The Muscles
Normal Structure — Skeletal Muscle

- Striated muscle fibers
- Primary function is to contract
- Contractile proteins: actin and myosin
  - Long, extensible and therefore return to normal length after muscular contraction
- Neuromuscular junction (NMJ)
  - Site of contact with motor neuron axon
  - Acetylcholine (ACh) — excitatory neurotransmitter
    - Causes depolarization and muscular contraction
  - Cholinesterase- enzyme that removes ACh
    - Allows muscle to relax
Normal Structure — Skeletal Muscle

- **Fibers**
  - **Type I: Slow twitch**
    - High resistance to fatigue (fatigue resistant)
    - Used in low level activity: aerobic, daily activities
    - Endurance events (marathon)
  - **Type II: Fast twitch**
    - Fast twitch — A
      - Intermediate resistance to fatigue
      - Long-term anaerobic activity (mile run, 400 meter swim)
    - Fast twitch — B
      - Low resistance to fatigue
      - Short-term anaerobic activity (100 meter dash, 50 meter swim)
Fibers (cont.)

- Type of fiber depends solely on nerve impulses!
- Can switch from one type to another
  - Innervation changes
Functions

- Contraction
  - Enables movement
  - Generates heat
  - Breathe
  - Maintain posture

- Metabolite storage site
  - Glycogen, fat
Facts to know

1. Muscle and peripheral nerves are a single unit
2. Transmission of nerve stimuli depends on binding of Ach
3. Muscle function depends on structural components
4. Muscle function depends on minerals (calcium, potassium)
5. Affected by hormones (thyroid, adrenal insulin)
Facts to know

6. Affected by autoimmune disorders (SLE, MG, RA, dermatomyositis)

7. Destruction of muscle fibers causes release of muscle specific enzymes (creatine kinase (CK))

8. Cannot regenerate properly
Terminology

- Weakness – inability to adequately contract
- Fatigability – inability to sustain action
- Tetany - muscle spasm
- Fibrillation – uncoordinated and irregular contraction of groups of fibers
  - Like a rapid muscle twitch
- Myalgia – muscle pain
Terminology

- **Hemiplegia**
  - Muscle paralysis on one side of the body

- **Paraplegia**
  - 2 extremities and +/- trunk paralyzed

- **Quadriplegia**
  - All 4 extremities and trunk paralyzed
Terminology

- **Sprain**
  - Stretching or tear in a ligament

- **Strain**
  - Stretching or tear in a musculotendinous unit

- **Avulsion**
  - Ligamentous or tendinous detachment/separation from bone
Pathology
Neurogenic Atrophy

- Atrophy caused by injury to nerves
  - Upper motor neuron
    - In the central cortex
    - Transection of the spinal cord (SCI)
    - Strokes, hemorrhage
    - Permanent and irreversible
  - Lower motor neuron
    - Anterior horn cell of spinal cord
    - Transection of the entire nerve
    - Poliomyelitis
Neurogenic Atrophy
Myasthenia Gravis (MG)

- Autoimmune disease involving the NMJ
  - Impaired nerve impulse transmission

- Etiology
  - Unknown
  - Auto immune disorder

- Rare
  - ♀ before age 40
  - ♂ over age 60
IMPAIRED TRANSMISSION IN MYASTHENIA GRAVIS

**NORMAL NEUROMUSCULAR TRANSMISSION**

1. Motor nerve impulses travel to motor nerve terminal.
2. Acetylcholine (ACh) is released.
3. ACh diffuses across synapse.
4. ACh receptor sites in motor end plates depolarize muscle fiber.
5. Depolarization spreads, causing muscle contraction.

