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Computer-based assessment of upper-limb incoordination in autosomal recessive spastic ataxia of Charlevoix-Saguenay patients: A pilot study

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ABSTRACT

Ataxia refers to a group of neurological disorders characterized by a lack of coordination during voluntary movements. One of the most commonly used tests to assess upper-limb coordination is the Archimedes spiral test. The purpose of this research is to present an innovative computer-based Archimedes spiral test that can accurately assess coordination. Forty nine individuals (age: 25.2 ± 7.1 years) were recruited including thirteen patients diagnosed with Autosomal Recessive Spastic Ataxia of Charlevoix/Saguenay (ARSACS). Participants were instructed to trace a spiral on the touch-screen with the tip of their index finger at a self-paced velocity by following an on-screen spiral template. Mean error and maximum error as well as frequency analysis were calculated to classify healthy and ARSACS participants. While mean and maximum errors provided good results, the highest classification success rate was obtained using frequency analysis, particularly between $f = 1.2$ Hz and $f = 1.7$ Hz. Interpretation of traditional paper-drawn Archimedes spirals is limited, and several computerized versions have been reported. Herein, we present a custom-made tool that allows discrimination of measures assessing ataxia in ARSACS. This utilizes a proposed frequency method that may have the potential to track the evolution of upper-limb incoordination in patients and therefore help clinicians and scientists to better monitor their patients.

1. Introduction

Recessive ataxias are a heterogeneous group of neurodegenerative disorders affecting the cerebellum and/or the spinal cord. Autosomal recessive spastic ataxia of Charlevoix/Saguenay (ARSACS) falls into this family of neurological diseases [1]. This particular neurological condition was first described by Bouchard and colleagues in 1978 [2]. Clinically, ARSACS is a mixed picture of progressive upper motor neuron, cerebellar and peripheral neuropathy with variable intensity of these three components [3]. The progressive loss of coordination in the upper limbs is a key feature of the disease [4]. The assessment of coordination is greatly influenced by the presence of the peripheral neuropathy where a progressive loss of strength is clinically observed in the thumb abductor and the intrinsic muscles of the hand. It often limits the choice of assessment methods as most of them imply some type of fine prehension. A common test for assessing upper-limb incoordination is the Archimedes spiral [5,6]. This test has been established to show quantitative changes in essential tremor, Parkinson Disease, Niemann-Pick disease, psychogenic movement disorders and ataxia [5,7–10].

1.1. The Archimedes spiral test

There are several ways of performing the Archimedes spiral test. The standard Archimedes spiral test used in neurology consists of drawing a free form spiral [11]. Another method proposes drawing between two standardized spiral lines [12]. Finally, the method used in this paper consists in tracing over a standardized spiral [13–16] In its original form, the spiral is traced using pen and the result is analyzed qualitatively to classify the participant in one of five possible levels ranging from 0 to 4 [17]. In addition, it is also possible to measure the deviations between the standardized shape and the drawn spiral by using a ruler. A participant's coordination can then be evaluated according to the number and magnitude of these deviations. However, this method has limitations: the use a standard ruler only allows for limited accuracy of the measurement and the evaluation of the spiral trace only considers the sections containing the largest errors and thus, provides an incomplete picture. Although this procedure is regularly used in the evaluation of several ataxic syndromes and other movement disorders, the level of

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accuracy in these scales is limited [5,6,10]. Since the evolution of ARSACS is typically slow [3], it is often difficult to accurately track the progression of the disease. However, the development of technologies, such as graphic tablets and scanners, could allow a significant improvement in measurement accuracy and thus ensure that even small changes can be detected [6,11,14–16,18,19]. While these devices have shown good validity [5,20], there are some improvements that can be made. For instance, Miralles and colleagues [15] propose using the Archimedes spiral test on paper and digitizing the results for processing afterwards. This increases the number of steps required which also increases the total test time. In addition, the resolution of the scanned image may have an impact on the result.

Others, working with graphic tablets, require that the test be performed using a pen. With ataxic patients, the decrease in muscle strength of the hand limit the use of an intermediate object, such as a pen, as it may increase the difficulty in realizing the task. Finally, to the best of our knowledge, no previous work has been performed on using a computerized Archimedes spiral test on ARSACS patients, although a recent study used similar technology with patients affected by other types of movement disorders [21].

The objectives of this paper are twofold: 1) to present a new frequency analysis method for the Archimedes spiral test involving a finger-based touch-screen testing environment and 2) to demonstrate its construct validity. Our hypothesis is that ARSACS patients will present lower scores than healthy controls on the computerized Archimedes spiral. This instrument will potentially help clinicians and scientists better monitor their patients.

