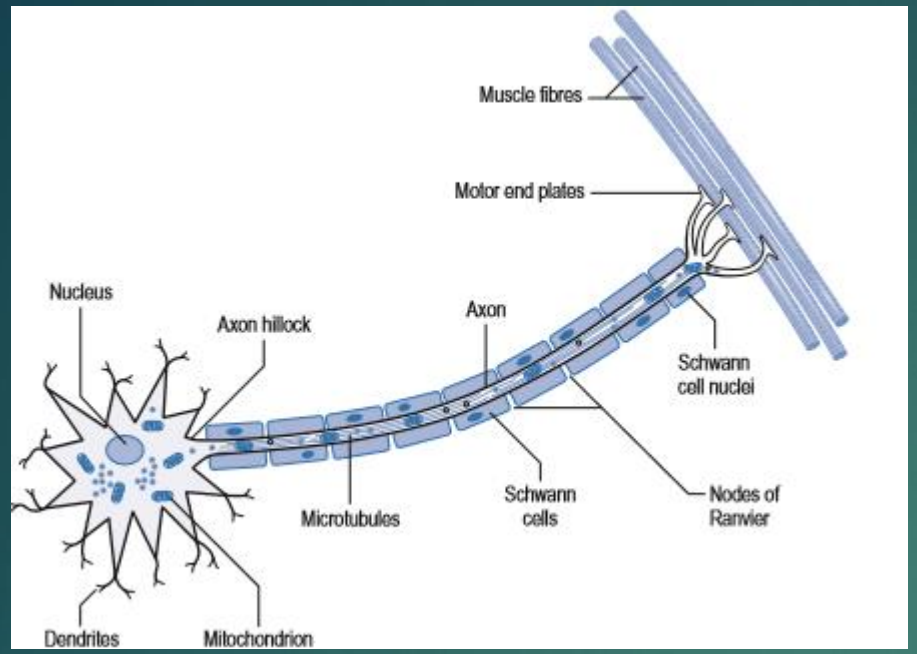
Polyneuropathies

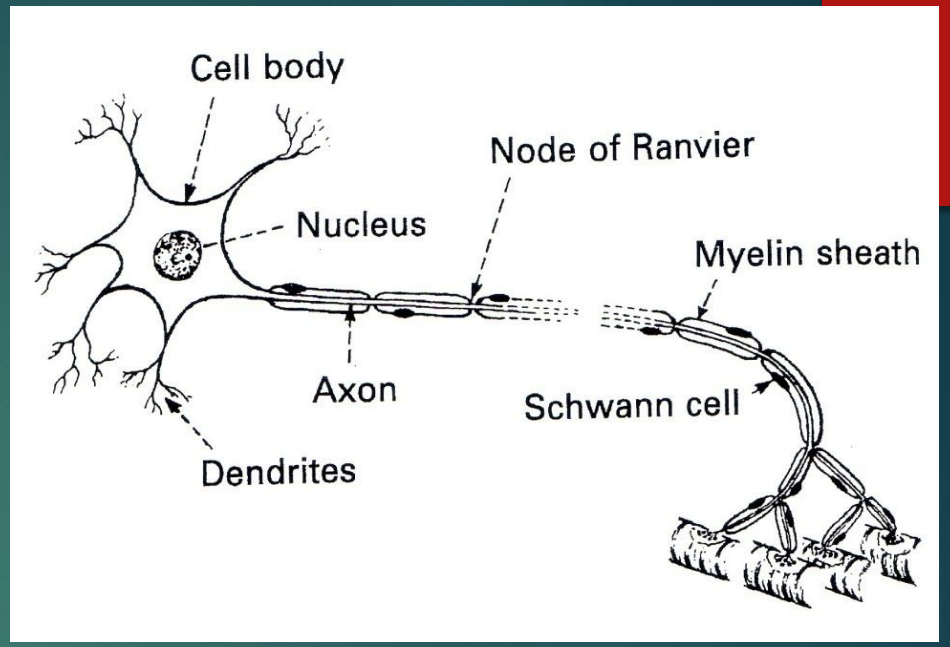
Daw Nyein Myat Noe
Demonstrator
Department of Physiotherapy
University of Medical Technology, Yangon

Learning Objectives

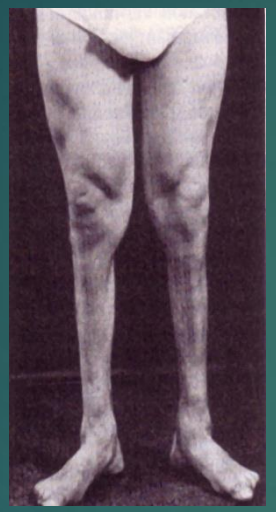
- Define polyneuropathies.
- Explain classification, pathology, aetiology and investigations of polyneuropathies.
- Describe most common acute and chronic polyneuropathies and their related management.
- Describe the role of physiotherapy in polyneuropathies.

