433 Teams

MEDICINE

24|Myopathies

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COLOR INDEX

Slides - Step-Up medicine - Kaplan Notes - Extre explanation - Doctor Notes

Objectives:

Not provided.

General classification

Acquired Myopathies

Inflammatory Myopathy
- Polymyositis
- Dermatomyositis
- Inclusion body myositis

Infection
- Viral infections (HIV, influenza virus, Epstein-Barr virus)
- Bacterial pyomyositis (*Staphylococcus aureus* and streptococci are common organisms)
- Spirochete (Lyme disease)
- Parasitic infections such as trichinosis

Toxic Myopathy
- Medications
  - Steroids
  - Cholesterol-lowering medications: statins, fibrates, niacin, and ezetimibe
  - Propofol
  - Amiodarone
  - Colchicine
  - Chloroquine
  - Antivirals and protease inhibitors
  - Omeprazole
  - Tryptophan
- Toxins
  - Alcohol
  - Toluene

Myopathy Associated with Systemic Diseases
- Endocrine disorders
  - Thyroid
  - Parathyroid
  - Pituitary or adrenal dysfunction
- Systemic inflammatory diseases
  - Systemic lupus erythematosus
  - Rheumatoid arthritis
  - Scleroderma
  - Sjögren's syndrome
  - Mixed connective disease
  - Sarcoidosis
- Electrolyte imbalance
  - Potassium or magnesium abnormalities
  - Hypophosphatemia
- Critical illness myopathy
  - Nondepolarizing neuromuscular blocking agents
  - Steroids
- Amyloid myopathy
  - Primary amyloidosis
  - Familial amyloidosis (TTR mutation)

Inherited Myopathies

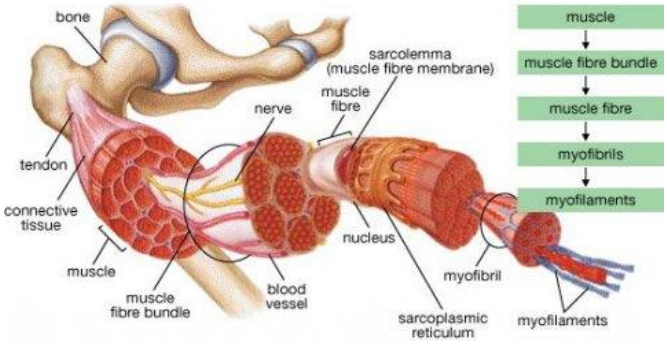
Muscular Dystrophy
- Dystrophinopathy (Duchenne muscular dystrophy,Becker muscular dystrophy)
- Myotonic dystrophy 1 and 2
- Facioscapulohumeral muscular dystrophy
- Oculopharyngeal muscular dystrophy
- Limb girdle muscular dystrophy

Congenital Myopathy
- Nemaline myopathy
- Central core myopathy

Metabolic Myopathy
- Acid maltase or acid alpha-1,4-glucosidase deficiency (Pompe's disease)
- Glycogen storage disorders 3-11
- Carnitine deficiency
- Fatty acid oxidation defects
- Carnitine palmitoyl transferase deficiency

Mitochondrial Myopathy
- Myoclonic epilepsy and ragged red fibers (MERRF)
- Mitochondrial myopathy, lactic acidosis, and strokes (MELAS)
- Mitochondrial neurogastrointestinal encephalomyopathy (MNGIE)
- Progressive external ophthalmoplegia (PEO)

Physiology

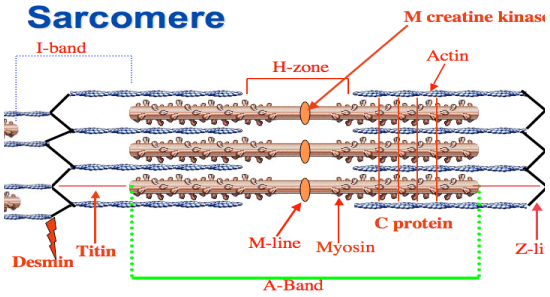


Muscle Fiber Components:

Sarcolemma muscle cell membrane	**Sarcoplasma** muscle cell cytoplasm	**T tubule**: cell membrane extension into the sarcoplasm (to reach the myofibrils)
Motor end plate: contact surface with axon terminal	**cisternae**: areas of the ER dedicated to Ca++ storage (located on each side of the T-tubules)	**Myofibrils**: organized into sarcomeres

The Sarcomere:

