Journal of Bangladesh College of Physicians and Surgeons
Vol. 40, No. 4, October 2022

Electromyographic Patterns in Children presented as Floppy Baby: Experience in a Neurophysiology Laboratory of a Tertiary care hospital in Bangladesh

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Abstract

Introduction: Electromyography (EMG) is an invaluable diagnostic tool to reach a diagnosis in patients with hypotonia. The objective of this study was to observe the EMG patterns in children presented as floppy baby.

Methods: It was a cross-sectional study conducted in the neurophysiology department of the National Institute of Neurosciences and hospital over 4 years. Floppy children, aged 1 month to 5 years, referred to the laboratory for EMG were enrolled in the study. Hypotonia due to acute flaccid paralysis or children having drugs that could cause decreased muscle tone were excluded from the study. Thereafter, children were categorized into groups of central, peripheral, and uncategorized/mixed hypotonia. EMG was done in all patients and an electro-diagnostic impression was made.

Results: Out of 72 patients, 54.17% were male and 45.83% were female. Their average age of performing EMG was 26.81±18.12 months. Clinically it was observed that central and peripheral hypotonia were present in 18.05% and 70.83% of cases respectively and in 11.11% of cases, hypotonia could not be differentiated. EMG revealed Spinal muscular atrophy (SMA) in 38.89% of cases, followed by myopathy (26.39%), peripheral neuropathy (9.72%), and neuromuscular disorder (2.78%). In 19.44% of cases, EMG findings remained normal.

Conclusion: Hypotonia of peripheral origin was the most common EMG finding in floppy children. SMA is the commonest electro-diagnostic impression followed by myopathy.

Keywords: Electromyography, Floppy baby, peripheral hypotonia, Spinal muscular atrophy, myopathy

(J Bangladesh Coll Phys Surg 2022; 40: 240-245)
DOI: https://doi.org/10.3329/jbcps.v40i4.61881

Introduction

The electrodiagnostic study provides an extension to the neurological examination for the evaluation of neuromuscular disease. The evaluation of children with electromyography (EMG) requires special care and technique because of the discomfort of the tests. EMG is often used to evaluate hypotonia in infants, assess the severity, and also helps to localize the lesion. EMG may contribute to the diagnosis of spinal muscular atrophy (SMA), myopathy, muscular dystrophy, disorders of neuromuscular transmission (NMJ), hereditary polyneuropathies, and myotonic disorders.[1]

The term floppy baby is commonly used to denote babies who have either low muscle tone (hypotonia) or low muscle power (weakness) or have ligamentous laxity and an increased range of joint mobility. Strictly speaking, this term should be used for hypotonic babies.[2]The hypotonia is usually evident at birth or identified during early infancy. These infants exhibit delayed motor milestones and poor control of the movement with hypotonic movement patterns. Weak infants always have hypotonia, but hypotonia may present without weakness.[3] This clinical diagnosis has numerous etiologies and establishing a specific diagnosis in the individual case may often be labor-intensive.

Hypotonia is encountered in the diseases of the central nervous system, anterior horn cell, peripheral nerves,

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Received: 5 Oct, 2021 **Accepted:** 25 August, 2022

muscle, and the neuromuscular junction.[4] Based on the clinical criteria hypotonia can be classified into two major groups: central and peripheral. Central causes are more frequent than peripheral disorders. Central hypotonia may be due to cerebral insult, brain malformation, genetic defects, congenital syndromes, inborn errors of metabolism, and neurometabolic disorders. Peripheral disorders include SMA, hereditary neuropathies, myasthenia, botulism, congenital myopathy, muscular dystrophy, and some metabolic and multisystem diseases.[5] The presence of both central and peripheral manifestations point toward mixed and more complex hypotonia such as acid maltase deficiency, and chromosomal disorders.[6]

Although the precise sensitivity of EMG in the evaluation of floppy children is not specified, it has overall 80% sensitivity for the correct diagnosis.[1] In skilled hands, EMG is particularly helpful in making a rapid electrophysiological diagnosis, localizing the site of involvement in the motor unit, as well as in instances where a genetic analysis has failed to make a specific diagnosis. Several studies have been done on the usefulness of EMG in evaluating floppy children worldwide but lacking in our country. The objective of this study was to observe the patterns of EMG in the evaluation of floppy children and also to categorize the neuromuscular diseases among floppy babies who were referred to the department of Neurophysiology for EMG.

