ORIGINAL ARTICLE

Consistency between referral diagnosis and post-ENMG diagnosis in children

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Abstract

Objective: To evaluate the degree of consistency between the referral diagnosis and that based on electroneuromyography.

Methods: The retrospective study was conducted at the Paediatric Neurology Laboratory of Mersin University School of Medicine, Turkey, and comprised all electroneuromyographies carried out between January 2005 and December 2010. Demographic data, referral diagnosis and post-procedure diagnosis were recorded for each patient, and were classified into groups. Consistency between the two groups was compared using SPSS 13.

Results: Of the total 294 patients, polyneuropathy was the reason for referral in 104 (35.4%), peripheral nerve injury in 54 (18.4%), brachial plexus injury in 52 (17.7%), myopathy in 52 (17.7%), hypotonia in 23 (7.8%), and facial paralysis in 9 (3.0%) patients. There was consistency between the two diagnoses in 179 (60.9%) patients.

Conclusion: Electroneuromyography is an uneasy, painful and stressfull procedure for children, and, therefore, it should be recommended only in cases where the result may be beneficial in the diagnosis, treatment and follow-up of a patient.

Keywords: Electroneuromyography, Children, Neuromuscular diseases, Diagnostic value. (JPMA 64: 179; 2014)

Introduction

Electroneuromyography (ENMG) is an electrophysiological method of examination for neurophysiological state of motor neuron, peripheral nerve and muscle functions. ENMG is used for diagnosis, treatment and monitoring of diseases with an impact on muscles, neuromuscular junctions, nerves and anterior horn motor neurons as well as determining their prognosis.[1,2]

ENMG still maintains its importance despite advanced diagnostic tools. While it is often used for the evaluation of neuromuscular diseases in adults, many technical difficulties restrict its use in children. The diagnostic value of this examination in children depends highly on the experience of the examiner and the tolerance level of patients. Furthermore, unlike other laboratory procedures, ENMG examination should not always be performed in a standard manner. It should rather be based on patient's history and preliminary diagnosis of the clinician.[3]

The present study was planned to evaluate the degree of consistency between the referral diagnosis and the post-ENMG diagnosis, the diagnostic value of ENMG and to determine the rate of unnecessary ENMG examination in patients referred to the ENMG laboratory concerned.

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Patients and Methods

The retrospective study was conducted at the Mersin University School of Medicine, Turkey, and comprised all procedures carried out at the Paediatric Neurology ENMG Laboratory between January 2005 and December 2010. Demographic data, techniques used during ENMG, referral diagnosis and post-ENMG diagnosis were all recorded for each patient. The referral diagnoses and post-ENMG diagnoses were classified into groups. The consistency between referring diagnosis and post-ENMG diagnosis was compared.

For the purpose of comparison, 'consistency' was described as a similarity between the referral diagnosis and the post-ENMG diagnosis. 'Inconsistency' was described as the difference between the referral diagnosis and the post-ENMG diagnosis or a normal ENMG result.

All laboratory procedures had been carried out using the EBNeuro Myto II instrument. The procedure was explained to the child, if he/she was capable of understanding, and to his/her parents. It was made sure that a parent was present in the room during the procedure. Motor and sensory nerve conduction studies were performed using electrodes appropriate for the patient's age.[4] If it was a needle ENMG, it was performed by the same experienced researcher with a concentric needle electrode appropriate for the patient's age. We followed the guidelines recommended on the basis of clinical findings and preliminary diagnosis for selection of nerve conduction studies (NCS) and the needle to be used.[1,2,5] All results

were interpreted as compared to the normal data according to the age groups.[5-7] ENMG reports included referral diagnosis, date of ENMG procedure, and findings as well as demographic data. The conclusion section included abnormal results, interpretation of the examiner, and technical difficulties and suggestions, if any.

The statistical demographic data and post-ENMG diagnoses were analysed using SPSS 13.0 programme.

Results

Of the total 294 patients, 180 (61.2%) were male, and 114 (38.8%) were female with an age range from 1 month to 17 years (mean: 7.41±4.99 years). Of them, 228 were referred from the Paediatric Neurology Polyclinic, and 66 from other polyclinics (Orthopedics, Physical Therapy and Rehabilitation, and Paediatrics).

Table-1: Consistency between the referral diagnoses and the post-ENMG diagnoses.

