Neuromuscular Conditions

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Neuromuscular Pathophysiology

Learning Objectives:

- Explain the etiology, pathophysiology, manifestations, diagnosis and general management for the following conditions:
  - myasthenia gravis
  - Guillain Barre syndrome
  - amyotrophic lateral sclerosis
  - muscular dystrophy
  - spinal muscle atrophy
  - critical illness polyneuropathy and myopathy
- Assess weakness of ventilatory apparatus through physical assessment, lung volume measurement, blood gases, inspiratory and expiratory pressures and sleep studies.

Requirements for ventilation

- neural ventilatory drive - stimulus to breathe
- neural transmission to muscles of ventilation
- contractility of ventilatory muscles
- feedback to CNS; e.g., from chemoreceptors

Learning Objectives:

- Apply non-ventilatory techniques to management of neuromuscular conditions.
- Apply mechanical ventilation to neuromuscular conditions.

Requirements for ventilation

- receptors- input to ventilation center
  - brainstem- pH sensor
  - carotid bodies- oxygen
  - lung stretch receptors - mechanics

Requirements for ventilation

- ventilatory drive- centers in medulla and pons (brainstem)
  - medullary ventilatory center- regularity of pattern
  - pneumotaxic & apneustic centers- frequency and tidal volume

Requirements for ventilation

- muscular contraction
  - transmission across myoneural synapse
  - muscular function

Requirements for ventilation

- nerve transmission from centers to ventilation muscles
  - phrenic nerve
    - from C3-C5
    - innervates diaphragm
  - intercostal nerves
    - from T1-T12
    - innervate intercostal muscles

Neuromuscular impairment of ventilation

- CNS (brain, spinal cord) failure
  - trauma
  - infection
  - inflammation
  - toxins
  - drugs
  - ischemia/hypoxia
  - congenital dysautonomia - inborn failure of breathing automaticity

Requirements for ventilation

- nerve transmission from centers to ventilation muscles
  - abdominal nerves
    - thoracic and lumbar spine
    - innervate abdominal muscles

Neuromuscular impairment of ventilation

- nerve transmission failure
  - trauma- transection of spinal cord
    - C1-C3- absence of nerve transmission to all ventilatory muscles
    - C6-C7- absence of transmission to intercostal & abdominal muscles

Neuromuscular impairment of ventilation

- nerve transmission failure
  - iatrogenic- laceration of phrenic nerve
    - occurs during heart surgery
    - may self-reverse (months)
  - infection
    - poliomyelitis
    - West Nile virus

Neuromuscular impairment of ventilation

- muscular failure (congenital)
  - spinal muscle atrophy
  - muscular dystrophy

Neuromuscular impairment of ventilation

- nerve transmission failure
  - degenerative disease
    - amyotrophic lateral sclerosis
    - multiple sclerosis
  - autoimmune disease
    - Guillain-Barre syndrome
    - myasthenia gravis
    - Lambert-Eaton syndrome

Neuromuscular impairment of ventilation

- muscular failure (acquired)
  - polymyositis (inflammation)
  - atrophy; e.g., ventilator-induced diaphragmatic dysfunction (VIDD)
  - fatigue - excessive WOB
  - rhabdomyolysis - breakdown of muscle
  - critical illness polyneuropathy

Neuromuscular impairment of ventilation

- nerve transmission failure
  - toxins - botulism (neuromuscular junction)
  - drugs
    - neuromuscular blockers
    - anticholinesterase agents

Components of ventilatory impairment

- Inspiratory muscle weakness - decreases lung volumes
- Expiratory muscle weakness - decreases expiratory flow (cough)
- Bulbar weakness - muscles of:
  - throat
  - jaw
  - tongue
  - face
  - predisposes to aspiration

Electrophysiology Tests

Nerve conduction studies

- Defined - percutaneous nerve stimulation with surface recording of conduction

Purposes

- Confirm presence of neuromuscular disorder
- Distinguish between nerve, muscle and neuromuscular junction disorders
- Identify specific diagnosis