- Myosin: thick filament
- Actin: thin filament
- Bands formed by pattern: A and I and H bands
- Z line: area of attachment of the actin fibers
- M line: Myosin fiber centers



Synaptic Events:

The Action Potential reaches the axonal bulb → Voltage-gated calcium channels open → The influx of calcium in the bulb activates enzymes → the vesicles containing the neurotransmitter molecule dock and release the neurotransmitter in the synapse

The neurotransmitter for skeletal muscles is always acetylcholine
The receptors on the muscle fiber are cholinergic receptors
These receptors are nicotinic (fast) acting receptors

Muscle Relaxation:

1. Ach is removed from the receptors by acetylcholinesterase
2. Ligand-gated Na+channels close
3. Na/K pumps reestablish the RMP
4. Ca++ ions leave troponin and are brought back into the cisternae (this process needs energy)
5. Tropomyosin moves back over the actin active site
6. The myosin heads release their binding to actin
7. The filaments passively move back into resting position

Myopathies

- Definition: Disorders in which there is a primary functional or structural impairment of skeletal muscle.

Approach to Myopathies:

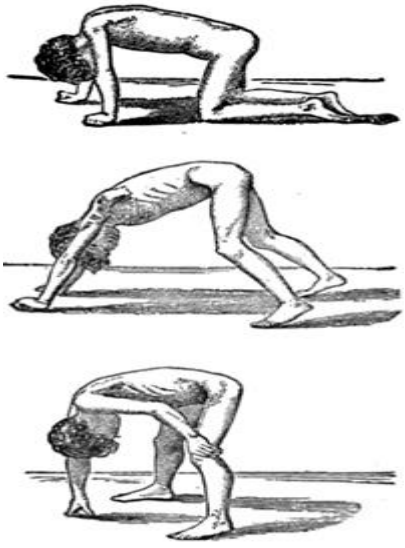
- Distinguishing true muscle weakness from asthenia or motor impairment not due to loss of muscle power.
- Localizing, within the neuromuscular system, the site of the lesion that is producing weakness.
- Determining the cause of the lesion

Symptoms:

POSITIVE:	NEGATIVE:
Myalgia Myotonia. Cramps. Contractures. Myoglobinuria.	Weakness. Atrophy. Exercise intolerance. Periodic paralysis

Gower's Sign: Patients with proximal leg weakness may rise from sitting on the floor by climbing up their legs with their hands.

Myotonia: Impaired relaxation after sustained voluntary contractions.



Weakness *

• Weakness is the cardinal symptom • The distribution of weakness is variable and may change over time . • Most of the time in the proximal muscles; muscles of the shoulder girdle. • Patients complain from difficulty arising from a chair or low toilet, difficulty climbing stairs, a waddling gait, difficulty lifting objects over the head, combing hair or brushing teeth. • Distal weakness is less common. • Patients with proximal leg weakness may rise from sitting on the floor by "climbing up their legs with their hands", This is termed Gower's sign.

-Exercise Intolerance

• A less reliable negative symptom, not a specific symptom. • Often reflects the general level of conditioning and health • In patients without any objective weakness depression should be considered. • Exclude certain metabolic myopathies or mitochondrial cytopathies. • Ask if it is elicited by brief or long term exercise, which orients towards a disorder of carbohydrates or lipid metabolism, respectively.

It is not a feature of {muscle diseases} if it is isolated.

Myalgias

• An infrequent symptom, seen in inflammatory and metabolic myopathies. • Orthopaedic or rheumatologic conditions are more frequent causes (we should rule them out).

• Constant proximal muscle pain often accompanies inflammatory myopathies. • Episodic myalgias after exercise point to metabolic myopathies. • In individuals with waxing and waning, diffuse myalgias, especially in neck and lower back anxiety should be ruled out.

Cramps

• Involuntary painful contractions of muscle that last for seconds to minutes, seen in metabolic myopathies. • Most are benign and occur predominantly in calves . • Risk factors are dehydration, old age, prolonged sitting, use of diuretics, hypothyroid state, DM • They are most common in motor neuron disease ,and in chronic neuropathies rather than in myopathies • Cramps are only common in metabolic myopathies such as myophosphorylase deficiency (McArdle's disease), and in hypothyroid myopathy

*Important points to ask:

- Onset. If hyper acute usually vascular. –
Course. Fixed course like in congenital myopathies or progressive like dystrophies. –
Limbs involved, all four limbs; either symmetrical or not. If in one side only think of CNS lesions. - Muscles involved. Proximal. if distal think of neuropathies. - Progression. - Presence of sensory/autonomic symptoms, they are not caused by myopathies, but may be associated with them (e.g. mitochondrial myopathy). - Patient's demographics

Myotonia

• Impaired relaxation after sustained voluntary contraction. • A painless phenomenon • Commonly involves intrinsic hand muscles and eyelids . • It is due to repetitive depolarization of the muscle fibers • It improves with repeated exercise. • Clinically, myotonia can be seen by tapping the muscle (percussion myotonia) or by voluntary contractions of muscle groups (action myotonia). • Typical tests are squeezing the hand of the examiner or forceful closure of the eye.

