacité de la rééducation, en particulier lors d'une ataxie dégénérative ou d'une sclérose en plaques. Il est à noter qu'il n'y a aucun essai randomisé multicentrique portant sur un grand nombre de patients ; c'est un défi pour les années à venir. Le programme de rééducation comportera des exercices intensifs d'équilibre et de coordination. L'utilisation de techniques de rééducation complémentaires tels que la réalité virtuelle, le biofeedback, la marche en suspension sur tapis roulant ou le lestage semble intéressante mais la place de ces techniques nécessitera d'être mieux définie. Les traitements médicamenteux ont été uniquement étudiés sur les ataxies dégénératives. Le niveau de preuve est faible quant à l'efficacité des traitements proposés.

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Mots clés : Ataxie cérébelleuse posturale ; Rééducation ; Posturographie

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1. English version

1.1. Introduction

For many years, it was thought that postural and balance disorders in cerebellar ataxia were not treatable. However, the results of several recent studies suggest that rehabilitation can relieve postural disorders in patients with cerebellar ataxia. This article is one of the first to review this recent literature. It covers 19 studies of rehabilitation (including three randomized trials) and eight studies of drug treatment. The aetiology of the postural disorders varied; in the rehabilitation studies, degenerative ataxia and multiple sclerosis (MS) were the most frequently studied conditions ($n = 5$ studies for each, including the three randomized trials). This may be a source of bias that prevents the results from being extrapolated to other types of cerebellar syndrome. In terms of drug treatments, only degenerative ataxia has been studied.

The results of the studies examined here suggest that quantitative evaluation of postural disorders in cerebellar ataxia has value for both clinical research and clinical practice. We shall successively address the evaluation of postural disorders, rehabilitation, drug treatments and surgical treatments in the context of postural ataxia. Treatment of the underlying disease and any associated impairments (orthopaedic deformations, sensorimotor impairments, cognitive disorders, etc.) will not be discussed here.

1.2. Evaluation of postural disorders in ataxia

Postural disorders in cerebellar ataxia can be evaluated both qualitatively and quantitatively. Qualitative evaluations are

based on a precise assessment of clinical symptoms. The combination of postural instability and a staggering, festinating gait is a sign of cerebellar ataxia. Accurate analysis of clinical symptoms is often omitted because of low levels of physician awareness (given that teaching about gait and gait disorders is not a high priority at medical schools) [35].

The recent observation of ipsilesional and contralesional deviations of the subjective visual vertical (SVV) in patients with acute, unilateral, cerebellar damage [2,3] suggests that the sense of verticality may be altered in cerebellar diseases. However, it is possible that this disorder is related to concomitant vestibular damage. An evaluation of several modes of perception of the vertical in patients with cerebellar ataxia should be able to resolve this question. In fact, it is widely acknowledged that vestibular disorders can alter the SVV without greatly affecting the subjective haptic and postural verticals [7,28]. In theory, there is no reason why perception of the postural vertical should be altered in patients with cerebellar damage alone [3].

Certain generic evaluations of balance disorders and ordinal scales evaluating the various components of ataxia can be used to quantify the severity of postural disorders in cerebellar ataxia. The generic evaluations of balance include the Berg Balance Scale (BBS), timed standing tests (on one or two legs) and posturography. The latter technique reveals marked oscillations (Fig. 1); a frequency-domain analysis of the posturographic signal shows a peak in the 3 to 5 Hz band that is characteristic of cerebellar tremor. These non-specific evaluations of cerebellar ataxia have been described in detail elsewhere [34]. Generic gait assessments are also useful. In cerebellar ataxia, the basic spatiotemporal gait parameters

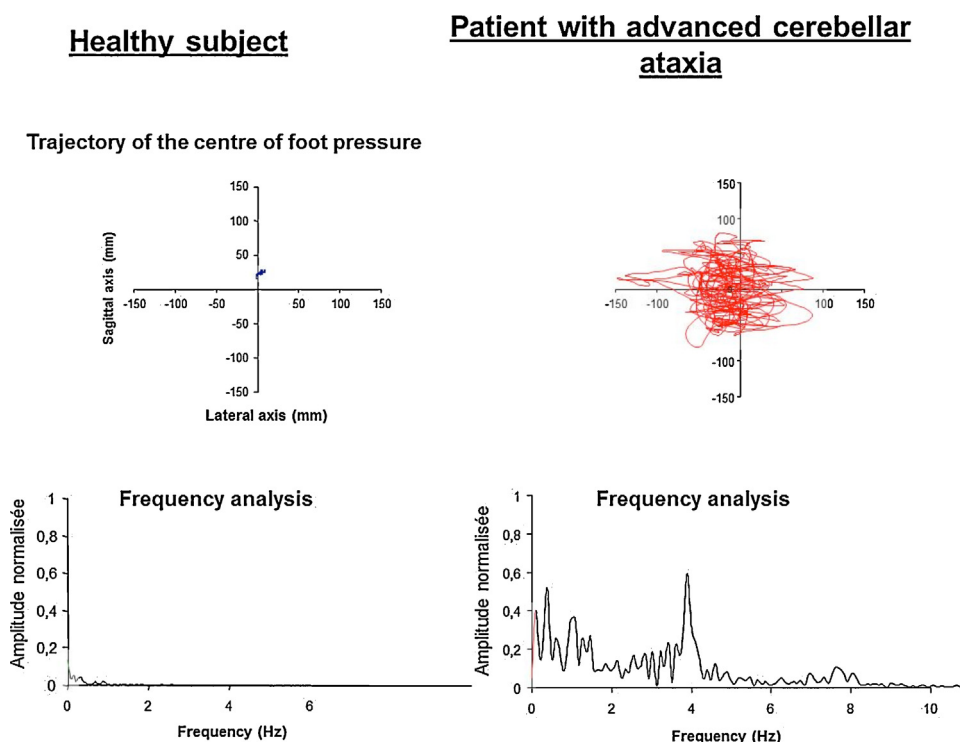


Fig. 1. Posturographic analysis of a healthy subject (left) and a patient with cerebellar ataxia (right).

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