Peripheral Neuropathies

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Objectives:

- Weren't given

References:

Slides - Black "Team 433"

Doctor's notes - Red

Step up / davidson - Blue

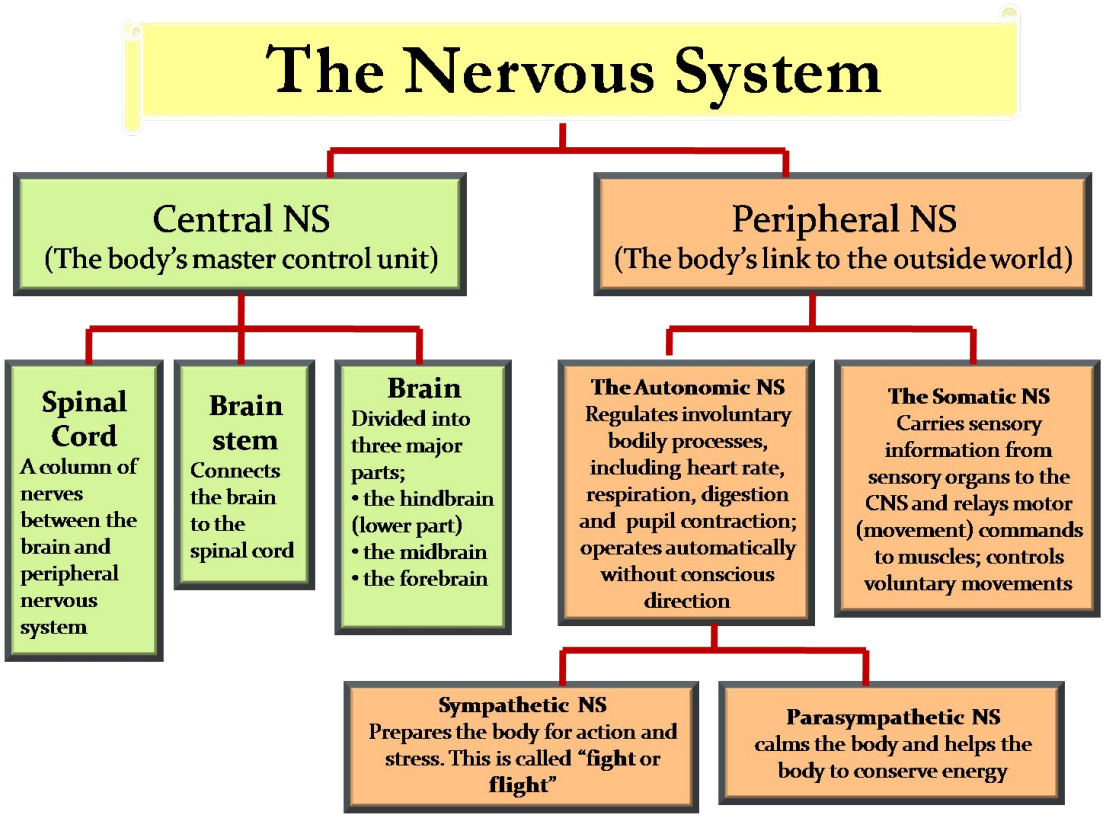
Extra explanation - Grey

Optional:

Chapter 5

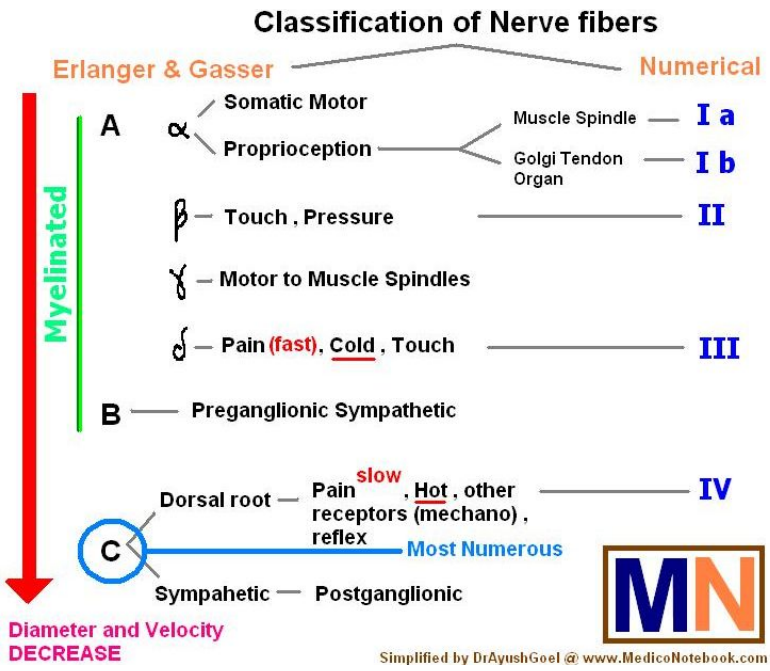
Editing file

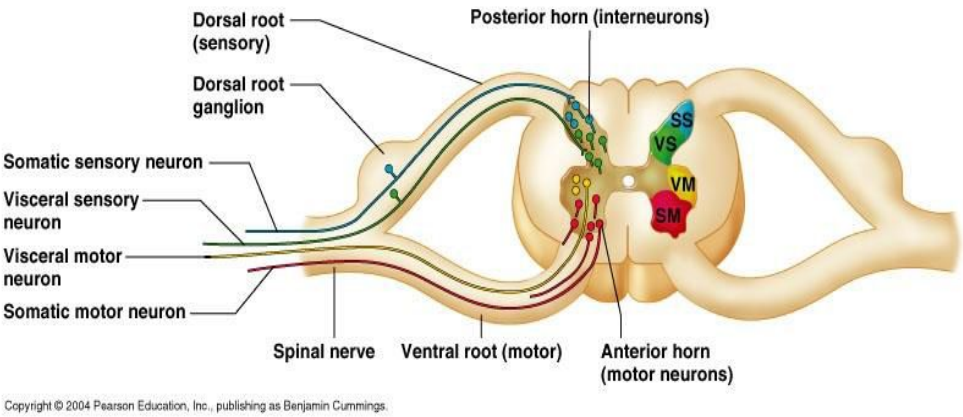
Anatomy



- **Peripheral Nervous System:**

There are two types of cells in the peripheral nervous system. These cells carry information to (sensory nervous cells) and from (motor nervous cells) the central nervous system (CNS). Cells of the sensory nervous system send information to the CNS from internal organs or from external stimuli. Motor nervous system cells carry information from the CNS to organs, muscles, and glands. The motor nervous system is divided into the somatic nervous system and the autonomic nervous system. The somatic nervous system controls skeletal muscle as well as external sensory organs such as the skin. This system is said to be voluntary because the responses can be controlled consciously. Reflex reactions of skeletal muscle however are an exception. These are involuntary reactions to external stimuli. The autonomic nervous system controls involuntary muscles, such as smooth and cardiac muscle. This system is also called the involuntary nervous system. The autonomic nervous system can further be divided into the parasympathetic and sympathetic divisions.





Motor Pathway	**Starts as the axons of anterior horn cells (in the spinal cord) come out through the ventral root. It's divided into somatic and autonomic nervous system)**
Sensory Pathway	**Starts from periphery where sensory cells receive stimuli and send it to the CNS. It enters the spinal cord through the dorsal root (passing through dorsal root ganglia)**

❖ Sensory tracts

Dorsal Column

(Gracile (inferior to t6) & Cuneate Fasciculi (superior to t6))

1. **Proprioception(Movement & Position of joints)**
2. **Discriminative touch from ipsilateral side**

- **Ipsilaterally**
- **Decussate 2nd order neuron Medulla (brainstem)**

Spinothalamic Tracts

1. **Lateral: Pain & Temperature.**
2. **Anterior: Crude Touch & Pressure sensation.**

- **Contralateral**
- **Decussate 2nd order Neuron Posterior grey horn (spinal cord)**

Spinocerebellar Tracts

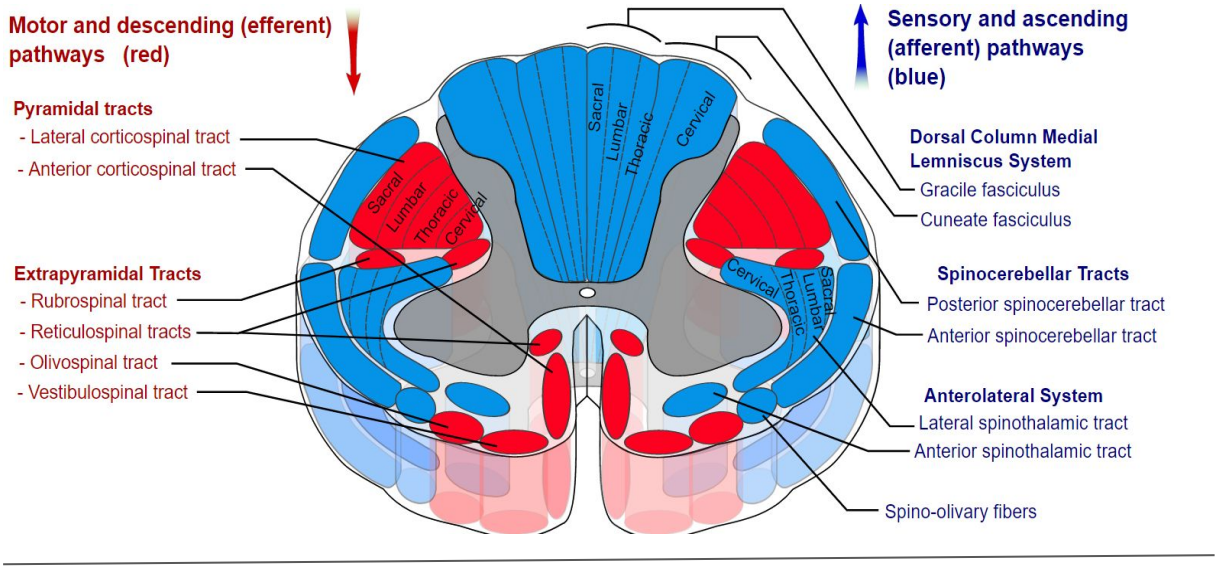
(posterior & Anterior)

Carry information derived from muscle spindles, Golgi tendon organs and tactile receptors to the cerebellum.

