**ORIGINAL ARTICLE** 

## Activities of Daily Living Scale - the tool for clinical state monitoring of spinocerebellar ataxia and Friedreich ataxia patients

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## ABSTRACT

## Background

Autosomal dominant spinocerebellar ataxia (AD SCA) and Friedreich ataxia (FRDA) are neurodegenerative and neurometabolic disorders whose characteristics include progressive impairment of balance and motor functions. The causes of these disorders vary, but both involve degeneration of the cerebellum and its connections. Until now, there has been no causative therapy for the disorders.

A number of research reports point out that undamaged cerebellar tissue can be able to cope with changes and learn new or different motor skills. It is for this reason that physiotherapy and other physical activities improving balance and motor coordination are essential for all patients. It is necessary to monitor and evaluate the results of therapy/physiotherapy in order to be able to suggest the most efficient symptomatic therapy concept and thereby extend the active life of patients. The Vestibular Disorders Activities of Daily Living Scale or ADLS is used for the evaluation of the daily activities of patients suffering from vestibular disorders. There is no similar scale for patients with cerebellar or sensoric ataxia. ADLS is used for patients with vestibular ataxia and for that reason we posit that it may also help AD SCA and FRDA patients.

Ataxic patients with AD SCA and FRDA are typically tested using the International Cooperative Ataxia Rating scale (ICARS); Our goal is to test AD SCA and FRDA

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Department of Paediatric Neurology Charles University in Prague 2nd Faculty of Medicine and Motol Hospital V Uvalu 84,Prague 5, 150 06 Czech Republic Phone no: + 420 224 433 301 Fax: + 420 224 433 322 E-mail: jarusch@email.cz patients using the ADLS. Posturography, a diagnostic tool for balance impairment, is then used an objective scale of measurement.

Methods

The observed group was created from thirty ataxic patients – thirteen diagnosed with FRDA and seventeen with AD SCA. All were assessed using the ICARS as well as ADLS. The FOOTSCAN pressure platform was used for evaluating postural stability. The patients were examined in normal standing positions and the following parameters were evaluated: Absolute deviation in front/back direction; Absolute deviation in a left/right direction; Total travelled way of the centre of pressure COP.

Results

ADLS showed a correlation with the ICARS (r=0,56) and selected parameters of postural stability.

Conclusions

It appears that ADLS is a reasonably effective tool for clinically monitoring the neurological state of FRDA and AD SCA patients. The correlation with the ICARS (International Cooperative Ataxia Rating Scale) scale is statistically significant. Posturographic examination as an objective diagnostic tool confirms correlations between the results of ADLS and all selected posturographic parameters.

Keywords: Spinocerebellar ataxias, Friedreich ataxia, quality of life, activities of daily living, postural equilibrium

## INTRODUCTION

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