Nerve conduction studies

- Parameters
  - compound muscle action potential (CMAP) - sum of the response of all stimulated muscle fibers
  - conduction velocity - if decreased, then neuropathy is present
  - F wave - absence indicates demyelination, impairs conduction

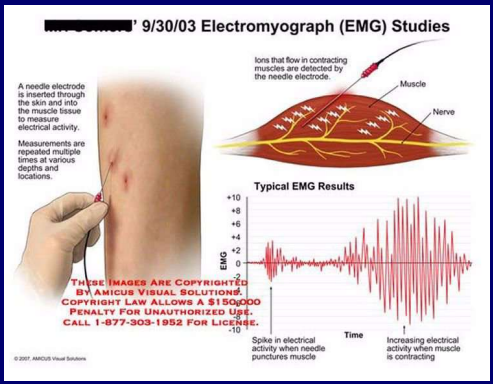
Types

- nerve conduction studies
- needle electromyography
- neuromuscular junction testing
- repetitive nerve stimulation
- single-fiber EMG
- train-of-four stimulation
- phrenic nerve conduction
- needle EMG of diaphragm

Needle electromyography

- Needle inserted in muscle and measures motor unit potentials (MUP) with voluntary contractions
- Decreased duration/amplitude of motor unit potential suggests a myopathy

Needle electromyography



Phrenic nerve conduction

- Technique
  - Surface electrodes, bilaterally on neck stimulate phrenic nerve
  - Diaphragm CMAP measured with electrodes placed at the xiphoid process and costal margin

Repetitive nerve stimulation

- Surface electrodes produce repetitive, strong stimuli
- Early decrement in compound muscle action potential (CMAP) indicates neuromuscular junction disorder; e.g., myasthenia gravis

Phrenic nerve conduction

- Significance
  - bilateral decreased amplitude - neuropathy
  - unilateral decreased amplitude - traumatic or surgical lesion

Train-of-four stimulation

- Used to titrate surgical neuro-muscular blockers
- Four stimuli of ulnar nerve, with observation of thumb twitches - desired is 1 or 2 twitches per 4 stimuli

Click for video of train-of-four testing
http://www.globalrph.com/neuromuscular.htm#Train_of_four

Guillain-Barre Syndrome

Description & demographics

- Group of five autoimmune peripheral neuropathies
  - acute inflammatory demyelinating polyneuropathy (90% of cases)
  - acute motor axonal neuropathy (young people)
  - acute motor sensory axonal neuropathy (uncommon)
  - Miller-Fisher syndrome (rare)
  - acute pandysautonomia (rarest)

Pathophysiology

- Antecedent infection - one to three weeks before GBS onset
- Interaction of pathogen and nerve tissue activates autoimmune response - immunity to self

Description & demographics

- Victim of acute pandysautonomia

Pathophysiology

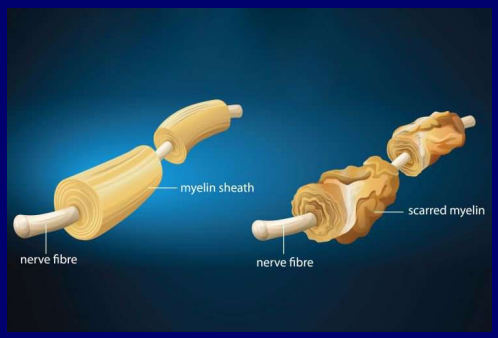
- Antecedent infection - one to three weeks before GBS onset
- Interaction of pathogen and nerve tissue activates autoimmune response - immunity to self
- Autoimmune response produces antibodies that cause demyelination of nerve cells
- Demyelination impairs neural conduction

Epidemiology

- Most common neuromuscular condition for ICU admission
- 1-3 cases/100,000/year
- All age groups
- No gender preference
- Incidence unrelated to current influenza vaccines

Pathophysiology

- Demyelination



Pathophysiology

- Common pathogens
  - campylobacteriosus - most common
  - cytomegalovirus
  - Epstein-Barr virus
  - varicella zoster virus
  - mycoplasma pneumoniae
  - Zika virus

