The Role of Electromyography and Nerve Conduction Studies in Ulnar Longitudinal Deficiency: A Case Report

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The Role of Electromyography and Nerve Conduction Studies in Ulnar Longitudinal Deficiency: A Case Report

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ABSTRACT

Background. Ulnar longitudinal deficiency (ULD) is a rare disorder that generally affects the entire upper limb, including the elbow, forearm, and hands

Case Report. We present the case of a 17-month-old girl with ULD affecting her right upper extremity, which was supported by radiological examination. The nerve conduction studies (NCS) indicated axonal sensory neuropathy of the right median and ulnar nerves, and cross-innervation from median to ulnar nerve. The median, ulnar, radial motor, and radial sensory nerves were all normal. Needle electromyography (EMG) revealed normal motor unit action potentials in all muscle samples. EMG and NCS serve as diagnostic screening tools providing valuable information to identify and locate nerve pathology.

Conclusions. Prompt and thorough knowledge and management of motor and neurological abnormalities are essential for ULD patients' future functionality.

Keywords: Postaxial longitudinal deficiency; Ulnar club hand; Nerve conduction study; Upper extremity anomaly; Bone deficiency; Electrodiagnosis.

Abbreviations:

APB : Abductor pollicis brevis

DIP : Distal interphalangeal joint

EDC : Extensor digitorum communis

EMG : Electromyography

FCR : Flexor carpi radialis

FCU : Flexor carpi ulnaris

MIP : Middle interphalangeal joint

MMT : Manual muscle testing

MUAP : Motor unit action potentials

NCS : Nerve conductions studies

PIP : Proximal interphalangeal joint

ROM : Range of motion

ULD : Ulnar longitudinal deficiency

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INTRODUCTION

Ulnar longitudinal deficiency (ULD) is also known as "congenital ulnar hemimelia," "postaxial longitudinal deficiency of the upper limb," "ulnar club hand," or "ulnar ray deficiency"[1]. This rare condition typically affects the entire upper extremity, including the elbow, forearm, and hands, and is characterized by the partial or complete absence of ulnar bone formation with complex carpal, metacarpal, and digital abnormalities [1,2]. The etiology of ULD is attributed to a deficiency in the Sonic Hedgehog, which is responsible for the formation of ulnar-sided forearm structures and four ulnar-sided digits [3]. It affects 1-2 in 100,000 children, with a predominant occurrence in males compared to females at a ratio of 3:2. Most cases (70%) present as right-sided and unilateral, characterized by ulnar deviation of the hand and shortened forearm [4–6] Electromyography (EMG) demonstrates substantial cross-innervation of the intrinsic muscles from both the medial and lateral nerves to the median nerve in congenital upper-extremity anomalies [7]. Physiatrists aid patients in adjusting to physical or cognitive constraints that impede function, crucially contributing to interdisciplinary teams, especially for children with upper-limb deformities [8]. EMG and nerve conduction studies (NCS) in patients with ULD have not been previously reported.

CASE REPORT

A 17-month-old female presented with a right upper extremity deformity. Born preterm at 32 weeks via cesarean section due to breech presentation with a malformed right forearm and hand with three fingers. Birth anthropometric parameters were normal. The mother was 36 years old during pregnancy, had no history of illness, medication use, or environmental exposure during gestation. The patient can grasp objects like milk bottles, mobile phones, and toys with her right hand. She exhibits normal mobility, feeding, and communication skills for her age and performs age-appropriate daily activities independently. There is no family history of congenital anomalies.

Physical examination revealed normal vital signs, and age-appropriate body weight, height, and body mass index (9.2 kg, 78 cm, 15.1 kg/m^2). The right upper extremity exhibited deformity with wrist in flexion and ulnar deviation position, and absence of 4th and 5th digits (Figure 1). The range of motion (ROM) of the right shoulder was full, stiff of the elbow joint, and limited ROM in the wrist and fingers. Manual muscle testing (MMT) of the right shoulder was functional, the elbow was non-functional, and the wrist and fingers were weak functional, with no signs of lower or upper motor neuron lesions. The patient had isolated hand involvement of the unaffected side and unable holding large or heavy objects of household items that necessitate bimanual ability.

