NEUROMUSCULAR DISORDERS & ANESTHETIC IMPLICATIONS PART I

MSNA534: Principles of Nurse Anesthesia for Patients with Co-existing Diseases

Kim Kujanek, Amber Kursch, & Hilary Pekarek
Student Registered Nurse Anesthetists
Adventist University of Health Sciences
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OBJECTIVES

- Describe the basic pathophysiology, diagnosis, treatment, and anesthetic implications of 24 neuromuscular disorders
- Increase the anesthesia provider's awareness and knowledge of neuromuscular diseases
- Present the most recent data related to neuromuscular diseases thereby allowing the opportunity for anesthesia providers to treat patients with current, evidence based research findings

NEUROMUSCULAR DISORDERS PART I

- Autoimmune
  1. Myasthenia Gravis (MG)
  2. Lambert-Eaton Myasthenic Syndrome (LEMS)
  3. Multiple Sclerosis (MS)
  4. Guillain-Barré Syndrome (GBS)
  5. Systemic Lupus Erythematous (SLE)*
  6. Scleroderma*
- Connective Tissue Disorders
  1. Systemic Lupus Erythematous (SLE)*
  2. Scleroderma*
  3. Marfan's Syndrome
  4. Ehlers-Danlos Syndrome (EDS)
- Autosomal Dominant Disorders
  1. Huntington's Chorea Disease
  2. Amyotrophic Lateral Sclerosis (ALS)
  3. Neurofibromatosis (NF)
  4. Myotonic Dystrophica
  5. Malignant Hyperthermia*

NEUROMUSCULAR AUTOIMMUNE DISORDERS

1. Myasthenia Gravis (MG)
2. Lambert-Eaton Myasthenic Syndrome (LEMS)
3. Multiple Sclerosis (MS)
4. Guillain-Barré Syndrome (GBS)
5. Systemic Lupus Erythematous (SLE)*
6. Scleroderma*

- ** Note: These disorders are BOTH autoimmune & connective tissue disorders*

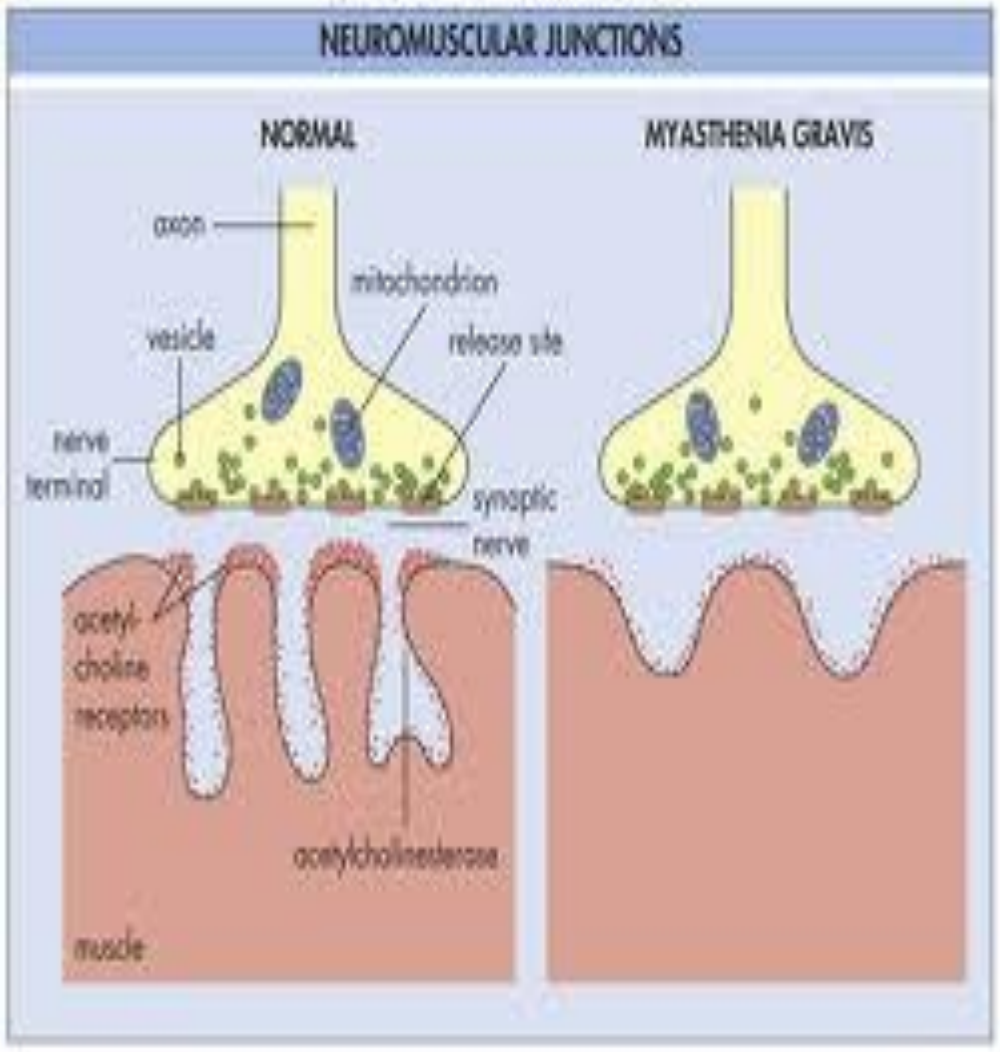
MYASTHENIA GRAVIS (MG)

MYASTHENIA GRAVIS (MG)

- MG is an AUTOIMMUNE disease
- Antibodies act against ACh receptors at the NMJ
- Resulting weakness and easily fatigued muscles
- Affects 1/20,000; Women > Men
- Classified into groups by severity and muscle groups involved:
  - **Group I**= Ocular muscles only (20% of MG patients)
  - **Group II, A**= Mild, generalized symptoms, usually EXCLUDING respiratory muscles
  - **Group II, B**= More severe, rapid progression of symptoms, MAY involved respiratory muscles
  - **Group III**= RAPID onset and progression, HIGH MORTALITY rate
  - **Group IV**= Severe form that's usually progression of IIA and IIB

MG PATHOPHYSIOLOGY

- T- helper cell activation in thymus leads to antibody formation
- Antibodies damage postsynaptic membrane via complement-mediated reaction (Type II Hypersensitivity) with ↕degradation and formation of ACh receptors
- Often associated with other autoimmune diseases
- May be *exacerbated* with pregnancy, viral infections, stress, **SURGERY, HYPERthermia**



MG DIAGNOSIS & TREATMENT

DIAGNOSIS

- Electromyography (EMG)
- Edrophonium test
- Presence of anti-ACh receptor antibodies

TREATMENT

- Cholinesterase inhibitors (aka anticholinesterase drugs) to ↑concentration of ACh at nicotinic postsynaptic membrane
- Thymectomy
- Immunosuppressants
- Corticosteroids
- Plasmapheresis (separation of blood cells and plasma; blood cells are returned to patient, plasma is not in order to ↓antibodies in circulation)

MG ANESTHETIC IMPLICATIONS

- Thorough **preoperative evaluation** to assess severity of disease and current treatment regimen
  - Current respiratory status
  - Presence of thymoma that could lead to anterior airway obstruction (awake intubation?)
  - Bulbar dysfunction= impairment of CN IV, V, XI, XII: patients are high risk for aspiration so premedicate with H2 blocker, PPI, etc.; RSI?
  - Steroid therapy
  - Patients with severe disease may continue Mestinon the day of surgery

MG ANESTHETIC IMPLICATIONS

- Intubation **WITHOUT** muscle relaxation is optimal:
  - *Patients are very SENSITIVE to NDMRs, usually requiring 10% of normal dose if absolutely necessary*
  - *Patients are usually RESISTANT to succinylcholine and will require two to three times the normal dose*
  - *Patients will have PROLONGED effect of succinylcholine if they are TREATED with anticholinergic agents as these drugs inhibit pseudocholinesterase*
  - *Volatile agents have muscle-relaxing properties that are exaggerated in MG patients so this alone may be sufficient for relaxation*
  - *REGIONAL ANESTHESIA is acceptable, but techniques that block accessory muscles and rib cage muscles will* ↑ respiratory dysfunction

MG ANESTHETIC IMPLICATIONS

- EXTUBATION CRITERIA:
  - Fully awake with complete reversal of muscular blockade (assess for potential cholinergic crisis vs. muscular weakness)
  - VC > 15ml/kg
  - A-a gradient O2 >350mmHg on 100% FIO2
  - PaCO2 <50mmHg
  - Negative inspiratory force (NIF) > -20cmH20
  - Stable hemodynamics
  - Normothermia

LAMBERT-EATON MYASTHENIC SYNDROME (LEMS)

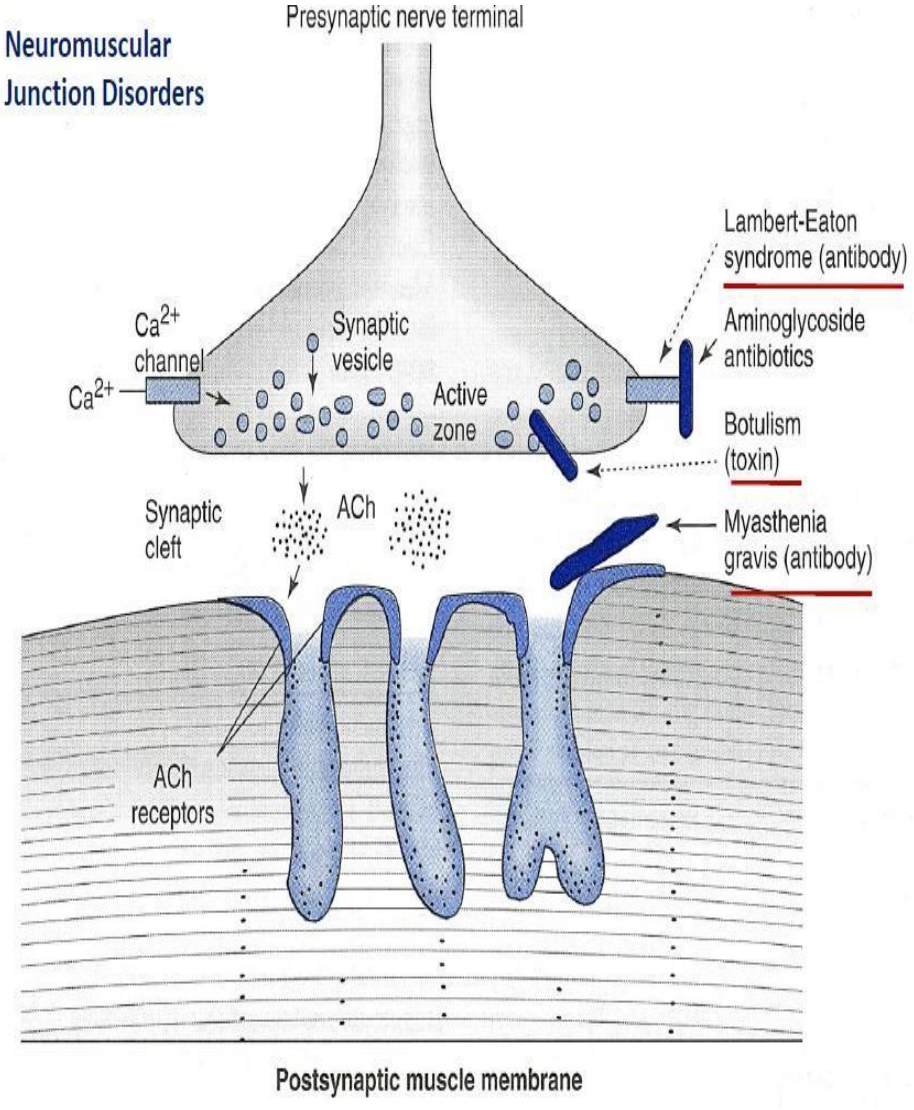
LAMBERT-EATON MYASTHENIC SYNDROME (LEMS)

- Paraneoplastic syndrome of neuromuscular transmission that resembles myasthenia gravis
- Acquired autoimmune disease resulting in skeletal muscle weakness
- May be associated with small cell carcinoma of lung
- Presence of immunoglobulin G antibodies to **voltage-sensitive calcium channels** that causes a deficiency of these channels at the **PRESYNAPTIC** motor nerve terminal
- Deficiency restricts calcium entry when the terminal is depolarized
- Anticholinesterases are NOT effective in the treatment of this syndrome
- 3,4-diaminopyridine does improve muscle strength (NOT available in the US)
- Immunoglobulin will improve muscle strength temporarily

LEMS ANESTHETIC IMPLICATIONS

- Sensitive to effects of BOTH succinylcholine and NDMR
- Reversal may be inadequate
- Potential presence of LEMS needs to be considered in patients undergoing bronchoscopy, mediastinoscopy, and thoracoscopy for suspected lung cancer

https://lookfordiagnosis.com/mesh_info.php?term=Lambert-Eaton+Myasthenic+Syndrome&lang=1



COMPARISON OF LEMS & MG

Characteristic	LEMS	MG
Manifestations	Proximal limb weakness (legs > arms), exercise improves strength, muscle pain common, reflexes absent or ↓	Extraocular, bulbar, & facial muscle weakness; exercise causes fatigue; muscle pain uncommon; reflexes normal
Gender	Males > Women	Women > Men
Co-existing pathologic conditions	Small cell lung cancer	Thymoma
Response to muscle relaxants	SENSITIVE to succs. & NDMR; poor response to anticholinesterases	RESISTANT to succs.; SENSITIVE to NDMR; good response to anticholinesterases

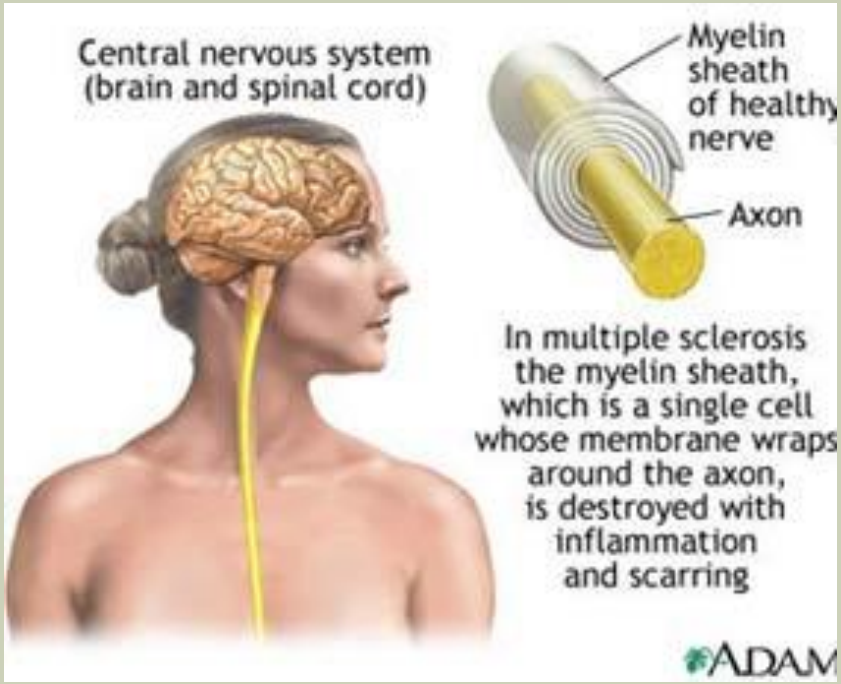
*Modified from Stoelting's Anesthesia and Co-existing Disease (6th edition), Table 21-5

MULTIPLE SCLEROSIS

MULTIPLE SCLEROSIS

- Autoimmune disease affecting CNS
  - *Peripheral nerves are NOT affected by MS*
  - Unpredictable course with exacerbations over several years
- Exact etiology= unknown
- Women 2x > Men
- Onset: Age 20 & 40
- High rate concordance among twins
- ↑ risk if 1st degree relative has disease
- More common in northern Europe, southern Australia & United States

MULTIPLE SCLEROSIS PATHOPHYSIOLOGY



- **Inflammation & demyelination in the brain & spinal cord**
  - Environmental stimulus activates T cells
  - Activated T cells cross the BBB➔ initiates inflammatory & immunologic attack on myelin
  - Demyelination causes CNS dysfunction
  - # of oligodendrocytes is ↓/absent & followed by formation of demyelinative plaques
- Early phases: neural tissue can repair itself explaining the relapsing nature of the disease

MULTIPLE SCLEROSIS CLASSIFICATION

- Classified in early phases
  - Relapsing-remitting (85%)
    - Many develop neurodegeneration & are categorized as secondary progressive MS
  - Primary progressive MS
    - Devoid of acute episodes
    - Develop progressive neurodegeneration

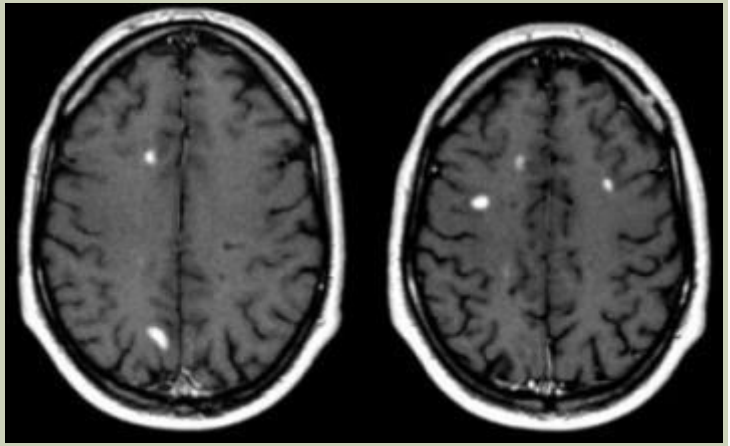
MULTIPLE SCLEROSIS SIGNS & SYMPTOMS

- Presenting symptoms depend on sites of demyelination
  - Brainstem
    - Nystagmus, diplopia, trigeminal neuralgia, autonomic dysfunction, & alterations in ventilation
    - **Can have pharyngeal or laryngeal dysfunction, ↑ risk aspiration**
  - Lesions of the spinal cord
    - Weakness & paresthesias
    - Legs affected more than arms
    - Bowel retention & urinary incontinence

- Unique Facts:
  - **Pregnancy is associated w/ improvement in symptoms**
  - Relapse often occurs w/in initial 3 months postpartum

MULTIPLE SCLEROSIS DIAGNOSIS

- S/S of CNS white matter disease
  - Numbness, tingling & swelling of limbs , visual loss, diplopia, spacticities & contractures, orthostatic hypotension, and bowel & bladder dysfunction
- ≥2 attacks separated by a month or more
- Involvement of ≥2 non-contiguous anatomic areas
- CSF analysis:
  - Elevated IgG & albumin
- MRI= sensitive diagnostic tool
  - Ability to locate demyelinated plaques in CNS



MULTIPLE SCLEROSIS TREATMENT

- NO CURE
- Goal: Modulate immunologic & inflammatory responses that damage the CNS
  - **Interferon preparations**
  - **Glatiramer**
  - **Mitoxantrone**
  - **Monoclonal antibodies**
    - Natalizumab
  - **Oral agents:**
    - Cladribine, fingolimod, laquinimod, teriflunomide, & dimethyl fumarate
  - *Corticosteroids (for acute relapses not responding to other medications)*
- Symptomatic therapy may include:
  - For spasticity: **Diazepam, Dantrolene, Baclofen**
  - For painful dysesthesias, tonic seizures, & ataxia: **Carbamezapine**
    - ***Note: MS is associated w/ ↑ incidence of seizure disorders***
- Nonspecific measures: Avoid excessive fatigue, emotional stress, & hyperthermia
  - "Demyelinated fibers are extremely sensitive to increases in temperature and hyperthermia can block impulse conduction." (Barash, 2014, p.619)

MULTIPLE SCLEROSIS ANESTHETIC IMPLICATIONS

- Debatable if regional or general anesthesia cause exacerbation
  - Increased exacerbation linked to: infection, hyperpyrexia, & emotional stress
  - GA= most frequently used technique for patients w/ MS
- Advisable to inform patient that surgery and anesthesia *could* lead to relapse
- **Regional Anesthesia:**
  - **Spinal anesthesia is associated with relapse of MS**
  - **Epidural analgesia CAN be safely administered to parturients**
- **Avoid hyperthermia & slight elevations in body temperature**
  - **Even a 1°C increase can cause exacerbation**
- Spasticity and possible contractures may make positioning of the patient difficult

MULTIPLE SCLEROSIS ANESTHETIC IMPLICATIONS

- Aspiration Precautions: If pharyngeal or laryngeal dysfunction
- Autonomic dysfunction can enhance hypotensive effects of VA
  - Invasive hemodynamic monitoring might also be necessary if there are signs of autonomic dysfunction
- NDNMB
  - Baclofen & Dantrolene: increases sensitivity to NDNMB
  - Anticonvulsants: Produces resistance to NDNMB
- AVOID SUCCINYLCHOLINE: Could produce exaggerated release of K+
- *Consider stress dose steroids intra-operatively to prevent adrenal insufficiency (if on corticosteroids)*
- ↑ likelihood of respiratory support immediately postoperatively 2° respiratory muscle weakness & respiratory control dysfunction

