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MEDICAL SCIENCES / DAHİLİ TIP BİLİMLERİ

How Accurately is the Pre-diagnosis of Electroneuromyography in Pediatric Patients?

Pediatrik Hastalarda Elektronöromiyografi Ön Teşhisi Ne Kadar Doğru?

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Abstract

Objectives: Electroneuromyography (ENMG) is a laboratory test commonly used in the diagnosis of neuromuscular diseases, which is a continuation of anamnesis and physical examination. The aim of this study is to analyze accordance between pre-diagnosis and electrophysiological diagnosis in pediatric patients.

Materials and Methods: Patients aged 0-16 years and underwent ENMG examinations between 2016 and 2019 were evaluated retrospectively.

Results: The ENMG results of a total of 526 patients were examined in the study. The mean age of the patients was 10.81±4.31 years. While the pre-diagnosis and the electrophysiological diagnosis were compatible in 32.31% of the patients, incompatibility with the pre-diagnosis was found in 67.69%. While no difference was found between concordant and discordant patients in terms of pre-diagnosis and outcome between genders (p=0.06), there was a difference between concordant and non-concordant patients in terms of categorized age and pre-diagnoses (p=0.01, p=0.02, respectively).

Conclusion: A high discrepancy was found between pre-diagnosis and electrophysiological diagnosis. With more detailed history and clinical examination of pediatric patients, unnecessary requests can be avoided and waiting times of real patients can be shortened.

Key Words: Electromyography, Children, Examination And Diagnoses

Öz

Amaç: Elektronöromyografi (ENMG), anamnez ve fizik muayenenin devamı niteliğinde olan nöromusküler hastalıkların tanısında yaygın olarak kullanılan bir laboratuvar testidir. Bu çalışmanın amacı, çocuk hastalarda ön tanı ile elektrofizyolojik tanı arasındaki uyumu incelemektir.

Gereç ve Yöntem: Çalışmada 0-16 yaş arası olan ve 2016-2019 yılları arasında ENMG tetkikleri yapılan hastalar retrospektif olarak değerlendirildi.

Bulgular: Çalışmada toplam 526 hastanın ENMG sonuçları retrospektif olarak incelendi. Hastaların ortalama yaşı 10,81±4,31 yıl idi. Hastaların %32,31'inde ön tanı ile elektrofizyolojik tanı uyumlu iken, %67,69'unda ön tanı ile uyumsuzluk saptandı. Cinsiyetler arasında ön tanı ve sonuç açısından uyumlu ve uyumsuz hastalar arasında fark bulunmazken (p=0,06), yaş ve ön tanı kategorisi açısından uyumlu ve uyumsuz hastalar arasında fark vardı (p=0,01, p=0,02, sırasıyla).

Sonuç: Ön tanı ile elektrofizyolojik tanı arasında yüksek bir tutarsızlık bulundu. Pediatrik hastaların daha detaylı öykü ve klinik muayenesi ile gereksiz isteklerden kaçınılabilir ve gerçek hastaların bekleme süreleri kısaltılabilir.

Anahtar Kelimeler: Elektromiyogarfi, Çocuklar, Muayene Ve Tanı

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Introduction

Electroneuromyography (ENMG) is a commonly used laboratory test in the diagnosis of neuromuscular diseases, which is a continuation of anamnesis and physical examination. ENMG examination provides benefits to the clinician such as diagnosis, prognosis, treatment follow-up, treatment selection, and guiding surgery (1). Clinicians need ENMG evaluations in pediatric patients as well as in adult patients (2). Different ENMG techniques such as nerve conduction studies, needle electromyography (EMG), single fiber EMG, evoked potentials, and cascade stimulation used in adult patients can also be applied in the pediatric age group (1,3). ENMG examinations performed in the pediatric patient group contain some differences compared to the adult patient group (2,3). Differences in normal values for each age group, the difference in the spectrum of neuromuscular diseases in the pediatric patient population from the adult population, and the inability to routinely apply standard ENMG protocols due to low tolerance of pediatric patients are some of these differences (2-4). Considering these, the ENMG examination in the pediatric population is a laboratory examination that requires clinical experience. There are studies in the literature examining the compatibility between pre-diagnosis and electrophysiological diagnosis in the pediatric patient population in their own ENMG laboratories (5-8).

Our aim in this study was to evaluate the distribution of examinations in pediatric patients by age, performed between 2016-2019 in our own ENMG laboratory, and to evaluate the accordance between pre-diagnosis and electrophysiological diagnosis.