Differential diagnosis

- Myasthenia gravis
- Botulism
- Polymyositis
- Poliomyelitis
- West Nile virus
- Heavy metal intoxication; e.g., arsenic
- Tick paralysis (Lyme disease)
- Porphyria - a hemoglobin disorder

Manifestations

- Severity is variable - weakness to total paralysis with autonomic dysfunction
- Evolves over hours to days
- Ascending paralysis - starts as 'rubbery legs'
- Tingling in extremities

Diagnosis

- Clinical manifestations; e.g.:
  - ascending paralysis
  - areflexia
  - pain
- Laboratory - elevated CSF proteins; 48H after onset

Manifestations

- Does not affect consciousness - patients are alert and frightened
- May involve cranial nerves - bulbar weakness
- Loss of deep tendon reflexes
- Pain in affected muscles; sometimes back pain
- Ventilatory failure in 30% of patients

Diagnosis

- Electrodiagnostic features
  - may be absent in early stages
  - decreased nerve conduction velocity
  - needle EMG - abnormal spontaneous activity
  - normal muscle response to direct stimulation

Management

- Supportive
  - respiratory care
  - nursing care
  - psychological, emotional support

Management

- Plasmapheresis vs. IVIg
  - they are equally effective for GBS
  - IVIg easier to administer
  - IVIg has fewer complications

Management

- Intravenous immunoglobulin (IVIg)
  - neutralizes antibodies
  - five daily infusions

Course, prognosis

- Worst clinical function usually in first week
- Nearly full function at four weeks
- Full recovery within months for 85% of patients
- Relapse occurs in 5-10%
- Mortality <5%

Management

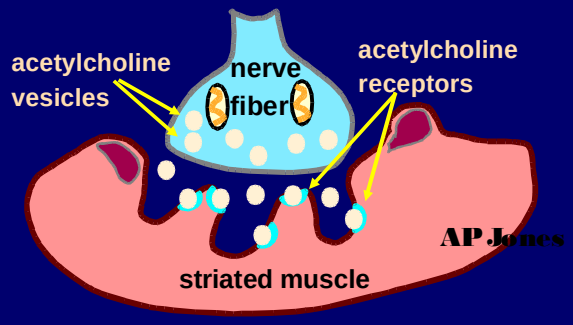
- Plasmapheresis
  - blood removed from patient, plasma separated, then treated to remove antibodies, then returned or replaced with substitute fluid
  - complications (uncommon)
    - hypotension
    - hemorrhage
    - sepsis - immunosuppression

Myasthenia Gravis

Description

- Autoimmune disorder that compromises transmission across the neuromuscular junction.
- Characterized by weakness and fatigability of striated muscles.
- Broad range of severity - from drooping eyelids (ptosis) to ventilatory failure.

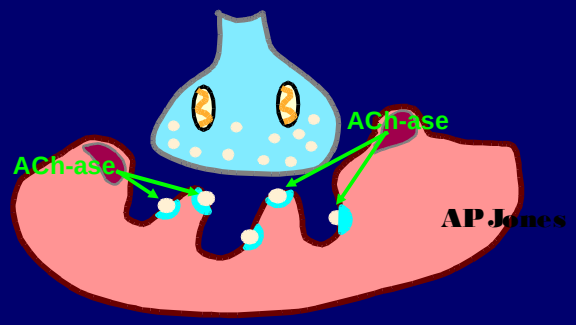
Nerve impulse ==> release of ACh ==> AChR ==> depolarization of muscle



Epidemiology

- 14 cases per million in US (not rare)
- Occurs in all age groups
- Females, peak occurrence - 20-40 YO
- Males, peak occurrence - 40-60 YO
- Congenital form - onset in utero, with decreased fetal movement
- Juvenile form - onset in 20s

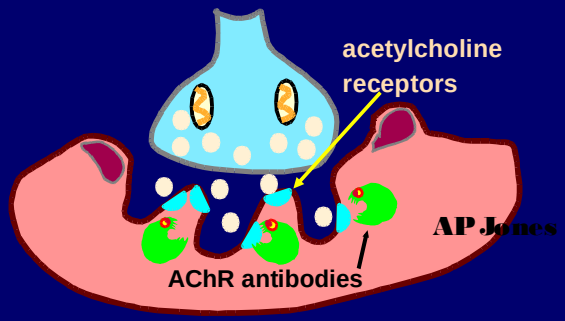
Release of ACh-ase ==> hydrolyzes ACh ==> repolarization of muscle for next contraction



