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Electrophysiologic features of ulnar neuropathy in childhood and adolescence

Ioannis Karakis [a,b,c], Wendy Liew [a,d], Heather Szelag Fournier [a], H. Royden Jones Jr [a,b,1], Basil T. Darras [a], Peter B. Kang [a,e,f,*]

[a] Department of Neurology, Boston Children's Hospital and Harvard Medical School, Boston, MA, USA
[b] Department of Neurology, Lahey Clinic, Burlington, MA, USA
[c] Department of Neurology, Emory University School of Medicine, Atlanta, GA, USA
[d] Department of Neurology, KK Women's & Children's Hospital, Singapore
[e] Division of Pediatric Neurology, Department of Pediatrics, University of Florida College of Medicine, Gainesville, FL, USA
[f] Department of Neurology, University of Florida College of Medicine, Gainesville, FL, USA

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HIGHLIGHTS

- We analyzed patterns of ulnar nerve injury in 49 patients with pediatric ulnar neuropathy (PUN).
- There is frequent axonal and fascicular injury in PUN, similar to adults.
- In proximal axonal lesions sensory fibers to digit V and motor fibers to distal muscles are affected.

ABSTRACT

Objective: To analyze patterns of nerve injury in pediatric ulnar neuropathy (PUN).
Methods: Retrospective analysis of 49 children with PUN.
Results: Sensory loss in digit V was the prevailing complaint (89%). Predominant localization was at the elbow (55%). Diminished ulnar SNAP was the most common abnormality (71%) with median axon loss estimate (MAXE) of 62%. Dorsal ulnar cutaneous (DUC) sensory nerve action potential (SNAP) was reduced in 55% with MAXE of 43%. Abductor digiti minimi (ADM) and first dorsal interosseous (FDI) compound muscle action potential (CMAP) were reduced half of the time, with MAXE of 30% and 28% respectively. There was high correlation between ulnar sensory MAXE and ADM MAXE ($r = 0.76$, $p < 0.0001$), FDI MAXE ($r = 0.81$, $p < 0.0001$) and DUC MAXE ($r = 0.60$, $p = 0.0048$). Neurogenic changes were seen in the ADM, FDI, flexor carpi ulnaris (FCU) and flexor digitorum profundus IV (FDP IV) in 79%, 77%, 25% and 35% respectively. Pathophysiology was demyelinating in 27%, axonal in 59% and mixed in 14%.
Conclusions: In proximal axonal lesions, sensory fibers to digit V and motor fibers to distal muscles are predominantly affected, whereas in demyelinating lesions, slowing occurs twice as frequently as conduction block.
Significance: There is frequent axonal and fascicular injury in PUN.

1. Introduction

Ulnar neuropathy is the most common upper extremity mononeuropathy in children and adolescents, constituting 22% of mononeuropathies in a recent study of 2100 pediatric EMG evaluations performed over 11 years (Karakis et al., 2014). In contrast, median neuropathies predominate in the upper extremities in adults (Stewart,1987a).

Abbreviations: EMG, Electromyography; NCS, nerve conduction studies; PUN, pediatric ulnar neuropathy; IRB, institutional review board; FDI, first dorsal interosseus; ADM, abductor digiti minimi; FCU, flexor carpi ulnaris; FDP IV, flexor digitorum profundus IV; CMAP, compound muscle action potential; DUC, dorsal ulnar cutaneous; SNAP, sensory nerve action potential.

* Corresponding author at: Division of Pediatric Neurology, University of Florida College of Medicine, P.O. Box 100296, Gainesville, FL 32610, USA. Fax: +1 352 294 8067.
E-mail address: pbkang@ufl.edu (P.B. Kang).
[1] H. Royden Jones, Jr, MD is deceased.

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The clinical and electrographic features of ulnar neuropathy have been extensively studied in adults (Jabre and Wilbourn, 1979; Pickett and Coleman, 1984; Stewart, 1987b; Dunselman and Visser, 2008). These studies highlighted heterogeneous presentations, even in lesions arising from common areas of compression, such as the elbow. Specifically, sensory nerve action potentials (SNAP) are more commonly impaired when recorded at digit V, as opposed to the dorsal ulnar cutaneous (DUC) branch. Within the intrinsic hand muscles, compound muscle action potentials (CMAP) abnormalities and neurogenic changes during needle examination are more frequently encountered in the first dorsal interosseous (FDI), compared to the abductor digiti minimi (ADM). Most importantly, proximal ulnar innervated muscles, such as the flexor digitorum profundus IV (FDP IV) and, even more, the flexor carpi ulnaris (FCU) are typically spared (Stewart, 1987b). These findings are attributed to selective vulnerability of specific nerve fascicles (Sunderland, 1945; Campbell et al., 1989, 1991).

There is a scarcity of clinical and electrophysiological information on pediatric ulnar neuropathy (PUN). In the largest study of 21 patients reported two decades ago, (Felice et al., 1996) electrodiagnostic testing revealed an abnormal ulnar SNAP to digit V and an abnormal CMAP recorded from the ADM in three quarters of the patients, and demyelinating features in one quarter of them. The dorsal ulnar cutaneous (DUC) branch was studied in a minority of patients and it was absent in 40% of them. Needle examination revealed neurogenic changes in three quarters of the children. Overall, 62% of cases were deemed to be predominantly axonal, 14% mostly demyelinating, 14% mixed and 10% indeterminate.

