Acute Neuro-myopathies

5. Acute neuro-myopathies *Wolfgang Grisold (Austria) - WFN*

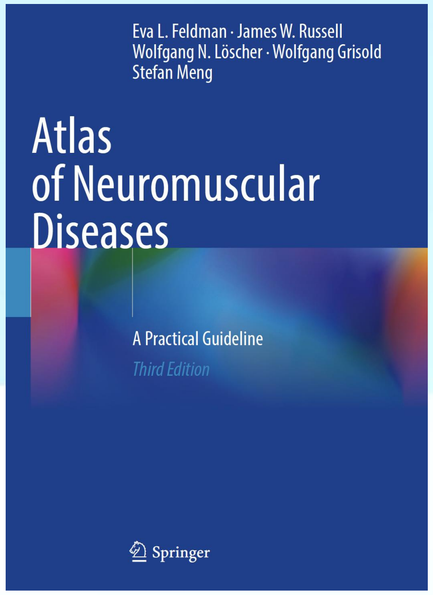
Anna Grisold, Dr. Dr., Stefan Meng Doz. Dr. (Vienna Austria)

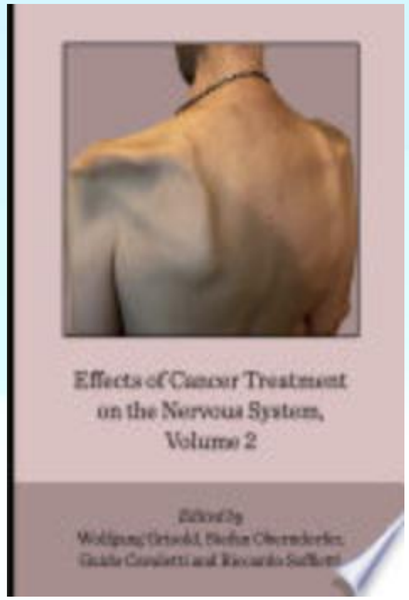
No COI

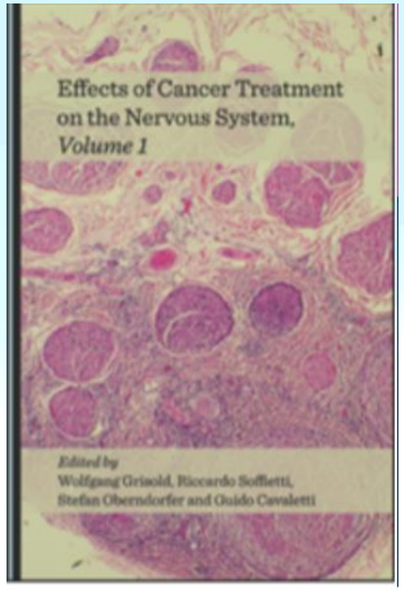
Authors contributed equally in their parts

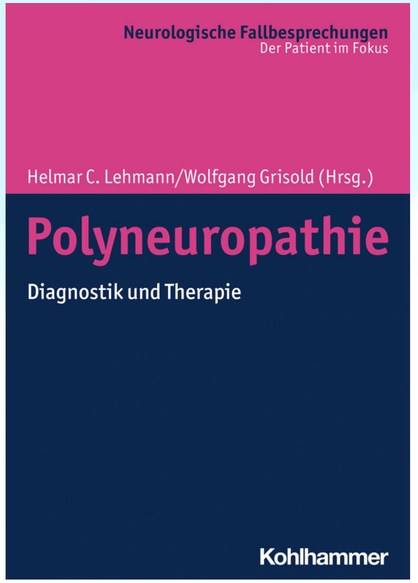
Images and slides were used from own

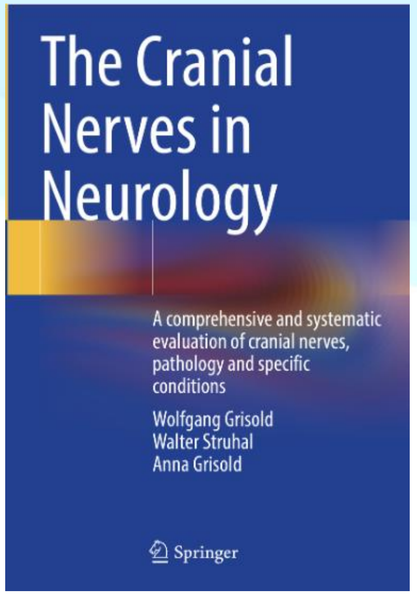
Neuromuscular Publications











3 rd edition

One Russian

translation

In Press

Content

- **Tools**
- **Clinical investigation, Status,**
- **Minimal biomarkers: Blood, Electrolytes CK, CSF**
- **NCV (later EMG)**

- Internal Medicine
- Imaging
- Lab- extended
- (Genetics)
- (Biopsy)
- Microbiology

- **Second line:**

Time -perspective

Acute

Acute, Sub Acute ?

Sensory ?

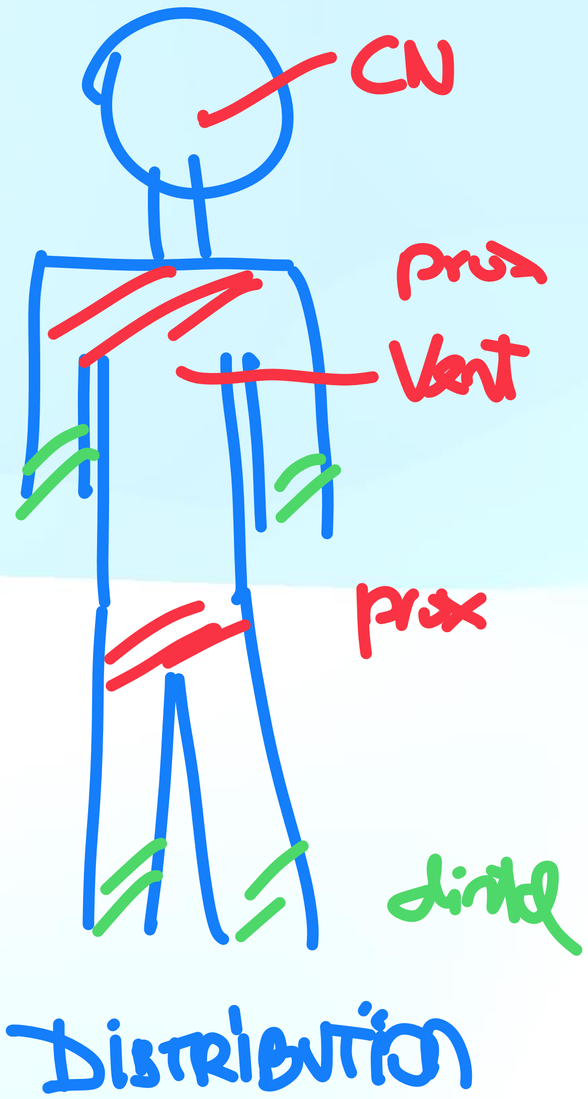
Symmetric /asymmetric

Additional: CNS, CN, Autonomic

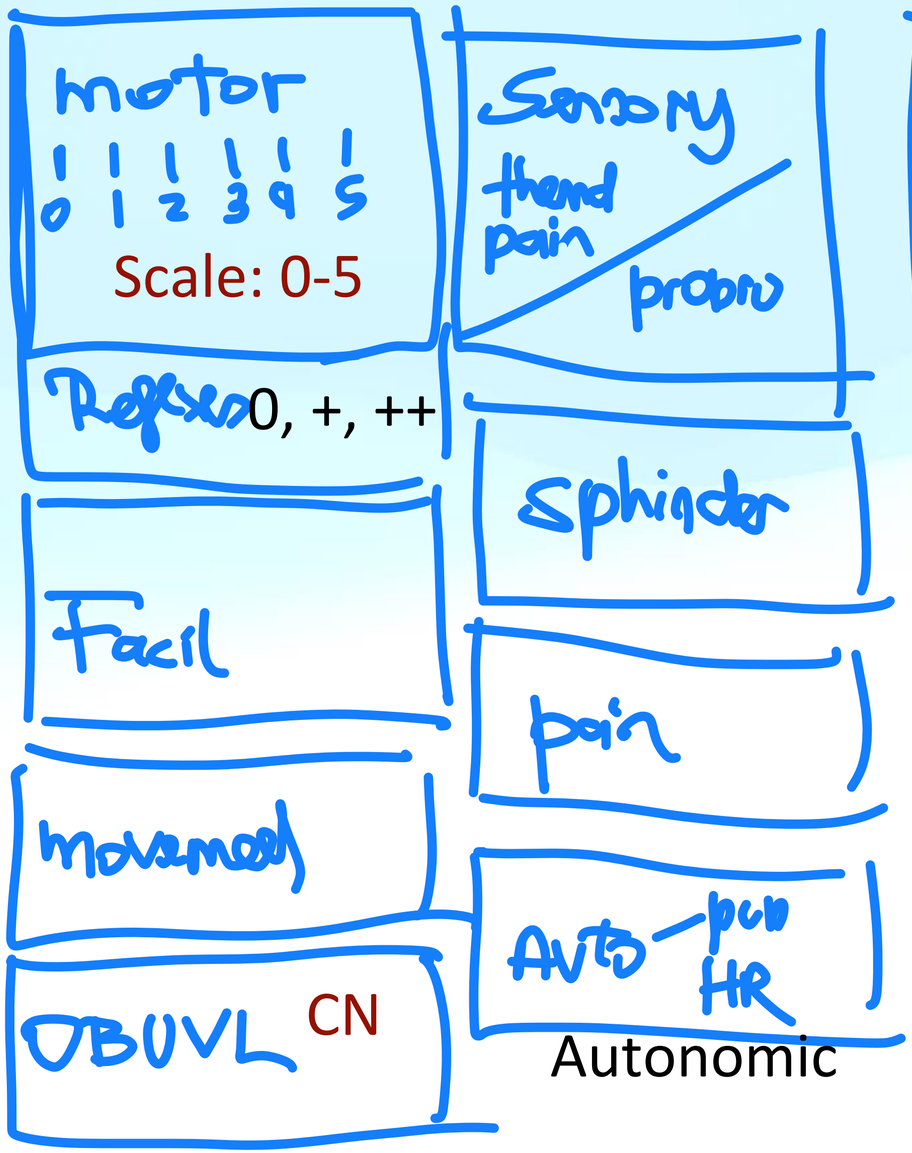
Examination and questions:

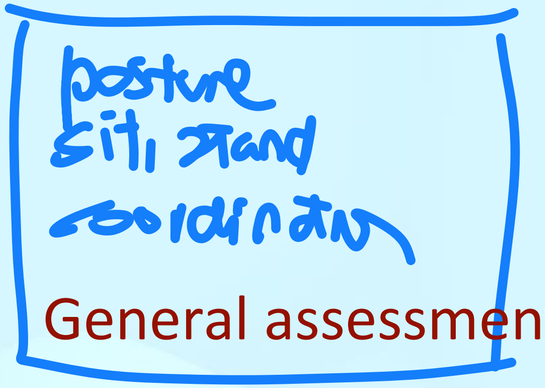
- **Motor acute paresis**
  - Distribution
- **Sensory** involvement
  - Sensory level
- **Reflexes** (motor/sensory = NCV)
- Respiration
- Sphincter control
- Cranial nerves
- Time acute, subacute progressive , waxing and waning.
- Pain, Sphincter function, Autonomic
- Movements, coordination, ataxia

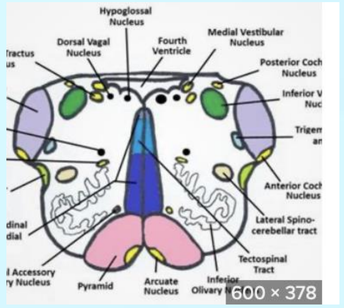
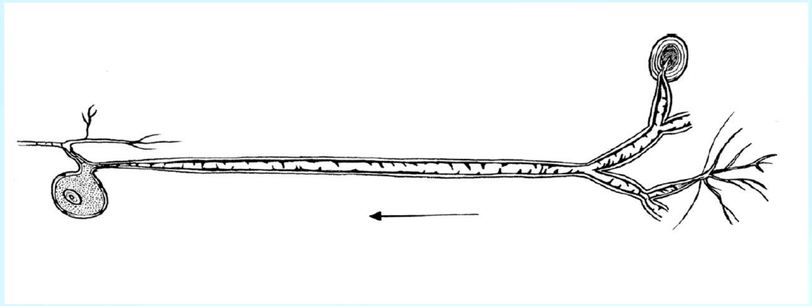
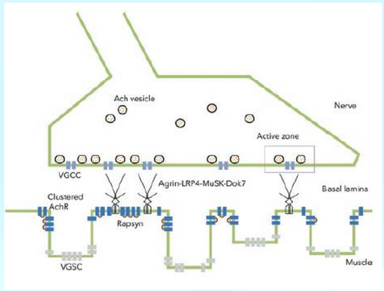
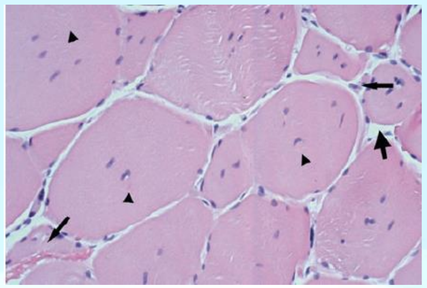
Distribution of weakness



Scales and other functions





Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMT	Muscle	Specific condition
				Sensory ataxia

Tracts

Neurons

Mot/Sensory roots

DRGs

Plexus

Peripheral nerves

Axons

Myelin

Vessels

Pre-

Post synaptic

Muscle fibers

Fibrous tissue

Fasciae

Vessels

Frequency of NM Weakness

	Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMT	Muscle	Specific condition
+++	Cervical spine	GBS and variants	Acute toxic, IT therapy	Myasthenia gravis	Necrotizing Inflammatory	ICU
++	Brainstem		Infections	Botulismus	Channelopathy Hypokaliemic	Sensory
+	Poliomyelitis	SSN	Metabolic	Combat toxins, warfare, bio terrorism	Metabolic	
		Neoplastic	Neoplastic			
			Genetic neuropathy and chemotherapy			