Contents



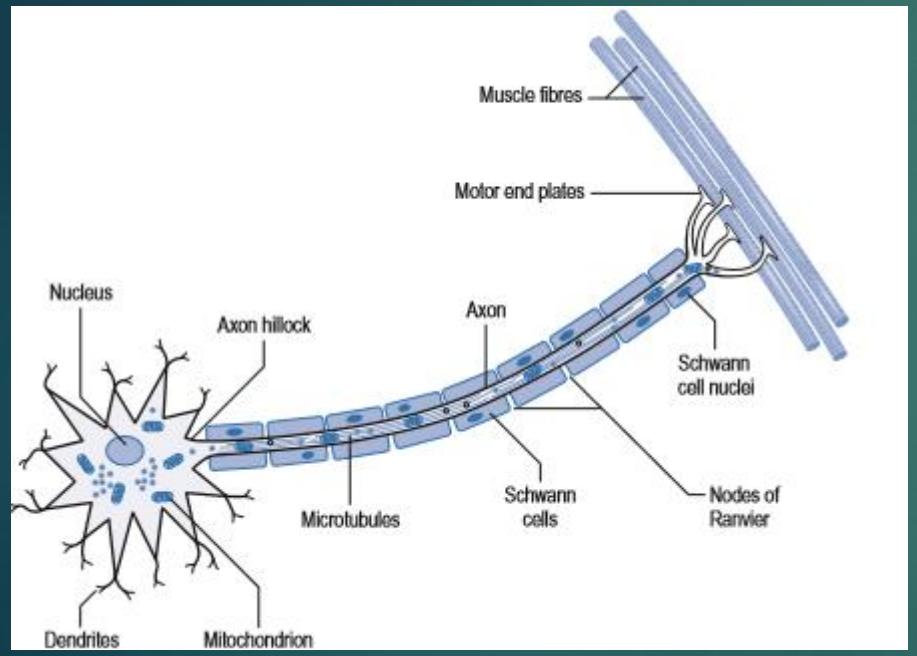


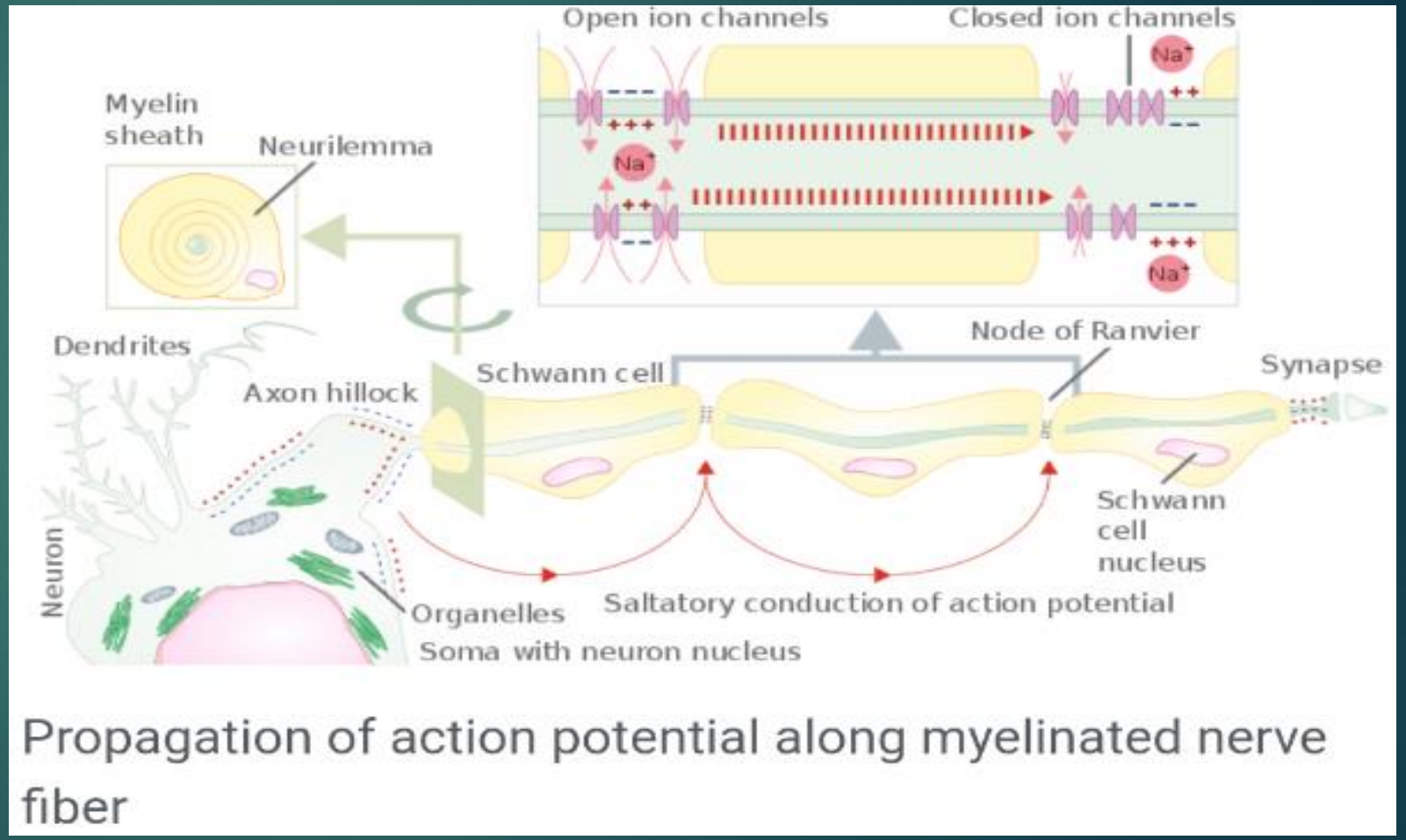
- Polyneuropathies are generalized disorders of peripheral nerves, affecting both motor and sensory neurons
- Impairment of transmission of nerve action potentials d/t disruption of axon or myelin sheath



- Begins in the hands and feet, progress to the arms and legs and other parts of the body where it may affect the autonomic nervous system
- Acute or chronic
- Usually seen in the young or middle-aged adult; men being affected more than women

