BHARATHIDASAN UNIVERSITY

Program: M.Sc., Biomedical Science

Course Title : Neurobiology

Peripheral Nerve Demyelinating Diseases

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Peripheral Nerve Demyelinating Diseases

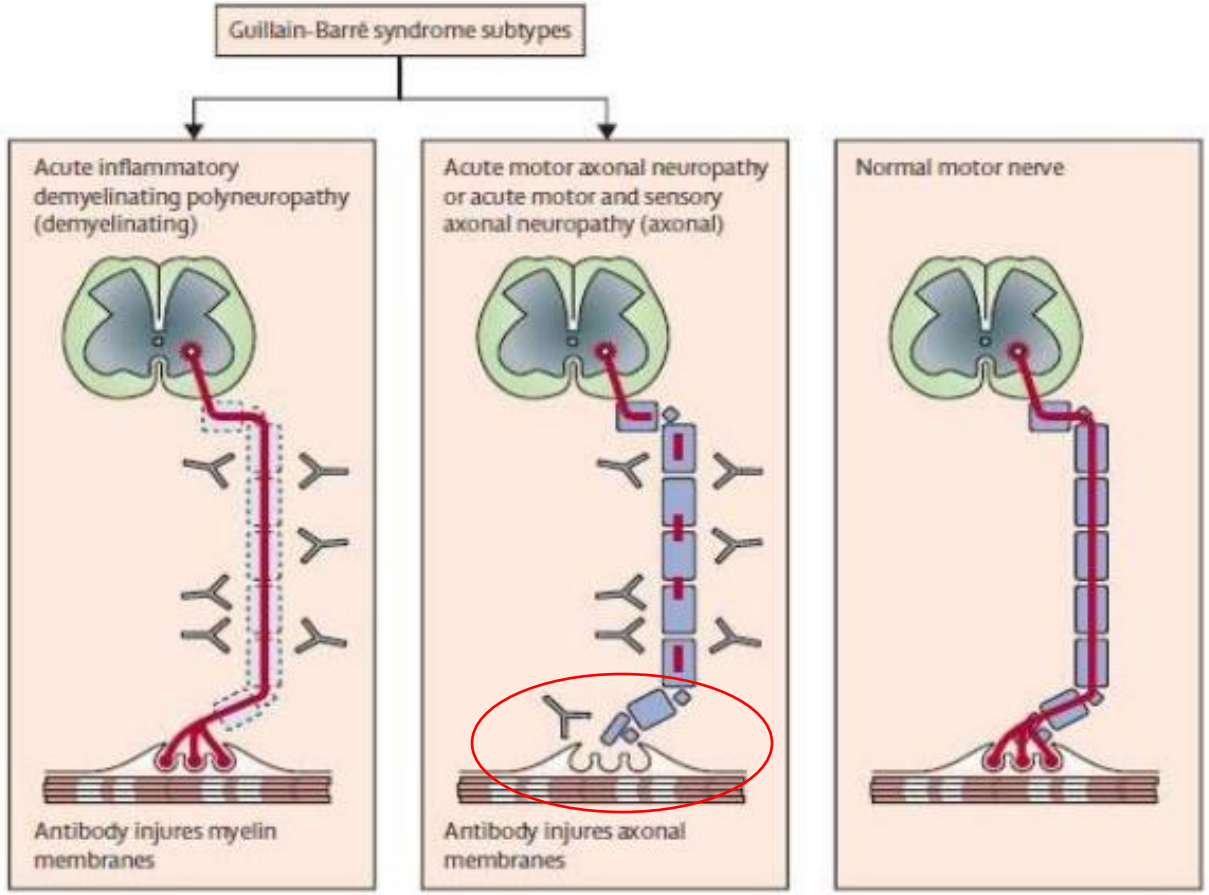
- The injury or a genetic disease of Schwann cells can result in the lack of support of peripheral neurons and/or a loss of efficient neuronal conduction.