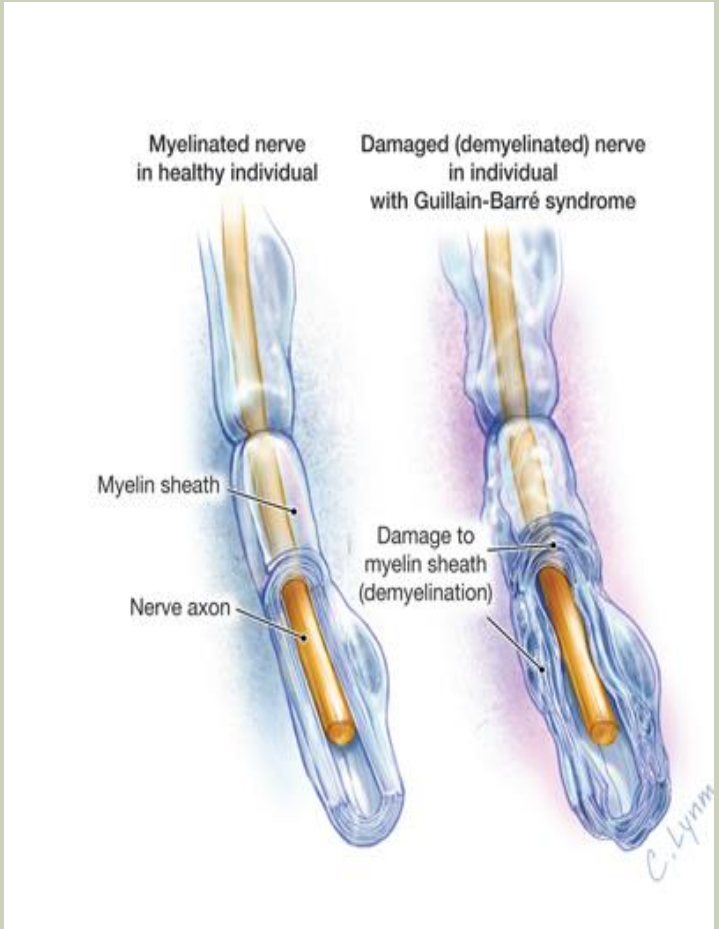
GUILLAIN-BARRÉ SYNDROME

GUILLAIN-BARRÉ SYNDROME

- AKA“Polyradiculoneuritis” or “Acute Idiopathic Polyneuritis”
  - AUTOIMMUNE DISEASE
    - Inflammatory neuropathy
  - A disease of the peripheral nerves which involves a cell-mediated response
  - Usually have a history of a respiratory or gastrointestinal infection within 4 weeks onset of neurological symptoms

GUILLAIN-BARRÉ SYNDROME PATHOPHYSIOLOGY

- Sporadic number of GBS or GBS-like cases reported postoperatively
- **Usually triggered by viral or bacterial infection**
- Infectious agent produces substance that causes immune reaction
  - This substance resembles neural component of the host
  - Autoantibodies develop that attack the host
    - Axons are primarily injured
    - Nerve fibers are cut/crushed
- Develop antibodies to gangliosides in the peripheral nerves

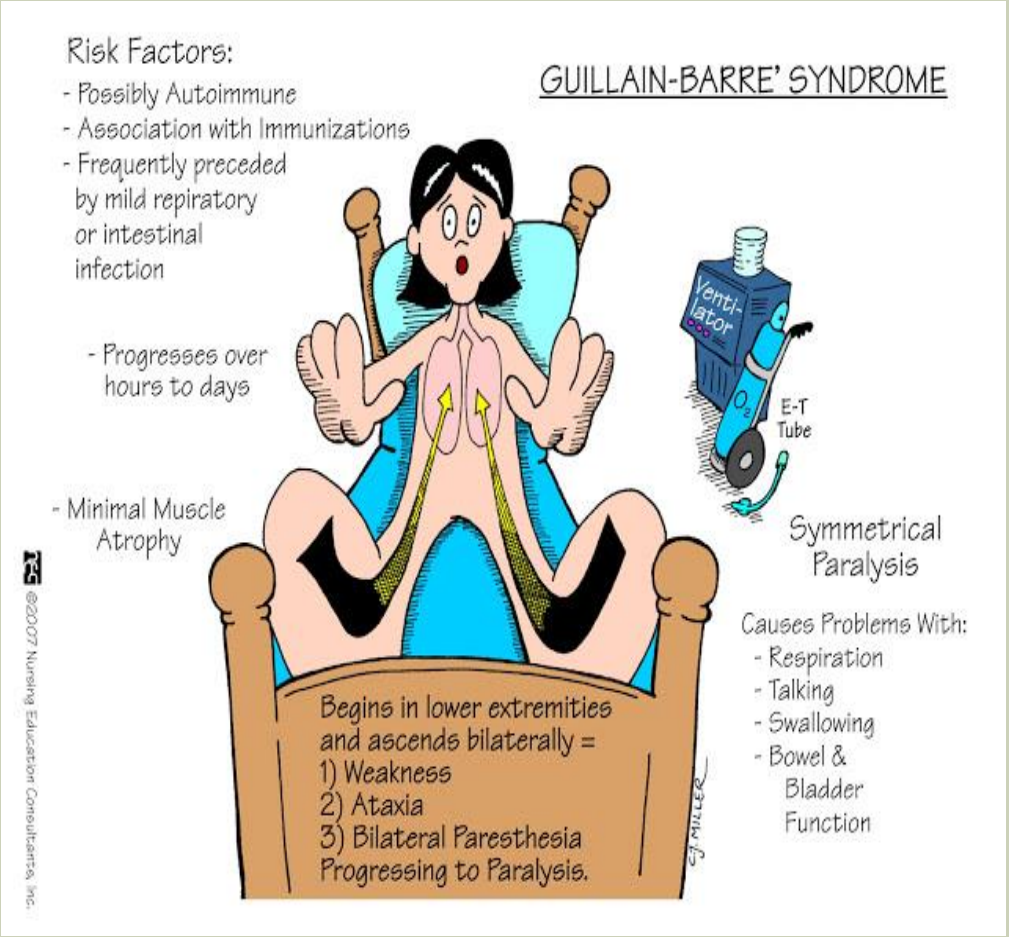


GUILLAIN-BARRÉ SYNDROME SIGNS & SYMPTOMS

- Characterized by onset of ***symmetrical*** skeletal muscle weakness in legs
  - Paresthesias may precede this
  - Paralysis progresses cephalad
    - Eventually involving muscles of trunk & arms
- Maximal weakness: 2-4 weeks after onset
- Plateau phase develops prior to recovery phase
- Autonomic nervous system dysfunction→ wide fluctuations in CV parameters
- Like autonomic hyperreflexia, physical stimulation can→ hypertension, tachycardia, & cardiac dysrhythmias

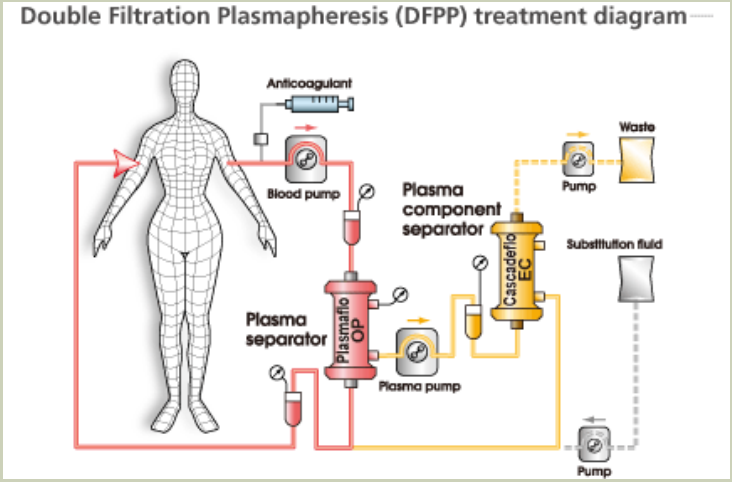
GUILLAIN-BARRÉ SYNDROME DIAGNOSIS

- ↑ protein concentration in the CSF
- Normal CSF cell count
- Essential criteria for diagnosis
  - Progressive bilateral weakness of limbs 2° neuropathy
  - Duration of progression of weakness < 4 weeks
  - Areflexia



GUILLAIN-BARRÉ SYNDROME TREATMENT

- Ventilatory support
  - **Most serious problem: ventilatory insufficiency**
  - 25% of BGS patients will require ventilatory support
    - Intubation
    - Tracheostomy
    - **Initiate when VC < 15ml/kg** (Hines & Marschall, 2012, p.270)
- Plasmapheresis
- Intravenous Immunoglobulin
- *Note: Corticosteroids NOT useful*
- Recovery
  - 85% achieve good recovery
  - 3-5% develop chronic, recurrent neuropathy



GUILLAIN-BARRÉ SYNDROME ANESTHETIC IMPLICATIONS

- Autonomic nervous system dysfunction
  - Unpredictable changes in BP, therefore ABP monitoring prudent
    - → Anticipate hypotension 2° postural changes, blood loss, or positive pressure ventilation
    - → Anticipate exaggerated increases in HR & BP with noxious stimuli (i.e. laryngoscopy & tracheal intubation)
    - May have exaggerated responses to indirect-acting vasopressors

- Regional Anesthesia
  - Controversial
  - Patients with pronounced sensory disturbances may benefit from neuraxial opioids
  - Has been used successfully in parturients

GUILLAIN-BARRÉ SYNDROME ANESTHETIC IMPLICATIONS

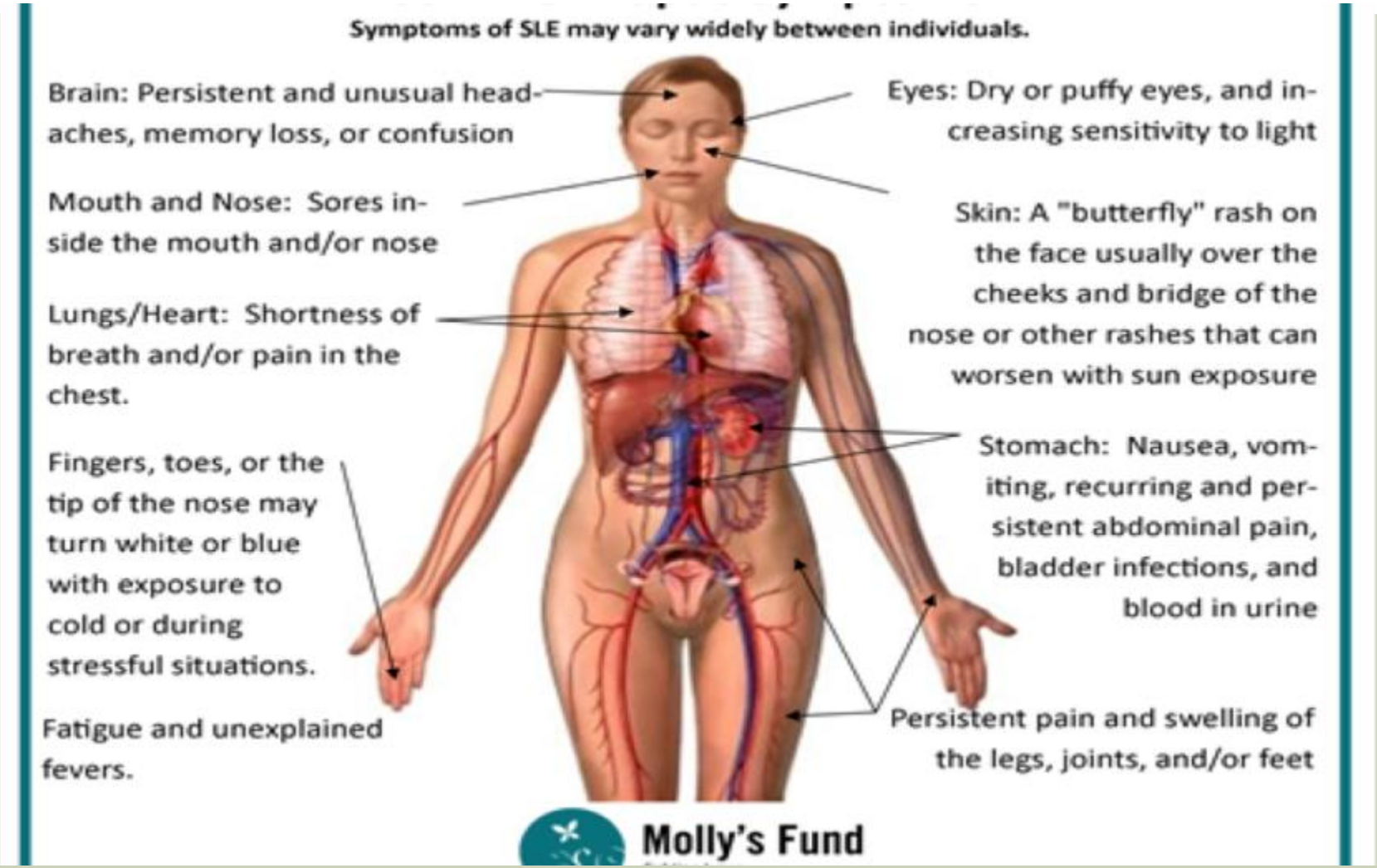
- **AVOID SUCCINYLCHOLINE**
  - Risk may persist even after clinical recovery from GBS
- **Caution with NDNMB**
  - Short acting NDNMB w/ minimal CV effects useful i.e. Cisatracurium or Rocuronium
  - **↑ SENSITIVITY**
    - Can vary from extreme sensitivity to resistance (depends on phase of disease)
- Mechanical ventilation may be required immediately postop

SYSTEMIC LUPUS ERYTHEMATOUS (SLE)

SYSTEMIC LUPUS ERYTHEMATOUS (SLE)

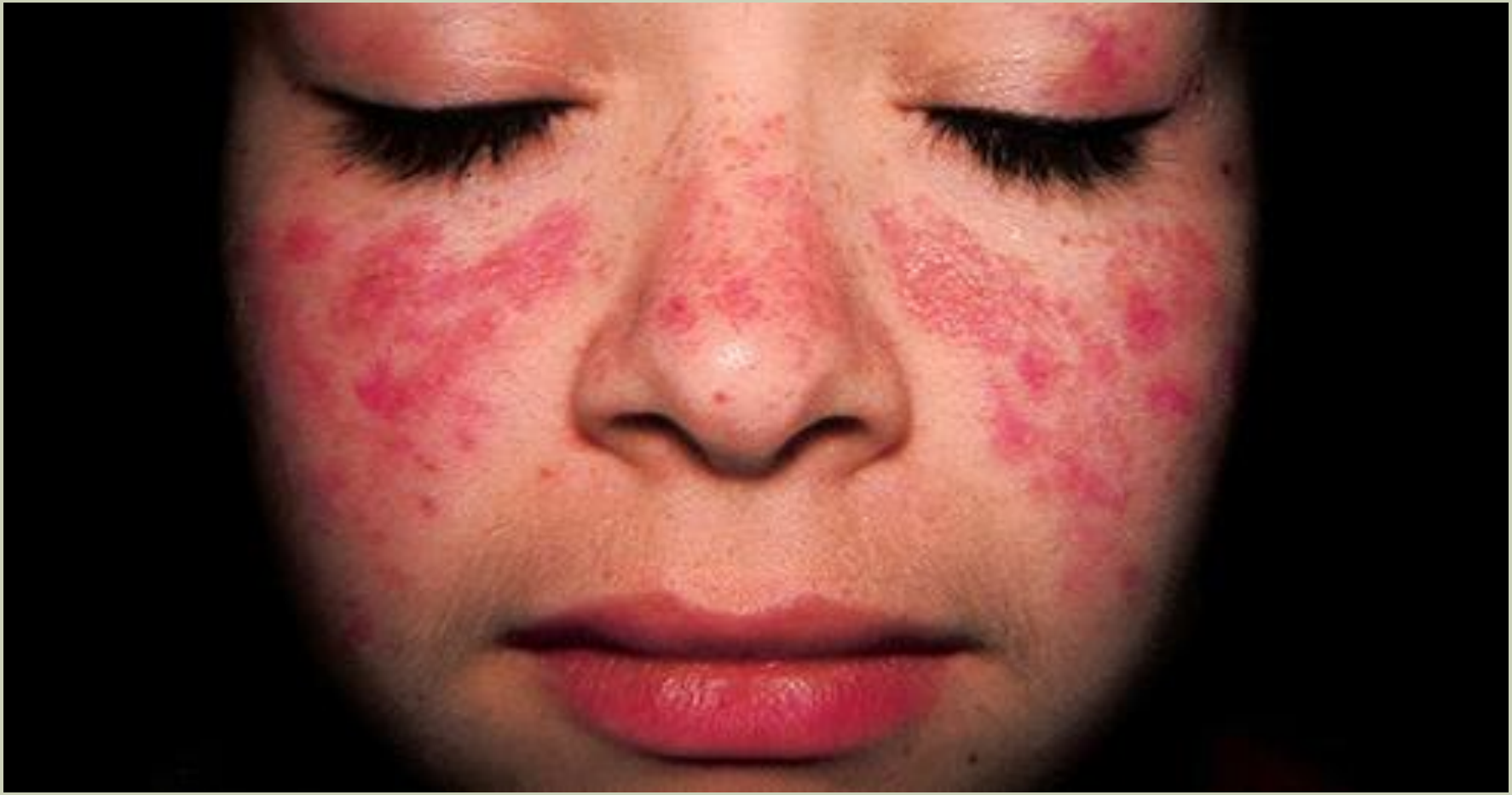
- Autoimmune disease of unclear etiology- thought to be r/t lymphocyte dysfxn. and immune complex formation
  - Antibodies to native double-stranded DNA are highly diagnostic for SLE
- Most common in African American women of childbearing age
- Stressors such as SURGERY, pregnancy, and/ or sepsis can cause exacerbations
- Drug-induced lupus (slower progression) caused by procainamide, hydralazine, & quinidine

SLE PATHOPHYSIOLOGY



http://www.mollysfund.org/2013/01/lupus-symptoms-and-signs/

SLE PATHOPHYSIOLOGY



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SLE PATHOPHYSIOLOGY

- Cardiac involvement includes:
  - Pericarditis
  - Myocarditis
  - Libman-Sacks Endocarditis (noninfectious)
    - Involves aortic and mitral valves, but usually without significant valvular dysfxn
  - Fetal congenital heart block in children of women with SLE
  - Predisposition to thromboembolic disease
- Pleuritic involvement may cause restrictive pulmonary pattern
  - Lupus pneumonia- dry cough, pleural effusions
- ↑ risk for pulmonary embolism due to hypercoagulable state
- Diffuse proliferative glomerulonephritis

SLE PATHOPHYSIOLOGY

- Autoimmune hemolytic anemia
- Acute immune thrombocytopenia & rare thrombotic thrombocytopenic purpura
- Leukopenia → ↑ risk infection
- Splenomegaly
- Antiphospholipid antibodies → hypercoagulable state → spontaneous abortion
- Cerebral “vasculitis” leading to mood disturbances, psychosis, dementia, and/ or seizures
- Neuropathy involving cranial and peripheral nerves

SLE DIAGNOSIS AND TREATMENT

- Diagnosis:
  - Detection of antinuclear antibodies is selective screening test
  - Diagnosis is likely if 3 out of the 5 manifestations are present:
    1. Antinuclear antibodies
    2. Characteristic rash
    3. Thrombocytopenia
    4. Serositis
    5. Nephritis

- Treatment:
  - ASA
  - NSAIDs
  - Antimalarial drugs (hydroxychloroquine and quinacrine)
  - Corticosteroid therapy
  - Danazol
  - Vincristine
  - Cyclophosphamide
  - Splenectomy
  - Plasmapheresis
  - Methotrexate
  - Azathioprine
  - Mycophenolate mofetil

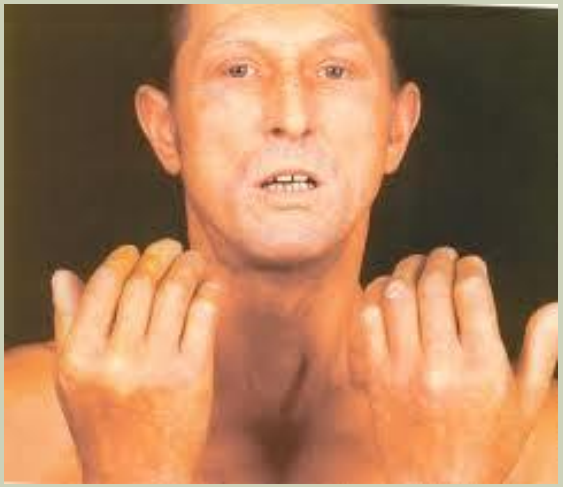
SLE ANESTHETIC IMPLICATIONS

- Thorough preoperative assessment to evaluate status of disease process
- Antibiotic prophylaxis for bacterial endocarditis
- Consider stress dose steroids
- Careful fluid replacement and medication selection
- Consider DVT prophylaxis
- Laryngeal involvement with mucosal ulceration, cricoarytenoid arthritis, and RLN palsy, may be present in 1/3 SLE patients

SCLERODERMA

SCLERODERMA

- AUTOIMMUNE, Multisystem disease, widespread symmetric, leathery induration of skin
- Systems affected: muscles, bones, heart, lungs, & intestinal tract
- Leads to organ fibrosis
- Presents between 20-40 years of age
- Women > Men



- ***Pregnancy can accelerate progression of disease:*** *Spontaneous abortion, premature labor in parturients with rapidly progressing scleroderma*

SCLERODERMA

- Begins with injury to vascular endothelial cells and cell death
- ➔Leakage of serum proteins into the interstitial space
- ➔Tissue edema and lymphatic obstruction ➔ fibrosis and atrophy of organs
- Poor prognosis linked to degree of organ involvement
- **Leading cause of death: Acute Renal Failure**

Often associated with CREST. This variant is usually less progressive & benign.