The current study enriches our knowledge on PUN by examining a large pediatric cohort of 49 patients tested over an 11-year period in a tertiary referral center. Correlations between clinical and electrophysiologic localizations, along with neurophysiological patterns, were of particular interest.

2. Methods

2.1. Subjects

The study was approved by the Committee on Clinical Investigation (institutional review board) at Boston Children's Hospital. Participants up to 21 years of age and/or their parents/guardians gave their informed consent for the diagnostic procedures prior to their electrodiagnostic studies. A total of 2100 electrodiagnostic studies performed in the Electromyography Laboratory at Boston Children's Hospital from January 1, 2001 to December 31, 2011 were reviewed retrospectively. Characterization of the overall cohort has been previously published, but the patients with ulnar neuropathy were not discussed in detail (Karakis et al., 2014). The majority were performed in the outpatient setting and without sedation. Patients who met criteria for PUN were subjected to further data extraction and analysis for the current study. Clinical data collected included age, gender, disease duration, symptoms and signs. Electrophysiologic data collected included ulnar nerve slowing/conduction block and onset latencies recorded from the ADM and FDI, ADM and FDI CMAP and axon loss estimate, digit V ulnar and DUC SNAP amplitude and axon loss estimate, and needle EMG neurogenic changes in the muscles supplied by the ulnar nerve. As dedicated imaging of the ulnar nerve via ultrasound or MRI was not performed systematically in the cohort, imaging data were not collected for this study.

2.2. Electrophysiological data

Standard protocols and reference values were used for nerve conduction studies (NCS) and needle EMG (Jones et al., 1996; Kang, 2007; Darras et al., 2015). Studies were performed primarily on TECA^{TM} Synergy EMG monitoring systems (Oxford instruments, Oxford, UK), with a minority performed on an XLTEK NeuroMax EMG machine (Excel-Tech, Ltd., Ontario, Canada). Upper extremity temperatures were generally maintained at or above 32 °C. The data were interpreted by experienced neuromuscular neurologists according to standard criteria (Darras et al., 2015). Given that all patients were ≥5 years old, we used normative data akin to the adult population (ulnar SNAP to digit V: ≥17 μV amplitude, ≥ 50 m/s conduction velocity; DUC SNAP: ≥8 μV amplitude, ≥50 m/s conduction velocity; ulnar to ADM CMAP: ≥6 mV amplitude, ≥49 m/s conduction velocity; ulnar to FDI CMAP: ≥7 mV amplitude, ≥49 m/s conduction velocity) (Preston and Shapiro, 2005).

The diagnosis of ulnar neuropathy was based on electrodiagnostic criteria. Nevertheless, the exact electrodiagnostic test protocol was determined by the indication, targeted history and physical examination, abnormalities identified during the study, and the patient's tolerance. When tolerated, comparisons with the contralateral side were performed.

Ulnar motor nerve conduction studies were performed recording from the ADM and/or the FDI. An estimate of ulnar motor nerve conduction block was obtained from the formula $(B-A)/B \times 100$, where B represents the ADM or FDI response amplitude on the affected side below the elbow and A represents the ADM or FDI response amplitude on the affected side above the elbow. An estimate of ulnar motor nerve axon loss was obtained from the formula $(U-A)/U \times 100$, where U represents the ADM or FDI response amplitude from the unaffected side or the standard control value for age (when testing of the unaffected side was not performed), and A is the ADM or FDI response from the affected side. This calculation has been used in adult (Brown and Watson, 1991; Watson and Brown, 1992; Kang et al., 2005) and pediatric EMG studies (Karakis et al., 2017). F responses were recorded infrequently due to patient discomfort in the pediatric age range. Antidromic sensory nerve conduction studies of the ulnar to digit V and DUC were typically obtained, with contralateral studies recorded when tolerated. An estimate of ulnar sensory nerve axonal loss was obtained from the same formula $(U-A)/U \times 100$, where U represents the ulnar SNAP amplitude from the unaffected side or the standard control value for age (when testing of the unaffected side was not performed), and A is the ulnar SNAP from the affected side (Kang et al., 2005). Concentric needle examination was performed in all patients in distal and proximal ulnar innervated muscles, as well as non-ulnar innervated muscles when appropriate and tolerated.

Inclusion criteria for the diagnosis of ulnar neuropathy were abnormalities confined to the ulnar nerve distribution. Exclusion criteria were concurrent abnormalities in the median or radial nerve distributions on NCS and in non-ulnar innervated muscles on needle EMG. The ulnar neuropathy was deemed primarily demyelinating when conduction block (defined as a 50% reduction in amplitude) or conduction slowing of >10 m/s by comparing stimulation above and below the elbow was present. In such cases, the degree of clinical weakness and recruitment reduction would be expected to outweigh signs of axonal loss. Similar to other mononeuropathies, a category of pure conduction block was not designated since any significant demyelination could be associated with mild axonal loss (Wilbourn, 1986). The ulnar neuropathy was deemed primarily axonal when the ulnar sensory study or the ulnar motor studies to the ADM and/or FDI identified a >50% amplitude drop throughout the course of the nerve compared to the contralateral side, or, when contralateral testing was not performed, of the normal value for age. Needle EMG in such cases demonstrated acute and/or chronic neurogenic changes in the form of abnormal spontaneous activity and motor unit remodeling