1. Acute inflammatory demyelinating polyradiculopathy (AIDC)
2. Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP)
3. Charcot-Marie-Tooth
4. Krabbe disease
5. Metachromatic leukodystrophy

Acute inflammatory demyelinating polyradiculopathy (AIDC)

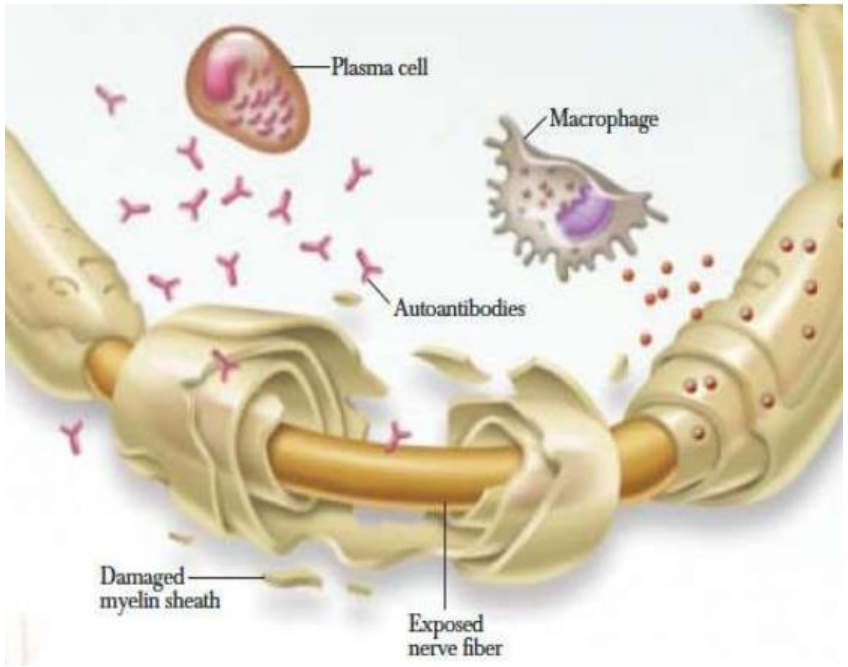
- autoimmune disorder in which the immune system attacks the peripheral nerves, often damaging the Schwann cells
- Autoimmune : immune system recognizes the myelin epitope as “foreign” and targets it for destruction.
- results in weakness, numbness, pain, and autonomic dysfunction, like respiratory failure, HTN, hypotension, Tachycardia, Bradycardia, gastric hypomotility and urinary retention.
- most common AIDC is Guillain-Barre syndrome (GBS).
- Infection of Mycoplasma pneumoniae, Epstein-Barr, cytomegalorvirus, Influenza A, Haemophilus Influenzae, Campylobacter jejuni, & Zika virus

Schwann cells



Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP)

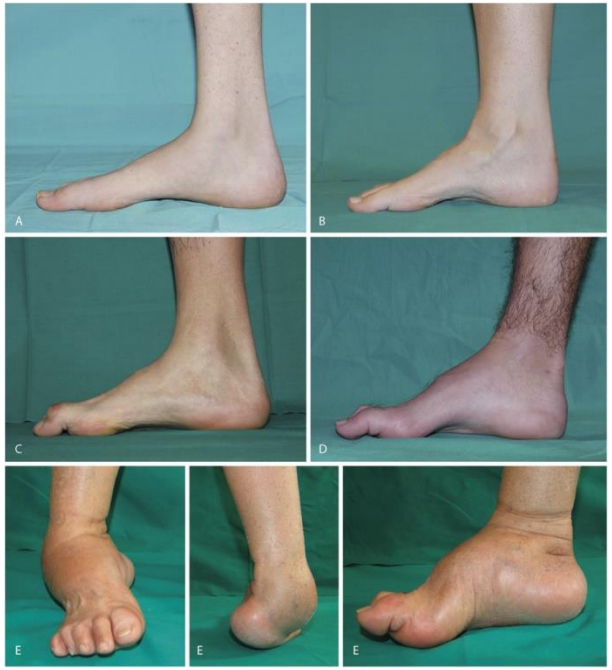
- as the name implies, is a chronic immune attack on the peripheral myelin
- Acquired demyelinating motor and sensory neuron
- All aged ppl get affected, but most ppt get affected at their 50-60 years.
- Both proximal and distal muscles are affected.
- Symptoms: numbing, tingling, pain, progressive muscle weakness, loss of deep tendon reflex, fatigue, & abnormal sensations

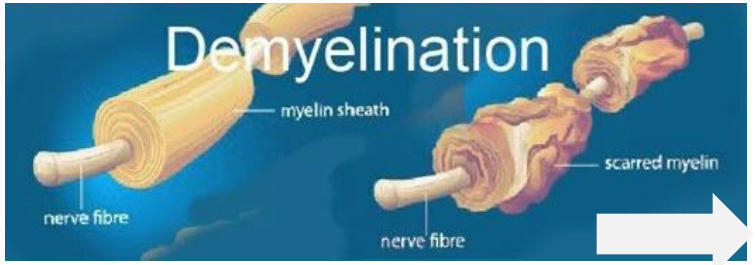


Charcot-Marie-Tooth

- hereditary demyelinating peripheral neuropathy that affects both sensory and motor neurons that control muscles

- due to the mutations in genes that produce number of proteins involved in the structure function of peripheral neuron axon or in myelin sheath.