Materials and Methods

This study was approved by the Local Institutional Ethics Committee of University of Health Sciences Turkey, Dışkapı Yıldırım Beyazıt Training and Research Hospital (approval date: 23.12.2019; approval number: 78/01) and was conducted in accordance with the Declaration of Helsinki guidelines. In this study, ENMGs, performed on children under 16 years of age in the ENMG laboratory of the physical medicine and rehabilitation clinic of our hospital between 2016-2019, were examined. The age of the patients and the year in which the test was performed were noted from their files. Pre-diagnoses were divided into anterior horn motor neuron disease (MND), radiculopathy, plexopathy, peripheral nerve lesion, polyneuropathy (PP), entrapment neuropathy of lower-upper extremity, myopathy, neuromuscular junction disease, facial paralysis, and Guillain-Barre syndrome (GBS). The patients were divided into 0-2 years, 3-6 years, and 7-16 years and pre-diagnosis frequency in these age groups were examined. The accordance between the pre-diagnosis and the diagnosis after ENMG was examined.

Statistical Analysis

Statistical analyzes were made using the SPSS 20.0 program. The chi-square test was used for examining statistical analysis. A value of $p<0.05$ was considered statistically significant.

Results

The ENMG results of 526 patients in total were examined in the study. The mean age of the patients was 10.81±4.31. Two hundred and thirty-three of the patients (44.3%) were female and 293 (55.7%) were male. Twenty-eight (5.3%) of the ENMG examinations were performed on patients between 0-2 years, 97 (18.4%) on 3-6 years, and 401 (76.2%) on 7-16 years old patients. The distribution of ENMG numbers by age was shown in Figure 1. Accordingly, EMNG was applied mostly to patients aged 15 years [71 (13.5%)], followed by patients aged 16 years [60 (11.4%)] and 13 years [55 (10.5%)], respectively. The distribution of ENMG numbers made by years was shown in Figure 2. ENMG was administered to 83 (15.8%) patients in the pediatric age group in 2016, 129 (24.5%) in 2017, 211 (40.1%) in 2018 and 103 (19.6%) in 2019.

The distribution of ENMGs according to pre-diagnoses was shown in Table 1. In our laboratory, ENMG examination was mostly performed with a pre-diagnosis of purine nucleoside phosphorylase (PNP) at a rate of 50.7%. The most common pre-diagnosis (32.1%) in patients aged 0-2 years is plexopathy. In patients between the ages of 2-6 and 7-16, the most common pre-diagnosis was PNP with rates of 42.2% and 54.3%, respectively.

The accordance between pre-diagnosis and electrophysiological diagnosis and diagnoses after electrophysiological examination were shown in Table 2. One EMNG was performed with the pre-diagnosis of MND, and it was found to be 100% compatible with the pre-diagnosis. A total of 20 patients were evaluated with the pre-diagnosis of radiculopathy and 23.8% were found to be compatible with the

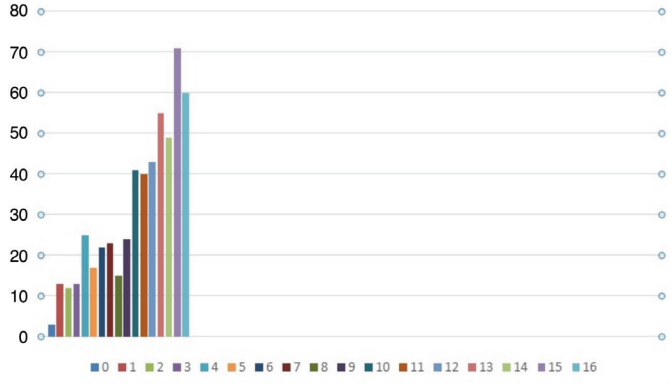

Figure 1: Distribution of ENMG numbers by age

ENMG: Electroneuromyography

pre-diagnosis; while the electrophysiological examination of the patients who presented with this pre-diagnosis revealed that 66.7% had normal results, 4.8% had plexus damage, and 4.8% had entrapment neuropathy. Twenty patients who presented with a pre-diagnosis of plexopathy were examined, 14 patients (74.1%) obtained a result consistent with the pre-diagnosis, while 5 (18.5%) patients had normal results and 1 (7.4%) patient had a peripheral nerve lesion. Sixty-six of the patients were evaluated with a pre-diagnosis of peripheral nerve lesion and an electrophysiological diagnosis compatible with the pre-diagnosis was found at a rate of 63.6%. The results of 21 (31.8%) incompatible patients were found to be normal, 1 (1.5%) had radiculopathy, and 2 (3%) had entrapment neuropathy.

