PLEXOPATHY

I. Brachial Plexopathies

A. Anatomy

1. Roots form trunks
   a) C5 + C6 form upper trunk
   b) C7 forms middle trunk
   c) C8 + T1 form lower trunk

2. Trunks divide into anterior and posterior divisions

3. Trunks also form cords
   a) Posterior divisions of upper + middle + lower trunks form posterior cord
   b) Anterior divisions of upper + middle trunks form lateral cord
   c) Anterior divisions of middle + lower trunks form medial cord

4. Cords form nerves
   a) Posterior cord forms radial nerve and axillary nerve
   b) Lateral cord forms musculocutaneous nerve and outer (lateral) portion of median nerve
   c) Medial cord forms ulnar nerve and inner (medial) portion of median nerve
5. Sympathetic preganglionic (myelinated) nerve fibers arise from intermediolateral column in the spinal cord from C8 (T1) to L1, exit with ventral roots and ascend rostrally along vertebra to ganglia

a) From superior cervical ganglia postganglionic (unmyelinated) fibers follow roots C1 – C4

b) Middle cervical ganglia variable

c) From inferior cervical ganglia (stellate) postganglionic (unmyelinated) fibers follow roots C6 – T1

B. Functional analysis

1. Division of trunks into anterior and posterior cords based on primitive anterior (preaxial) and posterior (postaxial) mesodermal masses.
2. All anterior limb compartment and clavicular + coracoid-based muscles are preaxial and innervated by nerves from anterior division cords (musculocutaneous-biceps muscle, median and Ulnar nerves).

3. All posterior limb compartment and scapular-based muscles (excluding coracoid-based muscles) are postaxial and innervated by nerves from posterior cords (long thoracic- serratus anterior muscle, dorsal scapular-rhomboid muscles, suprascapular-supraspinatus and infraspinatus muscles, thoracodorsal-latissimus dorsi muscle subscapular-subscapularis and teres major axillary-deltoid and teres minor muscle, and radial nerves).

C. Clinical Characteristics

1. Sensory loss always associated with disturbances of sweat secretion except root avulsion.

2. Horner’s syndrome associated with T1 involvement

3. Trunk lesions
   a) Upper trunk (most common, Duchenne-Erb, “porters tip” hand) = Axillary + Musculocutaneous + Suprascapular + lateral pectoral nerve + Lateral portion Median nerve (pronator teres and flexor carpi radialis) + Brachioradialis
   b) Middle trunk (rare) = thoracodorsal + subscapular + Radial (except brachioradialis)
   c) Lower trunk = Ulnar + medial median nerve + medial pectoral nerve

4. Cord lesions
   a) Posterior cord = radial + axillary nerves + thoracodorsal + subscapular
   b) Lateral cord = musculocutaneous + lateral portion of median nerve-PRT & FCR (spares hand)
   c) Medial cord = Ulnar + medial portion of medial nerve-median intrinsic hand (APB, OP) and all finger flexors (FPL, FDP, FDS)

D. Pathophysiology

1. Traumatic Brachial Plexopathies
a) Plexus vulnerable to trauma given superficial location in interscalene triangle, and close to mobile bony structures in shoulder that may compress plexus, or excess traction when head/neck and shoulder/arm forced in opposite directions.

(1) Traction injury: motor cycle, roll-over motor vehicle crash, or fall with suspension of body from upper limb

(2) Laceration injury: glass, weapons, surgical procedure

(3) Compressive injury: heavy downward force on clavicles—“pack palsy” or “rucksack paralysis”

(4) Traumatic lesions rarely limited to one trunk or cord.

(5) Trauma may also cause coexistent lesions with brachial plexopathy

(a) Spinal cord

(b) Individual nerves that arise from plexus (ex. axillary, radial, etc.)

(c) Root avulsion- usually lower cervical roots, and cannot spontaneously regenerate

(6) Management of Traumatic Brachial Plexopathy:

(a) Initial EDX test at least 2-3 weeks after injury

(b) Wait 3-4 months for spontaneous recovery from neurapraxia

(c) After 4 months if EDX test show severe axonal degeneration then surgical exploration with suturing, nerve grafting or both.

(i) Surgical goal to restore nerve supply to proximal arm muscles: Deltoid, Biceps, Triceps (distal/hand muscles no effective)

(ii) Focus on upper and middle trunks & lateral and posterior cords.

