Upper limb motor function, including the presence of motor incoordination, is routinely assessed as part of the neurological examination. The clinician relies on it as one of the preliminary diagnostic procedures. It can also be helpful in case management planning in progressive neuromuscular disorders. Generally, in this kind of disease, in a proactive management care plan, the role of the clinician is to monitor the progress of the disease and...
to decide when to refer the individual to rehabilitation and social services. In the context of limited evaluation time in clinical settings, measurement instruments and methods have to be carefully chosen in order to get an accurate picture of the patient.

Among clinical evaluations, measurement of coordination is of particular interest in neuromuscular disorders with predominant ataxic features, such as Friedreich’s ataxia. Coordination is defined as the capacity to execute a controlled movement with accuracy and rapidity. Its measure involves the appropriate activation of muscles with temporal and spatial components. Quantification of upper extremity coordination has been used as a diagnostic tool, to measure disease progression and to evaluate the impact of a given treatment. Desrosiers and collaborators have demonstrated moderate correlations between coordination and global upper extremity performance in an elderly population, which could be expected since coordination is a prerequisite of adequate upper extremity performance but not the only aspect to take into consideration.

Among disorders with ataxic symptoms, autosomal recessive spinocerebellar ataxia of Charlevoix–Saguenay (ARSACS) (MIM 270550) is an early-onset familial ataxia with pyramidal, cerebellar and distal neuropathic involvement of variable intensity. It is an inherited spinocerebellar degeneration found predominantly in the population of the northeastern part of the province of Quebec, Canada. Autosomal recessive spinocerebellar ataxia of Charlevoix–Saguenay is characterized by developmental defects in the fiber myelination of the central and peripheral nervous systems, with progressive axonal degeneration of the corticospinal and spinocerebellar tracts and axonal neuropathy. There are about 300 patients living today in the region of Saguenay–Lac-St-Jean and Charlevoix with equal prevalence between genders. Richter and colleagues have located the gene responsible for ARSACS on chromosome 13 (13q11) and two ancestral haplotypes were identified.

Clinical manifestations start early (around 18 months) usually with gait ataxia and history of falling reported by the parents. The early signs of the disease are slight ataxia and spasticity, predominantly in the legs. The disease progression becomes most obvious in the late teens and early twenties and the mean age at which patients become wheelchair-bound is 41 years old.

Although ARSACS mainly affects the lower limbs (incoordination, weakness and spasticity), upper extremity disabilities are also present and include incoordination, decreased dexterity and, for some patients, moderate to severe distal amyotrophy of the first dorsal interossei. Upper limb incoordination is a key characteristic of the assessment in this population.

Several methods have been used to quantify coordination. The traditional finger-nose test is a commonly used method and needs no specific apparatus. However, finger-nose tests are not standardized and several methods are used in clinical and research settings. For example, the finger-nose test can involve subjects alternately touching their nose and the evaluator’s finger in different positions or touching their nose and fully extending their arm in front of them. In addition, the traditional finger-nose test generates gross qualitative observations, which are not suited to the longitudinal observations needed to measure disease progression. Instrumental measures of coordination have also been developed but are rarely found in clinical practice as they take more time to administer. Moderate correlations between clinical and instrumental measures of coordination were demonstrated in people with brain damage and indicated a complementary role of the two measures.

The traditional finger-nose test generally includes measures of time of execution, dysmetria and tremor. The evaluation of dysmetria and tremor has shown poor test-retest and interrater reliability. The measure of time of execution, on the other hand, demonstrated excellent test-retest and interrater reliability. In order to increase reliability, Desrosiers and collaborators developed a protocol for a Standardized Finger-Nose Test (SFNT), and reference values for speed of execution are available for the older adult population (60 years and over). However, little is known about the validity of the test with a neuromuscular clientele with ataxic characteristics.

The validity of a test refers to its ability to measure the general and specific characteristics for which it was designed. Different types of validity of a test may be verified. Criterion validity is mainly studied by comparing a test to a gold standard measure. However, this benchmark measure is not always available. In such cases, construct validity is examined. This evaluates an instrument’s ability to confirm a hypothesis or theoretical construct related to the variable measured. Several types of construct validity can be studied, including convergent and discriminant validity. Convergent validity refers to the relationship with another instrument, itself reliable and valid, that measures a similar concept whereas discriminant validity refers to the ability of an instrument to differentiate between groups on a selected variable.

