

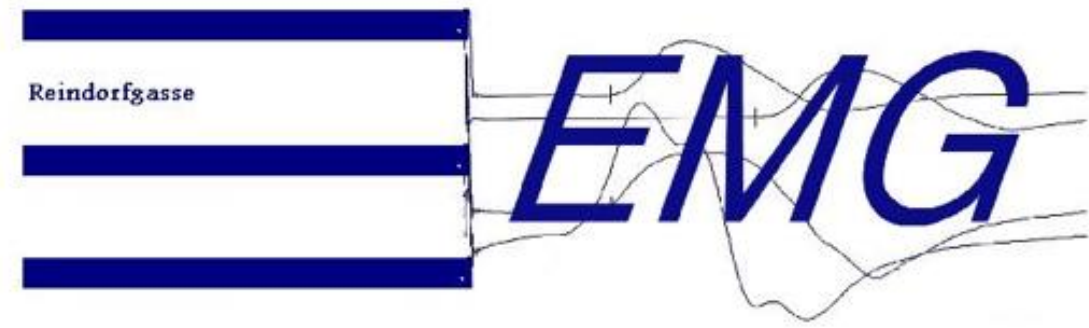
PART 2

CN

- V
- VII
- V
- XII



Horner Syndrome



Neuromuscular Disease

Clinical presentations, signs and symptoms

**Lecture presented at the EAN Teaching Course in
Burkinje Faso, Nov. 2017**

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COI

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- Ludwig Boltzmann Institute for Experimental und Clinical Traumatology, Vienna, Austria

Is it neuromuscular ?

Combinations ?

Case history

Clinical presentation

new concepts

examples

Neuromuscular System (A common construct)

- (Extracranial parts of the Cranial nerves)
- Nerve Roots
- Plexus
- Peripheral nerves
- Neuromuscular junction
- Muscle
- Autonomic system

Neuropathies: How often ?

Median (CTS)	103,00
Ulnar	25,00
Radial	2,50
Cut. Femoral	10,00

Disc (> 50 yrs)	
Cervical (aged)	600,00
Lumbar (aged)	1079,00

PNP DM II: 6400

Pro 100 000 Patient years

Source: Callaghan, Ann Arbor 2016, USA

Combinations of neuromuscular and central nervous system. Examples:

- MND/ALS
- Some hereditary neuropathies
- Myotonic dystrophy
- etc.

Case history



Family history (tree)

Diseases, medications,

Exposition, trauma

**Onset, duration, acute, chronic
progressive , fluctuating,
episodic**

Symptoms:

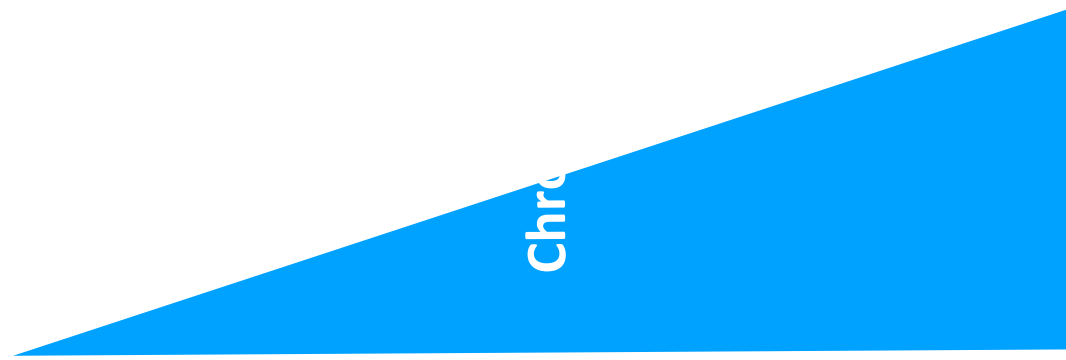
**motor, sensory, autonomic
movement, composite**

Acquired/genetic

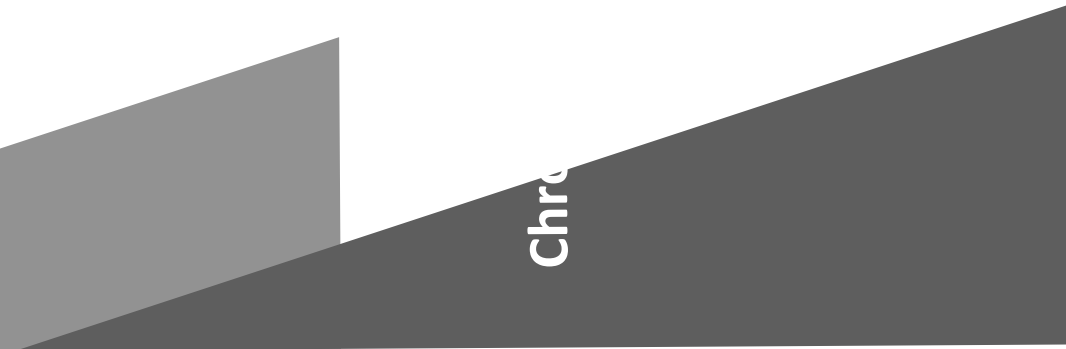
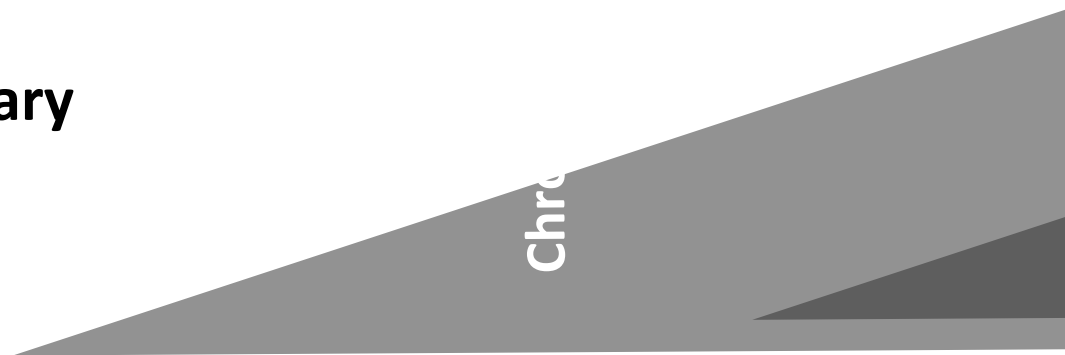
**Before examining the patient-
create hypothesis**

Time

Acquired



Hereditary



Symptoms and signs

Motor

Sensory

Composite function

Autonomic

Neuropathic Pain

Distribution

Symptoms and signs

Skin

Skeletal system

Motor

Distribution of weakness, atrophy

Strength (BMRC)