Causes

Vascular

Inflammatory and Autoimmune

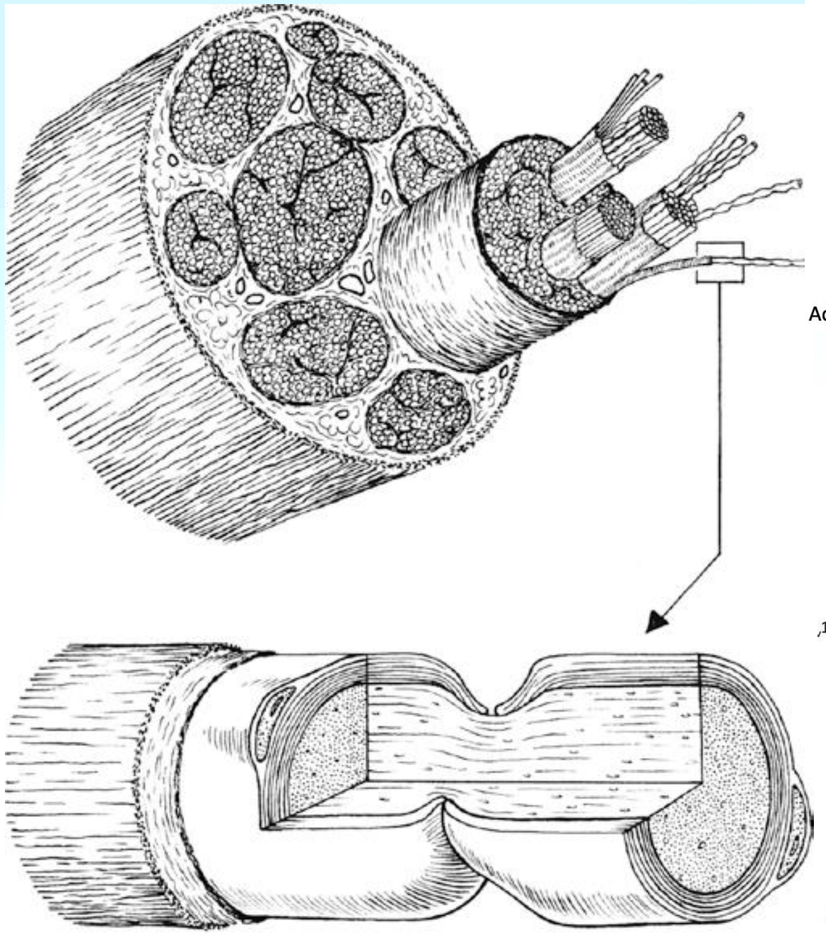
„Granulomatose“

Infection

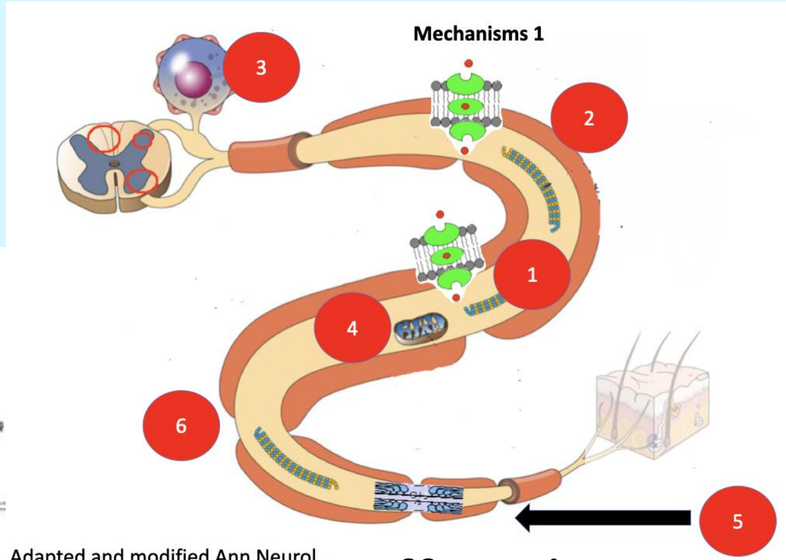
Toxic

Metabolic

Neoplastic

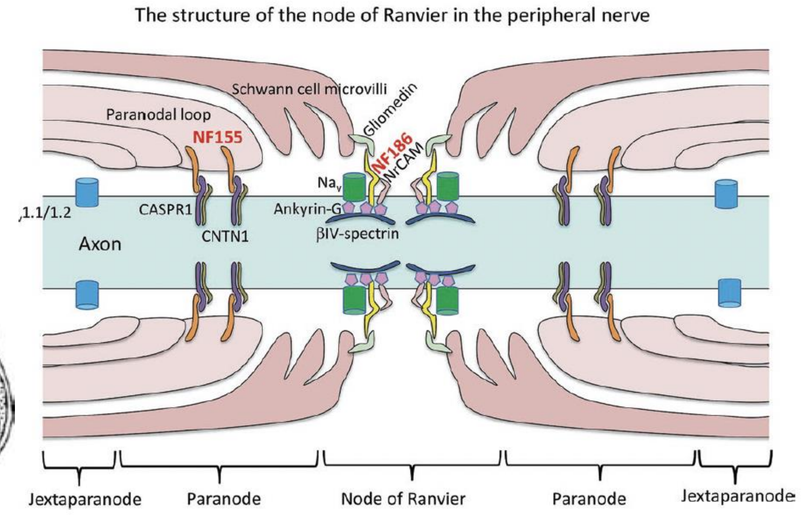


Feldman et al



Adapted and modified Ann Neurol

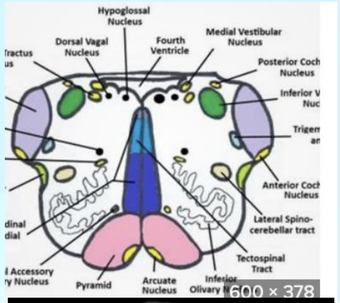
Staff et al.



cell adhesion molecule. (Reprinted from Neurochemistry International, Volume 130, Kira et al. Anti-neurofascin autoantibody and demyelination, Pages 104360, Copyright 2019, with permission from Elsevier)

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMT	Muscle	Specific condition

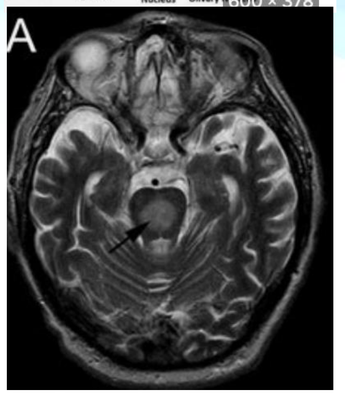
Central Causes as differential



Bilateral medial medullary infarction: a systematic review. J Stroke Cerebrovasc Dis 2013 Aug;22(6):775-80.

Vascular : pontine infarcts, bilateral anterior medullary infarction

Central Pontine Myelinolysis (CPM)

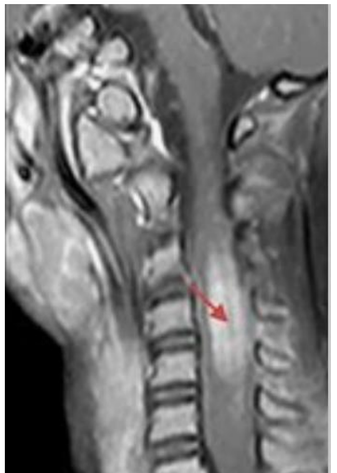


CPM

Transverse Myelopathy (inflammatory, parainfectious, paraneoplastic)

Spinal anterior and posterior artery syndrome

Myelopathy: Immune, paraneoplastic, parainfectious



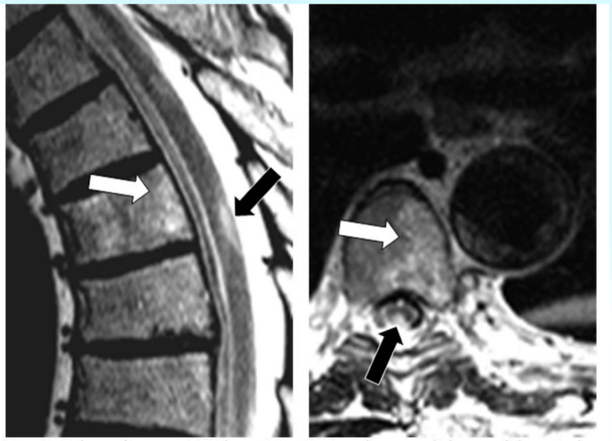
Transverse myelitis

Anterior horn cell disease: eg. Poliomyelitis, Acute Flaccid Myelitis, EV-D68

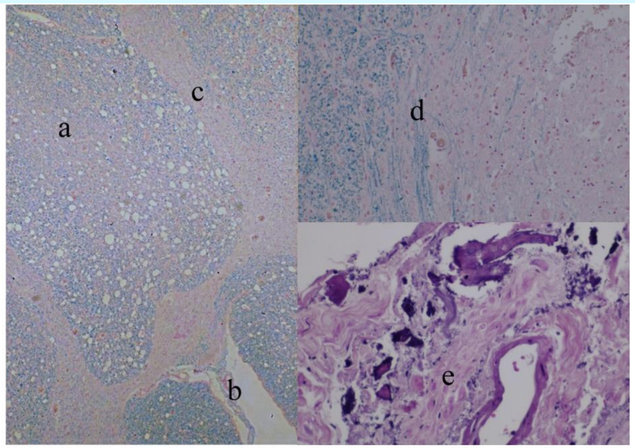
Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMT	Muscle	Specific condition

NO myelopathy

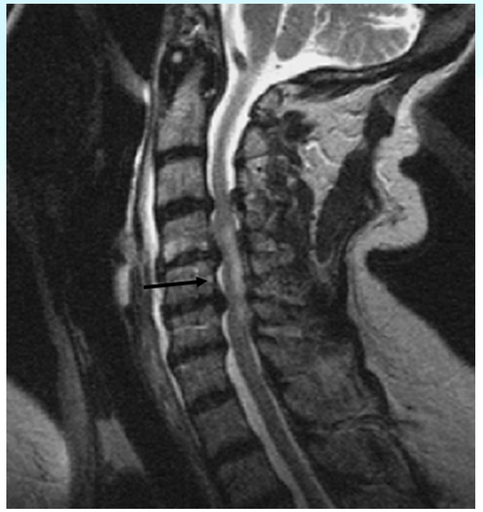
Spinal anterior and posterior artery syndrome

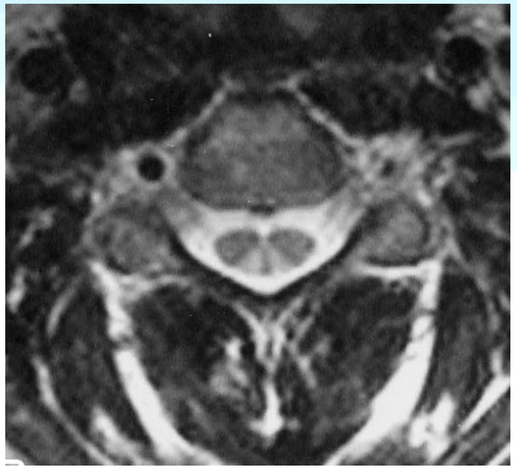


Case 1, 5 days after onset: T2-hyperintensity in a dorso-medial area of the spinal cor at level TH6/7 (a) and of the left lateral parts of vertebral body TH6 (b)



Case 2, histological workup: a, b, c, d: Kluever-Barrera staining, e) Hematoxilin Eosin staining; a) vacuolar ischemic myelopathy of dorsal column, b) anterior median fissure, c) dorsal horn with signs of ischemic lesion, d) borderline between healthy (left) and ischemic (right) area, e) left: calcerous embolism, right: arteriole





AJNR Am J Neuroradiol 1998, 19 (5) 894-896

r Neurol

actions

- Search in PubMed
- Search in NLM Catalog
- Add to Search

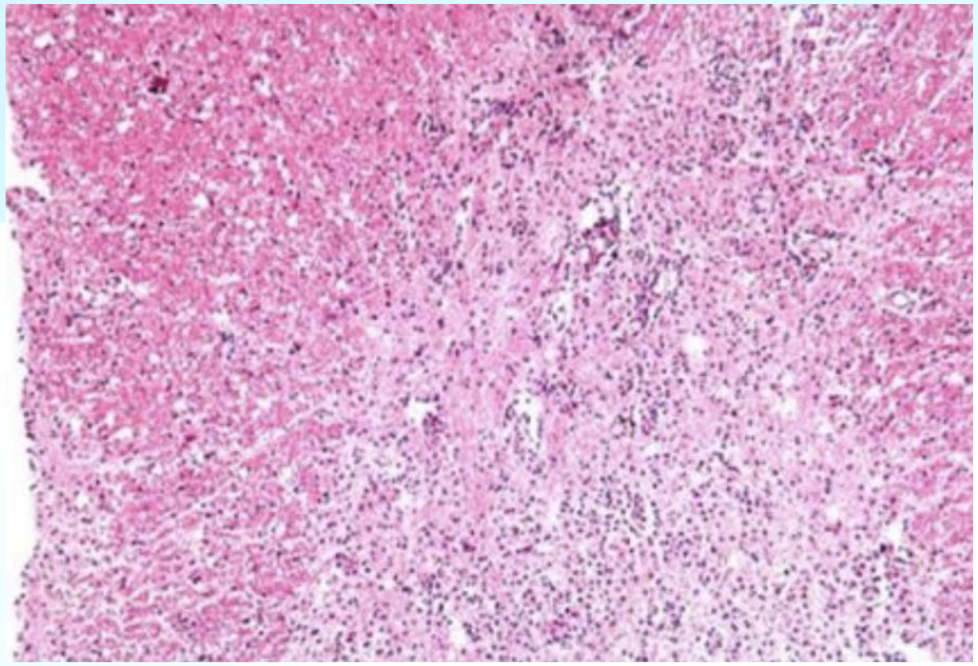
. 2011;65(4):183-6. doi: 10.1159/000324722. Epub 2011 Mar 10.

