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NEUROMUSCULAR DISORDERS

Preoperative Evaluation

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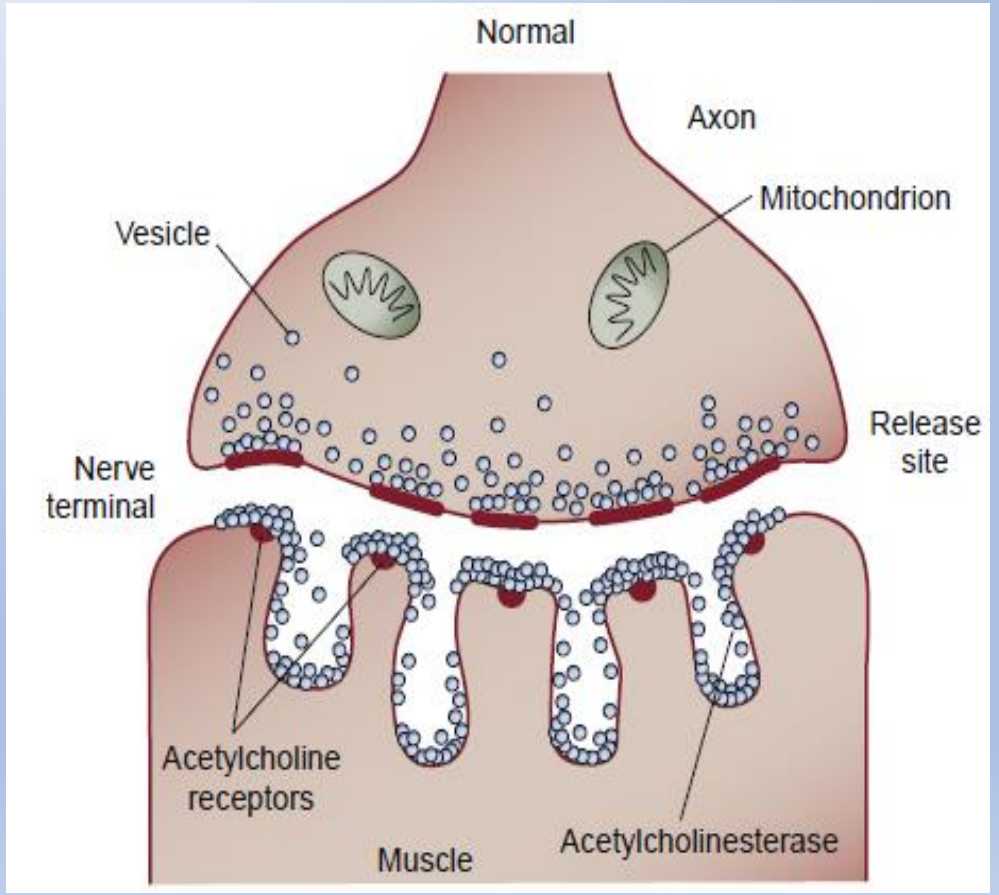

Neuromuscular disorders

- **Neuromuscular disorders** consist of conditions affecting any major component of the motor unit: motoneuron, peripheral nerve, neuromuscular junction, and muscle.
- Neuropathies may involve all components of the nerve, thereby producing sensory, motor, and autonomic dysfunction, or only one component.
- Myopathies may involve the proximal muscles, the distal muscles, or both.

Muscular Dystrophy

- Is a group of hereditary diseases ***characterized by painless degeneration and atrophy of skeletal muscles***.
- ***Progressive, symmetrical skeletal muscle weakness and wasting***
- ***No evidence of skeletal muscle denervation***
- ***Sensation and reflexes are intact***

PSEUDOHYPERTROPHIC MUSCULAR DYSTROPHY (***DUCHENNE'S MUSCULAR DYSTROPHY***)