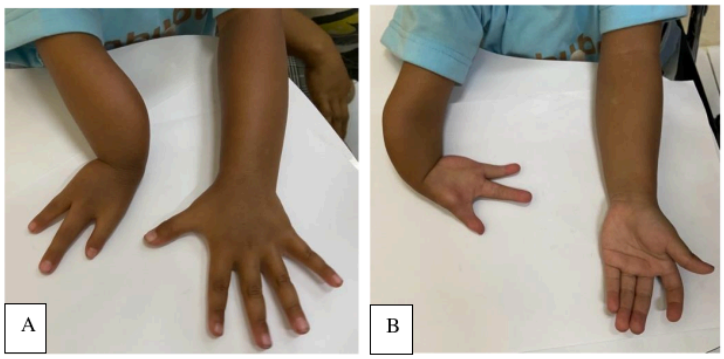
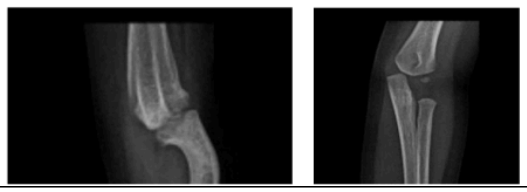


Figure 1. Clinical pictures of patient's upper extremity A) Dorsal side, B) Volar side

Radiographic examination of the right antebrachium showed radiohumeral synostosis, complete ulna absence, carpal bone deficiency, and missing MIP, PIP, and DIP joints in digits IV and V, alongside a bowed radius, with the left upper extremity appeared normal (Figure 2). EMG and NCS were performed to assess the muscular and neurological conditions of the upper extremity. NCS findings revealed axonal neuropathy in the right median and ulnar nerves. The median, ulnar, radial motor nerves, and radial sensory nerves were normal. EMG findings showed normal motor unit action potentials (MUAPs) of the deltoid, brachioradialis, EDC, FCR, APB, FCU, and 1st dorsal interossei muscles (Figure 3).