SCLERODERMA DIAGNOSIS

Based on symptoms:

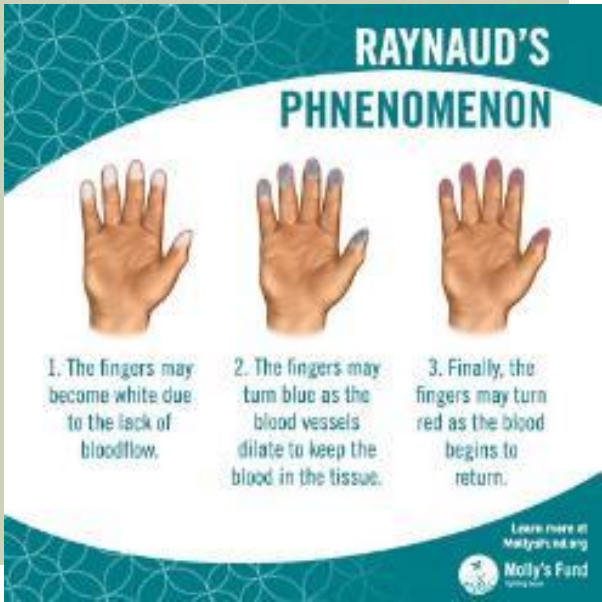
- Skin
  - Mildly thick, non-pitting edema ➔fibrotic & taut
  - Decreased mobility & restricted joint motion ➔ flexion contractures, particularly of fingers
- Musculoskeletal
  - Myopathy manifesting as weakness, especially of proximal skeletal muscles
  - Plasma creatine kinase usually ↑
  - Limited joint movement due to tautness & thickening of skin
  - Possible mild inflammatory arthritis
  - Avascular necrosis of the femoral head possible
- Nervous
  - Thickened nerve sheath ➔ nerve compression
  - Peripheral or cranial nerve neuropathy
  - Facial pain mimics trigeminal neuralgia
  - Keratoconjunctivitis sicca➔ corneal abrasions
- Cardiovascular
  - Sclerosis of small coronary arteries conduction system
  - Fibrosis of cardiac muscle
  - Pulmonary hypertension
  - Conduction delays, dysrhythmias, and heart failure
  - 2D Echo: ↓ EF or impaired diastolic fx
  - Pericarditis and pericardial effusion

SCLERODERMA DIAGNOSIS

- Pulmonary
  - Fibrosis of pulmonary arteries → cor pulmonale
  - Interstitial fibrosis in 80% of pts
  - Impaired diffusion capacity
  - Arterial hypoxemia
  - ↓ pulmonary compliance
  - Chronic aspiration pneumonitis → restrictive lung disease → M&M
- Renal
  - Renal artery stenosis & ↓ RBF
  - Systemic hypertension
  - Scleroderma renal crisis (rare)
    - Malignant hypertension, Retinopathy, & Irreversible renal failure
- Vascular
  - Peripheral vascular changes
  - **85% of patients develop Raynaud's phenomenon**
    - **Often initial presenting symptom**
  - Oral & nasal telangiestasias
- Gastrointestinal
  - Dysphagia
  - Xerostomia (dry mouth)
  - Hypomotility of lower esophagus, small intestine & colon → Pseudo-obstruction
  - Reduced lower esophageal sphincter tone → Frequent episodes of reflux
  - Malabsorption syndrome
  - Vitamin K deficiency → coagulation disorder.

SCLERODERMA TREATMENT

- Therapy is organ specific targeting several pathways
  - Side effects limit use of corticosteroids
  - Cyclophosphamide: greatest therapeutic effect
  - Endothelin receptor blockers, prostacyclin agonists; cytokine inhibitors, and stem cell transplantation
- Anesthesia often consulted for treatment of Raynaud's



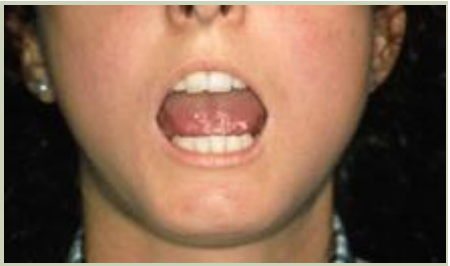
SCLERODERMA ANESTHETIC IMPLICATIONS

- **Preoperative**
  - No specific contraindications to any type of anesthesia
  - Need thorough preoperative evaluation
  - Assess coexisting organ dysfunction
  - Pre-op evaluation should guide anesthetic plan

- **Aspiration Precautions**
  - Pretreat with antacids or H2 receptor antagonists prior to induction
    - Goal: ↑ gastric pH & ↓ risk for aspiration pneumonitis.

- **Choice of Anesthetic Technique**
  - Consider MAC or regional anesthesia
  - Responses to local anesthetics may be prolonged with regional
  - Catheter placement may be challenging with spinal or epidural

SCLERODERMA
ANESTHETIC IMPLICATIONS



Limited mouth opening & decreased tongue mobility in a pt w/ Scleroderma

- **Airway**
  - Endotracheal intubation can be challenging
  - Consider awake FOI
  - Tracheostomy in severely affected patients
  - **Avoid nasotracheal intubation → risk for severe nasal hemorrhage**
  - **RSI with cricoid pressure ↓ risk for aspiration**

- **Ventilation**
  - Higher airway pressures for adequate ventilation
  - Moderate tidal volumes & low PEEP
  - Supplemental O_2 2° impaired diffusion & hypoxemia
  - Avoid ↑ in PVR by preventing respiratory acidosis & arterial hypoxemia

SCLERODERMA ANESTHETIC IMPLICATIONS

- **Pharmacodynamics**
  - Cautious administration of opioids
    - Sensitive to respiratory depressant effects
  - Caution w/ anesthetic drugs reliant upon renal elimination
  - ***Caution w/ use of muscle relaxants (↑ SENSITIVITY 2° to myopathies)***

- **Monitoring**
  - Invasive cardiovascular monitoring
    - Indicated w/ compromised myocardial function & coronary arteriosclerosis
  - Anticipate exaggerated hypotension during induction of anesthesia
  - Use etomidate for patient w/ compromised cardiac status
  - Eyes sufficiently protected to prevent corneal abrasions
  - Consider central venous catheter (IV access can be challenging)
  - Use caution if considering A-Line (2° Raynaud's Phenomenon)

NEUROMUSCULAR CONNECTIVE TISSUE DISORDERS

1. Systemic Lupus Erythematous (SLE)*
2. Scleroderma*
3. Marfan's Syndrome
4. Ehlers-Danlos Syndrome (EDS)

- **Discussed in previous slides*

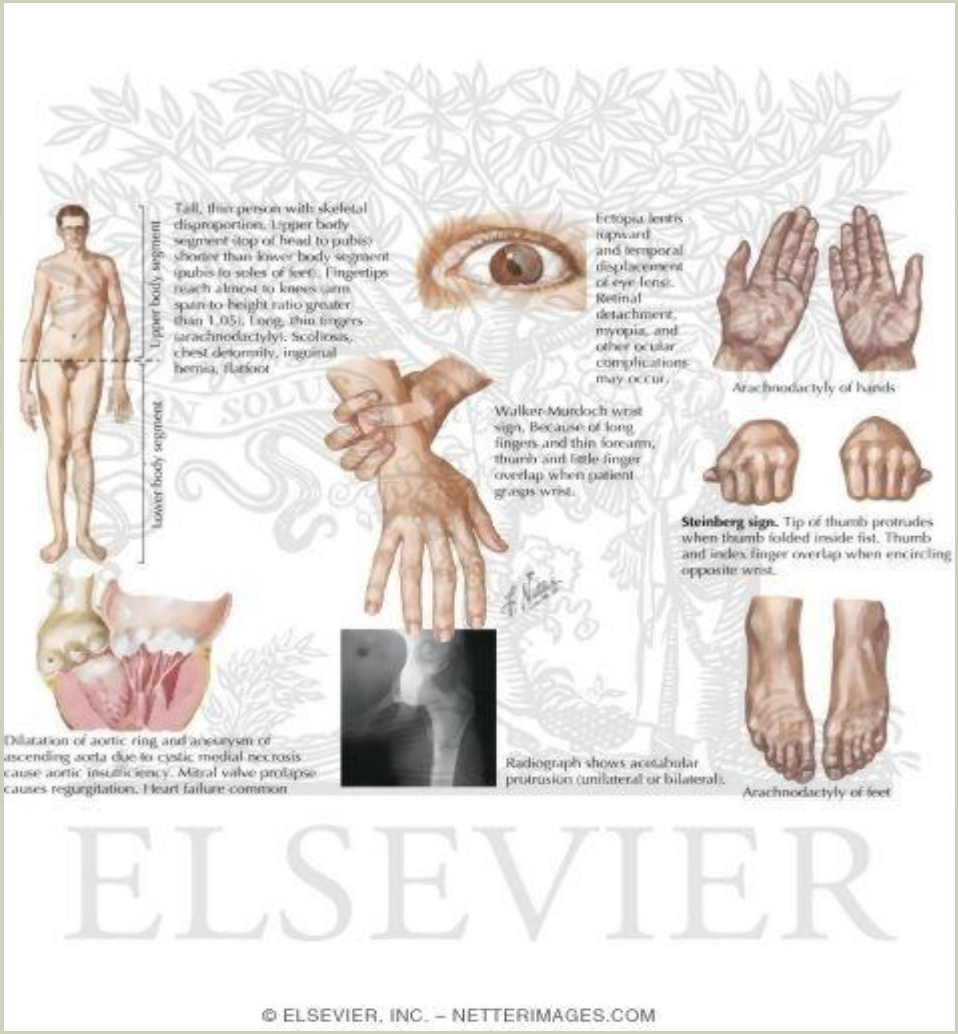
MARFAN'S SYNDROME

MARFAN'S SYNDROME

- Inherited autosomal dominant trait
- Specifically relates to the fibrillin-1 gene
- Fibrillin is a necessary component of elastin
- Genetic derangement of elastin in vital hemodynamic systems such as the lungs, heart and vasculature leads to complications such as pulmonary cysts/emphysema and valvular dysfunction

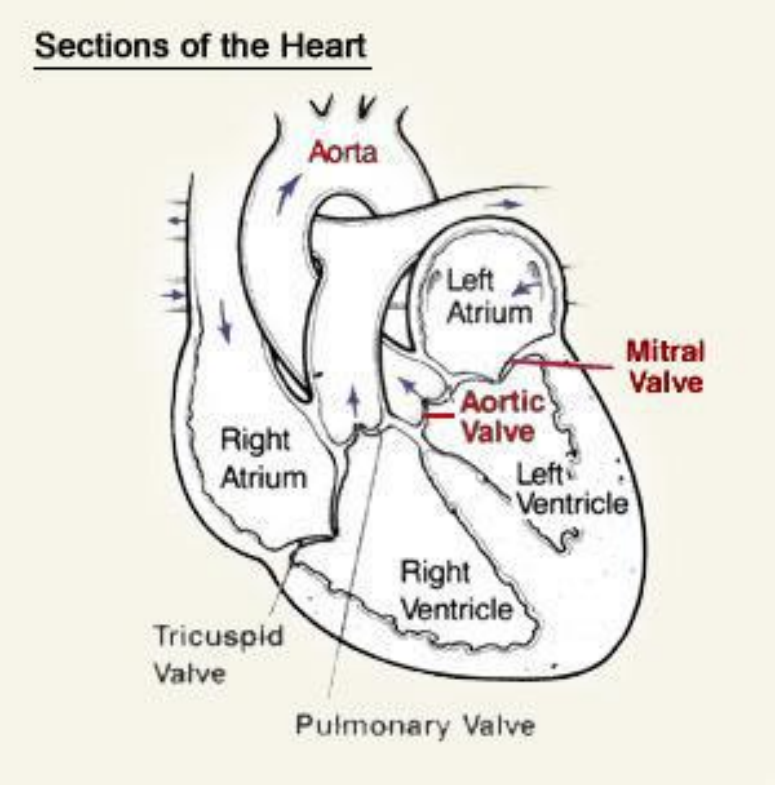
PHYSIOLOGICAL CHARACTERISTICS OF MARFAN'S

- Skeletal System
  - Tall, long slender arms, legs, fingers, and toes
  - Scoliosis, pectus excavatumor carinatum
  - Abnormal joint flexibility
  - High palate, small jaw
- Ocular Findings:
  - Myopia
  - Dislocated lens
  - Detachment of the retina
  - Glaucoma
  - Cataracts



PHYSIOLOGICAL CHARACTERISTICS OF MARFAN'S

- Cardiovascular System
  - Heart murmur
  - Aortic or mitral regurgitation
    - Mitral most commonly affected
  - Aortic root dilation or dissection
  - Pulmonary artery dilatation
  - Congestive heart failure
- Other
  - Spontaneous pneumothorax
  - Decreased muscle mass and subcutaneous fat
  - Obstructive sleep apnea
  - Lumbosacral dural ectasia
  - Early onset of emphysema



ANESTHETIC IMPLICATIONS

- Pre-operative work-up in these patients should include an extensive cardiac examination including electrocardiogram and echocardiogram to determine the extent of cardiac abnormalities.
- Restrictive lung disease requires pre-operative pulmonary function testing and most commonly pressure control ventilation
- Potential for difficult intubation given the skeletal abnormalities of these patients
  - avoid temporomadibular joint dislocation during intubation
  - avoiding hypertension secondary to direct laryngoscopy is imperative as the effects can be disastrous to weakened vasculature (aortic dissection/rupture)

ANESTHETIC IMPLICATIONS

- Managing patient positioning during the intraoperative setting is paramount given the hyperextensibility of the joints and potential for kyphoscoloisis
- Cardiovascular complications pose the largest risk to the patient with Marfan's
- Defective connective tissue in the aorta and heart valves can lead to aortic dilation, dissection, or rupture
- Maintaining a normal blood pressure is typically ideal in order to preserve adequate blood flow to the coronaries and prevent further damage to the aorta

ANESTHETIC IMPLICATIONS

- Pulmonary aspect of Marfan's syndrome must be addressed
- Barotrauma and pneumothoraxes can occur secondary to pulmonary cysts
- Strongly consider tightly controlled positive pressure ventilation in order to prevent further pulmonary damage

EHLERS-DANLOS SYNDROME (EDS)

EHLERS-DANLOS SYNDROME (EDS)

- Inherited connective tissue disorder
- Caused by abnormal production of procollagen and collagen formation
- Incidence= 1 in 5000 people
  - Variety of types
  - Type IV (vascular) syndrome associated with increased risk of death
  - Other types are most often undiagnosed
- **Signs and Symptoms:**
  - Musculoskeletal discomfort
  - Easy bruising and scarring
  - Osteoarthritis
  - Joint hypermobility
  - Skin fragility
  - Hyperelasticity
  - Collagen excess in GI tract, vasculature, uterus
    - Leads to arterial rupture in uterus, bowels, arteries
  - **Spontaneous vascular dissection**
    - **Most common cause of death**
  - Pregnancy
    - Increased risk of premature labor and excessive bleeding

EDS DIAGNOSIS & TREATMENT

- Often misdiagnosed or undiagnosed
  - Often mistaken as a psychiatric disorder
  - Patients often become depressed, drug users, suicidal
- Diagnosis based on clinical findings and presentation
- Genetic testing available (only way to have definitive dx)
- Treat symptoms
  - Efficient wound care
  - PT & OT to learn muscle control & body mechanics

EDS ANESTHETIC IMPLICATIONS

- ↑ risk of bleeding
  - Avoid IM injection
  - Avoid repeated instrumentation of the esophagus or nose
- Obtain adequate IV access
  - Increased risk of hematoma formation with placement
  - Infiltrated IV may go unnoticed due to laxity of skin
- **Avoid regional anesthesia (due to hematoma risk)**
- Ventilation
  - Tracheal damage
    - Watch cuff pressures
  - Increased risk of pneumothorax
    - Maintain low airway pressures
- Postoperative
  - Wound dehiscence and hemorrhage
    - Maintain BP control

NEUROMUSCULAR AUTOSOMAL DOMINANT DISORDERS

1. Huntington's Chorea Disease
2. Amyotrophic Lateral Sclerosis (ALS)
3. Neurofibromatosis (NF)
4. Myotonic Dystrophica
5. Malignant Hyperthermia*

- ** See class notes from previous semesters.*

HUNTINGTON'S CHOREA

HUNTINGTON'S DISEASE

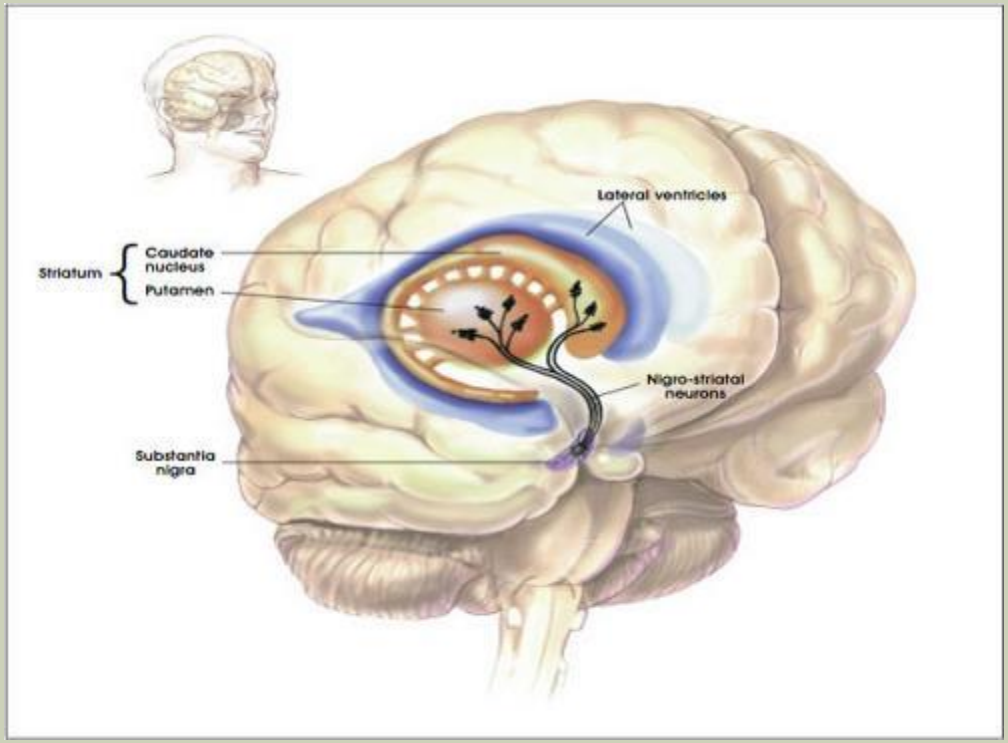
- Is a rare, autosomal dominant, inherited degenerative disorder of the nervous system
- Characterized by clinical hallmarks of choreathetoid movements, psychiatric disturbances and dementia
- Late onset (at 30-45 years)

PATHOPHYSIOLOGY

- HD appears to develop from the neurotoxicity of an abnormal variant of the protein *huntingtin,* arising from an autosomal dominant mutation involving a trinucleotide expansion (CAG) in the Huntington gene on chromosome

- Basal ganglia disease

- There is a marked decrease in GABA throughout the basal ganglia



COMPLICATIONS OF HUNTINGTON'S DISEASE

- The basal ganglia is associated with a variety of functions, including voluntary motor control, procedural learning relating to routine behaviors or "habits," eye movements, and cognitive, emotional function.
- Physical symptoms include:
  - Choreoathetoid movements, along with frequent and sudden jerk movements of any of the limbs or trunk
  - Cachexia with advanced disease
  - **Pharyngeal muscle involvement leads to dysphagia and increases the patient risk of pulmonary aspiration**
- Mental symptoms include:
  - Depression, obsessive-compulsive behavior, social withdrawal, irritability, and delusions
- NO TREATMENT TO STOP DISEASE PROGRESSION