**NEUROMUSCULAR TRANSMISSION IN MYASTHENIA GRAVIS**

1. Motor nerve impulses travel to motor nerve terminal.
2. ACh is released.
3. ACh diffuses across synapse.
4. ACh receptor sites, weakened or destroyed by attached antibodies, block ACh reception.
5. Depolarization and muscle contraction don’t occur. Neuromuscular transmission is blocked.
Myasthenia Gravis (MG)

- **Clinical symptoms**
  - Skeletal muscle weakness and fatigability
  - Extraocular muscle and facial muscle weakness
    - Ptosis and diplopia
    - Bland facial expression
  - Muscle weakness
    - Proximal musculature first
  - Muscle fatigability
  - Death due to respiratory compromise
  - Thymus enlargement
  - Speech abnormalities
Myasthenia Gravis (MG)
Myasthenia Gravis (MG)

- **Treatment**
  - Symptomatic (because disease incurable)
  - Inhibition of acetylcholinesterase (AChE)
    - AChE = enzyme that degrades ACh
    - Inhibition of AChE floods the neuromuscular junction with Ach
  - Plasmaphoresis
    - Removes antibodies from blood
    - Temporary relief
  - Thymus enlargement
    - Thymectomy
Myasthenia Gravis

- **PT**
  - No aggressive strengthening
    - Will cause more weakness
  - Symptomatic treatment
    - Endurance
    - Functional activities
  - Symptoms fluctuate throughout the day
    - Plan therapy at max energy time
Muscular dystrophy

- Group of muscle diseases characterized by
  - Primary muscle cell pathology of genetic origin
  - Progressive course
  - Symptoms related to muscle wasting
Muscular dystrophy

- Includes
  - Duchenne-type (girdle)
  - Becker’s (girdle, milder form)
  - Limb-girdle (shoulder, girdle)
  - Fascioscapulohumeral (face, shoulder)
  - Myotonic (eyelids, face, distal limbs)
Differences Between Types of Muscular Dystrophy

- Mode of inheritance
- Age at onset
- Muscle groups affected
- Severity of disease
Muscular dystrophy

- Duchenne-type
  - Most common type
  - Rapid progression
    - Typically loss of ambulation by 9-10 y/o
    - Death usually in the 20s
  - Pathogenesis
    - Lack of dystrophin in skeletal muscles
      - Muscle cell degeneration and loss
      - Compensatory hypertrophy of viable fibers
      - Ingrowth of fibrous tissue, and fat cells which replace lost fibers
Muscular dystrophy

- Duchenne-type
  - Clinical features
    - Progressive wasting of muscles
      - Proximal (hip girdle, lower extremities, neck flexors)
      - Gower’s sign
        - Difficulty getting up off the floor
    - Hyperlordotic, wide-based, waddling gait
    - Hypertrophy of weak muscles (neck extensors, PFs)
    - Contractures (heel cord, ITB, hamstrings, iliopsoas)
      - Walk on toes due to calf contractures
    - Myocardial weakness (signs of heart failure)

Adopted from: Jean Flickinger, *Childhood Neuromuscular Disorders*, USIP Department of PT, 09/05/06
Muscular Dystrophy
Duchenne-Type

shoulders and arms are held back awkwardly when walking

swayback

weak butt muscles (hip straighteners)

Knees may bend back to take weight.

thick lower leg muscles (the 'muscle' is mostly fat, and not strong)

tight heel cord (contracture); child may walk on toes

weak muscles in front of leg cause 'foot drop' and tiptoe contractures.

belly sticks out due to weak belly muscles (child is poor at sit-ups)

thin, weak thighs (especially front part)

poor balance; falls often

awkward, clumsy if walking

http://www.dinf.ne.jp/doc/english/global/david/dwe002/dwe002g/dwe00212g01.gif
Muscular Dystrophy

- **Duchenne-type**
  - **Medical Management**
    - Scoliosis
      - Spinal fusion
    - Tendon lengthening
      - Decreases falls, prolongs ambulation
    - Steroids
      - Improved strength and function
    - Cardiac
      - Cardiac myopathy, fibrosis, conduction abnormalities

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Muscular Dystrophy

Duchenne-type: PT

- All activities play related – they are still children!
- NO AGGRESSIVE STRENGTHENING
  - Will cause muscle cell death
- Submaximal endurance training
- Aquatic therapy
- Equipment prescription and training

