# Peripheral Neuropathies

**Objectives:**

- **Obtain informative history from a patient with peripheral neuropathy.**
- **Use clinical information to recognize different patterns of peripheral neuropathy**
- **Provide differential diagnosis for each pattern**

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**Resources:** 435 team + Davidson

- Editing file
- Feedback

# How to Approach patients?

★ **History Taking:** The goal of taking history from a patient with neuropathy is to classify their condition into one of the 10 patterns that we will discuss shortly after.

1. **Personal information**
2. **CC**

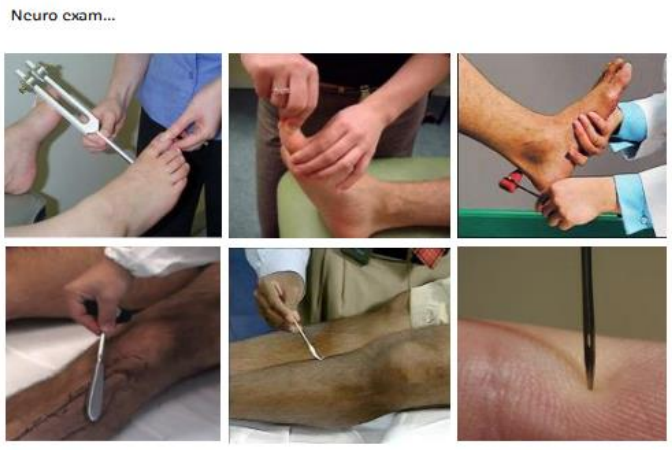
| | |
|---|---|
| **3. HPI** | **Symptoms:** Nerves have either motor, sensory or autonomic function.<br>Ask specific questions to identify if the lesion is related to which function.<br>• Tingling, burning, stabbing, throbbing, prickling, dead, icy, hot, clumsy, wooden sensory symptoms<br>• Weakness<br>• Distal leg:<br>-Tripping, stepping over curbs, uneven ground<br>• Proximal:<br>-Standing from sitting, walking up or down stairs<br>-Shaving, combing hair, brushing teeth<br>• Upper extremity predominant:<br>-Turning keys, opening jars distal , doing up buttons<br>• Autonomic: anhidrosis, excessive sweating, orthostatic light-headedness when he stands up , He gets annoyed from light for a long period of time because his pupil doesn't constrict , impotence, dry mouth, early satiety.<br>• Onset**:** <4 weeks Acute, 4-8 weeks sub-acute, >8 weeks Chronic<br>• Duration<br>• Were you a reasonable athlete as a child? Did you finish last in foot races? Were you able to skate or play soccer? To see if the pt. has inherited type of nerve disease<br>• Progression<br>-Chronic progression<br>-Acute deterioration to nadir then stability or improvement |
| **4.Past medical history** | • Diabetes (glucose intolerance) most common cause of neuropathy in SA<br>• Thyroid disease hypothyroidism can cause fluid retention which may exert pressure on peripheral nerves<br>• Renal failure / hepatic failure<br>• Malignancies<br>• Connective tissue disease<br>– SLE<br>– Rheumatoid arthritis<br>• Previous cervical or lumbar disc disease<br>• Previous entrapment neuropathies<br>– Multiple entrapments (consider HNPP hereditary neuropathy pressure palsy , amyloidosis)<br>• Orthopedic procedures on feet and ankles<br>• Exposure to toxin |

| | |
|---|---|
| **5.Family history**<br>They usually deny family history, so ask about each family member , if they have foot deformities for example and you will find a significant history . | • Detailed family history<br>– Walking difficulty, use of cane or wheelchair<br>– Postural or foot deformities<br>• Probe history of disabled or possibly affected individuals<br>• Do not necessarily accept what diagnoses other individuals have |
| **6.Social history** | • Exposure to alcohol common cause of neuropathy in western countries<br>• Occupation<br>• Tobacco<br>• Recreational drugs<br>• Vitamin and herb use<br>-In malnourished pts. Post gastric sleeve they get vit.b12 deficiency > nerve disease<br>-this table shows occupations and toxins that produce neuropathy. |
| **7. Review of systems** | • Joint pain, stiffness and swelling indicates connective tissue disease<br>• Fever<br>• Skin rash<br>• Other systems |

| Occupation | Neuropathy |
|---|---|
| Dentists | Nitrous oxide |
| Painters | Hexacarbons |
| Farmers | Organophophates |
| Welders | Lead |
| Jewelers | Arsenic |
| Plastic industry | Acrylamide |

## ★ Neurological examination:

- Confirm localization (LMN vs UMN reflexes are exaggerated + positive Babinski sign + increased muscle tone + different weakness pattern than LMN, myopathy vs neuropathy) **video**
- Recognize **pattern of neuropathy**
  - Motor vs sensory vs sensorimotor
  - Proximal vs distal
  - Symmetric vs asymmetric
- Recognize features of hereditary neuropathy
- Recognize features that narrows the differential diagnosis
  - Purpura and livedo reticularis may indicate renal failure
- Autonomic features
  - BP & HR supine and standing
  - Pupillary reaction to light and accommodation
- Other
  - Skin: trophic changes (such as thin, shiny, and discolored skin)
  - ulcerations or amputations.
  - peripheral pulses.



# Peripheral neuropathies

## ★ Concepts:

- Peripheral neuropathy is a disorder that occurs when your PNS malfunction because they're damaged or destroyed.
- Disorders of the peripheral nervous system are common and may affect the motor, sensory or autonomic components, either in isolation or combination.
- **The site of pathology may be:**
  - Nerve root (radiculopathy).
  - **Nerve plexus** (plexopathy)
  - **Nerve** (neuropathy).

## ★Pathophysiology:

- Damage may occur to the nerve cell body (axon) or the myelin sheath (schwann cell), leading to axonal or demyelinating neuropathies.
  - The distinction is requiring neurophysiology (nerve conduction studies and electromyography), and it is very important as only demyelinating disorders are usually susceptible to treatment.