Nerve Physiology

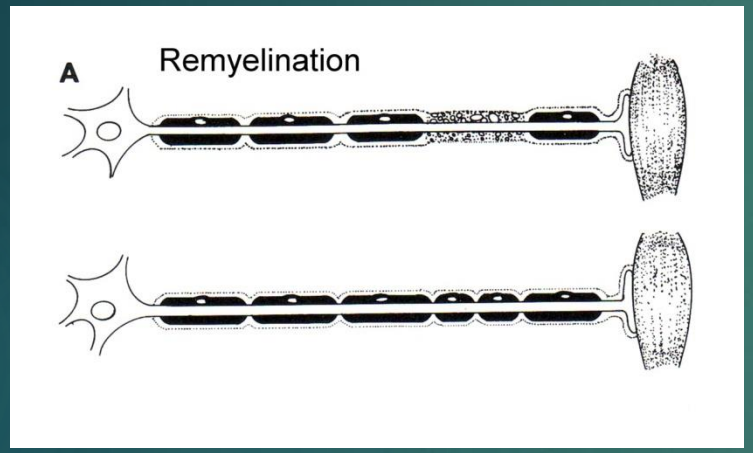




Propagation of action potential along myelinated nerve fiber

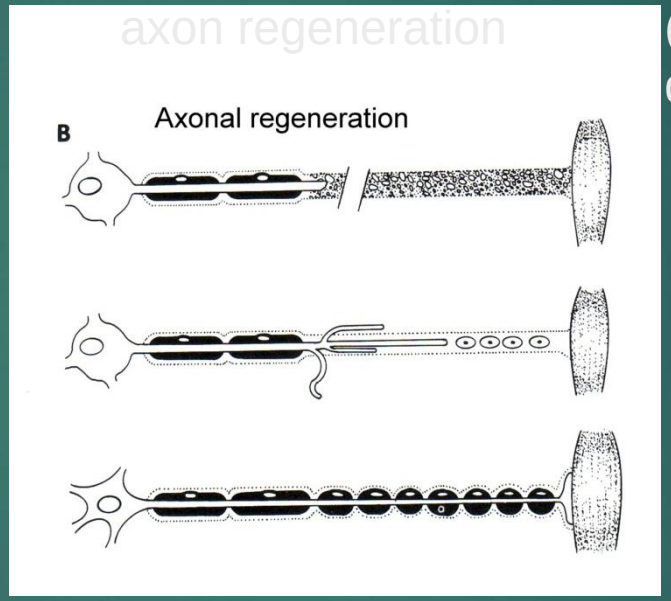
Nerve regeneration – reinnervation

Remyelination



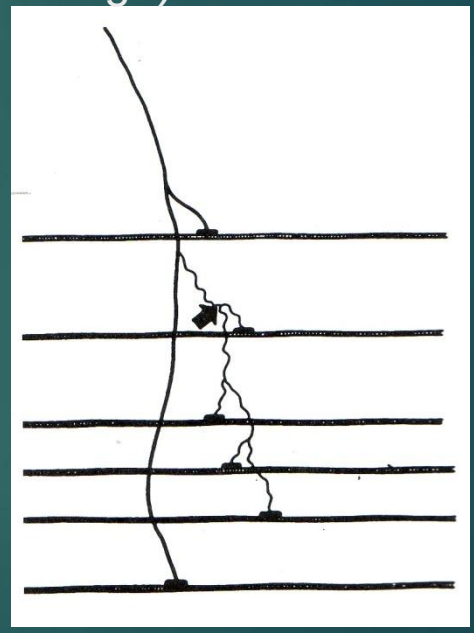
2-12 weeks

Proximo-distal axon regeneration



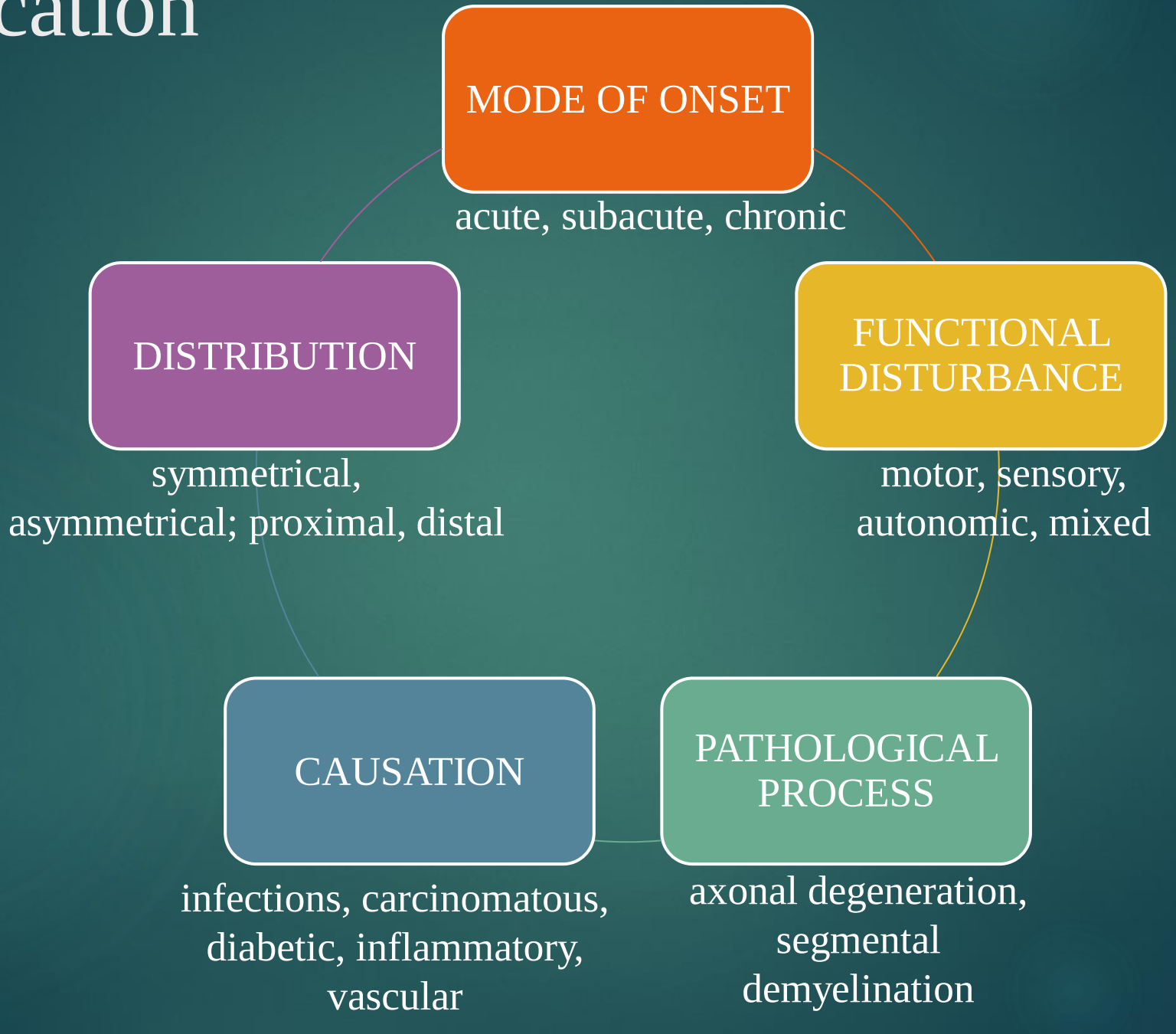
1 mm/day
Intact basal lamina/endoneurium is needed