- **Ipsilaterally**
- **Posterior crosses twice**

1. **Spinal cord**
2. **Cerbellum**

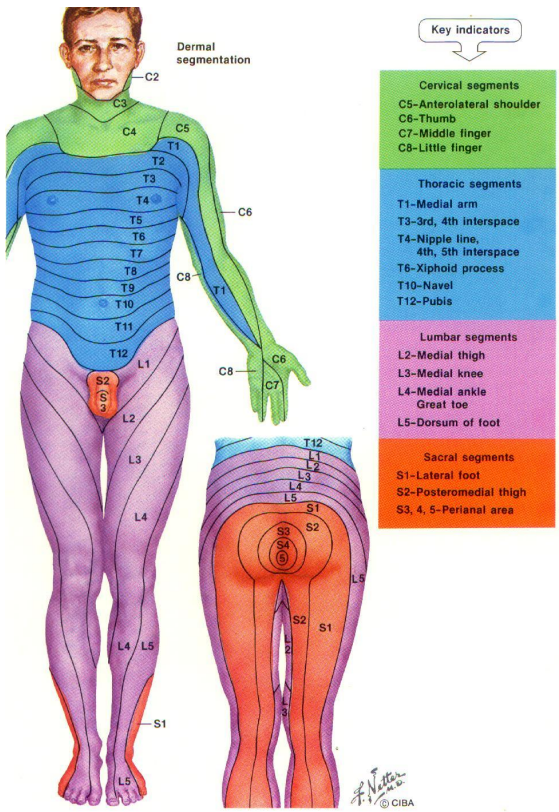
Sensory tracts Cont'd..



Dermatomes & Myotomes

***Important points:**

1. There is no C1 dermatome.
2. C4 and T2 dermatome are contiguous on trunk.
3. Thumb, middle finger, and fifth digits are innervated by C6, C7, and C8, respectively.
4. Nipple is at T4 level.
5. Umbilicus is at T10 level.
6. Lumbar and sacral dermatome are contiguous in the posterior axial line of leg.



- Remember each muscle is supplied by multiple roots but in this table the main ones are mentioned to help you in diagnosing or localizing the defect.
- A reflex called: supinator operator radialis reflex (brachioradialis reflex) can be decreased if there is a lesion at C6 and C7. (Assessment using a Hammer)
- After memorizing the dermatomes and myotomes try to link things in your mind so you get the whole picture. E.g. a patient presented with abnormal sensations in his thumb and weakness while trying to flex his elbow joint ⟶ C6 is probably affected.

Root	Muscle	Primary function
C3	Diaphragm	Respiration
C4	Diaphragm	Respiration
C5	Deltoid	Arm abduction
C5	Biceps	Forearm flexion
C6	Brachioradialis	Forearm flexion
C7	Triceps	Forearm extension
L3	Quadriceps femoris	Knee extension
L4	Quadriceps femoris	Knee extension
L4	Tibialis anterior	Foot dorsiflexion
L5	Extensor hallucis longus	Great toe dorsiflexion
S1	Gastrocnemius	Plantar flexion

Classifications

Definitions:

- **The terms "peripheral neuropathy," "polyneuropathy," and "neuropathy" are frequently used interchangeably, but are distinct.**
- **Peripheral neuropathy is a less precise term that is frequently used synonymously with polyneuropathy, but can also refer to any disorder of the peripheral nervous system including radiculopathies and mononeuropathies.**
- **Polyneuropathy is a specific term that refers to a generalized, relatively homogeneous process affecting many peripheral nerves, with the distal nerves usually affected most prominently.**
- **Neuropathy, which again is frequently used synonymously with peripheral neuropathy and/or polyneuropathy, can refer even more generally to disorders of the central and peripheral nervous system.**

Numerous inherited and acquired (most are acquired) pathological processes may affect peripheral nerves, targeting either: A. The nerve roots (radiculopathy). B. The nerve plexuses (plexopathy). C. And/or the individual nerves themselves (neuropathy).

Disorders may be primarily directed at : A. The axon. B. The myelin sheath (Schwann cells). C. Or the vasa nervorum (the vascular supply of the nerves).

An acute or chronic peripheral nerve disorder may be: A. Focal (affecting a single nerve: mononeuropathy).B. Multifocal (several nerves: mononeuropathy multiplex). C. Or generalized (polyneuropathy).

	Mechanism of damage	Example
Demyelination	**Schwann cell damage leads to myelin sheath disruption, This causes marked slowing of conduction**	**Guillain-Barre Syndrome** **HSMN (Hereditary sensory and motor neuropathy)**
Axonal degradation	**Usually associated with atrophy, most of chronic or metabolic diseases affect axons. Once the axons are affected. Its irreversible)**	**Toxic Neuropathies (Alcohol and excessive sugar)**
Wallerien degradation	**The axon starts to degenerate distally to the area of injury (opposing the cell body direction) and takes long time to regenerate**	**Trauma, cut**
compression	**Cause focal demyelination**	**Carpal tunnel Syndrome**
infarction	**Nerve have small blood vessels “vasa nervosum” these blood vessels affected in DM and arteritis, So nerves get infarcted**	**Arteritis, Polyarterisinosoda, DM, Atherosclerosis**
Infiltration	**Infiltration of peripheral nerves by inflammatory cells occurs in leprosy and granulomes (Nerves get damaged**	**Infiltration leprosy, Sarcoidosis**

History

Signs and symptoms

	Loss of function "Negative"	Altered function "positive"
Motor	**Wasting Hypotonia Weakness Hyporeflexia Orthopedic deformity**	**Fasciculations* Cramps**
Sensory (Large Fibers)	**↓ Vibration, ↓ proprioception, hyporeflexia sensory ataxia**	**Paresthesias**
Sensory (Small fibers)	**↓ Pain ↓ Temperature**	**Dysesthesias Allodynia****
Autonomic Nerves	**↓ Sweating Hypotension Urinary retention Impotence Vascular color changes**	**↑ Sweating Hypertension**

Note: the term positive or negative has nothing to do with the presence or absence of the symptoms. It only describes the symptom.
(*): Tiny movement caused by activation of single motor unit.
(**): Experience of pain from non-painful stimulation.

Important Notes: (Team 433)

- Motor nerves , sensory nerves, or autonomic nerves may be affected. More than one type of nerves may be affected at the same time (In GBS the patient has sensory/motor loss and autonomic too). The effect could be loss of function (negative) or inappropriate gain of function (Positive).
- Weakness isn't specific for LMN only, it can happen in UMN disorders as well, but in LMN lesions there are more sensory loss, hypotonia, hyporeflexia, and muscles atrophy which you don't find in UMN lesions.
- Fasciculation are found in LMN diseases and it's classical for ALS.

Q/ What's unique about peripheral nerve motor and sensory and sometimes autonomic?
Everything will fit in single peripheral nerve distribution, it doesn't fit with myotomal or radicular distribution and it doesn't fit with central distribution.

- What's central distribution?

Suppose you get a patient with hand numbness, how can I tell this hand numbness is median nerve (peripheral nerve), C6 (nerve root) or stroke, how can I tell the difference? By assessing the distribution, so If it is 3 fingers and a half and above the wrist is spared, I will think about median nerve and carpal tunnel syndrome, while If it is affecting also the forearm, I will think about nerve root, and If it is both sides of the hand but up to the wrist, so it doesn't fit with median, doesn't fit with ulnar, it can be stroke. So stroke or central pathology doesn't really go by single dermatomal or myotomal distribution, it will affect like half of the limb. So you have to think about the distribution.

A. Radiculopathy:

- **The location of the injury is at the level of the <u>nerve root</u>, This can result in pain (radicular pain), weakness, numbness, or difficulty controlling specific muscles. in a radiculopathy, the problem occurs at or near the root of the nerve, shortly after its exit from the spinal cord. However, the pain or other symptoms often radiate to the part of the body served by that nerve(the same root). For example, a nerve root impingement in the neck can produce pain and weakness in the forearm. Likewise, an impingement in the lower back or lumbar-sacral spine can be manifested with symptoms in the foot.**
- **Pain is a key finding.**
- **This affects a group of muscles supplied by a spinal root (myotome) and a sensory area supplied by a spinal root (dermatome). Therefore, the distribution of affected areas can help differentiate this from a peripheral neuropathy or a plexopathy.**
- **Patients may present with weakness, atrophy, and sensory deficits in a dermatomal pattern, may include fasciculations and diminished deep tendon reflexes.**

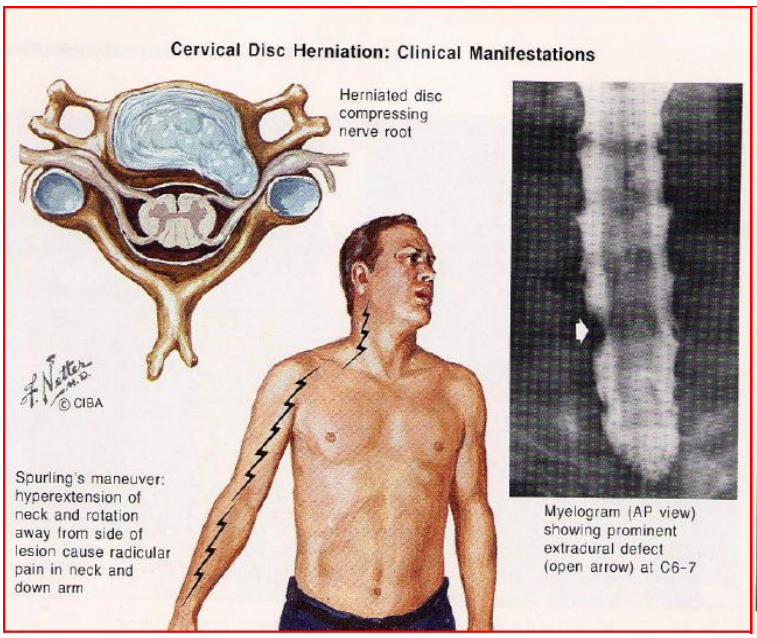
Common causes of Radiculopathies:

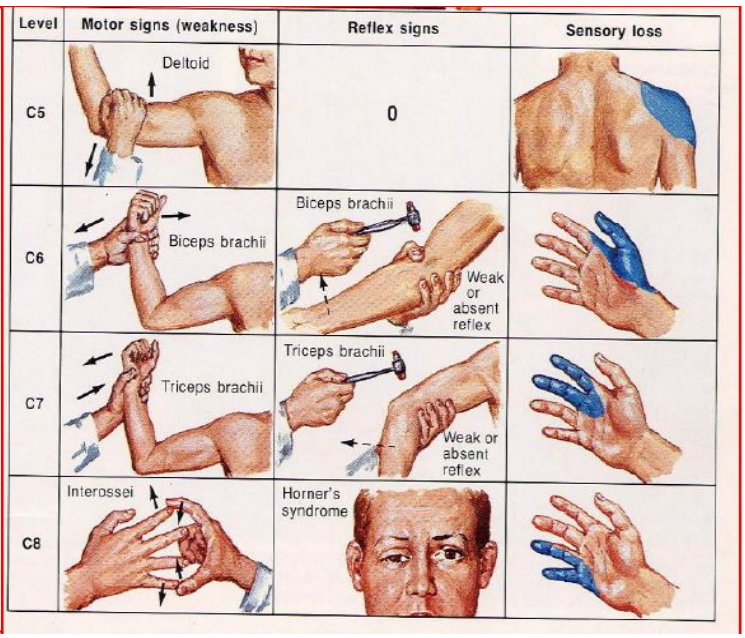
- **Compressive: herniated disc, spondylosis, tumor.**
- **Infiltrative: tumor seeding, infection.**
- **Inflammatory: immune-mediated.**

- To diagnose radiculopathy with through history and examination you must know :

1. **Dermatomes, part of skin supplied by single root.** **(page 5)**
2. **Myotomes, the muscles supplied by it.** **(page 5)**

1- Cervical Radiculopathy:





A. **Patients presented with weakness in abduction, numbness on shoulder area. Or Came with numbness on shoulder area only, you think of deltoid (C5) or it can be axillary nerve. To differentiate (by examination) check the myotome of C5 which is shoulder abduction and also it supplies the biceps (flexion) so biceps reflex weak or absent. But in axillary injury only the deltoid will be affected.**

B. **C7 triceps (extension) and supplies sensation to middle finger. Weak or absent reflex of the triceps.**

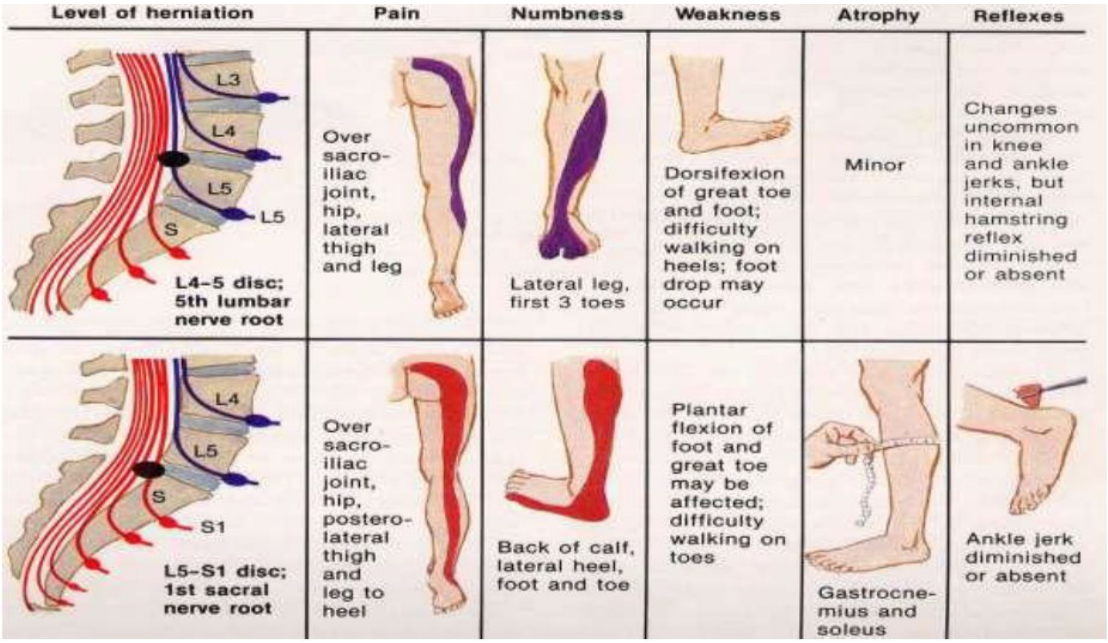
C. **C6 biceps (forearm flexion) and the sensation along the forearm laterally with thumb and index fingers. What else give sensation in this area? (Median! But median gives 3.5 fingers of the radial side, also control the flexors of the forearm and the muscles of the hands causing weakness).**

D. **C8 gives the interossei* (adduction and abduction and making a fist). Sensation: pinky and ring finger.**

(*) Group of muscles

***NOTE*: Supinator or brachioradialis reflex supplied by C6-C7 (can be decreased) you'd think of ulnar mononeuropathy too but the myotome supply is too small to distinguish ulnar clinically so you ask for EMG. EMG is also done in cases to know if the cause of damage is demyelination or axonal.**

2- Lumbar Radiculopathy:



- **Foot drop happens due to sciatic nerve injury or it can be L5-S1 radiculopathy .**
- **All branches come from the sciatic nerve so all the muscles here will be affected, dorsiflexion, plantar flexion, eversion, inversion.**
- **What also causes foot drop in UMN lesions are strokes in MCA "middle cerebral artery" (you differentiate through physical examination, if it's UMN lesion you'll find hypertonia, hyperreflexia..etc).**

Q/ How to differentiate between sciatic nerves injuries and L5-S1 radiculopathy? EMG

- **Sciatic nerve branches in the leg: Tibial nerve(plantar flexion and inversion),common fibular nerve(dorsiflexion and eversion).**
- **In lumbosacral radiculopathy L5-S1 are the most common injured ones.**

B. Plexopathy:

- **Deficits (motor and sensory) involve more than one nerve.**
- **Findings are variable depending on which part of the plexus is involved.**
- **Trauma is the most common cause overall, especially for the brachial plexus, while postsurgical hematoma in the pelvis is a more common cause in lumbosacral plexopathy.**
- **Plexuses that are commonly involved include:**

1. **Brachial plexus Erb–Duchenne type is the more common (upper trunk C5-6 roots) and less common lower trunk (C8-T1) .**
2. **Lumbosacral plexus (L5-S3).**

In plexopathy: the defect can be anywhere BUT ANYTHING PROXIMAL TO THE LESION WILL BE SPARED.
The nerve conduction for all sensory nerves will be normal in <u>radiculopathy</u> (why? because the cell body is spared) But in <u>plexopathy</u> all sensory and motor supply are affected cause the injury is after the dorsal ganglion (where the nerve becomes mixed).
When someone comes with mixed symptoms (sensory and motor), examine the proximal part, if intact you're probably dealing with plexopathy rather than radiculopathy .

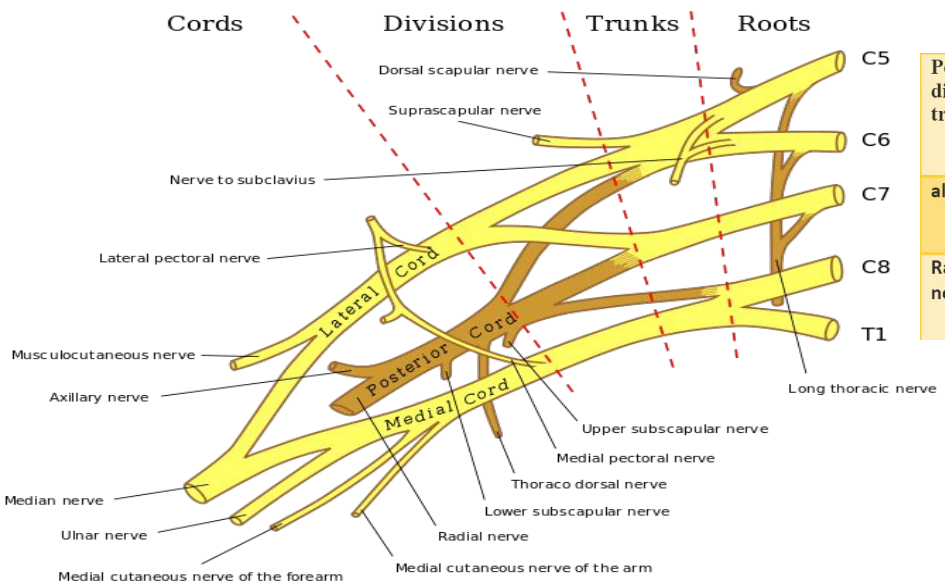
#Case: someone presents with numbness of both hands.
Put radiculopathy at the bottom of your differential.

Why?