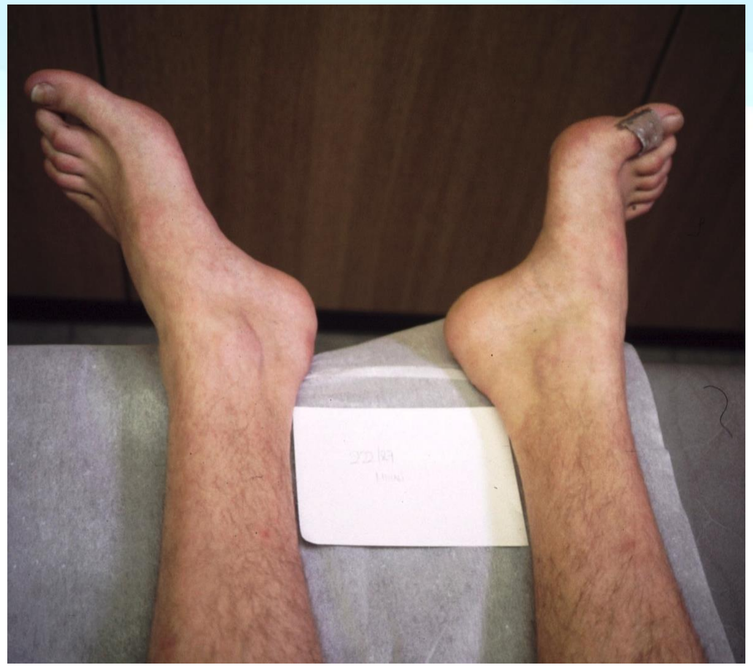
Clinical core symptoms of posterior spinal artery ischemia

Walter Struhal [1], Thomas Seifert-Held, Heinz Lahrmann, Franz Fazekas, Wolfgang Grisold

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMT	Muscle	Specific condition

Poliomyelitis & exacerbation of the Postpolio syndrome in some general conditions. (infection, metabolic issues)





Widdicombe

Rare: Respiratory initial presentation of MND

Central, Pons, Medulla, Spinal cord	**Nerve Roots DRG**	Peripheral Nerves	NMT	Muscle	Specific condition

GBS and variants

West Nile

Radiculopathies: Inflammatory, toxic, neoplastic

Genetic neuropathies and chemotherapy

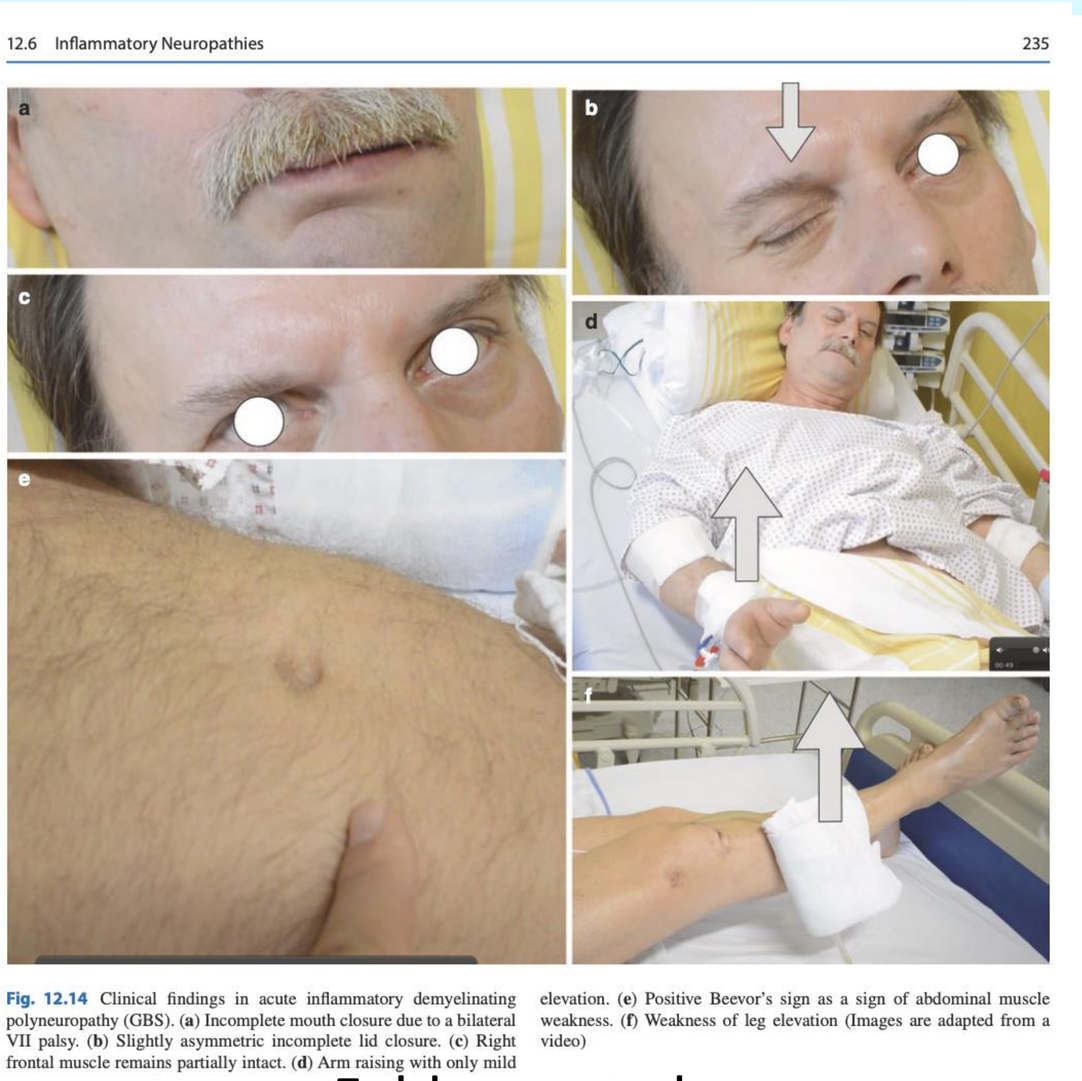


Fig. 12.14 Clinical findings in acute inflammatory demyelinating polyneuropathy (GBS). (**a**) Incomplete mouth closure due to a bilateral VII palsy. (**b**) Slightly asymmetric incomplete lid closure. (**c**) Right frontal muscle remains partially intact. (**d**) Arm raising with only mild elevation. (**e**) Positive Beevor's sign as a sign of abdominal muscle weakness. (**f**) Weakness of leg elevation (Images are adapted from a video)

Feldman et al

Central, Pons, Medulla, Spinal cord	**Nerve Roots DRG**	Peripheral Nerves	NMT	Muscle	Specific condition

GB

Abb. 8.1: Elektroneurografie: Untersucht wurden motorisch der N. tibialis und der N. ulnaris rechts, sensibel der N. suralis und der N. ulnaris rechts. Beide motorische Nerven zeigen verlängerte F-Wellenlatenzen, bei Ableitung des N. tibialis finden sich zusätzlich A-Wellen (Pfeil).

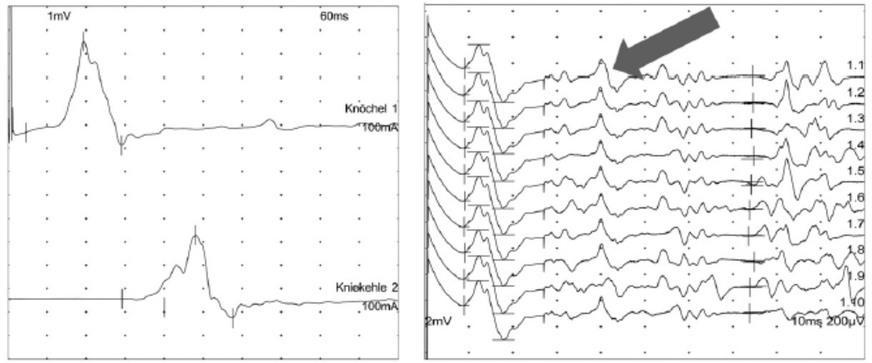
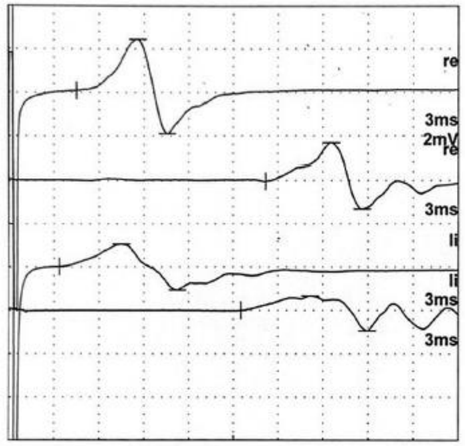
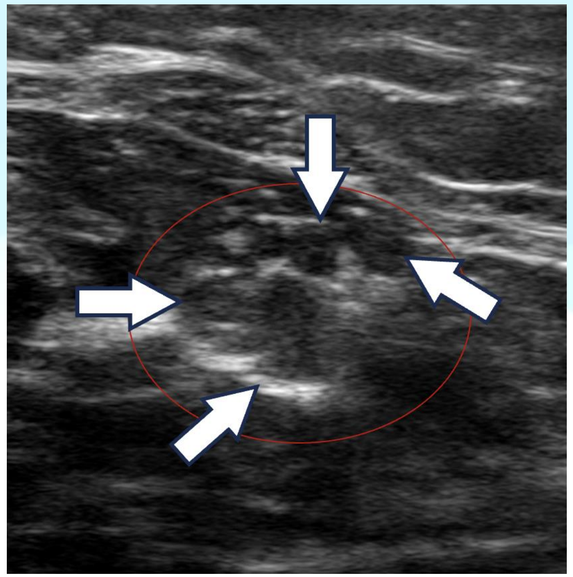


Abb. 7.1: N. tibialis bds. insbesondere proximal mit deutlicher Chronodispersion und verlangsamter mNLG



Lehmann et al



Ultrasound

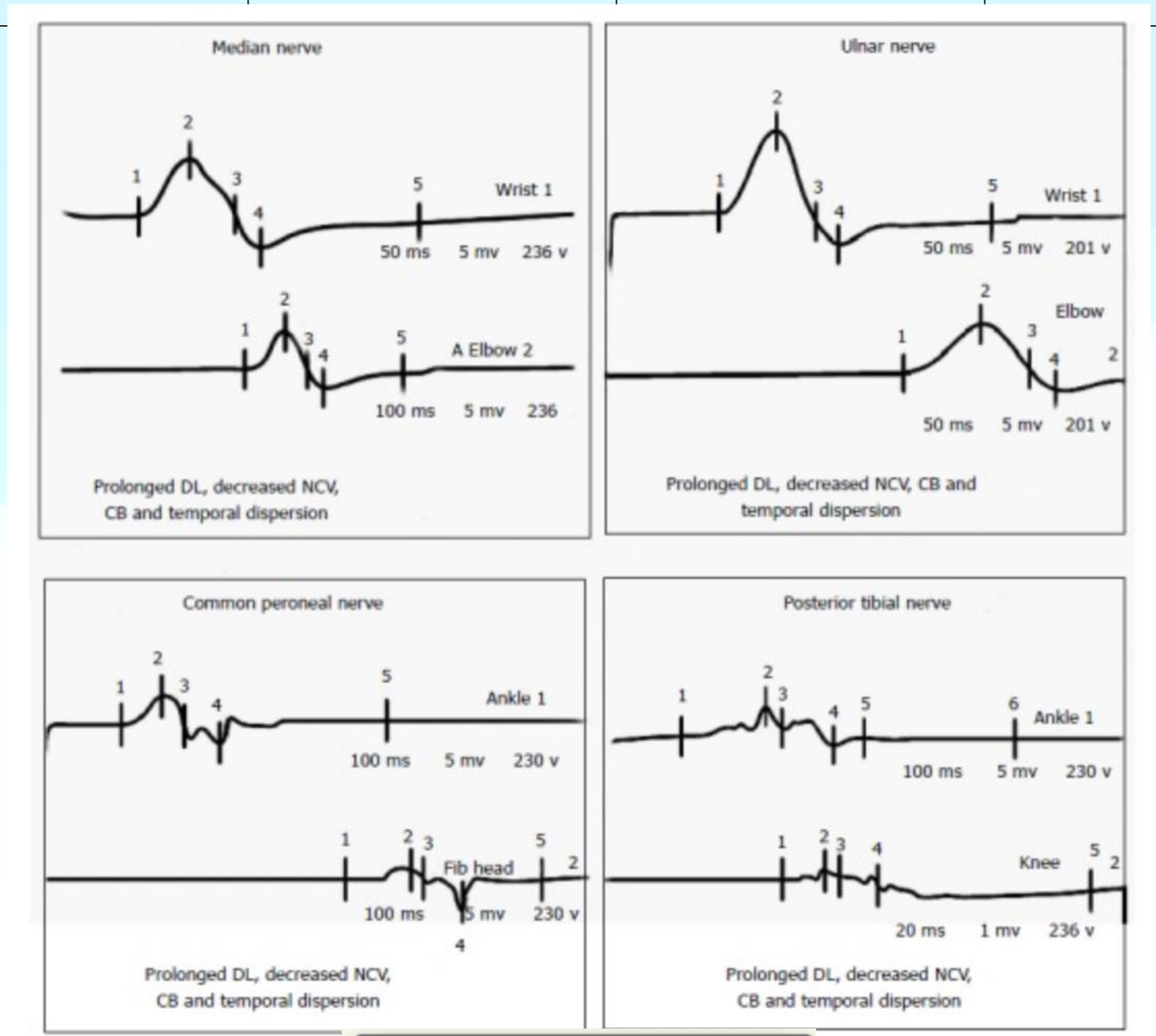
Median nerve

thickened

Dr Meng

Central, Pons, Medulla, Spinal cord	**Nerve Roots DRG**	Peripheral Nerves	NMT	Muscle	Specific condition