Collateral reinnervation (in case of partial nerve damage)

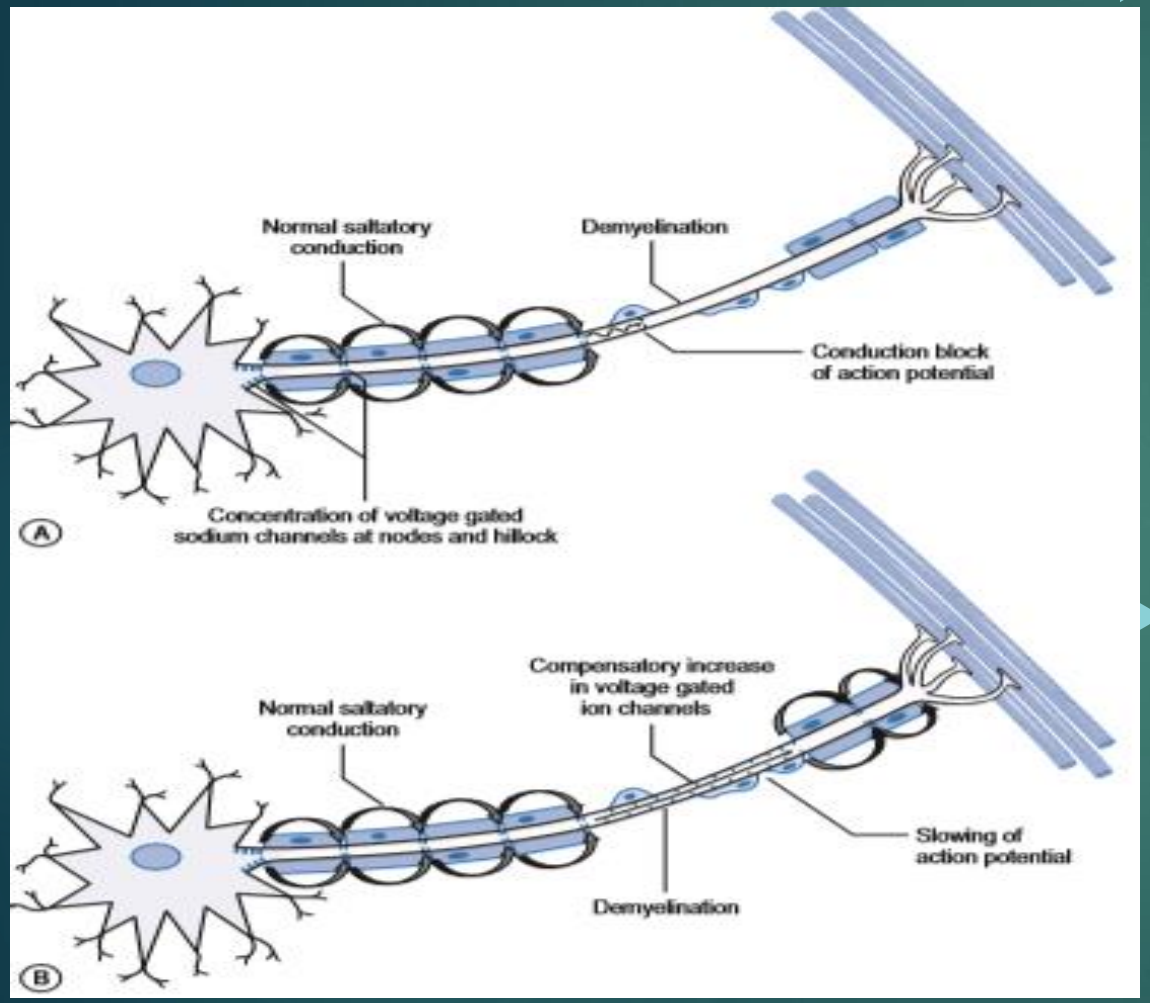


Starts within 4-6 weeks

Classification



Pathology



- Axonal degeneration
  - Nerve cell body & axon are affected
  - Similar to Wallerian degeneration
  - Recovery - slow and incomplete
  - Occurs in neuropathies of causes: poisons, nutritional deficiencies, ischaemia
- Segmental demyelination
  - Affects Schwann cell, result in demyelination
  - Recovery - rapid and complete
  - Occur with a diabetic neuropathy & Guillain-Barre syndrome

Aetiology

Infective condition

- local infections of peripheral nerves: e.g., virus – herpes zoster, bacteria -leprosy, brucellosis
- current infection, e.g. dysentery, influenza, mumps
- infections with organisms whose toxins - affinity for PNS, e.g. diphtheria, tetanus, botulism

Post-infective polyneuropathy

- e.g. Guillain-Barre Syndrome

Toxic substances

- e.g., heavy metals – mercury, lead, arsenic, gold and copper organic chemicals – aniline, cyanide, triortho-cresyl-phosphate
- drugs – isoniazid, thalidomide, and nitrofurantoin, vin-cristine

Aetiology

Deficiency, metabolic, blood disorders	• e.g., alcoholism, porphyria, leukaemia, DM, chronic uraemia, liver failure & vitamin deficiencies
Trauma	• e.g., physical (compression/stretching), electrical (earth shock) or radiation injury to nerves
Connective tissue disease	• e.g. RA, polyarteritis nodosa, SLE, amyloid disease, sarcoidosis & carcinoma