	Consistent (n)	Consistent (%)	Inconsistent (n)	Inconsistent (%)	Sex Male / female	Age (mean year)	Total (n)
Polyneuropathy	59	56.7	45	43.3	51/53	9.3	104
Peripheral nerve injury	40	74.0	14	26.0	43/11	9.7	54
Brachial plexopathy	46	88.4	6	11.6	34/18	3.6	52
Myopathy	14	26.9	38	73.1	35/17	7.7	52
Hypotonic infant	11	47.8	12	52.2	14/9	1.1	23
Facial paralysis	9	100	0	0	3/6	7.0	9
Total	179	60.9	115	39.1	180/114	7.35	294

Based on the referral diagnoses, ENMG was carried out for polyneuropathy in 104 (35.4%) patients, peripheral nerve injury in 54 (18.4%), brachial plexus injury in 52 (17.7%), myopathy in 52 (17.7%), hypotonia in 23 (7.8%) and facial paralysis in 9 (3.0%) patients (Table-1).

Polyneuropatic Diseases

Of the 104 with ENMG with the referral diagnosis polyneuropathy, ENMG findings were consistent in 59 (56.7%) patients. Based on the involved nerve fibre the ENMG results showed, type of lesions were demyelinating in 14 (23.7%) patients, axonal in 22 (37.3%), and both demyelinating and axonal in 23 (39.0%). Based on the nerve function, there was a damage to the motor nerves in 24 (40.7%) patients, to the sensory nerves in 3 (5.1%), and to both the motor and sensory nerves in 32 (54.2%).

Of the 54 patients with peripheral nerve injury (N. ulnaris, N. medianus, N. radialis, N.sciaticus and N.peronealis), 40 (74%) showed consistent results. Among them 22 (55.0%) out of 40 patients had upper limb peripheral nerve injury. Of these patients, 10 (45.5%) had a peripheral nerve injury of N.Ulnaris, 7 (31.8%) of N.Medianus (three associated with carpal tunnel syndrome), and 5 (22.7%) of both N.Ulnaris and N.Medianus. None of them had an injury of N.Radialis. Twelve out of 18 (45.0%) patients with a lower limb peripheral nerve injury had an injury of N.Sciaticus and 6 had an injury of N.Peronealis. None of the patients had an injury of N.tibialis.

Of the 52 patients with a referral diagnosis of brachial plexus injury, ENMG-based diagnosis was consistent in 46 (88.5%). Among these 46 patients, the lesion was in the left arm in 11 (23.9%), and in the right arm in 35 (76.1%). Further examinations in 6 (11.5%) patients with inconsistent results, demonstrated hemiplegic cerebral palsy.

ENMG results were consistent with myopathy in 14 of 52 (26.9%) patients. Among 23 patients with hypotonia, ENMG consistency was found in 11 (47.8%) patients: spinal muscular atrophy in 6 (26.1%), myopathic alterations in 2, polyneuropathic findings in 3 (13.0%), and normal findings in 12 (52.2%) patients.

There was 100 per cent consistency of ENMG results for all the 9 patients with a referral diagnosis of peripheral facial paralysis.

ENMG results of 109 (37.1%) patients who were required to undergo ENMG with different referral diagnoses were within normal range for age. Evaluation of the referral diagnosis for those patients whose ENMG results were considered within normal range showed that the reason for referral was polyneuropathy in 44 (40.4%) patients, myopathy in 36 (33.0%) patients, hypotonia in 12 (11.0%) patients, peripheral nerve injury in 11 (10.1%) patients and brachial plexus injury in 6 (5.5%) patients.

Overall, referral diagnosis and post-ENMG diagnosis were consistent in 179 (60.9%) patients.

Discussion

Although there are numerous studies that have evaluated the contribution of ENMG in adults, but such studies are

rare in children.[8-10] The number of studies that report on consistency rate between the referral diagnosis and the post-ENMG diagnosis is very little.[9-12] Therefore, the present study aimed at evaluating the consistency rate between the referral diagnosis and the post-ENMG diagnosis in children, who were required to undergo ENMG with a referral diagnosis of neuromuscular diseases.

We found a consistency of 60.9% between the referral diagnoses and the post-ENMG diagnoses. This rate varies between 39% and 55% in literature.[8-12] The consistency was over 50% in patients who were required to undergo ENMG with a referral diagnosis of polyneuropathy, brachial plexus injury, peripheral facial paralysis, peripheral nerve injury, while it was less then 50% in patients who were required to undergo ENMG with a referral diagnosis of hypotonia and myopathy.