GBS



https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5535318/figure/F2/

Central, Pons, Medulla, Spinal cord	**Nerve Roots DRG**	Peripheral Nerves	NMT	Muscle	Specific condition

Cranial

Nerves

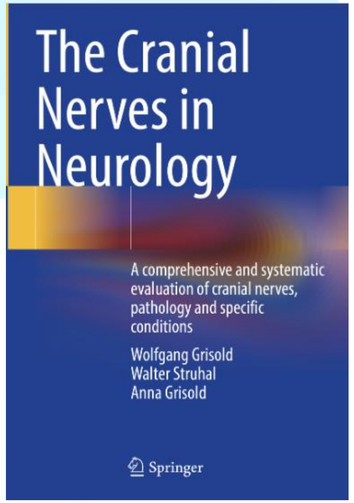


Table 3.7.1 Clinical subtypes of Guillain-Barré syndrome

	AIDP	AMAN	AMSAN	MFS	FDP
Frequency Distribution	Frequent in western countries	Rare	Rare	Rare	Rare
Symptoms	Progressive para-/tetraparesis Sensory deficits Weak or absent reflexes	Progressive para-/tetraparesis No sensory deficits Weak or absent reflexes	Often severe tetraparesis Severe sensory deficits Absent or reduced reflexes	Ophthalmoplegia Areflexia Ataxia	Paresthesias Absent or only minor motor deficit
Cranial nerve deficits	Frequent	Occasionally, but less frequent than in AIDP	Present	Specific pattern with absent eye movements, anisokoria, etc.	Bilateral facial weakness
Autoantibodies	None specific	GM1 IgG GD1a IgG	GM1 IgG GD1a IgG	GQ1b IgG	

AIDP, acute inflammatory demyelinating polyneuropathy; AMAN, acute motor axonal neuropathy; AMSAN, acute motor-sensory axonal neuropathy; FDP, facial diplegia with paresthesias; GD1a/GM1/GQ1b, gangliosides; IgG, immunoglobulin; MFS, Miller Fisher syndrome

Lehmann et al, 2023

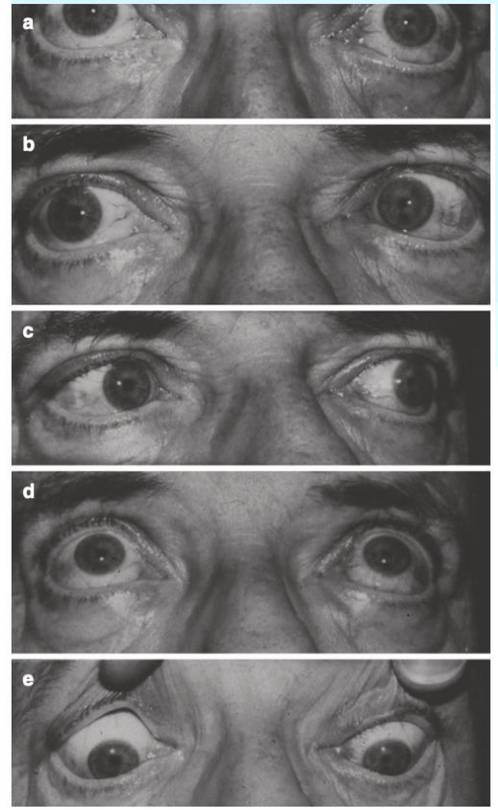


Fig. 12.15 Miller Fisher syndrome. (**a**) Patient with Miller Fisher syndrome and ophthalmoparesis with (**b**, **c**) restricted horizontal gaze, and (**d**, **e**) reduced upward and downward gaze

Zifko et al

Central, Pons, Medulla, Spinal cord	**Nerve Roots DRG**	Peripheral Nerves	NMT	Muscle	Specific condition

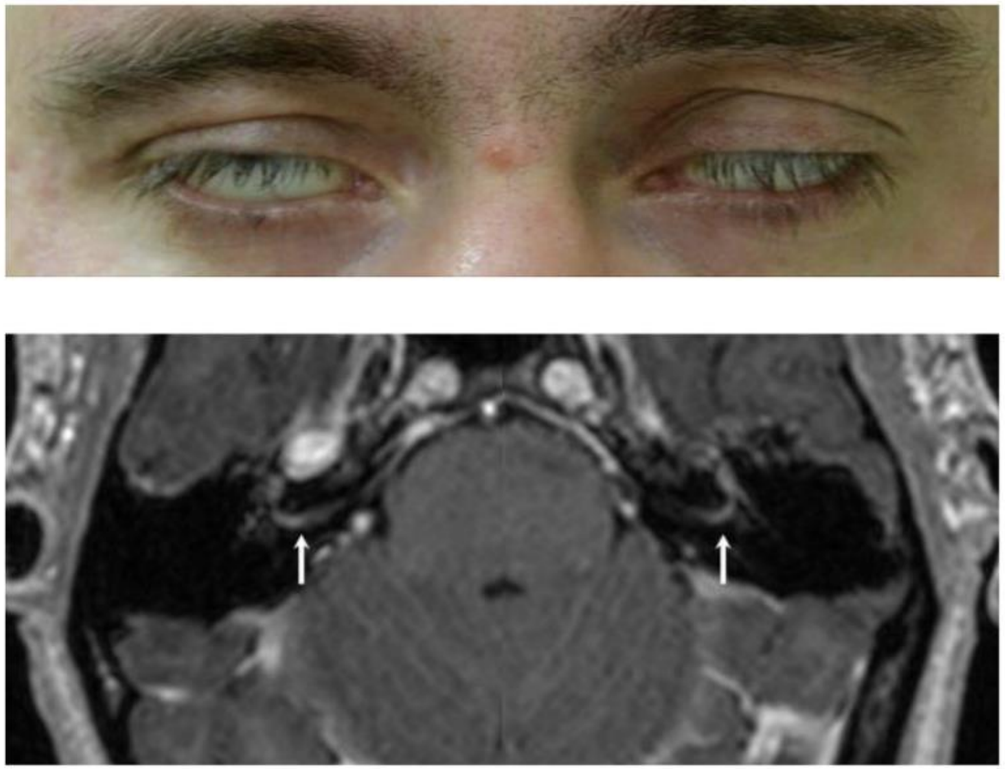


Fig 3.7.3 A. Bilateral Bell's palsy with inability for complete eyelid closure (bilateral lagophtalmus). B. After intravenous gadolinium administration, bilateral strong enhancement of the facial nerve in its extra- and intracanalicular segments is visible (white arrows), axial T1-weighted gradient echo sequence (reproduced with permission from [33]).

Lehmann et al, 2023

Central, Pons, Medulla, Spinal cord	**Nerve Roots DRG**	Peripheral Nerves	NMT	Muscle	Specific condition

GBS and CN VII

after

Covid vaccination

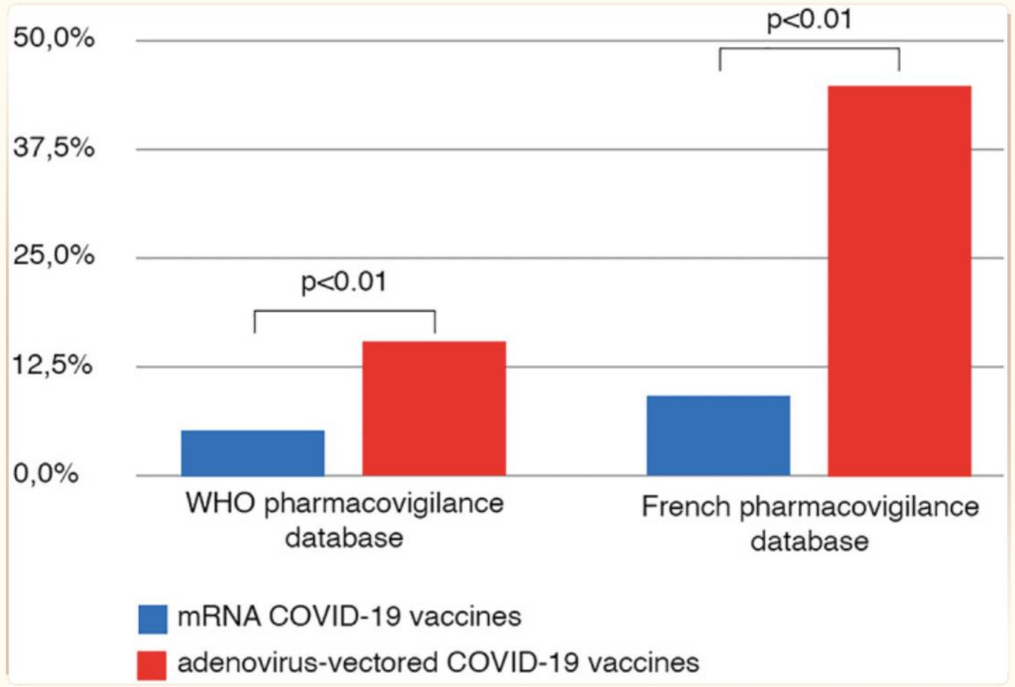


FIGURE 1

Frequency of facial paralysis associated with Guillain–Barré syndrome after COVID-19 vaccine administration. WHO = World Health Organization. [Color figure can be viewed at www.annalsofneurology.org]

Ann Neurol. 2022 Jan; 91(1): 162–163.
Published online 2021 Nov 12. doi: 10.1002/ana.26258
PMCID: PMC8652690
PMID: 34699065

Adenovirus COVID-19 Vaccines and Guillain–Barré Syndrome with Facial Paralysis

Among the 48,907 cases reported with COVID-19vaccines, there were 69 (0.1%) cases of GBS, of which 23 involved FP (33.3%).
This included 2 of 22(9.1%) GBS patients who received mRNA vaccines (Pfizer–BioNTech) and 21 of 47 (44.7%) who received adenovirus-vectored vaccines (20/44 [45.5%] Oxford–AstraZeneca, 1/3 [33.3%] Johnson & Johnson), also indicating a higher frequency of FP-GBS occurring after adenovirus-vectored vaccines (Fisher exact test:p = 0.0053; see Fig 1).

Central, Pons, Medulla, Spinal cord	**Nerve Roots DRG**	Peripheral Nerves	NMT	Muscle	Specific condition

World Health Organization (WHO) pharmacovigilance database, VigiBase:

1,257,497 cases reported with COVID-19 vaccines, 1,256 (0.1%) “acute polyneuropathies” (all cases correspo and 119 elsewhere).

0 ,09 %

Ann Neurol. 2022 Jan; 91(1): 162–163.
Published online 2021 Nov 12. doi: 10.1002/ana.26258
PMCID: PMC8652690
PMID: 34699065

Adenovirus COVID-19 Vaccines and Guillain–Barré Syndrome with Facial Paralysis

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https://who-umc.org/vigibase/

Central, Pons, Medulla, Spinal cord	**Nerve Roots DRG**	Peripheral Nerves	NMT	Muscle	Specific condition

GBS Differentials

8 Patientin mit Guillain-Barré-Syndrom

Erkrankung	Wegweisende Befunde
Transverse Myelitis	Gestörte Blasen- und Mastdarmfunktion, gesteigerte MER, Babinski-Reflex positiv
Myelopathie	Gestörte Blasen- und Mastdarmfunktion, gesteigerte MER, Babinski-Reflex positiv
Tetanus	Muskelkrämpfe, Spastik, Trismus
Rabies	Zentralnervöse Symptome, Affektstörungen (Angst), Orientierungs- und Vigilanzstörungen
Neuroborreliose	Reißende Schmerzen, Erythema migrans, pluriradikuläre Verteilung
Critical illness Neuropathie	Eher axonale Schädigung in der NLG/EMG-Untersuchung
Myasthenia gravis	Doppelbilder, tageszeitabhängige Fluktuation
Myopathien/-myositiden	Eher proximal betonte Schwäche, massive CK-Erhöhung
Psychogene Lähmung	Diskrepanz zwischen Schwere der Erkrankung und Besorgtheit (»belle indifference«), normal erhältliche Muskeleigenreflexe
Exazerbation einer länger bestehenden Polyneuropathie	Muskelhypotrophie

Tab. 8.3: Differenzialdiagnose des GBS mit wegweisenden klinischen Befunden

Transverse myelitis

Myelopathy

Tetanus Cramps, trismus

Rabies CNS

Lyme (?)