1. **Bilateral at the same time , it won't push the nerve at the same time with all distribution it's not possible.**
2. **Nerves don't come out as one root, there are rootlets (it's not likely to compress all rootlets together at once)**

1- Brachial plexus:

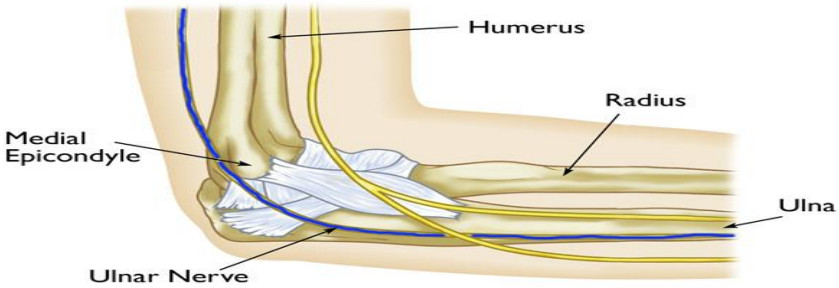
- **Roots:C5, C6, C7, C8, T1.**
- **Trunk: Upper (RootsC5&C6), Middle (RootC7), Lower (RootC8&T1).**
- **Division: Each trunk divides into anterior and posterior divisions.**



Posterior cord (post division of all trunks)	Medial cord (anterior division of lower)	Lateral cord (anterior division of upper and middle)
all roots	C8,T1	C5,C6,C7
Radial nerve Axillary nerve	Median nerve Ulnar nerve	Median nerve Musculocutaneous nerve

1- Brachial plexus Cont...

A- Ulnar nerve: most common injury cubital tunnel groove causing cubital tunnel syndrome (hanging the arm on a chair and sleeping on it).



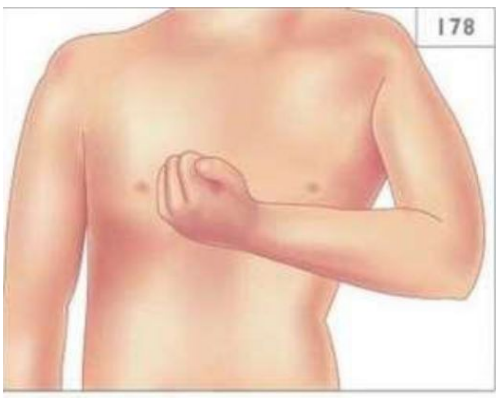
The second most likely site is at or near the wrist, especially in the area of the anatomic structure called the canal of Guyon. However, it can also occur in the forearm between these two regions, below the wrist within the hand, or above the elbow.

Most of the hand muscles are supplied by the ulnar except for the thenar.

So they will experience numbness and tingling along the little finger and the ulnar half of the ring finger accompanied by weakness of grip.

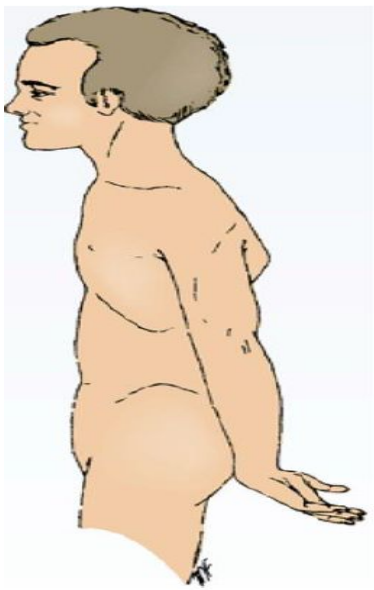
Causes of brachial plexopathy:

1. **Trauma**
2. **Tumor infiltration**
3. **Infection by viral**
4. **Immune-mediated**
5. **Delayed effects of radiotherapy**



178 Klumpke palsy.

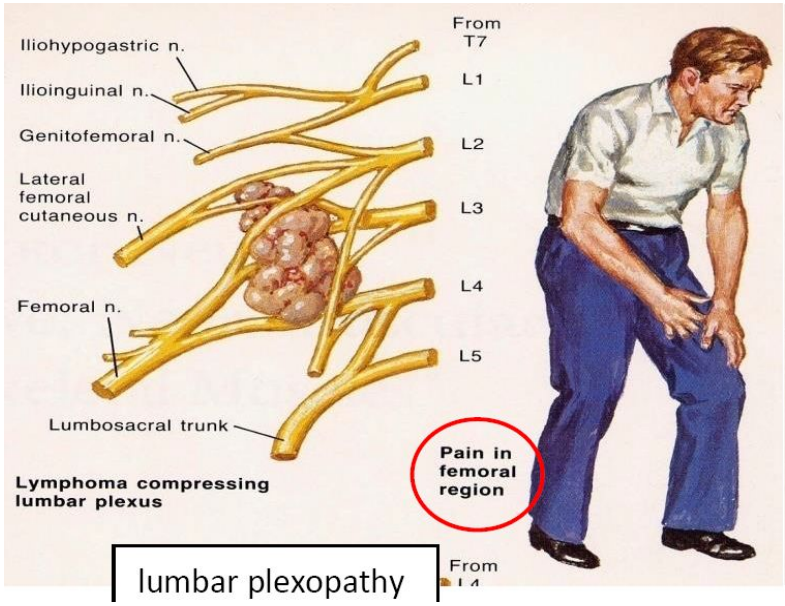
B- Klumpke's paralysis: Lower Lesions of the Brachial Plexus, (Klumpke Palsy)/Lower Trunk (C8,T1) Lesion.
The nerve fibers from this segment run in the ulnar and median nerves to supply all the small muscles of the hand. The hand has a clawed appearance due to ulnar nerve injury.

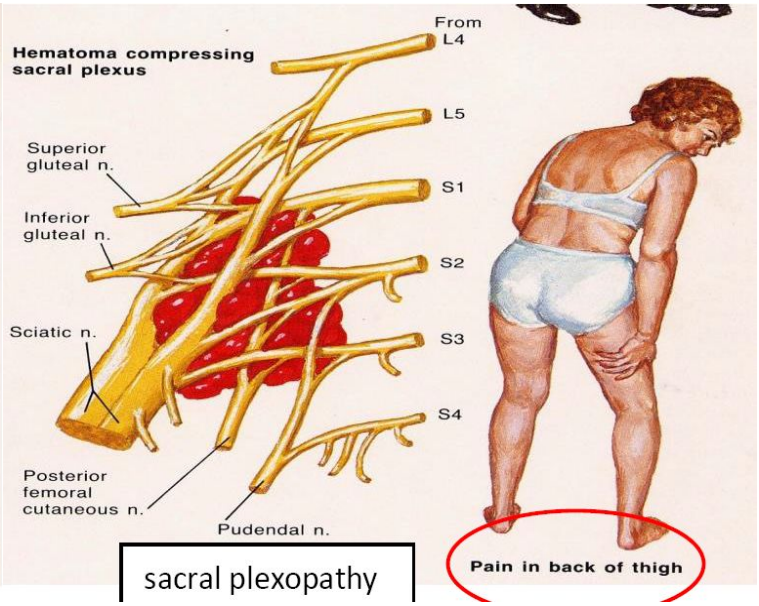


Erb's palsy

C- Erb's palsy: Upper Lesions of the Brachial Plexus Upper Trunk C5,6 (Erb-Duchenne Palsy "waiter's tip position". Resulting from excessive displacement of the head to the opposite side and depression of the shoulder on the same side (a blow or fall on shoulder).

2- Lumbosacral plexus:





Causes of lumbosacral plexopathy:

1. **Tumors : CA cervix, prostate, bladder, colorectal, kidney, breast, testis, ovary, sarcoma, lymphoma.**
2. **Compressed by aortic aneurysm.**
3. **Radiation plexopathy.**
4. **Plexitis : follow herpes zoster.**
5. **Diabetic amyotrophy.**
6. **Trauma (rare) .**

Clinical manifestations:

Patients usually present with asymmetric, focal weakness, numbness, dysesthesia, and/or paresthesia in multiple contiguous lumbosacral nerve root distributions.
Patterns of weakness usually help localize the "lesion" to a more specific area within the plexus.
Lumbar plexus lesions: tend to cause weakness of hip flexion and adduction and/or knee extension.
Lumbosacral trunk and upper sacral plexus lesions: result in foot drop or flail foot depending on the extent of involvement, and weakness of knee flexion or hip abduction.

Patterns of sensory disturbance are less reliable given the difficult clinical delineation between dermatomal and named nerve sensory loss. However, in general:
A. lumbar plexus: Sensory disturbance involving the anterior and medial thigh and medial leg.
B. lumbosacral trunk and/or sacral plexus lesion: Sensory disturbance involving the leg, dorsum of the foot, posterior thigh, and perineum.

C- lesions according to number of affected nerves:

A. (Mononeuropathy).
B. (Mononeuropathy multiplex).
C. (Polyneuropathy).

- Weakness is more prominent distally at the outset (as opposed to muscle myopathy usually asymmetric).
- Presents with diminished deep tendon reflexes, may include sensory changes
- (numbness, paresthesias, tingling), muscle atrophy, and fasciculations.
- Can be due to diabetes (nerve infarction), trauma, entrapment, or vasculitis.
- (The most common cause of peripheral neuropathy is diabetes mellitus).
- Common neuropathies include radial/ulnar/median/musculocutaneous nerves, long
- thoracic nerve, axillary nerve, common peroneal nerve, and femoral nerve.

A- Mononeuropathy:

- These neuropathies are recognized largely by clinical features. Diagnosis is confirmed by nerve conduction studies.
- People with these conditions can experience both weakness and numbness.
- Common causes: Diabetes, Vasculitis,Infection, Leprosy, Neoplasm, Brachial or lumbar plexopathy, Radiation, Sarcoidosis and Radiculopathies.

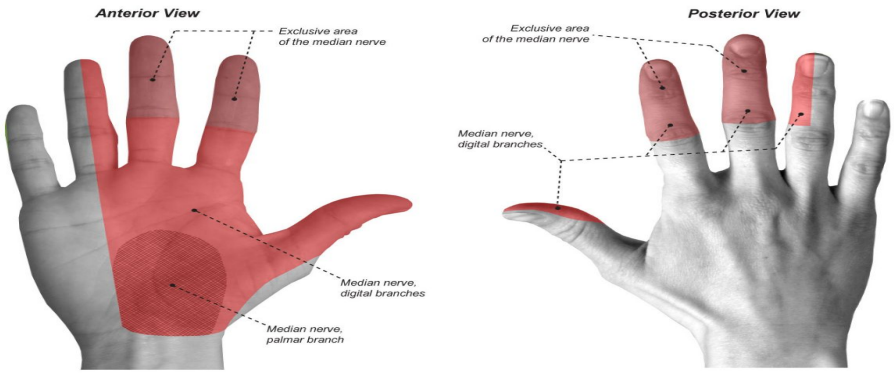
Table 21.52 Nerve compression and entrapment

Nerve	Entrapment/compression site
Median	Carpal tunnel (wrist)
Ulnar	Cubital tunnel (elbow)
Radial	Spiral groove (of humerus)
Posterior interosseous	Supinator muscle (forearm)
Lateral cutaneous of thigh	Inguinal ligament
Common peroneal	Neck of fibula
Posterior tibial	Tarsal tunnel (flexor retinaculum – foot)

In the next few slides will go through these nerves

1- Median nerve compression:

- The median nerve is derived from the medial and lateral cords of the brachial plexus.
- It contains fibres from all five roots (C5-T1).
- Three separate syndromes are recognised: Carpal tunnel syndrome, Pronator syndrome, and Anterior Interosseous syndrome.
- Hand of Benediction or Pop's Blessings (APE HAND) will result from median nerve injury.