Methods

This was a cross-sectional study. The study was conducted at the National Institute of Neurosciences and Hospital in the department of Neurophysiology for four years from January 2017 to December 2020. Patients aged 1 month to 5 years having generalized hypotonia (Fig 1) since birth or infancy with or without weakness referred from the department of pediatric neurology and other hospitals between that period were enrolled in the study. Detailed history and clinical examination were done by a pediatric neurologist. Hypotonia was classified as central or peripheral according to the clinical findings.[5] Acute flaccid paralysis of any etiology and children who were on medication that could cause decreased muscle tone were excluded from the study.

Clinical features suggestive of central and peripheral hypotonia[5]

Central hypotonia	Myopathic facies
Dysmorphic features	Normal head growth
Microcephaly	Motor delay with relatively normal social and cognitive function
Social and cognitive impairment with motor delay	Family history of neuromuscular diseases
Features of pseudobulbar palsy	Low pitched weak cry
Axial weakness is more than the limbs	Frog-leg posture
Brisk jaw jerks	A paucity of spontaneous movement
Scissoring	Limb weakness is more than axial
Seizures	Increased range of joint mobility
Normal or brisk tendon reflexes	Muscle fasciculation
Extensor planter response	Reduced or absent deep tendon reflexes

Fig 1: *Typical posture of limbs of a child with generalized hypotonia (Frog-leg posture)*

Electromyography (EMG)

EMG was performed on all patients by trained pediatric neurophysiologists. Nerve conduction study (NCS) of the right upper limb (median and ulnar nerves) and left lower limbs (tibial, peroneal, and sural nerves) were done using the Neuropath S1 EMG machine by Nihon Kohden, Japan. The latency, amplitude, and nerve conduction velocity of compound muscle action potential (CMAP) and sensory nerve action potential (SNAP) were recorded. These were compared to a standard table of reference value[1]. Needle EMG was performed in the muscles of mild to moderate clinical

weakness of both upper and lower limbs. Based on the EMG findings patients were categorized as having myopathy, neuropathy, anterior horn cell disorder, neuromuscular junctional disorder, central hypotonia, or normal according to standard criteria.[7,8]

EMG patterns

Myopathy:

Low amplitude, polyphasic, short-duration motor unit action potentials (MUAPs) with early interference pattern (Fig:2).

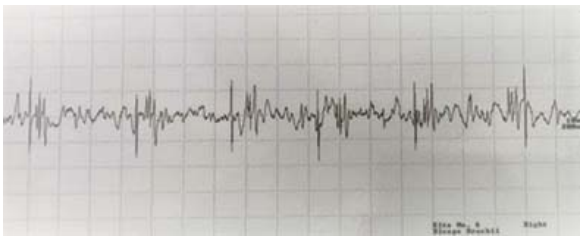

Fig 2: *EMG of a 3-year-old boy showing myogenic MUAPs*

Spontaneous fibrillation at rest, long-duration polyphasic MUAPs, decreased interference pattern (Fig: 3).