- ***Duchenne and Becker muscular dystrophies*** are **X-linked** recessive disorders that occur primarily in male patients(2- 5 year-old boys).
- Initial ***symptoms(waddling gait, frequent falling, and difficulty climbing stairs)***due to involvement of the proximal skeletal muscle groups of the pelvic girdle.
- Affected muscles become larger as a result of fatty infiltration (*pseudohypertrophic).*

PSEUDOHYPERTROPHIC MUSCULAR DYSTROPHY (***DUCHENNE'S MUSCULAR DYSTROPHY***)

- **Kyphoscoliosis** can develop.
- Skeletal muscle atrophy can predispose to **long bone fractures**.
- **Mental retardation** is often present.
- Serum ***creatine kinase concentrations are 20 to 100 times*** normal, reflecting increased permeability of skeletal muscle membranes and skeletal muscle necrosis.
- *Death usually occurs at 15 to 25 years* of age as a result of **congestive heart failure and/or pneumonia**.

PSEUDOHYPERTROPHIC MUSCULAR DYSTROPHY (***DUCHENNE'S MUSCULAR DYSTROPHY***)

- Degeneration of cardiac muscle (***tall R waves in V_1, deep Q waves in the limb leads, a short PR interval, and sinus tachycardia***)
- ***Mitral regurgitation*** may occur as a result of papillary muscle dysfunction or decreased myocardial contractility.
- **Chronic weakness of the respiratory muscles** and a ***weakened cough*** result in ***loss of pulmonary reserve*** and ***accumulation of secretions***.
- ***Sleep apnea*** may occur and may contribute to development of ***pulmonary hypertension***.

PSEUDOHYPERTROPHIC MUSCULAR DYSTROPHY (***DUCHENNE'S MUSCULAR DYSTROPHY***)

- ***Preoperative Evaluation***
- Anesthesia for muscle biopsy or correction of orthopedic deformities.

- The focus of the **preoperative history** is on **palpitations, dyspnea, chest pain, syncope, orthopnea, dependent edema, aspiration, and pneumonia**.

PSEUDOHYPERTROPHIC MUSCULAR DYSTROPHY (***DUCHENNE'S MUSCULAR DYSTROPHY***)

- ***Preoperative Evaluation***
- The physical examination generally concentrates on the **cardiopulmonary system**, whereas ***important additional preoperative tests include an ECG, PFTs, and echocardiography.***

- ***Hypomotility of the gastrointestinal tract*** may delay gastric emptying and, in the presence of **weak laryngeal reflexes**, can increase the **risk of pulmonary aspiration**.

PSEUDOHYPERTROPHIC MUSCULAR DYSTROPHY (***DUCHENNE'S MUSCULAR DYSTROPHY***)

- ***Management of Anesthesia***
- Use of **succinylcholine is contraindicated** because of the **risk of rhabdomyolysis, hyperkalemia, and/or cardiac arrest.**

- ***The response to nondepolarizing muscle relaxants is normal.***

PSEUDOHYPERTROPHIC MUSCULAR DYSTROPHY (***DUCHENNE'S MUSCULAR DYSTROPHY***)

- ***Management of Anesthesia***
- ***Rhabdomyolysis***, with or without cardiac arrest, has been observed in association with administration of volatile anesthetics to these patients even in the absence of succinylcholine administration.
- **Dantrolene should be available**, because there is an increased incidence of malignant hyperthermia in these patients.
- Regional anesthesia avoids the unique risks of general anesthesia in these patients.

PSEUDOHYPERTROPHIC MUSCULAR DYSTROPHY (***DUCHENNE'S MUSCULAR DYSTROPHY***)

- ***Management of Anesthesia***
- **Postoperative pulmonary dysfunction** should be anticipated and attempts made **to facilitate clearance of secretions**.
- **Delayed pulmonary insufficiency** may occur up to 36 hours postoperatively .

- ***Male patients with a family history*** of either Duchenne or Becker muscular dystrophy should be considered at risk, and they require precautions similar to those in patients with diagnosed disease.

