

Outcome of Neuromuscular Electrodiagnostic Testing in Children

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ABSTRACT

This study aimed to describe the spectrum of the results of neuromuscular electrodiagnostic investigations performed by age and pathology in paediatric patients. This retrospective observational study took place from January 2019 to December 2021. Electrodiagnostic records of paediatric patients (birth to 15 years) were referred to the electromyography (EMG) laboratory for EMG and nerve conduction study (NCS). Electrodiagnostic and demographic data were collected. Among these, 151 (56%) were males and 118 (44%) were females. Forty-four percent tests came out to be normal and 151(56%) tests revealed electrophysiological abnormality. Peripheral nerve neuropathy (n = 39, 14.5%), polyneuropathy (n = 35, 13%) and myopathy (n = 28, 10%) were the most common electrophysiological diagnoses. The electrodiagnostic test helps in differentiating the aetiologies, thus guiding the subsequent work-up logically in the paediatric population.

Key Words: Brachial plexus neuropathies, Electromyography, Duchene muscular dystrophy, Neuromuscular junction disorders, Nerve conduction studies, Acute inflammatory demyelinating polyneuropathy, Rehabilitation, Electromyography, Hereditary sensory motor polyneuropathy.

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Electrodiagnostic testing plays a significant role in diagnosing neuromuscular disorders among the paediatric population. The test is vital to localise the lesions of the peripheral nervous system, to differentiate between neurogenic, myopathic or neuromuscular junction (NMJ) disorders, to grade the severity and prognosis of disease, to monitor disease progression, and to rule out conditions which do not involve the peripheral nervous system.¹ Although the fundamental principles of the electrodiagnostic test are the same for adult and paediatric population, there are significant differences that electromyography needs to keep in mind when studying infants and children. These include variations in pathology, normative values, and technical factors.² Myelination is completed by the child's fifth birthday, due to which nerve conduction velocities in full-term infants are approximately half of the normal adults.² Paediatric patients have a smaller nerve diameter as compared to adults.² Given the short stature of paediatric patients, the nerve lengths of children are shorter than adults.²

Studies assessing paediatric patients electrophysiologically have either focused on a specific diagnosis, evaluated the clinical electromyography (EMG) compliance, or reviewed the compliance and consistency between the preliminary diagnosis and the electrophysiological diagnosis.^{1,3,4} Local data regarding paediatric electrodiagnostic tests in Pakistan are limited. The aim of this study was to investigate the diagnoses established as a result of electrophysiological investigation in the paediatric population and its distribution of age groups and gender.

This retrospective observational study was conducted in the electrodiagnostic department of Combined Military Hospital, Kharian, from January 2019 to December 2021. All patients up to 15 years of age referred to the electrodiagnostic department were included. Patients with incomplete medical records were excluded.

EMG and NCS reports of patients were the primary source of data. Electrodiagnostic reports were de-identified before analysis. The age, gender, and presenting symptoms of patients were noted down. The patients were classified according to age group and pathology; for example, plexopathy, nerve root lesions, polyneuropathy, myopathy, neuropathy, anterior horn disease, neuromuscular junction transmission disorder, facial paralysis, and other rare diseases. SPSS version 21 was used for statistical analysis. Frequencies and percentages were calculated for qualitative variables. Chi-square test was used for comparison of electrophysiological diagnosis between male and female patients. A p-value <0.05 was considered statistically significant.

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Table I: Gender distribution of electrodiagnostic pathologies in the paediatric population.

Diagnosis	Gender of Patient		p-value*
	Male (n = 151) n (%)	Female (n = 118) n (%)	
Normal Study	71 (47%)	46 (39%)	0.15
Polyneuropathy	16 (11%)	19 (16%)	
Myopathy	18 (12%)	10 (8%)	
Plexopathy upper limb	10 (7%)	13 (11%)	
Neuromuscular junction disorders	1 (1%)	1 (0.8%)	
Anterior horn cell disorders	7 (5%)	5 (4%)	
Peripheral nerve neuropathy	21 (14%)	18 (15%)	
Facial nerve paralysis	1 (1%)	0 (0)	
Polyradiculopathy	6 (4%)	6 (5%)	

*Chi-square test was applied; p-value of less than <0.05 was considered significant.

A total of 2215 NCS/EMG tests were performed in the department from January 2019 to December 2021. Out of these, 269 (12.14%) tests were conducted on paediatric population which met the inclusion criteria. Among these, 151 (56%) were males and 118 (44%) were females. Common symptoms of patients upon presentation to electrodiagnostic clinic were mono or hemiparesis (26.1%), weakness in all four limbs (19%), lower limb weakness (17%), delayed walking (13%), delayed milestones (7.3%), acute sudden weakness (6.3%), foot drop (5.2%), frequent falls (1.8%), pain lower limbs (1.7%), wrist drop (1.4%), and drooping of eye lids, regression of milestones, decreased sensation of limb (0.4% each).

