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Electromyography in Pediatric Population

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ABSTRACT

Introduction: Electrodiagnostic evaluation provides an important extension to the neurological examination for the evaluation of pediatric neuromuscular disease. Many pediatric neuromuscular diseases are analogous to those seen in the adult. However, the relative frequency of these illnesses varies greatly when different age populations are compared. The purpose of the present study is to provide a retrospective analysis of children referred to our electromyography (EMG) laboratory for electrophysiological examinations.

Methods: We retrospectively reviewed electrodiagnostic records of patients aged between 0–15 years, from January 2004 to June 2013. Patients were classified as having plexopathy, nerve root lesions, polyneuropathy, myopathy, mononeuropathy, anterior horn cell disease, neuromuscular transmission disorder, facial nerve palsy, and other rare disorders.

Results: We reviewed totally 5563 pediatric records, which was on the average 578 studies per year. It was about 14% of the all EMG examinations performed in our laboratory. When we looked at all the procedures, 3271 of the records included needle EMGs, 170 of them were single fiber EMGs, 100 of them were repetitive nerve stimulations, and 52 of them were evoked potentials. The results were normal in 55% of the cases. As a result of electrophysiological examinations, the common diagnoses were: plexopathy (28.6%), polyneuropathy (7.4%), and myopathy (6.6%) in patients aged 0–5 years (41.2% of all records); myopathy (9.4%), PNP (8.5%), mononeuropathy (6.4%), and plexopathy (5.9%) in 6–10 years (28.2% of all records); PNP (11.3%), myopathy (6.6%), and mononeuropathy (5.6%) in 11–15 years (30.6% of all records).

Conclusion: Infants and toddlers mostly suffered from brachial plexopathy which can be prevented by proper obstetrical management. Nerve conduction studies and EMG yielded diagnostic importance for demyelinating neuropathy and myopathy in patients older than 6 years of age.

Keywords: EMG, pediatric, neuromuscular, electrophysiology, differential diagnosis

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INTRODUCTION

Electrophysiological investigations are a follow-up to anamnesis and neurologic examination, and the scope of investigation is structured based on the patient's complaints and examination findings. While electrophysiological examination can be performed for each group, there are certain differences, particularly in pediatric electrophysiological investigations (1).

Mostly, electromyography laboratories (EMG) have been predominantly designed for the assessment of adult patients. If your laboratory is not suitable for assessment of pediatric patients or you are not experienced in this age group, you may not wish to assess the pediatric patient group, or may not comment on the results of your investigation. In fact, we can perform many investigation techniques that we use for adults, including nerve conduction studies, needle EMG, consecutive stimulation test, evoked potentials investigations, and even the single-fiber EMG, also in the pediatric patient group. In addition to technical difficulties, the fact that the normal values by age group are not known for all investigation methods also causes trouble in commenting of the results (2). Another point to consider is the difference in the incidence of these disorders by age group as well as the similarity of many pediatric neuromuscular disorders to those observed in adults.

Studies assessing pediatric patients electrophysiologically have either focused on a specific diagnosis and evaluated the clinical EMG compliance, or reviewed the compliance and consistency between the preliminary diagnosis referred to an EMG laboratory and the

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electrophysiological diagnosis (1, 3, 4) while this trial was designed to retrospectively document electrophysiological investigations performed in pediatric patients, referred to our laboratory with various preliminary diagnoses, taking into consideration only the EMG recordings, and asses their characteristics by age group. The study basically investigated the diagnoses, established as a result of electrophysiological investigation and how these diagnoses are distributed by age groups.

METHODS

Electrodiagnostic records of patients within the age range of 0–15 years, who had been referred to our electromyography laboratory (EMG) between January 2004 and June 2013 with various preliminary diagnoses from different departments and centers, were retrospectively assessed. The patients were classified to have the following disorders: plexopathy, nerve root lesions, polyneuropathy, myopathy, neuropathy, anterior horn disease, neuromuscular junction transmission disorder, facial paralysis, and other rare diseases. This classification was based on the results obtained from the patients' investigations and the EMG recordings, available in our system. In addition, the electrophysiological methods used for the investigations were also noted.