Pathophysiology

- Autoimmune activity against acetylcholine receptors by anti-AchR antibodies

Myasthenia gravis - AChR antibodies decrease available receptors, preventing depolarization



Manifestations

- Does not affect consciousness
- Weakness, initially in ocular muscles
  - ptosis (drooping eyelids)
  - diplopia (double vision)
- Difficulty chewing, speaking, singing swallowing
- Fatigability - improves with rest
- Descending paralysis

Click to see video of MG patient detained by police (2 min)
http://www.youtube.com/watch?v=ypwjG4J7JKM

Diagnosis

- Clinical presentation
- Ice pack test - ice improves eyelid fatigue
- Tensilon (edrophonium HCl) test - anticholinesterase agent increases ACh, increasing ACh-receptor interactions
  - onset 30 sec.
  - duration 5 min.

Click to see video of positive Tensilon test (4.75 min)
https://www.youtube.com/watch?v=qXGwdV1KXnI

Differential diagnosis

- Hypothyroidism
- Medications may induce or exacerbate MG:
  - antibiotics; e.g., aminoglycosides
  - cardiovascular agents; e.g., propanolol
  - anti-malarial agents; e.g., chloroquine
  - miscellaneous agents

Diagnosis

- Electrophysiology - repetitive nerve stimulation
  - rapid decline in amplitude of induced responses
  - specific for neuromuscular junction disorders

Differential diagnosis

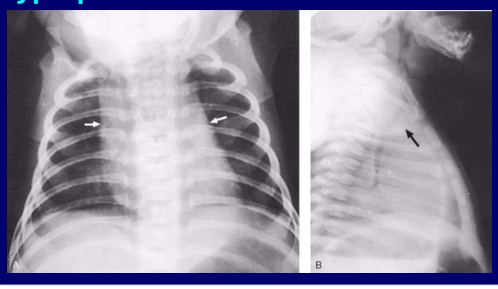
- Hypothyroidism
- Adverse effects of medications
- Amyotrophic lateral sclerosis
- Botulism - strong resemblance
- Guillain-Barre syndrome
- Polymyositis
- Lambert-Eaton syndrome - complication of carcinoma
- Multiple sclerosis

Diagnosis

- Laboratory - AChR-Ab
  - specific for MG
  - false negative results in 50% of MG patients who have only ocular weakness

Diagnosis

- Medical imaging - chest CT with contrast is indicated
  - 20% MG patients have thymoma
  - 70% MG patients have thymic hyperplasia



Management

- Anticholinesterase agents
  - oral pyridostigmine (Mestinon)
  - symptomatic improvement
  - bedside pulmonary function testing by RTs for dosage, duration of action

Management

- Supportive - severe weakness (myasthenic crisis) may require intubation, ventilation

Management

- Cholinergic crisis - side effect, due to overdose with pyridostigmine
  - manage with atropine

Management

- Evaluate for thymectomy
  - improvement (delayed) in 85%
  - remission in 35%
  - recommended for all MG patients between puberty and 55 YO
- Thymectomy - should be done in center with experience

Management

- Immunosuppression
  - severe weakness
    - plasmapheresis
    - IVIg
  - moderate weakness
    - glucocorticoids - improvement in most patients
    - azathioprine
    - mycophenolate mofetil
    - cyclosporine

Amyotrophic Lateral Sclerosis

Epidemiology

- Possible risk factors:
  - smoking
  - oxidative stress
  - head injury - soccer players

Description

- AKA 'Lou Gehrig's disease'
- Devastating progressive neurodegenerative condition, causing:
  - painless weakness
  - muscular atrophy

FYI - Click to see Lou Gehrig video (.5 min)
http://www.youtube.com/watch?v=_LFprYLW3_U