## ★ Pathological classifications:

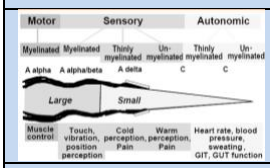
<table>
<tr><td colspan="5">Neuropathic disorders</td></tr>
<tr><td colspan="3">Neuronopathy (Pure; pure sensory or pure motor or autonomic)</td><td colspan="2">Peripheral neuropathy (mixed "sensorimotor")</td></tr>
<tr><td rowspan="2">Sensory<br>(ganglionopathies)</td><td rowspan="2">Motor<br>(motor neuron disease)</td><td rowspan="2">Autonomic</td><td>Myelinopathies</td><td>Axonopathies</td></tr>
<tr><td>e.g. GBS (Guillain–Barré syndrome)</td><td>e.g. Toxic neuropathies</td></tr>
</table>

## ★ Etiological classifications:

<table>
<tr><td>Acquired</td><td>Hereditary</td></tr>
<tr><td>Dysmetabolic states:<br>• Diabetes mellitus<br>• Neuropathy related to renal disease<br>• Vitamin deficiency states (eg, vitamin b12 deficiency)</td><td>Charcot-marie-tooth disease and related disorders</td></tr>
<tr><td>Immune mediated:<br>• Guillain-barre syndrome (GBS)<br>• Vasculitis e.g. (polyarteritis nodosa)<br>• Sarcoidosis</td><td>Hereditary sensory and autonomic neuropathy</td></tr>
<tr><td>Infectious<br>• Herpes zoster<br>• HIV</td><td>Hereditary neuropathy with liability to pressure palsy (hnpp)</td></tr>
<tr><td>Cancer related<br>• Malignant infiltration e.g (lymphoma)<br>• Primary amyloidosis</td><td>Familial amyloidosis</td></tr>
<tr><td>Drugs/ toxins<br>• Chemotherapy induced e.g. (cisplatin)<br>• Other drugs e.g. (amiodarone, INH)<br>• Heavy metals and industrial toxins e.g. (lead)</td><td>Motor neuron disease</td></tr>
<tr><td>Mechanical/compressive<br>• Radiculopathy<br>• Mononeuropathy</td><td></td></tr>
<tr><td>Unknown etiology<br>• Cryptogenic sensory and sensorimotor neuropathy</td><td></td></tr>
</table>

## ★ Clinical features:

Every nerve in your peripheral system has a specific function, so symptoms depend on the type of nerves affected.

| Symptoms | | |
|---|---|---|
|  | Loss of function "Negative" | Altered function "positive" |
| **Motor** | • Wasting<br>• Hypotonia<br>• Weakness:<br>  ○ **Proximal weakness:** difficulty to rise arm (to brush the teeth, comb the hair), as well as problems climbing stairs or rising from a chair.<br>  ○ **Distal weakness:** dragging of the foot while walking, لمن يرفع رجله تسقط<br>• Hyporeflexia<br>• Orthopedic deformity | • Fasciculations<br>• Cramps |
| **Sensory (Large Fibers)** | • ↓ Vibration,<br>• ↓ proprioception,<br>• Hyporeflexia<br>• Sensory ataxia | Paresthesia |
| **Sensory (Small fibers)** | • ↓ Pain<br>• ↓ Temperature | • Dysesthesias<br>• Allodynia |
| **Autonomic Nerves** | • ↓ Sweating<br>• Hypotension<br>• Urinary retention<br>• Impotence<br>• Vascular color changes | • ↑ Sweating<br>• Hypertension |

## ★ Investigations:

- The investigations required reflect the wide spectrum of causes. Neurophysiological tests are key in discriminating between demyelinating and axonal neuropathies, and in identifying entrapment neuropathies.
- Most neuropathies are of the chronic axonal type.

**26.101 Investigation of peripheral neuropathy**

**Initial tests**

- Glucose (fasting)
- Erythrocyte sedimentation rate, C-reactive protein
- Full blood count
- Urea and electrolytes
- Liver function tests
- Serum protein electrophoresis
- Vitamin $B_{12}$, folate
- ANA, ANCA
- Chest X-ray
- HIV testing

**If initial tests are negative**

- Nerve conduction studies
- Vitamins E and A
- Genetic testing (see Box 26.99)
- Lyme serology (p. 335)
- Serum ACE
- Serum amyloid

(ACE = angiotensin-converting enzyme; ANCA = antineutrophil cytoplasmic antibody; ANA = antineutrophil antibody)

# Pattern-recognition approach to neuropathy

I. Recognition of a clinical pattern.
II. There are 6 key questions the clinician should consider in arriving at the pattern that fits the patient best.
III. Most neuropathy and neuropathy patients can be placed into one of **10 patterns.**

**3 goals:**

1. To determine the **location** of the lesion.
2. To know the **cause** of the lesion.
3. To determine whether the **therapy** is possible?

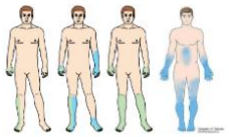
**6 Questions:** Easily defined clinical patterns of involvement are used to identify patients in need of neurologic consultation, whenever you're confronted with a case of peripheral neuropathy it is very necessary to ask yourself these 6 key questions.