{You will find delayed relaxation of the eyes/ Sustained contraction of the hand muscle.}

Myoglobinuria

Severe and relatively acute muscle fiber damage {Inflammatory myopathy, Toxic myopathy}.

Classification:

Hereditary	Acquired
Channelopathies	Drug-induced myopathies
Congenital myopathies	Endocrine myopathies
Metabolic myopathies	Inflammatory/immune myopathies
Mitochondrial myopathies	Myopathies associated with other systemic illness
Muscular dystrophies	Toxic myopathies
Myotonias	

Investigations:

- Muscle enzymes:
  - CK. (CK could be high because of IM injection, strenuous exercise, trauma
  - Aldolase.
  - LDH.
  - Aminotransferase.
  - ANA, ENA antibodies(anti Ro/SSA, anti La/SSB, anti Sm, and anti RNP)
- Myositis specific antibodies (anti-histidyl-t-RNA aynthase anti Jo-1).
- Genetic testing.
- Electromyography, nerve conduction studies.
- MRI.
- Muscle biopsy.

Congenital Myopathies: (Slow/Non-progressive)

Prenatal:	Postnatal: hypotonia, poor respiratory effort, difficulty feeding, reduced muscle bulk, weakness.	First year and beyond: hypotonia, weakness, delayed milestones, failure to thrive, recurrent respiratory infections, flaccid speech.
decreased fetal movement.		

Types:

- Multicore disease
- Centronuclear (myotubular) myopathy
- Congenital fiber-type disproportion
- Central core disease
- Nemaline (rod) myopathy - Myofibrillar myopathy

Management:

- Genetic Counseling.
- Detection and treatment of orthopedic complication.
- Prevention of complications (general anaesthesia)

Malignant Hyperthermia

- Hypermetabolic crisis that occurs when a MH- susceptible individual is exposed to a volatile anaesthetic or succinylcholine.
- Genetic skeletal muscle receptor abnormalities allowing excessive calcium accumulation in the presence of certain anaesthtic triggering agents. →

{Accumulation of Ca will causes sustained muscle contraction which lead to 1-depletion of co2 2- anaerobic metabolism 3-acidosis rhabdomyolysis }

Symptoms:

- Masseter spasm immediately following anaesthetic induction.
- Hypercarbia.
- Sinus tachycardia.
- Generalized muscular rigidity.
- Tachypnea.
- Cyanosis.
- Rapidly increasing temperature is a later sign of MH and is typically absent when the diagnosis is initially suspected.
- Sweating
- Cola- colored urine.
- Ventricular fibrillation.

Muscular dystrophies

- Muscular Dystrophy: is Inherited, progressive degeneration of the muscles with connective tissue replacing muscle fibers with systemic involvement

  (variable age of onset)

Types:

- Dystrophinopathies: Duchenne and Becker (DMD, BD). Most common
- Emery-Dreifuss muscular dystrophy.
- Autosomal dominant dystrophies:

  ♣ Facioscapulohumeral MD. ♣ Oculopharyngeal MD.

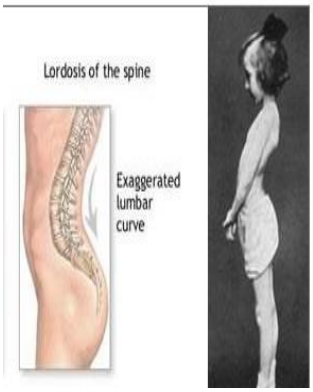
  ♣ Classic, Congenital and Proximal myotonic dystrophy.
- Limb girdle MD.

Dystrophinopathies

- X linked recessive disorders caused by mutation in the dystrophin gene. (so it affects males more than females)
- Duchenne and Becker (DMD, BD).
- Dystrophin provides mechanical reinforcement to the sacrolemma and stabilizes the glycoprotein complex.
- Its absence causes digestion of the glycoprotein complex. This initiates degeneration of the muscle fiber resulting in muscle weakness.