Myasthenia Gravis

- Myasthenia gravis (MG) is an autoimmune disorder : IgG antibodies against the α-subunit of the acetylcholine receptor destroy acetylcholine receptors of the neuromuscular junction .

- The severity of the disease correlates with the ability of antibodies to **decrease the number of available acetylcholine receptors.**
- The ***hallmarks of this disease is muscle weakness that worsens with activity and improves with rest.***

Myasthenia Gravis

- Skeletal muscles **(ocular, pharyngeal, and laryngeal muscles)** innervated by **cranial nerves** are especially **vulnerable**, as indicated by the appearance of **ptosis, diplopia, and dysphagia**, which are often the initial symptoms of the disease.
- **Women 20 to 30 years** of age are most often affected; **men** with myasthenia gravis are **often older than 60 years** of age when the disease presents.
- **Antibodies** are present in more than **80%** of patients with myasthenia gravis.

Myasthenia Gravis

- **Other autoimmune diseases may occur in association with myasthenia gravis.**
- **Hyperthyroidism**
- **Rheumatoid arthritis,**
- **Systemic lupus erythematosus**
- **Pernicious anemia.**
- **Polymyositis**

Myasthenia Gravis

- **Infection**, **electrolyte abnormalities**, **pregnancy**, **emotional stress**, and **surgery** ***may precipitate or exacerbate muscle weakness.***
- **Thymic hyperplasia** is present in **two thirds of patients** with myasthenia gravis, and 10% to 15% of these patients have **thymomas**.

Myasthenia Gravis

- ***TREATMENT***
- ***Anticholinesterase drugs*** are the first line of treatment for
- myasthenia gravis.(Pyridostigmin&Neostigmin)
- ***Higher dosages of pyridostigmine*** may actually induce more muscle weakness, the so-called ***cholinergic crisis.***
- ***Immunosuppressive therapy*** (corticosteroids, azathioprine, cyclosporine, mycophenolate) is indicated when skeletal muscle weakness is not adequately controlled by anticholinesterase drugs.

Myasthenia Gravis

- ***TREATMENT***
- ***Plasmapheresis*** removes antibodies from the circulation and produces short-term clinical improvement in patients who are experiencing *myasthenic crises* or are being prepared for thymectomy.
- The indications for administration of ***immunoglobulin*** are the same as for plasmapheresis. The effect is temporary
- Patients with generalized myasthenia gravis are candidates for ***thymectomy.***

Myasthenia Gravis

- ***Preoperative evaluation***
- Although controlled ventilation was frequently required for at least 24 to 48 hours postoperatively, immediate extubation has become more common.
- Therefore, it is important to advise these patients during the preoperative interview that they may be intubated and ventilated when they awaken.

Myasthenia Gravis

BOX 35.8 Risk Factors of Postoperative Ventilation for Patients with Myasthenia Gravis[396]

Vital capacity <2-2.9 L
Duration of MG >6 years
Pyridostigmine dosage >750 mg/day
History of chronic pulmonary disease
Preoperative bulbar symptoms
History of myasthenic crisis
Intraoperative blood loss >1000 mL
Serum antiacetylcholine receptor antibody >100 nmol/mL
Pronounced decremental response on low frequency repetitive nerve stimulation

Myasthenia Gravis

- ***Preoperative evaluation***
- Patients taking ***azathioprine*** require a ***CBC and LFTs*** because of drug-induced bone marrow suppression and liver dysfunction.

- Patients treated with ***steroids*** need ***glucose measurement***, as well as ***steroid supplementation*** in the perioperative period.

Myasthenia Gravis

- ***Preoperative evaluation***
- Because ventilatory function can be compromised***, preoperative PFTs*** indicated for those with severely affected ventilatory function.
- PFTs may be particularly helpful if patients are being considered for ambulatory surgery, especially in freestanding surgical centers.
- Drugs that can exacerbate myasthenic symptoms should also be avoided :
- **Medications**, especially the **(aminoglycosides, propranolol, ciprofloxacin,clindamycin),**, can aggravate the muscle weakness.

