Updates in Anesthetic Management for Patients with Muscle Diseases

Thesis
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Master Degree of Anesthesiology

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List of Contents

List of Abbreviations

Ach :	Acetylcholine.
ADP :	Adenosine Diphosphate.
ALS :	Amyotrophic Lateral Sclerosis.
ASA :	American society of anesthesiology
ATP :	Adenosine triphosphate.
Ca :	Calcium ion
CHCT:	Caffeine-halothane contracture test
ChEI :	Cholinesterase inhibitors
CPEO :	Chronic Progressive External Ophthalmoplegia
CT :	Computed tomography
DM :	Dystrophia myotonica
DMD :	Duchenne muscular dtstrophy
DMPK :	Myotonic Dystrophy protein kinase
GBs :	Guillain-Barre syndrome
ICU :	Intensive Care Unit
KSS :	Kerans-Sayre Syndrome
LEMS :	Lambert Eaton Myasthenic Syndrome
MEALS:	Myopathy, Encephalopathy, Lactic Acidosis and Stroke
MERRF:	Myoclonic epilepsy with ragged red fibers
MG:	Myasthenia Gravis
mRNA :	Messenger Ribonucleic Acid
MRI:	Magnetic Resonant Imaging
nAChRs:	Nicotinic Acetylcholine Receptors
NMJ:	Neuromuscular junction
NMBAs:	Neuromuscular blocking agents
PACU:	Post Anesthetic care unit
PCP :	Primary care physician
RYR1 :	Reynauld recetor type 1
SCh :	Succinylecholine
SR :	Sarcoplasmic Reticulum
TIVA :	Total Intravenous Anesthesia.

List of Figures

List of Tables

المعالجه التخديرية لامراض العضلات

رسالة
توطئة للحصول على درجة الماجستير فى التخدير

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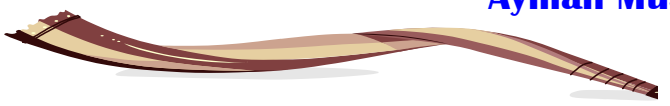
Ayman Mustafa Khalaf



سورة البقرة الآية: ٣٢

Introduction

Neuromuscular disorders are a heterogeneous group of diseases affecting skeletal muscles and have a pathophysiologic basis at the level of the central nervous system, peripheral nerves, neuromuscular junction, or muscle fiber. These disorders are often grouped together in relation to anesthesia due to common considerations in the perioperative period. In addition to abnormal responses to muscle relaxants, these diseases may be associated with systemic problems that may profoundly affect the conduct of anesthesia. Anesthetic management may be required either for problems relevant to the disorder or for comorbid conditions **(Haes et al., 2002).**

Anesthesia for patients with neuromuscular disorders may be challenging. Many disorders are associated with diminished cardiopulmonary reserve, difficult airway, deficient airway protection mechanisms or cardiac, respiratory, renal and metabolic problems. Thus thorough preoperative assessment and choice of anesthetic technique and monitoring should be made with consideration of the potential airway and cardiovascular problems **(Klingler et al., 2005).**

Neuromuscular blockers can have unpredictable effects in patients with neuromuscular disorders and should be monitored carefully when used. The differences in pharmacokinetics and pharmacodynamics observed when using

neuromuscular blocking drugs in patients with disorders of the nervous or muscular systems can be mostly explained in terms of an alteration in the distribution of nicotinic acetylcholine receptors. Normally these receptors are mainly confined to the neuromuscular junction but in disease states they can spread out from the neuromuscular junction along the length of the muscle fibers Succinylcholine is formally contraindicated as it may cause hyperkalemia sufficient to produce cardiac arrest **(Shieldsetal, 2006).**

Regional anesthesia is preferable to general anesthesia, especially in patients with respiratory and cardiac problems; however, there is a concern that complications of the procedure may be difficult to distinguish from progression of the disease process. In those with cardiovascular complications and autonomic dysfunction, severe hypotension may result from neuroaxial blockade **(Khan et al., 2005).**

Intravenous anesthetic agents can facilitate intubation without the use of neuromuscular blocking drugs and the use of total intravenous anesthesia with short acting intravenous anesthetic agents and opioids can accelerate recovery and reduce postoperative respiratory failure. Although general anesthesia with volatile anesthetics have a cardio depressant effect by decreasing release of calcium from the sarcoplasmic reticulum and decreasing the responsiveness of the contractile

filaments to calcium, inhalational anesthetics have been widely used **(Sambrook et al., 2001).**

Complications during and after anesthesia include rhabdomyolysis, malignant hyperthermia, malignant hyperthermia like reactions and primary or secondary changes relating to the underlying neuromuscular disorder such as cardiorespiratory problems, autonomic dysfunction, as well as hypothermia or hyperthermia. Admission to the intensive care unit for postoperative management should always be considered given the significant complications that may occur **(Steven et al., 2001).**

Aim of the Work

This thesis spot light on muscle diseases, the current concept of the pathogenesis, anesthetic considerations as well as anesthetic management and perioperative complications.