1-Duchenne Muscular Dystrophy (DMD)

Motor developmental delay.	Onset: age **3-6** years	Toe walking, as a compensation for the progressive weakness of the knee extensors.	Difficulty rising from sitting position. Difficulty rising from sitting position. Gower's sign.	Lumber lordosis, waddling gait, pseudo-hypertrophy of the calves.



https://www.youtube.com/watch?v=2Bxrt3NGCj4

Lordosis: refers to normal inward lordotic curvature of lumbar and cervical regions of the spine

At 12 years: loss of ambulation, marked wasting of muscles, contractures, kypho-scoliosis, exaggerated lumber lordosis.

Death due to respiratory complication between 15-30 years.

Systemic Involvement:

- Cardiomyopathy : CHF and arrhythmias.
- Malignant hyperthermia like reactions with rhabdomyolysis.
- Intestinal pseudo-obstruction.
- CNS involvement: mental retardation, learning disabilities.

Investigations:

- CK is markedly elevated early in the disease.
- Electromyography: myopathic potentials.
- Muscle biopsy: necrosis, replacement with connective tissue and fibrosis, variation in muscle fiber size, absent dystrophin.

Management:

- Early detection of systemic involvement:
- Assessing for evidence of cardiac dysfunction and treatment accordingly.
- Weight and growth monitoring.
- Screening for orthopedic complications to maintain function and prevent contractures
- Dietary calcium and vitamin D supplementation, and yearly DXA scanning.
- Avoidance of anaesthesia and sedation if possible.
- Glucocorticoids (are the amainstay)

Glucocorticoids

The effects	The side effects
For boys 5 years and older who are no longer gaining motor skills or whose motor skills are declining. ✓ It increases strength, muscle and pulmonary functions. ✓ Reduces cardiomyopathy and lower mortalitiy. ✓ Has an anabolic action in contrast to its catabolic action on normal skeletal muscle in unaffected people. ✓ Stabilizes sacrolemma.	➢ Weight gain ➢ Cushingoid facial appearance, acne. ➢ Short stature, compression fractures ➢ Delayed puberty. ➢ Excessive hair growth. ➢ Gastrointestinal bleeding. ➢ Psychosis, and behavioral changes.

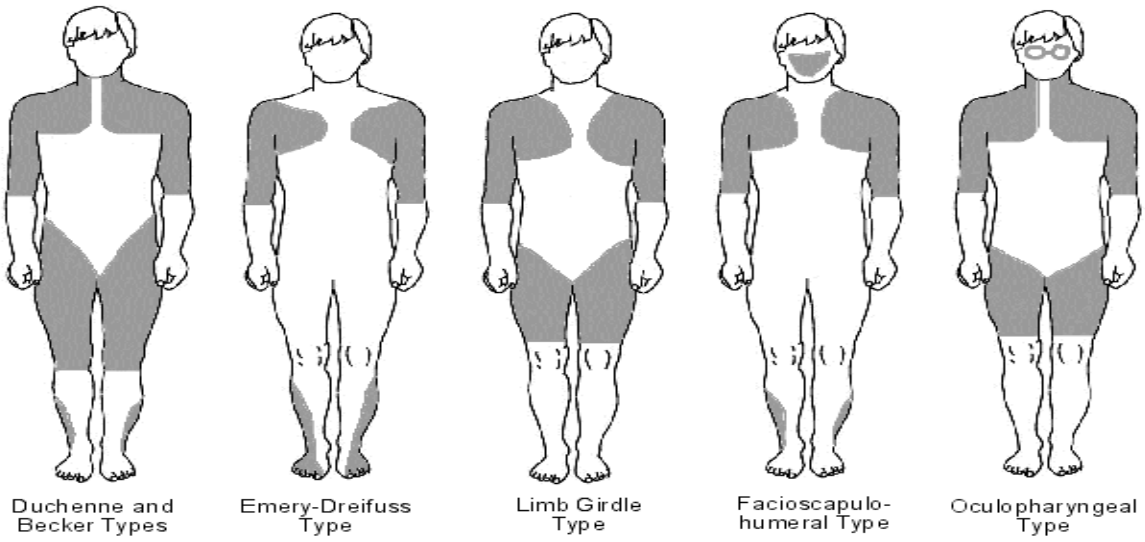
2-Becker Dystrophy			
Older age at onset. Most between ages 5 and 15. Onset in the third or fourth decade or even later can occur	Less severe symptoms.	Loss of ambulation is usually in the 4th decade.	Muscle biopsy shows decreased staining patterns rather than complete absence of dystrophin.