Myasthenia Gravis

- ***MANAGEMENT OF ANESTHESIA***
- Anticholinesterase drugs inhibit not only true cholinesterase but also impair plasma pseudocholinesterase activity, which introduces the possibility of a prolonged response to succinylcholine.
- All anticholinergic drugs may be withheld for 6 hours preoperatively, and medication should be reinstituted postoperatively

Myasthenia Gravis

- ***MANAGEMENT OF ANESTHESIA***
- Increased sensitivity to nondepolarizing muscle relaxants.
- The initial dose of a nondepolarizing neuromuscular blocker should be **decreased by one half to two thirds** and the response monitored using a peripheral nerve stimulator.
- Despite the increase in potency, the durationof action of intermediate-acting muscle relaxants is short enough and yet be predictably reversed at the conclusion of surgery.

Myasthenia Gravis

- ***MANAGEMENT OF ANESTHESIA***
- *There is resistance* to the effects of succinylcholine. The 95% effective dose (ED_{95}) is approximately 2.6 times higher than normal.
- Corticosteroid therapy produce resistance to the neuromuscular blocking effects of steroidal muscle relaxants such as vecuronium.

Myasthenia Gravis

- ***MANAGEMENT OF ANESTHESIA***
- The relatively short duration of action of intermediate-acting muscle relaxants is a desirable characteristic in this patient group.

- The respiratory effects of opioids, which can linger into the postoperative period, detract from their use for maintenance of anesthesia

Myasthenia Gravis

- ***MANAGEMENT OF ANESTHESIA***
- Potent volatile anesthetics have been used successfully in MG patients.

- A volatile anesthetic usually provides adequate muscular paralysis to allow most surgical procedures to be performed without the need for a neuromuscular blocking agent.

Myasthenic syndrome (Eaton-Lambert syndrome)

- Myasthenic syndrome is an acquired autoimmune disease.

- Caused by antibodies against voltage-gated calcium channels that result in a decrease in acetylcholine release.
- Lambert-Eaton syndrome is not associated with thymic abnormalities, but it commonly occurs in patients with malignant diseases, especially ***small cell lung cancer and gastrointestinal tumors.***
- The muscle weakness classically ***improves with activity and is worse after inactivity.***

Myasthenic syndrome (Eaton-Lambert syndrome)

- Patients with ELMS have muscle weakness and fatigability, generally of the proximal limb muscles, with the lower extremities affected more often than the extraocular and bulbar muscle groups.

- Unlike MG, patients with ELMS are usually worse in the morning with gradual improvement throughout the day.

Myasthenic syndrome (Eaton-Lambert syndrome)

- Anticholinesterase drugs effective in the treatment of myasthenia gravis do *not* produce an improvement in patients with myasthenic syndrome.
- a selective potassium channel blocker, namely 3,4 diaminopyridine, is used and should be continued perioperatively..
- Immunoglobulin also increases muscle strength temporarily (for 6 to 8 weeks).

Myasthenic syndrome (Eaton-Lambert syndrome)

TABLE 21-5 Comparison of myasthenic syndrome and myasthenia gravis

Characteristic	Myasthenic syndrome	Myasthenia gravis
Manifestations	Proximal limb weakness (legs more than arms), exercise improves strength, muscle pain common, reflexes absent or decreased	Extraocular, bulbar, and facial muscle weakness; exercise causes fatigue; muscle pain uncommon; reflexes normal
Gender	Affects males more often than females	Affects females more often than males
Co-existing pathologic conditions	Small cell lung cancer	Thymoma
Response to muscle relaxants	Sensitive to succinylcholine and nondepolarizing muscle relaxants	Resistant to succinylcholine, sensitive to nondepolarizing muscle relaxants
	Poor response to anticholinesterases	Good response to anticholine-sterases

Myasthenic syndrome (Eaton-Lambert syndrome)

- ***MANAGEMENT OF ANESTHESIA***
- Patients with myasthenic syndrome **are sensitive to the effects of both depolarizing and nondepolarizing muscle relaxants**.
- Antagonism of neuromuscular blockade with anticholinesterase drugs may be inadequate.
- The potential presence of myasthenic syndrome and the need to decrease doses of muscle relaxants should be considered in patients undergoing bronchoscopy, mediastinoscopy, or thoracoscopy for suspected lung cancer.