CIP

MG

Myopathy proximal weakness, CK elevation

Psychogenic

Exacerbation of a preexisting neuropathy

Central, Pons, Medulla, Spinal cord	**Nerve Roots DRG**	Peripheral Nerves	NMT	Muscle	Specific condition

Ganglionitis

SSN

Sjögren

Idiopathic

Nitrous oxide

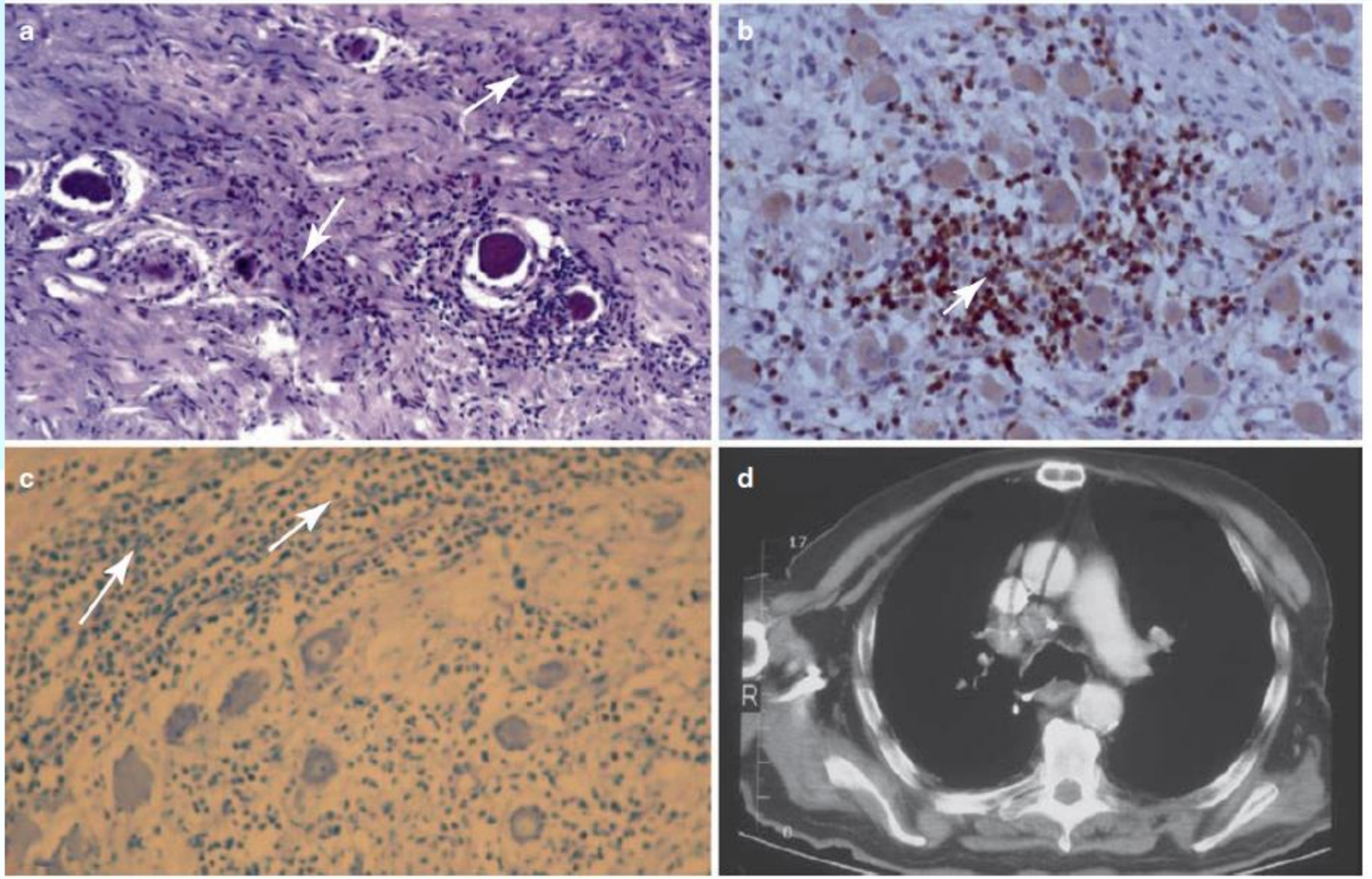
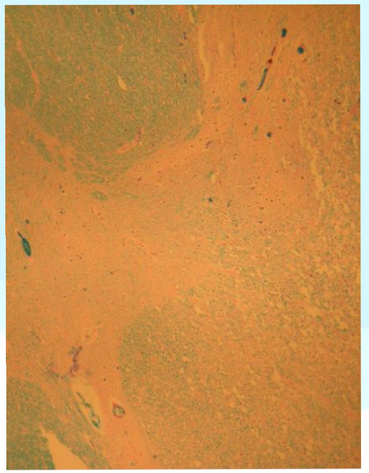
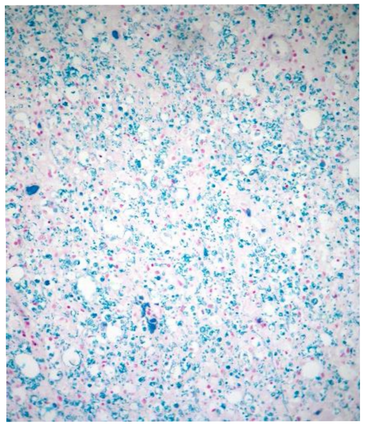


Fig. 12.28 Paraneoplastic ganglionopathy. (**a–c**) Dorsal root ganglion (DRG) pathology. (**a**, **b**) Examples of an inflammatory paraneoplastic ganglionitis (*arrows*). (**b**) An infiltrate that is immunostained for T-cells (*arrow*). (**c**) Rare example of neoplastic infiltration of a DRG by lymphoma cells (*arrows*) of a Burkitt-like lymphoma. This patient had additional meningeal infiltration. (**d**) Paraneoplastic ganglionopathy in a patient with small cell lung cancer. Chest CT shows enlargement of the mediastinal lymph nodes

Feldman et al





Posterior collumn degeneration

Image: Widdicombe, 77

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	**Peripheral Nerves**	NMT	Muscle	Specific condition

Acute Neuropathies

Porphyria

Diphtheria

POEMS Syndrome

Organophosphates (in stages)

Infections

Toxic

ICUAW

Vasculitis often asymmetric

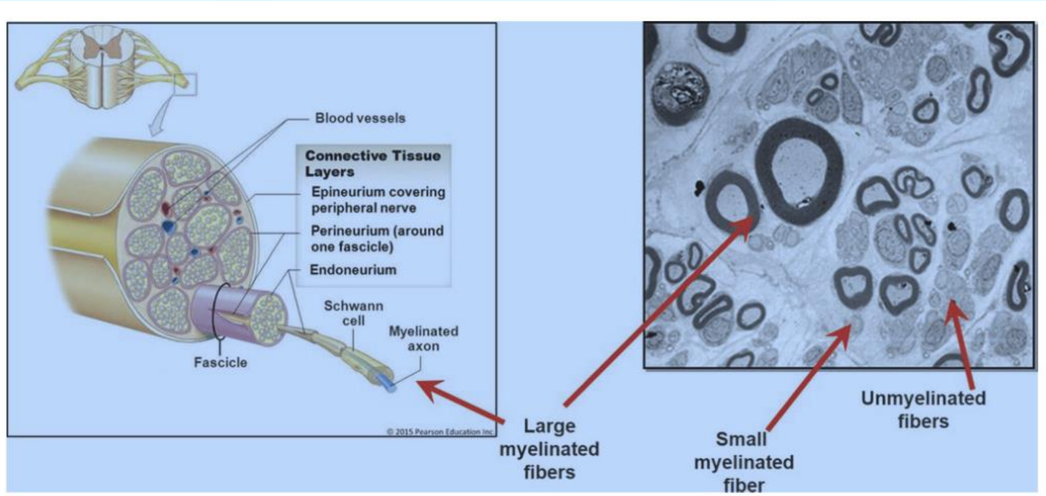
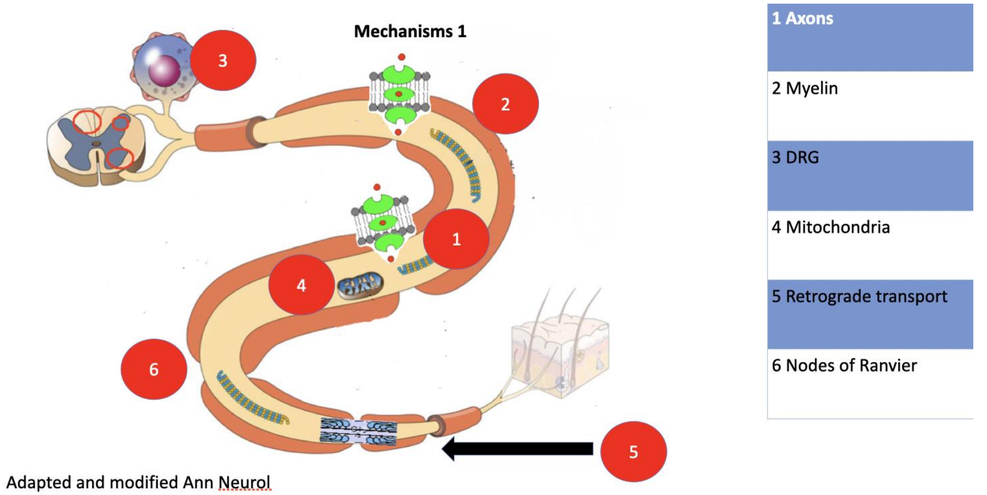


Fig. 12.1 Cross-section diagram (left) and electron micrograph (right) of a nerve fiber showing large and small fibers. Left panel reprinted from Surgery (Oxford), Volume 37, Issue 5, Nicholls and Furness, Orthopaedics– III: upper limb/Peripheral nerve compression syndromes of the upper limb, Pages 288–293, Copyright (2019), with permission



Adapted and modified Ann Neurol

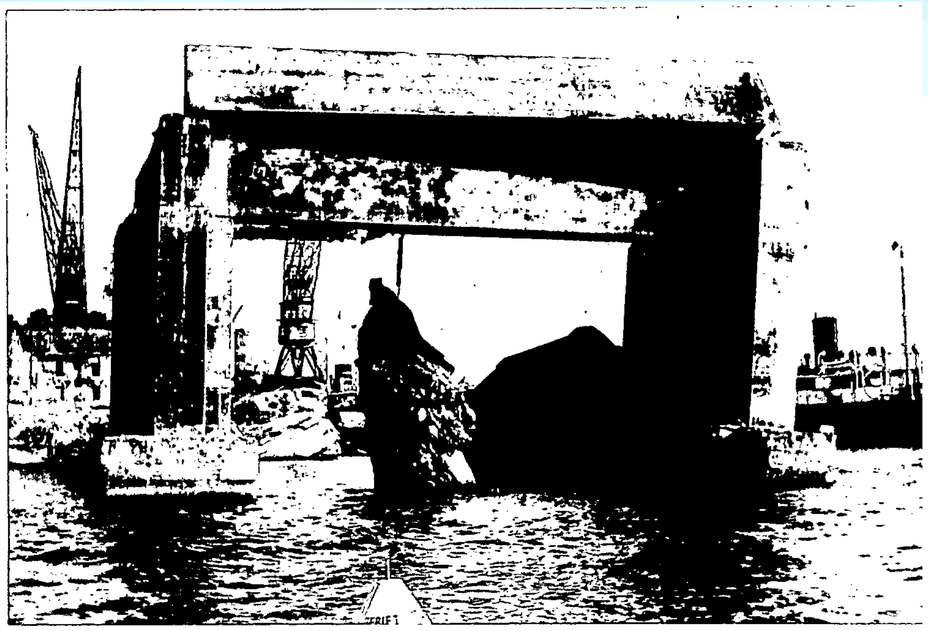
Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	**Peripheral Nerves**	NMT	Muscle	Specific condition

Acute Neuropathies

Organophosphates (Stages)

The case of TCE (triorthocresyl)- or U- boat (submarine or torpedo) oil

- Used as lubricant
- Ginger jake paralysis (US 1930)
- Used as oiler cooking (Switzerland,
- Germany) or transport of oil in
- contaminated jars.
- Self propagating, disabling, spinal involvement



Gesprengter U-Boot-Bunker in Kiel 1945: „Keine kriegseigentümliche Gefahr"

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	**Peripheral Nerves**	NMT	Muscle	Specific condition

Acute Neu

TRI-ORTHO-CRESYL PHOSPHATE (T.O.C.P.)

	Intermediate syndrome	Delayed neuropathy
Time of onset	1- 4 days	2- 3 weeks
Site of weakness		
Limb	Proximal	Distal
Neck	+	-
Cranial nerves	+	-
Respiratory muscles	+	-
EMG	Tetanic fade	Denervation
Recovery	4- 18 days	6 – 12 months
Agents	Fenthion Dimethoate Monocrotophos	Metamidophos Trichrolophon Leptophos

Courtesy: Faouzi Belahsen, mod . NEJM, 316, 761 (1987)

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	**Peripheral Nerves**	NMT	Muscle	Specific condition

Acute Ne

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	**Peripheral Nerves**	NMT	Muscle	Specific condition

Acute Neuropathies

Major outbreaks of TOCP poisoning

Years	*Place*	*No of cases*	*Vehicles of TOCP*
1898–1900	France	6	Phospho–creosote[6]
1900–28	Europe	43	Phospho–creosote[7]
1930–1	USA	30–50 000	Ginger extract[8]
1930–5	Europe	hundreds	Apiol(abortifacient)[4]
1938	Durban-1	68	Cooking oil[11]
1940	Basel (Switzerland)	80	Cooking oil[10]
1940–6	Germany	hundreds	Cooking oil[2]
1942–3	Verona (Italy)	41	Ground contamination (this study)
1945	Liverpool (United Kingdom)	17	Cooking oil[12]
1955	Durban-2	11	Contaminated water[13]
1956	Japan	6	Cooking oil[14]
1959	Morocco	10 000	Cooking oil[18]
1960	Bombay (India)	58	Cooking oil[19]
1962	West Bengal	400	Contaminated flour[16]
1966	Romania	12	Liquor[9]
1967	Fiji	56	Contaminated flour[17]
1971–2	Vietnam	15–20	Cooking oil[20]
1977–8	Sri Lanka	20	Sesame oil[15]

Courtesy: Faouzi Belahsen

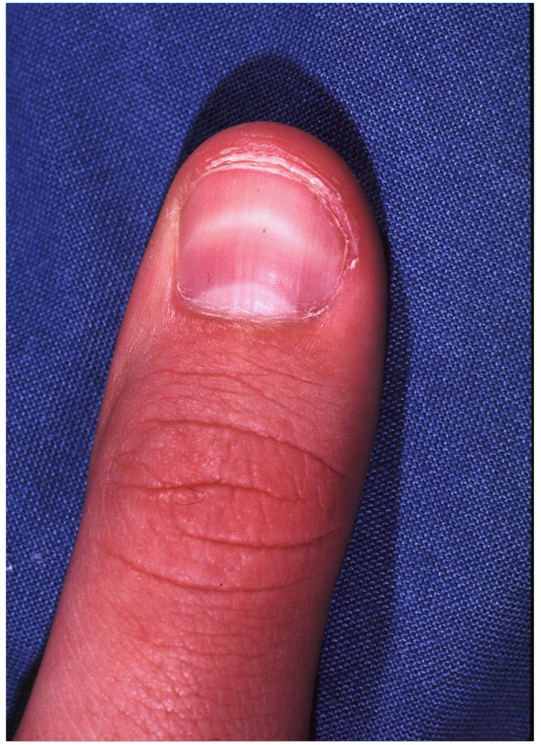
Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral **Nerves**	NMT	Muscle	Specific condition

Acute Neuropathies

Arsenic

- Acute: sensory , may resemble GBS (also coasting)

- Chronic
- Occupational
- Groundwater contamination (eg Bangladesh)
- Contamination of liquor



From EL Feldman et al
Atlas of neuromuscular disease
Springer 2014

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	**Peripheral** Nerves	NMT	Muscle	Specific condition

Acute Neuropathies

- **The Spanish Toxic Oil Syndrome 20 Years after Its Onset: A Multidisciplinary**

- Review of Scientific Knowledge
- Emilio Gelpí, et al, Environ Health
- Perspect 110:457–464 (2002) .