ANESTHETIC IMPLICATIONS

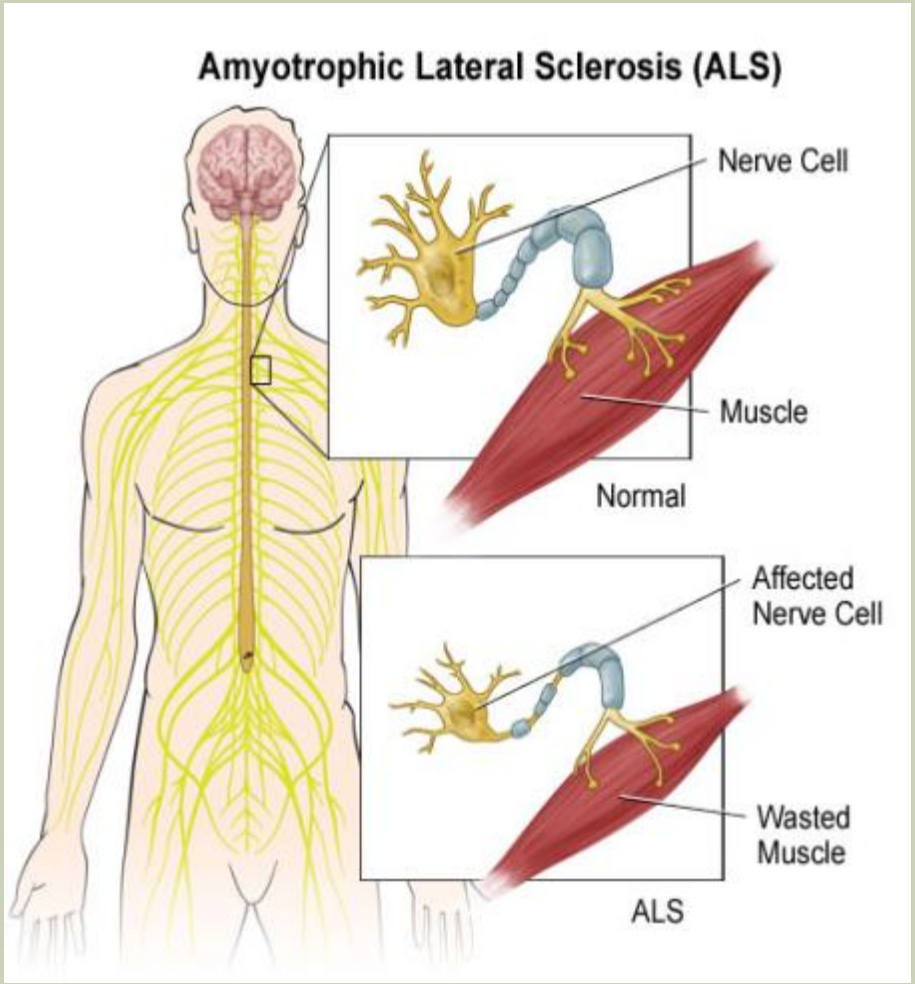
- Major concerns in anesthetic management of Huntington's disease are the following:
  - **Difficult airway**
  - **Risk of aspiration**
  - **Sleep apnea**
  - **Altered reactions to various drugs**
- A rapid sequence induction or modified rapid sequence induction with cricoid pressure be used if a general anesthetic with intubation is necessary.
- Use of TIVA technique to reduce the risk of post op shivering related to inhalation agents.
  - Post-op shivering= increase risk of general tonic spasms!!!
- Use of NDNMB(rocuronium) is safe in these patients.
- These patients may potentially have decreased levels of pseudocholinesterase leading to a prolonged effect of succinylcholine.

AMYOTROPHIC LATERAL SCLEROSIS

AMYOTROPHIC LATERAL SCLEROSIS

- "Lou Gehrig's Disease"
  - Recently gained public attention via *Ice Bucket Challenge*
  - Most commonly affects men 40-60 years of age
- Degenerative disease of motor neurons throughout CNS
  - Upper Motor Neurons
    - Corticospinal tracts (primary descending upper motor neurons)
  - Lower Motor Neurons
    - Anterior horn cells of spinal cord
    - Distal parts are affected first, then moves towards cell body
    - After this occurs the neuron is killed, and the muscle cells innervated by these neurons atrophies
    - Get: atrophy and muscle weakness.
- *Remarkable feature: Entire sensory system, autonomic nervous system, & intellect remain intact*

AMYOTROPHIC LATERAL SCLEROSIS PATHOPHYSIOLOGY



- Cause: Unknown
  - Many hypotheses:
    - Glutamate- induced excitotoxicity
    - Oxidative stress
- 10% of ALS cases are familial
- 90% of ALS cases are sporadic
- Associated with lung carcinoma
- Fast progressing
  - 50% mortality within 30 months diagnosis
  - 20% may survive 5- 10 years
- Most common causes of death: respiratory failure or circulatory collapse

AMYOTROPHIC LATERAL SCLEROSIS DIAGNOSIS

- 4 patterns of clinical presentation:
  - Limb onset
  - Bulbar onset
    - Dysarthria & dysphagia (Often causes pulmonary aspiration)
  - Pure upper motor neuron
  - Progressive muscular atrophy
- S/S influenced by affected neurons
  - Most common symptom: *asymmetric* limb weakness
- Initial symptoms: skeletal muscle weakness, atrophy & fasciculation
  - Often begins in the hands, progresses to all skeletal muscles (incl. tongue, pharynx, larynx & chest)
- Pulmonary function tests
  - Decrease in vital capacity & maximal voluntary ventilation
  - *Respiratory failure eventually develops with ventilatory support required*
- Autonomic dysfunction
  - Resting tachycardia, orthostatic hypotension, & elevated circulating levels of epinephrine & norepinephrine
- *Differential diagnosis: Plasma creatine kinase concentrations are normal in ALS, distinguishing this from chronic polymyositis*

AMYOTROPHIC LATERAL SCLEROSIS TREATMENT

- No cure
- Mostly symptomatic therapy
- Riluzole
  - Glutamate release inhibitor
  - Only specific drug for treatment of ALS
  - Modestly improves survival 3-6 months

Stephen Hawking was diagnosed with ALS >45 years ago & is a rare example of someone who has defied the mortality odds.

AMYOTROPHIC LATERAL SCLEROSIS ANESTHETIC IMPLICATIONS

- No preferred anesthetic technique
- Thorough pre-operative evaluation
- Muscle Relaxants
  - Neuromuscular transmission is markedly abnormal
    - ↑ SENSITIVITY TO NDNMB
    - AVOID SUCCINYLCHOLINE
- Consider TIVA if NMB not required
- ASPIRATION PRECAUTIONS:

Bulbar involvement w/pharyngeal muscle dysfunction ↑ risk of aspiration

- Regional Anesthesia
  - Usually avoided 2° fear of exacerbating disease symptoms
  - Epidural anesthesia has been used successful w/out neurologic exacerbation/pulmonary function impairment
- Postoperative ventilatory support likely 2° exaggerated respiratory depression

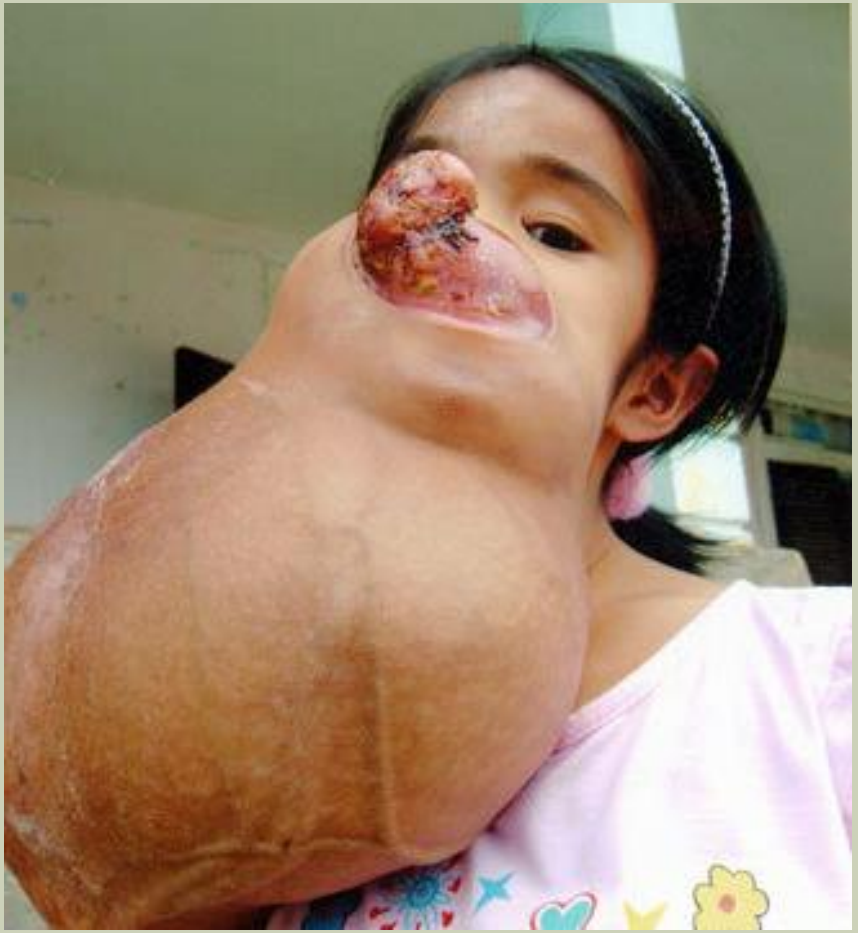
NEUROFIBROMATOSIS (NF)

NEUROFIBROMATOSIS (NF)

- Autosomal dominant mutation
- Incidence- both sexes affected
- Manifestation categories include:

1. Classic (von Reckling-hausen's disease)
2. Acoustic
3. Segmental

- Common feature= disease progression



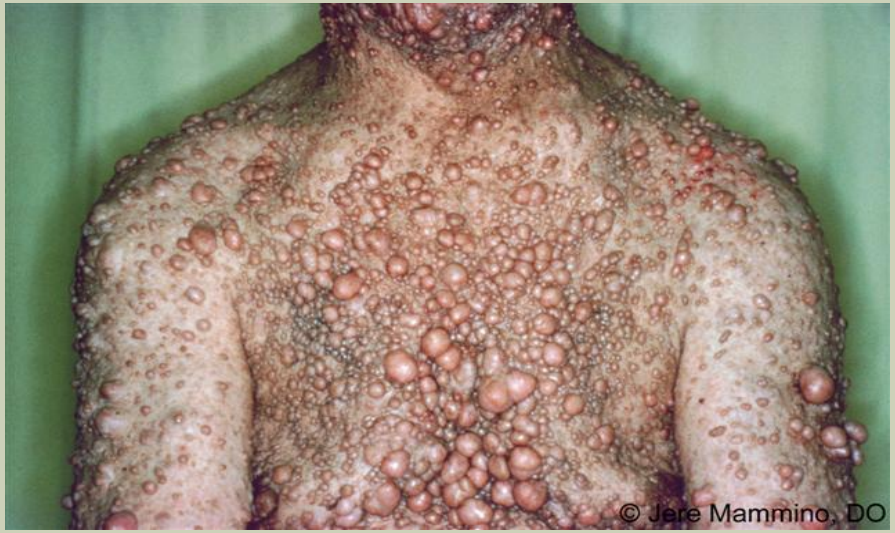
http://www.examiner.com/images/blog/wysiwyg/image/Paul_Provenza.jpg

NF PATHOPHYSIOLOGY

- Congenital pseudoarthrosis- spontaneous fracture progressing to nonunion injury requiring amputation
- Kyphoscoliosis
- Endocrine disorders
- Intellectual impairment seen in 40% patients
- Intracranial tumors occur in 5-10% of patients
  - ↑ morbidity and mortality
  - May manifest as seizure disorder
- Neurofibromas that involve the skin, but can occur in the peripheral nerves, nerve roots, and blood vessels innervated by autonomic nervous system
  - Compromise of airway anatomy?
    - Laryngeal
    - Cervical
    - Mediastinal

NF DIAGNOSIS & TREATMENT

- Diagnosis:
  - Café au lait spots (abnormal cutaneous pigmentation)
    - >/6 spots >1.5cm in diameter
    - Usually present at birth & ↑ during the first decade of life
    - Benign lesions, but patients have ↑ risk of cancer
    - Puberty and pregnancy= ↑number and size



http://www.aocd.org/?page=Neurofibromatosis

- Treatment
  - Management of symptoms
  - Surgical removal of neurofibromas
  - Surgical stabilization of progressive kyphoscoliosis

NF ANESTHETIC IMPLICATIONS

- Various presentations of NF
- Presence of pheochromocytoma?
- ↑ICP due to intracranial tumor?
- **Difficult airway due to expanding laryngeal neurofibromas → Awake, fiberoptic intubation**
- Patients with NF and scoliosis are likely to have cervical spine defects that impact positioning
- Regional anesthesia decisions must involve consideration of neurofibromas involving spinal cord
  - Epidural anesthesia is effective option during L&D
- Responses to MRs are variable (inconclusive)
  - Sensitive AND resistant to succs
  - Sensitive to NDMR

MYOTONIC DYSTROPHY

MYOTONIC DYSTROPHY

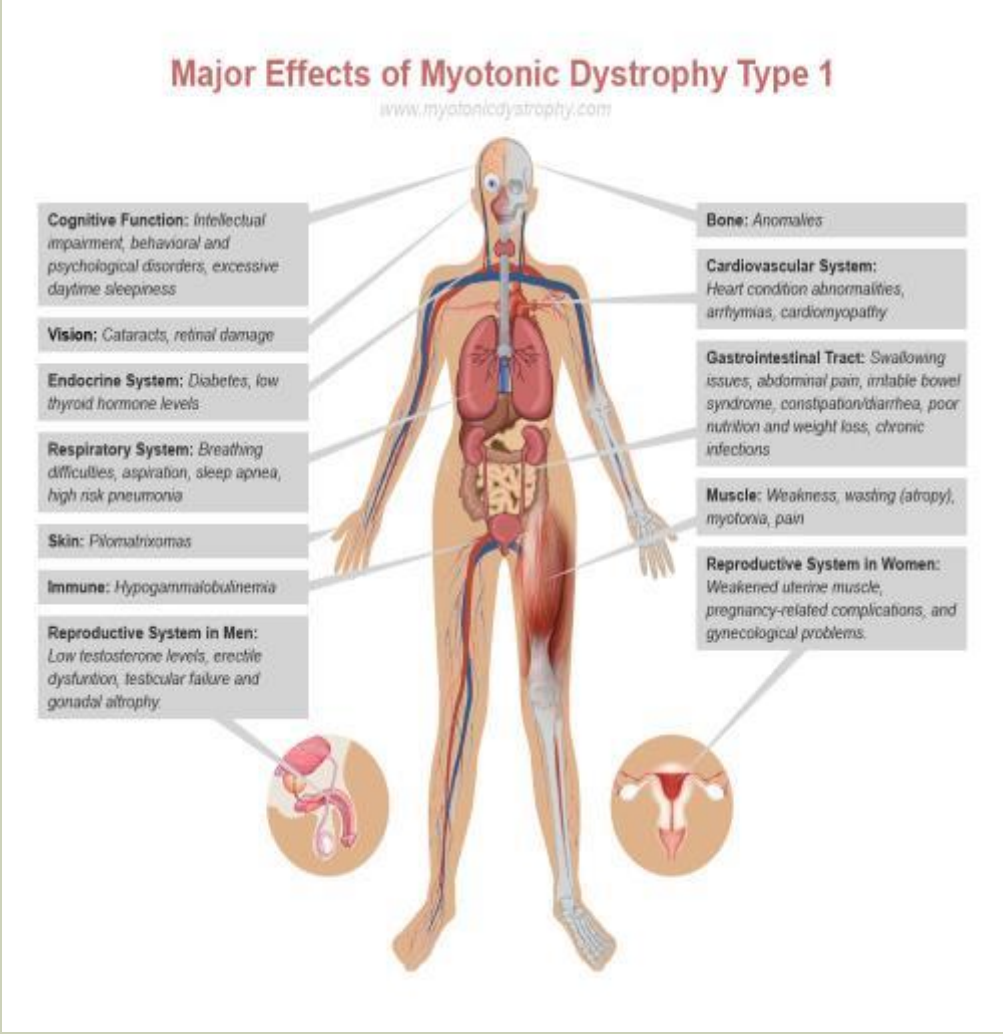
- Myotonic dystrophy is a term designated to describe a group of hereditary degenerative diseases of skeletal muscle characterized by persistent contracture following voluntary muscle contraction
  - Autosomal dominant inherited neuromuscular disorder
- The inability for muscles to relax is due to abnormal calcium metabolism
  - Calcium fails to return to the sarcoplasmic reticulum thus remaining available for continued skeletal contraction
- The most common and severe form of myotonic dystrophy is myotonic dystrophica

MYOTONIC DYSTROPHY CLINICAL MANIFESTATIONS

System	Manifestations
Neuromuscular	Myotonia, weakness Reduced deep tendon reflexes
Ocular	Cataract, ptosis Ophthalmoparesis, retinal pigmentation
Endocrine	Testicular atrophy, diabetes, pituitary dysfunction, hyperparathyroidism
Skin	Frontal balding, pilomatrixoma
Cardiovascular	Hypotension, syncope, palpitations, mitral valve prolapse, sudden death
Gastrointestinal	Dysphagia, pseudo-obstruction
Central nervous	Mental retardation
Immune	Reduced immunoglobulin levels

MYOTONIC DYSTROPHICA

- Inherited autosomal dominant trait with the onset of symptoms during the second or third decade
- Is a multisystem disease affecting cardiac, smooth and skeletal muscle
- Signs and Symptoms:
  - Usually manifests as facial weakness, wasting and weakness of the sternocleidomastoid muscles, ptosis, dysarthira, dyspagia, and inability relax the hand grip
  - Mental retardation, frontal baldness, and cataracts
  - Delayed gastric emptying
  - Cardiac dysrhythmias 1st degree HB is quite common
  - **Pharyngeal and thoracic muscle weakness makes these patients vulnerable to pulmonary aspiration**



MANAGEMENT OF ANESTHESIA

- **Preoperative evaluation**
  - Must consider the likelihood of cardiomyopathy, respiratory muscle weakness, and the potential for abnormal responses to anesthetic drugs
- **Intraoperative Management**
  - Treat all patients as if they have cardiomyopathy and conduction defects
  - Transthoracic pacing and antiarrhythmic medication should be available
  - Avoid inhalation agents if possible d/t their myocardial depressant and conduction system effects

MANAGEMENT OF ANESTHESIA

- **Intraoperative Management**
  - High risk of aspiration!!!
  - **Avoid succinycholine-→ can produce contractures lasting several minutes and interfere with intubation and ventilation**
  - **NDNMB are ok**
  - Caution should be used when using neuromuscular reversal agents as they could precipitate skeletal muscle contraction
    - Use short acting NDNMB and titrate carefully to avoid reversal agents
  - Patients are sensitive to the respiratory depressant effects of barbiturates, opioids, benzos, and propofol due central respiratory depression and weak respiratory musculature
  - When possible REGIONAL ANESTHESIA is the preferred anesthetic
  - Maintain normothermia and avoid shivering as both can induce myotonia
  - If needed, treat excessive muscle contractions with Phenytoin 4-6mg/kg/day or Quinine 0.3-1.5g/day

QUESTIONS?

NEUROMUSCULAR DISORDERS & ANESTHETIC IMPLICATIONS PART II

MSNA534: Principles of Nurse Anesthesia for Patients with Co-existing Diseases

Kim Kujanek, Amber Kursch, & Hilary Pekarek
Student Registered Nurse Anesthetists
Adventist University of Health Sciences
October 2014

OTHER NEUROMUSCULAR DISORDERS

- Neurological
  1. Seizure Disorder
  2. Arnold- Chiari Malformations
  3. Neuro-Ocular Disorders (NOD)
  4. Parkinson's Disease
  5. Post-polio Sequelae (PPS)

- Musculoskeletal
  1. Muscular Dystrophies
  2. Polymyositis
  3. Dwarfism
  4. Rheumatoid Arthritis (RA)
  5. Ankylosing Spondylitis (AS)
  6. Kyphoscoliosis
  7. Spinal Cord Disorders

SEIZURE DISORDER

SEIZURE DISORDER

- Caused by a synchronize discharged group of neurons in the brain.
- Most common neurologic disorder: 10% will experience a seizure in their lifetime.
- Epilepsy: Recurrent seizures resulting from congenital or acquired factors. Typically begins in childhood.
- Adults that have seizures most likely due to tumors, injury, metabolic dysfunction, infection, or cerebrovascular disease.

SEIZURE DISORDER

Partial Seizures

- Begins in focal area of one cerebral hemisphere

Simple Partial: No loss of consciousness. Pt has aura. Motor, sensory, or autonomic signs.

