THE VALUE OF ELECTROMYOGRAPHY AS A DIAGNOSTIC TOOL FOR FLOPPY INFANTS IN FAYOUM UNIVERSITY HOSPITALS

Thesis

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Dedication

I dedicate this work to the soul of my father and to my mother who will never be thanked as much as they deserve.

To my wife and to my little daughter who gave me lots of patience and support, I also dedicate this work.

I will never forget my brother whom I really wanted to be here.

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Abstract

Background: floppy infants represent a large part of the activity in the neurology clinic of Fayoum University Hospital. Neuromuscular disorders represent a common cause of floppiness in infancy period.EMG is an easy and cheap tool that can be used in diagnosis of these infants.

Objectives: To evaluate Electromyography as a diagnostic tool for floppy infants and to assess the value of its use to classify floppy infants according to the etiology.

Methods: A cross sectional descriptive study included ٤٢ floppy infants sought medical advice in the neurology clinic of Fayoum University Hospital during the period from May ٢٠١٥ till July ٢٠١٥.The study included infants blow the age of ٢ years with peripheral hyptonia.

Results: ٣٩ patients (٩٢.٧٥ %) out of ٤٢ patients performed EMG showed positive results.٢٣ cases (٥٥%) out of the ٣٩ patients showed neuropathic potentials discharging in a regular pattern denoting anterior horn cells affection (SMA) with normal Motor nerve conduction velocity study, ٢cases showed neuropathic potentials with axonal affection (diminished amplitude) on motor nerve conduction velocity studies and ١٣ cases (٣٢.٧٥%) showed myopathic potentials and normal motor nerve conduction velocity studies.٣ cases (٢١%) out of ١٣ patients showed myopathic motor unit action potentials were diagnosed according to clinical picture and other diagnostic criteria (liver & muscle biopsy, radiological imaging, ocular assessment and enzyme assay) into Arthrogryposis multiplex congenita, Muscle eye brain disease and glycogen storage type ٢ disease. ١٠ cases (٧٩%) out of ١٣ cases showed myopathic mptor unit action potentials were diagnosed as congenital myopathic diseases. One of these cases performed EMG twice at ٣ months interval and after the age of one year to be diagnosed as congenital myopathic disease. A case (٢.٥%) out of ٤٢ patients performed EMG was diagnosed as Nemaline Myopathy on muscle biopsy and showed normal ENMG results.

Conclusion: ENMG is a cheap diagnostic tool compared to new molecular diagnostic tools and gives a good clue for the next step for diagnosis of the cause of floppiness. Neuropathic potentials on ENMG are diagnostic for spinal muscular atrophy especially if clinical picture is going well with the diagnosis.

Keywords: floppy infants, EMG, neuromuscular disease, SMA, myopthic patients.

Abbreviations

AHCs: Anterior horn cells
AMC: Arthrogryposis multiplex congenita
CFTD: Congenital fiber type disproportion myopathy
CMAP: Compound muscle action potential
CMD: Congenital muscular dystrophy
CMS: Congenital myasthenic syndrome
CMT: Charcot marie tooth disease
CNS: Central nervous system
CPEO: Chronic progressive external ophthalmoplegia
CK: Creatinine kinase
CT scan: Computerized tomography
FCMD: Fukuyama muscle disease
FDA: US Food and Drug Administration
EEG: Electroencephalography
EMG-ENMG: Electromyography-Electroneuromyography
GSD: Glycogen storage disease
HIE: Hypoxic Ischemic Encephalopathy
HSAN: Hereditary sensory and autonomic neuropathy
KSS: Kearns-Sayre syndrome
LMN: Lower motor neuron
MD: Muscular dystrophy
MEB: Muscle eye brain disease
MELAS: Mitochondrial encephalopathy – myopathy – lactic acidosis – stroke
MERRF: Myoclonic epilepsy and ragged-red fibers
MND: Motor neuron disease
MRI: Magnetic resonance imaging
MUAP: Motor unit action potential
NCV: Nerve conduction velocity
NICU: Neonatal intensive care unit
NMD: Neuromuscular diseases
PBDs: Peroxisome biogensis disorders
PSW: Positive sharp waves
PICU: Pediatric intensive care unit
RCDP١: Rhizomelic chondrodysplasia punctata type ١
SMA: Spinal muscular atrophy
UMN: Upper motor neuron
WWD: Walker Wurberg Disease

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