A study by Yagci et al. had 20 patients with a referral diagnosis of polyneuropathy. Of them, 4 patients received the diagnosis of Guillain Barre Syndrome (GBS), 11 patients polyneuropathy, and 2 patients of radiculopathy. The consistency between the referral diagnosis and the post-ENMG diagnosis was 78.6%.[11] Hellmann et al. retrospectively evaluated the ENMG results of 195 patients with a final diagnosis of neuropathy and with a consistency of 99.5% between referral diagnosis and post-ENMG diagnosis.[12] However, this study evaluated the consistency between the post-ENMG diagnosis and the final clinical diagnosis. The present study analysed the consistency between the referral diagnosis and the post-ENMG diagnosis, and results showed that the results for 59 of 104 patients who were required to undergo ENMG with a referral diagnosis of polyneuropathy were consistent with polyneuropathy. We had a consistency rate of 56.7% between the referral diagnosis and the post-ENMG diagnosis. Although in literature, ENMG has a high reliability of diagnosis in patients with polyneuropathy, but our consistency rate between the referral diagnosis and the post-ENMG diagnosis was lower compared to the literature. This was attributed to the fact that infants with a gait disorder who were required to undergo ENMG for screening of polyneuropathy were included in the patient group as it was not possible to diagnose polyneuropathy simply based on the physical examination during infancy.

Electrophysiological studies are required to determine the exact localisation of lesions and to obtain information on the prognosis in patients with peripheral nerve injury. In a study by Kouyoumdjian, 456 adult patients with peripheral nerve injury were retrospectively evaluated. Most of the patients had upper limb peripheral nerve injury (73.5%). The most common injuries of upper limb peripheral nerves were of N. Ulnaris and N. Medianus, while the N. Peronealis was the most injured nerve in the lower limb.[13] Among 85 patients with peripheral nerve injury, Uzun et al. found that 48 patients had upper limb injury (56.5%), 36 had lower limb injury (42.3%) and one patient (1.2%) had both upper and lower limb nerve injury. The most common injury of the upper limb was observed in the ulnar nerve in 17 patients (35.4%), while the sciatic nerve was the most injured in the lower limb in 12 patients (66.6%).[14] In the present study, ENMG results of 40 of 54 patients who were required to undergo ENMG with a referral diagnosis of peripheral nerve injury were consistent. The consistency between the referral diagnosis and the post-ENMG diagnosis was 74.0%. Among these 40 patients, 22 (55%) had upper limb peripheral nerve injury. The most common injury of upper limb peripheral nerve was observed in N. Ulnaris in 10 of 21 patients (47.6%). In the Kouyoumdjian study,[13] the most often injured nerve of the lower limb was N. Peronealis, while it was N. Sciaticus in 12 patients (66.6%) in our study. It was found from the history of patients that the sciatic nerve injury was associated with injection in 9 of these 12 patients. In the present study, the upper limb injuries were more common than lower limb injuries which was in accordance with the literature.[13,14]

Electrophysiological studies are required to determine the localisation of lesions (lower, middle and upper truncus) and the presence of regeneration in the area of the lesion and the treatment plan in patients with brachial plexopathy. Yagci et al. showed that the results for 22 out of 26 patients who underwent ENMG with the referral diagnosis of brachial plexopathy were consistent with brachial plexopathy. The consistency between (84.6%).[11] Vredeveld et al. evaluated the reliability of electrophysiological testing in 184 patients with brachial plexopathy, and showed a reliability of 84% in electrophysiological testing for patients with brachial plexopathy.[15] In the present study, ENMG results of 46 of 52 patients who were required to undergo ENMG with a referral diagnosis of brachial plexopathy were consistent (88.5%). This rate was higher compared to the overall rate between the referral diagnosis and the post-ENMG diagnosis in our study. Further examinations showed that 6 patients who had inconsistency between the referral diagnosis and the post-ENMG diagnosis had hemiplegic cerebral palsy. Therefore, hemiplegic cerebral palsy should be excluded by a more comprehensive nerulogical examination prior to ENMG in infants with suspected brachial plexopathy.