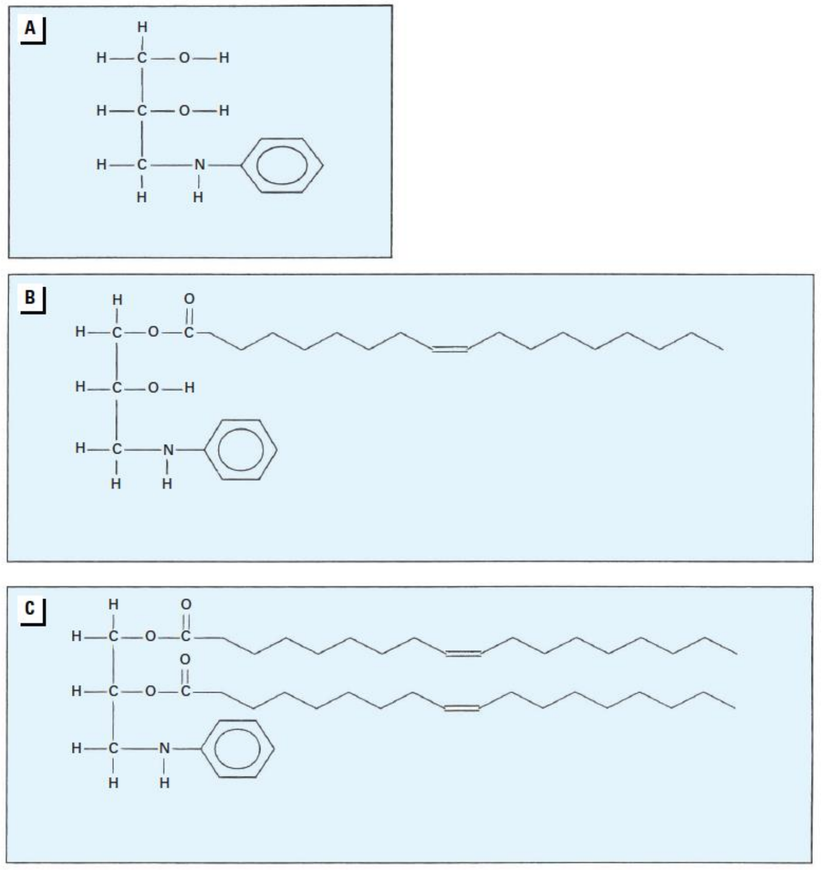


Figure 1. Structural formulas of derivatives of (*A*) PAP and its esters (*B*) O PAP and (*C*) OO PAP.

fatty acid esters of 3-(N-phenylamino)-1,2-propanediol (PAP).

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	**Peripheral** Nerves	NMT	Muscle	Specific condition

Jelly fish (Cnidarian peptide) (2000 types)

- Sensory symptoms
- Transient focal neuropathy of the motor and sensory nerves of the tongue and pharynx after swallowing water contaminated
- Isolated mononeuropathie – also multiplex
- GBS

- Vasospasm- compartment syndrome

Small Puffer Fish

Vascular
Direct toxic
n channels
euronal excitability
mune mediated

Journal of CLINICAL NEUROMUSCULAR DISEASE
Volume 14, Number 4
June 2013

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Biological toxins

- Porcine aerosol

- Marine toxins
- Ciguatoxin
- Tetrodoxin
- Jellyfish

- Shell fish
- Brevetoxin, saxitoxin

- Multiple : venoms (snakes, spiders)

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	**Peripheral** Nerves	NMT	Muscle	Specific condition

Associations with other infections

HIV

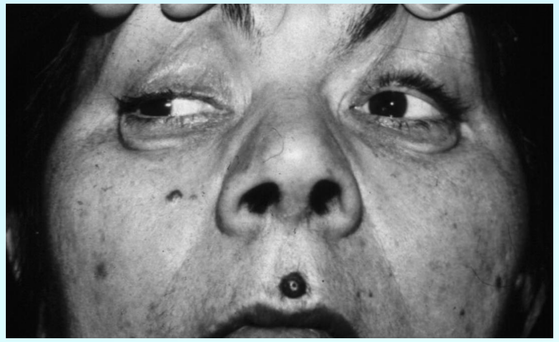
Clinical syndromes: Occur in specific phases of infection

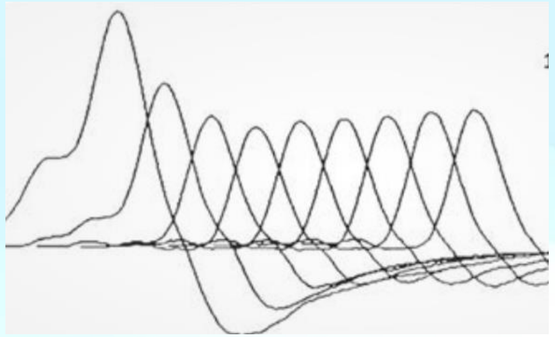
Early: Usually immune mediated
ALS-variant syndrome
Brachial plexopathy
CIDP
Guillain-Barré
Mononeuritis multiplex (Vasculitis)
Myasthenia gravis
Neuromyotonia
Immune myopathy
Cytoplasmic body (Rod) myopath

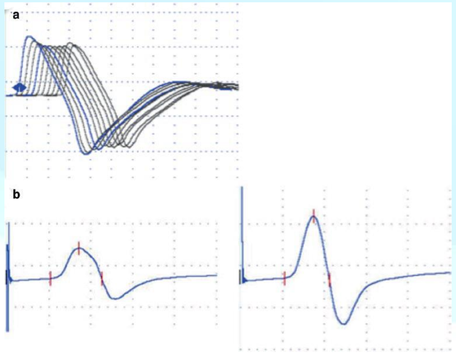
Hepatitis C
West Nile : neuropathy, myopathy, poliomyelitis
African tick bite fever
Dengue fever (acute neuropathy)
ZIKA
Covid SARS ?

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	**NMT**	Muscle	Specific condition

Myasthenia gravis

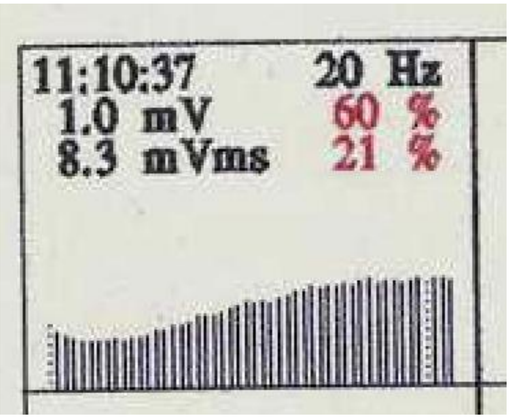






Botulism

Organophosphates „Nerve gas“, eg Sarin



One antidote, atropine, blocks acetylcholine receptors, sparing the body's muscles from overstimulation. The other, pralidoxime, or 2-PAM, removes sarin from the enzyme that stops acetylcholine from accumulating, Nelson said. However, both antidotes must be given within about

Image: Widdicombe, 77

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	**NMT**	Muscle	Specific condition

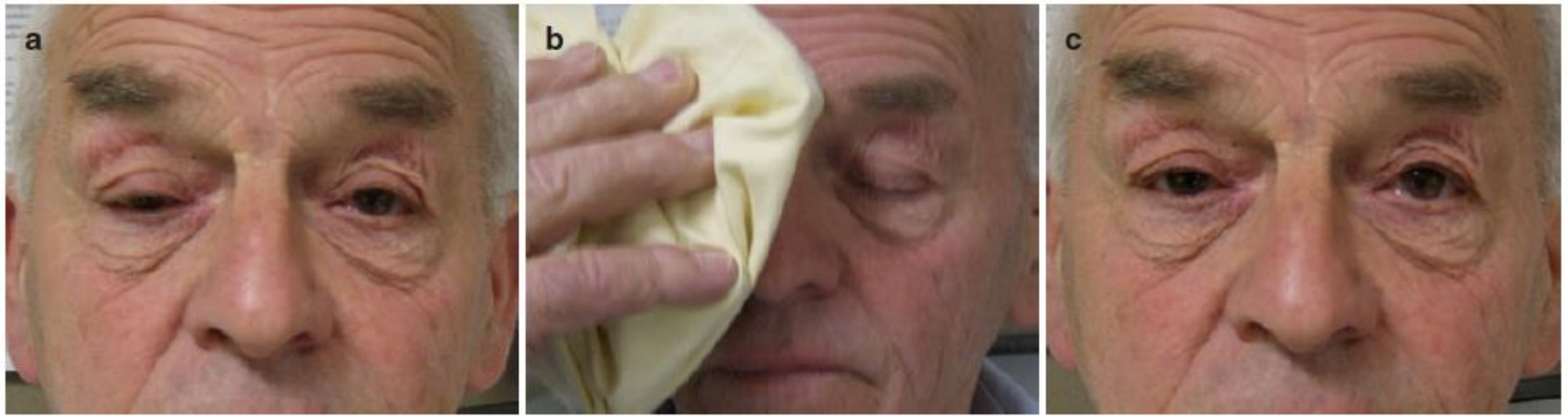


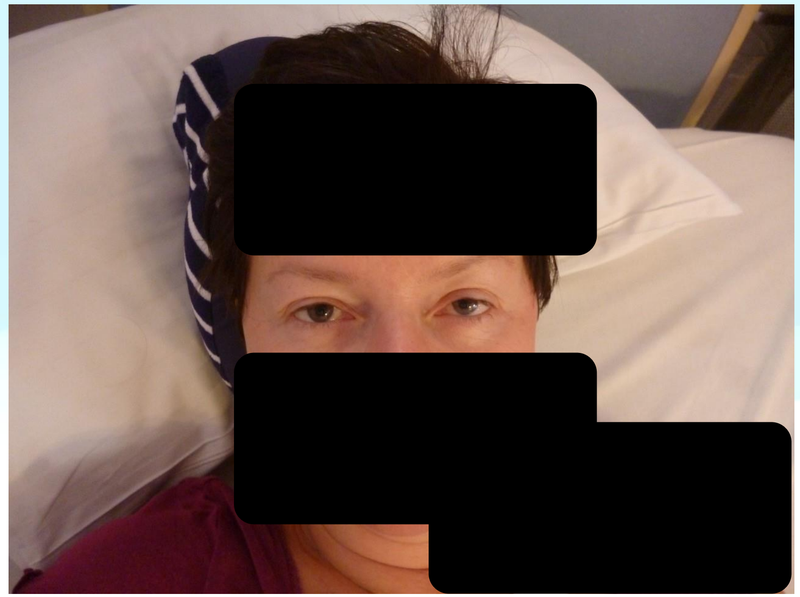
Fig. 13.6 Ice (pack) test. The ice pack test is a useful bedside test. (**a**) Patient with ptosis on the right. (**b**) A pack with ice is applied to one eye, usually 2 min is suggested. (**c**) Positive test with remission of ptosis. The effect is based on the fact that acetylcholinesterase activity is inhibited below 29 degrees Celsius

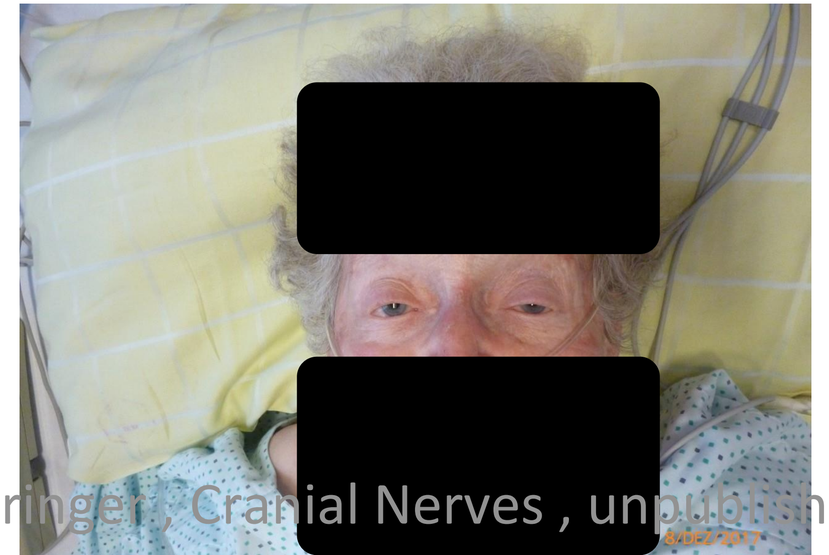
Feldman et al

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	**NMT**	Muscle	Specific condition

Botulism

- **Early symptoms**
  - Dysphagia Dry mouth , slurred speech
  - Vision blurred , diploma
  - Speech: Slurred; Difficult; Hoarse voice
  - Nausea, vomitting, constipation
    - Nausea & vomiting: More with food bourne botulism
    - Constipation: Childhood botulism
- **Weakness**
  - CN: Ptosis, EO muscles
    - Dyphagia, dysarthria
  - Diffuse weakness
    - Usually symmetric
    - Proximal > Distal
    - "Descending paralysis"
    - Respiratory
- **Sensory loss: Never prominent**
- Tendon reflexes
  - Reduced
- **Autonomic**: pupils, heart, hypotension
- hyperhydrosis , urinary retention





One antidote, atropine, blocks acetylcholine receptors, sparing the body's muscles from overstimulation. The other, pralidoxime, or 2-PAM, removes sarin from the enzyme that stops acetylcholine from accumulating, Nelson said. However, both antidotes must be given within about 10 minutes of exposure in order to be effective, he said.

