Global Academic Journal of Medical Sciences

Available online at www.gajrc.com
DOI: 10.36348/gajms.2022.v04i02.007

ISSN: 2706-9036 (P)
ISSN: 2707-2533 (0)

Original Research Article

Electromyogram (EMG) and Nerve Conduction Study (NCS) Findings in Children with Neuromuscular Disorders

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Article History
Received: 24.02.2022
Accepted: 31.03.2022
Published: 12.04.2022

Abstract: ***Background:*** An electromyogram (EMG) measures the electrical activity of muscles when they're at rest and when they're being used. On the other hand, nerve conduction study (NCS) is a medical diagnostic test used to evaluate the function, especially the ability of electrical conduction, of the motor and sensory nerves of the human body. Both are used to diagnose the status of neuromuscular disorders even in children. ***Aim of the study:*** The aim of this study was to assess the findings of electromyogram and nerve conduction study in children with neuromuscular disorders. ***Methods:*** This retrospective, observational study was conducted in the department of Paediatric Neuroscience, Dhaka Shishu Hospital & BICH, Dhaka, Bangladesh during the period from January 2010 to December 2016. In total 65 suspected children with neuromuscular disorders were selected as the study subjects. After elaborating the history taking and physical examination a provisional diagnosis was made and sent for NCS and EMG to trained personnel at Bangladesh Protibondhi Foundation along with other necessary investigations. All data were recorded and the "clinical and etiological" profile was correlated with the neurophysiologic findings retrospectively. ***Results:*** As investigation reports of EMG, among all participants, we found normal findings in 21 participants. Besides these, a variety of SMA and/or late-onset SMA were found in 16 participants. Moreover, diffuse fibrillation, biphasic complex, repetitive spontaneous discharge was found among 6 participants. On the other hand, as per the investigation reports of NCS among all the participants, the normal finding was in 34 participants. Besides this low amplitude CMAP and SNAP, low amplitude CMAP, variable sensory conduction velocity, and slow conduction velocity with temporal dispersion were found in 3, 6, another 3, and 4 participants respectively. The compatibilities of electrophysiological diagnosis were found at 68% for detecting neurogenic disease and 71% for detecting myogenic disease. On the other hand, the compatibilities of the Nerve Conduction Study were found 46% for detecting neurogenic disease and 50% for detecting myogenic disease. ***Conclusion:*** NCS and EMG are crucial investigations to reach confirmation as well as the exclusion of the diagnosis of neuromuscular disorder in children.

Keywords: Electromyogram, EMG, Nerve conduction study, NCS, Neuromuscular disorders.

Citation: Shaoli Sarker & A.Z.M. Azam, Humaira Rafiqa Quaderi (2022). Electromyogram (EMG) and Nerve Conduction Study (NCS) Findings in Children with Neuromuscular Disorders. *Glob Acad J Med Sci*; Vol-4, Iss-2 pp- 78-82.

1. INTRODUCTION

An electromyogram (EMG) assesses the electrical activity of muscles when they're at rest and when they're being used. On the other hand, nerve conduction study (NCS) is a medical diagnostic test used to evaluate the function, especially the ability of electrical conduction, of the motor and sensory nerves of the human body. In a study for discussion of the testing techniques and parts of the "peripheral nervous system" they test, the reader is referred to general references in clinical neurophysiology [1, 2]. The gold standard in diagnosing several acquired "neuromuscular disorders" is the nerve or muscle biopsy [3]. Needle EMG is such a technic. Basically, needle EMG (Electromyogram) may be considered an ''electrophysiologic biopsy'' of the muscle [4]. On the other hand, NCS (Nerve conduction study) is another useful tool in detecting patients who have pain, tingling, numbness, and hypo- or areflexic weakness [5]. Sensory NCS (Nerve conduction study) represents evoked sensory nerve action potentials (SNAPs) of the fastest conducting sensory nerve fibers [6]. They assess the integrity of this population of sensory fibers from the dorsal root ganglion to the periphery [7]. Common pitfalls in recording sensory nerve action potentials [6] are suboptimal stimulating or recording electrode position relative to the nerve being studied. The measurement of a motor nerve's response, or M wave, is achieved by recording a compound muscle action potential (CMAP) from a surface electrode at the end-plate region of a muscle [8]. Partial conduction block (PCB) manifests if a significant number of axons have blocked conduction to resulting in weakness and/or sensory losses [9]. The concept of temporal dispersion mainly is used in the same settings of a demyelinating lesion [10] and disseminates slowing of the conduction along a segment without producing clinical abnormalities [11].