The diagnostic reliability of ENMG is low in the myopathic

children. Even tough ENMG is considered normal, myopathy cannot be excluded.[16] Yagci et al. found myogenic alterations in 10 of the 24 patients who were required to undergo ENMG with a referral diagnosis of myopathy, and a consistency rate of 41.7%.[11] Hellmann et al. observed myogenic alterations in ENMG in 39 of 49 patients who were diagnosed as having muscular disease. However, this study included the patients with a final diagnosis of muscular disease; yet, ENMG showed myogenic alterations in 80% of patients.[12] In a similar study, Rabie et al. detected myogenic alterations in ENMG of seven of 15 patients (46.6%) who were diagnosed as having myopathic disease on the basis of muscle biopsy.[17] The present study showed myogenic alterations in 14 of 52 such patients. The consistency between the referral diagnosis and the post-ENMG diagnosis in myopathic children varies between 10% and 55% in literature.[16,17] The consistency in our study was 26.9%, which is in line with literature. The diagnostic reliability of ENMG is low in myopathic children. The result of a needle ENMG examination can be normal, myogenic or neurogenic in these patients.[16] Myopathy cannot be excluded even though the ENMG result is normal. Therefore, patients with suspected myopathy and normal ENMG results should be thoroughly evaluated with further examinations (such as muscle biopsy, gene analysis). On the other hand, in our clinic the patient with evident myopathy, such as Duchenne muscular dystrophy, has not been referred to ENMG laboratory. So the diagnostic reliability of ENMG is low in patients with suspected myopathy.

Electrophysiological examination is important in the diagnosis of hypotonic infants. However, hypotonia may occur due to many reasons during infancy. ENGM is used for the evaluation/screening of the motor unit in diagnosing hypotonic infants. Therefore, use of ENMG for evaluation/screening purposes provides beneficial information in all hypotonic cases with an unknown etiology.[18] Hellmann et al. who evaluated the ENMG results of 51 hypotonic infants, showed that the results were normal in 31 (61%) cases, and 14 (27%) had neurogenic involvement, and 6 (12%) had myogenic involvement. Consequently, the concordance between the referral diagnosis and the post-ENMG diagnosis was 39%.[12] Similarly, in an ENMG examination of 37 hypotonic infants by Cetin et al. the results of 16 (43.3%) were normal, whereas 5 (13.4%) had myogenic alterations, and 16 (43.3%) had neurogenic alterations. The concordance between the clinical diagnosis and the post-ENMG diagnosis was 56.7%. In this study, the consistency rate between the referral diagnosis and the post-ENMG diagnosis was higher as 7 patients had SMA.[19] In the present study, evaluations on ENMG reports for 23 patients who were required to undergo ENMG with a referral diagnosis of hypotonia showed that the results of 12 patients (52.2%) were normal, 8 (40%) had neurogenic alterations, and 2 (10%) had myogenic alterations. The concordance between the clinical diagnosis and the post-ENMG diagnosis was 47.8%. It was lower than our overall consistency rate between the referral diagnosis and the post-ENMG diagnosis. However, it was higher than that found in literature. It was attributed to the presence of SMA in 6 of our patients.

Electrophysiological examination is a difficult procedure in children with facial paralysis. However, ENMG is an important diagnostic tool in determining whether the paralysis is peripheral or central, and in providing information on prognosis. Sittel and Stenmert, evaluating the prognostic value of ENMG, found a consistency of 92.4% between ENMG and prognosis in 355 patients with facial paralysis.[20] In the present study, the results of all of the nine children who were required to undergo ENMG with the referral diagnosis of facial paralysis were consistent. However, we didn't evaluate the prognostic value in patients with peripheral facial paralysis.

We found that the consistency rate between the referral diagnosis and the diagnosis after electrophysiological examination was higher than that of other studies in literature. We believe that the higher rate was associated with the fact that all patients who would undergo electrophysiological examination were evaluated by a specialist experienced in ENMG prior to ENMG as well as comprehensive history-taking and neurological examination. This study's strength is that there are a few studies that have attemped to assess the consistency between referring diagnosis and post-ENMG diagnosis in children, and all ENMG were performed by a only one experienced physician (C.O). Hovewer, our study had several limitations. First, it is a retrospective chart review, thus patients were excluded for data could not be reached exactly. Also, it would be more usefull to give data related to control ENMG findings of these patients. Additionally, we based accuracy on the final diagnosis that is itself a subject of some uncertainty even when based on a reasonably long follow-up period.

Conclusion

ENMG is an uneasy, painful and stressfull procedure for children. Therefore, it shall be recommended in cases where ENMG result may be beneficial in the diagnosis, treatment and follow-up of a patient. In accordance with the literature, the consistency between the referral diagnosis and the post-ENMG diagnosis was over 50% for

facial paralysis, brachial plexopathy, peripheral nerve injury and polyneuropathy, while it was less than 50% for myopathy and hypotonia.

Disclosure

The present study was presented at the "12. Ulusal Cocuk Nöroloji Kongresi" 26-29 May 2010 Konya Turkey.

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