RESULTS

We screened the electronic archive of our Electromyography Laboratory of the Clinical Neurophysiology Department and evaluated the investigation results obtained between January 2004 and June 2013 (Table 1). We accessed the records of 5563 patients in total, who were investigated during that period. Considering this number, we calculated that 578 children had been investigated electrophysiologically on the average annually. Since the annual number of electrophysiological investigations was calculated to be 4500 on average, we observed that 14% of these were from pediatric patients. Of the 5563 patients investigated, 2987 were males and 2576 were females. Assessing according to the investigation method applied, all patients were detected to undergo nerve conduction studies: 3271 patients underwent needle EMG, 170 patients underwent single-fiber EMG, a method specific for investigating neuromuscular junction disorders. Consecutive stimulation tests had been performed in 100 patients to investigate neuromuscular junction diseases. We noticed that evoked potential investigations had been performed in 52 patients. We also observed that most of the patients underwent multiple investigation techniques so as to establish the accurate diagnosis.

Assessing the total patient population, in whom all electrophysiological methods were performed individually or as a combination, we detected that 55% of the patients had normal results.

Even if all the patients were in the pediatric age group, both the investigation plan and the resulting diagnoses differed between the subgroups. Classifying patients as being included in the 0–5 age group, 6–10 age group ad 11–15 age group, the primary diagnoses and the relevant ratios were as follows (Table 2):

Patients in the 0–5 age group represented 41.2% of all investigations, and the most common three diagnoses obtained in this group were as follows: 28.6% plexopathy, 7.4% polyneuropathy (PNP) and 6.6% myopathy.

As for the patients in the 6–10 age group, they represented 28.2% of all patients. The most common three electrophysiological diagnoses were as follows: 9.4% myopathy, 8.5% PNP, 6.4% mononeuropathy.

As for the patients in the 11–15 age group, they represented 30.6% of all patients. The most common three electrophysiological diagnoses were as follows: 11.3% myopathy, 6.6% myopathy, 5.6% mononeuropathy.

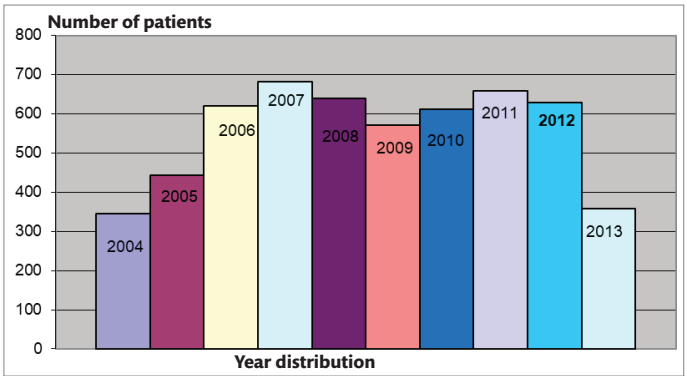

Figure 1. Distribution of the investigations by year

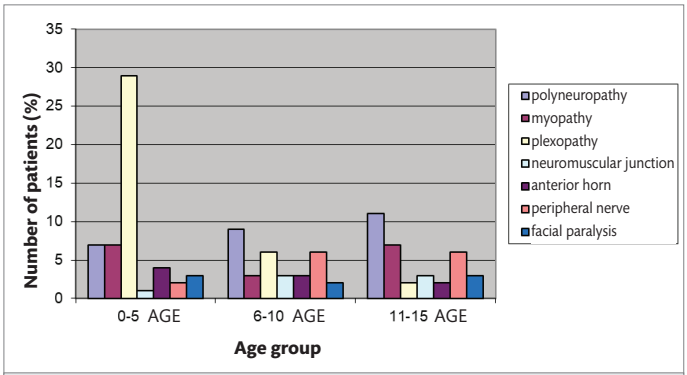

Figure 2. Distribution of the pathological diagnoses by age group

DISCUSSION

While electrophysiological investigations require more patience and caution in pediatric patients, they can also be a guide for the clinicians in relation to this age group. Electrophysiological investigations are helpful not only in establishing the diagnosis but also in monitoring the side effects and efficacy of treatment, and understanding and monitoring the progression process (5).