Time: Fluctuating, episodic

Muscle volume

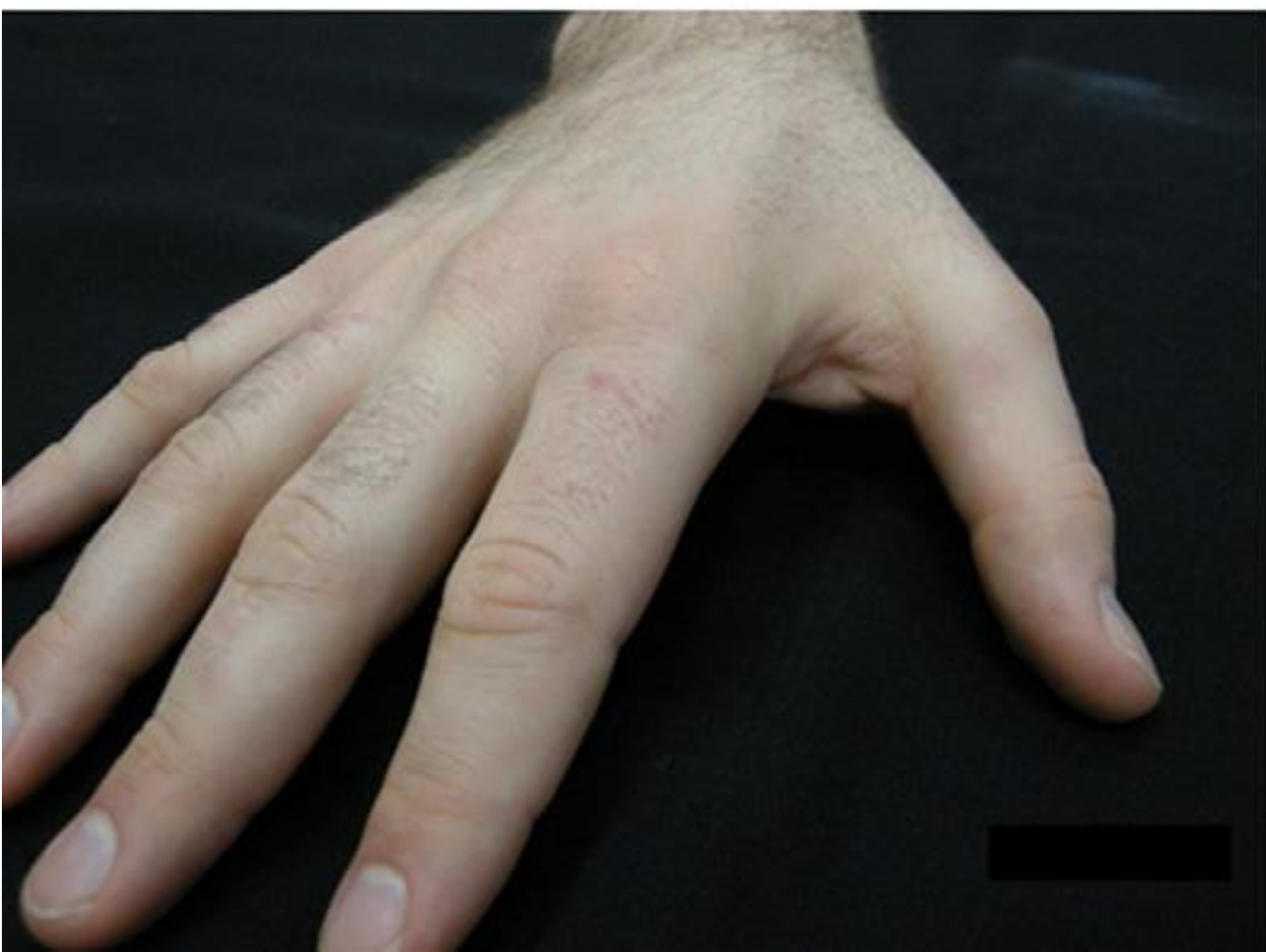
Atrophy

Hypertrophy

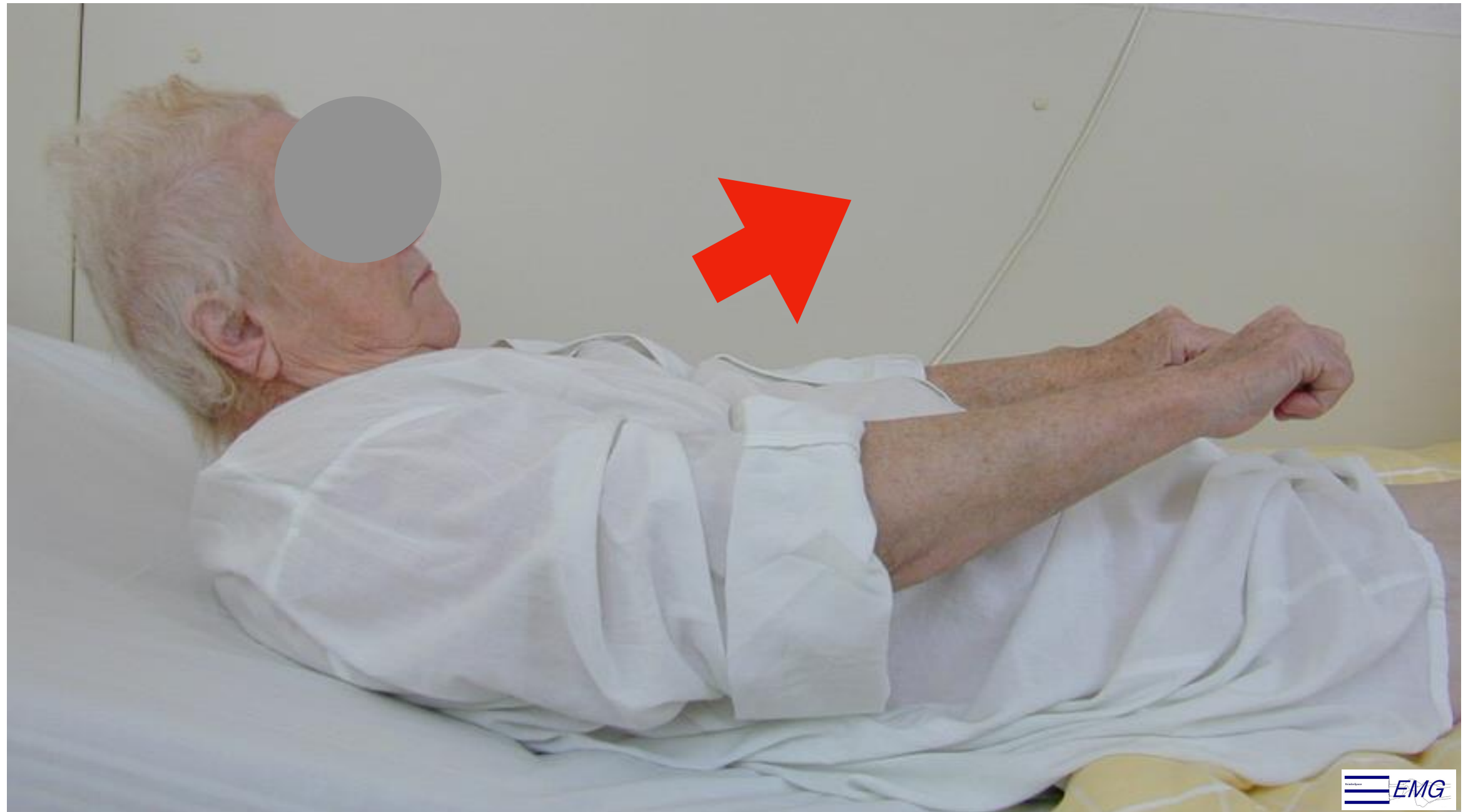
Phenotype distribution: myopathy, polio, club foot

