NEUROMUSCULAR DISEASES IN RELATION TO ANAESTHESIA

Essay

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M.Sc. of Anesthesiology

by

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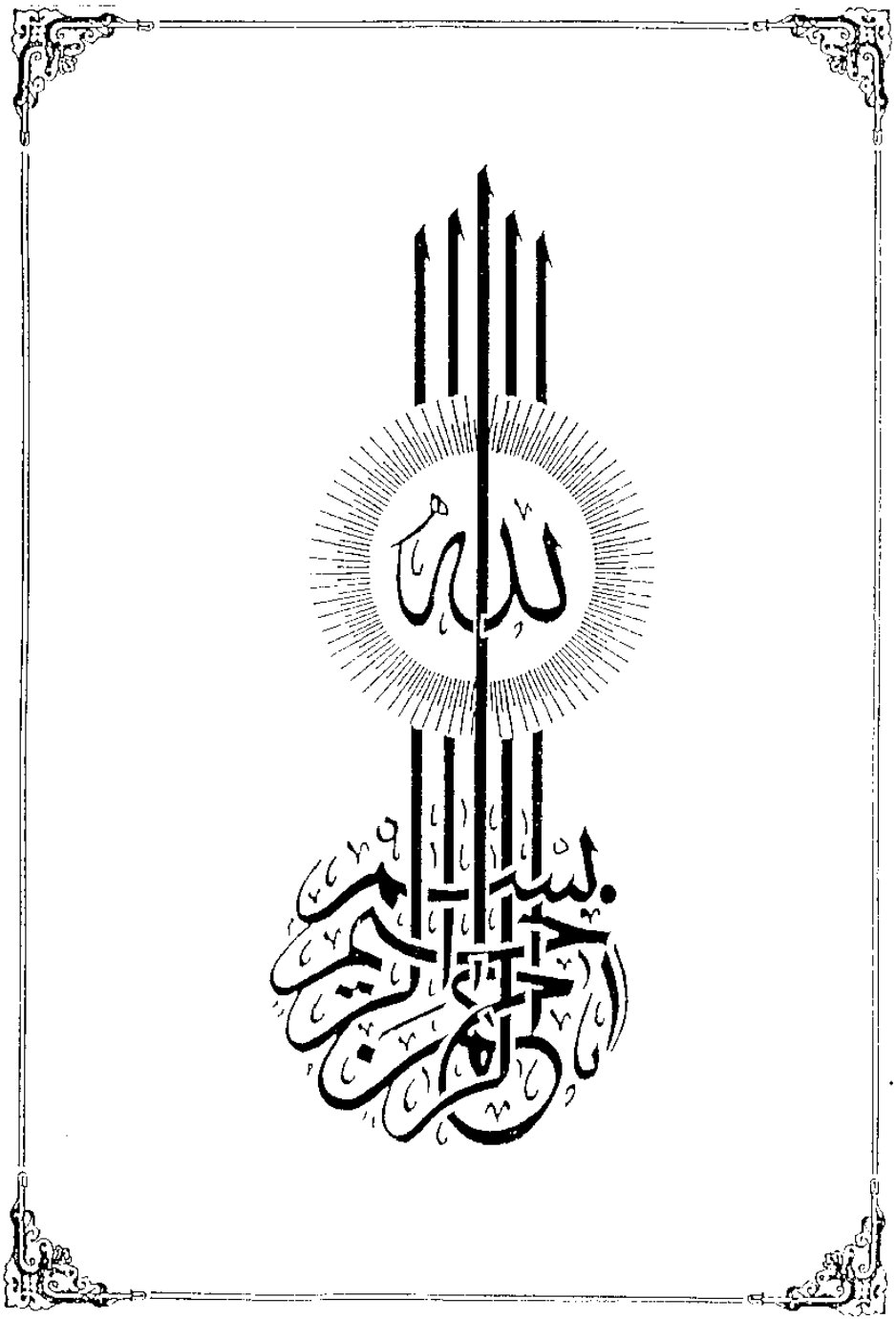
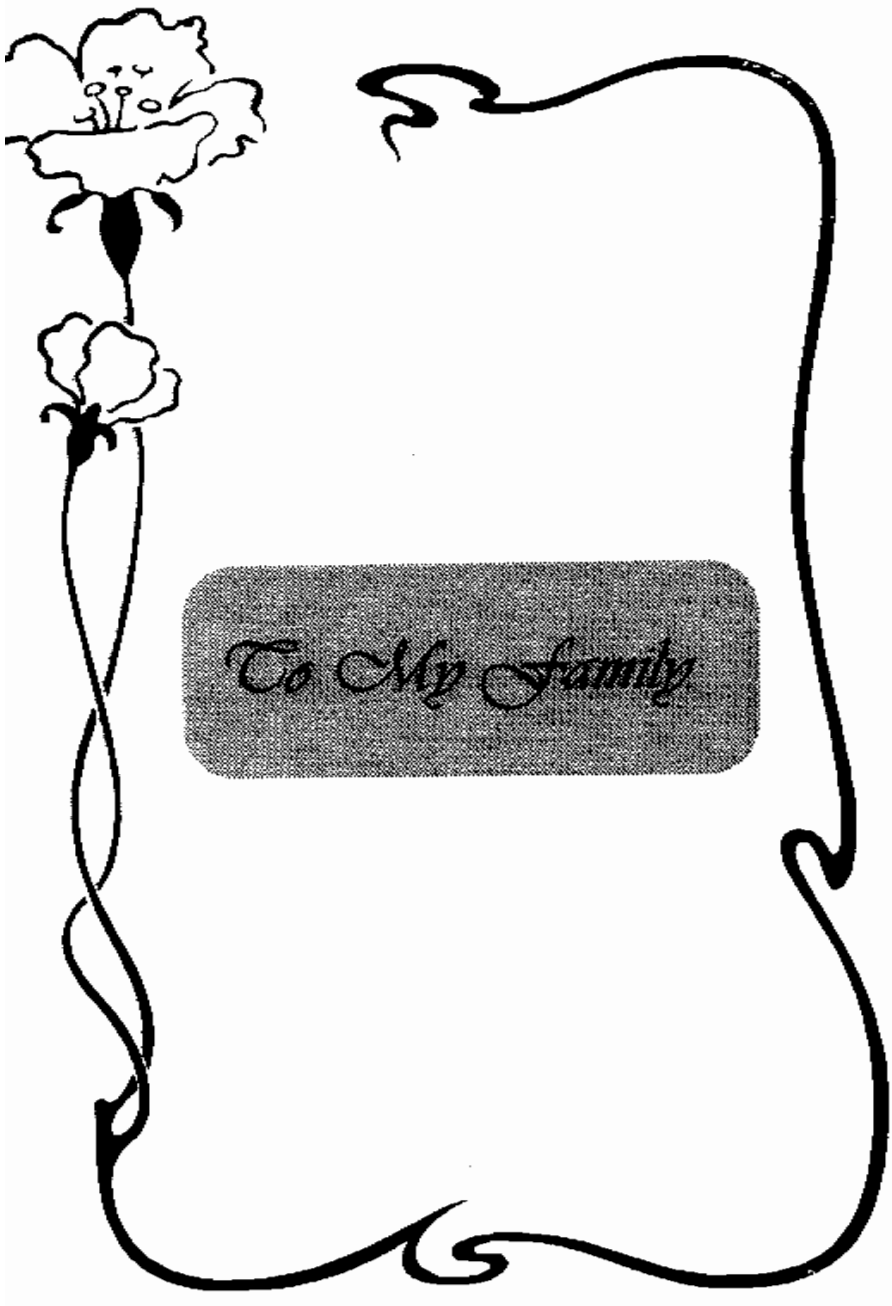
قالوا سبحانك لا علم لنا إلا ما علمتنا إنك أنت العليم الحكيم