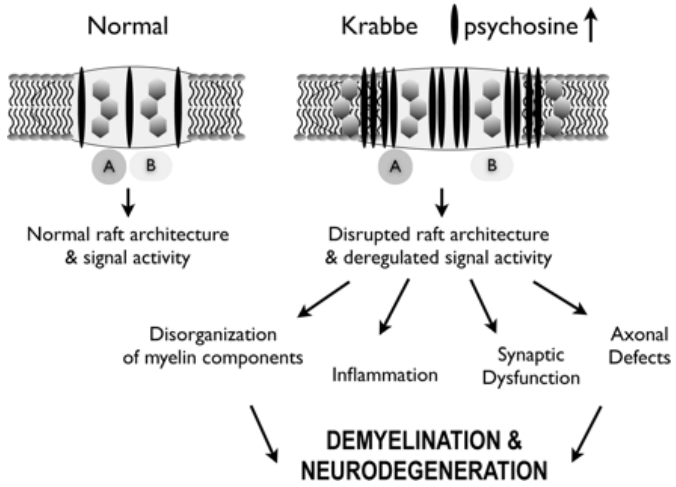


Krabbe disease

- hereditary lysosomal storage disease

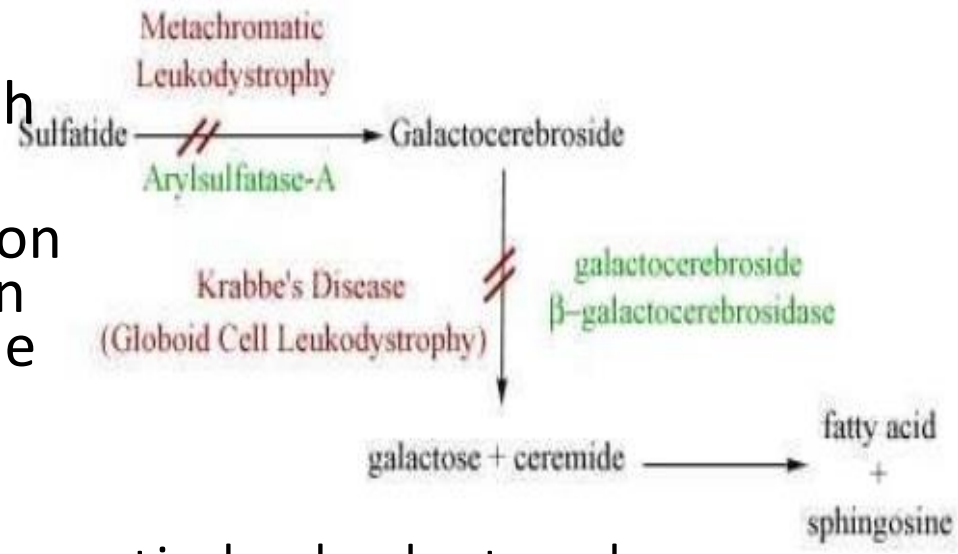
(i.e., deficient in the enzyme galactocerebrosidase or galactosylceramidase)

- Galactosylceramidase is a lysosomal enzyme
- Have no Galactosylceramidase activity
- No GALC leads to increase psychosine which in turn activate secretory Phospholipase A2
- PLA2 breakdowns lysophosphtidycholine and arachidonic acid.
- PLA2 results in death of oligodendrocytes which are responsible for myelin formation.
- in which dysfunctional metabolism of sphingolipids resulting in the destruction of proper myelin that can occur in the PNS and CNS.



Metachromatic leukodystrophy

- Another hereditary lysosomal storage disease that affects both the PNS and CNS
- in which there is an accumulation of sulfatides that destroy myelin (i.e., patients are deficient in the enzyme arylsulfatase A).



Krabbe disease and Metachromatic leukodystrophy

- have no known cure;
- bone marrow transplant can be an optional therapy.