Aetiology

Genetic disorders

- e.g., hereditary sensory radicular neuropathy, hypertrophic interstitial neuritis, peroneal muscular atrophy, Refsum’s disease

Pure vascular disorders

- e.g., atheroma, collagen disorders, diabetes mellitus, Burger’s disease

Unknown origin

General Investigations

- Urine – glucose, protein
- Haematology – FBC, ESR, vitamin B12, folate
- Biochemistry – fasting glucose, RFT, LFT, TSH
- Neurophysiology testing – nerve conduction studies, needle electromyography
- Nerve biopsy

Most common acute and chronic polyneuropathies

Demyelinating Polyneuropathies

- Occur due to degeneration/destruction of myelin
- Prolonged periods of demyelination can lead to axonal degeneration

<u>Acute demyelinating polyneuropathy</u>

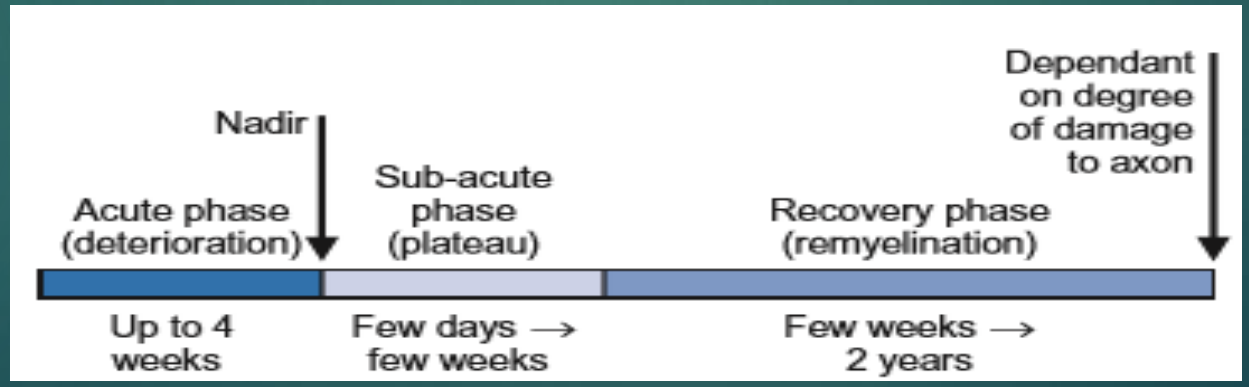
- Highest point of severity/nadir reaches in < 4 weeks
- Guillain Barre´ syndrome (GBS), affect any age group, more prevalent in men and in older people
- Rapidly progressive paralysis, sensory impairment and areflexia

Guillain Barre´ syndrome

- GBS is an autoimmune disorder where an immune response is directed towards unknown antigens triggered by the earlier infection
- This immune response leads to an inflammatory process and destruction of the myelin sheath in the larger diameter motor and sensory neurons

Presentation of Guillain Barre´ syndrome

- Acute inflammatory demyelination - disruption of saltatory conduction leading to a slowing or block of nerve conduction
- Proximal/distal weakness & sensory loss → ascends or descends from onset
- In severe cases - trunk weakness & bulbar dysfunction, respiratory function and vital capacity
- Autonomic involvement - fluctuations in BP & cardiac arrhythmias



Phases and progression of Guillain Barre ´ syndrome

Management

Initial management

- To address any serious complications, e.g. reduced VC and airway protection

Treatment

- Intravenous immunoglobulins (IVIg)
- Plasma exchange or plasmapheresis

Physiotherapy

- Respiratory interventions - sputum clearance techniques, maintenance of lung volumes, breathing exercises
- Prevention of secondary complications
- Early mobilization, splinting, positioning, stretches to maintain ROM & exercises to increase strength & endurance

Chronic demyelinating polyneuropathy

- Acquired or inherited
- Taking > 8 weeks to develop
- Most common causes - inflammatory and genetic pathologies

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP)

- Immunologically mediated neuropathy
- Multifocal demyelination, predominantly affects spinal nerve roots, proximal nerve trunks & plexuses leading to patchy regions of demyelination with some inflammatory infiltrates
- Primarily a motor neuropathy - affect distal & proximal m/s, distal ankle m/s - weakest
- Sensory impairment present with sensory ataxia
- Tendon reflexes (-), facial palsy, fatigue

Management

- Treatment with corticosteroids, intravenous immunoglobulins and plasma exchange are necessary over a prolonged period

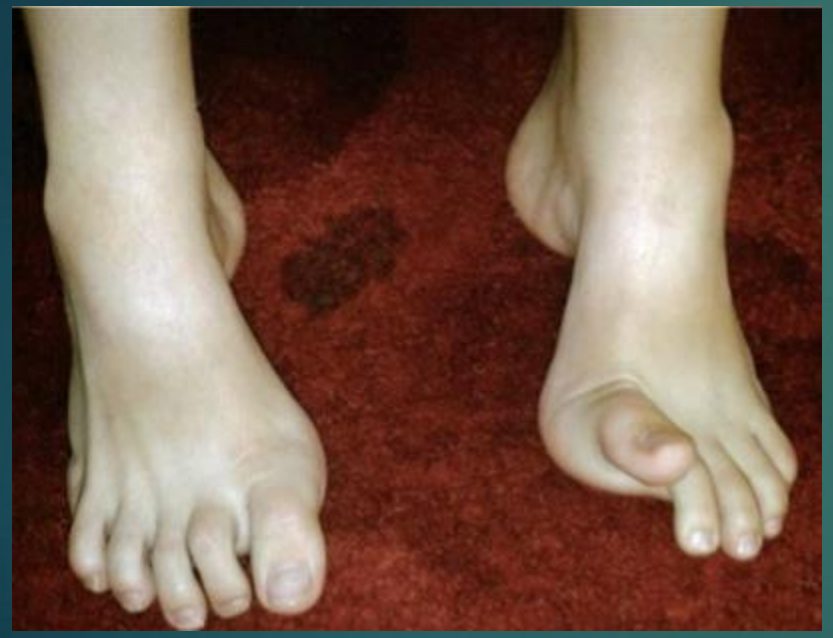
Charcot-Marie-Tooth disease type 1 (CMT1)

