Journal of the Neurological Sciences 316 (2012) 79–85

Contents lists available at SciVerse ScienceDirect

Journal of the Neurological Sciences

journal homepage: www.elsevier.com/locate/jns

Uses of the postural stability test for differential diagnosis of hereditary ataxias

J. Schwabova [a,*], F. Zahalka [c], V. Komarek [b], T. Maly [c], P. Hrasky [c], T. Gryc [c], O. Cakrt [d], A. Zumrova [b]

[a] Department of Neurology, Charles University in Prague, 2nd Faculty of Medicine and the Motol University Hospital, V Úvalu 84, Prague 5, 150 06, Czech republic
[b] Department of Paediatric Neurology, Charles University in Prague, 2nd Faculty of Medicine and the Motol University Hospital, V Úvalu 84, Prague 5, 150 06, Czech republic
[c] Research Sport Center, Charles University in Prague, Faculty of Physical Education and Sport, Jose Martího 31, Prague 6, 162 52, Czech republic
[d] Department of Rehabilitation and Sport Medicine, Charles University in Prague, 2nd Faculty of Medicine and the Motol University Hospital, V Úvalu 84, Prague 5, 150 06, Czech republic

ARTICLE INFO

Article history:
Received 19 June 2011
Received in revised form 16 January 2012
Accepted 24 January 2012
Available online 13 February 2012

Keywords:
Ataxia
Sensory ataxia
Cerebellar ataxia
Coordination impairment
Spinocerebellar ataxia type 2
Friedreich ataxia
Differential diagnosis
Centre of pressure

ABSTRACT

Friedreich's ataxia (FRDA) and spinocerebellar ataxia type 2 (SCA 2) are among the most commonly diagnosed hereditary ataxias in Czech Republic. Although criteria differentiate the ataxias, disorder onset symptoms may be similar.

Our goal was to determine whether and to what degree of validity posturographic examination may be utilized, with the aim of differential diagnosis; which specific posturographic parametres are suitable for differential diagnosis; and which differences in FRDA and SCA 2 patient posturographic findings may be established.

17 SCA 2 and 12 FRDA patients were examined with ten healthy controls. A multi-sensor tenzometric platform was used for posturographic examination. Toe standing position was added to basic tests, including standing position with and without visual control.

There was no difference between patients in standing position with visual control but there were distinct differences between FRDA and SCA 2 patients, based on upright stance without visual control and medio-lateral deviation.

There were no differences between patients in toe standing position, suggesting not only the cerebellum, but also deep sensation, helps to create the so-called adaptive controller.

Posturography is attested to as a useful method for differential diagnosis of hereditary ataxias and provides neurophysiological findings in cerebellar and sensoric ataxias.

1. Introduction

From a clinical and neurological perspective, ataxia is a syndrome appearing as a result of cerebellar affliction and/or its centripetal or centrifugal paths of all varieties of etiology. One of the causes of the manifestation of ataxia may be a group of hereditary ataxias with all types of inheritance. Friedreich ataxia (FRDA) and autosomal dominant spinocerebellar ataxia type 2 (SCA 2) are among the most commonly diagnosed hereditary ataxias in the Czech Republic [1,2]. FRDA was long thought to be a disorder particular to childhood age but greater data specification, resulting from the development of DNA analysis, now shows that FRDA belongs even among differential diagnosis of ataxias with onsets in adulthood [1].

In both FRDA and SCA 2 there is a progressive atrophy of the cerebellum as well as spinocerebellar and dorsal column paths [3]. In the begining, the clinical portrait, may be very similar. Genealogical data does not necessarily assist in the aims of DNA diagnosis.