## 1. What systems are involved?

- **Motor:** localized to AHC (anterior horn cell ), motor nerve roots, motor nerves . LMN are distal to AHC
- **Sensory:** DRG (dorsal root ganglion), sensory nerve roots, small nerves. Could be central.
- **Autonomic: autonomic nerves,** lateral **gray/white communicants**
- **Or Combinations.**

## 2. What is the distribution of weakness?

**A. Only distal versus proximal and distal**
**B. Focal/asymmetric versus symmetric**



| | |
|---|---|
| **Symmetric weakness** | • Represents a huge variety of DDx<br>• Symmetric **proximal** and **distal** weakness in a patient who presents with both motor and sensory symptoms -> CIDP and GBS. demyelinating disease<br>• Symmetric sensory and motor findings involving only the **distal** lower and upper extremities -> the disorder generally reflects a primarily **axonal peripheral neuropathy** and is much less likely to represent a treatable entity like diabetes |
| **Asymmetric /focal weakness** | • Motor neuron disease<br>• Radiculopathy<br>• Plexopathy<br>• Mononeuropathy or multiple mononeuropathies<br>• Compressive/entrapment mononeuropathies |

- The finding of weakness in both proximal and distal muscle groups in a symmetric fashion is the hallmark for acquired immune demyelinating polyneuropathies **(i.e. GBS guillian-barre) and the chronic form (CIDP chronic inflammatory demyelinating polyradiculoneuropathy).**

## 3. What is the nature of the sensory involvement?

- Pain: burning or stabbing
- Is the involved nerve fiber small or large? Small → Pain , light touch , pinprick & temperature .
- Large "faster than small" → Vibration, proprioception , ankle reflexes & joint position.
- Most neuropathies involve both small & large fibers.
- Severe proprioceptive loss

  – Central: dorsal column : Generally less profound proprioceptive loss , UMN signs

  -dorsal root ganglion is the first order neuron for all sensory modalities (large and small fibers) , so if a disease affects the dorsal root ganglia this will cause severe sensory loss .

  – Ganglionopathy: loss of all sensory modalities and reflexes.

**4. Is there evidence of upper motor neuron involvement?**

A. Without sensory loss (ALS, PLS)
B. With sensory loss (B12 def, copper, vit E, etc)

| | Upper motor neuron lesion | Lower motor neuron lesion |
|---|---|---|
| **Inspection** | Normal | Wasting, fasciculation |
| **Tone** | Increased with clonus | Normal or decreased, no clonus |
| **Pattern of weakness** | Preferentially affects extensors in arms, flexors in leg.<br>Hemiparesis, paraparesis or tetraparesis. | Typically focal, in distribution of nerve root or<br>peripheral nerve, with associated sensory changes |
| **Deep tendon reflexes** | Increased | Decreased/absent |
| **Plantar response** | Extensor (Babinski sign) | Flexor |

**5. What is the temporal evolution?**

A. Acute (days to 4 weeks)
B. Subacute (4–8 weeks)
C. Chronic (>8 weeks)
D. Preceding events, drugs, toxins

**6. Is there evidence for a hereditary neuropathy?**

A. Family history of neuropathy.
B. Skeletal deformities.
C. Lack of sensory symptoms despite sensory signs

## ★ 10 Phenotypic patterns: ★ PATTERN RECOGNITION SUMMARY

One can classify neuropathic disorders into several patterns based on sensory and motor involvement and the distribution of signs. Each syndrome has a limited differential diagnosis. A final diagnosis is arrived at by using other clues such as the temporal course, presence of other disease states, family history, and information from laboratory studies.

**Pattern1: Symmetrical proximal & distal weakness with sensory loss**

➢ **Inflammatory demyelinating polyneuropathy e.g. (GBS)**

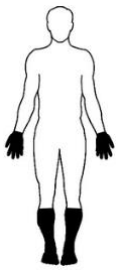
Guillain–Barré syndrome (GBS) is an immune-mediated condition (triggered by acute bacteria enteric infection).

- **Abrupt onset** (within less than 4 weeks) with rapidly ascending weakness/paralysis of all four extremities; frequently progresses to involve respiratory, facial, and bulbar muscles.
- Usually **symmetric** (but not always)
- Weakness:
  - Mild or severe.
  - Progresses from distal to central muscles.
- **If generalized paralysis is present, it can lead to respiratory arrest.**
- Extremities may be painful, usually show sensory loss on examination.
- Sphincter control and mentation are typically spared.
- Autonomic features (e.g., arrhythmias, tachycardia, postural hypotension) are dangerous complications.

**Pattern2: Symmetrical distal sensory loss with or without weakness.**

- **DM** diabetes is the most common cause of neuropathy

Frequently asymptomatic. The most common clinical signs (during examination) are,



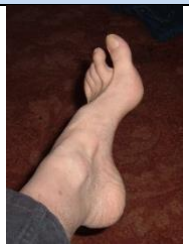
- **'Glove and stocking' impairment of all modalities of sensation (especially vibration)**
- Loss of tendon reflexes in the lower limbs.

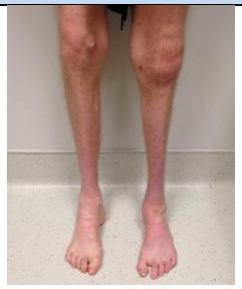
In symptomatic patients, sensory abnormalities are predominant. Symptoms (patient complain) include,

- Paresthesia in the feet (and, rarely, in the hands)
- Pain in the lower limbs (dull, aching and/or sharp, worse at night, and mainly felt on the anterior aspect of the legs)
- Burning sensations in the soles of the feet.
- Abnormal gait (commonly wide-based) often associated with a sense of numbness in the feet.
- Weakness and atrophy, in particular of the interosseous muscles, may develop, leading to,
  - Structural changes in the foot with loss of lateral and transverse arches.
  - Clawing of the toes and exposure of the metatarsal heads.
- This results in increased pressure on the plantar aspects of the metatarsal heads, with the development of callus skin at these and other pressure points.
- Electrophysiological tests demonstrate slowing of both motor and sensory conduction, and tests of vibration sensitivity and thermal thresholds are abnormal.
- **Note: autonomic neuropathy is common in diabetics & is not necessarily associated with peripheral somatic neuropathy. Parasympathetic or sympathetic nerves may be predominantly affected in one or more visceral systems. (e.g. postural hypotension, erectile dysfunction.)**

- CSPN (predominantly sensory polyneuropathy with no identifiable cause)
- Drugs induced peripheral neuropathy.
- **Charcot-Marie-Tooth (hereditary)**

Charcot–Marie–Tooth disease (CMT) is an umbrella term for the inherited neuropathies. This group of syndromes has different clinical and genetic features. The most common CMT is the autosomal dominantly inherited CMT type 1. Common signs are distal wasting (**'inverted champagne bottle' legs**), often with **pes cavus (high arched)**, and predominantly motor involvement.