Carpal tunnel syndrome:

Clinical features:

1. Nocturnal pain or paresthesia in the hand and/or forearm, thenar atrophy will be only in the hand (unlike Radiculopathy of C8 and T1)
2. Sensory loss in the palm and radial three-and-a-half fingers develops, followed by wasting of abductor pollicis brevis.
3. Tinel's sign*: Tinel's is elicited by tapping the flexor aspect of the wrist: this causes tingling and pain
4. Phalen's test positive*: In Phalen's, symptoms are reproduced on passive maximal wrist flexion.

- **Patient awakened by tingling and/or pain in the thumb, index and middle finger**
- **Gradual numbness of fingers while driving**
- **Atrophy of thenar muscles due to long- standing compression of median nerve**

Treatment:

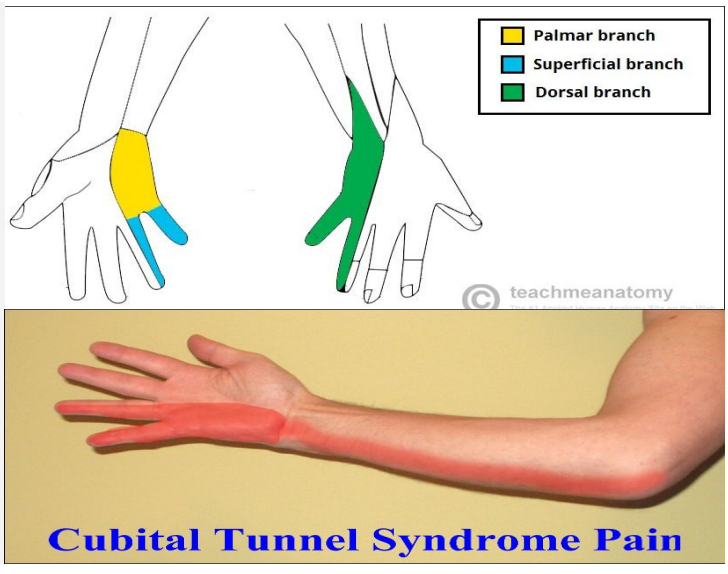
1. **Splinting – prevent wrist flexion.**
2. **Corticosteroid/anesthetic injection – give temporary relief.**
3. **Surgical decompression.**

2- Ulnar Nerve:

- **The ulnar nerve is derived from the brachial plexus. It is a continuation of the medial cord, containing fibres from spinal roots C8 and T1.**
- **Three lesions are recognized: Lesion at condylar groove, lesion at Guyon's canal and Lesion at wrist and hand.**
- **Lesion at condylar groove, which lead to (cubital tunnel)**

Cubital tunnel:

- **This follows prolonged or recurrent pressure and elbow fracture.**
- **Weakness and wasting of ulnar innervated muscles leads to clawing of the hand – hypothenar muscles, interossei and medial two lumbricals – with sensory loss in the ulnar one and a half fingers.**
- **Decompression and transposition of the nerve at the elbow is sometimes helpful.**

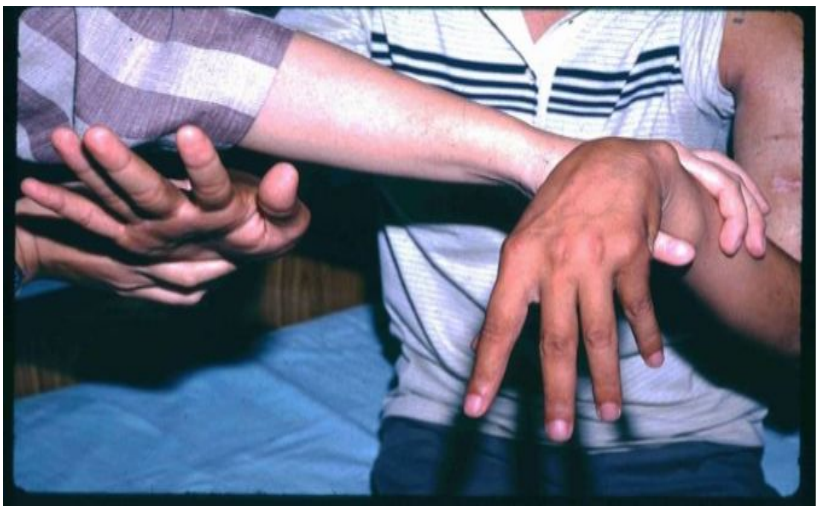


3- Radial Nerve:

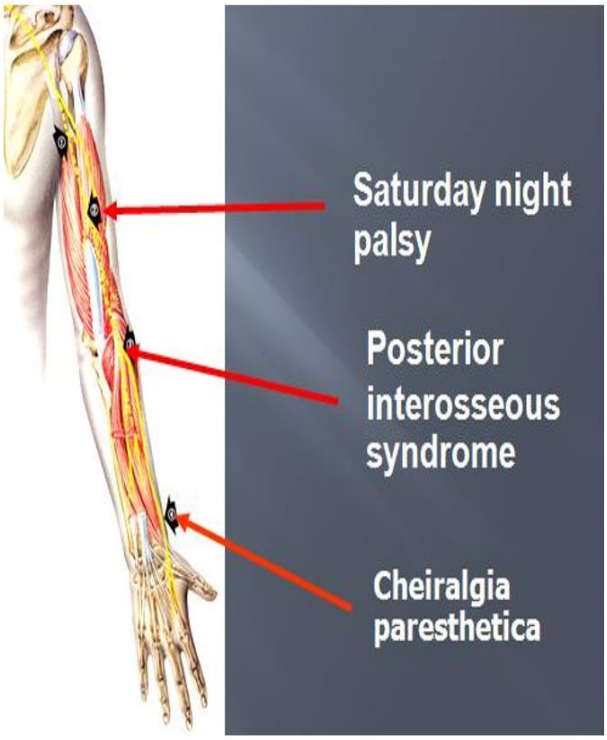
- **The radial nerve is a continuation of the posterior cord of the brachial plexus, containing nerve fibres from all 5 roots (C5 – T1).**

3 main lesions:

1. **Saturday night palsy: The terms Saturday night palsy and honeymooner's palsy refer to the concept of placing one's arm over another chair, with the resultant pressure causing injury to the radial nerve.**
2. **Posterior interosseous syndrome: (A pure motor branch of the radial nerve, no sensory loss)**
3. **Cheiralgia Paresthetica: (Sensory only).**



Wrist drop



Specific sites of lesions and its effects:

Axillary lesion: weak triceps and radial innervated muscles.

Mid-upper arm lesion: 'Saturday night palsy' (spiral groove or intermuscular septum), wrist drop, variable motor and sensory deficits (weakness of Brachioradialis and finger extension). with normal triceps, why ? because the affected part is always distal to the lesion site.

Posterior interosseous : weak extensor of thumb and other fingers, (wrist drop), no sensory loss without weakness of Brachioradialis.

Superficial radial nerve: terminal cutaneous branch:

A. Wrist drop of posterior interosseous nerve injury, the sensory supply will be intact.
B. Wrist drop of the spiral groove injury both sensory and motor of wrist and fingers will be affected.
C. In lower part, only sensory part of the radial will be affected at dorsum of the hand.

4- Femoral Nerve (L2,3,4)

- **Mix sensorimotor.**
- **Quadriceps femoris or knee extensor**

The lesions:

1. **Motor effect:**
   a) Wasting of quadriceps femoris.
   b) Loss of extension of knee Weak flexion of hip. (opposite to sciatic nerve).
   c) Weakness of hip flexor in intraabdominal lesion
2. **Sensory deficit: over anteromedial aspect of thigh and perhaps leg.**
3. **Absent or diminished knee jerk**

Lateral femoral cutaneous:

Meralgia Paresthetica: A painful mononeuropathy of the lateral femoral cutaneous nerve (LFCN), meralgia paresthetica is commonly due to focal entrapment of this nerve as it passes through the inguinal ligament, and it's pure sensory common condition seen in clinics "Pure sensory loss to the inferior iliac spine area"

Notes:

- **If it's sensory and motor defect at the same area (inferior iliac spine): It can't be femoral because the sensory branch of femoral which is the saphenous nerve extends till down so the sensation should be affected all the way down.**
- **But you think of radiculopathy (L2-L3) because they supply the motor and sensory part of that area but it should be asymmetrical to say it's radiculopathy, and if it was pure sensory it's meralgia paresthetica.**
- **Confirm by EMG.**
- **Whenever you have a problem in sensation in one area check it from other ways, it can be different nerve, it can radiculopathy, it can't be polyneuropathy here cause it's proximal defect not distal, neither mononeuritis multiplex because only one area is involved.**

5- Sciatic nerve (L4, S3):

- **The sciatic nerve is most frequently injured**
- **Composed of 2 main nerves of leg: common peroneal and tibial nerve.**
- **Paralysis of all muscles below knee plus hamstrings and for high lesion, external rotators of thigh.**
- **SCIATIC NERVE INJURY:**

1. **Motor effect: paralysis of hamstrings and all muscles of legs and foot. movement affected: Weak flexion of the knee, weak extension of hip and all movements of leg and foot.**
2. **Sensory effect:loss below knee except anteromedial aspect of leg and foot. Foot Drop (High steppage gait): which innervated by branch of femoral nerve.**

A. Common peroneal nerve:

- **Also called lateral popliteal nerve, affected by fracture or compressed against the head of the fibula after prolonged squatting, yoga, pressure from a cast, prolonged bed rest or coma, or for no apparent reason.**
- **Foot-drop and weakness of ankle eversion.**
- **Numbness develops on the anterolateral border of the dorsum of the foot and/or lower shin.**
- **Paralysis of anterior and lateral compartment of leg.**
- **Sensory loss over dorsum of foot and toes and anterolateral aspect of leg.**
- **Loss of dorsiflexion of ankle.**
- **The ankle jerk (S1) is preserved.**
- **Recovery is usual, though not invariable, within several months.**

B. Tibial nerve

- **Medial division of sciatic nerve.**
- **Lesions at ankle.**

Tarsal tunnel syndrome: when the tibial branch reaches the foot passing behind the medial malleolus and branches to medial and lateral tarsal nerves, there happens the common compression which causes burning and pain and discomfort.