﴿ سورة البقرة – الآية ٣٢ ﴾




To My family

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Khaled Mohamed Abdallah

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INTRODUCTION

Introduction

Many different diseases are classified as neuromuscular ieases, and the majority of them are rare and difficult to fferentiate, unless one is dealing with them on a daily basis. owever, patients with neuromuscular disorders often share the me three problems in relation to anaesthesia, independent on the pe of disorder; they may have a cardiomyopathy, a restricted spiratory capacity, and an abnormal response to muscle relaxants. , during preoperative evaluation of patients with neuromuscular sorder, the anaesthetist should keep in mind that those patients ve got some sort of cardiopulmonary insufficiency even not mptomatized. Unfortunately, neuromuscular diseases may be scovered accidentally on receiving drugs particularly in relation to aesthesia, nervertheless anaesthesia and surgical interventions ay induce the disease itself (myotonia) or other fatal complication alignant hyperthermia in patients with Duchenne muscular strophy).

For the purpose of clarity, neuromuscular disorders could be assified into five main groups according to site of primary lesion :

Intracranial lesions as * hemiplegia, * multiple sclerosis, and * diffuse intracranial lesions.

Spinal cord lesions as * paraplegia and * amyotrophic lateral lerosis.

Peripheral nerve lesions as * peripheral neuropathies, * muscular denervation, and * disuse atrophy.

Neuromuscular junction lesions as * myasthenia gravis and * myasthenic syndrome.

- Muscular lesions as * myotonia and * muscular dystrophy.

In this essay, we will consider mainly neuromuscular junction lesions and some of the muscular lesions such as myotonias and myasthenia gravis and their anaesthetic implication.