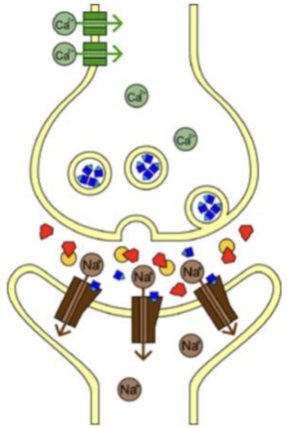
Springer , Cranial Nerves , unpublished

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	**NMT**	Muscle	Specific condition

Organophosphates „Nervengas“, eg Sarin

Warfare agents

- History: spear venoms
- Sarin and others
- Aerosols and skin !
- Tokyo incident



Schematische Wirkungsweise von Sarin am synaptischen Spalt. Sarin (rot), Acetylcholinesterase (gelb), Acetylcholin (blau)

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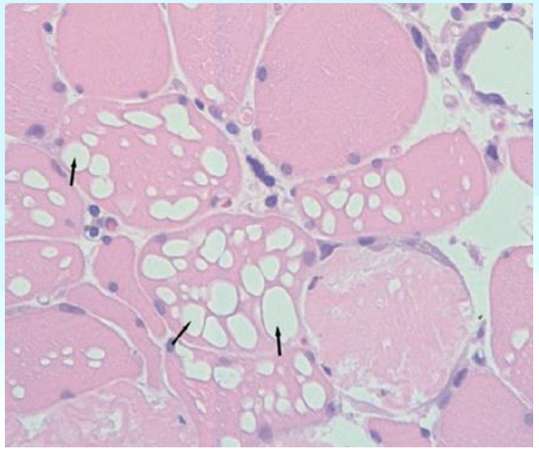
One antidote, atropine, blocks acetylcholine receptors, sparing the body's muscles from overstimulation. The other, pralidoxime, or 2-PAM, removes sarin from the enzyme that stops acetylcholine from accumulating, Nelson said. However, both antidotes must be given within about

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMT	**Muscle**	Specific condition

Myopathy



Inflammatory myopathies
Necrotizing myopathy
Rhabdomyolysis
Periodic Paralysis
Electrolyte Disorders, hypercalcemia, hypocalcemia
Myosin deficiency (Critical illness myopathy)
Carnitine Deficiency

Neuromuscular Blockade

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMT	**Muscle**	Specific condition

Hypokalemic Periodic Paralysis

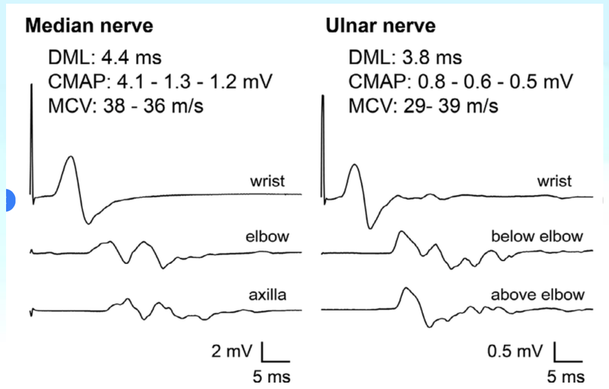


Figure1.Motor nerve conduction study in the median and ulnar nerves. Excessive temporal dispersions of proximal compound muscle action potentials (CMAPs) were recorded in the forearm segments of the median and ulnar nerves. Motor conduction velocities (MCVs) were also reduced (Normal, >50 m/s in the median and >52 m/s in the ulnar nerves). In contrast, distal motor latencies (DMLs) were nearly normal (normal: <4.1ms in the median nerve and <3.6ms in the ulnar nerve).

Electrodiagnostic
- EMG
  - Between attacks: Normal
  - During attacks: Irritability or Reduced insertional activity
- CMAP amplitude
  - Reduced during attacks
  - Increased immediately after sustained (5 min) maximal contraction
  - Progressively reduced (by 40%) during rest 20 to 40 min after initial increment (80% of patients)
  - Normals: Mild increase in CMAP amplitude after exercise
  - Epinephrine: Reduces size of CMAP

Hereditary

Ca^{++}
- Ca^{++} channel: CACNA1S; 1q32
- Na^{+} channel: SCN4A; 17q23
- Gitelman: SLC12A3; 16q13
- **K^{+} channel**
  - KCNE3: 11q13
  - KCNJ2: 17q24
  - KCNJ5: 11q24

- Renal tubular acidosis: SLC4A1; 17q21
  - **Gossypol myopathy**
- HOPP + CNS: ATP1A2; 1q23
- Thyrotoxic (TTPP)
  - 1: CACNA1S; 1q32
  - 2: KCNJ18; 17p11
  - 3: 17q24.3

Acquired
- K^{+} wasting
- Dengue fever

https://neuromuscular.wustl.edu/mother/myotox.htm

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMT	**Muscle**	Specific condition

Hypokalemic Paralysis

Barium myopathy (Hypokalemic)

Toxicology

Toxin: Soluble salts; Acetate, Carbonate, Chloride, Hydroxide, Nitrate, Sulfide Doses (Oral): Toxic 200 mg; Lethal 1 to 15 g Exposure

Oral: Suicide; Food contamination (Table salt, Flour, Potato meal substitution Inhalation
Burns: Molten barium chloride

Clinical
Acute toxicity

GI: Nausea; Vomiting; Diarrhea; Abdominal pain; Xerostomia

Perioral paresthesias (Occasional) Weakness

Quadriparesis: Flaccid
Muscle twitching
No involvement of cranial nerves or respiration Reflexes: Often absent; May be preserved Rhabdomyolysis: Occasional

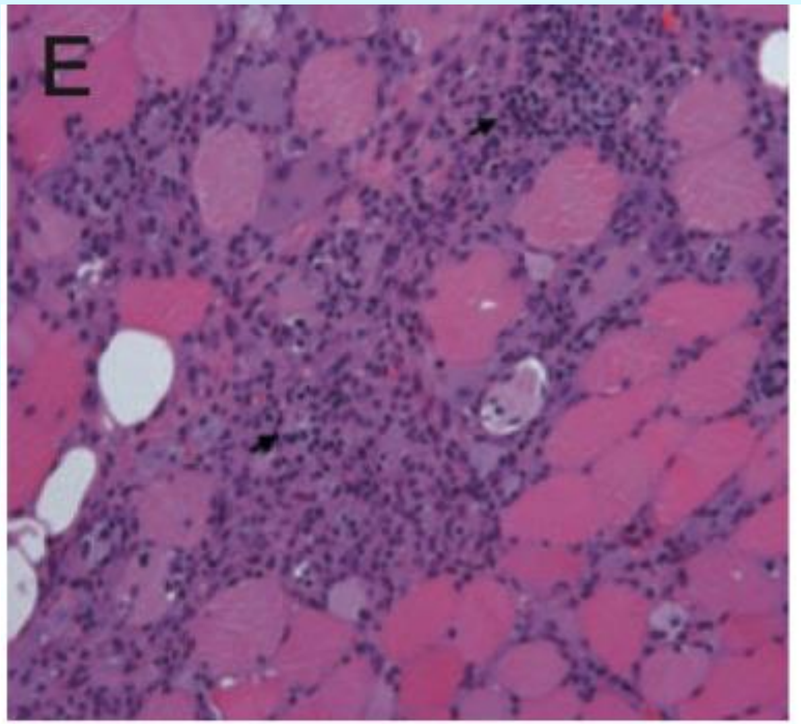
https://neuromuscular.wustl.edu/mother/myotox.htm

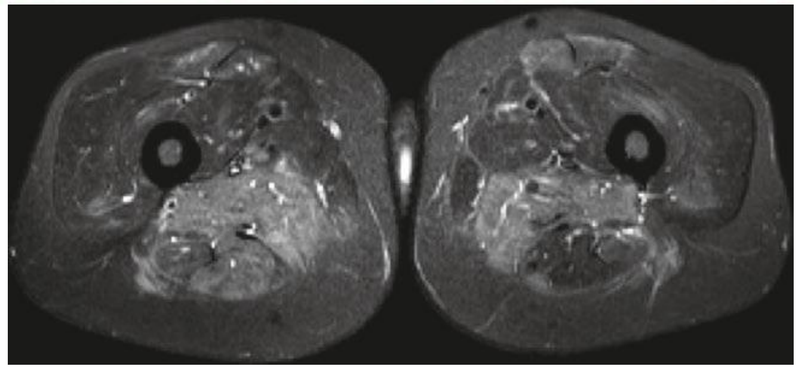
Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMT	**Muscle**	Specific condition

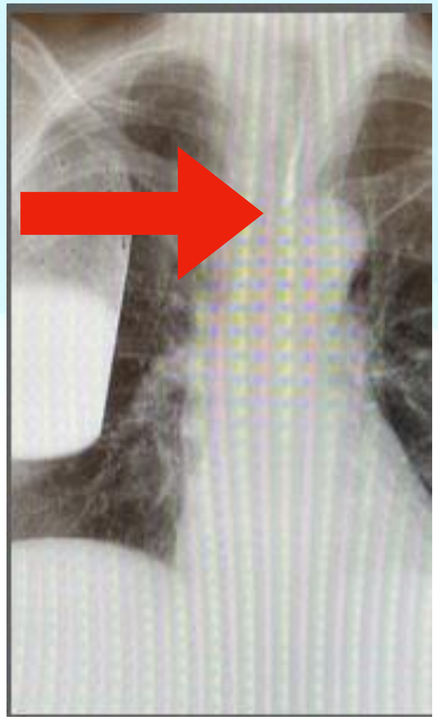
Myopathy

Inflammatory myopathies

Necrotizing myopathy
Immune mediated
Toxic myopathies







Case: Poliomyositis in remission.
Swallowing difficulties
Pseudosclerodermiform stenosis

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMT	**Muscle**	Specific condition

Myopathy

Inflammatory myopathies

SPECIFIC IMMUNE OR INFLAMMATORY MYOPATHIES

Immune myopathies (Polymyositis +...)
General aspects
- Clinical
- Laboratory
- Subtypes & Comparative features

Brachio-Cervical Inflammatory (BCIM)
Collagen vascular disease
Complement: C2 deficiency
Drug-induced
Graft-vs-host disease
HMGCR (200/100) antibody
Idiopathic
IMPP
MAS antibody
Nup antibody
Perimysial pathology (IMPP)
- Jo-1
- OJ
- t-RNA synthetase antibodies

PM + Mitochondrial (PM/COX-)
Regional ischemic (RIIM)
Sarcoidosis
Signal recognition particle Ab (SRP)
Systemic disorders & Myositis
U1-snRNP antibodies

Dermatomyopathies
Childhood (Juvenile)
Adult
- Drug-induced
- Malignancy-associated
  - p155 (TIF-γ) Ab
  - Regional ischemic (RIIM)
- MDA-5 antibody
- PM-Scl antibody +
- Amyopathic

Mi-2 antibody +
Perimysial pathology (IMPP)
- EJ antibody
- Jo-1 antibody

IM-VAMP
Inclusion body myositis (IBM)
IM-Mito (PM/COX-)

Other "Immune Myopathies"
Benign acute childhood myositis
Celiac disease
Chondroitin sulfate C deficient
Decorin antibody (BJ) myopathy
Eosinophilia myalgia syndrome
Fasciitis
Focal myositis
- Quadriceps; Other
- TRAPS

Granulomatous
Hemophagocytic lymphohistiocytosis
Hereditary
IM + abundant Macrophages
Infection
- Influenza
- Lyme myositis

Lupus
Macrophagic myofasciitis
Masticator myopathy
Mitochondrial antibody
Multinodular polymyositis
Myasthenia gravis
- Brachio-Cervial Inflammatory (BCIM)
- Myositis
- Lymphorrhages

Necrosis, Abundant
- with Encephalopathy
- Regional Ischemic Immune (RIIM)
- with Pipestem capillaries
- SRP antibodies

Neonatal perifascicular myopathy
Orbital
Perimyositis
Polymyalgia Rheumatica
Pyomyositis
Sarcocystis
Sarcoidosis
Toxoplasmosis
Trichinellosis (Trichinosis)

PATHOLOGIC CLASSIFICATION [68]

Perimysial pathology (IMPP)
Feature: Connective tissue pathology
- Aldolase high
- Dermatomyopathy with IMPP
  - Amyopathic (MDA5 Ab)
- Fasciitis
- Focal myositis
- Graft-vs-host disease
- t-RNA synthetase antibodies: Jo-1

IMPP + Necrosis
- HMGCR (200/100) antibody

Myovasculopathies
Feature: Damage to large or small vessels
- Dermatomyositis with Vascular Pathology (DM-VP; Childhood Dermatomyositis)
- Regional Ischemic (RIIM; Paraneoplastic)
- Pipestem capillaries

Immune Polymyopathies
Features: Necrosis; Little inflammation
- SRP antibody: Scattered necrosis

IM + Endomysial Pathology (IM-EP)
Features: C_{5b9} deposits; Glycoprotein Δ
- Brachio-Cervical Inflammatory (BCIM)
- Chondroitin sulfate C deficient
- Decorin antibody (BJ) myopathy

Histiocytic Myopathies
Histiocytic cells: Foci or Predominant
- Granulomatous
  - Mitochondrial antibody
  - Sarcoid
- Hemophagocytic lymphohistiocytosis
- IM + Abundant Macrophages (IMAM)
- IRIS
- Macrophagic myofasciitis

IM-VAMP syndromes
Features: Foci of T-cells but not B-cells
Vacuoles, **A**ggregates or **M**ito **P**ath
No response to immunomodulation
- Inclusion body myositis
- IM + Mitochondrial Pathology

https://neuromuscular.wustl.edu/mother/myotox.htm

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMT	**Muscle**	Specific condition

Myopathy

Inflammatory myopathies

Arq Neuropsiquiatr. 2022 May; 80(5 Suppl 1): 238–248.
Published online 2022 Aug 12. doi: 10.1590/0004-282X-ANP-2022-S131

Inflammatory myopathies: an update for neurologists

André Macedo Serafim Silva, [1] Eliene Dutra Campos, [1] and Edmar Zanoteli [1]

Table 2.