2. Methods

2.1. Design

This study assesses construct validity using the known-groups method. The comparison was made between healthy and ARSACS participants.

2.2. Participants

Forty nine (49) individuals (twenty eight males and twenty one females) with a mean age of 25.2 ± 7.1 years participated in this research. Of these participants, thirteen [13] were diagnosed with ARSACS while the other thirty six (36) participants were identified as being healthy (3 were left-handed). All participants were at least sixteen years of age, which is the legal age at which a person can give their consent to participate in a study according to the applicable local laws. Included in the healthy group were individuals exempt of upper body conditions that could distort a spiral trace made with the tip of the finger. To be included in the ARSACS group, participants must have a DNA molecular diagnosis (genetic test). All participants gave their informed written consent to participate in this research, which was approved by the University Ethics Committee (no.602.381.01).

The healthy participants were recruited from the kinesiology program at the University of Quebec in Chicoutimi whereas the ARSACS individuals were recruited from the Neuromuscular Clinic of Jonquiere Hospital. Both healthy participants and ARSACS patients performed the spiral test at the University Research Center using the same equipment. The tests were supervised by experienced research assistants.

2.3. Testing protocol

The computer-based Archimedes spiral test was performed on a computer equipped with an i5-2520M (2.5 GHz) processor with 4 GB of RAM running on Windows 7 Professional. The computer display is a 56 cm optical touch-screen monitor (Planar 2230MW) with a 5 min response time. The entire spiral test was created in MATLAB R2011b using a sampling rate of 100 Hz. When the test begins, the software draws a reference spiral on the screen. The dimension of this spiral was 15 cm in width by 15 cm in height with a gap of 2.5 cm separating each loop. As reported by Memedi et al. [10], when the spiral is too small, the hand of the participant would likely cover part of the spiral during the tracing motion, which interferes with the correct execution of the movement. It was found that the dimensions chosen were the most appropriate. The thickness of the spiral trace was chosen to be 1 mm, which corresponds to the thickness of a pencil trace on the paper version of the Archimedes spiral test.

To begin the procedure, the participant was asked to sit at an adjustable computer desk within an arm's length of the screen. In this position, it should be possible to touch the center of the screen with a slightly bent elbow. The individual is then instructed to trace the spiral on the touch-screen with the tip of the index finger of their preferred hand. While the participant follows the spiral on the screen at a self-paced speed, the pressure from their finger draws an image that is superimposed on the original drawing. Once the spiral is traced, the research assistant presses on the keyboard to signal the end of the test. The participants are only asked to trace one spiral. However, a 2-min period was provided to each participant before the beginning of the test in order to familiarize themselves with the setup.

Three main components of coordination can be extrapolated from the task:

1) Execution time. The execution time was measured because the speed at which the participant traces the spiral may have an impact on the quality of the drawing. Its value is displayed on the screen at the end of the test. Further breakdown of the execution time can be obtained for each portion of the spiral.
2) Spatial displacement. Spatial displacement, also known as radial error [15] or 1st order smoothness [21], is a renowned indicator of upper-limb incoordination. It is the measure that has traditionally been used in the Archimedes spiral test. To perform this measurement using the proposed software, data points are extracted from spiral trace at a rate of 100 Hz. As the user traces the spiral, the computer program associates each traced point to the nearest point on the reference spiral. This allows the software to determine which part of the reference spiral is being traced in real-time. For each of these traced points, the error is determined as the distance (in pixels) between the traced and the reference spirals. The result of this operation is a vector of error measurements from which the mean error and the maximum error can be calculated. These two measures can be used to characterize spatial displacement error.
3) Smoothness of movement. This characteristic can be determined by the amount of motion that is perpendicular to the trace of the spiral. When the movement is smooth, the motion perpendicular to the spiral trace is minimal. Equivalently, smoothness of movement can also be determined by measuring the variation in error between the traced spiral and the reference. Indeed, a smoother movement will tend to generate an error that fluctuates slowly. Similarly, movements that are more erratic will generate errors that change rapidly. Smoothness of movement can therefore be quantified using frequency analysis with proper windowing function [22]. In this work, the fast Fourier transform was used with a Hamming window. The results can then be traced on a graph with the X-axis representing the frequency and the Y-axis representing the amplitude. Large amplitudes close to the origin indicate that there are slow changes in measurement error whereas large amplitudes further away from the origin indicate high frequency changes in error. The amount of low and high frequency activities can be determined by performing an integral on a section of the frequency spectrum. The new method proposed in this paper is to perform a summation of all frequency components starting from a given frequency f up to the maximum available frequency, which corresponds to half the sampling frequency. In the case of this work, where the frequency spectrum ranges from 1 to 50 Hz, a frequency analysis with $f =$