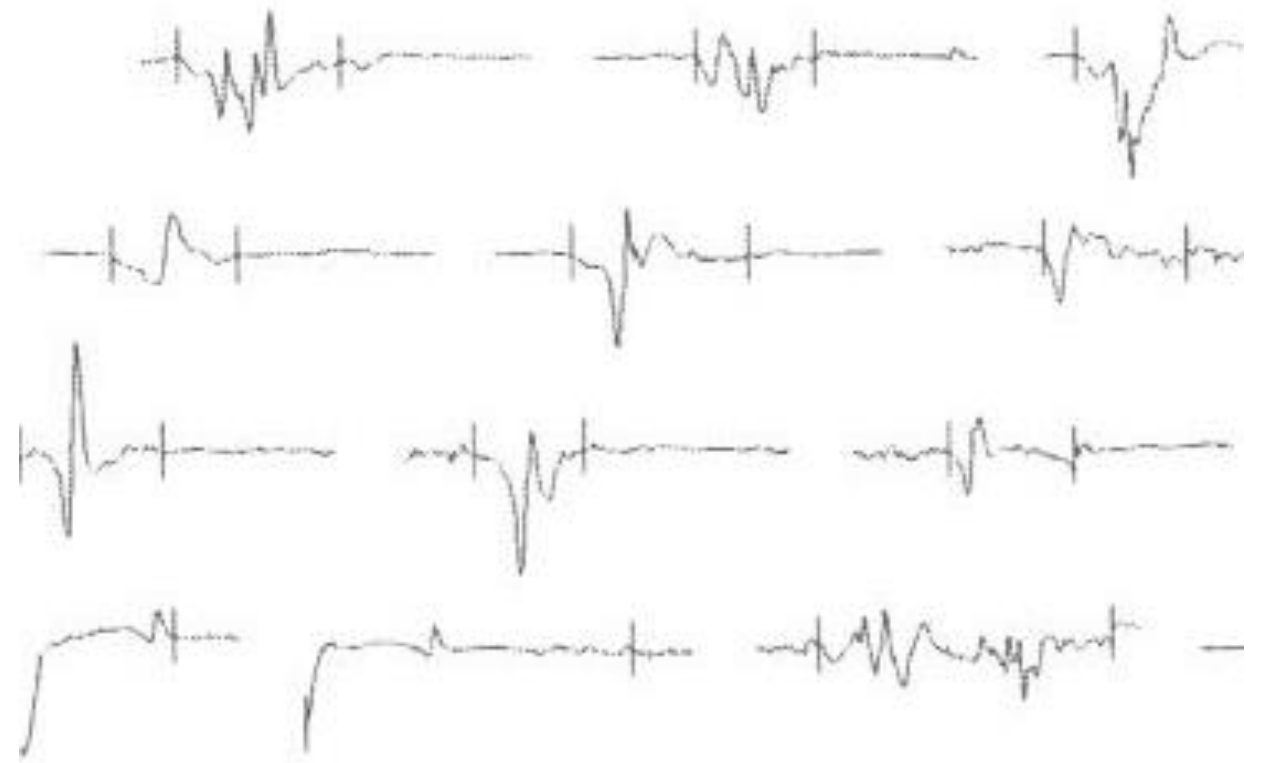
Winging





Proximal weakness





Carcinomatose Neuromyopathie

EMG

Percussion myotonia



Muscle hypertrophy

Examples

Pseudohypertrophy

Endocrinopathy

Focal muscle hypertrophy

Overuse

infections

Amyloid

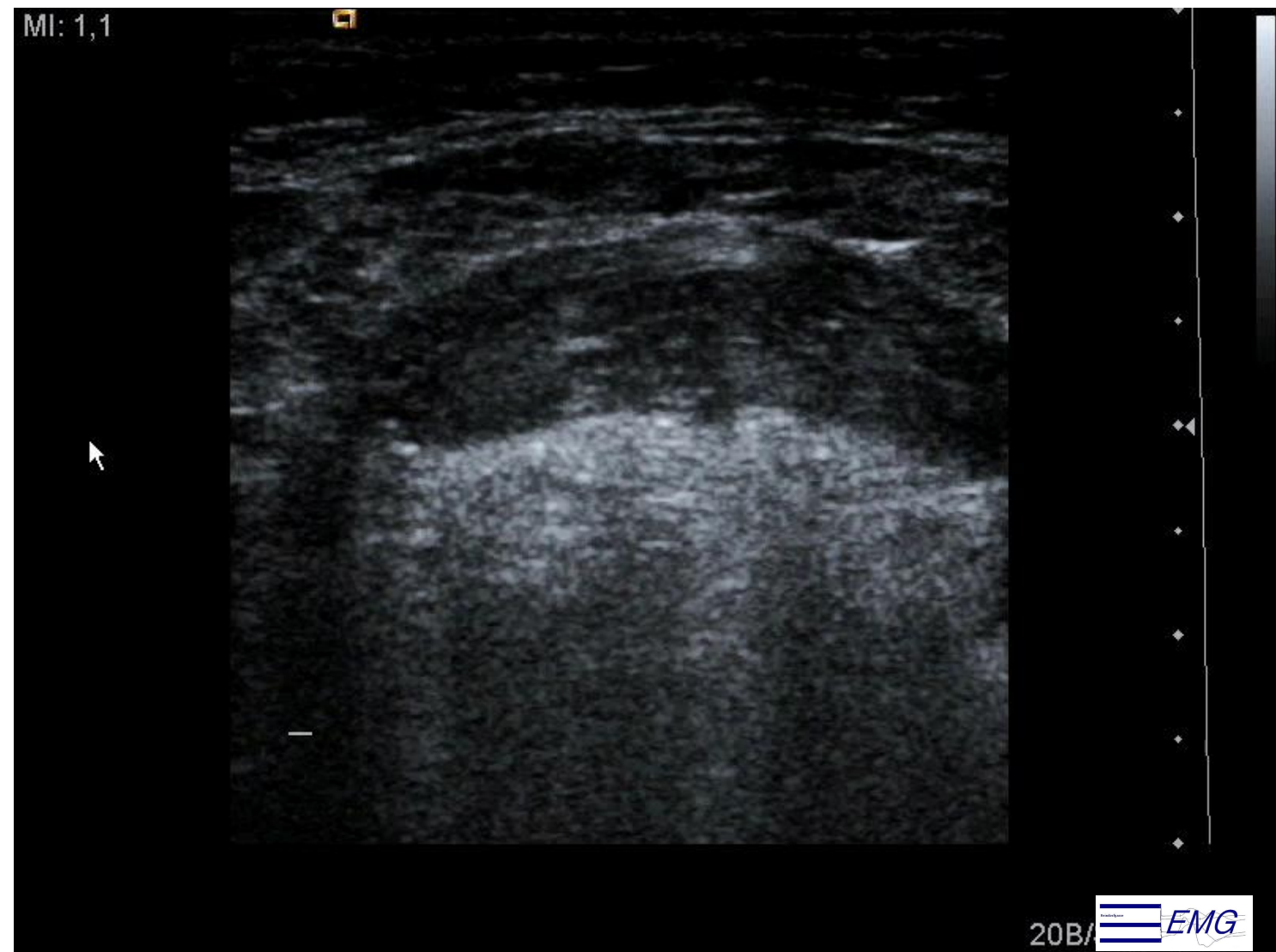


Steroid myopathy



Movement

- Fasciculation
- Myokymia
- Rippling
- „Stiffness“, myotonia
- Percussion myotonia
- Cramps