Pathophysiology

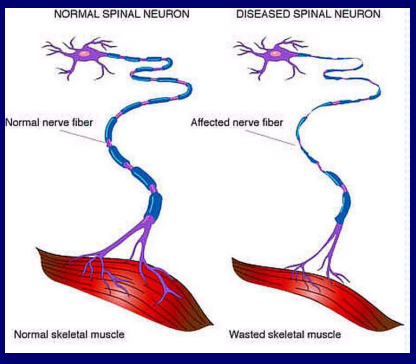
- Death of motor neurons:
  - anterior horn cells
  - brainstem motor neurons
  - corticospinal motor neurons
- Denervation causes muscular atrophy (amyotrophy)

Epidemiology

- Most common progressive neurodegenerative disease
- Incidence - 2 cases /100,000 people
- 5-10% of cases are inherited as autosomal dominant trait - familial ALS
- Male:female = 1.4:1
- 90% of ALS patients > 40 YO
- 5% of ALS patients < 30 YO

Pathophysiology

- Death of motor neurons



Manifestations

- Does not affect sensation or consciousness
- Manifestations vary, depending on which neurons are initially involved
  - lower neurons - limbs
  - bulbar neurons - face, tongue

Prognosis

- Common - death within 3 years of onset
- 20% survive 5 years
- 10% survive >10 years

Manifestations

- Does not affect sensation or consciousness
- Manifestations vary, depending on which neurons are initially involved
- Weakness
  - slow onset
  - asymmetric
  - starts distally in one limb
  - can start anywhere, progressing to everywhere

Differential diagnosis

- Importance - ALS has no cure; so, alternatives that may be treatable must be ruled out before accepting ALS as diagnosis.

Manifestations

- Cramping
- Fasciculation (twitching)
- Exaggerated motor expressions of emotion (laughing, crying)
- Progressive paralysis
- Difficulty swallowing
- Ventilatory failure

Differential diagnosis

- Other motor neuron diseases
- Structural disorders; e.g., CNS tumor, CNS radiation injury
- Toxic disorders; e.g., heavy metals
- Immune, inflammatory disorders; e.g,
  - multiple sclerosis
  - myasthenia gravis

Differential diagnosis

- Parkinson disease
- Thyrotoxicosis
- Infections; e.g.,
  - Lyme disease
  - syphilis
  - Creutzfeldt-Jakob disease- prion infection

Management

- Riluzole
  - glutamate antagonist
  - extends life by 2 months (average)
- Edavarone- FDA approved 2017
  - antioxidant
  - outcomes similar to riluzole
- Stem cell therapy - research in early stages

Diagnosis

- Clinical presentation
  - progressive course of weakness
  - hyperreflexia in weak, atrophied extremity
- Electrodiagnostics
  - denervation
  - fasciculation potentials

Muscular Dystrophy

Management

- Supportive
  - respiratory care
  - nutrition
  - physical therapy
  - nursing
  - end-of-life care

Description

- Muscular dystrophy - a group of hereditary, progressive conditions that cause muscle fiber degeneration.

Description

- Duchenne muscular dystrophy (DMD)
  - most common
  - X-linked recessive trait
  - males only

Pathophysiology

- Genetic defect for production of dystrophin
  - needed for muscle cell membrane integrity
  - deficiency permits leakage of muscle cell components ==> muscle cell death

Click to see video on histopathology of MD (3 min.)
http://www.youtube.com/watch?v=xwS3FtmvlRk

Description

- Becker MD
  - second most common
  - milder than DMD
  - X-linked recessive - males only
- Limb girdle MD - males & females
- Emery-Dreifus MD - males & females

Manifestations

- Usual onset of frequent falling at 3-5 YO
- Gait abnormalities; e.g., toe walking
- Gower sign - arising from sitting position by going prone
- Inability to ambulate 7-13 YO
- Contractures

Epidemiology - DMD

- 1 case per 3500 live male births
- 1/3 of cases are spontaneous mutations ==> mom was not born with the defective gene

Manifestations

- Progressive ventilatory failure
  - muscle weakness
  - kyphoscoliosis - restrictive lung dx
  - recurrent pulmonary infections