2. METHODOLOGY AND MATERIALS

This retrospective, observational study was conducted in the department of Paediatric Neuroscience, Dhaka Shishu Hospital and BICH, Dhaka, Bangladesh during the period from January 2010 to December 2016. In total 65 suspected children with neuromuscular disorders were selected as the study population. Ethical approval had been taken from the ethical committee of the mentioned hospital. Properly written consent was taken from all the participants before data collection. After elaborating the history taking and physical examination a provisional diagnosis was made and sent for NCS and EMG to trained personnel at Bangladesh Protibondhi Foundation along with other necessary investigations. In 5-10% of patients, deletion of both SMN1 and SMN2 genes was not detected. In such cases, point mutations have been identified. Nevertheless, detection of point mutation requires specialized techniques followed by DNA sequencing. Data regarding socio-demographic status, mode of presentations, clinical diagnosis, investigation reports of 'EMG and NCS' and 'compatibility between clinical and electrophysiological profile' of participants were recorded. Finally, the "clinical and etiological" profile was correlated with the neurophysiologic findings retrospectively.

3. RESULTS

In this study, among a total of 65 participants, the male-female ratio was 1.6:1. The mean (±SD) age of the participants was 3.75 ± 3.1 years. Most (80%) of the fathers of the participants were illiterate. In analyzing the mode of presentations, we observed that 68%, 56%, and 54% of patients were with hypotonia, motor skill regression, and motor developmental delay respectively. As clinical diagnosis SMA, muscular dystrophy, and myopathy were found among 35%, 25%, and 19% of patients respectively. As investigation reports of EMG, among all participants, we found normal findings in 16. Besides these, a variety of SMA and/or late-onset SMA were found in 16 patients. Moreover, diffuse fibrillation, biphasic complex, repetitive spontaneous discharge was found among 6 patients. On the other hand, as per the investigation reports of NCS among all the participants, the normal finding was in 15 patients. Besides this low amplitude CMAP and SNAP, low amplitude CMAP, variable sensory conduction velocity, and slow conduction velocity with temporal dispersion were found in 3, 6, another 3, and 4 patients respectively. The compatibilities of electrophysiological diagnosis were found at 68% for detecting neurogenic disease and 71% for detecting myogenic disease.

Table-1: Socio-demographic status of participants (N=65)

Basic demographic status	
Male-Female ratio	1.6 :1
Mean (±SD) age (in years)	3.75 ± 3.1
Rural-Urban ratio	1.8:1
Father's Education	
Literate	20%
Illiterate	80%
Mother's Education	
Literate	26%
Non literate	74%
Income group	
low	30%
Low middle & middle	43%
High	27%
Blood Status	
H/O Consanguinity	26%
H/O PNA	12%

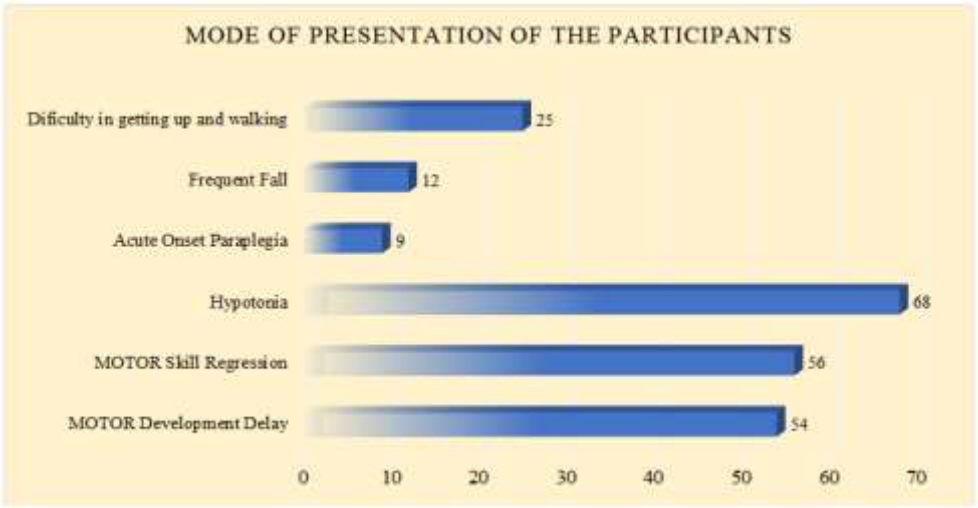

Fig-1: Mode of presentation of participants in % (N=65)

