ACUTE PERIPHERAL NEUROLOGIC LESIONS

DIVERSE GROUP OF DISORDERS → INVOLVE INJURY OR DISEASE IN SENSORY AND MOTOR FIBRES OUTSIDE OF THE C.N.S. EXTENDING TO THE NEUROMUSCULAR JUNCTION

MAY HAVE SYMPTOMS REFLECTING A DISORDER OF ANY OR A COMBINATION OF SENSATION, MOTOR OR AUTONOMIC FUNCTION

SENSORY SYMPTOMS → NUMBNESS, TINGLING, PAIN, ATAXIA, DYSESTHESIA

MOTOR SYMPTOMS → WEAKNESS

AUTONOMIC DISABILITY → ORTHOSTASIS, BOWEL/BLADDER DYSFUNCTION, GASTROPARESIS, SEXUAL DYSFUNCTION

APPROACH → DISTINGUISHING CENTRAL AND PERIPHERAL LESIONS:

- CNS and PNS neuroanatomy provides the best guide to distinguishing these lesions
    - Peripheral nerves contain a mix of motor and sensory fibres that follow well defined paths that render them prone to injury → hence peripheral nerve lesions are more likely to be confined to one limb and to present with involvement of multiple sensory and motor modalities
- Hyporeflexia more characteristic of PNS lesion, but can occur early in CNS disease, invariably followed by spasticity
- Most CNS lesions will lead to UMN signs → Babinski, hyperreflexia, hypertonia

Table 166-1 Differentiating Central from Peripheral Nervous System Disorders

	Central	Peripheral
History	Cognitive changes	Weakness confined to one limb
	Sudden weakness	Weakness with pain associated
	Nausea, vomiting	Posture- or movement-dependent pain
	Headache	Weakness after prolonged period in one position
Physical examination		
Reflexes	Brisk reflexes (hyperreflexia)	Hypoactive reflexes
	Babinski sign	Areflexia
	Hoffman sign	
Motor	Asymmetric weakness of ipsilateral upper and lower extremity	Symmetric proximal weakness
	Facial droop	
	Slurred speech	
Sensory	Asymmetric sensory loss in ipsilateral upper and lower extremity	Reproduction of symptoms with movement (compressive neuropathy)
		All sensory modalities involved
Coordination	Discoordination without weakness	Loss of proprioception

LOCALISING PERIPHERAL NEUROLOGIC DISORDERS:

- Precise diagnosis can be difficult

- The history required to define the process involves → symmetry, proximal vs distal symptoms, sensory, motor or autonomic involvement, mono vs polyneuropathy
- Look for muscle wasting, fasciculations, hyporeflexia
- Further studies → EMG, nerve conduction studies
- LP and CSF analysis are frequently required

TREATMENT → GENERAL CONSIDERATIONS:

- MANAGEMENT DEPENDS ON THE SPECIFIC DIAGNOSIS
- In patients with potential for respiratory failure, aspiration and cardiac dysrhythmia should be monitored appropriately
    - Consider FVC to assess need for respiratory support (although this is a clinical decision more often than not)

NEUROMUSCULAR JUNCTION DISORDERS:

BOTULISM:

- Toxin-mediated illness than can cause acute weakness leading to respiratory failure, offending organism is CLOSTRIDIUM BOTULINUM
- Now very rare, although isolated events do occur with improperly preserved canned foods, wound contamination of heroin users
- Botulinum toxin works by binding irreversibly to presynaptic membrane of peripheral and cranial nerves inhibiting the release of acetylcholine atht e peripheral nerve synapse → with time, new receptors are generated and patients improve
    - In classic botulism, there is NO SENSORY DEFICIT AND NO PAIN, occurring 6-48 hours post ingestion of tainted food → preceded by N+V, abdominal cramp and diarrhoea (i.e. like viral gastro)
    - Classically produces a DESCENDING, SYMMETRIC PARALYSIS → first affects cranial nerves and bulbar muscles (diplopia, dysarthria, dysphagia)
    - Anticholinergic signs → constipation, urinary retention, dry skin, hyperthermia
    - Pupils often dilated (differentiates from Myasthenia gravis)
- Diagnosis is clinical → ANTITOXIN CAN SHORTEN DURATION OF SYMPTOMS
- IV IG decreases duration of ventilation

TICK PARALYSIS:

- Uncommon, but multiple tick species implicated
- Begins as ataxia (2-6 days post attachment) → progressive lower then upper extremity weakness
- Mortality of untreated tick paralysis ~10%
- Treatment is supportive and complete removal of the attached tick → local wound care and supportive care