Complex Partial: Impairment of consciousness. Starts in one hemisphere (Temporal) but soon involves same area in 2nd hemisphere. Pt has déjà vu, automatisms, hallucinations.

Generalized Seizures

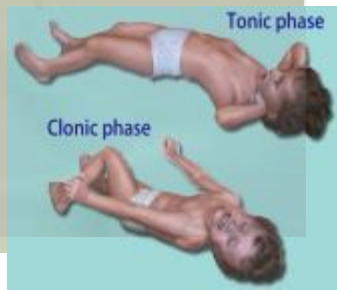
- Involves both cerebral hemispheres. + LOC. Can be convulsive/not.

Absence seizures (petit mal): Loss of consciousness. "Blank stare"

Atonic: Sudden loss of muscle tone."Drop attacks"

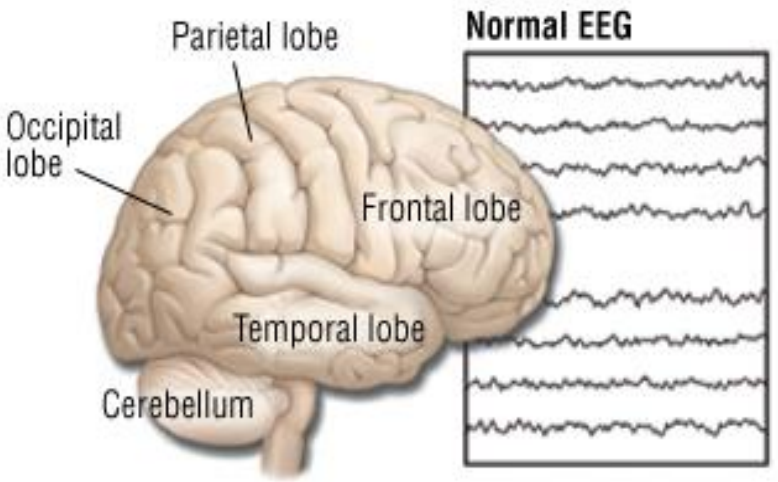
Myoclonic: Bilateral jerking of muscles

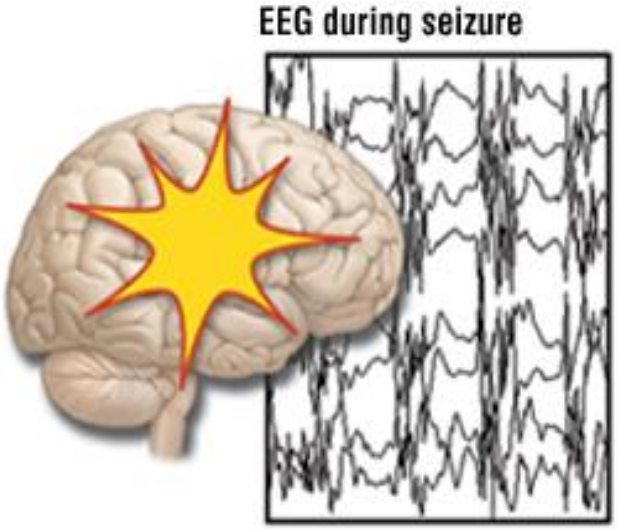
Grand Mal: Tonic/clonic. Incontinence seen (Porth, 2010).



Status Epilepticus: Seizures that do not stop spontaneously or occur in Succession without recovery.

SEIZURE DISORDER DIAGNOSIS





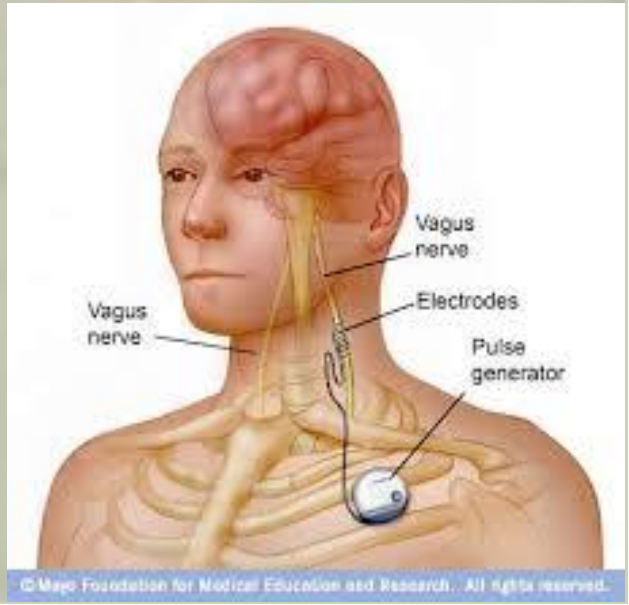
- Differential diagnosis:
  - Hypoglycemia, Hyponatremia, Other electrolyte abnormalities, Drug intoxication, Intracranial hemorrhage, ↑ ICP, Cyclosporin-induced, ETOH W/D
- MRI
- EEG
- Electrocorticography
  - Electrodes surgically placed directly on cerebral cortex
  - More accurate focus identification
  - Allows mapping of electrical events

SEIZURE DISORDER TREATMENT

- Medications:
  1. **Antiepileptics/ Anticonvulsants**
     - ~ 30% patients still poorly controlled with antiepileptics
     - Carbamazepine
     - Phenytoin
     - Levetiracetam
     - Valproate
     - Eslicarbazepine
     - Lamotrigine
     - Gabapentin
  2. **Barbiturates**
     - Phenobarbital, Thiopental
  3. **Benzodiazepines**
     - Midazolam, Lorazepam, Diazepam
  4. **Propofol**
     - MOA: GABA mediated inhibition of Cl- channels
     - Can be used in hospital setting to treat status epilepticus & is OK to use for ECT

SEIZURE DISORDER TREATMENT

- Support airway and ventilation
- Vagal Nerve Stimulator
  - Implanted on left side 2° significant cardiac innervation by right vagal nerve
  - Complications: VC paralysis, airway obstruction, bradycardia, asystole, & facial palsy
- Surgical Treatment
  - Resection of pathological section
  - Corpus Callosotomy
  - Hemispherectomy



SEIZURE DISORDER ANESTHETIC IMPLICATIONS

- Preoperative exam
  - Nature, cause & frequency of seizures
  - Date of last seizure
  - *Note: Important to maintain treatment w/ preoperative antiepileptic drugs throughout perioperative period*

- Impact of seizure medications
  - ***CYP inducers*** **: Carbamazepine, phenytoin, barbiturates**

  - **Consider effect on organ function**
    - Phenytoin
      - S/E: Hypotension, cardiac dysrhythmias, gingival hyperplasia, aplastic anemia, erythema multiforme, Stevens-Johnson syndrome
    - Valproate
      - S/E: Hepatic failure in 1/10,000, pancreatitis
      - Long term use= ↑ surgical bleeding (esp. in children)
    - Carbamazepine
      - S/E: Diplopia, dose-related leukopenia, hyponatremia, alterations in hepatic metabolism of various drugs

SEIZURE DISORDER ANESTHETIC IMPLICATIONS

- Muscle Relaxants
  - **Caution with Atracurium & Cisatracurium**
    - Active metabolite: Laudanosine is proconvulsant
  - **↑ RESISTANCE TO NMB (esp. phenytoin)**
    - Antiepileptic drugs can ↓ DOA of NDNMB

- For anesthetic induction, consider effect on CNS electrical activity
  - i.e. Methohexital can activate epileptic foci & is desirable for certain procedures (i.e. ECT)

- Anesthetic drug choice
  - Anti-epileptogenic drugs: Thiobarbiturates, opioids, & benzodiazepines preferred
  - Isoflurane, desflurane, & sevoflurane acceptable

- Alfentanil, ketamine, enflurane, isoflurane, & sevoflurane → epileptiform spike and wave EEG activity in patients w/out hx seizures
- *Ketamine may produce seizure activity, reasonable to AVOID its use*

SEIZURE DISORDER ANESTHETIC IMPLICATIONS

- For Intraoperative Electrocorticography
  - Inform patient of the increase risk of awareness during preop
  - Medications to maintain anesthesia
    - Precedex, droperidol, narcotics (e.g. remifentanil), N_2O, & diphenhydramine
    - Methohexital 0.3mg/kg, Etomidate 0.05-1mg/kg, & high doses of alfentanil 50mcg/kg can be given to induce cerebral electrical activity

- Regional anesthesia
  - Local anesthetics can ↓ seizure threshold, but no evidence that RA ↑ risk of seizures

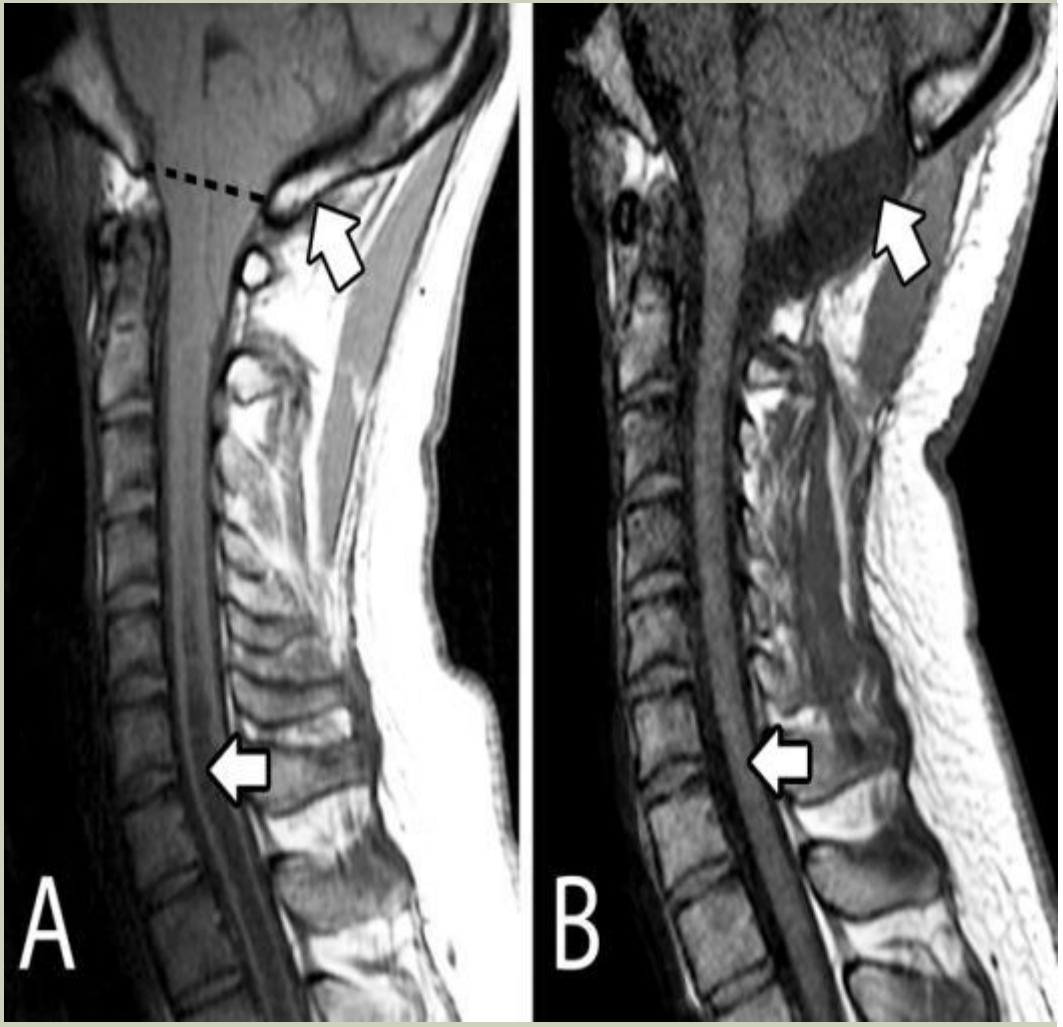
ARNOLD-CHIARI MALFORMATIONS

ARNOLD-CHIARI MALFORMATIONS

- Commonly known as Chiari malformations
- Congenital displacement of the cerebellum
- May also compress the brain stem
- Incidence of Chiari I= 1/1000 births
- Women > Men
- Four grades of malformation:
  - Chiari I- downward displacement of cerebellar tonsils over the cervical spinal cord
  - Chiari II- downward displacement of cerebellar vermis (meningomyelocele is often present)
  - Chiari III- displacement of cerebellum into an occipital encephalocele (EXTREMELY RARE)
  - ***Chiari IV- Involves an incomplete or underdeveloped cerebellum (cerebellar hypoplasia)= incompatible with life (fetal demise)***

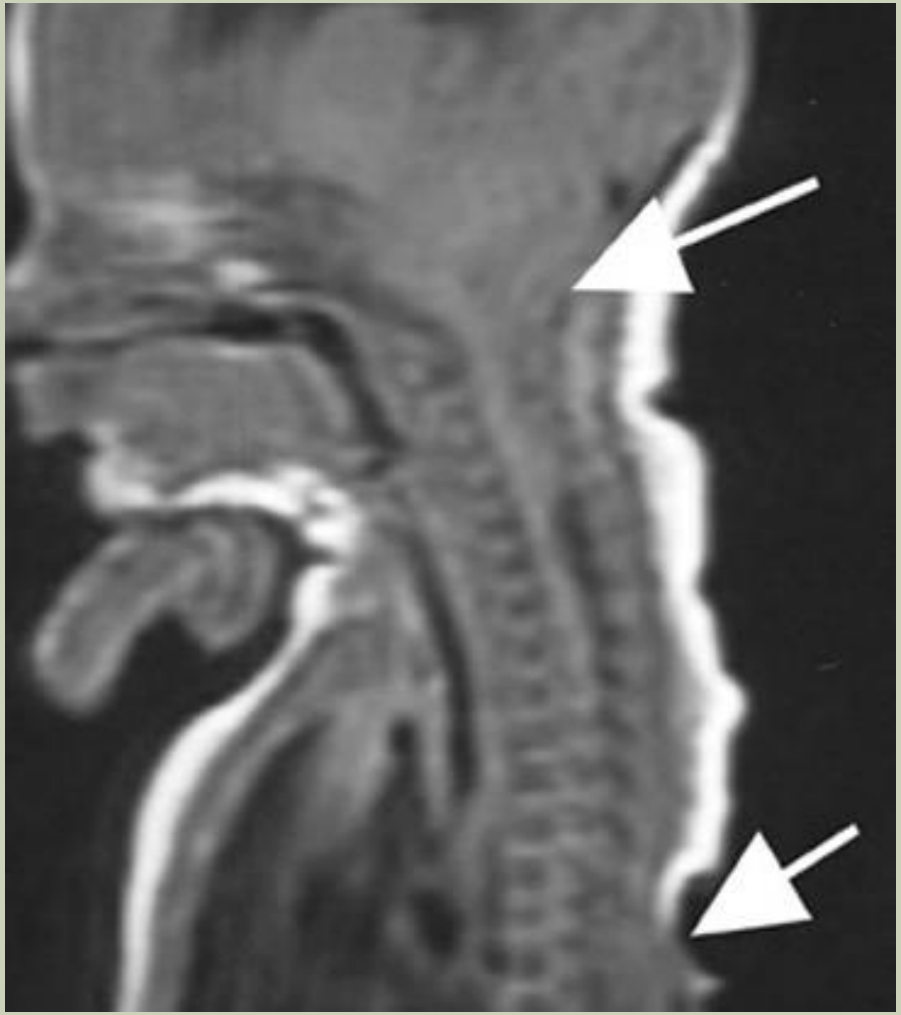
CHIARI I PATHOPHYSIOLOGY

- S/S can occur at any age
- Occipital HA is most common (aggravated by moving head, coughing, etc.)
- Visual disturbances, intermittent vertigo, ataxia can also occur
- Syringomyelia (spinal cord cyst)

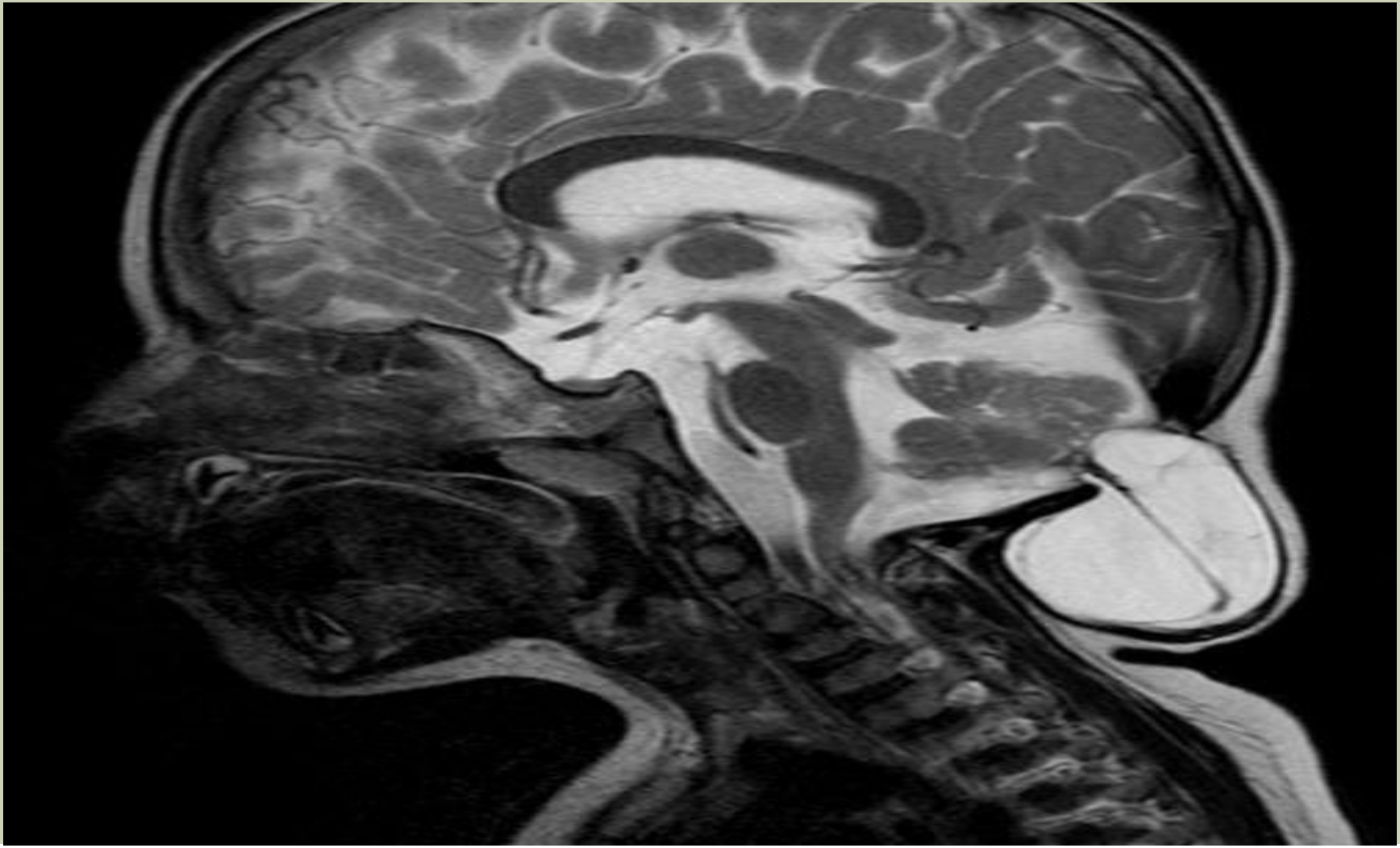


CHIARI II PATHOPHYSIOLOGY

- Cerebellar vermis and brainstem displaced into cervical spinal canal
- Presents at birth or early infancy
- Associated with spina bifida
- Obstructive hydrocephalus requiring shunting develops in 90% of infants
- Symptoms are difficulty swallowing, recurrent aspiration, stridor, & apneic episodes
- Surgical intervention within first 48 to 72 hours of life
- ↑ prevalence of clinical latex allergy



CHIARI III PATHOPHYSIOLOGY



CHIARI MALFORMATIONS

Diagnosis

- Signs of ↑ ICP= vision changes, headache, etc.
- Differential diagnosis
- Patient history
- Neurological examination
- Magnetic Resonance Imaging (MRI)

Treatment

- Based on symptomology
- Surgical decompression via enlarging the foramen magnum
- Supportive therapy thereafter

CHIARI ANESTHETIC IMPLICATIONS

- Careful documentation of baseline sensory and motor deficits
- Cervical spine assessment evaluating possible associated cervical spine abnormalities involving limited range of motion → **fiberoptic intubation is recommended** to avoid further displacement and/or compression of neural structures
- Neurologic assessment focused on evaluation of brainstem compression or cranial nerve involvement- vocal cord dysfunction, breathing disorders, dysphagia → risk for aspiration → awake extubation (to ensure intact airway reflexes) with smooth emergence (avoid coughing as this will ↑ ICP)
- **Avoid Succs. in order to prevent further ↑ ICP**