- **It can happen in both feet.**
- **Pain and paresthesia in sole.**
- **Paralysis of intrinsic muscles of foot.**
- **Tenderness of Tinel's sign at flexor retinaculum.**

Sacral nerve compression syndrome:

- **Pure sensory.**
- **Numbness on lateral aspect of foot**

6- Facial nerve:

The lesions:
Bell's palsy: idiopathic, HSV1
Ramsay Hunt syndrome: external ear pain with presence of herpes zoster vesicles in auditory canal and pinna, VZV.
Trauma: blunt impact to temporal bone.
Middle ear infection: otitis media, mastoid pain persist after acute infection resolved.
Neoplasm: rarely compressed by CPA (Cerebellopontine Angle) tumor but due to surgery for tumor removal.

- **Bell's palsy: (idiopathic facial paralysis)**
  - **Bell's palsy is not the result of a stroke or a transient ischemic attack (TIA). While stroke and TIA can cause facial paralysis, there is no link between Bell's palsy and either of these conditions.**
  - **But sudden weakness that occurs on one side of your face should be checked by a doctor right away to rule out these more serious causes.**

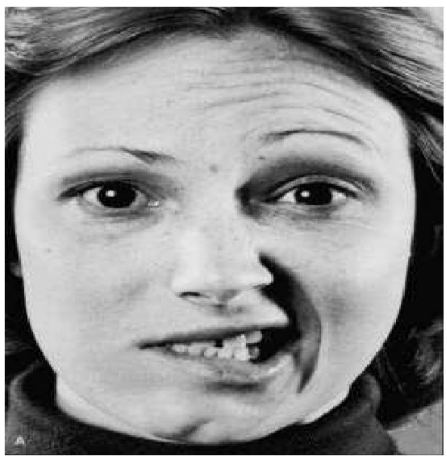
Always remember it's a mononeuropathy.
Facial nerve is LMN nerve (differentiate between upper and lower facial weakness)

- **Clinical features:**
  - **Postauricular pain (few days).**
  - **Lower motor neuron facial weakness (where all muscles of facial expression are affected on the side of the lesion).**
  - **Impaired taste.**
  - **Hyperacusis: caused by paralysis of the stapedius muscle, defined as intolerance of loud or high pitch sounds.**
  - **Difficulty with speaking.**
  - **Dryness of the eyes and mouth (decrease lacrimation and salivation).**
- **Diagnostic Tests:**

No test is usually done because of the characteristic presentation of paralysis of half of the face.
The most accurate test (if asked) is electromyography and nerve conduction studies.

- **Management:**

60% of patients have full recovery even without treatment.
The best initial therapy is prednisone. Acyclovir does not help.



2) Mononeuropathy Multiplex:

Definition: characterised by lesions of multiple nerve roots, peripheral nerves or cranial nerves

- **Mononeuritis multiplex: sometimes they're not explained by compression.**
- **Not only involve compressive nerves, but also the non-compressive.**
- **put in mind systemic diseases.**
- **This occurs in: DM, Leprosy, Vasculitis, Sarcoidosis.**
- **Diagnosis is largely clinical, supported by electrical studies.**
- **Several nerves become affected sequentially or simultaneously, e.g. ulnar, median, radial and lateral popliteal nerves.**
- **When multifocal neuropathy is symmetrical, there is difficulty distinguishing it from polyneuropathy. Where leprosy is prevalent, e.g. in India, a single nerve lesion can be the presenting feature.**

3) Polyneuropathy:

Q: How can we differentiate between axonal and demyelinating? A: by EMG nerve conduction study

1. **Axonal.**
2. **Demyelinating.**

- **Polyneuropathy is a generalized pathological process occurring in the longest peripheral nerves first, affecting the distal lower limbs before the upper limbs, with sensory symptoms and signs of an ascending 'glove and stocking' distribution.**
- **This is particularly true with axonal neuropathies where the disorder affects the metabolic processes required for axonal transport in the peripheral nerves.**
- **In inflammatory demyelinating neuropathies, the pathology may be patchier and variations from this ascending pattern occur.**

1- Axonal:

- **Acute:**
  - **AXONAL VARIANT OF GBS "Guillain-Barre Syndrome"**
  - **PORPHYRIA "disease of hemoglobin metabolism lead to sever abdominal pain and neuropathy"**
  - **Medication: NITROFURANTOIN**

- **Sub-acute:**
  1. **Toxins.**
  2. **Malnutrition.**
  3. **Systemic disease: DM, hypertension, HIV.**

- **Chronic: HMSN-II ' Hereditary Motor Sensory Neuropathy'**

2- Demyelination:

A) Uniform: -All nerve demyelinated . -Genetic\Hereditary diseases mainly cause UNIFORM.

B) Non-uniform: -Patchy demyelinating.

- **Acute AIDP 'acute inflammatory demyelinating polyneuropathy' > Diphtheria: serious bacterial infection.**
- **Chronic CIDP 'chronic inflammatory demyelinating polyneuropathy'**

Q: How can we differentiate between them?
A: By EMG. electromyography.

Q:Patient come polyneuropathy, What is the most important test?
A: EMG. electromyography.

Acute inflammatory demyelinating Polyneuropathy (AIDP):

Guillain-Barre Syndrome "in acute only":

- **Guillain-Barré syndrome (GBS) is an acute, frequently severe, and fulminant, polyradiculoneuropathy that is autoimmune in nature. Males are at slightly higher risk for GBS than females, and in Western countries adults are more frequently affected than children.**
- **The most common acute polyneuropathy.**
- **Acute inflammatory polyneuropathy is most commonly demyelinating (acute inflammatory demyelinating neuropathy, AIDP), but occasionally axonal and probably has an autoallergic basis. There is a predominantly cell-mediated inflammatory response directed at the myelin protein of spinal roots, peripheral and extra-axial cranial nerves. The resulting release of inflammatory cytokines blocks nerve conduction and is followed by a destruction of the myelin sheath and the axon. Paralysis follows 1–3 weeks after an infection that is often trivial and seldom identified. Campylobacter jejuni and cytomegalovirus infections are well-recognized causes of severe GBS. Infecting organisms induce antibody responses against peripheral nerves.**
- **The patient complains: weakness of distal limb muscles and/or distal numbness. These symptoms progress proximally, over several days to 6 weeks. In some cases respiratory and facial muscles become weak, sometimes progressing to complete paralysis. Autonomic features sometimes develop.**
- **GBS (AIDP) is the most common condition we see.**
- **Ten years later, it'll affect all the nerves even the proximal ones, CSF proteins will be high because it'll affect the roots and the roots are covered by protein which will reach the CSF in case of inflammation.**
- **Characteristics of GBS: cells will be normal and CSF proteins will be high.**

Subtype	Features	Electrodiagnosis	Pathology
Acute inflammatory demyelinating polyneuropathy (AIDP)	Adults affected more than children; 90% of cases in western world; recovery rapid; anti-GM1 antibodies (<50%)	Demyelinating	First attack on Schwann cell surface; widespread myelin damage, macrophage activation, and lymphocytic infiltration; variable secondary axonal damage
Acute motor axonal neuropathy (AMAN)	Children and young adults; prevalent in China and Mexico; may be seasonal; recovery rapid; anti-GD1a antibodies	Axonal	First attack at motor nodes of Ranvier; macrophage activation, few lymphocytes, frequent periaxonal macrophages; extent of axonal damage highly variable
Acute motor sensory axonal neuropathy (AMSAN)	Mostly adults; uncommon; recovery slow, often incomplete; closely related to AMAN	Axonal	Same as AMAN, but also affects sensory nerves and roots; axonal damage usually severe
M. Fisher syndrome (MFS)	Adults and children; uncommon; ophthalmoplegia, ataxia, and areflexia; anti-GQ1b antibodies (90%)	Demyelinating	Few cases examined; resembles AIDP

A. Clinical features:

1. Abrupt onset with rapidly ascending weakness/paralysis of all four extremities; frequently progresses to involve respiratory, facial, and bulbar muscles.
a. Usually symmetric (but not always).
b. Weakness may be mild or severe.
c. Weakness usually progresses from distal to central muscles.
d. If generalized paralysis is present, it can lead to respiratory arrest
2. Extremities may be painful, but sensory loss is not typical.
3. Sphincter control and mentation are typically spared.
4. Autonomic features (e.g., arrhythmias, tachycardia, postural hypotension) are dangerous complications.

B. Diagnosis:

- **Diagnostic Tests.**
- **The most specific diagnostic test is nerve conduction studies/electromyography. These will show a decrease in the propagation of electrical impulses along the nerves, but it takes 1–2 weeks to become abnormal.**
- **CSF shows increased protein with a normal cell count.**
- **Tests of Respiratory Muscle Involvement,when the diaphragm is involved, there is a decrease in forced vital capacity and peak inspiratory pressure. Inspiration is the "active" part of breathing and the patient loses the strength to inhale.**
- **PFTs tell who might die from GBS.**

C. Treatment:
Carefully monitor pulmonary function. Mechanical ventilation may be necessary.
Intravenous immunoglobulin (IVIG) or plasmapheresis are equal in efficacy.
Do not give steroids(Prednisone). They are usually harmful and never helpful in Guillain–Barré syndrome.