Manifestations

- Progressive ventilatory failure
  - muscle weakness
  - kyphoscoliosis - restrictive lung dx
  - recurrent pulmonary infections
- Cardiomyopathy - major cause of death
- Cognitive deficit - IQ about 85
- Death before 30 YO, due to cardio-pulmonary failure

Management

- Supportive
  - respiratory care - lots of it
  - nutrition
  - physical therapy
  - nursing
  - end-of-life care

Diagnosis

- Clinical presentation
  - frequent falling
  - gait abnormalities
  - Gower sign
- Electromyography - decreased action potential amplitude

Management

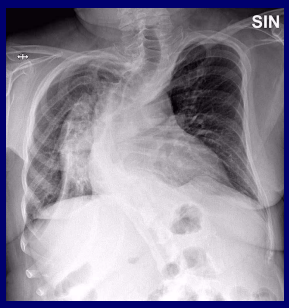
- Special problem - kyphoscoliosis renders chest imaging difficult
  - standard chest radiographs - difficult to position and interpret
  - echocardiography - poor images

Diagnosis

- Laboratory studies
  - elevated creatine phosphokinase (CPK) - DMD specific
  - muscle biopsy for histology
  - gene testing
    - prenatal diagnosis
    - carrier identification

Management

- Special problem - kyphoscoliosis renders chest imaging difficult.



Management

- Cardiac care
  - problems
    - cardiac dysfunction is masked by other limitations
    - dysrhythmias
    - ventricular failure
    - thromboembolic events

Management

- Steroids - prednisone
  - prolongs walking
  - improves cardiac function
  - but -- side effects

Management

- Cardiac care
  - early cardiac evaluation
  - diuretics, as indicated
  - angiotensin converting enzyme (ACE) inhibitors, as indicated
  - beta blockers, as indicated
  - anticoagulants, as indicated

Management

- Interventions under study
  - PTC 124 (Ataluren)
    - interferes with expression of defective genes
    - FDA orphan status for DMD & CF
  - gene replacement
  - stem cells - replace dystrophin

Management

- Surgical procedures
  - surgical release of severe contractures
  - spinal fusion for kyphoscoliosis

Spinal Muscle Atrophy

Description

- An autosomal recessive hereditary disease characterized by progressive hypotonia (floppiness) and muscular weakness.

Pathophysiology

- Absence of a neuron survival gene allows programmed cell death (apoptosis)
- Progressive degeneration of motor neurons from anterior horn cells in the spinal cord.

Description

- SMA major types:
  - Type I (Werdnig-Hoffmann disease) - identified in patients from birth to age 6 months.
  - Type II chronic infantile SMA - diagnosed in infants aged 6-12 months.
  - Type III (Kugelberg-Welander disease) - diagnosed in children aged 2-15 years.

Manifestations

- Type I - shows at birth to 6 mo.
  - suspected with decreased fetal activity
  - floppy (hypotonic) newborn
  - unable to control head or roll over
  - tongue fasciculations - cardinal sign

Epidemiology

- The most common degenerative neurologic disease in children
- The leading heritable cause of infant mortality
- Incidence - 1 case per 15,000-20,000
- 3-10 times more common in North Dakota
- Male:female = 2:1

Manifestations

- Type II - shows at 6-12 mo.
  - weakness in lower extremities
  - able to control head and sit up
  - upper extremity tremors
  - tongue fasciculations - cardinal sign

Manifestations

- Type III - shows at 2-15 yrs.
  - tongue fasciculations - cardinal sign
  - can walk early in life, then weaken
  - wheelchair bound by 40 YO
- SMA does not affect cognition

Prognosis

- Type I SMA
  - respiratory management has increased longevity
  - 1 YO without ventilation
  - >10 YO with noninvasive ventilation
- Type III SMA - may have normal life span

Differential diagnosis

- Cerebral palsy
- Muscular dystrophy
- Myasthenia gravis
- Polymyositis
- Inflammatory polyneuropathy
- Infant botulism - honey

Management

- Supportive
  - respiratory care
  - nutrition
  - physical therapy
  - nursing
  - end-of-life care - especially for parents