Two hundred and sixty-seven patients were evaluated with PNP pre-diagnosis, and PNP was detected in 42 patients (15.73%). The result was normal in 214 (80.1%) of these patients, radiculopathy in 2 (0.7%), peripheral nerve lesion in 2 (0.7%), myopathy in 5 (1.9%), and entrapment neuropathy in 2 patients (0.7%) were determined. As a result, 84.2% of the patients were found to be inconsistent with the pre-diagnosis after the electrophysiological examination.

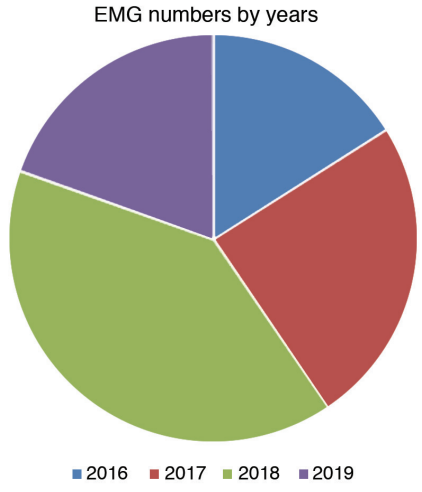

Figure 2: Distribution of ENMG numbers by years
ENMG: Electroneuromyography

Thirty-eight of the patients were referred to our laboratory with a pre-diagnosis of myopathy and results consistent with the pre-diagnosis were obtained in 9 (23%) patients; the results of 23 (60.52%) of the patients referred with this pre-diagnosis were normal, and PNP was found in 6 (15.78) of them.

Electrophysiological diagnosis compatible with the pre-diagnosis was found in 18.8% of entrapment neuropathy of the upper extremity and in 66.8% of entrapment neuropathy of the lower extremity. 58.6% of the patients referred with the pre-diagnosis of GBS had a normal result; on the other hand, 41.37% of patients had electrophysiological findings compatible with GBS.

Among the years examined, 335 (63.7%) of EMNG examinations performed with any pre-diagnosis were found to be normal. While the pre-diagnosis and the electrophysiological diagnosis were compatible in 32.31% of patients, incompatibility with the pre-diagnosis was found in 67.69%.

In the chi-square analysis, no difference was found between concordant and discordant patients in terms of pre-diagnosis and outcome between genders (p=0.06), while there was a difference between concordant and non-concordant patients in terms of categorized age and pre-diagnoses (p=0.01, p=0.02, respectively).

Discussion

This study examines the distribution of EMNG studies conducted in the pediatric patient population between 2016

Table 1: Pre-diagnoses by age

	Total N=526	0-2 years N=28	2-6 years N=97	7-16 years N=401
MND	1 (0.2%)			1 (0.2%)
Radiculopathy	21 (4%)	2 (7.1%)	4 (4.1%)	15 (3.7%)
Plexus damage	27 (5.1%)	9 (32.1%)	6 (6.2%)	12 (3.0%)
Traumatic peripheral nerve lesion	66 (12.5%)		19 (19.6%)	47 (11.7%)
PNP	267 (50.7%)	7 (25%)	41 (42.2%)	219 (54.6%)
Myopathy	39 (7.4%)	5 (17.9%)	6 (6.2%)	28 (7%)
Myopathy + neuropathy	10 (1.9%)		5 (5.2%)	5 (1.2%)
Facial Palsy	15 (2.9%)		4 (4.1%)	11 (2.7%)
Entrapment neuropathy of the upper extremity	32 (6.1%)		1 (1%)	31 (7.7%)
Entrapment neuropathy of the lower extremity	19 (3.6%)			19 (4.7%)
GBS	29 (5.5%)	5 (17.9%)	11 (11.3%)	13 (3.2%)

MND: Motor neuron disease, GBS: Guillain-Barre syndrome, PNP: Purine nucleoside phosphorylase

and 2019 in our clinic according to age, age groups, pre-diagnoses, and the consistency between pre-diagnoses and electrophysiological diagnoses based on each pre-diagnosis.

In our study, it was observed that most pediatric patients who applied to our electrophysiology laboratory for ENMG examination were patients between the ages of 7-16. PNP was the most common pre-diagnosis in all pediatric patients and with this pre-diagnosis, patients between 2-6 years and 7-16 years of age were referred to the clinic most frequently. On the other hand, the situation was found to be slightly different between the ages of 0-2, the most common pre-diagnosis in this age group was found to be plexopathy. 67.69% of ENMG examinations performed between these years were found to be inconsistent with the pre-diagnosis. When the pre-diagnoses are examined separately, a high rate of non-compliance is observed, especially in patients referred to the laboratory with a PNP pre-diagnosis.