CHIARI ANESTHETIC IMPLICATIONS

- REGIONAL ANESTHESIA CONSIDERATIONS:
  - Parturients WITHOUT ↑ ICP, or WITHOUGHT significant neurologic symptomatology, have been reported to have safe and effective epidural and spinal anesthetic throughout labor and delivery
  - In patients with ↑ ICP and neurologic deficits, the risks and benefits for any form of anesthesia should be carefully weighed, as dural puncture may result in the sudden neurologic deterioration caused by further cerebellar herniation
  - A combination of cervical and pudendal blocks, supplemented by parenteral opioids, may be the safest approach during labor and delivery
  - GETA WITHOUT the use of Succs is advised for caesarean section

NEURO-OCULAR DISORDERS (NOD)

NEURO-OCULAR DISORDERS (NOD)

- Disorders involving the visual system that affect the retina, optic nerve, and intracranial optic system:

1. Leber's Optic Atrophy
2. Retinitis Pigmentosa
3. Kearns-Sayer Syndrome
4. Ischemic Optic Neuropathy (Anterior and Posterior) is THE MOST COMMON CAUSE OF NEW-ONSET BLINDNESS DURING THE POSTOPERATIVE PERIOD
5. Cortical Blindness
6. Retinal Artery Occlusion
7. Ophthalmic Venous Obstruction

(#1-3 are degenerative diseases)

NOD
LEBER'S OPTIC ATROPHY

- Also known as Leber's hereditary optic neuropathy
- First human disorder for which a mitochondrial pattern was definitively described
- RARE
- Degeneration of the retina and atrophy of optic nerves → central blindness that occurs in adolescence or early adulthood
- Usually associated with other neuropathology including MS and dystonia

NOD
RETINITIS PIGMENTOSA

- Genetically and clinically heterogeneous group of inherited retinopathies
- Degeneration of the retina
- Occurs in 1 in 3000
- Retinal pigmentation of peripheral areas upon exam
- Vision loss occurs from the periphery of the retina toward the center until total blindness occurs

NOD
KEARNS-SAYER SYNDROME

- Retinitis pigmentosa associated associated with progressive external ophthalmoplegia
- RARE
- Potentially a viral etiology
- Typically manifests before third decade of life
- Cardiac conduction abnormalities are common (BBB to complete AV block) ➔ sudden death
- Generalized degeneration of CNS has been reported

NOD
ISCHEMIC OPTIC NEUROPATHY (ION)

- Should be considered in patients c/o visual loss during the first week following any surgery
- After injury, vision improvement prognosis is poor and urgent ophthalmologic consultation imperative
- Ischemic injury to the optic nerve can result in both central and peripheral vision
  - Anterior= central retinal artery & small branches of the ciliary artery
  - Posterior= small branches of the ophthalmic & central retinal arteries

*Incidence= Posterior ION > Anterior ION

NOD
ANTERIOR ION

- Caused by infarction within the watershed perfusion zones between small branches of the short posterior ciliary arteries
- Sudden, painless, monocular visual deficit varying in severity
- Asymptomatic optic disk swelling may be earliest sign (congenitally small optic disk is usually present)
- An arteritic anterior ION is less common and associated with inflammation and thrombosis (high dose steroids can be given to ↓ inflammation and prevent damage to contralateral eye)
- A nonarteritic form of anterior ION is more common and likely to manifest during postop period
  - Attributed to ↓ O2 delivery to optic disc during hemorrhagic hypotension, anemia, cardiac surgery, ENT surgery, cardiac arrest, & hemodialysis

NOD
POSTERIOR ION

- Acute vision loss despite initially negative opthalmoscopic findings → retrobulbar involvement of optic nerve
- Presumed cause is ↓ O2 delivery to posterior portion of optic nerve *due to hypotension, anemia, congenital absence of central retinal artery, altered optic disc anatomy, air embolism, venous obstruction, & infection*
- Posterior ION is more common than anterior ION as a cause of visual loss in the perioperative period
  - HIGH RISK SETTINGS/ SITUATIONS INCLUDE:
    - Prolonged spine surgery in prone position
    - Cardiac surgery
    - Radical neck dissection
    - Hip arthroplasty
    - Cardiac arrest
    - Treatment of hypertensive crisis
    - Blunt trauma
    - Severe anemia

NOD
CORTICAL BLINDNESS

- Loss of vision, but retention of pupillary reactions to light and normal findings upon funduscopic exam
- May occur after profound hypoperfusion and infarction of watershed areas in the parietal or occipital lobes
- Observed after cardiac surgery (due to emboli from CPB), craniotomy, laryngectomy, & caesarean section
- Patients may not be aware of focal vision loss, which typically improves over time
- Abnormal parietal and/or occipital lobes via CT and/or MRI confirms the diagnosis

NOD
RETINAL ARTERY OCCLUSION

- Painless monocular blindness that is severe]
- Due to occlusion of a branch of the retinal artery caused by ulcerated atherosclerotic plaque in the ipsilateral carotid artery
- Vasospasm and thrombosis may precipitate this after radical neck surgery that is complicated by hemorrhage and hypotension
- This can occur following intranasal injection of alpha-adrenergic agonists (i.e. phenylephrine, afrin, etc.)
- Ophthalmoscopic exam reveals a pale edematous retina
- Stellate ganglion block may improve vision

NOD
OPHTHALMIC VENOUS OBSTRUCTION

- Obstruction of venous drainage from eyes can occur intraoperatively when patient positioning results in external pressure on the orbits
- Diagnosis is confirmed by ophthalmoscopic examination revealing engorgement of the veins and edema of the macula
- Prone positioning and use of headrests during neurosurgical procedures require special attention to ensure eyes and orbits are free from pressure

PARKINSON'S DISEASE

PARKINSON'S DISEASE

- Is a neurodegenerative disorder of unknown cause characterized by resting tremor, bradykinesia, rigidity, and postural instability

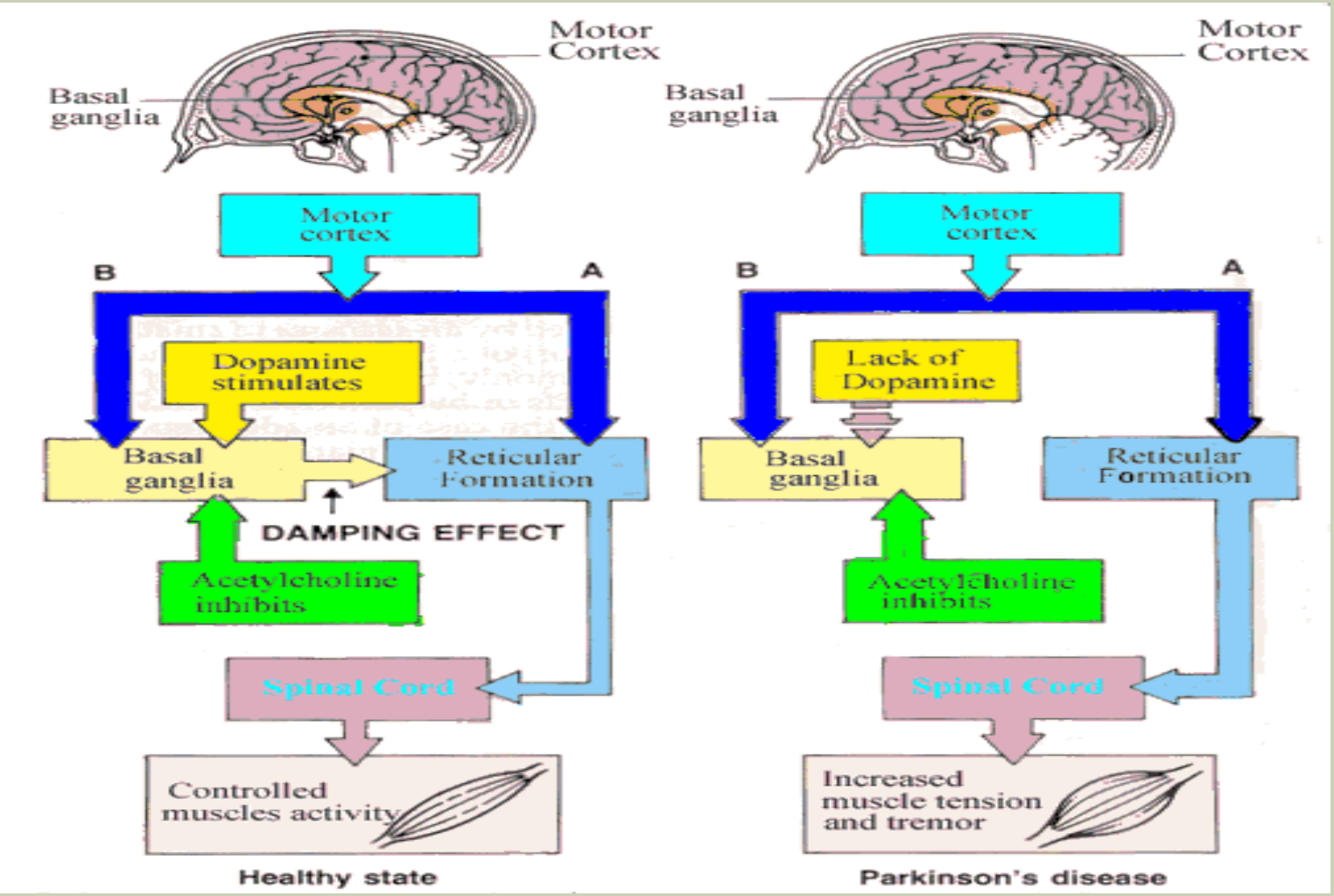
- Along with the mentioned cardinal signs of Parkinson's Disease patients will also likely experience some of the following:
  - Dementia(common), fatigue, depression, hallucinations, psychosis, anosmia(lack ability to perceive odor), and autonomic instability

- Older age has been persistently associated with increased risk of Parkinson's Disease

PATHOPHYSIOLOGY

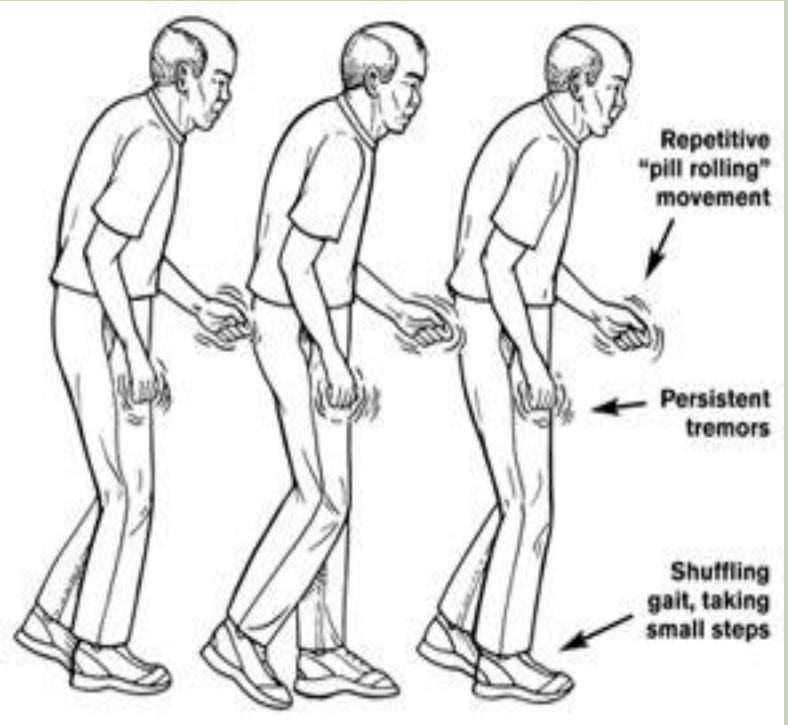
- Coordination of movement depends on a complex feedback loop in which the cortex sends information to the basal ganglia and cerebellum and in turn receives information from these structures through the thalamus
- Parkinson's is characterized by a neuronal loss, depigmentation, and gliosis(dense fibrous network replaces previous healthy nerve tissue) in the substantia nigra
- Along with a neuronal loss there is characteristic loss of dopaminergic fibers normally present in the basal ganglia.
  - This results in a loss of regional dopamine concentrations leading to unopposed stimulation of acetylcholine and excessive extrapyramidal motor system stimulation

PATHOPHYSIOLOGY



CLINICAL MANIFESTATIONS

- **Classic Triad:**
  - Skeletal muscle tremors, rigidity, and akensia
- Earliest manifestations may include a loss of arm swings when walking
- Facial immobility will include infrequent blinking and lack of emotional expressions
- Tremors alternate rhythmically between flexion and extension of digits(pill rolling).
  - Tremors are more prominent during periods of rest



ANESTHESIA MANAGEMENT

- Understanding how Parkinson's is treated is imperative to the anesthetic management of these patients
  - Treatment is focused on increasing the amount of dopamine available in the basal ganglia
  - The use of Levodopa is the most effective treatment for Parkinson's disease
- The elimination half life of Levodopa and the dopamine is produces is brief, therefore interruption of drug therapy for more than **6-12 hours** can result in an abrupt loss of therapeutic effects
  - Loss of therapeutic effects can include skeletal muscle rigidity leading to potential ventilation complications
- **Levodopa therapy must be continued throughout the perioperative period**
  - Oral doses can be given via OGT/NGT if needed

ANESTHESIA MANAGEMENT

- **Preoperative Evaluation**
  - Cognitive impairment may predispose patients to postoperative delirium, and baseline mental status should be assessed
  - More susceptible to respiratory complications, and the pre-op evaluation should elicit evidence of swallowing dysfunction, retained or excessive oropharyngeal secretions, dyscoordination or rigidity of accessory muscles of respiration, and recent or active respiraotry infection
  - Evaluate for arrhythmias, hypertension as well as orthostatic hypotension(common S/E of medications)
  - Autonomic dysfunction may also present in the form of micturition, salivation, GI function, and temperature regulation

ANESTHESIA MANAGEMENT

- **Intraoperative Management**
  - Potential challenges include: increase risk of aspiration, potential for autonomic instability, arrhythmia, and relatively high incidence of orthostatic hypotension
  - Regional anesthesia may reduce the risk of aspiration and respiratory complications, minimize the risk of PONV and facilitate earlier ability to resume PO medications
  - Propofol is good choice for induction if there is an abscence of hypovolemia and/or cardiac dysfunction
  - **Nondepolarizing neuromuscular blockers may mask tremor, but patient with PD appear to have normal sensitivity to these drugs.**
  - **Succinylcholine only caused hyperkalemia in one case**

ANESTHESIA MANAGEMENT

- **Intraoperative Management**
  - Anticholinesterases are safe for use in reversing NMB
  - Glycopyrrolate does not cross the BBB and is the preferred anticholinergic
  - Opioids should be used with caution due the potential of respiratory depression and potential to exacerbate muscle rigidity
  - **Medications C/I: phenothiazines, butyrophenones, and reglan due to the increase risk of EPS or dystonic reactions**
  - Treat hypotension with direct acting agents and volume resuscitation

ANESTHESIA MANAGEMENT

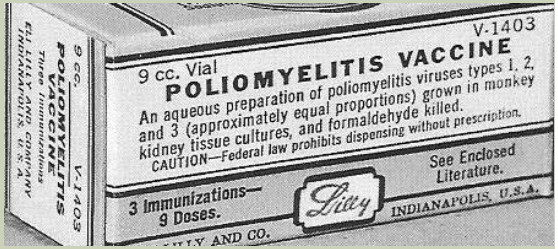
- **Postoperative Management**
  - Watch closely for postextubation respiratory failure
  - Increased risk of laryngospasm, inability to handle oral and respiratory secretions, and aspiration pneumonia
  - Patients should be fully awake prior to extubation
  - Respiratory reserve is likely to be diminished due to intraoperative atelactasis and medication effects

POSTPOLIO SEQUELAE

POSTPOLIO SEQUELAE

- AKA "POLIOMYELITIS" (***Attention: Not to be confused with polymyositis***)

- Etiology
  - Caused by enterovirus
  - Initially infects reticuloendothelial system
    - In minority of patients, virus enters CNS & targets motor neurons in brainstem & anterior horn of spinal cord ➔ Paralysis

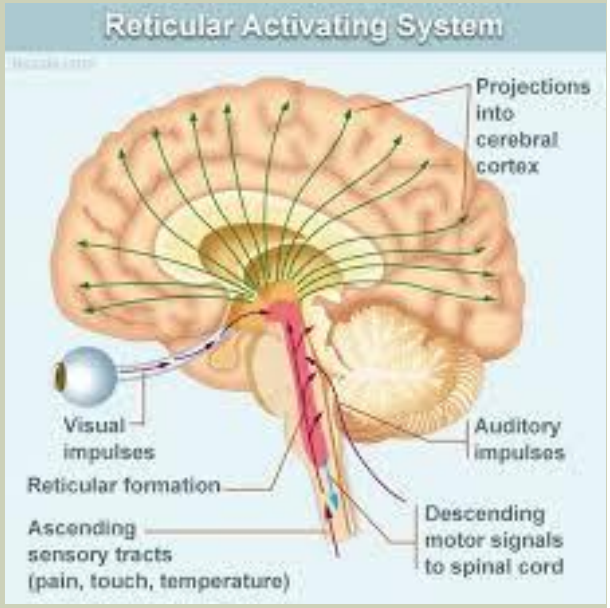


- Incidence
  - Previously a worldwide pandemic
  - Mostly eradicated in USA 2° vaccinations
  - Significantly ↓ since institution of Polio vaccine
    - Henrietta Lacks?

- More likely to see a patient with postpolio sequelae than acute polio
  - ~1.63 million survivors in USA
  - 10-40 years post-polio exposure, 25-40% of survivors develop postpolio syndrome

POSTPOLIO SEQUELAE SIGNS & SYMPTOMS

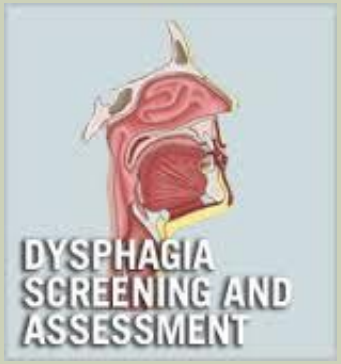
- Usually manifest as neurological damage from original poliovirus
  - Fatigue
  - Skeletal muscle weakness
  - Joint pain
  - Cold intolerance
  - Dysphagia
  - Sleep/breathing problems (i.e. OSA)
- Can damage reticular activating system



POSTPOLIO SEQUELAE ANESTHETIC IMPLICATIONS

- Can damage reticular activating system
  - → Increased sensitivity to sedative effects of anesthetics
  - → Delayed awakening from general anesthesia

- Thorough preoperative assessment
  - 40% have pulmonary dysfunction



- Aspiration risk
  - Dysphagia present in ~ 20%

- OUTPATIENT SURGERY USUALLY NOT APPROPRIATE
  - Have ↑ risk of complications, particularly respiratory muscle weakness & dysphagia

POSTPOLIO SEQUELAE ANESTHETIC IMPLICATIONS

- **Muscle Relaxants**
  - **↑ sensitivity to NDNMB**
  - **AVOID Succinylcholine (Potential for ↑K+)**
- **Regional Anesthesia**
  - **Consider ultrasound for regional blocks due to spinal deformities**
- Severe back pain postoperatively 2° co-existing skeletal muscle atrophy & scoliosis

- Anticipate profound postoperative ***shivering*** 2° sensitive to cold
- Abnormal postoperative pain perception 2° poliovirus damage to endogenous opioid-secreting cells in brain & spinal cord

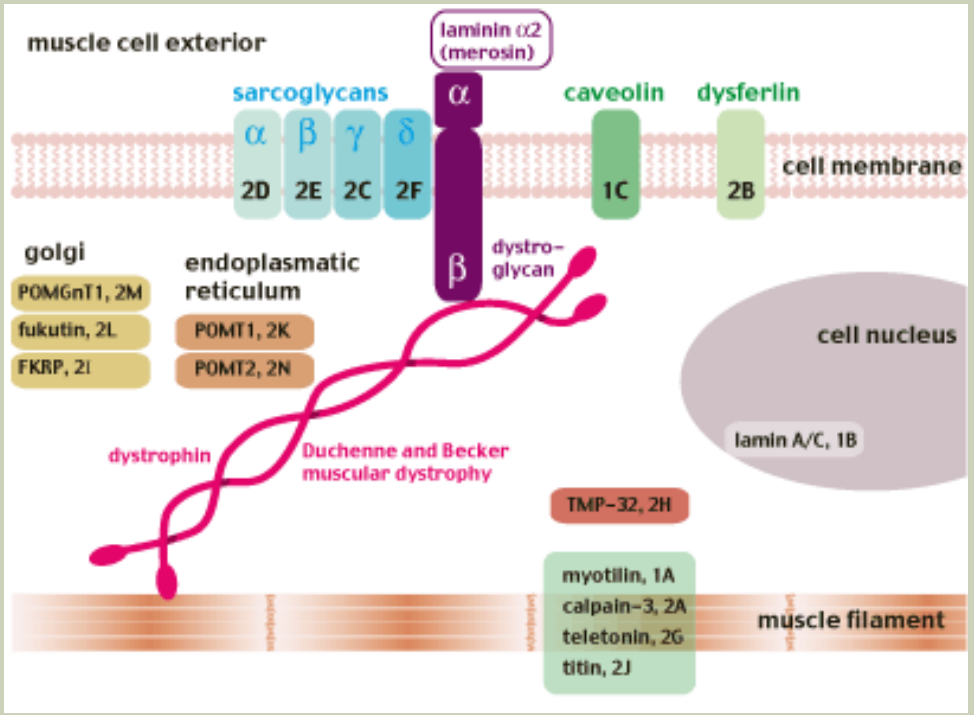
MUSCULAR DYSTROPHIES (DUCHENNE)

