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An examination of motor unit number index in adults with cerebral palsy

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ABSTRACT

Spinal motor neuron loss may be a factor contributing to weakness in central disorders. The aim of this study was to assess whether motor unit numbers are reduced in the hand musculature of adults with cerebral palsy (CP) using the motor unit number index (MUNIX) technique. In this prospective, case-control study, 10 adults with CP were matched with healthy controls. MUNIX was computed using area and power of voluntary surface hypothenar electromyographic (EMG) signals and the compound muscle action potential (CMAP) recorded with ulnar nerve stimulation. The motor unit size index (MUSIX) was calculated based on maximum CMAP amplitude and MUNIX value. Gross Motor Function Classification Scale (GMFCS) and Manual Abilities Classification Scale (MACS) levels were rated for CP subjects. MUNIX was significantly lower for CP participants (Mean 167.8 vs. 214.4, $p = .022$). MUNIX values did not correlate with GMFCS or MACS. MUSIX values were higher, though not significantly, for CP subjects ($p = .11$). MUSIX increased with increasing MACS levels ($r^2 = .4017$, $p = .049$). Thus, motor unit numbers in ulnar hand muscles may be decreased with CP. MUSIX values are associated with greater hand impairment. Therefore, peripheral motor unit loss as a component of the weakness found with CP deserves further evaluation.

1. Introduction

Cerebral palsy (CP) is being found with increasing prevalence in the adult US population as greater numbers of children with significant impairments from CP are surviving to adulthood (Oskoui et al., 2013; Strauss et al., 2008). Though generally described as a non-progressive motor disorder, a number of impairments appear to progress as adults with CP age with their disability (Murphy et al., 2009). Individuals with CP have been noted to develop loss of function earlier in adulthood or even as older adolescents, a phenomenon known as “early ageing” (Murphy et al., 1995). Evaluation of the potential factors that may impact the health status is needed, as quantitative assessments of the functional changes in these adults are limited (Tosi et al., 2009).

The pathophysiology of CP includes both pathological muscular changes with an increase in adipose tissue and alterations in muscle activation, both of which may potentiately contribute to functional decline (Fowler et al., 2010; Johnson et al., 2009; Ohata et al., 2008; Rose and McGill, 2005). Whether muscle weakness and atrophy after central disorders secondarily involves motor neuron degeneration has been a subject of debate. Studies have provided conflicting evidence with regard to the process of motor neuron death and/or trans-synaptic degeneration after central lesions apart from CP (Chang, 1998; Li et al., 2011; Terao et al., 1997).

Various electrophysiological techniques have been used to evaluate pathological changes in α motor neurons or to monitor disease progression or assess the effectiveness of interventions. Traditionally, intramuscular electromyography (EMG) that relies on invasive needle or fine wire electrodes has been used to obtain physiological and diagnostic information about motor unit properties. Various techniques have been described to provide estimates of motor unit numbers in vivo (Brown et al., 1988; Daube, 1995; Doherty and Brown, 1994; McComas et al., 1971; Rashidipour and Chan, 2008; Shefner et al., 2011). A less laborious or time-consuming electrophysiological technique using surface EMG recordings was described by Nandedkar in early 2000s, called the motor unit number index, or MUNIX (Nandedkar et al., 2003, 2004). The MUNIX value is calculated using data obtained from the compound muscle action potential (CMAP, or M-wave) in

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response to supramaximal electrical stimulation, in conjunction with different levels of surface EMG recordings during voluntary muscle contractions. The MUNIX value is approximately proportional to the motor unit numbers in a muscle and can be used to characterize their changes (Li et al., 2012b). MUNIX has been used in recent studies to quantify motor unit numbers in several upper and lower motor neuron disorders (Ahn et al., 2010; Boekestein et al., 2012; Bromberg, 2013; Furtula et al., 2013; Li et al., 2011, 2014; Nandedkar et al., 2011; Zhou et al., 2014) This technique has been shown to be reproducible and reliable in adults (Kaya et al., 2013, 2014; Nandedkar et al., 2011; Neuwirth et al., 2011a,b) The motor unit size index, or MUSIX can also be derived by dividing the MUNIX value into the CMAP amplitude. MUSIX values that are increased in conjunction with reductions in MUNIX values are suggestive of spinal motor neuron loss (Nandedkar et al., 2010; Li et al., 2012a).

The overall objective of this study was to assess neuromuscular changes of CP subjects using noninvasive electrophysiological recordings. Specifically, from evoked and voluntary surface EMG signals, MUNIX and MUSIX values were calculated and compared between CP and matched control subjects. The possible role of spinal motor neuron involvement in pathological changes or functional loss of CP subjects was evaluated through the examination of their affected hand muscles. The methods used in this study are non-invasive, easy to apply, and can provide supplementary information for other electrophysiological techniques.