ACUTE PERIPHERAL NEUROPATHIES:

GUILLAIN-BARRE SYNDROME:

- An acute polyneuropathy characterised by immune-mediated peripheral nerve myelin sheath destruction
- Exact cause unknown, but many associations:
    - Viral or febrile illness
    - Campylobacter jejuni infection
- Monophasic illness, at its worst at 2-4 weeks, and recovery can take up to a year
- SUBACTUE ASCENDING SYMMETRIC WEAKNESS OR PARALYSIS AND LOSS OF DEEP TENDON REFLEXES THAT FOLLOWS A VIRAL ILLNESS
    - Paralysis may ascend to the diaphragm
    - Specific diagnostic criteria listed below:

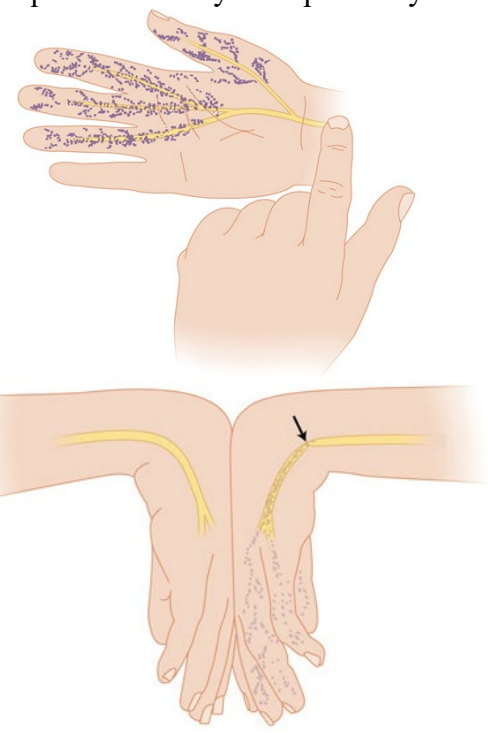
Table 166-2 Diagnostic Criteria for Classic Guillain-Barré Syndrome
Required
Progressive weakness of more than one limb
Areflexia
Suggestive
Progression over days to weeks
Recovery beginning 2–4 wk after cessation of progression
Relative symmetry of symptoms
Mild sensory signs and symptoms
Cranial nerve involvement (Bell's palsy, dysphagia, dysarthria, ophthalmoplegia)
Autonomic dysfunction (tachycardia, bradycardia, dysrhythmias, wide variations in blood pressure, postural hypotension, urinary retention, constipation, facial flushing, anhydrosis, hypersalivation)
Absence of fever at onset
Cytoalbuminologic dissociation of cerebrospinal fluid (high protein and low white cell count)
Typical findings on electromyogram and nerve conduction studies

- Diagnosis is MOSTLY HISTORICAL
- Miller-Fisher variant associated with C. jejuni and is charactgerised by ophthalmoplegia, ataxia and decreased or absent reflexes with less severe weakness (has abrupt commencement, progresses rapidly and has prolonged course with poor prognosis)
- LUMBAR PUNCTURE → high protein, low WCC (predominant mononuclear cells)
- First step in management is ASSESSMENT OF RESPIRATORY FUNCTION → airway protection in advance of respiratory compromise decreases the incidence of aspiration and other complications
- IV IG and plasmapheresis are used to treat GBS → provide equivalent but not additive benefit in terms of reduction of duration of symptoms
    - Corticosteroids are of no benefit and may be harmful

Table 166-3 Managing Respiratory Failure in Guillain-Barré Syndrome
Indications for intubation
Vital capacity <15 mL/kg
PaO_2 <70 mm Hg on room air
Bulbar dysfunction (difficulty with breathing, swallowing, or speech)
Aspiration
Indications for admission to intensive care unit
Autonomic dysfunction (**Table 166-2**)
Bulbar dysfunction
Initial vital capacity <20 mL/kg
Initial negative inspiratory force <−30 cm of water
Decrease of >30% of vital capacity or negative inspiratory force
Inability to ambulate
Treatment with plasmapheresis

FOCAL NEUROPATHIES:

- Mononeuropathies are MOST LIKELY DUE TO FOCAL COMPRESSION → diabetes can cause noncompressive mononeuropathy
- MEDIAN MONONEUROPATHY (CARPAL TUNNEL SYNDROME):
  - Classic signs → pain, paraesthesias and numbness in distribution of the median nerve
  - Tinel sign and Phalen manoeuvre are used to assist diagnosis but have poor sensitivity and specificity