(iii) Neurotization: Take intercostals nerve grafts, most often to musculocutaneous nerve
(iv) Avulsed roots can be reimplanted into cord with some functional improvement.

(v) Muscle transfers

2. Shoulder Injuries

a) Usually damage nerves arising from plexus then plexus itself

b) Shoulder dislocation: anterior, posterior-uncommon and inferior; Proximal humeral fractures, Clavicle Fractures

c) Frequency

(1) Anterior shoulder dislocation: Axillary-42%, Suprascapular -14%, musculocutaneous-12%, Ulnar-8%, Radial-7%, Median-4%--Most Recover when associated with dislocation

(2) Proximal humeral fracture: Axillary-58%, Suprascapular -48%, Musculocutaneous-29%, Ulnar-6%, Radial-32%, Median -17%--More likely more than one nerve involved in fractures

(3) Clavicle Fractures: immediate damage to plexus from bone fragment, hematoma, but may be delayed from scar tissue/callus or pseudoaneurysm.

3. Perioperative Brachial Plexus Injuries: In general most patients have complete recovery in 2 months.

a) Plexopathy related to malposition during general anesthesia

(1) Hyperabduction of arm, particularly if head rotated in opposite direction while muscle tone low

(2) Head tilted down, arm abducted and pressure on shoulder

b) Plexopathy related to median sternotomy

(1) 2.7-14% of coronary artery bypass surgeries

(2) Usual lower trunk/medial cord lesion (confuse with Ulnar nerve)

(3) Likely related to degree of sternal spread
(4) Restricted arm abduction recommended for prevention. Also can monitor with median nerve SEP during surgery.

c) Plexopathy related to shoulder surgery

(1) 1-8% of arthroscopy

d) Plexopathy from regional anesthesia or arteriography

(1) Axillary or Interscalene brachial plexus block

(2) Puncture axillary artery- blood leaks into fascial sheath-hematoma or pseudoaneurysm, often progressive symptoms, diagnosis with ultrasound

e) Postoperative Acute Brachial Plexus Neuropathy (ABN)

(1) Develops a few days after surgery and associated with a new shoulder/arm pain

(2) May mimic an anterior interosseous neuropathy (ABN-variant)

4. Malignant disease and Brachial Plexopathy

a) Lymph node or bone metastasis compressing lower trunk/medial cord

(1) Breast, Lung and Lymphoma

(2) Serve medial arm pain

b) Pancoast syndrome

(1) Carcinoma at apex of lung invading lower trunk/medial cord

(2) Severe medial arm pain

(3) Often have Horner’s syndrome

(4) Weakness and sensory changes involving lower trunk in 1/3rd

c) Primary tumor of brachial plexus

(1) Peripheral nerve sheath tumors (PNST)

(2) Neurofibromas: often upper trunk/ lateral cord
(a) Pain, supraclavicular mass, plexiform

(3) Intraneural Perineurioma – rare

5. Radiation-Induced Brachial Plexopathy

(1) Uncertain pathophysiology but changes seen include: demyelination, vasa nervosa alterations, and fibrosis.

(2) May develop 6 months to 34 years after radiotherapy

(3) Breast cancer survivors: 56% developed brachial plexopathy after 19 years, and 92% after 34 years.

(4) Sensory and motor deficits, often with edema, pain is not common

(5) Rare, radiation induced nerve sheath tumor, 4-40 yrs later, usually malignant, painful, enlarging mass, and detect with imaging.

(6) Often upper trunk or diffuse plexus and rarely just lower trunk/medial cord

(7) EDX

(a) EMG- Myokymic discharges: if present, distinguish from tumor/met etiology; Fasciculations more often seen after radiation

(b) NCS- supraclavicular/root stimulation may reveal conduction block

(8) Imaging

(a) MRI is superior to CT, but tumor can be present without MRI changes, MRI epidural lesions argue for metastatic disease.

(b) If MRI inconclusive can get plain x-ray or bone scan to detect mets in bony structures.

(c) If MRI inconclusive consider PET scan.