The objectives of this study were to explore 1) the convergent construct validity of the SFNT by correlating its score with upper extremity function tests that require coordination, functional independence and social participation, and 2) the discriminant construct validity of the SFNT by verifying its ability to differentiate between two age groups.

METHODS

Participants

A random sample of 24 participants with ARSACS, aged 18 and over were recruited from the Neuromuscular Clinic of the Centre de réadaptation en déficience physique in Jonquière, Québec, Canada. All the participants were homozygous for the mutation causing ARSACS on chromosome 13q11. For discriminant construct validity, the participants were stratified by age (< 40 years old and ≥ 40 years old) into two groups, based on the clinical observation that involvement is much more severe after age 40.

All participants were free of any other form of motor or cognitive deficits that could affect upper extremity function. They signed an informed consent form approved by the Complexe Hospitalier de la Sagamie Ethics Committee.

Data collection procedure

Participants were evaluated by the same occupational therapist in two sessions. Upper extremity function, including coordination, was evaluated during the first session at the Clinic, while functional independence and social participation were evaluated during the second session at the participant’s home.
Upper extremity strength was measured using three different measurement instruments. First, shoulder abductors, elbow flexors, wrist extendors and 1st dorsal interossei were evaluated with the Microfet-2 (Hoogan Health Industries) and belt-resisted method following Desrosiers’ specifications for upper extremity evaluation. Test-retest reliability coefficients varied between 0.88 and 0.96, depending on the muscle group. Second, grip strength was measured using the Jamar dynamometer (Asimow Engineering Co) following standardized procedures (mean of three measures). Test-retest reliability was estimated at 0.96 for the right hand and at 0.95 for the left. Finally, a pinch gauge (B&L Engineering) was used to measure the strength of the 1st dorsal interses following Mathiowetz and colleagues’ recommendations (mean of three measures). Test-retest reliability has been estimated at 0.83 for the right side and at 0.87 for the left.

The TEMPA was used to measure global upper extremity performance. It consists of nine standardized tasks (five bilateral and four unilateral), representing daily activities. The speed of execution and functional rating of each task are recorded for one trial. The functional rating corresponds to the person’s independence in performing each task and is measured on a 4-point scale from 0 (the task is successfully completed without hesitation or difficulty) to -3 (the task is not completed). Intraclass correlation coefficients ranged from moderate to high (0.70-1.0) for test-retest reliability of speed of execution and functional rating.

Functional independence

The Functional Independence Measure (FIM) was administered to evaluate functional independence. This scale includes 18 items which assess, on a seven-level scale, independence in basic daily activities in six domains: self-care, sphincter control, transfers, locomotion, communication and social cognition. Based on a meta-analysis, the test-retest reliability median is 0.95. In this study, only items that require major use of the upper extremity were used: eating, grooming, bathing and upper extremity dressing, and this subscore was called FIM-upper extremity. The score varies from 4 to 28, with the latter indicating a higher level of independence. This subscore was developed specifically for this study in order to focus on activities related to upper extremity use. In the context of this study, this subscale appeared more valid than using the measure’s entire motor subscore.

Social participation

Social participation is a conceptual domain of the new International Classification of Functioning and the Disability Creation Process. In this model, social participation is operationalized by the concept of life habits, which refers to “daily activities and social roles that ensure the survival and development of a person in society throughout his or her life”. Social participation was evaluated using the short version of the Assessment of Life Habits (version 3.0) composed of 69 items. These items are grouped into 12 categories: nutrition, fitness, personal care, communication, housing, mobility, responsibility, interpersonal relationships, community life, education, employment and leisure. The first six categories refer to daily activities while the others are associated with social roles. The measure of each item is based on two specific elements: 1)
degree of difficulty, and 2) type of assistance (technical assistance, physical arrangements, human help). The scale ranges from 0 to 9 where a score of 9 indicates complete independence without human or technical assistance (maximum level of social participation). Conversely, a score of 0 indicates that the activities or role cannot be accomplished because of tremendous difficulty or environmental factors. The reliability of the global score recently studied with older adults with physical disabilities is excellent (ICC and 95% confidence intervals: 0.95 (0.91 to 0.98) for test-retest).32