PHYSIOLOGY OF NEUROMUSCULAR TRANSMISSION

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Anatomy of the Neuromuscular Junction :

The motor neuron runs without interruption from the ventral orn of the spinal cord to the muscle as a large myelinated axon. As ıe axon approaches the muscle, it branches repeatedly to contact ıany muscle cells and to gather them into a functional group known s a motor unit. The synapse between the motor nerve and the ıuscle is termed the neuromuscular junction (NMJ) (Figure 1) *Standaert, 1986)*.

In the human body, each muscle fibre is supplied by only one xon branch and it receives its innervation at a discrete point on its ırface, that is to say, it is focally innervated. The extraocular ıuscles, and few others are exceptions in that many of their muscle bres receive a dense innervation, and hence possess multiple NMJs istributed over their surface *(Bowman, 1989)*.

When the motor nerve fibre reaches the muscle fibre, it loses s myelin sheath and the neurolemma blends with the sarcolemma. Ieanwhile, the axoplasm of the nerve fibre spreads out in close ontact with the sarcoplasm of the muscle fibre, being separated 'om it by a potential gap of about 20 nm, the junctional cleft *Baraka, 1982)*.

The whole neuromuscular junction is surrounded by ıembrane which is closely adherent to the nerve termed the chwann cell membrane. It separates the junctional cleft from the xtracellular fluid and may serve to mechanically preserve the euromuscular junction. The muscle surface is heavily corrugated at

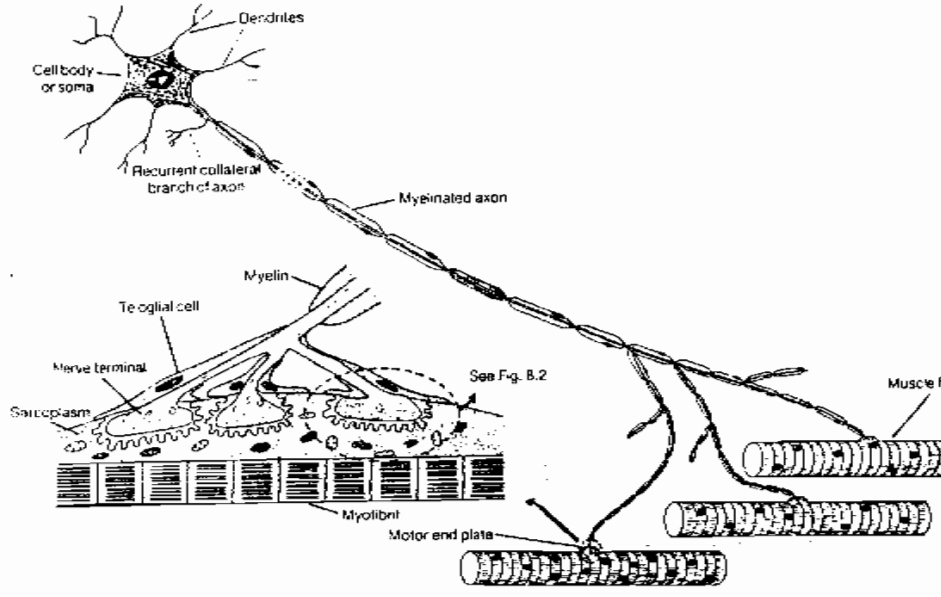

Figure (1) : Diagram of a motor unit containing focally innervated muscle fibres. A motor end plate is enlarged as the inset on the left *(Bowman, 1989)*.

he neuromuscular junction and deep invaginations of the junctional left into the muscle, the secondary clefts, are separated by folds, the unctional folds, in the surface. Around the crest of each of these econdary clefts is a zone rich in cholinesterase. In these areas lie he cholino-receptors *(Durant, 1984)*.

The nerve terminal is filled with mitochondria and material of chemical transmission. It contains structures called synaptic vesicles, that are round bodies about 500 A° in diameter and contain acetylcholine molecules. The acetylcholine content of each vesicle s referred to as a quantum. The contents of the nerve endings are not homogenous. The vesicles are congregated in the portion oward the junctional surface while microtubules, mitochondria, and other support structures are aligned toward the opposite side. The vesicles are ordered in a repeating pattern of triangles arrays with he apex near thickened transverse bands of the terminal axonal nembrane, so called "active zones". There are small particles arranged alongside the active zones between vesicles, these are pecial proteins that form channels which allow calcium to enter the nerve and cause the release of vesicles (Figure 2) *(Fahim et al., 984)*.

As the membrane on the muscle side is highly convoluted, it exposes a very large area to the junctional cleft. The surface is highly organized. The top of each fold runs exactly between two active zones in the nerve ending. The shoulders of each peak are aligned with the vesicles release sites at the sides of active zones *Feldman, 1991)*.

Receptors are concentrated on the shoulders, almost completely covering the surface. Their numbers diminish as the distance of the shoulder increases, so that they are sparse deep in the cleft between two folds. In contrast, acetylcholinesterase spreads