(d) May need surgical exploration or biopsy to diagnosis.
Management

(a) No known treatment, and in 2/3 relentlessly progressive

(b) Surgery not effective and may be harmful

(c) Physical therapy to prevent frozen shoulder, contractures

6. Thoracic Outlet Syndrome (TOS)

a) True Neurologic TOS

(1) 1: million, F-9: M-1 ratio

(2) Associated with down-turned C7 transverse process and fibrous band from C7 to rudimentary 1st rib; the C8/T1 rami or lower trunk are stretched over this fibrous band resulting in axonal loss, however, presence of above fibrous band does not mean thoracic outlet syndrome exists

(3) Abnormal musculotendinous anomalies found in 62% of cadaver dissections without causing a known, neurologic compression

(4) Pain leads paresthesias, weakness/atrophy later

(5) Pain aching, poorly localized (whole arm) worse with use (especially carrying heavy items)

(6) Sensory loss and paresthesias in Ulnar and medial cutaneous nerve distributions

(7) Motor dysfunction vague weakness/clumsiness of fingers later atrophy of all hand muscles (thenar atrophy noted first but Ulnar hand muscles usually weak as well) and may even cause atrophy flexor forearm

b) Thoracic outlet syndrome, vascular insufficiency:

(1) NO NEUROLOGIC abnormalities

(2) Sensations of coldness and aching

(3) Pallor with dependency
(4) Weakness with continued use (reduced exercise tolerance, not fixed weakness)

(5) Subclavian artery narrowed with post-stenotic dilatation where thrombus can form cause distal emboli

(6) Axillary/subclavian vein thrombosis, associated with arm edema

c) Thoracic outlet syndrome, work up

(1) Usual vascular testing maneuvers doubtful specificity, Adson’s maneuver (abduction and external arm rotation) and EAST test result in false positives in many healthy subjects

(2) Nerve conduction studies (distal) mainly to exclude focal median/Ulnar entrapments;

   (i) Ulnar, and medial cutaneous sensory, amplitude may be reduced

   (ii) Reduced median, and sometimes Ulnar, motor CMAP amplitude

(3) F-response minimally prolonged (WNL, reduced compared to opposite)

(4) EMG-abnormal lower trunk innervated muscles, particularly APB muscle should be abnormal


   a) Any age, usually unilateral, but can be bilateral

   b) Not uncommon to be preceded by infection or immunization (particularly tetanus toxoid)

   c) May be seen in patients with Diabetes Mellitus, systemic lupus, and polyarteritis nodosa

   d) 10% of HNPP patients may have brachial plexopathy

   e) Onset with shoulder area pain, often severe, lasting hours to 4 weeks
f) Weakness develops 24 hrs to 4 weeks after pain onset.

g) Areas of weakness and severity are very variable

(1) Serratus anterior, supraspinatus and infraspinatus muscles, biceps, brachioradialis and deltoid most commonly involved, but any muscle can be involved

(2) Suggest upper trunk with most common nerves: long thoracic, suprascapular, axillary, radial, anterior interosseous and phrenic nerves

h) Sensory abnormalities less marked than weakness and pain, but 2/3 complain of paresthesias and even more have objective sensory loss on exam with most common area of sensory loss in axillary nerve distribution.

i) Variants of sporadic ABN:

(1) Anterior interosseous neuropathy- suspect from motor fascicular involvement of the proximal median nerve, may also include Pronator teres muscle and lateral antebrachial cutaneous nerve (not part of AIN)

(2) Phrenic neuropathy- usually unilateral with hemidiaphragm involved, but often dyspnea and/or orthopnea, consider diagnosis of ABN in acute onset of dyspnea

(3) ABN with cranial neuropathy- most common facial palsy

(4) Sensory symptoms only with on the Median nerve and medial antebrachial cutaneous nerve involved (cohort of 8 patients)

j) Management

(1) Differentiate from cervical radiculopathy with EDX

(2) If concern for malignancy, chest image

(3) Analgesics for pain control and physical therapy to prevent frozen shoulder
8. Hereditary Acute Brachial Plexopathy

(1) Rare, autosomal dominant, mapped to 17q25 SEPT9 gene

(2) Onset 20-30’s, recurrent episodes may be triggered by pregnancy, infections, strenuous exercise, stress

(3) May involve cranial nerves: VII, VIII, X

(4) Dysmorphic features: hypotelorism, epicanthic folds, cleft palate, syndactyly, dysmorphic ears, short stature