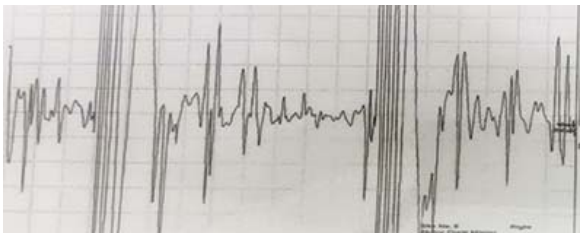

Fig.-3: *EMG of a 2-year-old boy showing neurogenic MUAPs*

Anterior horn cell disorder:

Spontaneous fibrillation at rest, long-duration polyphasic MUAPs, decreased interference pattern), normal latency, and SNAPs in sensory NCS.

In central disorders:

Either a normal pattern or decreased activation (although the number of MUAPs is reduced, it is appropriate for the level of firing).

Neuromuscular junctional disorder:

A decremental response at 2–3 Hz rates of stimulation in at least one muscle was considered as defective neuromuscular transmission in Congenital myasthenic syndrome (CMS).

All demographic, clinical, and EMG data were recorded in a prefixed questionnaire. All data were collected, presented, and analyzed by using an appropriate statistical package program (Statistical Package for Social Science (SPSS), version, 20). Continuous data were presented as means and standard deviations, whereas categorical data were presented as numbers and percentages.

Written informed consent was taken from the parents before inclusion in the study. The study was approved by the ethical review committee of the institute.

Results

A total of 72 children were enrolled in this study and the ages to presentation in the neurophysiology laboratory range between 2 months and 60 months. Their mean age was 26.81±18.12 months. Among the studied population, 54.17% were male and 45.83% were female. In the present study, consanguinity was present in 20.83% and a family history of a similar condition was found in 15.28% of cases. In addition to hypotonia, facial dysmorphism was seen in 8.33%, and 4.17% of cases and was associated with other congenital anomalies. Bulbar palsy and fasciculation were found in 5.56% and 15.28% of cases respectively (Table-1).

Table I

Clinical characteristics of floppy children (n=72)

Variables		
Age(months)	Range	2-60
Mean±SD		26.81±18.12
		Number (%)
Sex	Male	39 (54.17)
	Female	33 (45.83)
Consanguinity		15 (20.83)
Positive family history		11 (15.28)
Decreased fetal movement		8 (11.11)
Polyhydramnios		2 (2.78)
History of perinatal asphyxia		6 (8.33)
Facial dysmorphism		6 (8.33)
Associated with other congenital anomalies		3 (4.17)
Bulbar involvement		4 (5.56)
Fasciculation		11 (15.28)
Seizure		8 (11.11)

Out of 72 cases, 18.05% had central hypotonia, 70.83% had peripheral hypotonia and in 11.11% of cases, hypotonia could not be differentiated (Table 2). Concerning the EMG impression of floppy babies, anterior horn cell disorder (concomitant with the clinical diagnosis of SMA) was the most frequent diagnosis followed by myopathy. A decremental response was found in only 2.78% of cases in a background of fatigable weakness and negative AchR antibody. This finding pointed towards a diagnosis of CMS. EMG was within the normal limit in 19.44% of cases (Table-3).

Table II

Clinical categorization of hypotonia (n=72)

Pattern of hypotonia	Number (%)
Central hypotonia	13 (18.05)
Peripheral hypotonia	51(70.83)
Uncategorized / mixed	8 (11.11)

Table III

Distribution of cases according to EMG findings (n=72)

Diagnosis	Number (%)
SMA (anterior horn cell disorder)	28 (38.89)
Myopathy	19 (26.39)
Peripheral neuropathy	7 (9.72)
Neuromuscular junctional disorder (NMJ)	2 (2.78)
Central hypotonia	2 (2.78)
Normal findings	14 (19.44)

Discussion:

The floppy children often represent a diagnostic challenge for clinicians. A well-organized systemic approach is essential for evaluating a child with hypotonia. History and clinical examination should be more emphasized as it is very essential in localizing the problem to a specific region of the nervous system.