Tongue Ultrasound

Sensory

Quality

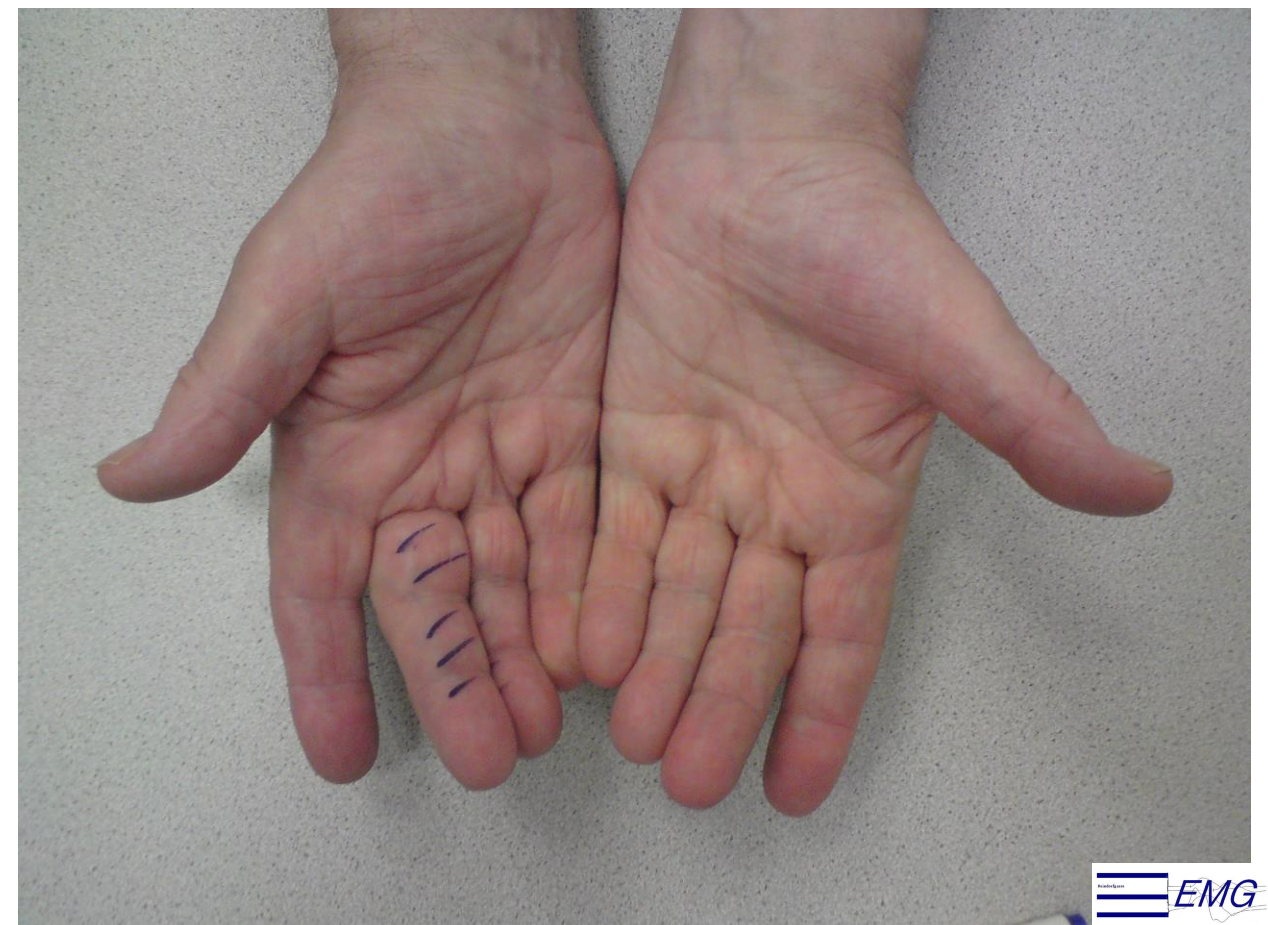
Sensory assessment

Instruments

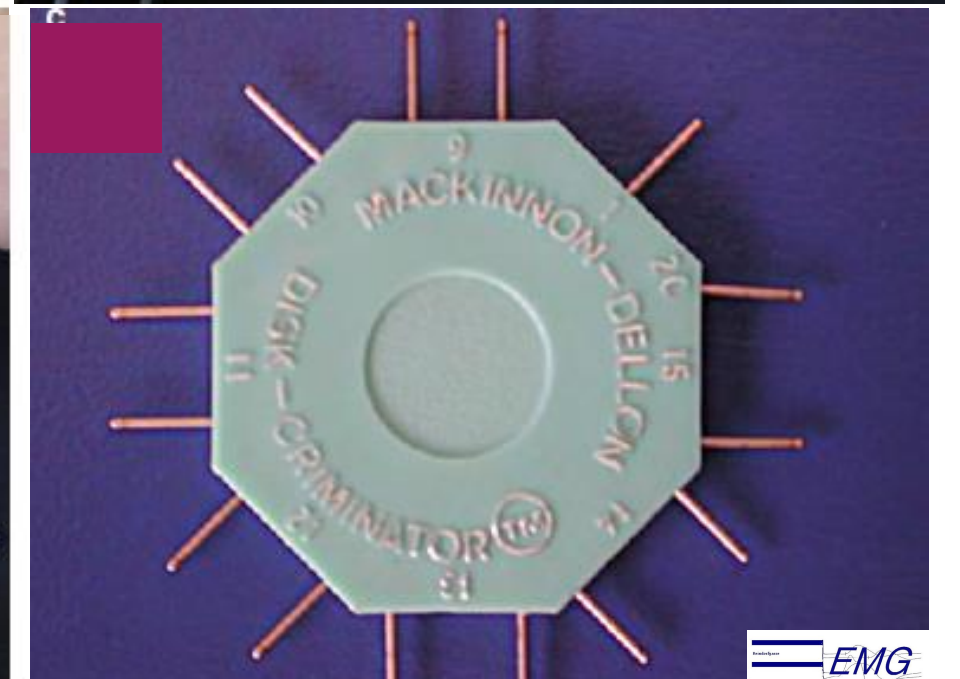
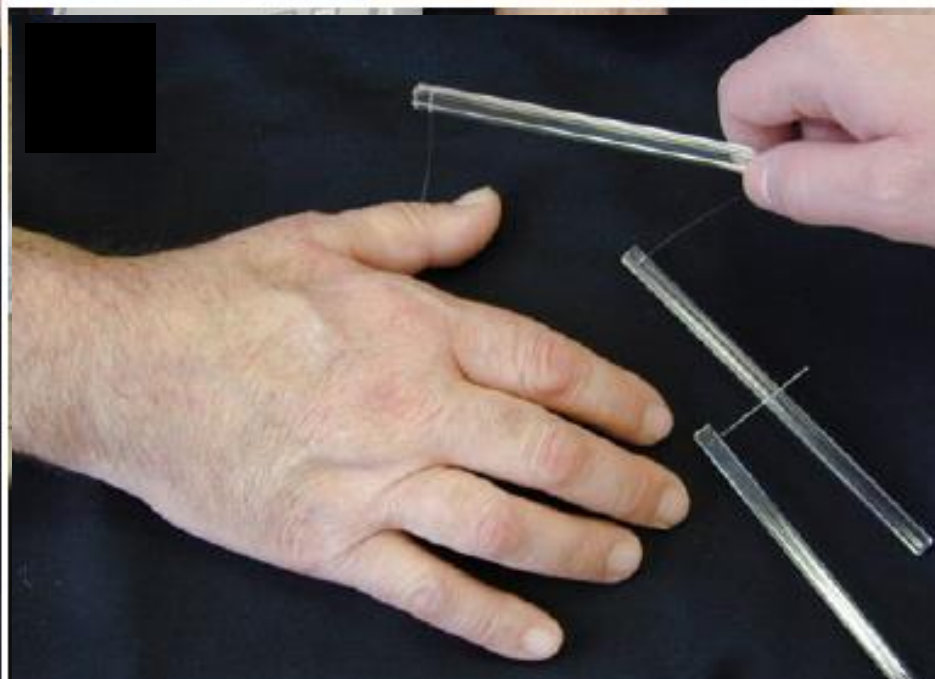
Type of fiber lesion

Tinel Hofmann sign

Sclerotoma,
myotoma,
„neurotoma“



Instruments





A



B

A: superficial peroneal nerve

B. Infrapatellar nerve

Tinel Hofmann: red circle



Foot

Test Composite function !

Simple (but complex) functions:

Opening and closing buttons

Recognition of objects

Balance: Romberg

Stance on one leg

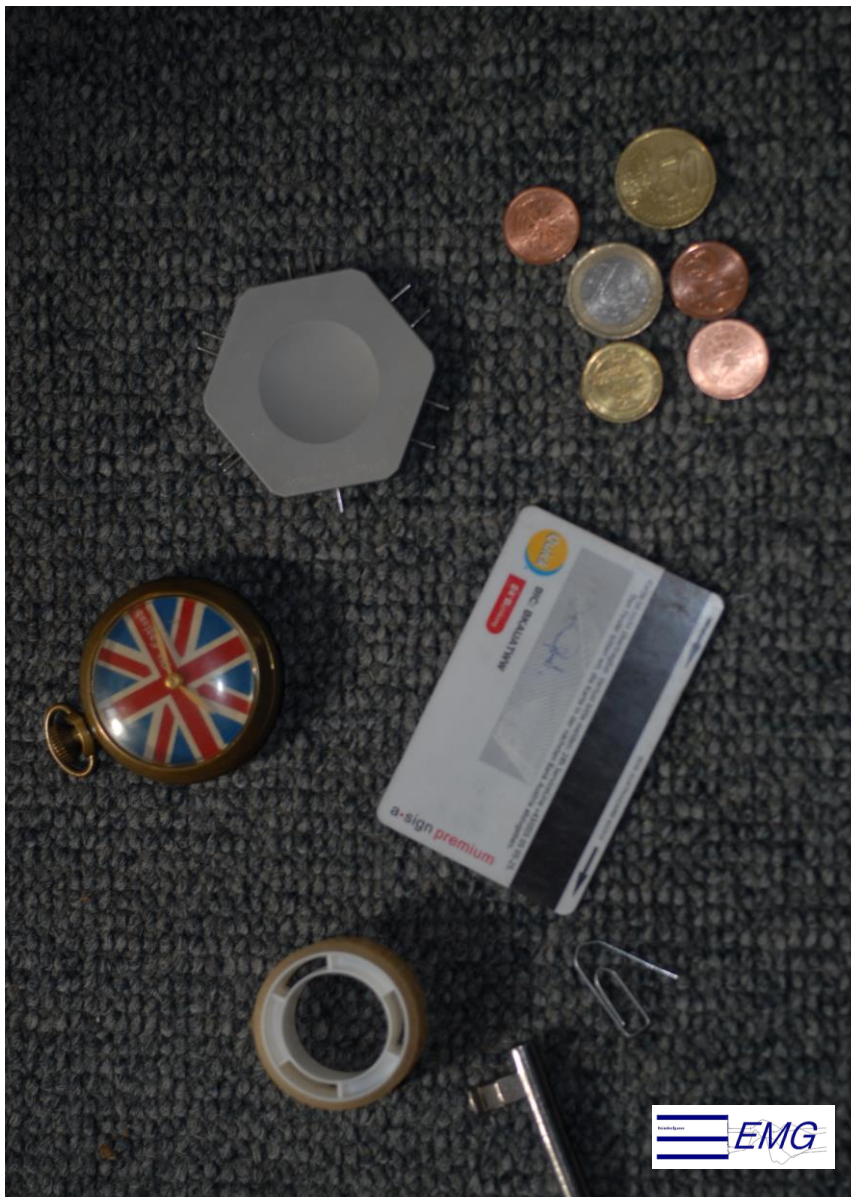
Can you put on your trousers standing
freely one leg ?

Muscle:

„Hands up“

Standing up from squat

Push ups



**Ask for
Autonomic
dysfunction**

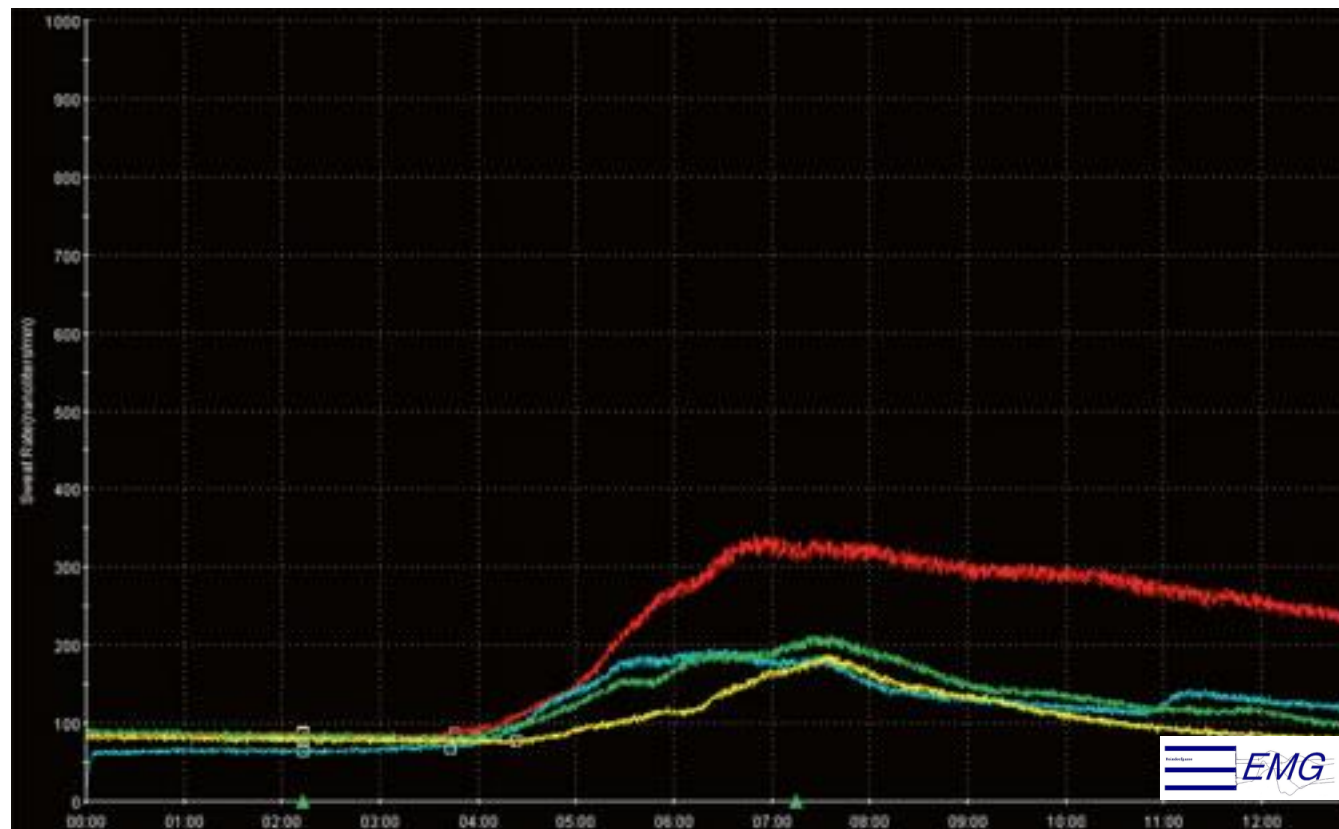
Dry mouth, dry eyes ?