Inability of the patients to cooperate during investigation may occasionally challenge the investigating physician technically. The principle of repeatability for ensuring data reliability cannot be fulfilled for certain patients. Both technical challenges, and the mental status of the patient and his/her family cause a failure to perform the electrophysiological assessment in an optimum way (6). Along with the technical challenges, we can mention certain clues and facilitating factors for accurately assessing the numeric data obtained.

Some of these include earning the trust of the child and the family, making explanation on the investigation using a language that is easy for them to understand, if possible, inviting the family members to the site of investigation during the assessment and using mother-child cooperation while choosing and practicing the investigation method (7). What probably requires the utmost experience is to know which investigation should be performed in which patients and decide where to stop. These are actually applicable for all EMG investigations and require more attention in the pediatric patients.

At our laboratory, up to 4500–5000 electrophysiological investigations are being performed annually, and one seventh of these investigations are performed in the pediatric age group. The number of patients, we obtain by screening a period of approximately 10 years, appears to be

a quite good value for pediatric population assessments. This study also demonstrates our experience in pediatric patients, considering the numeric data from our laboratory, and the characteristics of the investigations performed.

The most remarkable point to consider during assessment of the results was the fact that more than half of the patients (55%) had normal electrophysiological investigation results. These normal results may have multiple causes. In an attempt to list these causes, we can first mention the presence of a disease that is not associated with the patient's peripheral nervous system (such as central causes-central hypotonia); secondly, we can consider the fact that the disease may not have yet exhibited electrophysiologically (such as early-stage polyneuropathy); and as a third cause, we can consider the fact that the investigation method may have remained inadequate for the investigated disease (fe.g. there is a suspicion of myopathy, however no needle EMG was performed in the patient); fourth, the clinician may have wanted to conduct a situation determination; and fifth, EMG indication may be unnecessary for that patient. Taking into consideration that it is a demanding investigation both for adults and children, determination of the accurate indications, and the questions asked to the investigating team are very important. The question of the clinician would also determine the extent to which the electrophysiological investigation should be extended.

Another interesting point in relation to our data which revealed that more than half were normal electrophysiological assessment results was the difference in diagnoses, observed upon classification of the pediatric patients by age group. The group of patients, included in the 0–5 age range (the smallest children) represented the biggest group among (41.2%) all patients. The most common electrophysiological diagnosis in this group was plexopathy, as indicated also in the literature. Unfortunately, congenital brachial plexus damages are not only observed in our country but also worldwide; while there are different data on its incidence, it is considered to be 1–2 cases per 1000 live births (8). While cesarean section decreases the risk of brachial plexopathy, it may occur also in babies born with this method (9). In addition, maternal diabetes, multiparity, fetal macrosomia, presence of obstetric brachial plexus stroke in the previous child, intrauterine torticollis, breech presentation, assisted delivery, extended delivery period may be included in the other intranatal risk factors (8). While the timing of EMG for these babies remains to be controversial in the literature, a majority of these babies are observed to recover within the first 3 months using conservative methods. EMG is a helpful method to determine the babies with an approximate sequel of 20–25%, who could or could not improve. We can recommend making decision in relation to EMG by following the first 3-month period (5). At our laboratory, we prefer to perform EMG usually for babies with a preliminary diagnosis of plexopathy when they are 3 month-old.