MUSCULAR DYSTROPHIES

- Muscular dystrophies are a group of hereditary myopathic diseases characterized by progressive weakness
- The dystrophies are all best characterized by painless degeneration and atrophy of skeletal muscles without evidence of muscle denervation
- Sensation and reflexes are intact
- The most common form of muscular dystrophy is Duchenne's muscular dystrophy (DMD, also called pseudohypertrophic muscular dystrophy)

PATHOPHYSIOLOGY

- Muscular dystrophies are characterized by degeneration of the skeletal muscle fibers and replacement with fibrous and fatty connective tissue
- The absence of dystrophin results in a loss of skeletal muscle membrane integrity and subsequent breakdown of the sarcolemma with an influx of extracellular calcium, activation of cellular proteases, inflammation, necrosis, and replacement fibrosis
  - **Dystrophin stabilizes the muscle surface membrane during contraction and relaxation**

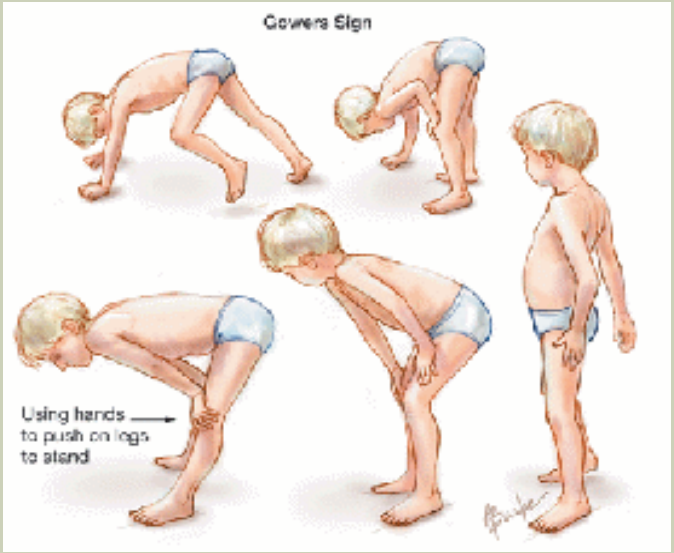


DUCHENNE'S MUSCULAR DYSTROPHY

- Most common form
  - seen in 1 per 3500 births
- Inherited as a sex-linked recessive trait; most of the reported cases are male
- Proximal limb muscle biopsy remains the standard for diagnosis
- Short life expectancy
  - 15-25years

CLINCIAL MANIFESTATIONS

- The initial clinical presentation involves a waddling gait, frequent falling, and difficulty climbing stairs because of proximal muscle weakness in the pelvic girdle
- Certain muscles, particularly the calves, demonstrate early hypertrophy
- Pelvic girdle and proximal leg muscle weakness is responsible for ***Gowers' sign***, with the child climbing up the legs to stand up
- Kyphoscoliosis can develop
- Serum creatine kinase concentrations are 20 to 100 times normal reflecting increased permeability of skeletal muscle membranes and skeletal muscle necrosis
- Mental retardation is often present



CLINICAL MANIFESTATIONS

- Loss of dystrophin and an increase in calcium leads to inflammation and myocardial cell death and fibrosis, producing a ***dilated cardiomyopathy***
- **Cardiomyopathy is the major cause of death in DMD patients**
- EKG reveals tall R waves in V_1, deep Q waves in the limb leads, a short PR interval, and **sinus tachycardia(resting)**
- **"Gold standard" for assessing cardiac function in DMD patients is the two-dimensional echocardiogram**
- Hypomotility of the gastrointestinal tract may delay gastric emptying
- Duchenne's dystrophy produces respiratory muscle weakness, which places these patients at increased risk for perioperative pulmonary complications.
  - Progressive decrease in total lung capacity and vital capacity. Inability to cough and clear secretions predisposes these patients to pneumonia
- As the disease progresses, kyphoscoliosis contributes to further **restrictive lung disease**
- 30% of deaths in individuals with DMD are due to respiratory causes

ANESTHESIA MANAGEMENT

- **Preoperative Management**
  - Patients should have a recent 2D Echo and EKG
  - Preoperative pulmonary function studies are valuable in determining the postoperative course of these patients.
    - Patients with a vital capacity >30% of the predicted value can usually be extubated immediately after surgery.
    - Vital capacity < 30% of predicted and the added morbidity of kyphoscoliosis, which can contribute to a restrictive respiratory pattern, postoperative ventilatory support will be required.
  - Delayed pulmonary insufficiency may occur up to 36 hours postoperatively, even if the patient's skeletal muscle strength apparently returned to preoperative level
    - May require a longer period of time on the ventilator

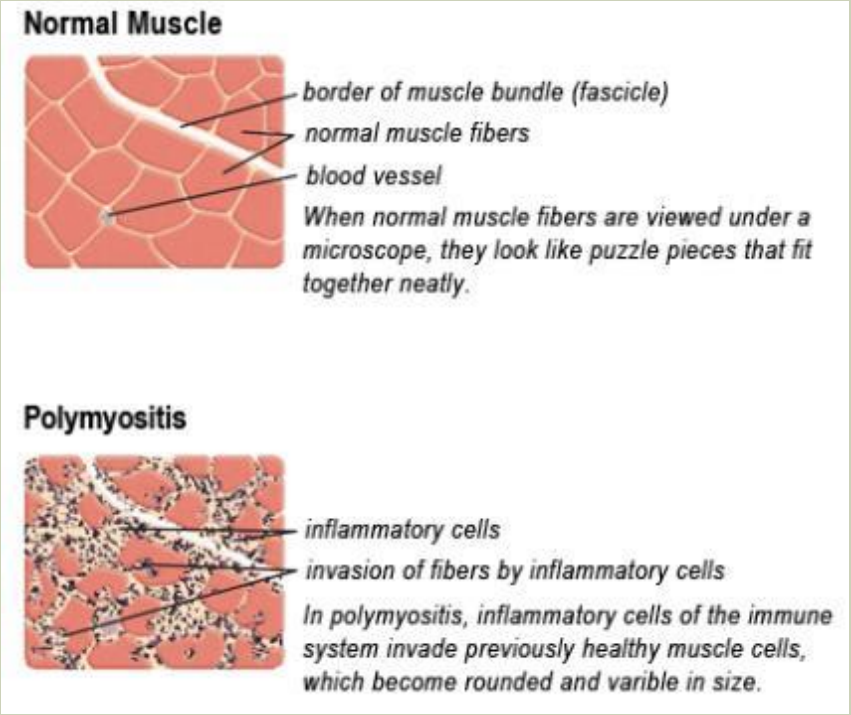
ANESTHESIA MANAGEMENT

- **Intraoperative Management**
  - NDNMB are **OK!**
    - Effects maybe prolonged in these patients
  - Hypomotility of the gastrointestinal tract may delay gastric emptying and, in the presence of weak laryngeal reflexes, can **increase** the risk of pulmonary aspiration
  - Avoid inhalational agents and succinylcholine as both have the potential to lead to rhabdomyolysis, hyperkalemia, and/or cardiac arrest
  - Avoid inhalation agents also due to the increase risk of MH
    - Have dantrolene readily available!
    - If general is required a TIVA is strongly recommended
  - Regional anesthesia if possible is the anesthetic of choice in order to avoid potential complications related to general anesthetics
    - Beneficial with postoperative pain control & pulmonary toileting
  - Monitoring is directed at early detection of malignant hyperthermia
  - Avoid cold/shivering

POLYMYOSITIS

POLYMYOSITIS

- An inflammatory myopathy, multisystem disease of unknown etiology
- **Does NOT affect neuromuscular junction**
- Women > Men
- After 20 years of age
- ***Attention: Not to be confused with "Poliomyelitis"***



POLYMYOSITIS
SIGNS & SYMPTOMS

- Muscle pain and proximal weakness, specifically, neck flexor, proximal arm & leg weakness
- ***Dysphagia, pulmonary aspiration, & pneumonia 2° paresis of pharyngeal & respiratory muscles***
- Diaphragmatic & intercostal muscle weakness → ventilatory insufficiency
- Heart block 2° myocardial fibrosis/atrophy of conduction system, LV dysfunction, & myocarditis can occur
- Often associated w/: SLE, Scleroderma & RA
- Widespread necrotizing vasculitis can be present in childhood forms of the disease (Hines & Marschall, 2012, p.444)

POLYMYOSITIS DIAGNOSIS

Diagnosis

- Based on proximal skeletal muscle weakness, ↑ serum creatine kinase.
- Speed of skeletal muscle destruction parallel ↑ in serum creatine kinase.

- Electromyography studies
- Skeletal muscle biopsies can also support clinical diagnosis

Treatment

- Usually managed w/ corticosteroids
- Immunosuppressive therapy when corticosteroid fails
- Treatment with methotrexate, azathioprine, cyclophosphamide, mycophenolate, or cyclosporine may be useful.
- IV immunoglobulin in refractory cases

POLYMYOSITIS
ANESTHETIC IMPLICATIONS

- Anesthetic management is complex
- Vulnerability to aspiration with general anesthesia
- Use ET tube rather than a LMA to protect airway from potential regurgitation.
- Intubation *not* usually difficult

- Airway Management
  - Mechanical ventilation postoperatively with coexisting muscle weakness and lung disease
  - Mechanical ventilation & corticosteroids when respiratory muscle weakness is accompanied by hypercapnea
  - Aggressive treatment for severe respiratory failure

POLYMYOSITIS
ANESTHETIC IMPLICATIONS

- **Muscle Relaxation**
  - ***Responses to NDNMB & Succinylcholine are NORMAL***

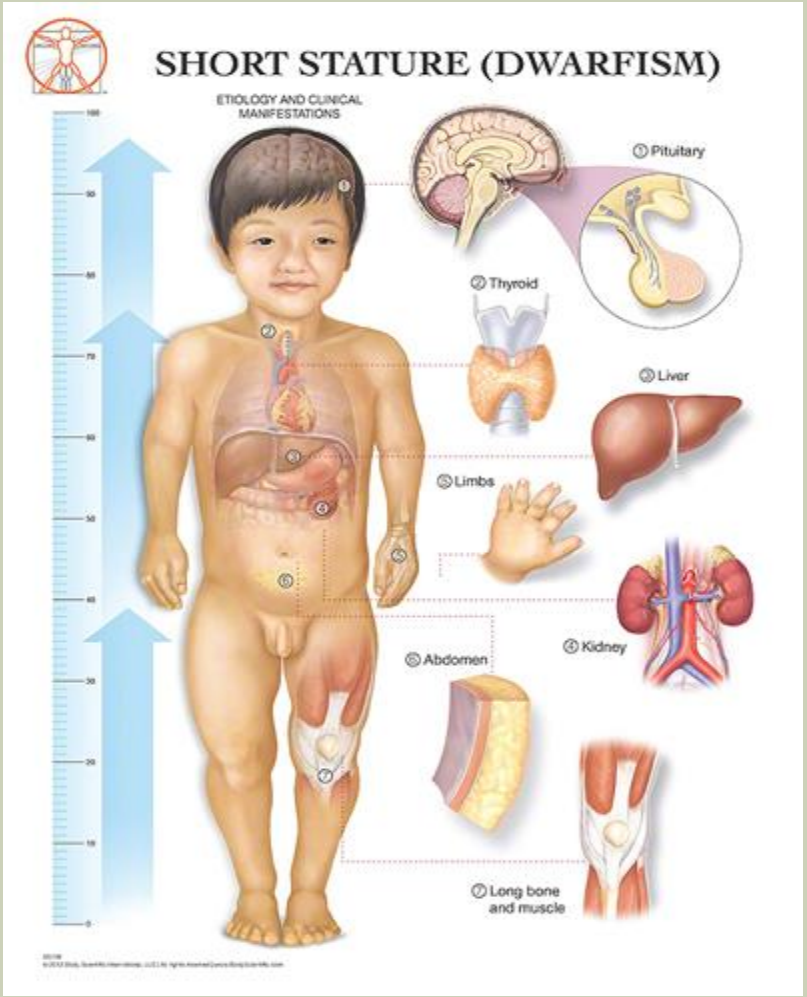
- Choice of Anesthetic Technique
  - Consider degree of coexisting cardiopulmonary disease
    - Preoperative echocardiogram, anesthetic drugs, anesthetic technique & cardiac monitors based on coexisting cardiopulmonary disease
  - Regional anesthesia is of consideration
  - Regional techniques prevent risk and complications associated w/ NMB & inhaled agents

- ***Consider stress dose steroids if on corticosteroids***

DWARFISM

DWARFISM

- Caused by abnormal cartilage cell structure and formation in the epiphyseal growth plate.
- Classified as proportionate & disproportionate
  - Proportionate: the proportions of the limbs, trunk and head size are relatively same as in a normal adult.
  - Disproportionate: features are not proportionate as compared to a normal adult.
- Adult male potential height is approximately 132 cm & adult females are approximately 122 cm.



ACHONDROPLASIA

- Achondroplasia is most common form & is more common in females.
- Results in an abnormality in the size of the trunk, limbs and skull, resulting in short stature.
- Occurs by an autosomal dominant gene transmission with the main underlying cause being a reduction in endochondral ossification rate.

PHYSIOLOGICAL ALTERATIONS

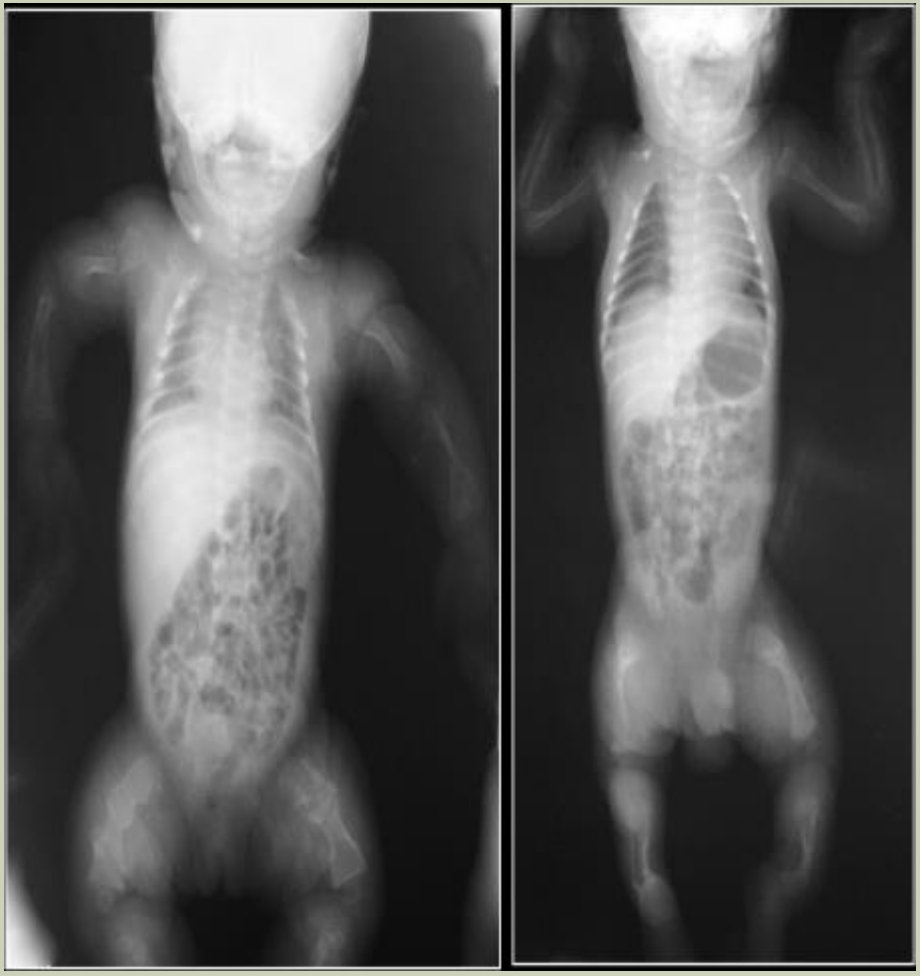
- Shortened stature can result from premature fusion of the bones at the base of the skull.
- Many characteristics are neurologically related:
  - Functional fusion of the atlanto-occipital joint with odontoid hypoplasia
  - Atlantoaxial instability, bulging disks and severe cervical kyphosis also possible
  - Spinal stenosis can present as nerve root compression, cauda equena syndrome, or thoracolumbar kyphosis
  - hydrocephalus, malformation of skull, mental retardation
- Cardiovascular involvement is most commonly related to pulmonary hypertension, which may eventually lead to cor pulmonale.

ANESTHETIC COMPLICATIONS

- Airway Complications:
  - atlantoaxial instability, dental abnormalities
  - obstructive sleep apnea
  - tracheomalacia, laryngomalacia
  - reduced vital capacity and functional residual capacity resulting in ventilation-perfusion mismatching and intrapulmonary shunts.
  - abnormal chest mechanisms
  - restrictive lung patterns
- Strongly consider awake intubation and/or use of fiberoptic bronchoscope
- Avoid any situation where urgent airway management is necessary
- Cases reported of cervical spine injuries and death related to routine airway management

PREOPERATIVE PREPARATIONS

- Chest radiography, electrocardiogram, and transthoracic echocardiography
- Flexion-extension lateral cervical spine radiographs
- Magnetic resonance imaging (MRI) of Computed tompgraphy (CT) in cases where cervical radicular signs are present
- Pulmonary testing is indicated as well as blood gas assays if severe kyphoscoliosis is observed.



INTRAOPERATIVE CONSIDERATIONS

- General and regional anesthesia have been used in this patient population
- Strongly consider endotracheal intubation. Administer neuromuscular blockers only after mask ventilation has proved possible.
- Pressure control ventilation may combat restrictive lung disease
- Routinely assess for stridor secondary to tracheomalacia
- Spinal and regional anesthesia is possible in this population but requires adequate pre-operative assessment

OBSTETRICS

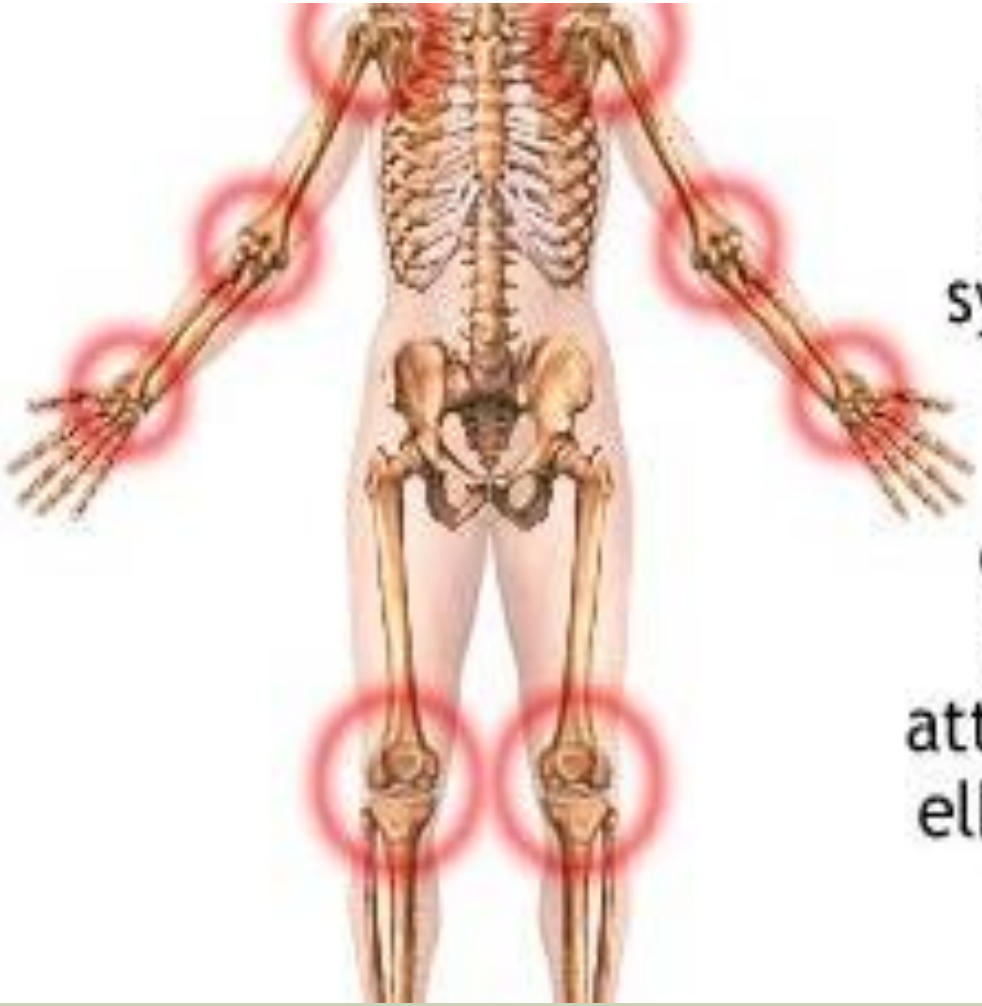
- Higher incidence of cardiorespiratory problems than a normal gravid patient
- Spinal anesthesia is possible but requires adequate pre-operative assessment
- Cesarean section is very common as a greater incidence of cephalopelvic disproportion due to a small pelvis and a normal size infant exists.