- Scoliosis
  - Pain, positioning, respiratory compromise
- Tendon lengthening
  - Serial casting, bracing
- Respiratory compromise
  - Incentive spirometer, respiratory muscle retraining

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Myopathies

- Used to describe nonspecific muscle weakness secondary to an identifiable disease of condition.
- Causes include many metabolic and hormonal diseases, autoimmune diseases.
- Classified as hereditary or acquired.
- Characterized by progressive muscle weakness with pain and tenderness.
- Treatment directed at cause.
Myositis

- = inflammatory muscle diseases

- Infectious or immune
  - Infectious
    - HIV, bacteria, viruses, protozoa, worms
    - Often caused by S. aureus and parasites (Taenia solium)

Myositis

- **Immune**
  - **Polymyositis**
    - limited to muscles
  - **Dermatomyositis**
    - not limited to muscles; involves other organs
  - **Myositis of SLE**
    - Most common around blood vessels
    - Vessels narrow, cause muscle cell atrophy
  - **Sarcoidosis**
    - Systemic disease; type IV hypersensitivity reaction

Myositis

- **Pathogenesis**
  - Inflammatory changes causing damage ranging from significant functional losses to minor self-limiting conditions
  - If left untreated, risk of tissue necrosis or muscle tissue damage
  - Dermatomyositis and polymyositis
    - Chronic inflammation of the muscles
    - Infiltrated muscles attempts to regenerate
      - Hypertrophy of unaffected fibers occurs

Myositis

- Clinical features
  - Symptoms observed with the inflammatory process
  - Pain
  - Muscle weakness usually bilateral; proximal > distal
  - Malaise
  - Fever
  - Muscle swelling
  - Tenderness
  - Lethargy

Myositis

- Clinical features (cont.)
  - Dermatomyositis and myositis
    - Dysphagia, vasculitis, Raunaud phenomenon, cardiomyopathy, interstitial pulmonary fibrosis
  - Dermatomyositis
    - Purple skin rash on eyelids, face, chest, extensor surfaces of extremities
    - Eyelid edema

Dermatomyositis
Myositis

- **Medical Management**
  - Aggressive early treatment
  - Immunosuppressive therapy

- **PT**
  - Presentation: Muscle weakness and extensive skeletal muscle damage
  - Submax exercise
    - No eccentrics of intense exercise

Tumors

- **Soft tissue tumors**
  - Skeletal muscles, fasciae, tendons, nerve sheaths, and interstitial fibrous tissue

- **Benign**
  - Small, rarely develop into malignant tumor
  - Neurofibromatosis type I, neurofibromas, rhabdomyomas
Tumors

- Locally invasive
  - Locally aggressive
  - Low-grade malignancy, but have a high recurrence rate after surgery

- Malignant (sarcomas)
  - Invade local tissues
  - Metastasize to distant sites
    - Lungs
  - ↑ mortality
  - Rhambdomyosarcoma, synovial sarcoma, MFH, liposarcoma
Tumors

- Clinical Features
  - All age groups
  - Masses in soft tissues of extremities or the body

- Symptoms
  - Pain
  - Functional disturbances
  - Destruction of normal tissues
Tumors

- **Treatment**
  - Surgery and chemotherapy, in conjunction with radiation therapy (XRT)

- **Prognosis**
  - Primarily dependant on size and location
  - 5 year survival rate: 40%
Tumors

- PT
  - Address psychosocial and musculoskeletal issues
Fibromyalgia

- Chronic pain syndrome
- Multiple areas of muscle tenderness and joint pain

**Etiology:**
- Genetic predisposition?
- Dysfunction of hormonal linkage system
- Onset related to trauma
Fibromyalgia

- Symptoms:
  - Specific sites of tenderness (11 of 18 points)
  - Headache
  - Fatigue
  - Chest pain
  - Depression
  - Sweating
  - Poor memory, concentration difficulties
  - Morning stiffness
  - Sleep disturbances
Fibromyalgia

- PT
  - Light exercise
  - Aerobic exercise