Pathophysiological Consideration of Skeletal Muscle

Muscle is a tissue that shortens and develops tension so that movement is brought about. Muscles are divided into two types based on their appearance in light micrographs. Striated muscles, which include skeletal and cardiac muscle and are characterized by alternating light and dark bands and Smooth muscles that have no distinguishing surface features **(Mackenzie, 2001).**

Through sustained contraction or alternating contraction and relaxation, muscle tissue has five key functions; producing body movements such as walking and running, force production for maintaining posture and stabilizing joints, heat production, moving substances within the body and regulating organ volume **(Baechle, 2000).**

Muscle tissue has four special properties that enable it to function and contribute to homeostasis; electrical excitability, which is the ability to respond to certain stimuli by producing electrical signals, contractility which is the ability of muscle tissue to contract forcefully when stimulated by an action potential, extensibility which is the ability of muscle to stretch without being damaged and elasticity which is the ability of muscle tissue to return to its original length and shape after contraction or extension (**Mackenzie, 2001).**

Each skeletal muscle is a separate organ composed of hundreds to thousands of cells called fibers because of their elongated shapes. Connective tissues surround muscle fibers, whole muscles, blood vessels and nerves penetrate into muscle (**Saladin, 2010).**

Three layers of connective tissue extend from the deep fascia to protect and strengthen skeletal muscle. The outermost layer is the epimysium, the perimysium surrounds groups of 10 to 100 or more individual muscle fibers, separating them into bundles called fascicles. Both epimysium and perimysium are dense irregular connective tissue. penetrating the interior of each fascicle and separating individual muscle fibers from one another is the endomysium, a thin sheath of areolar connective tissue. Deep fascia, epimysium, perimysium and endomysium are all continuous with the connective tissue that attaches skeletal muscle to other structures, such as bone or another muscle (**Nagiub et al, 2002).**

Microscopic anatomy of a skeletal muscle fiber:

Each individual muscle fiber contains very fine, long protein strands called myofibrils, which are aligned side by side and extend the length of the fiber. They are the units which lengthen and contract the muscle. The myofibrils are "actually chains of tiny contractile units" called sarcomeres, which are aligned end to end like boxcars in a train along the length of the myofibrils (**Naguib et al, 2002).**

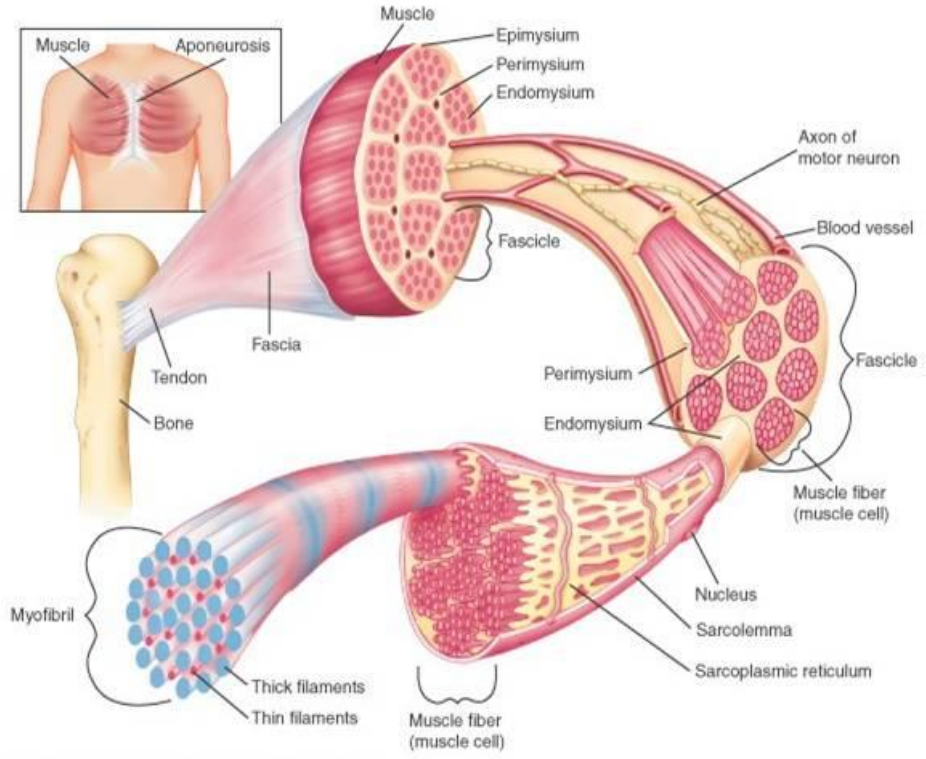


Figure (1): Showing structure of skeletal muscle **(Gerard, 2008).**

The multiple nuclei of a skeletal muscle fiber are located just beneath the sarcolemma, the plasma membrane of the fibers. Thousands of tiny invaginations of the sarcolemma, called T tubules, tunnel from the surface toward the center of each muscle fiber. T tubules are open to the outside of the fiber and thus are filled with extracellular fluid. This arrangement ensures that all parts of the muscle fiber become excited by an action potential simultaneously **(Mc Ardle, 2000).**

The sarcomeres are formed by even finer strands known as myofilaments. The myofilaments are composed of proteins