

## Neuropathy and Myopathy

1. Recognize the clinical signs, symptoms, evaluation, and treatment of myasthenia gravis  
Occur any age, associated w/ thymic tumors, thyrotoxicosis, RA, and lupus. More women, cause fluctuating weakness and easy fatigability of voluntary muscles, commonly affects external ocular muscles and cranial muscles, may also affect respiratory and limb. Due to immune mediated decrease in functioning Ach receptors. Concurrent infections exacerbate symptoms, slowly progressive course. Pt presents w/ ptosis, diplopia, difficulty swallowing/chewing, respiratory difficulty. Characteristic feature is sustained activity that leads to temporary increased weakness. Diagnosis confirmed by giving Achesterase drug, most commonly use edrophonium test (10mg IV) get obvious improvement in strength in myasthenia patients, also use neostigmine (1.5 mg IM). X-ray/CT may show thymoma. Treatment: (a) anticholinesterase drugs - . symptomatic relief, use pyridostigmine 30-180 mg four times a day, may use atropine to counteract side effects. (b) thymectomy - patients under 60 years, give symptomatic benefit or remission (c) corticosteroids - w/ poor response to anticholinesterase and thymectomy, initial high dose then taper to lower dose (d) azothioprine - severe or progressive disease (e) plasmapheresis - temporary improvement in myasthenia crisis
2. Recognize the clinical signs, symptoms, evaluation and treatment of Guillain Barre syndrome  
Acute or subacute polyneuropathy, follows infective illness (especially Campylobacter jejuni), immunologic basis. Clinical features - present w/ symmetric weakness starting in legs more proximally, may get muscle wasting if axonal degeneration has occurred, reflexes absent, autonomic dysfunction (tachycardia, cardiac irregularities, labile BP, sweating, sphincter disturbance). Investigational studies - CSF has increase protein, but normal cell count, EMG shows slow motor and sensory conduction velocity/evidence of demyelination and axonal loss. Treatment - plasmapheresis reduced time for recovery and likelihood of residual neurological deficits 1 IV 19 as effective as plasmapheresis, therapy otherwise symptomatic, prevent complications like respiratory collapse, best managed in ICU. Prognosis - symptoms/signs cease to progress by 4 wks, self-limiting, improvement over wks-months, 70-75% recover completely, 5% die
3. Recognize the clinical signs and symptoms of a generalized peripheral neuropathy
4. Understand the significance and differential diagnosis of mononeuritis multiplex  
Involvement of various nerves in asymmetric manner and at different times, presence of asymmetric, and multifocal motor/sensory/reflex deficits. DDX includes: Vasculitis (i.e. polyarteritis nodosa), DM, leprosy, sarcoidosis, HIV, lymphoma, lyme disease, hereditary liability to pressure palsies, multifocal motor neuropathy, chronic inflammatory demyelinating polyneuropathy.
5. Recognize the clinical signs, symptoms, evaluation and treatment of polymyositis  
Destruction of muscle fibers and inflammatory infiltration of muscles, occurs at any age, progress at variable rate, leads to weakness and wasting especially of proximal limb and girdle muscles, often associated w/ muscle pain, tenderness, dysphagia, and respiratory difficulty. May also see Raynaud's, arthralgia,-malaise, wt loss, and low grade fever. Associated w/ autoimmune diseases, serum CK elevated, EMG shows short/low

amplitude polyphasic motor unit potentials, muscle biopsy shows fiber necrosis and infiltration of inflammatory cells. Treatment w/ anti-inflammatory cells, often use Prednisone (start 60-80 mg then taper to 10-20 mg) can also use cytotoxic drugs (azathioprine, methotrexate), physical therapy to prevent contractures.

6. Recognize the common clinical signs and symptoms resulting from radiculopathies at the C5-6 C7, C8, L3-4, L5 and S 1 levels

C5-6 - affect deltoid and biceps, sensory change over lateral upper arm and thumb, depressed biceps reflex

C7....affect triceps and finger extensors, sensory change over middle fingers, depressed triceps reflex

C8 - affect finger extensors and abductors of index/5th finger, sensory change over little finger, no depressed reflex

L3-4 - affect quadriceps muscle, sensory change over medial shin, depressed knee reflex L5 - affect great toe extension, sensory change over medial foot and great toe, no depressed reflexes

S 1 - affect planter flexion, sensory change over lateral foot and small toe, depressed ankle reflex

7. Recognize the pattern of a dystrophinopathy

8. Know the major categories of muscular dystrophies

Group of inherited disorders, progressive muscle weakness and wasting, skeletal muscle genes encode sarcolemmal, cytoskeletal, cytosolic, extracellular matrix, and nuclear membrane proteins, no specific. treatment, deformities/contractures common, inactivity worsens disability.