Diagnosis

- Clinical presentation
  - progressive course of weakness
  - tongue fasciculations
- Electrodiagnostics
  - fasciculation potentials
  - denervation
- Laboratory - prenatal DNA testing

Critical Illness Polyneuropathy & Myopathy

Description

- First described in 1986
- Critical illness polyneuropathy (CIP) and critical illness myopathy (CIM) are complications of critical illness characterized by limb weakness and prolonged ventilator weaning.
- AKA - Intensive care unit-acquired weakness (debilitation)

Pathophysiology

- Complex and unclear
- The neuromuscular system may be an additional organ in multiple organ failure (MOF)
- Muscle inactivity ==> atrophy

Epidemiology

- Incidence - patient experiencing:
  - sepsis or systemic inflammation - 70%
  - multiple organ failure - 100%
  - ventilation for 4-7 D - 25-33%
  - ICU for 7 D - 49-77%

Pathophysiology

- Disruption of neuromuscular microcirculation by:
  - hyperglycemia
  - cytokines
- decreased anabolic hormones
- increased catabolic hormones

Epidemiology

- Additional risk factors
  - corticosteroids
  - neuromuscular blocking agents
  - aminoglycosides; e.g., gentamycin
  - hyperglycemia
  - parenteral nutrition (TPN)

Manifestations

- Muscle weakness, paralysis
- Absent deep-tendon reflexes
- Prolonged weaning from ventilator
- Longer duration of stay in ICU and hospital; therefore:
  - increased costs ($66,000./patient)
  - increased risk for morbidity; e.g., VAP

Differential diagnosis

- Myasthenia gravis
- Guillain-Barre' syndrome
- Amyotrophic lateral sclerosis
- Polymyositis
- Multiple sclerosis
- Et cetera

Management

- Intensive insulin therapy - maintain glucose 80-110 mg/dL
- Avoidance of potential triggers; e.g., steroids, neuromuscular blockers
- Mobilization in ICU

Diagnosis

- Primary aim is to rule out other diagnoses that may be treatable
- Clinical presentation
  - generalized weakness
  - weaning difficulty
- Electrodiagnostics - results may not justify the costs
- Laboratory - muscle biopsy to exclude myopathy

Respiratory Assessment For Weakness

Management

- Supportive
  - respiratory care
  - nutrition
  - nursing
  - physical therapy for recovery

Respiratory muscle evaluation

- Important point - respiratory muscle weakness can be detected by bedside evaluations before any blood gas abnormality is present.

Physical signs

- rapid, shallow breathing - early sign
- accessory muscle recruitment
  - sternocleidomastoids
  - scalenes
  - external intercostals
  - abdominal - active expiration
- abdominal paradox - diaphragmatic fatigue

Lung volumes and flows

- Lung volumes - restrictive pattern with initial normal compliance
  - decreased VC, TLC
  - normal FRC & RV
  - worsening with kyphoscoliosis - true restriction

Blood gas analysis

- Oxygenation deficits - advanced disease
  - hypoventilation
  - ventilation-perfusion defects
    - atelectasis
    - pneumonia

Lung volumes and flows

- Comparison of erect, vs. supine VC
  - can be done at bedside
  - normal supine is 10% less than erect
  - greater difference ==> muscle weakness

Blood gas analysis

- Hypercapnia - advanced disease
  - weakness ==> rapid, shallow breathing
  - decreased sensitivity to CO_2 (important)

Lung volumes and flows

- Lung volumes are not sensitive measures of respiratory muscle strength
- Lung volumes are compromised by reduced compliance; inspiratory pressure is not compromised - pressures are better indicators of strength

Lung volumes and flows

- Expiratory flow rates
  - decreased FEV_1
  - normal FEV_1/FVC
  - decreased PEF ==> impaired cough effectiveness
  - peak cough flow >160 L/min - effective cough for adults

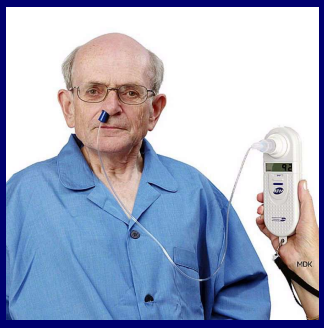
Sniff nasal inspiratory pressure (SNIP)