Out of the 269 tests, 118 (44%) tests were normal, and 151 (56%) tests revealed an electrophysiological abnormality. Among patients with normal electrodiagnostic tests, 32 (27%) patients had upper motor neuron signs suggesting central pathology (cerebral palsy). Peripheral nerve neuropathy (39, 14.5%), polyneuropathy (35, 13%), and myopathy (28, 10%) were the most common electrophysiological diagnosis among paediatric population in this study. The most common diagnosis in children aged 0-5 years was upper limb plexopathy (19, 14%). Polyneuropathy (14, 18%) was more frequently diagnosed in children aged 6-10 years. Meanwhile, peripheral nerve neuropathy (11, 20%) was the most common diagnosis in children aged 11-15 years. Gender-wise distribution of electrodiagnostic pathologies can be seen in Table I.

Of the total, 39 (14.5%) patients who had peripheral nerve neuropathy, 15 (38%) had it because of injection neuritis and 24 (62%) had it due to trauma. Nerves involved in injection neuritis were sciatic nerve (n = 14, 93%) and radial nerve (n = 1, 7%). Nerves injured in cases of traumatic peripheral nerve neuropathy were common peroneal nerve and ulnar nerve (n = 5; 21% each); radial nerve, combined median, ulnar and radial neuropathy (n = 4; 17% each); combined common peroneal and tibial in 2 (8%); and combined median and ulnar nerve median nerve and sciatic nerve involvement in one (4%) patient each.

A total of 35 (13%) patients had polyneuropathy. The distribution of polyneuropathy was; hereditary sensorimotor neuropathy (HSMN) in 20 (59%), Guillian-Barre syndrome in 7

(20%), sensory polyneuropathy in 4 (12%), and motor polyneuropathy in 4 (12%). The types of Guillian-Barre syndrome observed in this study were acute inflammatory demyelinating polyneuropathy (n = 4; 57%), acute motor sensory axonal neuropathy (n = 2; 28%) and acute motor axonal neuropathy (n = 1; 14%). Among patients with HSMN observed in this study, the common types were HSMN Type-II (n = 8; 40%) and HSMN Type-I (n = 4; 20%).

Of the total, 28 (10%) patients who had an electrophysiological diagnosis of myopathy, 8 (29%) patients had dystrophic myopathy while 20 (71%) patients had non-dystrophic myopathy.

Out of 12 (4.5%) patients who had anterior horn cell disorders, 10 (83%) had spinal muscular atrophy and 2 (17%) patients had poliomyelitis. In addition, 12 (4.5%) patients had lumbosacral polyradiculopathy. Furthermore, 2 (0.7%) patients had neuromuscular junction disorder. Finally, 1 (0.4%) patient had facial nerve paralysis (lower motor neuron type).

Nearly, half of the patients (44%) had a normal electrophysiological investigation. It may be because diseases of central pathology like cerebral palsy have normal peripheral nerves and muscles, which would result in a normal study. However, it is also possible that the disease may not have yet been exhibited electrophysiologically (such as early-stage polyneuropathy). Moreover, technical issues like the cooperation of the patient and adequate muscle activation may result in false negative results.

In the 0-5 years age group, plexopathy was the most common electrophysiological pathology, which was consistent with the study conducted by Orhan *et al.*³ Unfortunately, congenital brachial plexopathies exist worldwide with incidence ranging from 0.15 to 3 per 1000 live births.⁴ Caesarean section decreases the risk of brachial plexopathy.⁴ Risk factors include maternal diabetes, multiparity, foetal macrosomia, intrauterine torticollis, breech presentation, assisted delivery, and extended delivery period.⁴ Timing of EMG for these babies was controversial because most patients recovered within the first three months with conservative management.⁴ The authors preferred to conduct an electrodiagnostic test in these patients at three months.

The most common diagnosis in this study's 6-10 years age group was polyneuropathy (18%). Hereditary sensory motor polyneuropathy was the most common polyneuropathy in this group. The diagnosis was made based on clinical and electrophysiological findings. Genetic studies were not performed due to their non-availability. Demyelination was more prevalent in children with GBS in this study, which is consistent with the study conducted by Zia *et al.* in Pakistan.⁵ Myopathy (9.4%) was the most common electrophysiological diagnosis in this age group as reported by Orhan *et al.*³

In the age group of 10-15 years, mononeuropathy (20%) was the most common diagnosis in this study. Orhan *et al.* reported that polyneuropathy was the most common diagnosis in this age group.³ Mononeuropathy is more prevalent in authors' setup because of iatrogenic injection neuritis and post-traumatic nerve injuries. Injection neuritis exists worldwide, but the incidence is very high in this study. The reason may be that majority of this patient population belonged to lower socioeconomic and rural areas where health facilities are not up to the mark. Many healthcare workers were not adequately trained in nursing and safe injection practices.⁶

The study design was one of the limitations of this study because one cannot comment on the prognostic value of electrodiagnostic tests and consistency between the preliminary diagnosis and electrophysiological findings. The electrodiagnostic test helps differentiate the possible aetiology and guides subsequent workup more logically. Based on the age group of the paediatric patient, one can make a diagnosis by considering the electrophysiological findings.

ETHICAL APPROVAL:

The study was approved by Ethical Committee of Combined Military Hospital, Kharian (Letter No. 2021/09).

COMPETING INTEREST:

The authors declared no competing interest.

AUTHORS' CONTRIBUTION:

UA: Conception of study design, collection of data, manuscript writing, critical review.

MTK: Conception of study design, collection of data, manuscript writing, statistical analysis, critical review.

NA, AA: Manuscript writing, critical review.

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