Chronic inflammatory demyelinating polyneuropathy (CIDP):

- **Multifocal motor neuropathy (MMN).**
- **Lewis-Sumner syndrome, also known as multifocal acquired demyelinating sensory and motor neuropathy (MADSAM).**
- **Distal demyelinating neuropathy with IgM paraprotein, with or without anti-myelin associated glycoprotein (anti-MAG).**
- **Demyelinating neuropathy with IgG or IgA paraprotein.**
- **POEMS syndrome (osteosclerotic myeloma: Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal protein, Skin changes).**
- **Sensory predominant demyelinating neuropathy.**
- **Demyelinating neuropathy with central nervous system demyelination.**
- **Demyelinating neuropathy associated with systemic disorders, including: Hepatitis B or C, HIV infection, lymphoma, diabetes mellitus, SLE/CTD, thyrotoxicosis, organ or bone marrow transplants, inherited neuropathies, nephrotic syndrome, inflammatory bowel disease.**
- **Presents with a relapsing or progressive generalized neuropathy. Sensory, motor or autonomic nerves can be involved but the signs are predominantly motor; a variant causes only motor involvement (multifocal motor neuropathy, MMN).**
- **CIDP usually responds to immunosuppressive treatment, corticosteroids, methotrexate or cyclophosphamide, or to immunomodulatory treatments (plasma exchange or intravenous immunoglobulin, IVIg); MMN is best treated by IVIg.**
- **Some 10% of patients with acquired demyelinating polyneuropathy have an abnormal serum paraprotein, sometimes associated with a lymphoproliferative malignancy.**
- **Symmetrical numbness and tingling occurs in hands and feet, spreading proximally in a glove and stocking distribution. Distal weakness also ascends. Tendon reflexes are lost.**
- **Nerve conduction studies show either axonal degeneration or demyelination, or features of both, and no underlying cause is found.**
- **Steroids isn't recommended except in CIDP.**

TABLE 4-3 Chronic Inflammatory Demyelinating Polyneuropathies: Classification By Weakness and Sensory Loss Patterns

Clinical Features	Classic Chronic Inflammatory Demyelinating Polyneuropathy	Distal Acquired Demyelinating Symmetric Neuropathy	Multifocal Acquired Demyelinating Sensory and Motor Neuropathy	Multifocal Motor Neuropathy
Weakness and sensory loss	Symmetric, distal and proximal	Symmetric, mild distal	Asymmetric, mostly distal	Purely motor, asymmetric, mostly distal
Elevated CSF protein	Yes	Yes	Yes	No
M protein	Uncommon, may have IgG or IgA	Usually IgM, rarely IgG	Uncommon	Uncommon, rarely IgM
Antineural antibodies	Uncommon	Anti–myelin-associated glycoprotein (MAG) in 50% of patients	Uncommon	Anti-GM_1
Motor nerve conduction studies	Demyelination	Demyelination	Demyelination	Demyelination
Sensory nerve conduction studies	Absent or small sensory nerve action potentials (SNAPs)	Absent or small SNAPs	Absent or small SNAPs	Normal
Treatments	Prednisone, IV immunoglobulin (IVIg), plasmapheresis, immunosuppressants	Possibly rituximab Some patients without anti-MAG may respond to IVIg	IVIg, immunosuppressants	IVIg, possibly cyclophosphamide

Electrodiagnostic Criteria for CIDP: These criteria are applied by testing the median, ulnar (stimulated below the elbow), peroneal (stimulated below the fibular head), and tibial nerves on one side of the body. During testing, limb temperature should be no less than 33°C at the palm and no less than 30°C at the external malleolus.

Definite CIDP
At least one of the following demyelinating parameters are necessary:
≥51 percent prolongation of motor distal latency above the ULN in two nerves
≥31 percent reduction of motor conduction velocity below the LLN in two nerves
≥21 percent prolongation of F-wave latency above the ULN in two nerves, or>50 percent if the amplitude of the distal negative peak CMAP is <80 percent of the LLN.
Absence of F-waves in two nerves, if these nerves have amplitudes of distal negative peak CMAPs ≥21 percent of the LLN, plus at least one other demyelinating parameter (meeting any of the definite criteria) in at least one other nerve
Partial motor conduction block, defined by a ≥51 percent amplitude reduction of the proximal negative peak CMAP relative to distal, if distal negative peak CMAP is ≥21 percent of the LLN, in two nerves, or in one nerve plus at least one other demyelinating parameter (meeting any of the definite criteria) in at least one other nerve.
Abnormal temporal dispersion, defined by a >30 percent duration increase between the proximal and distal negative peak CMAP in at least two nerves.
Distal CMAP duration (interval between onset of the first negative peak and return to baseline of the last negative peak) increase in at least one nerve (median ≥6.6 ms, ulnar ≥6.7 ms, peroneal ≥7.6 ms, tibial ≥8.8 ms) plus at least one other demyelinating parameter (meeting any of the definite criteria) in at least one other nerve.
Probable CIDP
≥31 percent amplitude reduction of the proximal negative peak CMAP relative to the distal, excluding the posterior tibial nerve, if the distal negative peak CMAP is ≥21 percent of LLN, in two nerves, or in one nerve plus at least one other demyelinating parameter (meeting any of the definite criteria) in at least one other nerve
Possible CIDP
As in "Definite CIDP" but in only one nerve.

Neuropathies with HIV infection:

A.Seroconversion:

- **Guillain Barre syndrome**
- **Chronic inflammatory demyelinating polyneuropathy (CIDP)**

B.Symptomatic stage: Mononeuritis multiplex axonal type subacute or chronic

C.Late symptomatic stage:

- **Distal symmetrical sensory polyneuropathy, most common neuropathy frequently coexists with symptomatic encephalopathy and myelopathy**
- **toxic polyneuropathy.**
- **subacute asymmetrical polyneuropathy of cauda equina, caused by cytomegalovirus**

Lesions localization

3 Functions of spinal cord: Motor, Sensory and Autonomic.

- **What's unique about spinal cord lesion localization?**
- **What's the hallmark When you bring a patient with a spinal cord injury?**
- **What is the most important question?**
- **Sensory level or the motor level (the level of injury).**
- **So the hallmark for the spinal cord injury to have a motor, sensory and autonomic deficit, this is the complete classic picture.**
- **Anything affecting the legs and sparing the arm, or it has a sensory level to the nipple it means spinal cord injury.**
- **Whenever you examine a patient and you are suspecting a spinal cord injury, try to identify the spinal level of injury, whether it is sensory level, anterior or posterior.**
- **Suppose a patient have weakness in both legs and normal upper, he also has spasticity in the legs with hyper-reflexia, where is the lesion? Dorsal or thoracic. It is not lumbar, because there is no spinal cord in the lumbar area, it is caudaequina (it is nerve roots so no spinal cord injury), so it will not cause hypertonia, it will cause hypotonia – nerve root injury.**
- **If you see the legs are affected with spasticity, then you know it is CNS lesion.**
- **If the arms are spared, then you know that the C spine is spared.**
- **What's the part of the CNS is below the C spine and above the lumbar? Dorsal or thoracic spine.**
- **You can identify it by the sensory examination, you do the sensory examination and you go anterior posterior for like pinprick and temperature until you identify the level.**

Continues...

- **You have to go by the map for the dermatomal and sensory level. The level for the nipple is T4 – T5. The level for the umbilicus is T10.**

- **Q: What about anterior horn cells, where are they located?**
- **A: Anterior horn of the spinal cord.**

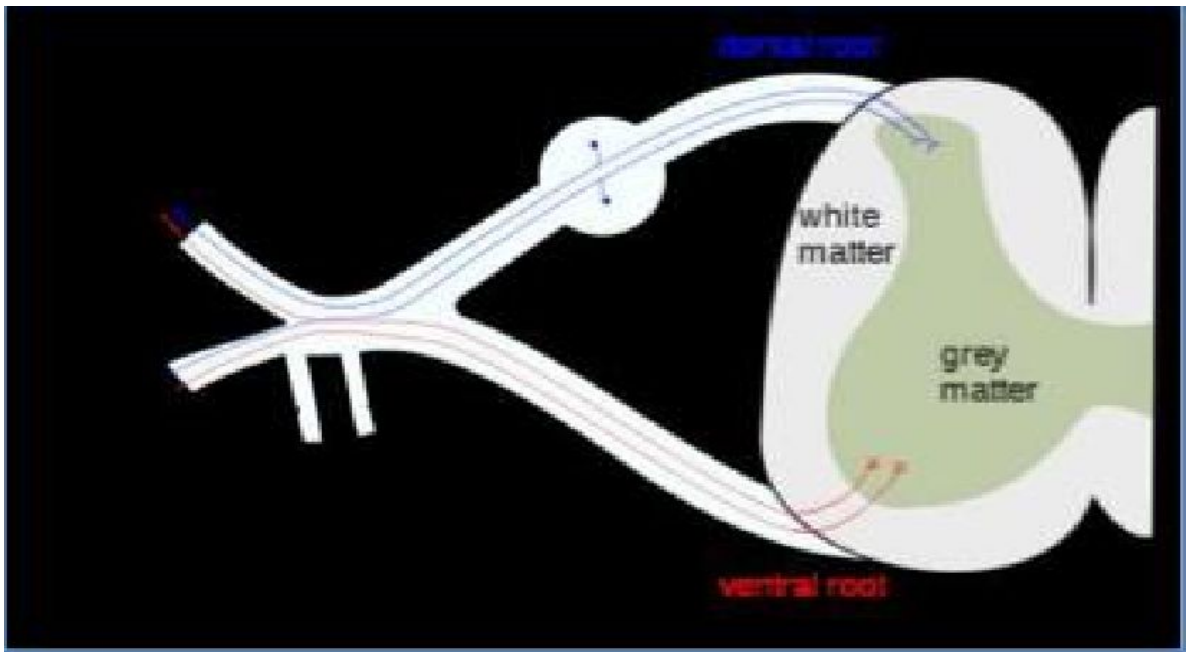
- **Q: What's the function?**
- **A: Motor.**