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Tinel sign

Phalen sign

- Initial treatment is conservative → splinting, behavioural modification, weight loss, NSAIDS. Consider steroid injection and surgical release if no response to above

ULNAR MONONEUROPATHY:

- Cubital tunnel syndrome → compression behind the medial epicondyle. Tingling in fifth and lateral fourth fingers and eventuall intrinsic muscle weakness
- Can also be compressed at GUYON'S CANAL → handlebar compression
- Treatment in ED consists of adequate analgesia and arranging follow up

ENTRAPMENT OF DEEP PERONEAL NERVE:

- TRAUMA, RAPID WEIGHT LOSS OR HABITUAL CROSSING OF THE LEGS → patients develop foot drop or numbness of the web between the great and second toes
- Entrapment can occur at fibular head, anterior to ankle joint (under extensor retinaculum)

MERALGIA PARAESTHETICA:

- Entrapment of lateral femoral cutaneous nerve within the inguinal canal → numbness and pain of the anterolateral thigh
    - Conservative management with NSAIDs, weight loss and physio are usually successful

MONONEURITIS MULTIPLEX:

- A group of disorders that have in common the dysfunction of multiple peripheral nerves separated both temporally and in anatomic location (diabetes is the most common causes)
- Treatment is management of underlying disorder

PLEXOPATHIES:

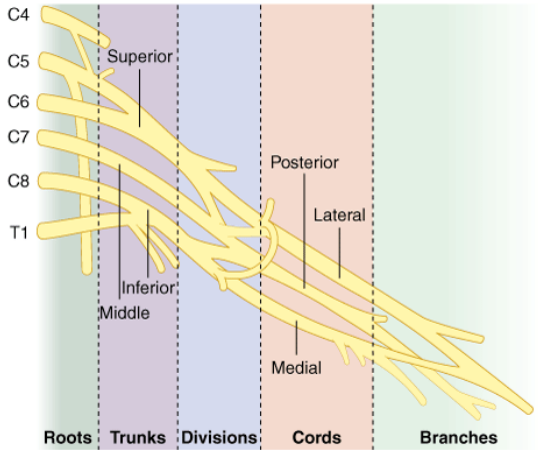
CERVICAL PLEXOPATHY:

- Concern for a neoplastic process → causes deep/boring pain that is constant → prompt imaging with CT/PET

BRACHIAL PLEXOPATHY:

- Most common site of plexopathy
- Anatomy outlined below → finally forms five nerves
- Upper trunk is most common site of involvement, affecting strength of proximal arm and shoulder musculature
    - Infraclavicular plexopathy due to trauma is frequently associated with injury to the axillary vessels
- Causes of brachial plexopathy → trauma (penetrating, humeral neck fracture or dislocation), shoulder relocation, neoplasm (Pancoast tumour) radiation or surgery
- BURNER SYNDROME → sudden axial impact of shoulder → short-lived burning and anaesthesia of affected limb that resolves spontaneously

- Most managed conservatively but those due to cervical rib, clavicle fracture and penetrating trauma require exploration to exclude associated injuries



LUMBOSACRAL PLEXOPATHY:

- More likely due to radiation, diabetic amyotrophy, aortic aneurysm or retroperitoneal haemorrhage than due to trauma
- Result in weakness of hip adduction and flexion and knee extension, decreased sensation at top/inner thigh

HIV-ASSOCIATED PERIPHERAL NEUROLOGIC DISEASE:

- Primary infection, complications and medications of HIV can all affect peripheral nervous system
- HIV infected patients have a high rate of mononeuritis multiplex and an inflammatory myopathy resembling polymyositis
- Treatment is conservative
- CMV radiculitis → CMV retinitis is almost always present → become acutely weak with primarily lower extremity involvement and may have variable degrees of bowel and bladder dysfunction
- Antiviral treatment with ganciclovir is effective (5mg/kg q12h)

DIABETIC PERIPHERAL NEUROPATHY:

- Half of patients with diabetes have symptoms of neuropathy and 15% require treatment
- The most common manifestation of diabetic peripheral neuropathy is a symmetric distal polyneuropathy, but focal neuropathies and mononeuritis multiplex can occur
- The most common cause of nontraumatic amputation is injury resulting from impaired sensation due to diabetic peripheral neuropathy that fails to heal because of the impaired blood flow
- There is a 60% reduction in the risk of developing neuropathy with tight glycaemic control
- Tricyclic antidepressants, anticonvulsants used to manage pain associated with neuropathy