The patients in the 6–10 age range (the second group) are those, in whom electrophysiological investigation is performed under the most difficult conditions. Explaining the investigation to the children in this age group using a language they can understand, and ensuring cooperation is of vital importance; otherwise, it is possible that you may not be able to perform the investigation. Myopathy is the most common electrophysiological diagnosis in this patient group. In patients presenting to the physician with symptoms such as difficulty of walking emerging explicitly at this age, remaining underdeveloped in physical activities relative to friends, the preliminary diagnosis of myopathy is highly consistent with electrophysiological diagnosis (1). However, particularly in the small age group and in case of congenital myopathies, myopathy may occasionally not be identified by electrophysiological findings. Electrophysiologically, this false negativity may have multiple causes. Myopathy may not be diagnosed particularly in small children (smaller than 1-year-old) due to technical reasons, lack of experience, patch distribution of the pathology over the muscle fibers, and findings that appear to be neurogenic (5). One should also note that electrophysiological assessment is a follow-up to anamnesis and physical examination. In case of strong clinical suspicion, other investigation methods should be used to establish diagnosis.

While polyneuropathies are observed at all ages, it ranked first among our patients in the 11–15 age group. Electrophysiological investigation is very valuable and helpful for diagnosis in patients with suspected "Guillain Barre Syndrome", potentially established through albuminocytological disassociation detected in the cerebrospinal fluid (CSF), who present with complaints such as acute-onset weakness, numbness extending upward from the legs, and who fail to show deep tendon reflexes at neurological examination (10). In patients, who are detected through atypical findings, nerve conduction studies may be informative starting from the first day. EMG does not only give information on the involvement pattern (sensory, motor or both) but it may also elucidate whether PNP is acquired or hereditary. Thus, it guides the clinician in his/her further investigations and treatment plan (5, 11).

Mononeuropathies included in our second and third group are actually among the diagnoses, which are not commonly observed in childhood. However, grouping effects on the nerves of the upper and lower extremities (ulnar, median, radial, peroneal, tibial) mostly from trauma, entrapment neuropathies accompanying certain metabolic disorders, and facial paresis in this group; we observed that it was the third most common electrophysiological diagnosis for our patient group, in children between 5 and 15 years of age.

Assessing by age group, "hypotonic baby" represents a significant patient population, which requires differential diagnosis electrophysiologically. The reason why it is not included in the first 3 in our data may be the fact that hypotonia occurs most commonly due to central causes. Since electrophysiological investigations would yield normal results in hypotonicity associated with central causes, it is possible that a portion of these patients are included in the 55% normal EMG investigations. On the other hand, a portion of patients with a preliminary diagnosis of "hypotonic baby" may have been classified as neurogenic or myogenic involvement. In addition, while EMG still has an important place in the diagnosis of spinal muscular atrophy that is also significant for hypotonic baby etiology, accurate and rapid diagnosis can be made by genetic investigation. Since we performed our trial retrospectively by considering the electronic report records, we cannot give an opinion on the consistence between the diagnoses that result in EMG requests. Reviewing the possibilities, we can only mention these probabilities.

In conclusion, this study we performed by taking into consideration the electrodiagnostic records irrespective of what has been requested showed that certain diagnoses could come to the forefront, based on age in the pediatric age group. Also, this study is significant in that it includes the largest number of patients for which electrophysiological investigations were assessed in the pediatric population.

This study was presented as a poster report at the 30th International Clinical Neurophysiology Congress (ICNN) held in Berlin (Germany) between 20 and 23 March, 2014. Poster no: 420

Ethics Committee Approval: Data in this study are written according to the Declaration of Helsinki. Ethical committee approval was not obtained because our study was a retrospective evaluation with an electronic database.

Informed Consent: Before the review was made, parents were informed that the data could be used for scientific purposes.

Peer-review: Externally peer-reviewed.

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Conflict of Interest: No conflict of interest was declared by the authors.

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