Summary and Conclusion

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The muscle diseases are all intrinsic disorders of skeletal muscle cell. So, skeletal muscle anatomy and physiology should be carefully understood. Skeletal muscles are accounting for most of the voluntary muscle mass of the body. Each skeletal muscle fiber is long, cylindrical, multinucleated and striated. Depending on the quantity of myoglobin, number of mitochondria, concentration of various, enzymes, and rate of contraction, the muscle fibers are red slow twitch (type I), white fast twitch (type II), or intermediate (type II B).

The final number of fibers in a muscle is attained before birth. Limited regeneration occurs in human skeletal muscle, but if damage is great, regeneration may not occur; lost muscle is then replaced by connective tissue.

Muscle contraction effectively reduces the resting length of the muscle fiber by an amount that is equal to the sum of all shortenings that occur in all sarcomeres of that particular muscle cell. The process of contraction, usually triggered by neural impulses, obeys the 'all- or-none- law" in that a single muscle fiber will either contract or not contract as a result of stimulation.

Because the process of muscle contraction consumes a great deal of energy, skeletal muscle cells maintain a high concentration of the energy- rich compounds ATP and creatine phosphate.

The neuromuscular junction (NMJ) is specialized, to transmit and receive chemical messages. Each motoneuron runs without interruption

from the ventral horn of the spinal cord to the neuromuscular junction as a large myelinated axon. As it approaches the muscle, it branches repeatedly to contract many muscle cells and to gather them into a functional group known as a motor unit.

The nerve is separated from the surface of the muscle by an approximate 20nm gap "the junctional cleft". The nerve and muscle are held in tight alignment by protein filaments, which span the cleft between nerve and end plate. The muscle surface is heavily corrugated. With deep invaginations of the junctional cleft, thus, the end plate's total surface area is very large. The shoulders of the folds are densely populated with acetylcholine receptors, about 5 millions of them in each junction.

Since all the muscle cells in a unit are excited by a single neuron, stimulation of the nerve either electrically or via an action potential originating from the ventral horn, or by any agonist, including depolarizing relaxants (e.g. succinylcholine), causes all muscle cells in the motor unit to contract synchronously. The synchronous contraction of the cells in a motor unit is fasciculation and often is vigorous enough to be observed throughout the skin.

The physiology and pharmacology of the NMJ is pivotal to many aspects of the practice of anesthesiology, including intraoperative care, intensive care unit (ICU) treatment, and pain management. Diseases such as the Lambert-Eaton syndrome, myasthenic syndrome, as well as exogenously administered magnesium and certain antibiotics, results in reduced presynaptic release of acetylcholine. Myasthenia gravis (MG) and rare congenital nicotinic channelopathies produce postsynaptic abnormalities of skeletal muscle receptor function. For instance, sodium,

and chloride channelopathies are now linked to myotonia and periodic paralysis. Mutations of channels at the sarcoplasmic reticulum have been identified in some cases of malignant hyperthermia.

Myotonic muscular dystrophy is a multisystem disorder characterized by slowing of relaxation after muscle contraction in response to electrical or percussive stimuli. Myotonic muscular dystrophy usually manifests in 2^{nd} to 3^{rd} decades. Myotonia is the principal manifestation early in the disease, but as the disease progresses, muscle weakness and atrophy become more prominent.

Multiple organ systems are involved in the disease as evidenced by perisenile cataract; premature frontal baldness; hyersommolence with sleep apnea; and endocrine dysfunction leading to pancreatic, adrenal thyroid, and gonadal insufficiency. Respiratory involvement leads to decreased vital capacity. Alveolar hypoventilation is caused by either pulmonary or central nervous systems dysfunction. Chronic hypoxemia may lead to corpulmonale. Gastrointestinal hypomotility can predispose patients to pulmonary aspiration. Uterine atony can prolong labor and increases the incidence of retained placenta. Cardiac manifestations, which are often present before other clinical symptoms appear, consist of atrial arrhythmias, heart block and depression of ventricular function. The myotonia is usually described by patients as a 'stiffness' that may ease with continued activity, the so-called 'warm- up" phenomenon.