Statistical analysis

The statistical analysis was performed using the SPSS 10.0 software package.33 Means and standard deviations for upper extremity function tests, functional independence and social participation scores are presented. For upper extremity function tests, a combined total score was computed in order to reduce the number of correlations by averaging the scores of the right and left upper extremities since no significant differences were found between the two performances. Spearman’s correlation coefficient was used to estimate the convergent validity of the SFNT test with the other upper extremity tests, functional independence and social participation. Wilcoxon’s signed rank test was used to demonstrate the discriminant construct validity of the SFNT. Significance for all the statistical analyses was fixed at the 0.05 level.

RESULTS

The distribution of gender (50% female) and age (37.5% < 40 years old) is equivalent to that in the population with ARSACS. The Table shows means and standard deviations for each upper extremity function, functional independence and social participation as well as Spearman’s correlation coefficients between these measures and coordination. The convergent validity of the SFNT was supported by moderate to strong correlations with the measures of dexterity (0.82), global upper extremity performance (0.74-0.79), upper extremity functional independence (0.74) and social participation (0.78). The moderate to low correlations found with measurements of muscle strength (0.19-0.56) also supported the validity of the test since minimal strength is a prerequisite for coordination.

The coordination score measured with the SFNT is significantly lower in patients over 40 years of age (6.7; SD: 3.4) than in patients below 40 (12.7; SD: 2.2) (p < 0.001), supporting the discriminant validity of the SFNT.

DISCUSSION

Coordination is known to affect several aspects of daily living such as the ability to execute basic activities of daily living, to work and to engage in recreational activities.34 Fast and simple quantitative tests of neurological function need to be developed and/or validated since neurological examination is an essential part of the neurologist’s curriculum.35 The SFNTmeets these two criteria since it takes less than two minutes to administer and can easily be set up in any clinician’s office as it requires only a few items. The objectives of this study were to explore the convergent and discriminant construct validity of the SFNT in a population with ataxic features. The convergent validity of the test was demonstrated by correlating the SFNT with other tests measuring variables that were expected to be associated. The strong correlations (> 0.80) found between coordination and dexterity demonstrated their close relationship. By definition, dexterity requires coordination, sensitivity and prehension.36 It is known that sensitivity is usually intact in ARSACS6 and prehensions are possible. Therefore, the disturbance of coordination or ataxia in this disease seems to be the most important factor in the decrease in dexterity. These relationships support the use of the SFNT as an adequate measure of coordination and dexterity, that can be used with confidence in the clinical evaluation of this population.

The SFNT was also moderately correlated (rho: 0.48-0.56) with shoulder strength, grip strength and strength of the 1st interossei (pinch strength). The analysis of the movements executed during the SFNT provides an understanding of these relationships. First, the SFNT requires good stability of the shoulder as the subject has to stay at a precise height during the elbow extension and flexion movements. Secondly, sufficient upper limb strength is necessary in order to be able to execute rapid and precise movements. Therefore, the moderate

Table: Scores of upper extremity functions, functional independence and social participation of 24 patients with ARSACS and their correlations with the SFNT