3-Emery-Dreifuss Muscular Dystrophy (EDMD):		
X-linked: Emerin mutations most common, FHL1 mutation also similar phenotype. AD: Mutations LMNA gene for lamin A/C (LGMD1B), but clinical symptoms are closely related	Onset in early childhood & teenage years	Clinical Presentation: Prominent and early contractures (elbows, neck) often preceding muscle weakness. Muscle weakness is in a limb- girdle distribution. Dilated cardiomyopathy may occur and may result in sudden death, arrhythmia, & conduction defects.

4-Limb-Girdle Muscular Dystrophy (LGMD):

Represents more than 1 genetic disorder

Inheritance: Autosomal dominant/ recessive Systematic classification is based on inheritance pattern: Autosomal dominant (LGMD1) Autosomal recessive (LGMD2)	Onset – late 1st to 4th decade	Clinical Presentation: Progressive weakness of pelvic & shoulder girdle muscles Diaphragmatic weakness & cardiomyopathy may also occur intellectual function is intact	Natural History: -Slow progression. -After onset > 20 years of contracture & disability -Rarely significant scoliosis	Treatment: Supportive.



Main areas of muscle weakness in different types of dystrophy

5-Myotonic Dystrophy

https://www.youtube.com/watch?v=Wg1SVoa-8JE

The most prevalent inherited neuromuscular disease in adults.

Autosomal dominanat.

Age of onset average is 29 years

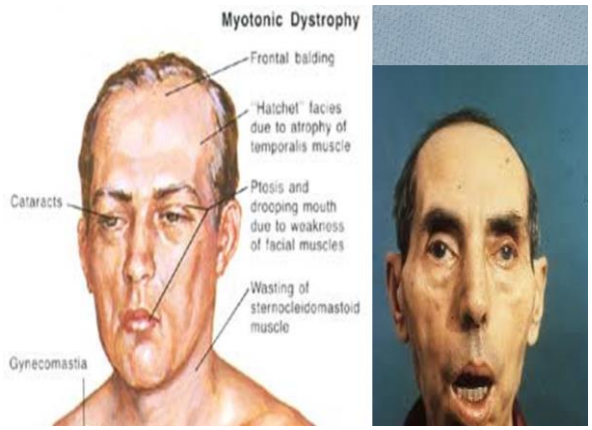
Diagnosis: can usually be made clinically in a patient with the characteristic presentation and a positive family history.

Genetic testing for an expanded CTG repeat in the DMPK gene is the gold standard for confirming the diagnosis of DM1.

Testing for the CCTG repeat in the ZNF9 gene is appropriate if DM1 testing is negative.

Clinical features:

- Myotonia.
- Weakness of the forearms and peroneal muscles.
- Ptosis and weakness of other facial muscles.
- Characteristic facial appearance: The face is long and narrow and the palate is high arched. The cheeks are hollowed and the jaw sags.
- Frontal bolding.
- Mild axonal neuropathy.
- Heart involvement.
- GIT dysmotility, constipation and diarrhea.
- Cataract.
- Endocrine abnormalities.
- Low IQ.



6- Fascioscapulohumeral Muscular Dystrophy

ETIOLOGY	ONSET	CLINICAL PRESENTATION:
-AUTOSOMAL DOMINANT -GENE DEFECT (*FRG1*) -CHROMOSOME 4Q35	late childhood/early adulthood	Muscle weakness of the face, shoulders and upper arms. Sparing of the deltoid, distal pectoralis major, and erector spinae Winging of the scapula. Markedly decreased shoulder flexion and abduction. Horizontal clavicles.

7-Distal muscular dystrophy

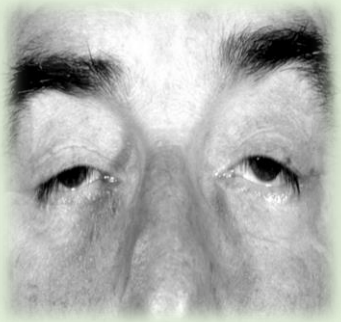
Slow progression therefore the patient may not know that they have it until they are in their late 40's or 50's.

Types:

- Welander's distal myopathy,
- Finnish (tibial) distal myopathy,
- Miyoshi distal myopathy,
- Nonaka distal myopathy,
- Gowers-Laing distal myopathy,
- Hereditary inclusion-body myositis type 1,
- Distal myopathy with vocal cord and pharyngeal weakness,
- ZASP-related myopathy.