- **Q: Is it upper motor neuron or lower motor neuron?**
- **A: Of course lower motor neuron.**

- **When we say first and second order neuron, suppose I want to move my thumb here, the order has to come from the motor cortex in my right hemisphere, it will go down through the brainstem until it reaches C6 in the anterior horn level in the c spine (neck), then the anterior horn cell from there will make the radial nerve to the extension of the thumb. So all what I need is two levels, only single synapse in the spinal cord.**

- **What's the hallmark of anterior horn cell lesion or pathology like lower ALS (Amyotrophic lateral sclerosis) – common disease in the states- ?Flaccidity and muscle atrophy and sensory examination will be normal.**

- **This is the way you can differentiate between anterior horn cells or motor neuron disease from peripheral neuropathy. The sensory examination is normal, but in different neuropathy we will see motor and sensory deficit going with single peripheral nerve distribution or multiple nerve distribution. In anterior horn cells you will see pure weakness, fasciculation and atrophy, but sensory examination is totally normal, why? (see the pic.).**



Pattern recognition

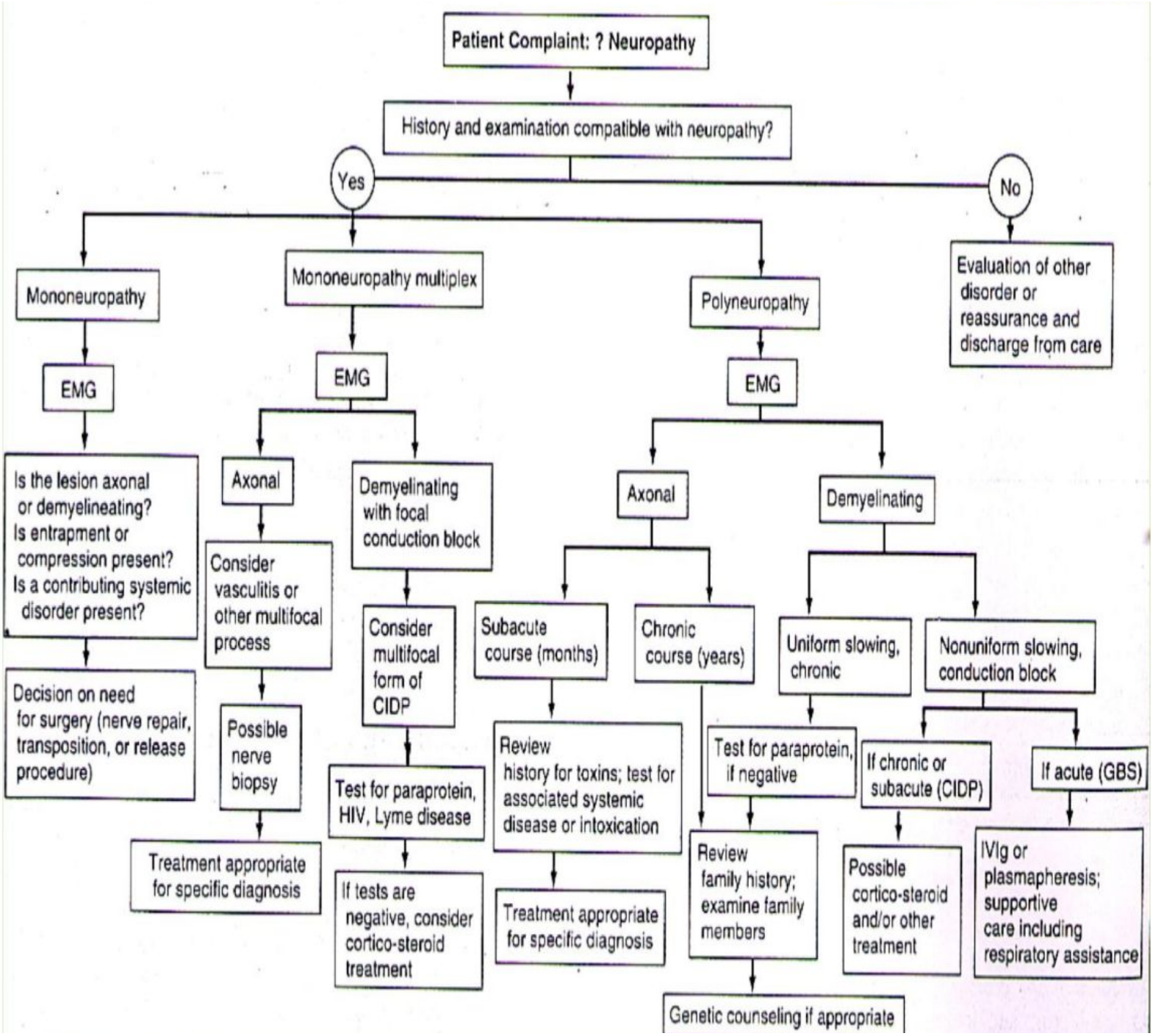
	Weakness								
	Prox	Dis	Asymm	Symm	Sensory Symptoms	Severe Proprioceptive Loss	UMN Signs	Autonomic Symps/Signs	Diagnosis
Pattern 1 - Symmetric prox & Distal weakness w/sensory loss	+	+		+	+				GBS/CIDP
Pattern 2 - Distal sensory loss with/without weakness		+		+	+				CSPN, metabolic, drugs, hereditary *
Pattern 3 - Distal weakness with sensory loss		+	+		+				Multiple – Vasculitis, HNPP, MADSAM, infection Single – Mononeuropathy, radiculopathy
Pattern 4 - Asymmetric prox & distal weakness w/sensory loss	+	+	+		+				Polyradiculopathy, plexopathy
Pattern 5 - Asymmetric distal weakness w/out sensory loss		+	+				+/-		+ UMN – ALS Pure UMN - PLS - UMN – MMN *
Pattern 6 – Symmetric sensory loss & upper motor neuron signs		+		+	+	+	+		B12 defic; Friedreich's, ALD
Pattern 7* Symmetric weakness without sensory loss	+/-	+		+					Prox & Distal SMA Distal Hereditary motor neuropathy
Pattern 8* Focal midline proximal symmetric weakness	+ Neck/extensor + Bulbar			+ +			+ +		ALS ALS
Pattern 9 – Asymmetric proprioceptive loss w/out weakness			+		+	+			Sensory neuronopathy (ganglionopathy)
Pattern 10 – Autonomic dysfunction								+	Diabetes, GBS, amyloid, prophyria *

*** = DM and hereditary neuropathy or chemotherapy**

*** = Multi focal neuropathy**

*** = hereditary autonomic neuropathy or DM**

Summary


Patient Complaint: ? Neuropathy
History and examination compatible with neuropathy?
Yes
No
Evaluation of other disorder or reassurance and discharge from care
Mononeuropathy
EMG
Is the lesion axonal or demyelineating? Is entrapment or compression present? Is a contributing systemic disorder present?
Decision on need for surgery (nerve repair, transposition, or release procedure)
Mononeuropathy multiplex
EMG
Axonal
Consider vasculitis or other multifocal process
Possible nerve biopsy
Treatment appropriate for specific diagnosis
Demyelinating with focal conduction block
Consider multifocal form of CIDP
Test for paraprotein, HIV, Lyme disease
If tests are negative, consider cortico-steroid treatment
Polyneuropathy
EMG
Axonal
Subacute course (months)
Review history for toxins; test for associated systemic disease or intoxication
Treatment appropriate for specific diagnosis
Chronic course (years)
Review family history; examine family members
Genetic counseling if appropriate
Demyelinating
Uniform slowing, chronic
Test for paraprotein, if negative
Nonuniform slowing, conduction block
If chronic or subacute (CIDP)
Possible cortico-steroid and/or other treatment
If acute (GBS)
IVIg or plasmapheresis; supportive care including respiratory assistance

MCQ's

Q1: A 31-year-old woman presents to accident and emergency with progressive difficulty walking associated with lower back pain. A few days ago she was tripping over things, now she has difficulty climbing stairs. She describes tingling and numbness in both hands which moved up to her elbows, she is unable to write. On examination, cranial nerves are intact but there is absent sensation to vibration and pin prick in her upper limbs to the elbow and lower limbs to the hip. Power is 3/5 in the ankles and 4−/5 at the hip with absent reflexes and mute planters. Her blood pressure is 124/85, pulse 68 and sats 98 per cent on air. She has a past medical history of type I diabetes and recently recovered from an episode of food poisoning a month or two ago. What is the diagnosis?

A. MS
B. Guillain–Barré syndrome (GBS)
C. Myasthenia gravis
D. Diabetic neuropathy
E. Infective neuropathy

Q2: A 52-year-old woman presents complaining of a two-month history of increasing fatigue and numbness in both of her arms and legs. She lives at home with her husband and has found it difficult coping with the daily activities of living. She suffers from hypothyroidism which is well controlled with thyroid replacement medication. A peripheral blood smear shows hypersegmented neutrophils. Which of the following is most likely the diagnosis?

A. Diabetic neuropathy
B. Brown-Sequard syndrome
C. Motor neurone disease
D. Hereditary neuropathy
E. Vitamin B12 deficiency

Q3: A female presents with diplopia. On closer examination, when asked to look right, her left eye stays in the midline but her right eye moves right and starts jerking. What is the diagnosis?

A. Myasthenia gravis (MG)
B. Vertigo
C. Cerebellar Syndrome
D. MS
E. Peripheral Neuropathy

Q4: On examination, a patient has 5/5 power in his upper limbs, 0/5 power in his lower limbs. Further examination reveals a sensory level at the umbilicus. Cranial nerves are intact. Where is the lesion?

A. C4
B. T4
C. T10
D. L1
E. L3

Answers: 1.B, 2.E, 3.D, 4.C

Thank you

If you have any question please contact with us at:
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