II. Lumbosacral Plexopathies

A. Anatomy – Lumbar

1. L1, 2, 3 and part of 4, ventral rami = lumbar plexus within psoas muscle.

2. Anterior (ventral) division L2-4 forms Obturator nerve.

3. Posterior (dorsal) division of L2-4 forms femoral nerve.

4. Lumbar plexus gives off the iliohypogastric, ilioinguinal, genitofemoral and lateral cutaneous nerve of the thigh.

5. Lumbosacral Trunk- axons from L4 and all of L5- pass over ala of sacrum adjacent to SI joint to join sacral plexus

B. Anatomy – Sacral

1. S1, 2, 3, 4 ventral rami joining Lumbosacral trunk to become Lumbosacral plexus

2. Dorsal L4, 5 lumbosacral trunk and dorsal S1, 2 form lateral Sciatic nerve which forms peroneal nerve
3. Ventral L4-S2 from medial Sciatic nerve which forms Tibial nerve

4. Superior (L5-S1) and inferior (S1,2) gluteal nerves, pudendal nerve (S2, 3), posterior cutaneous nerve of the thigh (S1,2,3), external anal sphincter (S4) arise from sacral plexus

C. Pathophysiology

1. Retroperitoneal Hemorrhage: if awake, patient reports severe abdomen & groin pain often radiating into thigh with development of lower limb paresthesias and weakness
   a) Anticoagulant-warfarin, heparin, etc.
   b) Coagulopathy-hemophilia, thrombocytopenia, DIC
   c) Femoral artery catheterization-angiogram (I’ve had a case from a femoral vein catheterization from a central line)
   d) Aneurysm rupture- aorta or iliac artery
   e) Trauma- with abdominal injury or spinal fracture and hematoma
   f) Clinical Syndromes

   (1) Femoral Neuropathy-hemorrhage in iliacus compartment, most frequent syndrome
       (a) Weakness quadriceps, patellar reflex lost, sensory loss anterior thigh and anteromedial lower leg (iliopsoas may have pain limited weakness)

   (2) Lumbar plexopathy- hemorrhage within psoas
       (a) Weakness quadriceps, iliopsoas, hip adduction, reduced/absent patellar reflex and sensory loss in femoral, obturator and lateral femoral cutaneous nerves

   (3) Lumbosacral plexopathy-larger hematoma extension into sacral plexus
       (a) Weakness as described with lumbar plexopathy (above) plus L5 and S1 innervated- anterior and posterior tibialis, glutei, hamstrings, gastrocnemius with sensory loss in L5, S1 dermatomes
2. Diabetic lumbosacral radiculoplexus neuropathy (DLRPN) result from multifocal damage of spinal nerve roots, lumbosacral plexus and nerves derived from plexus (femoral); Immune mediated inflammatory disordered producing micro-vasculitis of the vasa nervosa resulting in nerve ischemia.

a) Type 1 or 2 diabetics, more often periods of poor control or weight loss but not exclusive to these periods, and may even be presenting feature of DM diagnosis

b) More common in older age and with coexistent nephropathy or retinopathy

c) Abrupt onset, severe pain, often unilateral and proximal

d) May have systemic symptoms of anorexia, malaise, weight loss

e) Unilateral weakness of proximal leg and hip girdle muscles but distal lower limb muscles may be affected in later course;

f) Femoral nerve innervated muscles most frequently affected, but with EDX and careful exam more extensive involvement can be found.

g) A few have bilateral lower limbs involved at onset; Involvement of the other side develops over median time of 3 months.

h) Numbness is variable and not as prominent as weakness

i) EDX testing: frequently demonstrates symmetric diabetic polyneuropathy

(1) Reduced or absent SNAPS

(2) Reduced or absent CMAPS with minimal conduction velocity slowing

(3) EMG fibrillations/p-waves in many muscles, weak and normal, often with paraspinal muscle involvement

j) Treatment

(1) Pain severe and may require opioids

(2) Strict blood sugar control advocated, but no evidence if improves recovery
(3) Most spontaneously improve, no clear role for immunotherapy

(4) Will need AFOs, walking aids and physical therapy

3. Nondiabetic Lumbosacral Radiculoplexus Neuropathy (LRPN)

a) Same clinical and EDX findings and clinical course as DLRPN (diabetic)

b) Reports of patients with LRPN having impaired glucose tolerance (IGT)

4. Pregnancy

a) Variety of disorders (L5 radiculopathy, lumbosacral trunk, sciatic, femoral or peroneal mononeuropathy) secondary to positioning, regional anesthesia or fetus

b) Lumbosacral trunk

(1) Pressure exerted on L-S trunk where it passes over the brim of the pelvis and becomes compressed between head of child and bone.