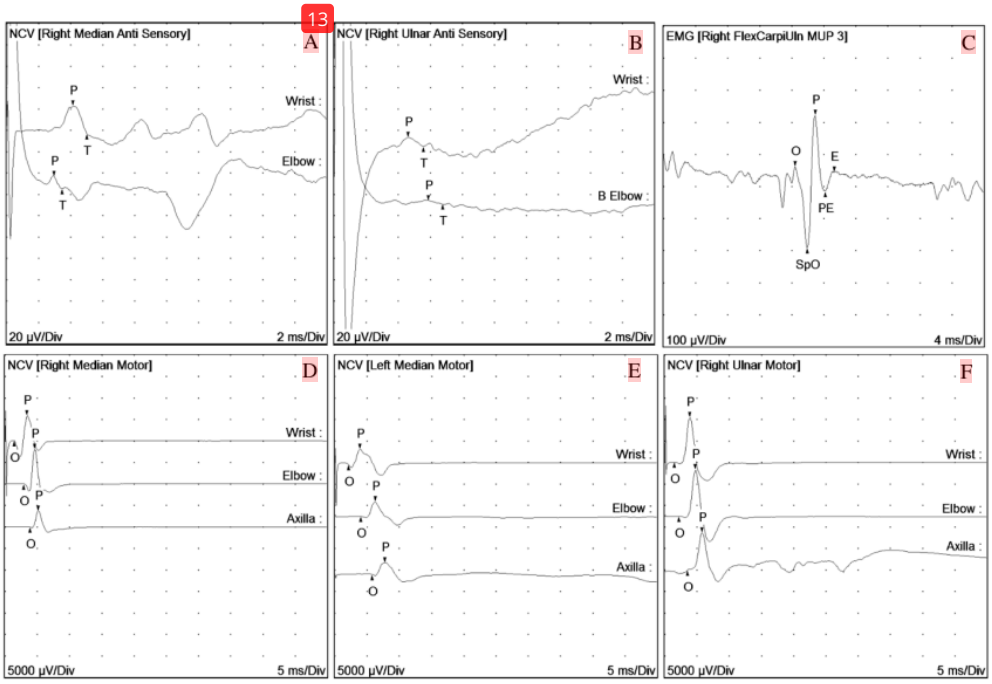


Figure 3. EMG and NCS waveforms A) & B) Axonal neuropathy sensory of right median and ulnar nerve, C) Normal MUAP of muscle sample (FCU), D, E, F) Median and ulnar motor

After 6 months of rehabilitation, the patient more frequently used her right hand to play and do activities, able to carry large and heavier household objects using both hands, ride and steer bicycle, with no reported complications. Surgical intervention will be considered once the patient reaches two years of age, allowing for a larger hand size that facilitates more precise surgery and better functional outcomes.

DISCUSSION

Patients with ULD manifest varying degrees of severity. The widely adopted Bayne classification categorizes ULD describe the elbow and forearm, while Cole and Manske classify based on the thumb and first web. The categorization directs the medical experts' attention to the most crucial deficits that need addressing to restore functionality [9]. The patient exhibited type-IV Bayne and type B Cole and Manske classification.

Congenital hand and nerve anomalies have been under-researched, with neural abnormalities linked to ULD scarcely documented. Surgical observation in one case revealed a forearm with a single bone (radius) and a hand with one finger, showing only one artery (radial artery) and one nerve (median nerve) [9]. A case study detailed a patient with intact median and ulnar nerves and two forearm bones. However, this anatomical setup may vary among individuals with ULD, necessitating further research to explore ulnar nerve anatomy in different ULD severities [10]. EMG and NCS are diagnostic tools for medical experts and surgeons to assess current and potential function. They validate nerve pathology diagnoses, identify concurrent issues, locate neurological lesions, and clarify clinical findings [11,12]. The same NCS waveforms of the ulnar and median motors were found, suggesting cross-innervation from the ulnar to the median. This EMG study can be informative for pre-surgical planning, to identify anatomical variations, locate the site and degree of nerve dysfunction, and determine functional recovery potential before performing surgical interventions, if necessary.

The approach in managing ULD between operative and nonoperative treatments is determined by the function of the limb [2]. Rehabilitation, child psychology, and parental counselling are important concepts to remember when dealing with the upper limb deformity population. The treatment goal is to restore function to the maximum extent possible, especially in bimanual ability, enhancing limb functionality, and overall quality of life [8,13,14]. Conservative options include stretching and splinting beginning at a young age [8,15]. A hand splint is suggested to limit ulnar deviation, increase radial deviation, protect wrist structures, train wrist muscle balance, and assist functional hand movements. Psychologists provided acceptance and commitment therapy and parental counseling to help the patient and family cope with psychological impairments. Surgical procedures are being considered if hand function does not improve and ulnar deviation progresses.

CONCLUSION

ULD is a rare congenital condition involving longitudinal deficiency and digital anomalies. This report details the EMG and NCS findings in a ULD patient, contributing to existing knowledge and offering clinical and research insights. A thorough understanding of these

conditions aids medical teams and surgeons in planning interventions to improve functionality and reduce complications.

PATIENT CONSENT

The authors obtained written consent of legal guardians for presentation of the cases within the present scientific paper

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest regarding the publication of this manuscript.

AUTHOR'S CONTRIBUTIONS

Conceptualization, AS, SMW, RDH. Methodology, AS, NAR, DFC. Formal analysis, VAJ. Investigation, AS, SMW, RDH. Writing original draft preparation, AS, VAJ. Writing—review and editing, AS, NAR, DFC. Visualization, AS, NAR, DFC. Supervision, SMW, RDH. Project administration, SMW, RDH. All authors have read and agreed to the published version of the manuscript".

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