ENMG applications in pediatric patients are evaluations that require clinical experience and can be guiding in the diagnosis, treatment planning, and follow-up (9). Due to difficulties such as the difference in normal values in every age group due to the incomplete development of the nervous system, difficulties of working in smaller areas, and the limited tolerance of the pediatric patient group, it is aimed to reach the most information with the least pain by going beyond the protocols applied in adults from time to time (10).

In a study examining EMNG applications in adults in our country, the most common pre-diagnoses in adults were carpal tunnel syndrome, PNP, and radiculopathy (11). In a study by Yagci et al. (5) in which they shared their own electrophysiological experiences in a group of pediatric patients, the most common pre-diagnosis was plexopathy, flaccid infant, and spinal atrophy during the infancy and toddler period, while PP and peripheral nerve damage were found to be the most common pre-diagnosis in later ages. In another study conducted in our country, the frequency of electrophysiological diagnoses in the pediatric

Table 2: Compatibility between clinical pre-diagnoses and electrophysiological diagnoses

	Compatible		Incompatible	
	N	%	N	%
MND	1	100%		
Radiculopathy	5	23.8%	Normal	14 (66.7%)
			Plexus	1 (4.8%)
			Entrapment neuropathy	1 (4.8%)
Plexopathy	20	74.1%	Normal	5 (18.5%)
			Peripheral nerve lesion	2 (7.4%)
Traumatic peripheral nerve lesion	42	63.6%	Normal	21 (31.8%)
			Radiculopathy	1 (1.5%)
			Entrapment neuropathy	2 (3%)
PNP	42	15.73%	225	(84.2%)
			Normal	213 (80.1%)
			Radiculopathy	2 (0.7%)
			Peripheral nerve lesion	2 (0.7%)
			Myopathy	5 (1.9%)
			Entrapment neuropathy	2 (0.7%)
Myopathy	9	(23.6%)	Normal	24 (60.52%)
			PNP	6 (15.78%)
Myopathy + neuropathy	Myopathy	1 (10%)	Normal	8 (80%)
	PNP	1 (10%)		
Facial palsy	13	(86.7%)	Normal	2 (13.3%)
Entrapment neuropathy of the upper extremity	6	18.8%	Normal	25 (78.1%)
			Peripheral nerve lesion	1 (3.1%)
Entrapment neuropathy of the lower extremity	12	66.8%	Normal	6 (27.8%)
			Radiculopathy	1 (5.6)
GBS	12	41.37%	Normal	17 (58.6%)

MND: Motor neuron disease, PNP: Purine nucleoside phosphorylase, GBS: Guillain-Barre syndrome

patient population was examined according to age groups, and the most common diagnosis was found to be plexopathy between the ages of 0-5, myopathy between the ages of 6-10 and PP between the ages of 11-16 (8).

In our study, we found that plexopathy was the most common between the ages of 0-2, and children between the ages of 2-6 and 7-16 years were most frequently examined with a pre-diagnosis of PP and peripheral nerve lesion. Although this situation is similar to other studies conducted in the pediatric patient group, it shows that pediatric patients apply to the ENMG laboratory with quite different pre-diagnoses from studies conducted with adults.

Looking at the distribution of ENMG numbers by age, it was observed that more EMNG examinations were performed at younger ages in two studies (5,8). More children between the ages of 7-16 years were studied in our laboratory. The reason for this difference can be attributed to the difference between hospitals in the age spectrum of children referred to our laboratory.

In a study conducted with adult patients, 49.1% of the ENMG results were found to be compatible with the pre-diagnosis (7). In another study examining the consistency between clinical and electrophysiological diagnoses in adult patients, the pre-diagnosis and result compatibility was found to be 53.6% (8).

In a study conducted on a pediatric patient population, this rate was found to be higher than in adults, as 60%; compatibility of more than 60% was found in disease groups such as brachial plexopathy, facial nerve damage, GBS, PP, and radiculopathy (5).

In a study by Hellmann et al. (6), evaluating the electrophysiological examination of 468 children, 98% of all diagnostic groups, 99.5% of neurogenic diseases, and 80% of myogenic diseases were found to be consistent with pre-diagnosis.

In our study, the pre-diagnosis/electrophysiological diagnosis ratio was found to be quite low (32.31%). Low compatibility with the diagnosis was found in the cases with pre-diagnoses of PNP, radiculopathy, myopathy, and entrapment neuropathy of the upper extremity (15.73%, 23.8%, 23.6%, 18.8% respectively); a high compliance rate was found for MND, plexopathy, peripheral nerve lesions and entrapment neuropathy of the lower extremity (100%, 74.1%, 63.6%, 66.8%).