RHEUMATOID ARTHRITIS (RA)

RHEUMATOID ARTHRITIS (RA)

- Most common type of chronic inflammatory arthritis causing joint and cartilage deformities
- Etiology is not fully understood
- Approximately 1% of worldwide population is affected
- Incidence= Women > Men
- SYSTEMIC disease with generalized debility:
  - Anemia
  - Muscle wasting
  - Poor nutrition
  - Susceptible to infection, hypothermia

RA PATHOPHYSIOLOGY

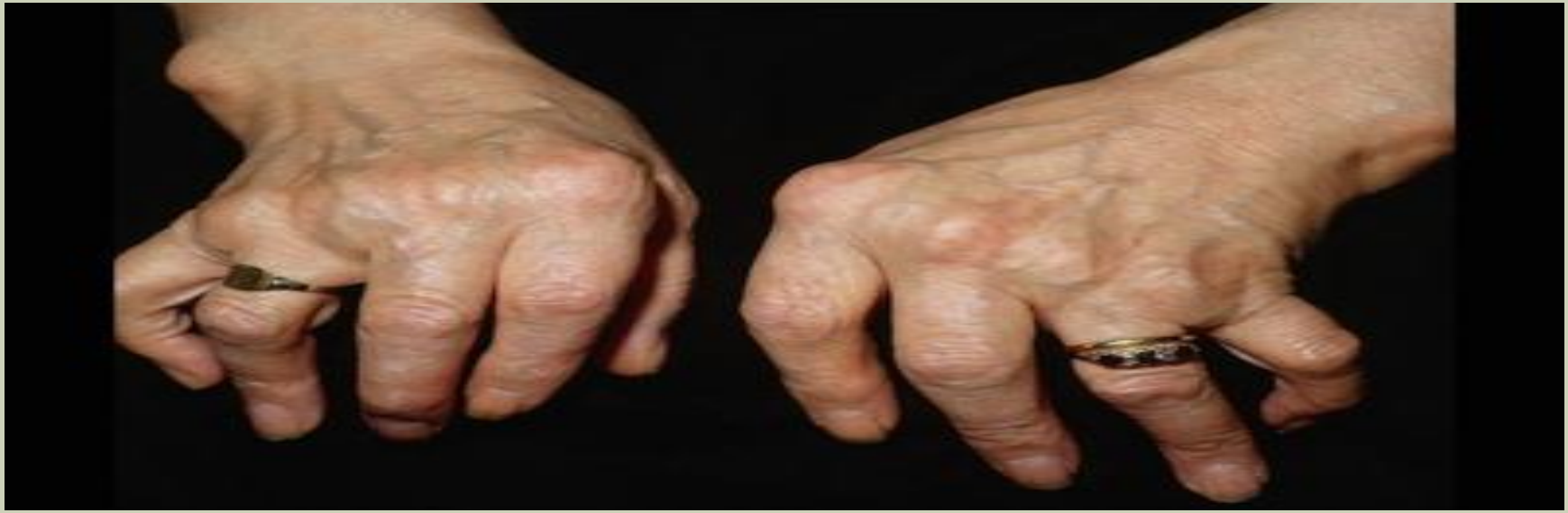


Rheumatoid arthritis usually affects joints symmetrically (on both sides equally), may initially begin in a couple of joints only, and most frequently attacks the wrists, hands, elbows, shoulders, knees and ankles

http://www.healthcentral.com/rheumatoid-arthritis/causes-000048_1-145.html

RA PATHOPHYSIOLOGY

- Systemic disease that presents with exacerbations and remissions
- Usually diagnosed between ages of 30-50 years
- Onset can be insidious or acute
- **Hallmark symptom is morning stiffness**



http://www.essortment.com/initial-symptoms-rheumatoid-arthritis-62402.html

RA DIAGNOSIS & TREATMENT

- Diagnosis:
  - Difficult to dx in early stages
  - ↑ erythrocyte sedimentation rate (ESR)= inflammatory process
  - Rheumatoid factor & anti-cyclic citrullinated peptide (anti-CCP) antibody testing
  - Radiologic studies comparing joints
- Treatment:
  - ASA
  - NSAIDs
  - Steroids
  - Azothiaprine (Imuran)
  - Cyclophosphamide (Cytoxan)
  - Methotrexate

RA ANESTHETIC IMPLICATIONS

- **Airway:**
  - Cervical spine deformities may present a challenge when instrumenting airway
    - Atlantoaxial subluxation- disorder involving C1-C2
    - Avoidance of displacing odontoid process is essential so spinal cord and blood vessels are not affected
  - Cricoarytenoid arthritis may minimize laryngeal view and compromise ability to extubate postoperatively
  - Potentially a difficult airway with advanced disease*
  - Avoid deep extubations

RA ANESTHETIC IMPLICATIONS

- **Pulmonary**:
  - Patients may have **restrictive lung disease**
    - Avoid excessive tidal volumes, may need to ↑ RR to maintain adequate minute ventilation
  - Extended mechanical ventilation may be necessary
  - **Pleural effusions are also common with advanced disease**

RA ANESTHETIC IMPLICATIONS

- **Cardiovascular:**
  - Pericarditis, myocarditis, cardiac valve fibrosis, & nodule formation in cardiac conduction system may cause arrhythmias
  - Chronic anemia may impact oxygenation
  - Chronic ASA and/or NSAID use may increase coagulation times
    - STOP ASA 7-10 days prior to surgery
    - STOP NSAIDS 24 hours prior to surgery

RA ANESTHETIC IMPLICATIONS

- Special care during intraoperative positioning and padding
- Keratonconjunctivitis sicca (dry eyes)
- Xerostomia (dry mouth)
- Mild hepatic and renal insufficiencies
- Avoid hypothermia as this can worsen joint discomfort

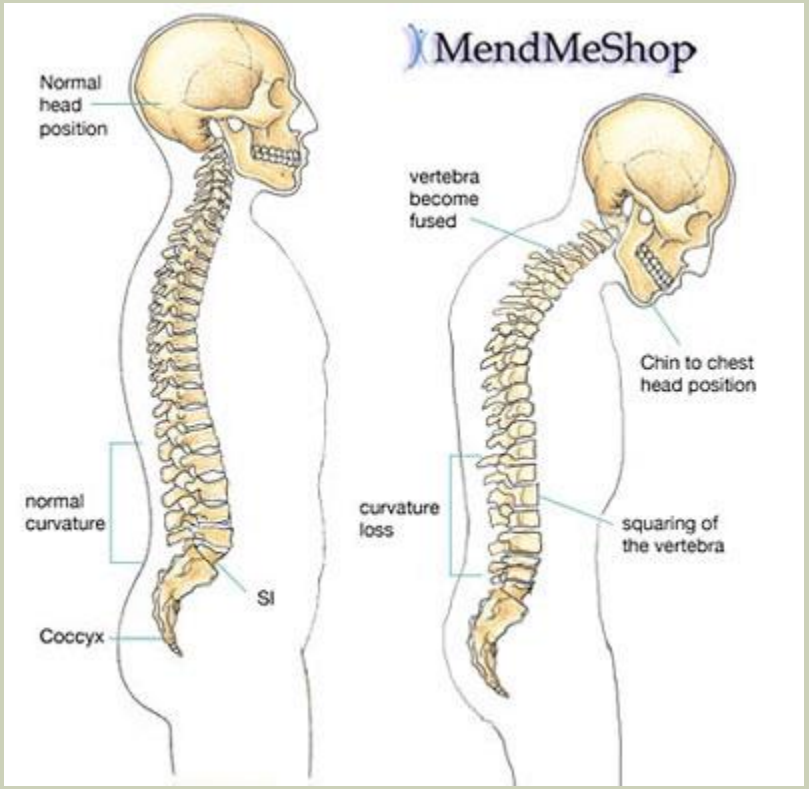
ANKYLOSING SPONDYLITIS

ANKYLOSING SPONDYLITIS

- Ankylosing Spondylitis is a subcategory of Spondyloarthropathies which is a group of nonrheumatic arthropathies
- It is a chronic, usually progressive, inflammatory disease involving the articulations of the spine and adjacent soft tissues
- Spinal disease begins in the sacroiliac joints and moves cranially
- Primarily effects men and often begins in young adulthood

DISEASE MANIFESTATIONS

- Inflammatory changes at the ligamentous insertions onto bone, the vertebrae begin to grow into each other, forming outgrowths known as syndesmophytes
  - This process typically begins at the sacral and lumbar regions, with cervical involvement occurring later in the disease process
- Involvement of the sternocostal, costovertebral, and thoracic spine results in decreased mobility of the thoracic cage and results in a **restrictive ventilatory pattern**
- Extraskeletal manifestations may occur such as; aortic insufficency, cardiac conduction abnormalities, iritis, and pleural effusions



ANESTHESIA CONCERNS

- Assess cardiopulmonary function
- Review radiographic imaging to determine the significance of cervical spine disease before airway management and positioning that require movement of the neck
- Use cautious manipulation of the neck because of instability and mobility limitations
- **These patients should be regarded as potential difficult airway**
- Neuroaxial anesthesia is extremely challenging
- Pay special attention to positioning

ANESTHESIA MANAGEMENT

- Preoperative evaluation should include radiographs of lower and cervical spine to assess the extent of fusion and an assessment of range of motion
- **Decreased range of motion and poor mouth opening can make direct laryngoscopy difficult**
- Atlantoaxial subluxation is also present in some patients
- If advanced disease is present **alternative and conservative airway management is strongly recommended**
  - **i.e. awake intubation**
  - **Alternative airway devices:** LMA, fiberoptic bronchoscope, and videolaryngoscope
- Maintaining spontaneous ventilation is highly recommended
- Neuraxial anesthesia is challenging due to smaller intervetebral spaces
  - Lateral approach may increase chances of success

ANESTHESIA MANAGEMENT

- **Intraoperative management**
  - Pay special attention to positioning!
  - Restrictive airway disease may lead to ↑peak pressures with positive pressure ventilation
  - Due to decreased thoracic cage mobility these patients compensate with increased diaphragmatic excursion, therefore maintaining diaphragmatic excursion is imperative.
    - Avoid trendelenberg position if possible, use a larger ETT when appropriate, avoid interscalene blocks that could lead to diaphragm paralysis
  - **Strict extubation criteria!**
    - Heavy reliance on diaphragmatic excursion increases the risk of postoperative respiratory insufficiency and possible reintubation

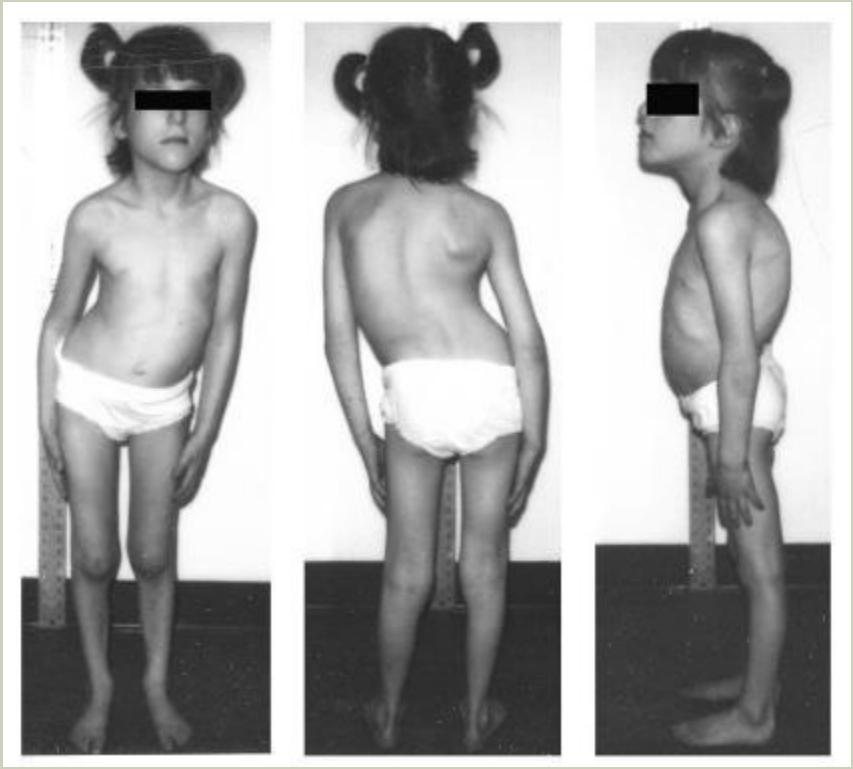
KYPHOSCOLIOSIS

KYPHOSCOLIOSIS

- Spinal deformity with anterior flexion (kyphosis) & lateral curvature (scoliosis) of the vertebral column

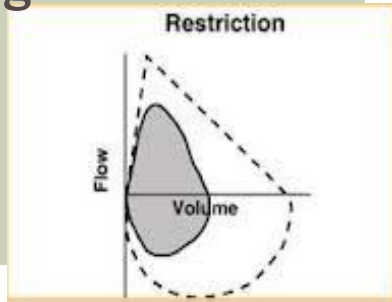
- Idiopathic Kyphoscoliosis
  - 80% of cases
  - Begins during late childhood
  - Can progress in severity during periods of rapid skeletal growth
  - Incidence: 4:1000
  - Familial disposition
  - Females 4x > Males

- Diseases often associated: Poliomyelitis, cerebral palsy, & muscular dystrophy



KYPHOSCOLIOSIS SIGNS & SYMPTOMS

- Spinal curvature >40° considered severe
  - Often associated w/ cardiac & respiratory dysfunction
- RESTRICTIVE LUNG DISEASE & PULMONARY FUNCTION → COR PULMONALE (Primary cause of death)
- As curve worsens, more lung tissue is compressed
  - ↓ vital capacity
  - Dyspnea on exertion
  - ↑ work of breathing 2⁄ abnormal mechanical properties of distorted thorax & ↑ airway resistance from small lung volumes
  - ↑ Alveolar-arterial O_2 difference
  - Pulmonary HTN 2⁄ ↑ PVR from compression of lung vasculature & response to arterial hypoxemia
  - PaCO2 usually maintained
  - Poor cough → frequent pulmonary infection



KYPHOSCOLIOSIS ANESTHETIC IMPLICATIONS

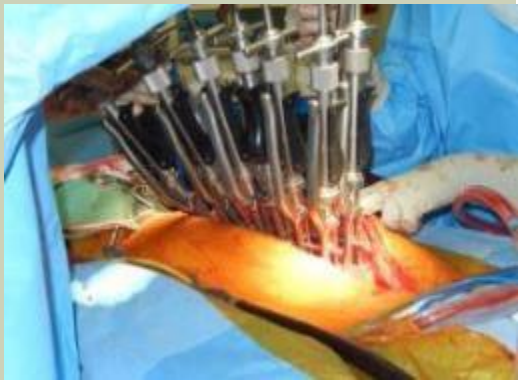
- Preoperative Evaluation
  - Assess severity of scoliosis
  - Review pulmonary function tests to assess degree of restrictive lung disease
  - ABG= helpful to detect unrecognized hypoxemia/acidosis can could → PHTN
  - May have preoperative pulmonary infection 2⌗ chronic aspiration

- Intraoperative Management
  - No specific drug or drug combination recommended
  - N_2O may ↑ pulmonary vascular resistance
    - Want to avoid in patients with PHTN
  - CVP monitoring helpful for monitoring PVR

KYPHOSCOLIOSIS ANESTHETIC IMPLICATIONS

- Considerations For Spinal Curvature Correction Surgery: Potential for large blood loss
  - Risk of spinal cord damage
  - Controlled hypotension
    - To ↓ blood loss
    - Caution 2° risk of ischemic optic neuropathy & spinal cord ischemia
      - Prolonged surgery & low transfusion threshold can ↑ risk of ischemia
  - Intraop, when spinal curvature is straightened, excess traction on spinal cord can → spinal cord ischemia → paralysis

- Monitoring for spinal cord ischemia
  - Wake-up test
  - Motor evoked potentials (MEP)
  - Somato-sensory evoked potentials (SSEP)



SPINAL CORD DISORDERS

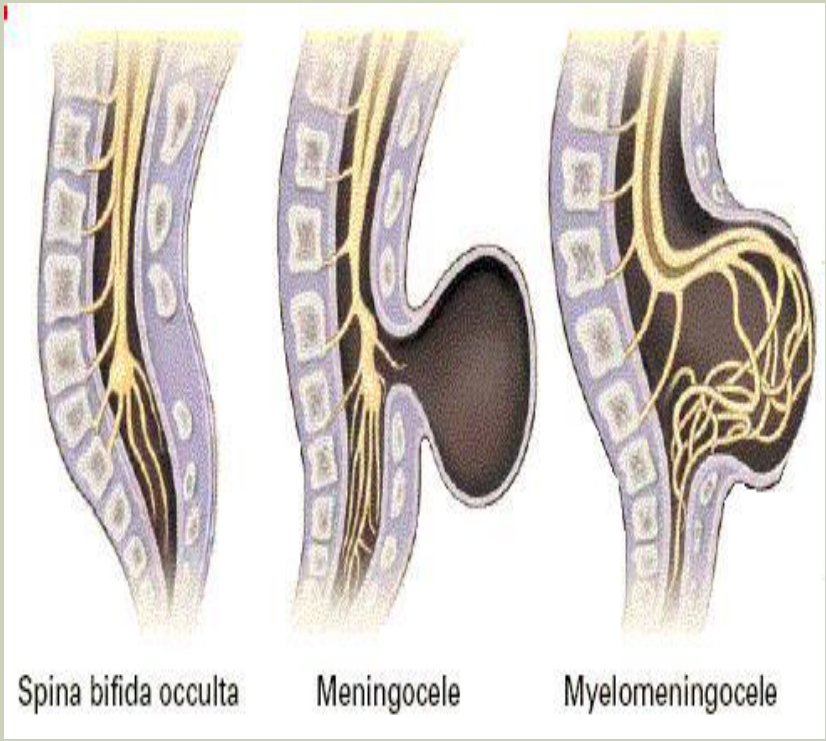
SPINA BIFIDA

- Spina Bifida is most common form of neural tube defect, characterized by a cleft in the spinal column
  - Defect is a result from abnormal fusion of one or more vertebral posterior arches.
- This cleft can be covered by normal-appearing skin, which results in a hidden defect ***spina bifida occulta*** without involvement of the underlying neural structures
- Meninges can herniate through the spinal cleft, creating a CSF-filled sac(***meningocele***) with or without skin covering
- In some cases both the spinal cord and meninges herniate through the spinal cleft(***myelomeningocele***) forming a defect that lacks a skin, and sometimes dural, covering.

SPINA BIFIDA OCCULTA

- Spina bifida occulta (incomplete formation of single lamina in the lumbosacral spine without other abnormalities) is a congenital defect that is present in an estimated 20% of individuals

- Usually produces no symptoms and is typically found by radiographic studies being completed for another disease process



CLINICAL MANIFESTATIONS OF SPINA BIFIDA

- Spina Bifida Occulta
  - Is sometimes discovered only incidentally, because normal skin hides the defect and the mild spinal cleft does not usually cause neurologic deficits. In most cases the overlying skin displays an abnormal lesion such as a dimple, hair patch, dermal sinus tract, hemangioma, or lipoma
- Meningoceles
  - By definition, only the meninges are affected without nerve tissue involvement. For this reason, these patients generally do not have neurologic deficits and are not at noticeable risk of developing long-term neurologic sequelae.
- Myelomeningocele
  - Have varying degrees of motor and sensory deficits as well as bowel and bladder dysfunction. Any damage to the cord and spinal nerves evident at birth is usually irreversible

ANESTHESIA MANAGEMENT

- Is regional an acceptable form of anesthesia in these patients?
  - **YES!** An increased risk with spinal anesthesia is not expected
  - Location of lesion is typically located between L5-S1
- Latex present during the perioperative period?
  - Those with spina bifida have a higher than average risk of latex allergy
  - Why? It is thought to be a reflection of frequent exposure to latex devices
- Positioning is one of the most critical aspects in the perioperative care of a patient with an open lesion(meningocele and myelomeningocele)

QUESTIONS?

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