Because of limited response to increased carbon dioxide, controlled ventilation during anesthesia and also postoperatively even after minor surgery is required.

The production of muscle relaxation in a myotonic patient is one of the most difficult problems facing anesthesiologist. As muscle relaxants

and nerve blocks, block only the motor nerve impulses, stimulation beyond the neuromuscular junction may still cause myotonia.

Cardiovascular monitoring is mandatory intra and post operatively. Care of aspiration should be taken care of.

Myasthenia gravis is characterized by weakness and easy fatigability of skeletal muscle. The weakness is thought to be due to autoimmune destruction or inactivation of postsynaptic acetylcholine receptors at the neuromuscular junction, leading to a reduced number of receptors and loss of folds n the post synaptic membrane.

The course of the disease is marked by exacerbations and remissions. The weakness can be asymmetric or generalized. Ocular muscles are most commonly affected, resulting in fluctuating ptosis and diplopia.

With bulbar involvement laryngeal and pharyngeal muscle weakness can result in dysarthria, difficulty in chewing and swallowing, difficulty in clearing secretions, or pulmonary aspiration. Muscle strength characteriscally improves with rest but deteriorates rapidly with exertion. Infection, stress, surgery and pregnancy have unpredictable effects on the disease but often lead to exacerbations. Fifteen percent of patients develop thymoma, while 65% have thymic hyperplasia. Other autoimmune disorders are also present in 10% of patients as (hypothyroidism, hyperthyroidism or rheumatoid arthritis).

Treatment is with anticholinesterase drugs, immunosuppressant, glucocorticoids, plasmapheresis, intravenous immunoglobulins and thymectomy.

Patients with myasthenia may present for thymectomy or for unrelated surgical or obstetric procedures. In all cases, patients should be under the best possible medical control prior to surgery. Patients scheduled for thymectomy often have deteriorating muscle strength; while those undergoing other elective procedures should be well controlled or in remission. Adjustments in anticholinesterase medication, immunosuppressants, or steroid therapy may be necessary.

Potential problems in continuing such therapy include altered patients requirements following surgery, increased vagal reflexes, and the possibility of disrupting bowel anastomoses secondary to hyperperistalsis. More over because these agents also inhibit plasma cholinesterase, they can prolong the duration of ester type local anesthetics and succinylcholine. Conversely, Patients with advanced generalized disease may deteriorate significantly when anticholinesterase agents are withheld.

Preoperative evaluation should focus on the recent course of the disease, the muscle groups affected; drug therapy, and coexisting illness. Patients with respiratory muscle or bulbar involvement are at increased risk for pulmonary aspiration. Premedication with metoclopramide or an H2 blocker may decrease this risk. Because some patients with myasthenia are often very sensitive to respiratory depressants, premeditation with opiods, benzodiazepines, and similar drugs is usually omitted. With the exception of muscle relaxants, standard anesthesic agents may be used in patients with myasthenia. Marked respiratory depression, however, may be encountered following even moderate doses of barbiturates or opiods. Propofol may be preferable because of its short duration of action. A volatile agent- based anesthetic is generally most satisfactory.

Deep anesthesia with a volatile agent alone in patients with myasthenia may provide sufficient relaxation for endotracheal intubation as well as most surgical procedures. The response to succinylcholine is unpredictable; patients may manifest a relative resistance, a prolonged effect or an unusual response. The dose of succinylcholine may be increased to 2mg/kg to over come resistance, but a prolonged effect should be anticipated. Many patients are sensitive to nondepolarizing muscle relaxants. Even a defasciculating dose in some patients can result in nearly complete paralysis. If a muscle relaxant is necessary, small dose of relatively short- acting muscle relaxants is used as cisaracurium and mivacurium. Neuromuscular blockade should be monitored carefully with a nerve stimulator. Ventilatory function should be examined and evaluated before extubation. Patients with bulbar involvement may be at greatest risk of postoperative respiratory failure.