The initial approach is to determine whether the problem is due to central or peripheral origin. This is of utmost importance when planning diagnostic investigations. In central hypotonia, Magnetic Resonance Imaging (MRI) or Computed tomography (CT) of the brain should be considered. A karyotype is indicated when dysmorphic features are present. A metabolic workup is needed for the evaluation of neurometabolic disorder. For the evaluation of peripheral hypotonia, creatinine phosphokinase (CPK) should be measured first as it is highly elevated in muscular dystrophy but remains normal or slightly elevated in SMA and congenital myopathy.[9] EMG is an important diagnostic tool to evaluate a child with peripheral hypotonia and has particular strengths in the diagnosis of SMA, various myopathies, Charcot-Marie-Tooth disease (CMT), and disorders of NMJ.[10] It has 80% sensitivity for the diagnosis of SMA and 75% sensitivity to conform to a myogenic pattern. [11] In a study, Guillen D, et al. found that the sensitivity of EMG for possible or confirmed neuromuscular disease was 72%, specificity 79%, positive predictive value 72%, and negative predictive value 79%. Thus, EMG could be a good screening test before considering a genetic or muscle immuno-histochemical study in our country where healthcare facilities are limited.

In our study, the majority of the floppy children are male (54.17%) and the mean age of presentation to the neurophysiology laboratory for EMG is 26.81±18.12 months. The youngest age is of 2 months. Some studies have shown that majority of hypotonic children presented at birth or early infancy.[13,14]This discordance may be due to our poor referral system or the scarcity of pediatric neurophysiologists. In a study conducted in Iran[15], the mean age was 18.2 ± 9.76 months which is in concordance with our study.

The results of this study have shown that most of the children had hypotonia of peripheral origin rather than central. In a few previous studies, it was described that central hypotonia was the most frequent cause of hypotonia among floppy children and mostly due to cerebral palsy, structural malformation of the brain, and genetic or metabolic disorders.[15-17] This difference is because our institute is a tertiary neurology hospital and there is a well-organized pediatric neurology department. The department runs various clinics (Neuromuscular, Cerebral palsy. Epilepsy, etc.) from where the patients are scrutinized and patients having central hypotonia were first investigated with imaging or metabolic screening. Patients suspecting neuromuscular disorders, mixed hypotonia, or hypotonia of unknown origin are referred for EMG to provide critical

assistance to locate the site of lesion, narrow the differential diagnosis, and therefore help the clinicians to plan further investigations.

Electrophysiological data of our study reflects that SMA is the most common electro-diagnostic impression followed by myopathy. Peripheral neuropathy and Disorders of NMJ are relatively uncommon among the studied population. Similar etiological profiles are also described in studies of various countries.[11,13,18-19] In this study, EMG findings were normal in 19.44% of cases. This is probably because hypotonia is not located within the peripheral nervous system. A previous study has stated that except for a few myopathies, normal EMG findings are suggestive of hypotonia of central origin.[6] Birdi K et al, in a study, did not find a clear etiology of hypotonia in 31% of cases.[20] However, our study has revealed that patients with evidence of peripheral hypotonia with or without central causes benefited the most from EMG. One study suggests that EMG is very accurate in SMA and is often used as supportive evidence to establish the diagnosis of SMA.[21]

Our study has shown that although EMG does not give a definite diagnosis still it is indispensable in deciding whether the weakness is due to neuropathy, myopathy, neuronopathy, or NMJ disorders. The categorization of floppy children is very important to consider which tests should be done next. Therefore, EMG complements other diagnostic test modalities. In resource-poor countries like ours where genetic tests are expensive and difficult to obtain, the information that an EMG gives is appreciated.

Conclusion

In this study, peripheral hypotonia is the most common cause of requesting EMG among children of 5 years or less. SMA is the most frequent cause of peripheral hypotonia followed by myopathy, peripheral neuropathy, and NMJ disorders. Further studies are recommended on a larger scale to explore the causes of floppy babies in our population.

Conflict of interest: There is no conflict of interest relevant to this paper to disclose.

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