8-Ocular muscular dystrophy

(Autosomal dominant myopathy with complete penetrance)

Onset	Treatment	Diagnosis:
Middle age with ptosis	Supportive Cricopharyngeal myotomy for the dysphagia . Doxycycline treatment. Cystamine treatment.	➢ Serum CK levels may be elevated. ➢ Genetic testing reveals (GCN)12-17 trinucleotide repeat expansions in exon 1 of PABPN1, the polyadenylation-binding protein nuclear 1 gene. ➢ Variation of fiber size and "rimmed" vacuoles on light microscopy and 8.5 nm intranuclear tubular filaments on electron microscopy.
Clinical features		
Ocular and pharyngeal muscle involvement**.** **Typically presents with ptosis, dysarthria, and dysphagia. It can also be associated with proximal and distal extremity weakness.**		

Mitochondrial Myopathies

Can present with a number of clinical pictures:

As chronic progressive external ophthalmoplegia (with or without mild proximal muscle weakness) or KearnsSayren syndrome	As an isolated myopathy with or without exercise intolerance and/or myalgia	As a severe myopathy or encephalomyopathy of infancy	As a predominantly multisystem disease with myopathy (eg, MELAS and MERRF)

Diagnosis:

- Lactate: Elevated lactate levels in plasma or cerebrospinal fluid (CSF) can be a supportive feature for the diagnosis of mitochondrial disease.
- Exercise testing: Can aid the diagnosis of mitochondrial myopathy when the phenotype is nonspecific, particularly for patients presenting only with exercise intolerance and fatigue.
- Muscle biopsy: Remains one of the main tools for diagnosing a mitochondrial disorder. Even biopsies of clinically asymptomatic patients can show pathologic abnormalities that are specific for mitochondrial dysfunction. The classic hallmark of mitochondrial diseases is "ragged red fibers" (RRF).
- Biochemical analyses: Measurement of respiratory chain function
- Molecular genetic studies

Endocrine Myopathies

- Adrenal dysfunction (as in Cushing disease or steroid myopathy)
- Thyroid dysfunction (as in myxedema coma or thyrotoxic myopathy)
- Parathyroid dysfunction (as in multiple endocrine neoplasia)
- Pituitary dysfunction
- Islands of Langerhans dysfunction (as in diabetic myopathy from ischemic infarction of the femoral muscles).

Frequency decreases after age 30; may become attack free in 40s and 50s

Muscle Channelopathies:

Nondystrophic Myotonias and Periodic Paralyses

A group of rare inherited diseases caused by mutations in muscle ion channels (sodium, chloride, potassium, and calcium). Mutations cause an increase or decrease in muscle membrane excitability.

THE NONDYSTROPHIC MYOTONIAS are characterized by delayed relaxation after muscle contraction, causing muscle stiffness and pain; the periodic paralyses are characterized by episodes of flaccid muscle paralysis.

Non dystrophic myotonias:	**Periodic paralyses:**	**Clinical Features:** Last several hours-more than a day
➢ Myotonia congenita (CLCN1) ➢ Paramyotonia congenita (SCN4A) ➢ Sodium channel myotonias (potassium aggravated myotonias) (SCN4A)	➢ Hypokalemic (CACNA1S/ SCN4A) ➢ Hyperkalemic (SCN4A) ➢ Anderson Tawil syndrome (KCNJ2)	➢ Flaccid paralysis – mild focal weakness to severe generalized weakness ➢ Occur anytime of the day; more common in morning ➢ Absence of myotonia ➢ Proximal > distal weakness; legs > arms ➢ Sparing of facial, ventilatory and sphincter muscles ➢ Attacks may be preceded by sensation of heaviness and or aching in the low back

Precipitating factors:

- Strenuous physical activity followed by rest or sleep
- High carb diet
- ETOH consumption
- Emotional stress
- Concurrent viral illness
- Lack of sleep
- Medications like beta agonists, corticosteroids, and insulin

Diagnosis:

- Serum K < 3.0mEq/L
- Serum CK level elevated
- EKG changes – U waves, flattening of T waves
- Provocative testing - Intravenous glucose load/ insulin
- Electrophysiology
- Sensory and motor NCS normal between attacks
- During attacks – small CMAP. Reduced insertional activity, fibs and positive sharp waves
- No myotonia on EMG
- Short/ long exercise test

Treatment:

- Reducing exposure to known triggers
- Acute treatment: replacement of K
- Acetazolamide: prevent attack recurrence and severity (may precipitate weakness in HypoKPP2)
- Dichlorphenamide – no longer available
- Triamterene and spironolactone

Inflammatory Myopathies (Polymyositis and Dermatomyositis)

Definitions:

The Term POLYMYOSITIS is used when the condition does not involve the skin.