**Other signs:**

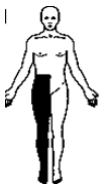
- Muscle weakness (especially in the foot and leg)
- Other foot deformities (hammertoes "curled toes")
- Muscle wasting (shrinking and weakness) in the legs
- Curved spine (scoliosis)
- Loss of pain sensitivity (upon examination)

**Pattern3: Asymmetric distal weakness with sensory loss.**

| | | |
|---|---|---|
| Multiple lesions: | ➢ Vasculitis<br>➢ Hereditary Neuropathy with Liability to Pressure Palsy (HNPP)<br>➢ Infection | |
| Single lesion: | ➢ **Radiculopathy** | Spinal **root** lesions (radiculopathy) **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.<br>**Etiology**<br>• Compressive: herniated disc, spondylosis, tumor.<br>• Infiltrative: tumor seeding, infection.<br>• Inflammatory: immune-mediated. |

**Pattern4:** Asymmetric proximal & distal weakness with sensory loss

- 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. A postsurgical hematoma in the pelvis is a more common cause in lumbosacral plexopathy.

**If it's not following the root pattern, then think of plexopathy.**

In plexopathy, the defect can be anywhere BUT ANYTHING PROXIMAL TO THE LESION WILL BE SPARED. One important thing, nerve conduction for all sensory nerves will be normal in radiculopathy (why? Because the cell body is spared) But in plexopathy 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.

**Plexuses that are commonly involved include:**

- Brachial plexus—Erb–Duchenne type is the more common (upper trunk—C5–6 roots). Lower trunk (C8-T1) is less common.
- Lumbosacral plexus (L5-S3)

**Pattern 5: Asymmetric distal weakness without sensory loss.**

| ➢ **With UMN:** | ➢ **Without UMN:** |
|---|---|
| ➢ **ALS (Amyotrophic lateral sclerosis)**<br>  <br>A disorder affecting the anterior horn cells and corticospinal tracts at many levels. **Corticobulbar involvement is common as well**. **The presence of upper and lower motor neuron signs is a hallmark of ALS.** Note that only the motor system is involved.<br>**Signs & symptoms:**<br>• Progressive weakness in legs & arms.<br>• Trouble with swallowing or speaking.<br>• Frequent muscle twitching & spams.<br>**Onset is usually between 50 and 70 years of age.** Occurrence of ALS before age 40 is uncommon. Only 10% of cases are familial, with the remainder being sporadic.<br>Prognosis is dismal: 80% mortality rate at 5 years; 100% mortality rate at 10 years. | MMN (Multifocal motor neuropathy) |

**Pattern 6: Symmetric sensory loss & upper motor neuron signs with proprioceptive loss.**

➢ **B12 deficiency (Subacute combined degeneration of the cord)**

Usually with underlying disorder that interferes with B12 absorption → Pernicious anemia

**Signs & symptoms:**

- Tingling, numbness & weakness in legs, arms & trunk.
- Change in mental status.
- Diminished sensations → Sensory ataxia.
- Positive Babinski sign
- Positive Rhomberg sign

**Treatment:** Reversible with B12 replacement

➢ ALD (Adrenoleukodystrophy)

**Pattern 7: Symmetric weakness proximal & distal without sensory loss.**

➢ SMA (spinal muscular atrophy)

**Pattern 8: Focal midline proximal symmetric weakness with upper motor neurons.**

➢ ALS

**Pattern 9: Asymmetric proprioceptive loss without weakness.**

➢ Ganglionopathy

**Pattern 10: Autonomic dysfunction.**

➢ (seen in DM & GBS)

| | Weakness | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|
| | Proximal | Distal | Asymmetric | Symmetric | Sensory Symptoms | Severe Proprioceptive Loss | UMN Signs | Autonomic Symptoms/ Signs | Diagnosis |
| Pattern 1: symmetric proximal and distal weakness with 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<br>Single: mononeuropathy, radiculopathy |
| Pattern 4: asymmetric proximal and distal weakness with sensory loss | + | + | + | | + | | | | Polyradiculopathy, plexopathy |
| Pattern 5: asymmetric distal weakness without sensory loss | | + | + | | | | ± | | LMN and UMN – ALS<br>Pure UMN – PLS<br>Pure LMN – MMN, PMA, BAD, LAD, MAMA |
| Pattern 6: symmetric sensory loss and upper motor neuron signs | | + | | + | + | + | + | | $B_{12}$ deficiency, copper deficiency, Friedreich ataxia, adrenomyeloneuropathy |
| Pattern 7[a]: symmetric weakness without sensory loss | ± | + | | + | | | | | Proximal and distal SMA<br>Distal Hereditary motor neuropathy |
| Pattern 8[a]: focal midline proximal symmetric weakness | +<br>Neck/extensor<br>+<br>Bulbar | | | +<br>+ | | | +<br>+ | | ALS |
| Pattern 9: asymmetric proprioceptive loss without weakness | | | + | | + | + | | | Sensory neuronopathy (ganglionopathy) |
| Pattern 10: autonomic dysfunction | | | | | | | | + | HSAN, diabetes, GBS, amyloid, porphyria, Fabry |

## Entrapment syndrome:

- Focal compression or entrapment is the usual cause of a mononeuropathy.
- Symptoms and signs of entrapment neuropathy are listed in Box 26.102. Entrapment neuropathies may affect anyone, but diabetes, excess alcohol or toxins, or genetic syndromes may be predisposing causes. Unless axonal loss has occurred, entrapment neuropathies will recover, provided the primary cause is removed, either by avoiding the precipitation of activity or by surgical decompression.