- Hereditary neurological condition
- Slow decline in distal m/s strength and sensation that predominantly affects longer peripheral nerves
- Type 1 CMT (CMT1) presents with demyelination of more thickly myelinated, fast-conducting axons, e.g. alpha motor neurons and 1a afferent sensory neurons

Presentation

- Muscle wasting with ‘inverted wine bottle’ appearance of distal lower limb and ‘claw hand’ of upper limb - weaken first & slow decline in strength over decades
- Degree & extent of weakness is correlated with axonal loss than demyelination
- Proximal limb m/s are less affected
- Principal impairment of thickly myelinated large diameter sensory nerves (light touch and vibration)
- Sensations by smaller diameter fibres (pain, temperature or pin prick) may be reduced

Presentation

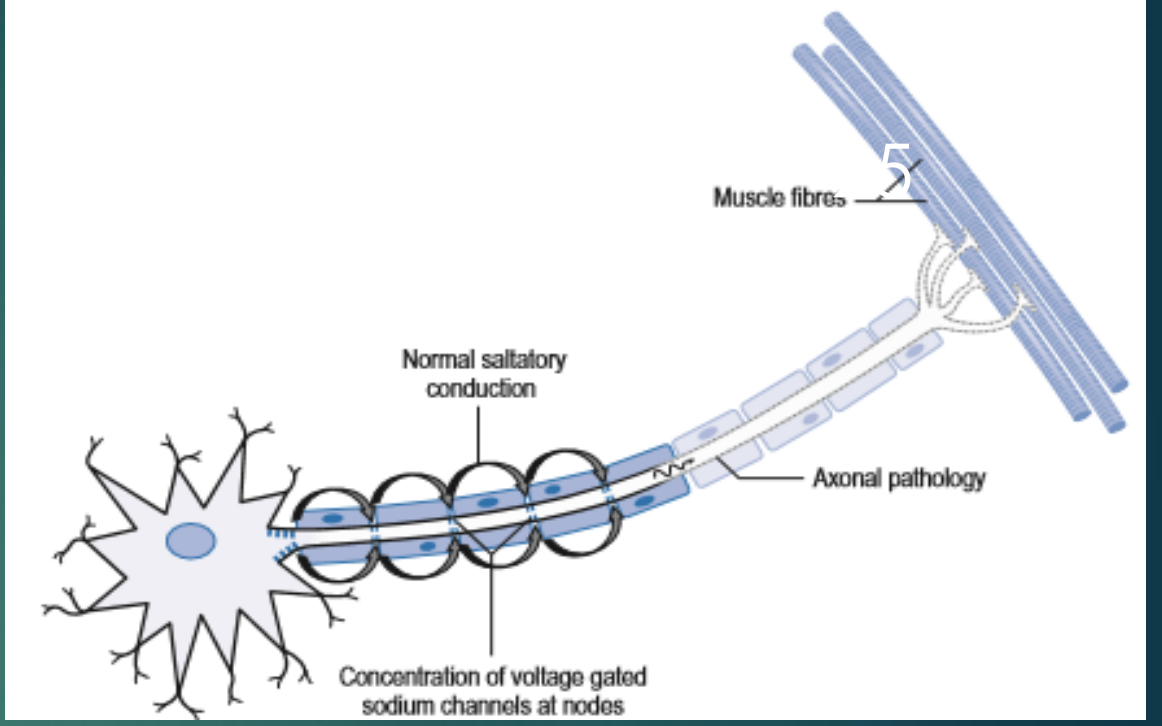


- Pes cavus , hind foot varus, toe clawing and dorsiflexion of MTPJ
- Foot drop and failure of plantarflexors influence the pattern of gait
- Aerobic deconditioning and disuse muscle atrophy impact on fatigue and prolonged performance of daily tasks.

Management

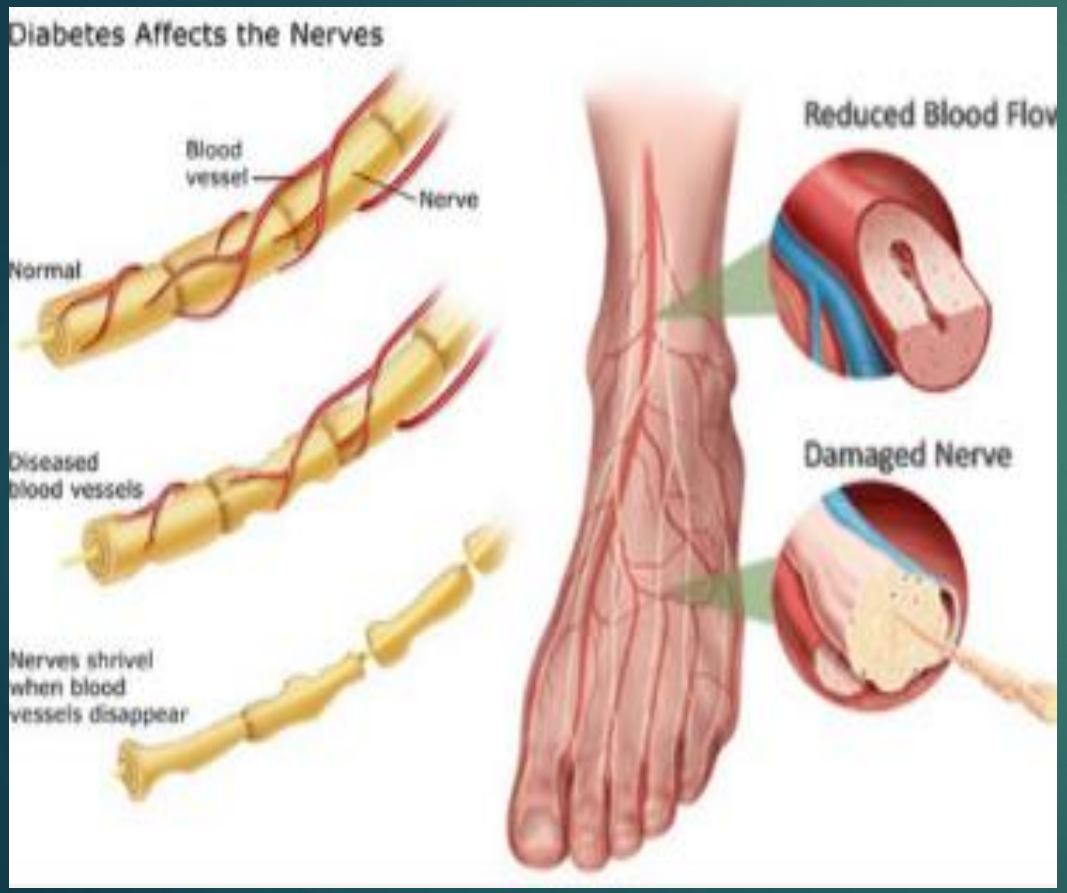
- There is no drug therapy for people with CMT1