- manometer connected via tube to one nostril
- other nostril remains open
- patient instructed to sniff maximally

Inspiratory/expiratory pressures

- PI_{MAX}
  - should be measured at FRC
  - large range for normals
  - sources of error
    - patient effort
    - leaks
    - technique - need to standardize

Sniff nasal inspiratory pressure (SNIP)



Inspiratory/expiratory pressures

- PE_{MAX}
  - should be measured at TLC
  - reflects coughing capability

Sniff nasal inspiratory pressure (SNIP)

- Advantages
  - natural maneuver - easy to do
  - reliability better than PI_{MAX}
  - validated for all age groups
- Prognostic information - SNIP < 40 cm H2O ==> nocturnal hypoxemia and risk for mortality (ALS patients)
- Predicted values - based on age and gender

Sleep studies (polysomnography)

- Rationale
  - to detect sleep-related hypo-ventilation
  - determine need for nocturnal non-invasive ventilation

Quality of life and decisions

- Noninvasive ventilation has increased survival time for NM patients
- Patients have their own perceptions on quality-of-life - likely to be better than we might think

Sleep studies (polysomnography)

- Baseline study - early in course of dx
- Follow-up
  - appearance of signs of sleep apnea
  - daytime hypercapnia
  - nocturnal desaturation
  - periodically, to evaluate therapeutic regimen

Quality of life and decisions

- Noninvasive ventilation has increased survival time for NM patients
- Patients have their own perceptions on quality-of-life - likely to be better than we might think
- Decisions about support should be made before any crisis
- Decisions should involve a health-care team, patient and family

Respiratory Management

Mechanical ventilation

- Noninvasive ventilation indicated for:
  - sleep disordered breathing
  - hypercapnia
  - pulmonary infections
  - perioperative management
  - pregnancy
  - palliative, end-of-life care

Non-ventilatory care

- Goal - delay need for ventilatory support
- Problem
  - ventilatory muscle weakness impairs cough ==>
  - secretion retention ==> recurrent pneumonia

Non-ventilatory care

- Treatment strategies
  - manual cough assistance - tussive squeeze
  - in/exsufflator - indicated for MEP < 60 cm H2O
    - positive pressure for inflation
    - negative pressure for increased expiratory (cough) flow
    - usual pressures 40 to -40 cm H2O
    - may reverse atelectasis
    - improves symptoms and SPO2

Non-ventilatory care

- Treatment strategies not used
  - percussion and postural drainage - not routinely used
  - bronchodilators - not indicated and may be harmful
  - aerosolized mucolytics - ineffective
  - deep suctioning

Therapeutics

- In/exsufflator - cough assistance

Image Courtesy of Philips Respironics

Non-ventilatory care

- Treatment strategies
  - manual cough assistance - tussive squeeze
  - in/exsufflator device - indicated for MEP < 60 cm H2O
    - positive pressure for inflation
    - negative pressure for increased expiratory (cough) flow
    - usual pressures 40 to -40 cm H2O

Mechanical ventilation

- Goals for ventilation
  - increase survival time
  - rest ventilatory muscles
  - increase chemoreceptor sensitivity - decreases daytime PCO2
  - improve sleep

Mechanical ventilation

- Negative pressure ventilation
  - as effective as NIPPV
  - cumbersome, difficult
  - types:
    - iron lung
    - cuirass; e.g., Pneumo-Wrap
    - Hayek™ RTX (cuirass)

Mechanical ventilation

- Continued airway and ventilatory care
- Support for communication
- Education for home care
- End-of-life support

Mechanical ventilation

- Noninvasive positive-pressure ventilation (BiPAP)
  - treatment of choice for NM disease
  - progression - nocturnal to intermittent daytime use
  - mouthpiece during daytime
  - limitation - pressure injury from masks with continuous use

END

Mechanical ventilation

- PPV with tracheostomy
  - decision to continue support
  - required when:
    - risk for aspiration increases
    - NIPPV becomes ineffective
    - patient develops intolerance or injury from face mask

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