Classification e key characteristics of the IIM subtypes.

DM	- Inflammatory myopathy accompanied by skin changes. - Some patients may have amyopathic or hypo myopathic presentations - There are five known autoantibodies associated: anti-Mi2, anti-TIF1-y, anti-NXP-2, anti-MDA-5 and anti-SAE - CD4 lymphocytes infiltrates, with a perivascular and interfascicular location and atrophy of the perifascicular fibers
PM	- No skin or pulmonary involvement - Good response to immunosuppressive treatment - No association with specific antibodies - Now considered an exclusion diagnosis - CD8 lymphocytes predominate, invading the endomysium and intact fibers
IMNM	- Associated with systemic conditions (cancer, statin, viral infections) - Presence of autoantibodies: anti-SRP and anti-HMGCR - In children, it may present as slowly progressive, and mimic muscular dystrophy - Abundant fibers in necrosis and macrophage predominance, which can be identified by labeling for CD68
IBM	- Slowly evolving weakness with distal atrophy in the hands and atrophy in the thighs - Individuals over 45 years - Association with anti-NT5C1A autoantibody - CD8 lymphocytes predominate, invading the endomysium and intact fibers and presence of marginated vacuoles
ASS	- Inflammatory myopathy, interstitial lung disease and joint involvement - Other findings: fever, "mechanic's hands" and Raynaud's phenomenon - All the patients have antibodies directed against aminoacyl-tRNA synthetases - The most common autoantibodies are anti-Jo-1, anti-PL-7, and anti-PL-12 - The muscle biopsy demonstrates T-cell and macrophage infiltrations and perifascicular atrophy and necrosis
OM	- Association of inflammatory myopathy with other connective tissue disorder - The most common antibodies are anti-PM /Scl and anti-U1-RNP - Perivascular inflammation, perifascicular necrosis and MHC-I increase

Open in a separate window

DM: dermatomyositis; PM: polymyositis'; IMNM: immune-mediated necrotizing myopathy; IBM: inclusion body myositis; ASS: antisynthetase syndrome; OM: overlap myositis.

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMT	**Muscle**	Specific condition

Toxic Myopathy

e-Aminocaproic acid	Daptomycin	Penicillamine
Amiodarone	20,25-Diazacholesterol	Pentaborane
Apamin	Emetine	Procainamide
Arsenic trioxide	Ethanol	Propofol
AZT	Fibrates	5-α Reductase
Barium	Gemcitabine	Selumetinib
Chlorophenoxy	Germanium	Statins
Chloroquine	Gold	Taipoxin
Ciguatoxin	Gossypol	TNF-α
Clofibrate	Interferon-α	Toxic oil
Colchicine	Ipecac	L-Tryptophan
Corticosteroids	Isotretinoin	Valproate
Crotamine	Lithium	Vecuronium bromide
Crotoxin	Minocycline	Vinca alkaloids
Cyclosporine	Mojave toxin	Zidovudine

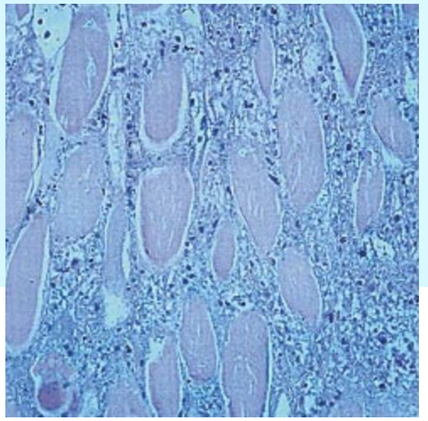
Review > QJM. 2018 May 1;111(5):307-311. doi: 10.1093/qjmed/hcy031.

Acute steroid myopathy: a highly overlooked entity

https://neuromuscular.wustl.edu/mother/myotox.htm

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMT	**Muscle**	Specific condition

Rhabdomyolysis



Medications and Toxic Substances That Increase the Risk of Rhabdomyolysis

Direct myotoxicity

HMG-CoA reductase inhibitors, especially in combination with fibrate-derived lipid-lowering agents such as niacin (nicotinic acid; Nicolar)
Cyclosporine (Sandimmune)
Itraconazole (Sporanox)
Erythromycin
Colchicine
Zidovudine (Retrovir)
Corticosteroids

Indirect muscle damage

Alcohol
Central nervous system depressants
Cocaine
Amphetamine
Ecstasy (MDMA)
LSD
Neuromuscular blocking agents

HMG-CoA = 3-hydroxy-3-methylglutaryl coenzyme A; LSD = lysergic acid diethylamide; MDMA = 3,4-methylene dioxymethamphetamine.

Am Fam Physician. 2002;65(5):907-913

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMJ	**Muscle**	Specific condition

Rhabdomyolysis

Critical Care 2005, **9**:158-169 (DOI 10.1186/cc2978)

Drugs that may induce rhabdomyolysis

Antipsychotics and antidepressants	Drugs of addiction
Amitriptyline	Heroin
Amoxapine	Cocaine
Doxepine	Amphetamine
Fluoxetine	Methadone
Fluphenazine	D-lysergic acid diethylamide (LSD)
Haloperidol	Antihistamines
Lithium	Diphenhydramine
Protriptyline	Doxylamine
Phenelzine	Other drugs
Perphenazine	Alcohol
Promethazine	Amphotericin B
Chlorpromazine	Azathioprine
Loxapine	Butyrophenones
Promazine	Emetics
Trifluoperazine	Epsilon-aminocaproic acid
Sedative hypnotics	Halothane
Benzodiazepines	Laxatives
Diazepam	Moxalactam
Nitrazepam	Narcotics
Flunitrazepam	Oxprenolol
Lorazepam	Paracetamol
Triazolam	Penicillamine
Barbiturates	Pentamidine
Gluthetimide	Phencyclidine
Antilipemic agents	Phenylpropanolamine
Lovastatin	Quinidine
Pravastatin	Salicylates
Simvastatin	Strychnine
Bezafibrate	Succinylcholine
Clozafibrate	Theophyline
Ciprofibate	Terbutaline
Clofibrate	Thiazides
	Vasopressin

TABLE 3

Enlarge Print

Infectious, Inflammatory, Metabolic and Endocrinologic Causes of Rhabdomyolysis

Infectious causes

Viruses: influenza virus B, parainfluenza virus, adenovirus, coxsackievirus, echovirus, herpes simplex virus, cytomegalovirus, Epstein-Barr virus, human immunodeficiency virus

Bacteria: Streptococcus, Salmonella, Legionella, Staphyloccus and Listeria species

Inflammatory causes

Polymyositis

Dermatomyositis

Capillary leak syndrome

Snake bites (mostly in South America, Asia and Africa)

Metabolic and endocrinologic causes

Electrolyte imbalances: hyponatremia, hypernatremia, hypokalemia, hypophosphatemia, hypocalcemia

Hypothyroidism

Thyrotoxicosis

Diabetic ketoacidosis

Nonketotic hyperosmolar syndrome

Genetic Causes of Rhabdomyolysis

Lipid metabolism

Carnitine palmitoyltransferase deficiency

Carnitine deficiency

Short-chain and long-chain acyl-coenzyme A dehydrogenase deficiency

Carbohydrate metabolism

Myophosphorylase deficiency (McArdle's disease)

Phosphorylase kinase deficiency

Phosphofructokinase deficiency

Phosphoglycerate mutase deficiency

Lactate dehydrogenase deficiency (characteristic elevation of creatine kinase level with normal lactate dehydrogenase level)

Purine metabolism

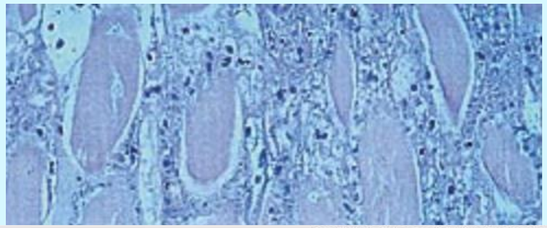
Myoadenylate deaminase deficiency

Duchenne's muscular dystrophy

Am Fam Physician. 2002;65(5):907-913

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMJ	**Muscle**	Specific condition

Rhabdomyolysis



Drugs, toxins, and venoms	Ethanol	
	Recreational drugs and stimulants	Use of heroin, lysergic acid diethylamide, cocaine, *N*-methyl-D-asparate (ecstasy), phencyclidine, caffeine, aminophyline, pseudoephedrine; sniffing glue
	Toxic plants and animals	Ingestion of hemlock, toxic mushrooms; effects of blowpipe dart poison, snake venoms, hymenoptera stings, envenomation by giant desert centipede
	Pharmaceutical agents	Use of benzodiazepines, corticosteroids, narcotic analgesics, immunosuppressants, salicylates, lipid-lowering statins, paralytics, antibiotics, antidepressants, antipsychotics, thromobolytics, chemotherapeutic agents

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMT	Muscle	**Specific condition**

Other Causes

Acute sensory loss

Intensive Care conditions

Musculoskeletal pain

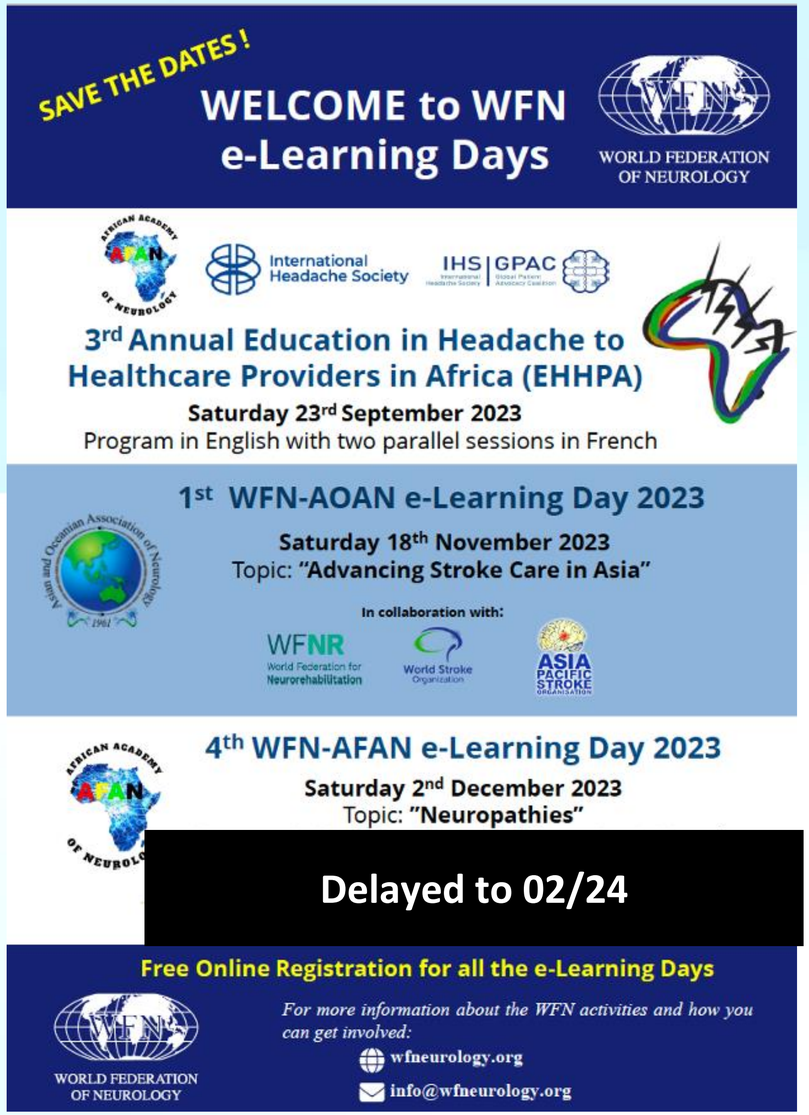
Insulin „Neuritis“

Rabies (paralytic form)

Tetanus

Exacerbation of chronic neuro-muscular disease

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMT	Muscle	Specific condition



Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMT	Muscle	**Specific condition**

2024

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMT	Muscle	**Specific condition**

2025

Central, Pons, Medulla, Spinal cord	Nerve Roots DRG	Peripheral Nerves	NMT	Muscle	**Specific condition**

2026

2026

ICNMD

Vancouver (Canada)

Copenhagen (Denmark)

Florence (Italy)

?

2027

Questions :	General : Wolfgang Grisold grisoldw@gmail.com	Immune Neuropathies and ICI: Anna Grisold anna.grisold@gmail.com	Nerve Ultrasound: Stefan Meng stefan.meng@meduniwien.ac.at
	Please specify precise question, please use English	Specific question for ICI	Nerve ultrasound
	You can add an image or video		
EMG Reindorfgasse, Plankensteiner Neurologie Seminare, Otto Knaus Foundation, JD Widdicombe			

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