Axonal Polyneuropathies



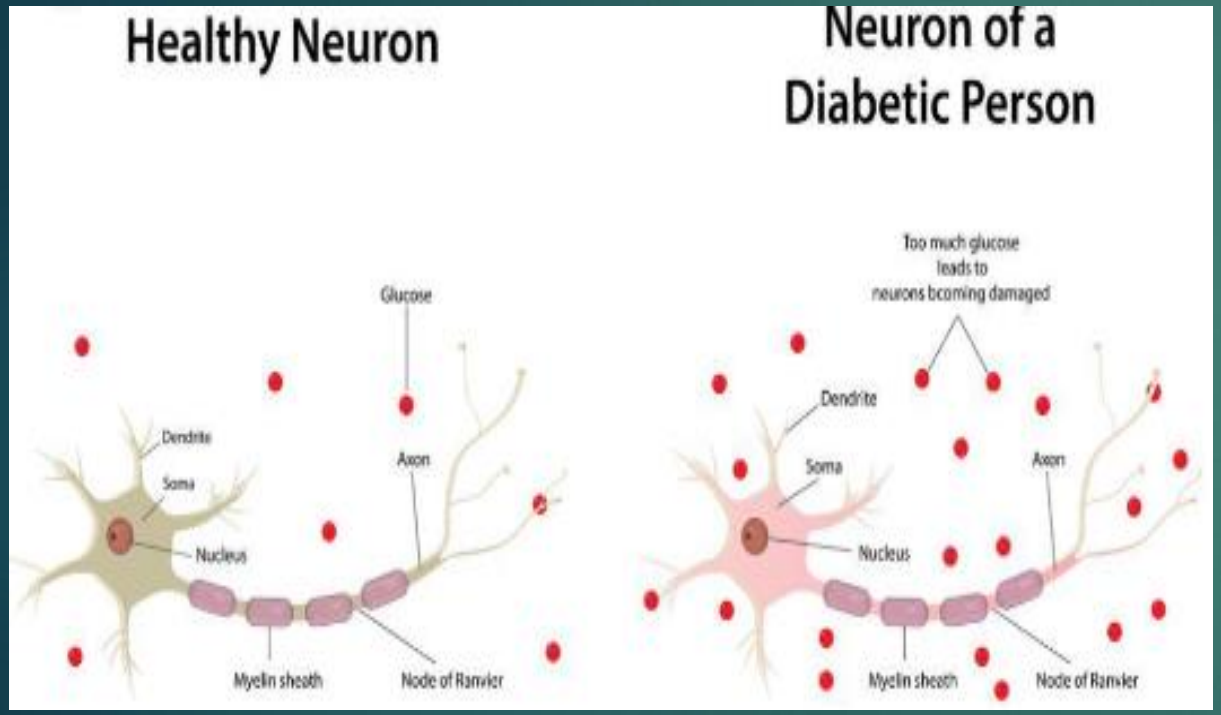
- Chronic axonal neuropathies are the most common type
- Causes vary from metabolic disorders, such as chronic renal failure and malignancy, to toxicity from chemical agents
- Characterized by abnormality and degeneration of nerve axons, so can affect nerves of any diameter and modality
- Diabetic neuropathy and Charcot-Marie-Tooth disease type 2 (CMT2)

Diabetic neuropathy



- Just sensory or sensorimotor in presentation
- Burning, numbness in feet d/t involvement of smaller diameter, unmyelinated fibres from cutaneous pain receptors
- Skin biopsies → reduced density of unmyelinated fibres in epidermis (degeneration)

Diabetic neuropathy



Pathological process

- Initiated through hyperglycaemia which has a toxic effect on nerves via oxidative stress, impaired axonal transport and accumulation of end products from glycation
- Effect on microvascular structures supporting the nerves with defects in the capillary endothelia

Presentation

- Diagnosis is from neurological examination & glucose tolerance testing
- Progression is slow and prognosis depends on diabetic management
- Gait pattern changes and increased double support → Increase plantar pressures → Risk of injury to skin and ulceration, so protective footwear is recommended
- Consequence of sensory loss is postural instability and falls due to reduced proprioception and cutaneous sensation

Management

- The standard therapy for IGT and diabetes is diet and lifestyle advice

Charcot-Marie-Tooth disease type 2

- Degeneration of nerve axon, less common than type 1; the most common mutation is of mitofusin 2 gene, a mitochondrial protein - important role in the process of mitochondrial fusion within a cell
- Long axon has high energy requirements far away from nerve cell body which is provided by the mitochondria
- Deficiency of mitofusin 2 affects the transport of mitochondria down the axon, result in degeneration of distal axon that is initially seen in longer nerves such as those supplying the foot and ankle muscles

Presentation

- Similar phenotype as CMT1
- Slow progression of distal weakness, wasting and sensory loss
- Onset of symptoms may be later in life so the pes cavus foot type is not always present