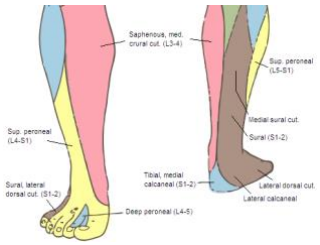
**26.102 Symptoms and signs in common entrapment neuropathies**

| Nerve | Symptoms | Muscle weakness/ muscle-wasting | Area of sensory loss |
|---|---|---|---|
| **Median** (at wrist) (carpal tunnel syndrome) | Pain and paraesthesia on palmar aspect of hands and fingers, waking the patient from sleep. Pain may extend to arm and shoulder | Abductor pollicis brevis | Lateral palm and thumb, index, middle and lateral half 4th finger |
| **Ulnar** (at elbow) | Paraesthesia on medial border of hand, wasting and weakness of hand muscles | All small hand muscles, excluding abductor pollicis brevis | Medial palm and little finger, and medial half 4th finger |
| **Radial** | Weakness of extension of wrist and fingers, often precipitated by sleeping in abnormal posture, e.g. arm over back of chair | Wrist and finger extensors, supinator | Dorsum of thumb |
| **Common peroneal** | Foot drop, trauma to head of fibula | Dorsiflexion and eversion of foot | Nil or dorsum of foot |
| **Lateral cutaneous nerve of the thigh** (meralgia paraesthetica) | Tingling and dysaesthesia on lateral border of thigh | Nil | Lateral border of thigh |

**Key-words:**

- Proximal weakness + Distal weakness + Symmetric + Sensory symptoms = **inflammatory** (GBS or CIPD – with acute presentation).
- Proximal weakness + Distal weakness + Asymmetric + Sensory symptoms = **Radiculopathy** (nerve roots supply multiple proximal and distal muscles
  - C7 nerve root supplies which muscle in the arm? Triceps, extensor digitorum communis (weakness → finger drop).
- Distal weakness + Symmetric + Sensory symptoms = **Metabolic** (vitamin deficiency), **Diabetic neuropathy**, **Idiopathic**.
  - length dependent, numbness, and شوية weakness.
- Distal weakness + Asymmetric + Sensory symptoms = **vasculitis with nerve infarct** (that's why it's asymmetric), **compressive**
- Upper motor neuron signs? **Cord involvement** (Myelopathy: deficiency or compressive) or motor neuron disease such as **ALS** that gives proximal & distal weakness.
- Autonomic symptoms = **Autonomic neuropathy**.
- Severe proprioceptive loss = **dorsal root ganglia** or **spinal cord** (dorsal column).

## Dermatomes of the foot: the dr. said the dermatomes are important he stressed on L5 , C7 , S1 !!



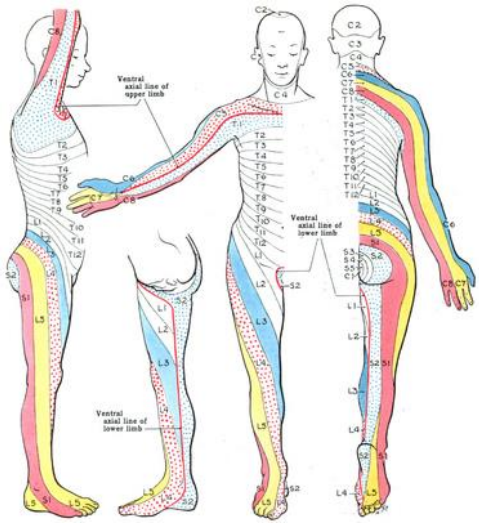

A dermatome is an area of skin that is mainly supplied by a single spinal nerve. There are 8 cervical nerves, 12 thoracic nerves, 5 lumbar nerves and 5 sacral nerves. Each of these nerves relays sensation (including pain) from a particular region of skin to the brain.

## Myotomes

- 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 reflex (associated with brachioradialis) can be decreased if there is a lesion at C6 and C7
- 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.
- Generally, you rule out UMN lesions by physical examination: absence of hypertonia and hyper-reflexia, no muscles atrophy, EMG (electromyograpgies) as well.

| TABLE 1: DERMATOMES | |
|---|---|
| C1 | Top of head |
| C2 | Face |
| C3 | Lateral neck |
| C4 | Lower neck and top of shoulder |
| C5 | Shoulder to base of thumb outside of arm |
| C6 | Front of shoulder, down arm, into thumb and back of hand |
| C7 | Back of shoulder, down back of arm, into back of hand |
| C8 | Little finger and wrist |
| T1 | Inside elbow down to wrist |
| L1 | Back and over trochanter |
| L2 | Back and anterior thigh to knee |
| L3 | Back, upper buttocks, anterior thigh and knee, inner lower leg |
| L4 | Inner buttocks, outer thigh inside of leg, and dorsum of foot and big toe |
| L5 | Buttock, back, side of thigh, lateral leg, dorsum of foot, inner half of sole of foot, and second and third toes |
| S1, S2 | Buttock, posterior thigh and lower leg |
| S3 | Groin and inner thigh to the knee |
| S4 | Genitals |

| TABLE 2: MYOTOMES | |
|---|---|
| C1, C2 | Cervical flexion |
| C3 | Cervical side flexion |
| C4 | Scapula elevation |
| C5 | Shoulder abduction |
| C6 | Elbow flexion and wrist extension |
| C7 | Elbow extension and wrist flexion |
| C8 | Thumb extension |
| T1 | Finger abduction |
| L1, L2 | Hip flexion |
| L3 | Knee extension |
| L4 | Ankle dorsiflexion |
| L5 | Big toe extension |
| S1 | Ankle plantiflexion |
| S2 | Knee flexion |