2. Methods

2.1. Study design

This was a prospective case-control study. Our local Institutional Review Board approved this study, and all subjects gave written consent before study procedures.

2.2. Subjects

Ten adults with CP (3 male,7 female), aged from 20 to 45 years (mean ± standard deviation: 31.4 ± 2.4 years), were recruited from physician and therapy clinics at our urban rehabilitation facility and through the Cerebral Palsy Research Registry of the Department of Physical Therapy and Human Movement Sciences of the Northwestern University (Chicago, USA). Ten healthy control subjects were recruited from a prior registry of control subjects and the local community of employees of our institution or university. Control subjects were matched for age (31.2 ± 2.7 years, $p > 0.5$), sex, and side of assessment with the individuals with CP.

The following inclusion criteria were used for the study: (a) age between 18 and 65 years old; (b) medically stable with no acute illness; (c) diagnosis of CP; (d) no history of significant trauma involving the upper limbs, diabetes mellitus, renal disease, alcoholism or other disease that might affect the peripheral nerves; (e) ability to provide informed consent; (f) no history of botulinum toxin injections in the studied muscles and (g) some level of voluntary control of upper limb and hand muscles as assessed using the Upper Extremity Selective Motor Evaluation (range 0–12/limb) (Krosschell et al., 2011) Exclusion/withdrawal criteria used for the study were: (a) sensory and/or severe cognitive impairment, or other medical illnesses in the opinion of the physician investigator that would interfere with the study procedures; (b) examination or history showing clinical or neurophysiological evidence suggesting superimposed radiculopathy or peripheral neuropathy including sensory examination, motor examination and reflex testing of the upper limbs; (c) inability to provide informed consent; (d) women who were pregnant or nursing.

2.3. Clinical assessments

2.3.1. Cerebral palsy subjects

Subjects with CP were assessed for pattern of CP, (hemiplegic, quadriplegic, diplegic or triplegic) and with the following scales, commonly used to rate activity level and hand function in CP, being rated:

2.3.1.1. The Gross Motor Function Classification System – Expanded and Revised (GMFCS). The GMFCS is ordinal, five-level classification system that describes an individuals usual activity level (Palisano et al., 2007). Though initially developed for children, the GMFCS has been validated for adults. Excellent agreement between self-reported and professionally determined GMFCS levels (ICC 0.93–0.95) has been found (Jahnsen et al., 2006; McCormick et al., 2007; Sandstrom et al., 2004) Levels range from Level 1 (walks without limitations) to Level V (transported in a wheelchair).

2.3.1.2. Manual Abilities Classification Scale (MACS). The MACS is a 5 level scale that decribes an individual's usual ability to handle objects (Eliasson et al., 2006). The levels range from Level 1: Handles objects easily and successfully, to Level V: Does not handle objects and has severely limited ability to perform even simple actions.

All subjects were assessed with the following measures:

2.3.1.3. Grip strength. Subjects were seated comfortably in a chair. The torso was in an erect position with shoulder adducted and neutrally rotated, elbow flexed in 90°, and forearm in the neutral position. This is a standard posture recommended by the American Society of Hand Therapists for measuring grip strength (Fess and Moran, 1981). A portable Jamar Plus digital hand dynamometer (model EN-120604) was used to measure the maximum strength of the hand, which was determined as the highest value in three measurements. For participants with CP, the grip force was measured bilaterally and the weaker hand was used for the MUNIX and MUSIX measurements. Upper limb strength was also assessed using the Medical Research Council 0–5 scale for muscle strength.

2.4. CMAP and surface EMG measurements

Prior to the test of electrical stimulation, recording electrodes were arranged as the following: the active electrode (Ag–AgCl electrode, 1.0 cm in diameter) was placed over the hypothenar eminence with the reference electrode at the distal phalanx of the pinky finger and the ground electrode on the dorsal side of the hand. A standard bar electrode was used as the stimulating electrode and positioned over the ulnar nerve, 2 cm proximal to the wrist crease. A single pulse electrical stimulus of 200 μs was delivered to the ulnar nerve via the cathode of the bar electrode during the experiments. The compound muscle action potentials (CMAPs) were displayed and stored in the Sierra Wave EMG system (Cadwell Lab Inc, Kennewick, WA, USA).

In the beginning of the test, a low intensity of stimulus was applied to help subjects get familiar with the electrical stimulation. Individual responses were saved and displayed in the same plot for comparison convenience. The stimulation intensity increased at a rate of 2 mA per step until there was no more increase in the amplitude of muscle responses. To guarantee activation of all the motor units in the hypothenar muscle, a supramaximal electrical stimulation (with an intensity 20% higher than that achieved the maximal response amplitude) was applied. The CMAP was determined as the muscle response elicited by supermaximal stimulation.