(2) Prolonged labor, cephalopelvic disproportion

(3) Foot drop (confused with peroneal) but careful exam & EDX confirm L5 abnormalities with abnormal posterior tibialis, glutei, EDH and preservation of paraspinal muscles.

(4) Full recovery in 5 months

5. Arterial Disorders

a) Distal Aortic aneurysms

(1) More often affect spinal cord but may cause ischemia to plexus associated with ischemic psoas

b) Common and internal iliac artery aneurysms

(1) Acute or chronic presentation with back/buttock/leg pain with plexus involvement

(2) May have vascular insufficiency in leg(s), often pulsatile mass palpated on pelvic or rectal examination
c) Thrombotic occlusion- distal aorta or iliac arteries
   (1) Acute low back pain and unilateral leg pain, numbness, weakness (confuse with radiculopathy)
   (2) Signs of arterial occlusion, i.e. cold/pallor often absent, but femoral and distal pulses are absent

d) Claudication of lumbosacral plexus
   (1) Exertional unilateral or bilateral pain in pelvis/buttock followed by numbness, paresthesias, weakness (confuse with lumbar spinal stenosis)
   (2) Stopping and resting rapidly resolves symptoms
   (3) No focal symptoms on exam at rest and EDX testing normal, as would see exam findings or abnormal EDX testing with lumbar spinal stenosis
   (4) Definitive test is angiography; often treated with stents/angioplasty

6. Trauma
   a) Fractures of the pelvic girdle, particularly sacrum
   b) Gunshot, stab wounds

7. Endometriosis
   a) Can infiltrate the S1-3 roots, sacral plexus, and proximal sciatic nerve
   b) Low back/gluteal pain, bladder dysfunction, sensory/motor in leg worse with menstrual cycle

8. Tumors
   a) Malignant tumors: uncommon and usually affect spinal cord, nerve roots or cauda equina
   b) If happens, tumors usually from abdomen/pelvic structures-colon, uterus, prostate, bladder, sacrum/psoas, lymphoma

9. Radiation Plexopathy
   a) Less frequent then radiation brachial plexopathy
b) 1 month - 31 yrs after XRT, median 5 yrs

c) Painless, weakness with less marked numbness/paresthesias

d) Have chronic neurologic dysfunction but symptoms do stabilize

e) EDX

(a) SNAP may be reduced or normal

(b) CMAP reduced with normal or slight slowing CV

(c) EMG features compatible with anterior horn cell or motor axonal damage, occasionally myokymic discharges

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- Lower trunk and medial cord may be difficult to distinguish, except medial pectoral nerve lower half of pectoralis muscle is in the lower trunk and not the medial cord
- Thoracodorsal nerve- Latissimus dorsi and Subscapular-Teres major are posterior cord and usually middle trunk.

- **Upper Trunk** = all the C5/6 innervated muscles plus Rhomboids, and may include PRT & FCR; SNAP(s) the median sensory to D1 (thumb) and lateral antebrachial cutaneous (LABC) are reduced. The musculocutaneous and axillary CMAPs may also be reduced.
• **Middle Trunk** = all the Radial innervated muscles excluding the Brachioradialis, FCR, PRT (lateral portion of median nerve) and latissimus dorsi; SNAP the median sensory to D2 (index) and D3 (ring finger) usually abnormal, and the radial sensory may also be abnormal.

• **Lower Trunk** = all the Ulnar innervated muscles and medial portion of the median nerve being all the median intrinsic hand muscles and FDP, FDS, FPL; SNAP Ulnar sensory to D5 (little finger) is usually abnormal. Can also add medial antebrachial cutaneous but abnormal only 65% or try the median sensory to D4 (ring finger) to differentiate lower trunk from Ulnar mononeuropathy.

• **Posterior cord** = Axillary nerve + All Radial innervated muscles, only radial SNAP abnormal

• **Lateral cord** = Musculocutaneous nerve and PRT and FCR, SNAP median to D1-3 and LABC responses affected with preservation of radial response.

• **Medial cord** = all Ulnar and medial portion of Median nerve, so same as lower trunk above.

REFERENCES