Management

- No specific medical interventions or under investigation

PHYSIOTHERAPY INTERVENTIONS

Exercise

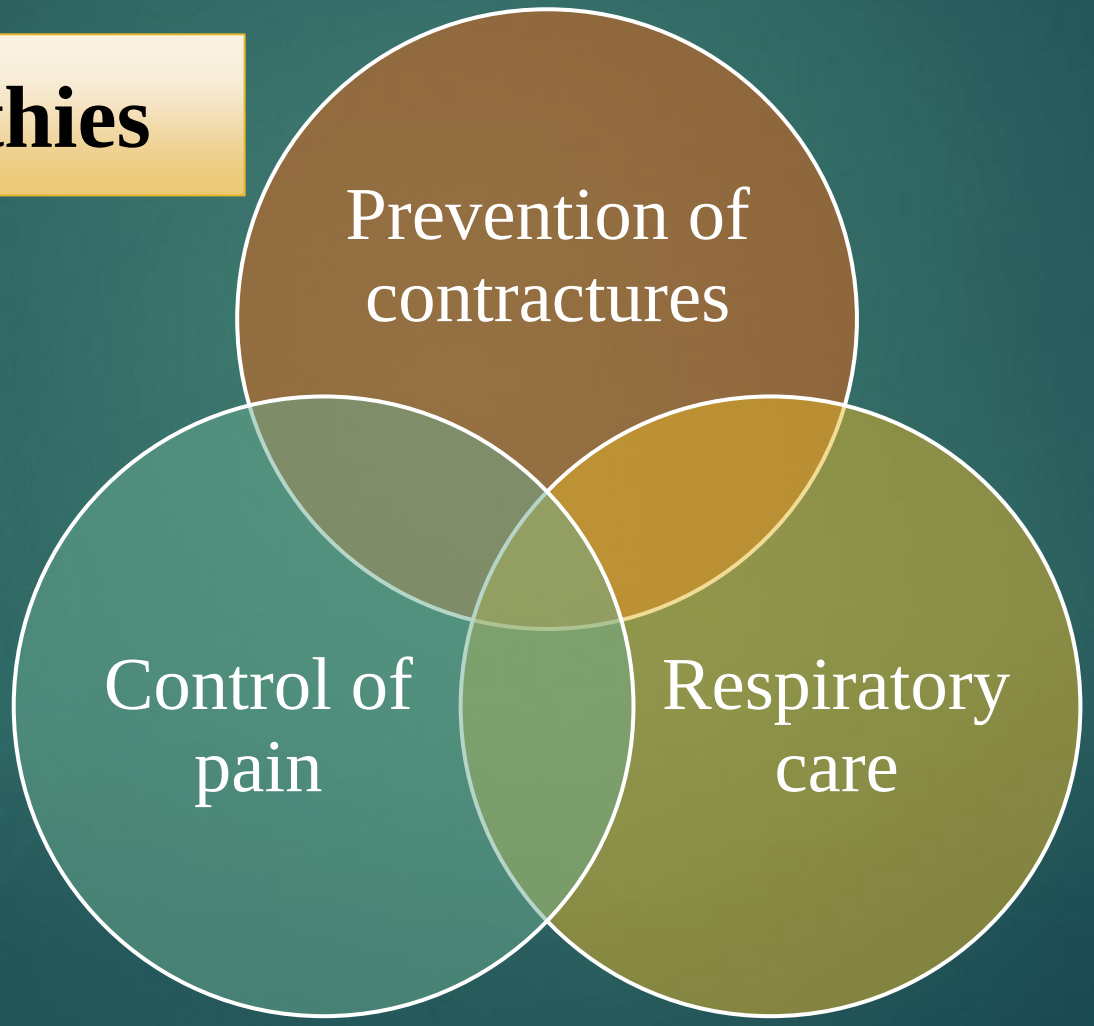
- improve aerobic fitness and cardiopulmonary function

Orthotic management

- For foot deformity to distribute pressure under plantar surface or correct flexible pes cavus using lateral posting
- To reduce plantar pressures foot orthoses, custom-made shoes or casts are used to redistribute pressure, and to provide soft padding and shock absorbance through total plantar contact
- Total contact casts reduce postural stability

Principles of physiotherapy intervention

Acute neuropathies



Stretching to prevent contractures

Muscles
(not stretched adequately, shortened position)

Structural changes
(loss of sarcomeres)

- All structures, including nervous system - moved through full range
- Encouraged to join in with movement and use, be taught self stretches in a weight-bearing position
- In bed-bound pt - foot drop splints & gentle stretches

Positioning

- Frequent changes - prevent selective muscle shortening and pressure sores

Pain

- Patients with acute GBS enjoy large-amplitude mid-range movements because of their pain-relieving properties
- In other neuropathies, massage and ice packs are helpful

Respiratory care

- Patient – ventilated, facial muscle weakness, ANS is affected, disturbed blood pressure, especially when using suction or when attempting early sitting

Chronic neuropathies

- Role of the physiotherapist is largely one of management in chronic cases
- Early referrals are important to advise on activities to maintain ambulation and prevent avoidable complications such as foot deformities

Strengthening exercises

- Rate of deterioration may be reduced or recovery hastened without affecting the underlying disorder
- Where muscles are severely weakened, strengthening is slim and exercise could cause further damage to the motor unit

Stretches

- Gentle stretches for muscle groups that are liable to shorten
- If real shortening occurred, attempts to stretch cause damage to other related structures

Pain relief

- Malalignment of joints due to muscle imbalance leads to pain
- Ice, massage and vibration diminish painful chronic sensory neuropathies
- Transcutaneous electrical nerve stimulation

Functional and mobility aids

- Orthoses and wheelchairs

References

- Thomson, et al., (1991). Tidy's Physiotherapy. 12th eds.
- Stokes, M. and Stack, E. (2011). Physical Management for Neurological Conditions. 3rd eds. China: Elsevier Limited.
- Cash's textbook of neurology for physiotherapists. 4th eds.
- http://www. Polyneuropathies. Physiopedia.com.

Thank You