<table>
<thead>
<tr>
<th></th>
<th>Standardized Finger-Nose Test [8.9 (SD: 4.2)] 1.5 – 16.5</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean (SD)</td>
</tr>
<tr>
<td><strong>Upperextremity function tests</strong></td>
<td></td>
</tr>
<tr>
<td>Gross dexterity (BBT)</td>
<td>32.2 (11.8)</td>
</tr>
<tr>
<td>Fine dexterity (PP)</td>
<td>4.2 (2.9)</td>
</tr>
<tr>
<td><strong>Upperextremity performance</strong></td>
<td></td>
</tr>
<tr>
<td>(TEMPA)</td>
<td></td>
</tr>
<tr>
<td>Functional rating (/39)</td>
<td>13.4 (8.5)</td>
</tr>
<tr>
<td>Speed of execution (sec)</td>
<td>290.4 (216.2)</td>
</tr>
<tr>
<td><strong>Strength (kg)</strong></td>
<td></td>
</tr>
<tr>
<td>Pinch strength (B&amp;L)</td>
<td>5.7 (1.9)</td>
</tr>
<tr>
<td>Grip (Jamar)</td>
<td>29.1 (11.2)</td>
</tr>
<tr>
<td>Shoulder abductors</td>
<td>25.1 (12.1)</td>
</tr>
<tr>
<td>Elbow flexors</td>
<td>39.4 (14.4)</td>
</tr>
<tr>
<td>Wrist extensors</td>
<td>32.3 (12.4)</td>
</tr>
<tr>
<td><strong>Functional independence</strong></td>
<td></td>
</tr>
<tr>
<td>(FIM-UE) (/28)</td>
<td>24.7 (4.3)</td>
</tr>
<tr>
<td><strong>Social participation</strong></td>
<td></td>
</tr>
<tr>
<td>(LIFE-H) (/9)</td>
<td>7.6 (1.0)</td>
</tr>
</tbody>
</table>

BBT: Box and Block Test; PP: Purdue Pegboard; TEMPA: Test Évaluant les Membres supérieurs des Personnes Agées; FIM-UE: Functional Independence Measure-Upper Extremity; LIFE-H: Assessment of Life Habits
correlations found between grip strength and coordination support the role of strength in the execution of precise movements.

Global upper extremity performance, upper extremity functional independence and social participation were highly correlated with SFNT (r: 0.74-0.79), which demonstrated the important role coordination plays in everyday activities. These relationships suggest that incoordination may play a significant role in the decline of function observed in ARSACS and needs to be better defined. Correlations were stronger than those reported in Desrosiers’s study with older adults, which could be explained by the greater variability of coordination in our population with cerebellar involvement. Indeed, statistics involving correlation can be influenced by the magnitude of variation within a specific population. Generally, a population with an ataxic disorder could be expected to demonstrate a greater variance in the score than the general population on a coordination test. Coordination was found to play a similar role in another study carried out with people with multiple sclerosis. In that study, the measure of coordination along with the measure of verbal intelligence and FIM admission score explained 56% of the variance in the patient’s functional improvement (FIM discharge score).

Therefore, the SFNT could possibly be a predictor of decline in functional independence and help clinicians refer people with increasing difficulty to rehabilitation and social services early in the process, without having to go through a sophisticated evaluation.

The discriminant construct validity of the SFNT was demonstrated as the scores were statistically lower for the older group, which was expected given the progressive nature of the disease and clinical findings. Indeed, despite the cross-sectional design of the study, the SFNT scores showed a progressive decrease with age. Upper limb motor coordination is known to decrease with age but the rate of progression is faster in ARSACS than in an older adult population (60 years of age and over). Therefore, the SFNT could generate more reliable and sensitive objective data on a longitudinal basis than the traditional subjective clinician’s finger-nose test.

Finally, the reader should bear in mind that the study sample comprised only one form of ataxic syndrome. Therefore, any inferences regarding the usefulness of the SFNT in other forms of ataxic syndromes should be made with caution.

The convergent validity of the SFNT was demonstrated by its relationships with related upper extremity functions tests, functional independence and social participation. These correlations also suggest that the SFNT could potentially be used as a measure of upper extremity function in a screening evaluation. Discriminant validity was demonstrated by the significant differences between the younger and older groups on the SFNT scores. The SFNT is a simple test that can be administered in any setting, requires no instrumentation and has been shown to be useful in other populations. In addition, the SFNT combines a standardized target with timing, which could help to reduce the interrater variability observed in other types of coordination tests and improve the longitudinal follow-up of population with ataxic features. The validation of the test should continue but its potential as a clinical tool for upper extremity function in a neuromuscular disorder with ataxic features was demonstrated.

ACKNOWLEDGEMENTS

The authors thank physiotherapist Louise Laroche and all the Neuromuscular Clinic staff of the Centre de réadaptation en déficience physique in Joliette. We also thank all the study participants. This study was carried out with the financial support of the Réseau provincial de recherche en adaptation-réadaptation of the Fonds de la recherche en santé du Québec.

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