The Term DERMATOMYUOSITIS is used when polymyositis is associated with a characteristic skin rash.

Causes:

Hypothesis: **A genetically susceptible individual + an environmental trigger → immune activation → chronic inflammation.**

Pathological changes in muscle:

a. Polymyositis: cell mediated process
b. Dermatomyositis: humoral immune mechanisms

Clinical features

Features common to both polymyositis and dermatomyositis:

a-Symmetrical proximal muscle weakness that develops subacutely over weeks or several months.

- The earliest and most severely affected muscle groups are the neck flexors, shoulder girdle, and pelvic girdle muscles.
- Distal extremity weakness is less frequent and less severe.

a. Myalgia in 33% of patients.
b. Dysphagia (involvement of esophageal muscles) in up to 30% of patients.

Features unique to dermatomyositis:

a. Heliotrope (butterfly) rash
b. Gottron's papules: popular, erythematous, scaly lesions over the knuckles.
c. V sign: rash on the face, neck, and anterior chest.
d. Shawl sign: rash on shoulders and upper back, elbows, and knees.
e. Periungual (around the nails) erythema with telangiectasis.
f. Subcutaneous calcifications in children

Associated findings:

a. In both polymyositis and dermatomyositis:
- Arthralgias (common)
- CHF and conduction defects (rare)
- Interstitial lung disease (in minority)

b. In dermatomyositis only:
- Vasculitis of the GI track, kidneys, lungs and eyes (more common in children).
- ↑Incidence of malignancy in older adults (lung, breast, ovary, GIT, and myeloproliferative disorders) → make effort to uncover occult malignancy.

In PM: 15% risk of non-hodgkin's lymphoma and lung CA

Diagnosis:

- The best initial test is CPK and aldolase.
- The most accurate test is a muscle biopsy.
- ANA is frequently positive, but nonspecific.
- Anti-Jo antibodies are associated with lung fibrosis.
- MRI detects patchy muscle involvement.
- Electromyography is often abnormal
- Other labs that are occasionally abnormal are the ESR, C-reactive protein, and rheumatoid factor. Like the occasional presence of anemia, none of these tests will help establish the diagnosis.

Management:

- Steroids are usually sufficient.
- When patient is unresponsive or intolerant of of steroids, use:

a. Methotrexate
b. Azathioprine
c. IV immunoglobulin
d. Mycophenolate

- Hydroxychloroquine helps the skin lesions.

Inclusion Body Myositis

More common in men and those above 50.
Insidious onset of slowly progressive proximal **and** distal weakness.
Early weakness and atrophy of quadriceps, forearm flexors, and tibialis anterior muscles. Involvement is asymmetrical.
Facial weakness in 1/3 of patients and dysphagia in ½ of patients.
There may be loss of deep tendon reflexes.
Diagnosis: slight elevation of CK levels (relatively low), muscle biopsy.
Poor response to therapy.

Toxic Myopathies

- Alcohol, cocaine.
- Lipid lowering agents.
- Steroids.
- Antimalarials, antiretroviral.
- Antipsychotic.
- Chemotherapy.

Statin Induced Myopathy

Clinical features:

- Myalgia
- Myopathic weakness
- Myositis.
- Myonecrosis
- Rhabdomyolysis

Prevention:

- Pravastatin and fluvastatin.
- A baseline CK level prior to starting statin.
- Patients should be alerted to report the new onset of myalgia and weakness.
- Caution in patients with renal failure, hypothyroidism and liver failure.

Glucocorticoid-induced myopathy

- Can occur with the initiation of systemic therapy as well as in chronic maintenance therapy when the dose is increased.
- Gradual onset over several weeks of proximal muscle weakness accompanied by muscle wasting. A common manifestation is difficulty getting up from a chair or climbing stairs.
  Pathophysiology: Glucocorticoids have a direct catabolic effect on skeletal muscle via effects on intermediary metabolism that provide amino acids as a substrate for gluconeogenesis.
  Diagnosis:

- absence of other causes of myopathy.
- demonstrating improved strength within three to four weeks after appropriate dose reduction.
  Treatment: Muscle strength begins to improve within three to four weeks after appropriate dose reduction and eventually resolves in virtually all patients if glucocorticoid therapy can be discontinued.