Yagci et al. (5) had 20 patients with a prediagnosis of PNP and found that the consistency between the referral diagnosis and the post-ENMG diagnosis was 78.6%. Also, Komur et al. (7) had found of consistency 59 of 104 patients with prediagnosis of PNP.

In particular, we think that the low compliance rate of our PNP pre-diagnosis with our electrophysiological diagnosis was due to the referral of pediatric patients with diabetes to our laboratory for PNP screening at that time. We think that, before the referral of pediatric patients with diabetes for screening, questioning the neuropathic symptoms may prevent unnecessary ENMG examinations in these patients.

In our study, compliance with the pre-diagnosis/electrophysiological diagnosis was found to be lower in the patient group referred with the pre-diagnosis of GBS compared to the studies in the literature (5). ENMG may be as normal, especially in the first week of GBS. Since GBS is easy to diagnose clinically, we expected a higher compliance rate with pre-diagnosis and electrophysiological diagnosis. We think that one of the reason of our low compliance with the pre-diagnosis in patients with GBS pre-diagnosis, it may be that the ENMG examination was performed within first week.

Examination findings may be confused with each other in patients with myelitis and GBS. Clinicians sometimes refer to ENMG examinations when myelitis and GBS cannot be distinguished by clinical examination. We think that another reason for the low compliance rate in patients with a pre-diagnosis of GBS is that clinicians refer to the ENMG laboratory with a pre-diagnosis of GBS in patients whose clinical examination cannot distinguish between GBS or myelitis.

Another group in which we found a low compliance rate compared to the literature is patients with a pre-diagnosis of plexopathy. The number of patients in the 0-2 years of age is very low with only 28 patients; in this age group plexopathy is the most consistent diagnosis before and after ENMG as addressed in other studies. As this study has a low number of patients in this age group, the chance of consistency will be lower because plexopathy is very easy to diagnose on neurological examination.

In a retrospective study of Türkel et al. (11) with ENMG examinations in adult patients, it was found that the correlation between pre-diagnosis and electrophysiological diagnosis changes significantly according to the pre-diagnosis.

It was stated that the consistency between pre-diagnosis and electrophysiological diagnosis in pediatric patients did not differ by age groups and gender (5). In our study, although the consistency between pre-diagnosis and electrophysiological diagnosis did not differ between genders, it was found that there was a difference according to age groups and pre-diagnoses. In our study, the emergence of such a different result can be attributed to the fact that the electrophysiological detection of PNP in patients presenting with a pre-diagnosis of PNP was at a very low rate, the fact that a larger proportion of the patient group was evaluated with the pre-diagnosis of PNP and also the number of patients in the 7-16 age group was high.

Study Limitations

We think that the retrospective nature of our study may create some limitations that may affect the results, so we think that prospective studies are required, especially in the pediatric patient group. Another limitation is that when planning the study, patients with or without symptoms were not separated, and patients were included in the study whether or not they had symptoms. We think that this situation affects consistency between pre-diagnosis and electrophysiological diagnosis rates, especially in the PNP group. Our advice to researchers is to consider this issue in future studies.

Conclusion

As a result of our study with a high participation number, the spectrum of neuromuscular diseases in the pediatric patient group is quite different from that of adults. Electrophysiological diagnoses of children referred for ENMG examinations were largely incompatible with their pre-diagnoses. Considering this situation and the difficulty of the electrophysiological examinations mentioned above in pediatric patients, these patients can be evaluated more accurately with more accurate pre-diagnoses by performing detailed history and physical examination before the electrophysiological examination. Hereby, unnecessary investigations can be reduced and children who really need this evaluation can be diagnosed faster and their treatment can be started earlier.

Ethics

Ethics Committee Approval: Ethical approval was obtained from the Ethics Committee of University of Health Sciences Turkey, Dışkapı Yıldırım Beyazıt Training and Research Hospital (aproval date: 23.12.2019; approval number: 78/01).

Informed Consent: This study was a retrospective study.

Peer-reviewed: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: E.Ü., D.C., Z.K.Ü., N.T., Concept: E.Ü., D.C., Z.K.Ü., N.T., Design: E.Ü., D.C., Z.K.Ü., N.T., Data Collection or Processing: D.C., Z.K.Ü., Analysis or Interpretation: E.Ü., D.C., Z.K.Ü., N.T., Literature Search: D.C., Z.K.Ü., Writing: E.Ü., D.C., Z.K.Ü., N.T.

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