Voiding

Sexual dysfunction

Orthostasis

Gastrointestinal



Neuropathic Pain

Pain characteristics

(stabbing, shooting, electric)

Distribution- cave Allodynia

Sensory, motor involvement

Elicited by : eg Tinel Hofmann

Neuropathic Pain conditions

- Acute episodic
- Painful neuropathy
- Small fiber
- Trauma causalgia
- Localized
- Surgery
- Erythromelalgia
- Phantom- pain
- PRURITUS

Intercostobrachial nerve



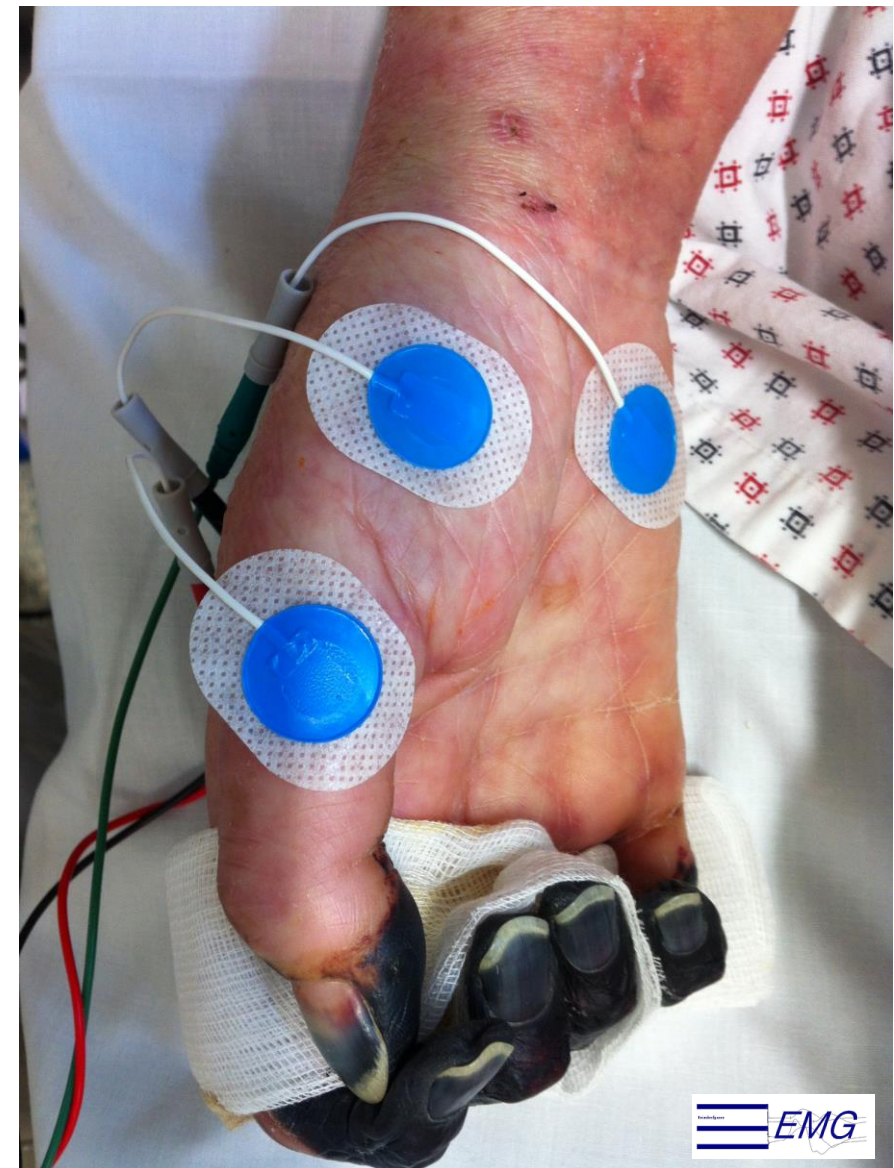
Painful neuropathies

- Alcohol
- Metals
- Drugs (eg. CIPN)
- Immune-mediated, Vasculitis
- Hereditary (eg Fabry)