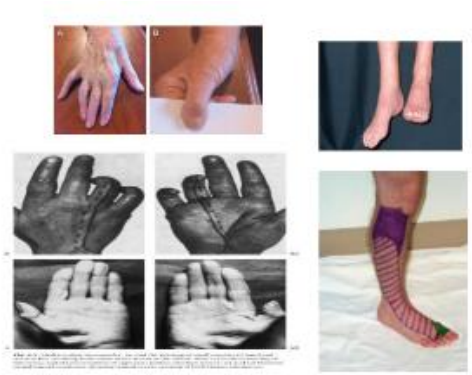


# Cases

-In the exam I will not ask you to give me the pattern, but I will ask you to give the DDx. The best way to reach the differential diagnosis is to identify the pattern of distribution before.
In any peripheral nerves case answer these questions to reach the pattern then the DDx.
1.What are the systems involved? (motor, sensory or both)
2. Where is the disruption of the weakness? (proximal, distal or both. Symmetrical or asymmetrical)
3. What is natural of sensory involvement?
4. Is there evidence of upper motor neuron involvement? (upper motor neurons or lower)
5. Onset (acute, subacute, chronic)
6. Is there any history of hereditary neuropathy?
-After answering these questions now you have to answer:
1. What is the pattern?
2. What is the DDX?
3. What is cause of that condition?

**Case 1:** A 65-year-old woman presented with a 3 month history of right-hand numbness, grip weakness, and vague elbow pain. Examination demonstrated diminished sensation of the medial hand and fourth and fifth digits, and weakness of finger abduction and adduction, associated with intrinsic hand muscle atrophy. Froment can't hold the paper as demonstrated in the pic and Wartenberg signs were evident.

**-DIAGNOSIS: Mononeuroathy (ulnar involvement)**



**1.What are the systems involved? (motor, sensory or both)**
Sensory and motor

**2. Where is the disruption of the weakness? (proximal, distal or both. Symmetrical or asymmetrical)**
Asymmetrical distal.

**3. What is nature of sensory involvement?**
Numbness, grip weakness, and vague elbow pain.

**4. Is there evidence of upper motor neuron involvement?**
No, Lower motor neuron only.

**5. Onset (acute, subacute, chronic)**
3 months = chronic.

**6. Is there any history of hereditary neuropathy?**
No.

So we have Asymmetrical distal weakness with Sensory loss. What is the pattern? Pattern 3.

What is DDx of pattern 3?

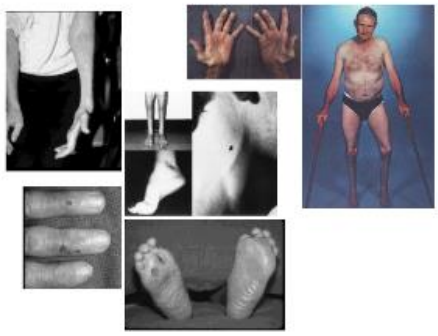
a. Vasculitis like (Mononeuritis multiplex) which is infarction to the nerve "nerve stroke " **severe and sudden**
b. Mononeuropathy : focal neuropathy due to compressive lesion and the most common is carpal tunnel syndrome "compression of median nerve" + ulnar nerve compression in the elbow which causes claw hand + common peroneal nephropathy which causes foot drop .
c. HNPP (hereditary neuropathy with liability to pressure palsy ): which is a hereditary dysfunction of the nerve .
How to differentiate between Vasculitis and Mononeuropathy or radiculopathy?

Vasculitis is **sudden** in onset and very tender (the patient will remember exactly when the pain started because its sudden and will come immediately because of severe pain)

**Case 2:** A 25-year-old man with no family history of neuropathy had been weak early childhood. He remembers he was unable to keep up with his peers when running. He is currently only able to walk if wearing ankle-foot orthosis. He denied sensory symptoms.

- Neurological examination :

symmetric severe weakness in distal leg muscles with power of 1-2/5 with bilateral drop feet. proximal leg muscles were 4/5 as well as intrinsic hand muscles,Proximal upper limb muscles were normal. Reflexes were absent. Vibration and proprioception sensation were absent over the toes bilaterally and Pinprick and temperature were decreased to the knees and wrists.



**1.What are the systems involved? (motor, sensory or both)**
Sensory and motor.

**2. Where is the disruption of the weakness? (proximal, distal or both. Symmetrical or asymmetrical)**
Symmetrical distal.

**3. What is nature of sensory involvement?**
Vibration and proprioception were absent + pain and temp were decreased .

**4. Is there evidence of upper motor neuron involvement?**
No.

**5. Onset (acute, subacute, chronic)**
Not mentioned by doctor or in the history.

**6. Is there any history of hereditary neuropathy?**
Yes .

-So we have symmetrical distal Sensory and motor loss. What is the pattern? Pattern 2.

What is DDx of pattern 2?

- CSPN(cryptogenic neuropathy>no apparent reason) , which is most common
- Metabolic : diabetes , thyroid .
- **The DIAGNOSIS: Hereditary .**

the patient denied family history + sensory loss . discrepancies between my examination and the patient's history + the pattern distribution direct you towards HERIDITARY neuropathy.

| | Weakness | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|
| | Proximal | Distal | Asymmetric | Symmetric | Sensory Symptoms | Severe Proprioceptive Loss | UMN Signs | Autonomic Symptoms/ Signs | Diagnosis |
| Pattern 2: distal sensory loss with/ without weakness | + | | + | + | | | | | CSPN, metabolic, drugs, hereditary |

the pt. denied any sensory loss because since he knew himself he doesn't feel anything, that's his norm !

**Case 3:** A 25 year-old woman developed numbness and tingling of the feet and hands followed by progressive leg more than arm muscle weakness over the last week. She experienced a diarrheal illness 3 weeks ago that had resolved within 10 days.