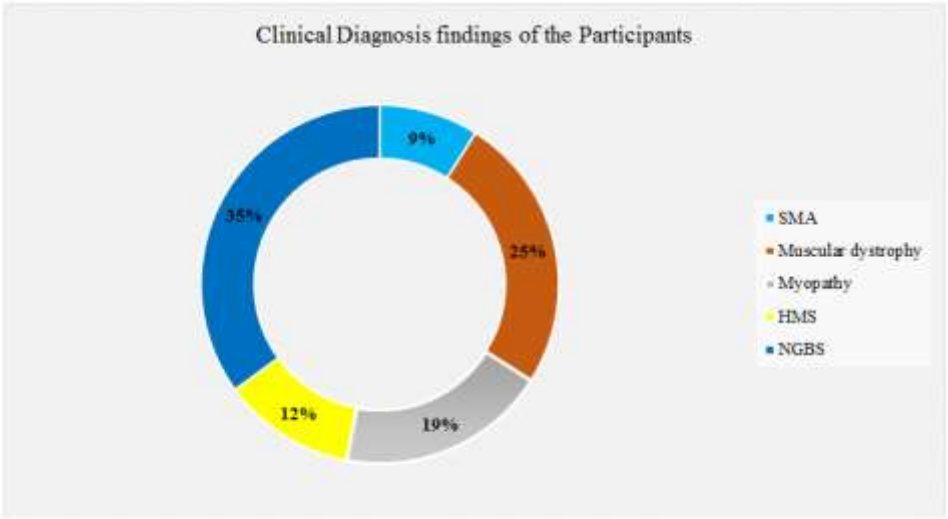

Fig-II: Clinical diagnosis Findings of The Participants (N=65)

Table-2: Investigation reports of EMG among participants (N=65)

Findings	Comment	n	%
Normal finding		21	32.31
Denervation and re-innervation	Variety of SMA, Late-onset SMA	16	24.61
Low amplitude scanty MUAPS	Myopathic disorder	14	21.53
Diffuse fibrillation, biphasic complex, repetitive spontaneous discharge	Severe myopathy mixed with a neurogenic component	6	9.23
Not elicited		8	12.32

Table-3: Investigation reports of NCS among participants (N=65)

Findings	n	%
Normal finding	34	52.3
Low amplitude CMAP and SNAP	3	4.62
Low amplitude CMAP	6	9.23
Variable sensory conduction velocity	3	4.62
Slow conduction velocity with temporal dispersion	4	6.15
No recognizable F wave latency	4	6.15
Not elicited	11	16.93

Table-4: Compatibilities of EMG in diagnosis among participants (N=65)

Type of disease	Clinical Diagnosis	Electrophysiological Diagnosis	Compatibility
Neurogenic disease	37	25	68%
Myogenic disease	28	20	71%

Table-5: Compatibilities of NCS in diagnosis among participants (N=65)

Type of disease	Clinical Diagnosis	Nerve Conduction Study	Compatibility
Neurogenic disease	37	17	46%
Myogenic disease	28	14	50%

4. DISCUSSION

In this study, among a total of 65 participants, the male-female ratio was 1.6:1. The mean (±SD) age of the participants was 3.75 ± 3.1 years. Basically, a single fiber electromyogram (EMG) is more sensitive in detecting and assessing neuromuscular transmission disorders (NTD), repetitive motor nerve stimulation may not be needed [12]. In this study, in analyzing the mode of presentations, we observed that 68%, 56%, and 54% of patients were with hypotonia, motor skill regression, and motor developmental delay respectively. As clinical diagnosis SMA, muscular dystrophy, and myopathy were found among 35%, 25%, and 19% of patients respectively. As investigation reports of EMG, among all participants, we found normal findings in 21. Myasthenia may be variable in its electrodiagnostic manifestations and presentations. Occasionally, fibrillation potentials may be found. If the disease is severe in condition, the MUAPs may be complex but small, suggesting a myopathy [13]. Although a quantitative electromyogram (EMG) often is not in needed in the diagnosis, it is beneficial in understanding the electrophysiologic aspects of neuromuscular disorders [14]. Single fiber EMG generally is a useful electrodiagnostic tool in detecting disorders of neuromuscular transmission which is more sensitive than repetitive motor nerve stimulation [15]. Single fiber EMG may be abnormal even before clinical weakness is present [12]. In this study, the compatibilities of EMG among the total 65 participants of electrophysiological diagnosis were found 68% for detecting neurogenic disease and 71% for detecting myogenic disease. In another study [16] the sensitivity rate was found 88%. On the other hand, the compatibilities of NCS were found at 46% for detecting neurogenic disease and 50% for detecting myogenic disease.

Limitation of the study

Though this was a single-center study with a limited sample size, so findings of this study might not reflect the exact scenario of the whole country

5. CONCLUSION & RECOMMENDATION

During the past six decades, electrodiagnosis (EDX) has played an increasingly important role in the clinical evaluation of patients who have neuromuscular disorders. NCS (Nerve

conduction study) and EMG (Electromyogram) are crucial investigations to reach confirmation as well as the exclusion of the diagnosis of neuromuscular disorder in children. For getting more specific findings we would like to recommend conducting similar studies with larger-sized samples in several places.

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