Skin

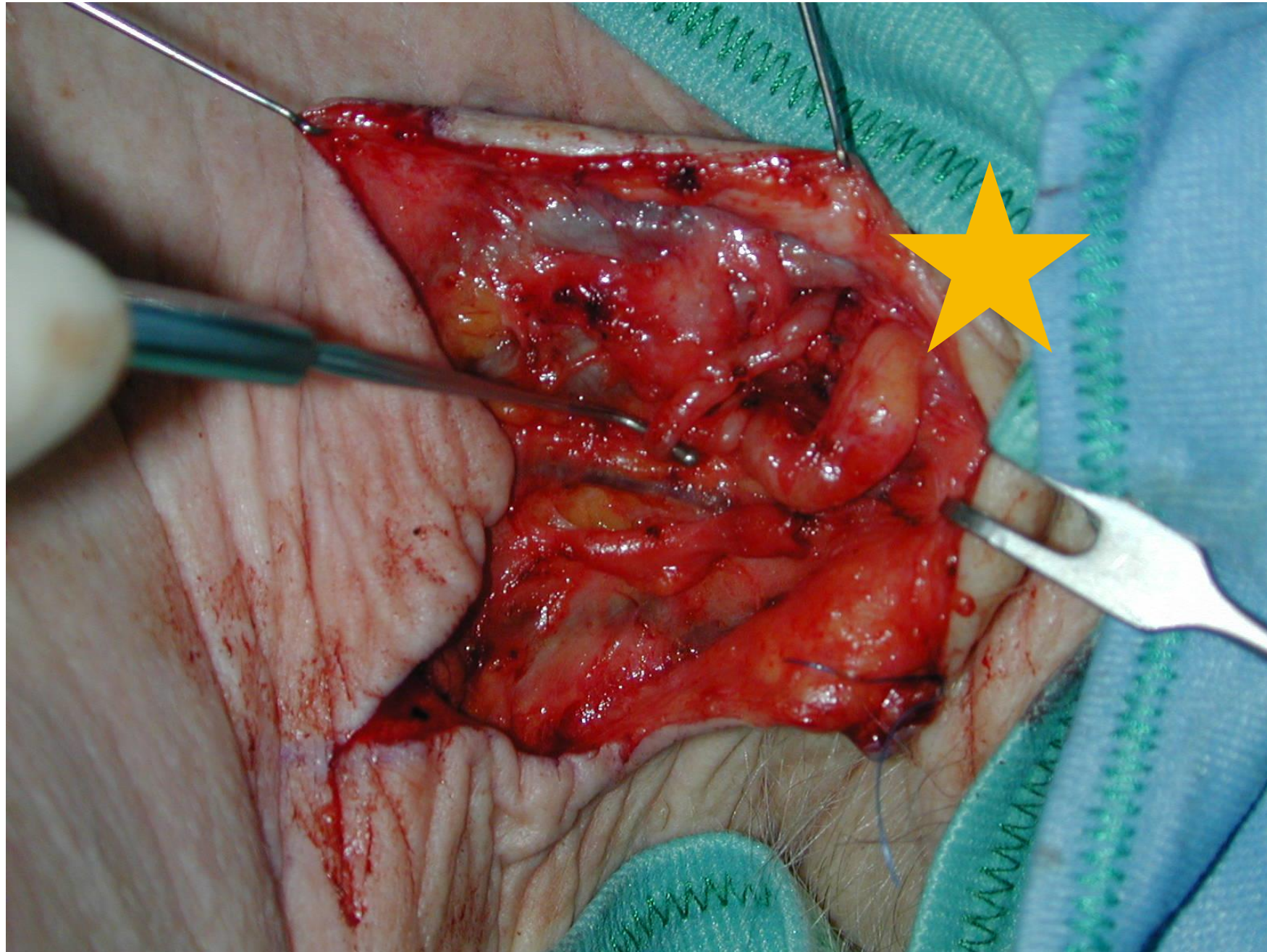


Haemangioma



Waterhouse F. syndrome

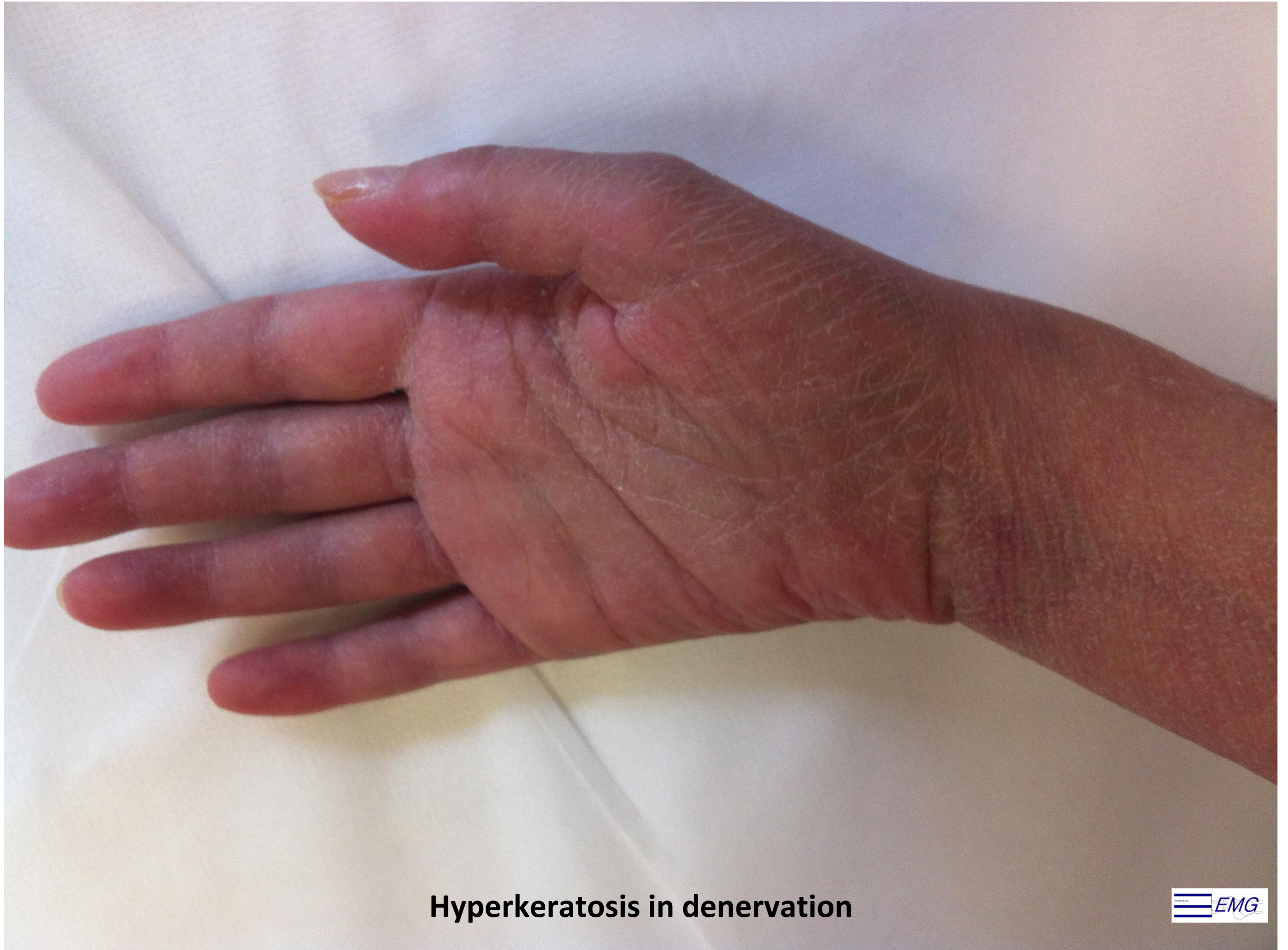
Thickening of nerves



Greater auricular nerve (intraoperative)