- Examination showed marked bifacial weakness and absent muscle stretch reflexes. She had normal pinprick, light touch and proprioception but vibration was reduced at the toes. Muscle power in the lower limbs was 2/5 and in the upper limbs 3/5, with equal proximal and distal weakness. She could not stand up or walk with assistance.
- Laboratory studies including vitamin B12 level and 2 hour glucose tolerance test were normal and there was no serum monoclonal protein. Forced vital capacity was 2.0 liters. • Cerebrospinal fluid evaluation showed no white cells but protein was 82 mg/dl. Nerve conduction studies showed 50% delay in tibial and median F wave latencies. Sensory conductions showed normal sural and absent median potentials

| | Weakness | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|
| | Proximal | Distal | Asymmetric | Symmetric | Sensory Symptoms | Severe Proprioceptive Loss | UMN Signs | Autonomic Symptoms/ Signs | Diagnosis |
| Pattern 1: symmetric proximal and distal weakness with sensory loss | + | + | | + | + | | | | GBS/CIDP |

**1.What are the systems involved? (motor, sensory or both)**

Sensory + motor.

**2. Where is the disruption of the weakness? (proximal, distal or both. Symmetrical or asymmetrical)**

Symmetrical proximal and distal.

**3. What is natural of sensory involvement?**

numbness and tingling of the feet and hands

**4. Is there evidence of upper motor neuron involvement?**

No.

**5. Onset (acute, subacute, chronic)**

1 week = acute

**6. Is there any history of hereditary neuropathy?**

No

So we have symmetrical proximal and distal sensory loss. What is the pattern? Pattern 1.

What is DDx of pattern 1?

- **THE DIAGNOSIS** GBS , peaks at week 4 . WHENEVER ITS ACUTE CONSIDER GBS IN YOUR DDX , AND 90% YOU WILL BE RIGHT !
- CIDP . It's simply chronic GBS that peals at week 8 !

    - The diarrhea was before the symptoms so probably there is no autonomic involvement. most probably he got an infection then developed Abs that attacked his nerves.
    - GBS patients has (Albumino-cytologic disassociation ) , which means there CSF fluid has high protein and and normal WBC count .
    - And on nerve conduction study they show demyelinating nerve roots .

**Case 4:** A 42-year-old man developed numbness and tingling in the toes, progressing up to the ankles over 2 years. He describes burning pain in his feet, mainly at night. He recently started noticing symptoms of numbness and tingling in distal fingers. He denies any weakness.

- Examination showed normal strength, with decreased pinprick and light touch sensations to the ankles and distal fingers. Vibration was absent at the toes and decreased at the ankles, and proprioception is normal at the toes. Reflexes are normal in the arms and at the knees but ankle reflexes are absent. Gait is normal.

**1.What are the systems involved? (motor, sensory or both)**

Sensory.

**2. Where is the disruption of the weakness? (proximal, distal or both. Symmetrical or asymmetrical)**

Symmetrical distal.

**3. What is natural of sensory involvement?**

numbness and tingling in the toes and burning pain in his feet.

**4. Is there evidence of upper motor neuron involvement?**

No.

**5. Onset (acute, subacute, chronic)**

2 years = **chronic** , progressive . they can't tell when it exactly started.

**6. Is there any history of hereditary neuropathy?**

No

-So we have symmetrical distal sensory loss. What is the pattern? Pattern 2.

What are the DDx of pattern 2? predominantly sensory polyneuropathy

**THE DIAGNOSIS : DM**

**Case 5:** A 36-year-old man, Eight weeks ago, he had bent down to lift a chair and developed acute pain in the right back and buttock with radiating paresthesia into the calf and lateral foot.

- Neurologic examination:

– normal muscle bulk and tone in the lower extremities.

– Straight-leg raising elicited pain and paresthesias into the right leg at 45 degrees.

– Power: weakness in right hip extension, knee flexion, and ankle plantar flexion

– Sensory examination: mild sensory loss on the right sole and lateral foot. – DTR: right ankle reflex was absent, other DTRs were normal.

**1.What are the systems involved? (motor, sensory or both)**

Sensory + motor

**2. Where is the distribution of the weakness? (proximal, distal or both. Symmetrical or asymmetrical)**

Asymmetrical proximal and distal.

**3. What is nature of sensory involvement?**

**Pain**

**4. Is there evidence of upper motor neuron involvement?**

No.

**5. Onset (acute, subacute, chronic)**

8 weeks = subacute

**6. Is there any history of hereditary neuropathy?**

No

So we have asymmetrical distal and proximal sensory loss. What is the pattern? Pattern 4.

-What are the DDx of pattern 4? Plexopathy and polyradiculopathy

**-THE DIAGNOSIS IS : S1 RADICULOPATHY , BECAUSE THE SYMOTOMS FOLLOW THE DERMATOMES + MYOTOMES OF S1 !**

**It's not the sciatic nerve, because if it was the sciatic there would be impairment in all feet movements + the sciatic doesn't supply gluteus maximus**

| | Weakness | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|
| | Proximal | Distal | Asymmetric | Symmetric | Sensory Symptoms | Severe Proprioceptive Loss | UMN Signs | Autonomic Symptoms/ Signs | Diagnosis |
| Pattern 4: asymmetric proximal and distal weakness with sensory loss | + | | + | + | + | | | | Polyradiculopathy, plexopathy |

- Check slide 11.
- STUDY L5 AND C7(IN THE NECK ) CAREFULY !
- L5 , WILL PRESENT WITH IMPAIRMENT IN :
  - -HIP ABDUCTOR
  - -ANKLE DORSI-FLEXION
  - -EVERSION .