Myasthenic syndrome (Eaton-Lambert syndrome)

- ***MANAGEMENT OF ANESTHESIA***

- As in patients with MG, those with ELMS should be carefully evaluated for the risk of postoperative respiratory failure and the need for prolonged respiratory monitoring in the postoperative period.

Periodic Paralysis

- Periodic paralysis is a spectrum of diseases characterized by intermittent acute attacks of skeletal muscle weakness or paralysis and associated with hypokalemia or hyperkalemia.
- The hyperkalemic form is much rarer than the hypokalemic form.
- Attacks generally last for a few hours but may persist for days. Muscle strength is normal between attacks.

Periodic Paralysis

TABLE 21-3 Clinical features of familial periodic paralysis

Type	Serum potassium concentration during symptoms (mEq/L)	Precipitating factors	Other features
Hypokalemic	<3.0	High-carbohydrate meal, strenuous exercise, glucose infusion, stress, menstruation, pregnancy, anesthesia, hypothermia	Cardiac dysrhythmias, electrocardiographic signs of hypokalemia
Hyperkalemic	>5.5	Exercise, potassium infusion, metabolic acidosis, hypothermia	Skeletal muscle weakness may be localized to tongue and eyelids

Periodic Paralysis

- ***MANAGEMENT OF ANESTHESIA***
- A principal goal of anesthetic management is avoidance of any events that can precipitate skeletal muscle weakness.
- Hypothermia must be avoided.
- It may be necessary to maintain normothermia during cardiopulmonary bypass.
- Nondepolarizing muscle relaxants can be safely administered.

Periodic Paralysis

- ***MANAGEMENT OF ANESTHESIA***
- ***Hypokalemic Periodic Paralysis***
- Maintenance of **carbohydrate balance**, correction of electrolyte abnormalities, and avoidance of events known to trigger hypokalemic attacks (psychologic stress, cold, carbohydrate loads).
- High-carbohydrate meals can trigger hypokalemic episodes and should be avoided during the 24 hours preceding surgery. Glucose-containing solutions and drugs known to cause intracellular shifts of potassium, such as β-adrenergic agonists, must also be avoided.
- ***Mannitol*** can be administered in lieu of a potassium-wasting diuretic should the operative procedure require diuresis.

Periodic Paralysis

- ***MANAGEMENT OF ANESTHESIA***
- ***Hypokalemic Periodic Paralysis***
- Frequent perioperative monitoring of serum potassium concentration (every 30 to 60 minutes) is useful, and aggressive intervention to increase the serum potassium concentration (infusion of potassium chloride at a rate of up to 40 mEq/hr) may occasionally be needed.
- Hypokalemia may precede the onset of muscle weakness by several hours, so timely potassium supplementation may help to avoid muscle weakness.
- Short-acting neuromuscular blockers are preferable if skeletal muscle relaxation is required for the surgery.
- Succinylcholine with its ability to increase serum potassium concentration transiently is acceptable in these patients.
- Regional anesthesia has been safely used.

Periodic Paralysis

- ***MANAGEMENT OF ANESTHESIA***
- ***Hyperkalemic Periodic Paralysis***
- Management of anesthesia in patients with hyperkalemic periodic paralysis includes:
- Preoperative potassium depletion with diuretics,
- Prevention of carbohydrate depletion by administration of glucose-containing solutions,
- Avoidance of potassium-containing solutions and potassium-releasing drugs such as succinylcholine.
- Frequent monitoring of serum potassium concentration is indicated, as is the ready availability of calcium for intravenous administration should signs of hyperkalemia appear on the electrocardiogram.


Thank you for your considerations