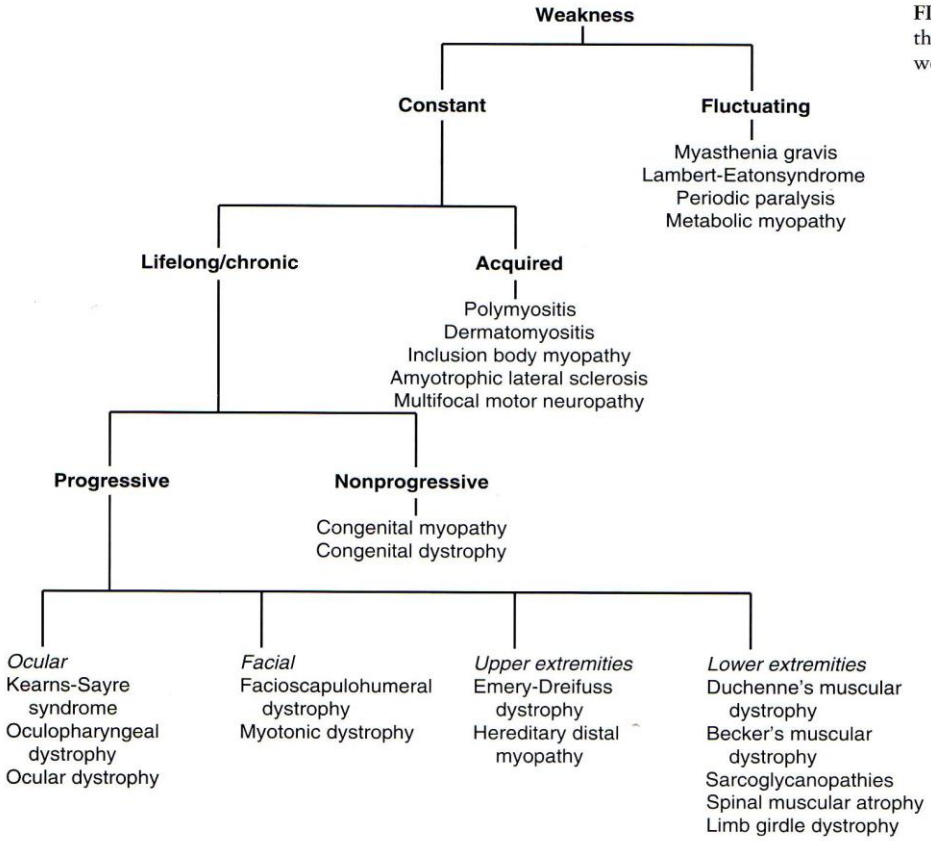


FIGURE 28.6 An algorithm for the approach to the patient with weakness.

MCQS

A patient complains of a history of generalized muscle weakness. On examination, his facial muscles show marked atrophy, and when you ask him to shake your hand, he appears to be unable to relax his grip for an extended period.
What is the likely diagnosis?

a. Myasthenia Gravis.

b. Inclusion body myositis.

c. Becker's dystrophy.

d. Myotonic Dystrophy.

2) The parents of a 3-year-old boy are concerned that he is not walking as well as other boys his age. Both parents are healthy, and there is no family history of neuromuscular disease. He has three older brothers who are healthy. Physical examination shows that he has large calf muscles and lower extremity proximal muscle weakness, as demonstrated by the need to use his arms and hands to assist in standing from a seated position. CK levels are elevated. If you are suspecting Duchenne dystrophy, what will the results of dystrophin staining show?

a. Reduced dystrophin staining.
b. Normal dystrophin staining.
c. Increased dystrophin staining.
d. Absent dystrophin staining.

3) A 30-year-old man complains of bilateral leg weakness and clumsiness of fine movements of the right hand. Five years previously he had an episode of transient visual loss. On physical examination, there is hyperreflexia with Babinski sign and cerebellar dysmetria with poor finger-to-nose movement. When the patient is asked to look to the right, the left eye does not move normally past the midline. Nystagmus is noted in the abducting eye. A more detailed history suggests the patient has had several episodes of gait difficulty that have resolved spontaneously. He appears to be stable between these episodes. He has no systemic symptoms of fever or weight loss. Which of the following is the most appropriate next test to order?

a. Lumbar puncture
b. MR scan with gadolinium infusion
c. Quantitative CSF IgG levels
d. Testing for oligoclonal bands in cerebrospinal fluid
e. CT scan of the head with intravenous contrast

Done By:

Rawan alabdillah	Mohammad alshehri
Shahad almuhaideb	Sara habis
Faroq Abdulfattah	Areej Alwahaib