One of the first symptoms in patients tends to be the loss of stability during regular day-to-day activities, both in static situations such as standing and, especially, in dynamic activities such as stair-climbing [4,5]. Neurological findings, as with neuroimaging method findings, may be weak at this stage. It is possible to differentiate between nosological units by means of typical clinical features such as Romberg sign, limb ataxia, truncal ataxia, spasticity and plantar reflex, typical among FRDA patients [6] and SCA 2 patients with cerebellar ataxia and slow eye movements [5]. MRI is also helpful in differential diagnosis: ponto-cerebellar atrophy may be found on the MRI among SCA 2 patients, while spinal atrophy without cerebellar atrophy is typical among FRDA patients.

For quantification and the possibility of comparing clinical findings, a range of clinical testing scales were developed. Such scales are unfortunately to some extent subjectively influenced. The most commonly used scales are the International Cooperative Ataxia Rating Scale (ICARS) [7] and the newer SARA scale (Scale for the assessment and rating of ataxia) [8] Test results are in the later stages of the disorder mostly in accordance with morphological findings. The clinical portrait of SCA 2 patients is framed namely by cerebellar ataxia. Such is not the case with FRDA as it is mostly a sensoric ataxia. Results in differentiation are not overly positive or beneficial in the early stages of the disorder [3].

Among electrophysical methods, posturography has been used more and more in recent years. Posturography helps to eliminate

* Corresponding author. Tel.: +420 224 436 801; fax: +420 224 436 820.
E-mail address: schwabova@gmail.com (J. Schwabova).

0022-510X/$ – see front matter © 2012 Elsevier B.V. All rights reserved.
doi:10.1016/j.jns.2012.01.022

the subjectivity of examiner and examined. On the basis of posturographic examination of ataxia patients, greater COP (centre of pressure) deviation with a typical cycle frequency and an inability to adequately react to deviation in position, with a tendency to overcorrect movement, was demonstrated [9,10]. The importance of the Romberg's test was confirmed [11].

On the basis of basic posturographic examination, sensoric ataxia is differentiated on the basis of the Romberg´s test [12]: Upright stance without visual control. No provocative maneuvres are typically applied for cerebellar symptomology. After having analyzed the situation, we focused on the possibility of expanding the testing programme by including toe standing position on tiptoes.

From a neurological perspective, upright stance shares three main systems: visual appartus, somatosensor and vestibular system. The cerebellum is thought to be critical for postural coordination and probably plays several different roles in control of posture involving sensorimotor integration [13]. Lesions in different regions of the cerebellum produce very different effects on postural control [14]. For examaple lesions of the lateral hemispheres can produce profound disorders of timing for arm and hand coordination without significant effects on posture and gait [15]. Toe standing position on tiptoes is apparently a common motor activity, during which relatively great changes occur in the centre of balance [16,17]. It is a basic motor skill whose consequence determines the quality of the motion carried out [18]. An important role in the coordination of balanced reaction is played by the cerebellum.

The lower limbs – namely the feet in the frontal foot and heel areas – are used in upright stance. The centre of pressure working under each foot is found closer to the heel area and in ideal cases, a symmetrical burden is to be expected.

When in toe standing position, pressure on the heel area is lessened and the burden is gradually transferred to the frontal foot area. In that moment, the heel is no longer in contact with the surface underfoot.

Medio-lateral deviation of the centre of pressure on a flat surface typically changes position towards the frontal foot. As in every free movement, the combination of opposing forces – agonist and antagonist – is important as regards muscle engagement in the toe standing position. The basic consideration is the assumption that the combination of agonist and antagonist is manifest in the change of pressure on the flat surface, from the heel area of the foot to the frontal foot. Incoordination is typical of patients with cerebellar lesion [13] and is identified by means of irregular changes in the centre of pressure [19]. Since the action has its own clear beginning and end, it is called a closed-loop motor skill. From the perspective of motor learning, such a skill is expected to be stable and automatic in healthy individuals [20]. It is, however, a difficult action for patients with stability and coordination problems [21].