# Summary

| Peripheral neuropathy | |
|---|---|
| **Definition** | disorders of the peripheral nervous system that are common and may affect the motor, sensory or autonomic components, either in isolation or combination |
| **Site of lesion** | Nerve root (radiculopathy) |
| | Nerve plexus (plexopathy) |
| | Nerve (neuropathy) |
| **Pathophysiology** | Damage that may occur to the nerve cell body or the myelin sheath |
| **Clinical features** | Motor: Wasting, Hypotonia, Weakness and Hyporeflexia |
| | Sensory: ↓ Vibration,↓ proprioception, ↓ Pain, ↓ Temperature |
| | Autonomic nerves: ↓ Sweating, Hypotension, Urinary retention and Impotence |
| **Classification** | 1- Neuronopathy: motor, sensory and autonomic |
| | 2- Peripheral neuropathy: myelinopathies and axonopathies |
| | Or could be classified as hereditay or acquired |

- **Approach to peripheral nervous system examination**

**1- Start with Hx [symptoms, onset, duration and progression]**

**2- Neurological examination**

- Confirm localization (LMN vs UMN, myopathy vs neuropathy)
- Recognize pattern of neuropathy (1)
- Recognize features of hereditary neuropathy
- Recognize features that narrows the differential diagnosis.
- Autonomic features

**(1) Pattern-recognition approach to neuropathy**

The whole idea of this lecture is to apply the 6-10-step clinical approach to neuropathy: 6 key questions, 10 phenotypic patterns.

| | |
|---|---|
| Questions | What systems are involved? |
| | What is the distribution of weakness? |
| | What is the nature of the sensory involvement? |
| | Is there evidence of upper motor neuron involvement? |
| | What is the temporal evolution? |
| | Is there evidence for a hereditary neuropathy? |
| Phenotypic pattern | 1-Symmetrical proximal & distal weakness with sensory loss,ex:GBS |
| | 2-Symmetrical distal sensory loss with or without weakness, ex: DM and drug induced neuropathy |
| | 3-Asymmetric distal weakness with sensory loss, ex: vasculitis and infection |
| | 4-Asymmetric proximal & distal weakness with sensory loss, ex: Plexopathy |
| | 5-Asymmetric distal weakness without sensory loss, ex: ALS and MMN |
| | 6-Symmetric sensory loss & upper motor neuron signs with proprioceptive loss,ex: B12 deficiency |
| | 7-Symmetric weakness proximal & distal without sensory loss, ex: SMA |
| | 8-Focal midline proximal symmetric weakness with upper motor neurons, ex: ALS |
| | 9-Asymmetric proprioceptive loss without weakness, ex: Ganglionopathy |
| | 10-Autonomic dysfunction seen in both DM & GBS |

# Questions

**1. A 55 years old female presented with sensory loss and incoordination in both upper and lower limbs for 5 months. Her neurological examination showed normal muscle power and absent reflexes. She had sensory loss to pinprick, vibration and position in both upper and lower limbs. Which one of the following localization describe pattern is associated with?**

A. Anterior horn cell.
B. Diffuse Peripheral Nerves.
C. Dorsal root ganglia.
D. Neuromuscular Junction.

**2. A 18 years old male presented with weakness and numbness for 5 years. On examination he had high arched feet. Reflexes was absent. Sensory examination showed abnormal sensation to pinprick and vibration. Muscle power was 2/5 distally, 4/5 proximally in lower limbs. And 3/5 distally, 5/5 proximally in upper limbs. Which one of the following is the most appropriate description for his neuropathy?**

A. Diabetic Neuropathy.
B. Inherited Neuropathy.
C. Toxic Neuropathy.
D. Vitamin B12 Deficiency.

**3. 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.

**4. A 49 years old male came to your clinic complaining of weakness in his right arm and hand with loss of sensation for the past 3 weeks. Which one of the following can cause his symptoms?**

A. Long standing DM.
B. Guillain–Barré syndrome (GBS).
C. Trauma.
D. Amyotrophic lateral sclerosis (ALS).

**5. Which one of the following will be affected incase of small fiber damage?**

A. Loss of proprioception.
B. Loss of sensation of temperature.
C. Loss of sensation of vibration.
D. A and C.

**6. What is the most common cause for peripheral neuropathy?**

A. DM.
B. GBS.
C. CMT.
D. ALS.

**7. Which one of the following will be associated with ascending weakness and loss of reflexes over a short period of time?**

A. Diabetic neuropathy.

B. ALS.

C. GBS.

D. CMT.

**8. Which one of the following can cause asymmetrical distal weakness with sensory loss?**

A. Charcot-Marie-Tooth (CMT).

B. Multifocal motor neuropathy (MMN).

C. Hereditary Neuropathy with Liability to Pressure Palsy (HNPP).

D. B12 deficiency.

**9. Which one of the following can affect the autonomic nervous system?**

A. GBS.

B. CMT.

C. DM.

D. A and C

**10. A 55-year old female presented with ascending weakness and sensory loss that started 2 weeks ago after having upper respiratory tract infection. Her neurological examination showed weakness in upper and lower limbs that was symmetric and graded as 3/5. Reflexes were diminished. She had sensory loss to pinprick, vibration and joint position in both upper and lower limbs. Which ONE of the following localizations is consistent with this pattern?**

a) Anterior horn cell

b) Diffuse peripheral nerves and nerve roots

c) Dorsal root ganglia

d) Neuromuscular junction

**11. Patient presented with 2 months of Right leg pain. He also has back pain. On physical examination he had muscle power of 4/5 in dorsiflexion, eversion and inversion. And had 5/5 in ankle flexion, knee flexion and extension, hip flexion and extension, and hip adduction. And had 4/5 in hip abduction. He also showed sensory deficits with pinprick test but had normal vibration test. What is the most likely diagnosis?**

a) Common peroneal injury

b) L5 radiculopathy

c) Femoral nerve injury

d) Popliteal injury

Answers:

1-C 2-B 3-B 4-C 5-B 6-A 7-C 8-C 9-D 10-B 11-B