

NEUROPATHIES, MYOPATHIES

- **NEUROPATHIES (7)**
 - Inflammatory
 - Infectious
 - Hereditary (HMSN-I)
 - HMSN-II, HMSN-III
 - Acquired (Toxic/Metabolic)
 - Traumatic
 - Neoplasms

- **MYOPATHIES (9)**
 - Denervation
 - Dystrophies
 - Ion Channel
 - Congenital
 - Genetic Metabolic
 - Inflammatory
 - Toxic
 - NeuroMuscular Junction
 - Neoplasms

GENERAL Reactions

- **NERVE**
 - **DEMYELINATION (segmental)**
 - **AXONAL DEGENERATION**
 - **NERVE REGENERATION**
 - **REINNERVATION**
- **MUSCLE FIBER**
 - **NECROSIS**
 - **VACUOLIZATION**
 - **REGENERATION**

- **ATROPHY**
- **HYPERTROPHY**

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- NEUROPATHY, Inflammatory**

- **Guillain-Barré**
 - Preceded by “influenza”-like illness
 - NO actual specific etiologic agent isolated, autoimmune disease to myelin gangliosides most likely
 - Inflammation of a peripheral nerve
 - **DEMYELINATION**
 - “ASCENDING” paralysis

NEUROPATHY, Infectious

- **Leprosy**
- **Diphtheria**
- **V/Z (Varicella-Zoster)**

**NEUROPATHY, Hereditary
(defective myelination)
(Hereditary Motor and Sensory Neuropathy)**

- **HMSN-I (Charcot-Marie-Tooth)**
- **HMSN-II (Like CMT of the neurons)**
- **HMSN-III (Palpable Nerves) (aka, Dejerine-Sottas)**

NEUROPATHY, Toxic/Metabolic

Symmetric, Asymmetric
Sensory, Sensorimotor
Somatic, Autonomic
Focal, Multifocal

NEUROPATHY, Toxic/Metabolic

Diabetes Mellitus
Vitamin Deficiencies (many Bs, E)
Heavy Metals, Pb, As, etc.
Organic Compounds
CHEMO

NEUROPATHY, Traumatic

- **Laceration regeneration rate = 1mm/day or 1 in/mo.**
- **Avulsion**
- **Carpal Tunnel**
- **Traumatic (amputation) “Neuroma”**

- **“Saturday Night” Palsy (radial n.)**
- **Morton “Neuroma”**

NEUROPATHY, Neoplastic

Benign: Schwannoma

Malignant: Malignant Schwannoma

QUIZ:

- **Why are Schwannomas the ONLY tumors of peripheral nerve?**

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 - **Denervation (SMA)**

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MYOPATHY, Denervation

SPINAL MUSCULAR ATROPHY

- Childhood diseases
- Chromosome #5 that harbors the survival motor neuron gene (*SMN1*)
- Anterior Horn Cells
- Often PAN-fascicular
- Shoulder, hip muscles

MYOPATHY, “Dystrophic”

- Jerry’s kids, no “DYSTROPHIN”

- **DUCHENNE (DMD), x-linked**
- **BECKER (BMD) (less common, less severe, same chromosome)**
- **Many others also, all of which have complex genetic patterns which have all been precisely defined**
- **MYOTONIA is a common feature**

MYOPATHY, Ion Channel “Channelopathies”

- **MYOTONIA/HYPOTONIC PARALYSIS**
- **FAMILIAL, (genetic) DISEASES**
- **TRIGGERED BY:**
 - Exercise
 - Cold
 - Carb Intake
- **Classified by K+, \uparrow K+, \downarrow K+**
- **MALIGNANT HYPERTHERMIA can be triggered off by anesthetic halogenated inhalation agents in some of these patients!!!**

MYOPATHY, Congenital

“Floppy Babies”

- **HYPOTONIC**
- **FAMILIAL, (genetic) DISEASES**
- **MANY TYPES, in most of which the precise genetic defects have been identified**
MYOPATHY, Metabolic
(genetic also)

- **LIPID**
 - **Mitoch. Enz. Def. → LIPID ACCUMULATION**

- **MITOCHONDRIAL**
 - **“PARKING LOT” mitochondria**

MYOPATHY, Inflammatory

- **DERMATOMYOSITIS**
- **POLYMYOSITIS**
- **INCLUSION BODY MYOSITIS**
- **ALL HAVE UNCLEAR ETIOLOGIES**

MYOPATHY, Toxic

- **THYROTOXICOSIS**
- **ETHANOL**
- **DRUGS (steroids, chloroquine)**
- **DRUGS (MANY MANY others)**
MYOPATHY,
NeuroMuscular Junction
- **Myasthenia Gravis**
 - **Associated with thymomas**
 - **Thymectomies often useful Rx:**
 - **AUTOIMMUNE DISEASE, CLEARLY**
 - **Ab's to ANTI-CHOLINESTERASE RECEPTORS**
 - **Anticholinesterase test is very diagnostic (edrophonium)**
 - **YOUNG WOMEN WITH EYE MUSCLE:**
 - **Ptosis →**
 - **Diplopia→**
 - **General Weakness**
- **Lambert-Eaton Syndrome (paraneoplastic), 60% have malignancies, auto-antibodies against NMJx**
MYOPATHY, Neoplastic
- **Benign**

—Rhabdomyoma



Malignant

—Rhabdomyosarcoma