Radiofibrosis syndrome





Hyperkeratosis in denervation



Skin



Lymphoma

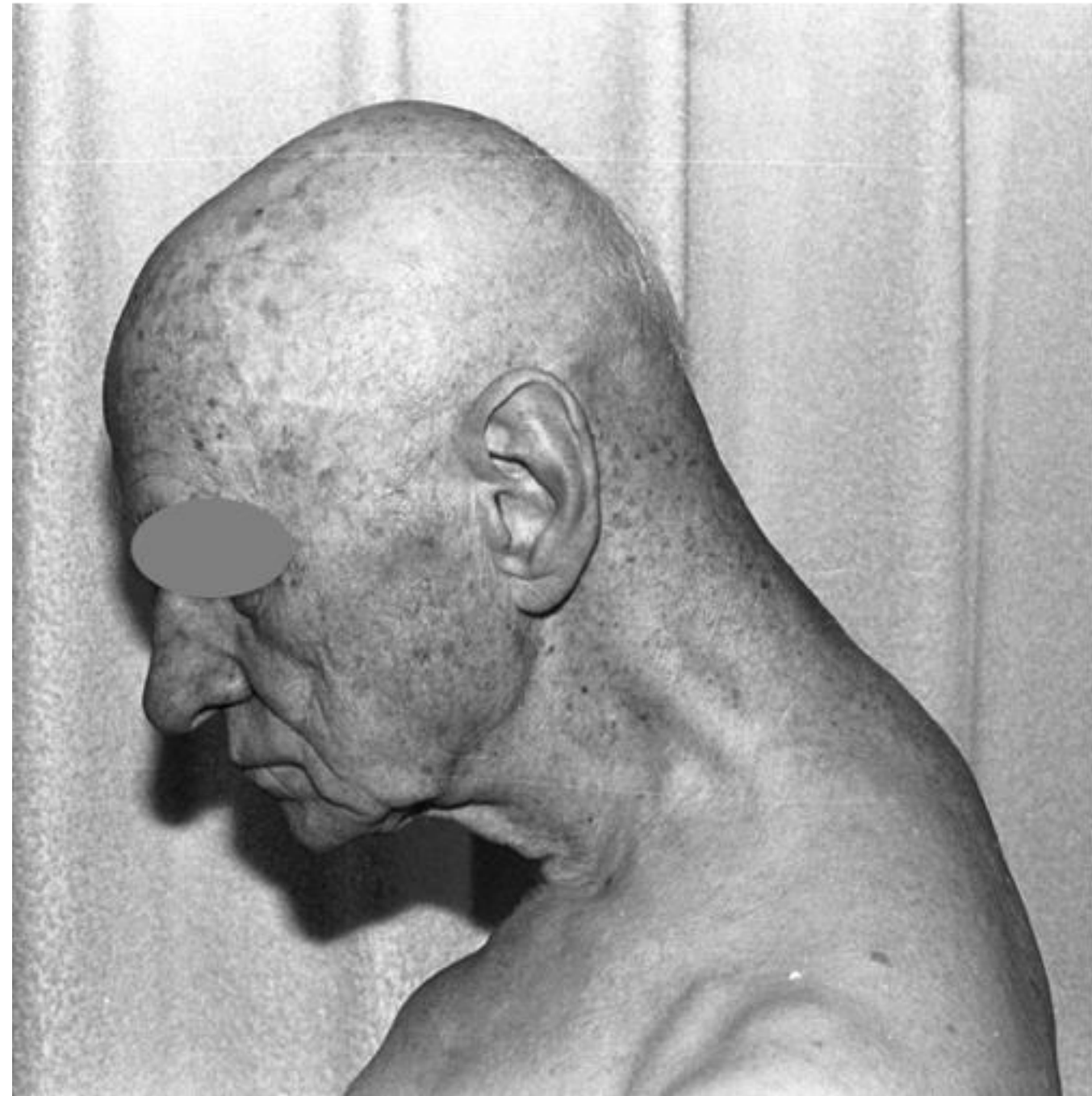
Hand Foot Syndrome



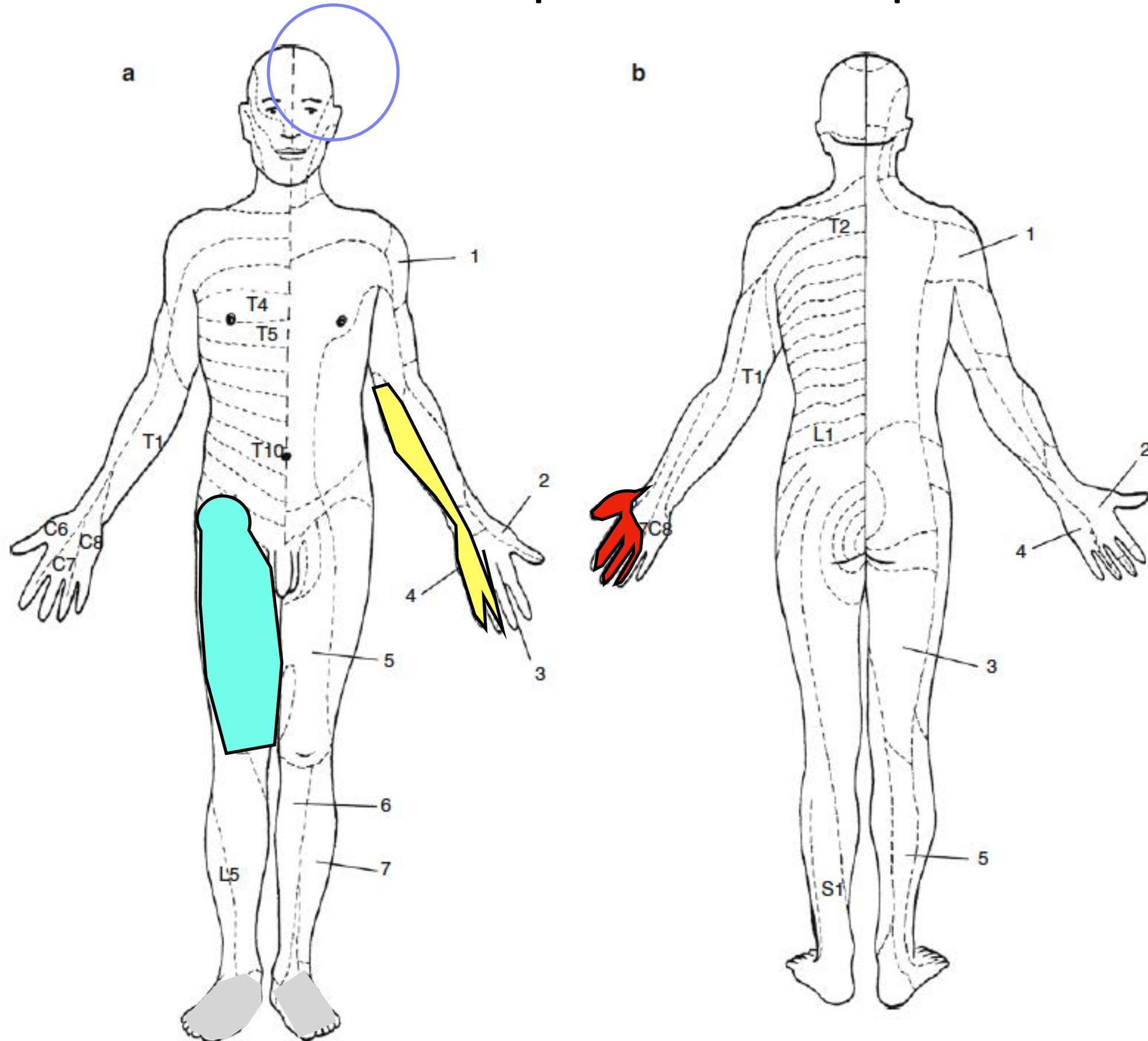
Skeletal system



Focal dysfunction/Weakness



Distribution: Neuropathies and Radiculopathies



Further Ancillary Tests

- NCV/EMG
- Imaging: ultrasound, MRI, CT
- Autonomic test
- Biopsy
- Laboratory
- Genetics

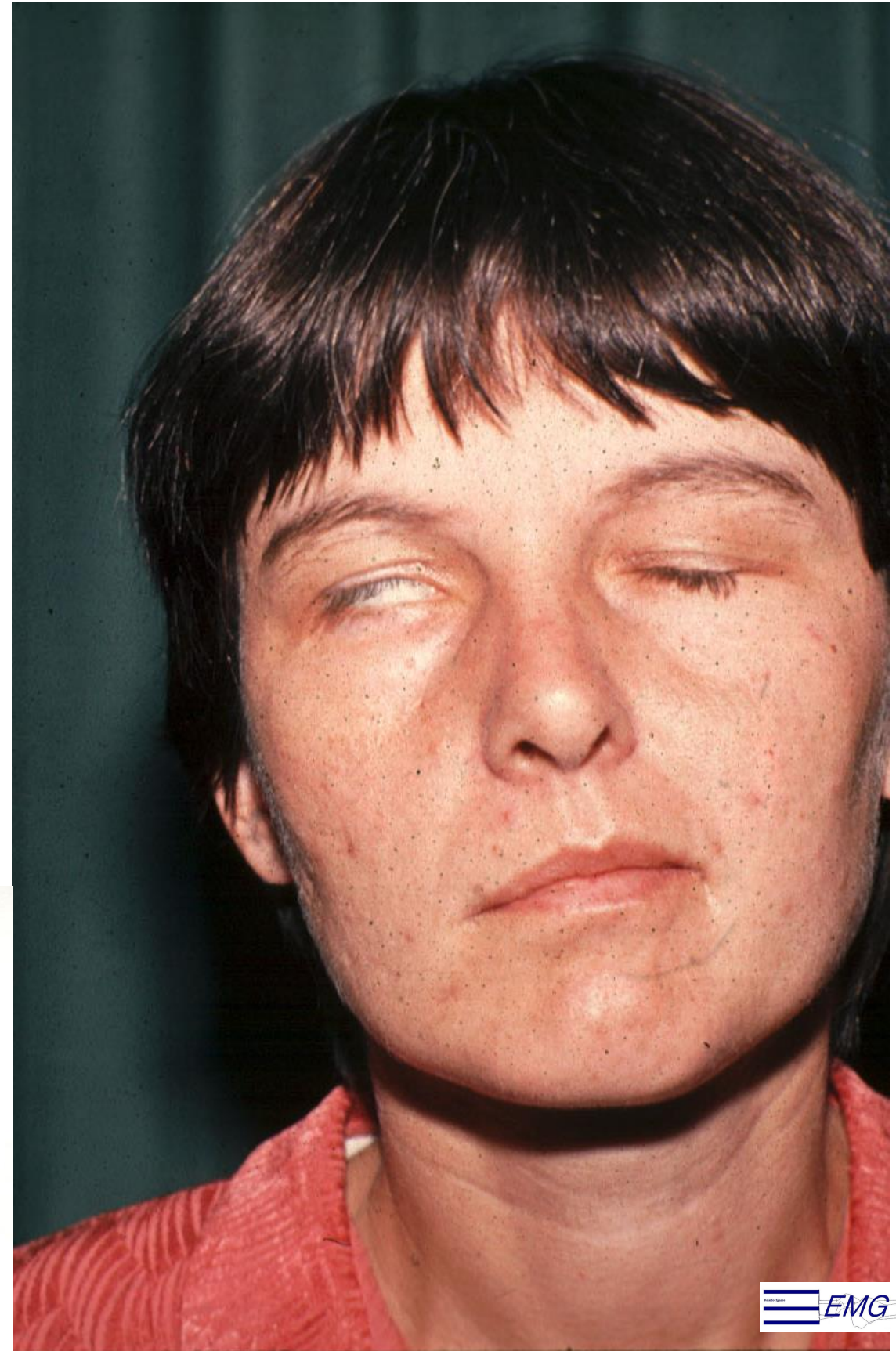
Nerve Ultrasound



Neuromuscular lesions

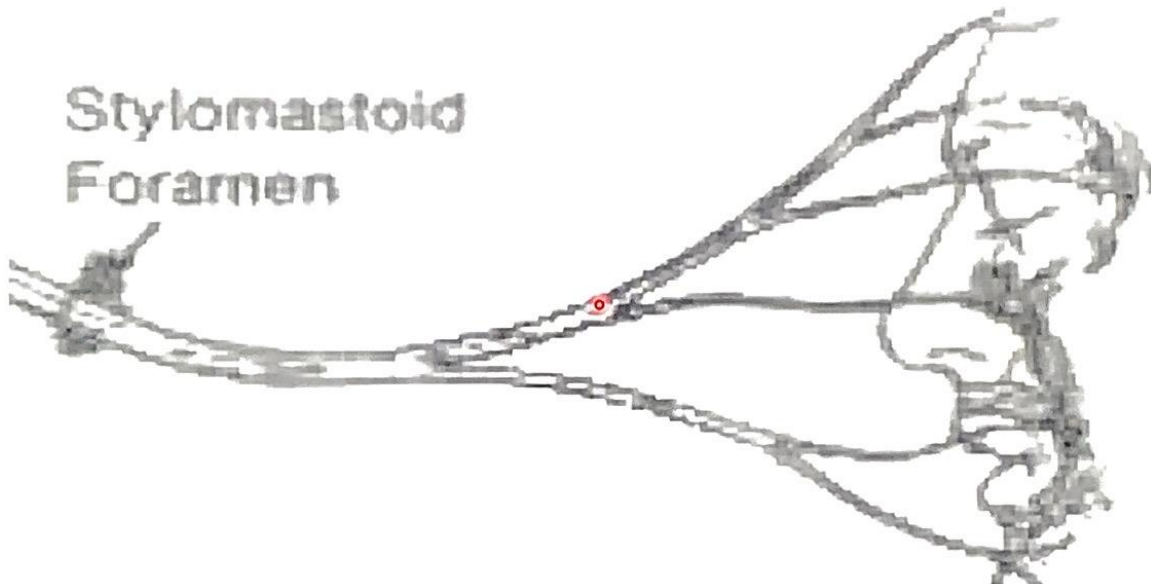
- CN
- Plexus lesions
- Mononeuropathies
- Polyneuropathies
- Neuromuscular transmission
- Muscle