There exists currently a wide range of work comparing clinical tests with postural stability parameters [22–25]. It has been demonstrated that, according to selected parameters, it is possible to clearly differentiate healthy and unhealthy individuals, although intercomparison does not lead to clear conclusions [24,25]. One cause may be that provocative maneuvers in changes of position and the bodily movement of patients are a component part of clinical scales and in some works not a part of posturographic examination.

Measuring dynamic body responses during simple motor maneuvers creates objective evidence about changes in time and space and bodily reactions to internal and external subjects. Initiated subjects are in most cases expected to arise from bodily movements carried out during regular day-to-day activities, since they are expected to increase the risk of loss of stability and, alongside loss of stability, increase the related risk of falling [4]. Several provocative maneuvers lower standing position stability. Among these is loss of eye control during measurement. It is possible in the same way to increase test difficulty – even when performing motor maneuvers such as toe standing position on tiptoes while standing, raising arms, or a combination of both movements [16,17,26].

The goal of our study was to test a) whether and to what extent posturographic examination is a helpful and useful procedure in the differential diagnosis of hereditary ataxias prior to DNA analysis; aa) and to test the value of toe standing position as a suitable and useful maneuver in the testing of ataxia patients; and aaa) to determine differences in postural stability parameters in patients with a preponderance of cerebellar rather than sensoric elements of ataxia.

2. Methods

2.1. Study sample

17 SCA 2 patients and 12 FRDA patients (Table 1) were tested with ten health controls. The disorder was verified on a molecular level. Patients chosen were able to stand, without the need of support, with or without visual control. Clinical findings were evaluated by means of the ICARS and SARA scales. No further criteria was brought to bear in order to best simulate the regular approach of a physician in practice. The average age of the SCA 2 patients was 43.8 (18–58) and the group was comprised of eleven men and six women with an average period of clinical symptoms stretching back 10.8 years (1–29). The average age of the FRDA patients was 31.9 (19–59) and the group was comprised of seven men and five women with an average period of clinical symptoms stretching back 10.91 years (5–27). Patients were divided into two groups – ICARS<30 = GM (Group of Mild Ataxia) and ICARS>30 = GS (Group of Severe Ataxia) – to compare postural stability results in relation to disability.

Healthy controls had neither orthopedic nor neurological case-history. Neurological findings were normal. ICARS was 0.

2.2. Assessment of postural stability parameters

The pressure method was used for posturographic examination, measuring pressure on sensors in the FOOTSCAN platform (RSscan, Belgium). The FOOTSCAN is 0.5 m by 0.4 m and has approximately 4100 sensors sensitive from one-tenth of N/cm^2 and a 500 Hz sensing frequency. Pressure on individual sensors is measured and the centre of pressure calculated on the contact area, or COP. Standard standing position with wide base was the selected standard stance. Hip width delimited stance, but was measured out afterwards by antropometer transferred to the base, determining instep distance. Transparent sheeting for tracing foot position was placed between feet and the FOOTSCAN platform to ensure individual conformity during repeated examination. Stance was measured according to standard practice at a length of 30 s [27]. The following parameters were used to evaluate COP centre of pressure: medio-lateral directional deviation (Delta X); anterio-posterior directional deviation (Delta Y); the whole course of total travelled way (TTW); and standard deviation of COP velocity (VelocitySD).

Parameters were viewed after a one-second time period after initial movement during toe-standing position, without the help of arms, in order to evaluate pressure changes in toe-standing position.

TTW was chosen among evaluated parameters, as well as trajectory change of centre of pressure in medio-lateral directional deviation: Delta X from initialization to stabilization in toe-standing position. Size of confident elipse was a further evaluated parameter and was calculated at intervals from the initialization of movement until the stabilization of toe-standing position (set at 1 s). The confident elipse is determined by the surface, which will in all probability locate the actual position of designated central pressure points. The final evaluated parameter was pressure under the right and left lower limb during initialization of toe-standing position, expressed by co-relation, the Right to Left ratio, or R:L.