Malignant hyperthermia is the anesthesiologists' disease. It is a subclinical myopathy that is unmasked upon exposure to the potent agents or succinylcholine; then skeletal muscle acutely and unexpectedly increase its oxygen consumption and lactate production, resulting in greater heat production, respiratory and metabolic acidosis, muscle rigidity, sympathetic stimulation, and increased cellular permeability. The best- accepted theory is that MH is caused by an inability to control calcium concentrations within the muscle fiber and that it may involve a generalized alteration in cellular or subcellular membrane permeability. Diagnosis is made on the basis of extraordinary temperature and acid-base and muscle aberrations. Specific treatment is the action of dantrolene on muscle calcium movements; symptomatic treatment is by reversal of acid-base and temperature change. Evaluation of affected families is guided by measurements of circulating creatine phosphokinase

and by analysis of drug-induced contractures in muscle biopsy specimens. Either general or regional anesthesia is safe for patients susceptible to MH, provided that if a general technique is chosen, care is taken to specially prepare the anesthesia machine and to avoid all anesthetic trigger agents. The patient should be warned of the dangerous nature of the syndrome and should be advised to carry and identification band all times.

Muscular Dystrophies are a group of hereditary disorders characterized by progressive weakness and degeneration of muscle.

Duchenne's muscular dystrophy is the most common and most severe form. It is an X-linked recessive disorder, affecting males almost exclusively. Patients characteristically develop symmetric proximal muscle weakness that is manifested as a gait disturbance. Fatty infiltration causes enlargement (pseudohypertrophy) of muscles, especially the calves. Progressive weakness and contractures eventually result in kyphoscoliosis. Degeneration of respiratory muscles interferes with an effective coughing mechanism and leads to retention of secretions and frequent pulmonary infections. The combination of marked kyphoscoliosis and muscle wasting produces a severe restrictive ventilatory defect. Degeneration of cardiac muscle is common, but results in dilated or hypertrophic cardiomyopathy in only 10% of cases. Electrocardiographic abnormalities and atrial arrhythmias are common. Death is usually due to recurrent pulmonary infections, respiratory failure, or cardiac failure by the age of 15- 25 years.

Becker's Muscular Dystrophy is a less common disorder; it is also an X-linked recessive muscular dystrophy. Manifestations are virtually identical to those of Duchenne's muscular dystrophy except that they

usually present in adolescence and progress more slowly. Mental retardation is less common. Death is usually from respiratory complications. Cardiomyopathy may occur in some cases and may precede severe skeletal weakness.

Other major variants include ***Facioscapulhumeral, and Limb-girdle Dystrophies.*** Patients with these diseases generally have normal responses to anesthetic agents. Nonetheless, because of the great variability and overlap between the various forms of muscular dystrophy, nondepolarizing muscle relaxants should be used cautiously, and succinylacholine should probably be avoided.

Regardless of the type of periodic paralysis either ***voltage-sensitive sodium channelopathies or voltage- gated calcium channelopathies,*** anesthetic management of the disorder is directed toward preventing the attacks. Careful ECG monitoring is necessary to detect attacks and arrhythmias during anesthesia. Frequent intraoperative measurements of plasma potassium concentration are advisable whenever possible. Glucose-containing intravenous fluids should not be used in patients with hypokalemic paralysis, whereas such solutions may benefit patients with hyperkalemic periodic paralysis.

Neuromuscular function should be carefully monitored during general anesthesia. The response to muscle relaxants is unpredictable. Succinylacholine is contraindicated in hyperkalemic paralysis and perhaps other variants as well. Because shivering and hypothermia may trigger attacks, maintenance of core temperature intraoperatively is important.

Conclusion

So, it is now understood that muscle diseases although very rare, but it could unexpectedly surprise the anesthesiologist intraoperatively. Cardiac arrest could be the first presentation of muscle diseases which could occur following induction of anesthesia that is why we should keep those diseases always in mind. Any patient suspected to have muscle disease, either by history or clinical examination, should be well investigated prior to surgery and a safe anesthetic technique should be planned for. The nature of the disease, complications and dangerous sequel should be explained to the patient.