(a) Duchenne - most common, x-linked, males, symptoms start at age 5, severe disability by adolescence, death by 3rd decade, toe walking/waddling gait/inability to run are early symptoms, weakness in proximal lower extremities, use arms to stand up, pseudohypertrophy of calves (fatty infiltration), may involve heart later, associated w/ mental retardation, serum. CK very high, no definitive treatment; prednisone may help, gene defect in dystrophin gene, absent or profound reduction in dystrophin,

(b) Becker-x-linked, weakness similar to Duchenne, onset and death later, no cardiac or cognitive dysfunction, Ck not as high, dystrophin levels normal, but protein qualitatively altered

(c) Emery-Dreifuss - x-linked recessive or autosomal dominant, onset in childhood, slow progression, weakness/wasting of triceps/biceps and peroneal/tibialis anterior, cardiomyopathy, Ck mildly elevated

(d) Limb-Girdle - autosomal recessive, late childhood, shoulder and pelvic girdle muscles equally effected, no pseudohypertrophy, CK less elevated

(e) Facioscapulohumeral- autosomal dominant: onset in adolescence, normal life span, severity variable, weakness confined to face, neck, and shoulder girdle, winged scapula common, no heart involvement, Ck normal or slightly elevated

- (f) Distal myopathy - autosomal dominant, present > 40 years old, small muscles of hands/feet, wrist extensors, and foot dorsiflexors affected, slow progression
- (g) Ocular - autosomal dominant, onset <30 years old, ptosis first sign, progressive ophthalmoplegia, facial weakness, slow progression
- (h) Oculopharyngeal- autosomal dominant, begin 3-5th decade, ptosis, external ophthalmoplegia, dysphagia, facial weakness, proximal limb weakness, serum: Ck mildly elevated.
- (i) paraspinal- after age 40, progressive paraspinal weakness, family history, back pain and kyphosis, serum CK mildly elevated, CT shows fatty replacement

## Spinal and Peripheral Nerve Disorders

1. Describe the anatomy of the bony spine and its relationship to the cervical, thoracic, lumbar, and sacral nerve roots

7 cervical, 12 thoracic, 5 lumbar, 5 sacral vertebra; 8 cervical (C1 sensory only, others mixed nerves), 12 thoracic (mixed nerves), 5 lumbar (mixed nerves), and 5 sacral nerve roots (mixed nerves). In cervical region nerve roots exit rostral to corresponding pedicle, in thoracic and lumbar regions nerve roots exit caudal to corresponding pedicle. In sacral spine roots emerge via neural foramina in sacrum.

2. Differentiate the conus medullaris and the cauda equina

conus medullaris = point where spinal cord ends at vertebral level L1

cauda equina = bundle of spinal nerve roots in the spinal canal that arise from lumbar enlargement and conus medullaris, comprise all the roots from L1 and below.

3. Discuss the surgical work up for surgery for disorders of the spinal cord, spinal roots, and peripheral nerves

Primary indication for workup = abnormal neurologic exam points to unexplained problem of cord, roots, or peripheral nerve.

Secondary indication for workup = instability of spine (cord/roots), or acute trauma to extremity (peripheral nerves)

- . Workup for cord/roots = MRI in all cases, EMG/neurophysiologic studies in selected cases, if hemorrhage/infection suspected do LP, if ischemia suspected do spinal angiography, if spinal instability present do CT scan.

Workup for peripheral nerves = EMGINCV in all cases, MRI in selected cases, if tumor suspected do MRI, if ischemia suspected do angiography.

4. Discuss the indications for surgery of the spinal cord, spinal roots, and peripheral nerves

Cord/roots = instability of spine, loss neurologic function, tumor that requires tissue diagnosis.

Peripheral Nerves = visibly divided nerve in open wound, loss of neurologic function localized by EMG, tumor that requires tissue diagnosis

5. Identify the major mixed nerves

Median, Ulnar, Radial, musculocutaneous, femoral, obturator, sciatic

6. Locate the major entrapment sites

Carpal tunnel - median nerve at wrist, paresthesia of hand, pain in forearm, may get weakness and atrophy of thenar muscles.

Cubital tunnel - ulnar nerve at elbow, paresthesia, hypesthesia pain in little finger and ulnar border of hand, weakness of adductor pollicis/ deep flexors/ intrinsic hand muscles

Tarsal tunnel- posterior tibial nerve at ankle, complain of burning in foot, weakness of intrinsic foot muscles

7. Relate the major causes of acute peripheral nerve injury

bullets, knives, tourniquets, fractures (i.e. colles' and mid-humerus)