Facial nerve: 3 different problems



Nervus Fazialis VII

Stylomastoid Foramen



Nervus Fazialis VII Topographie

Cases of tongue involvement



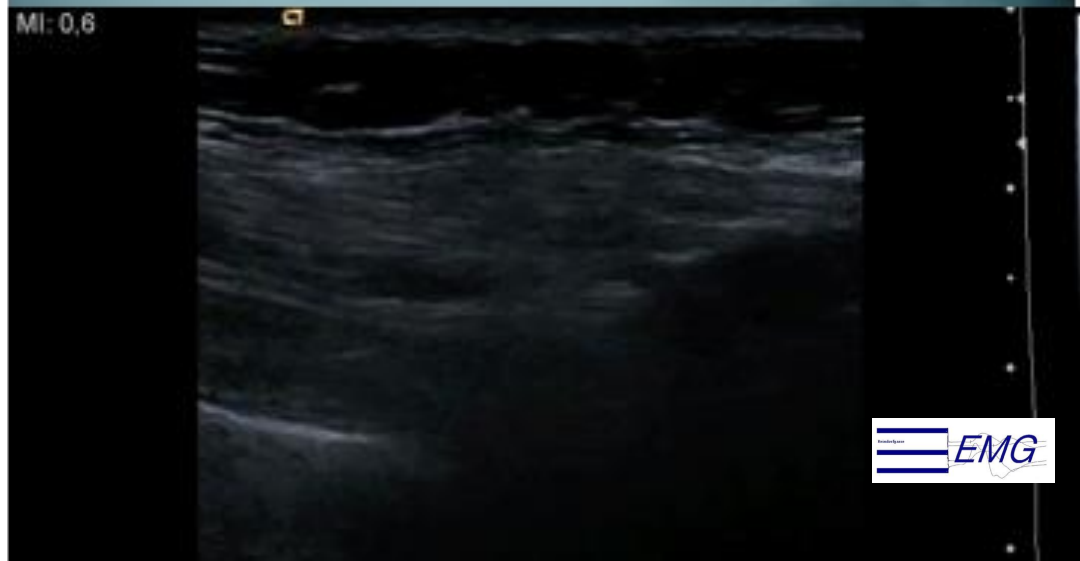
The Plexus

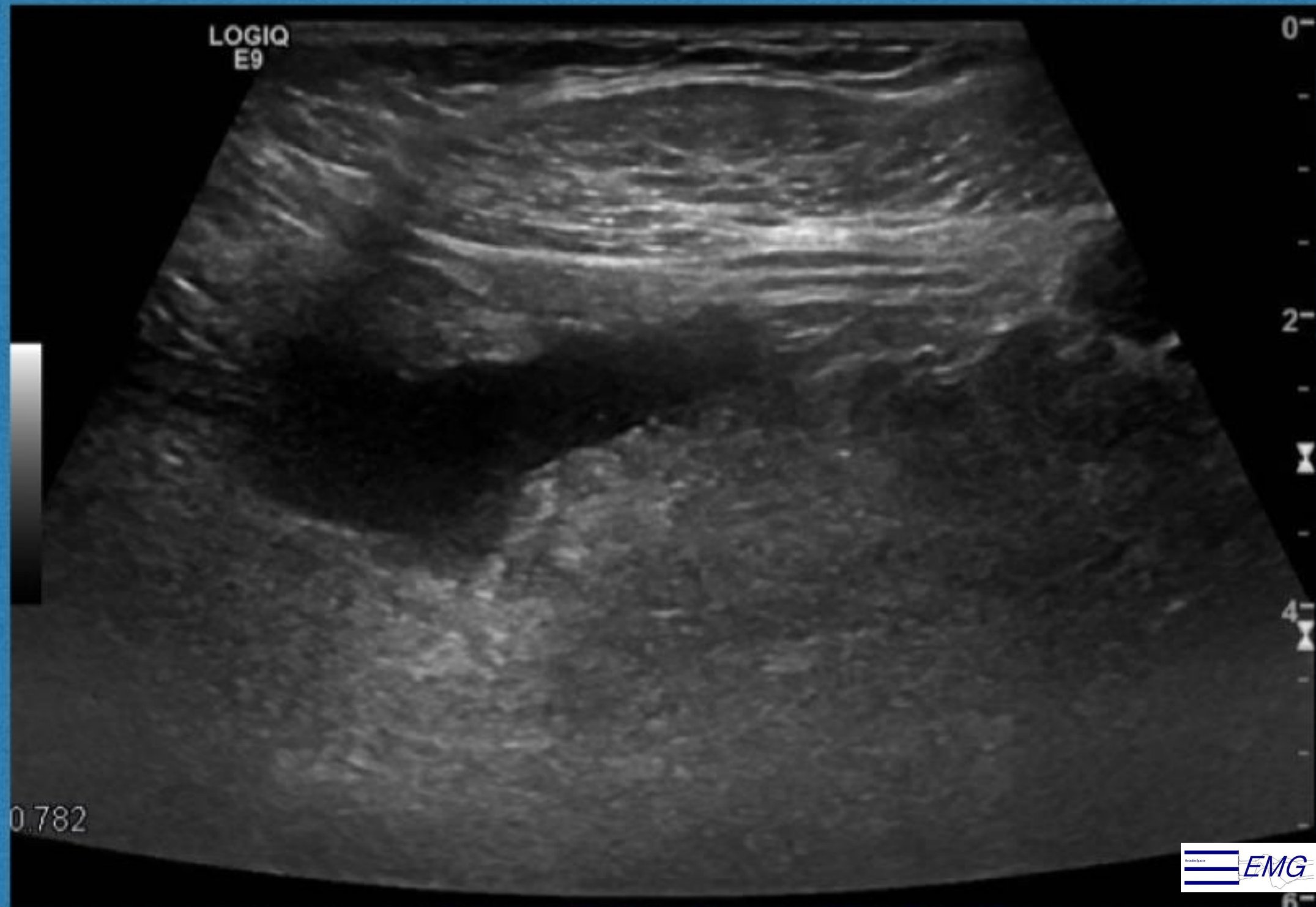
- Cervical
- Brachial
- Lumbar
- Sacral
- (Coccygeal)

4 Plexus cervikalis

Phrenic nerve palsy

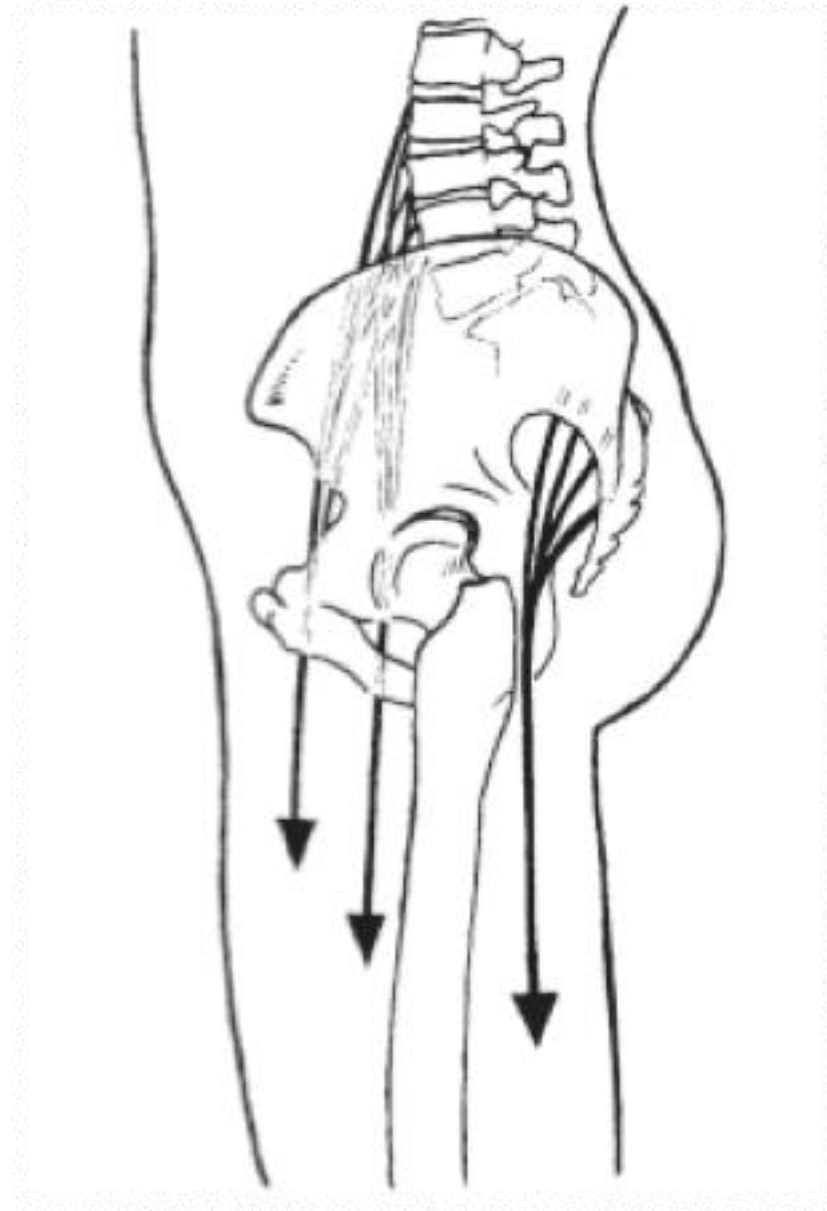
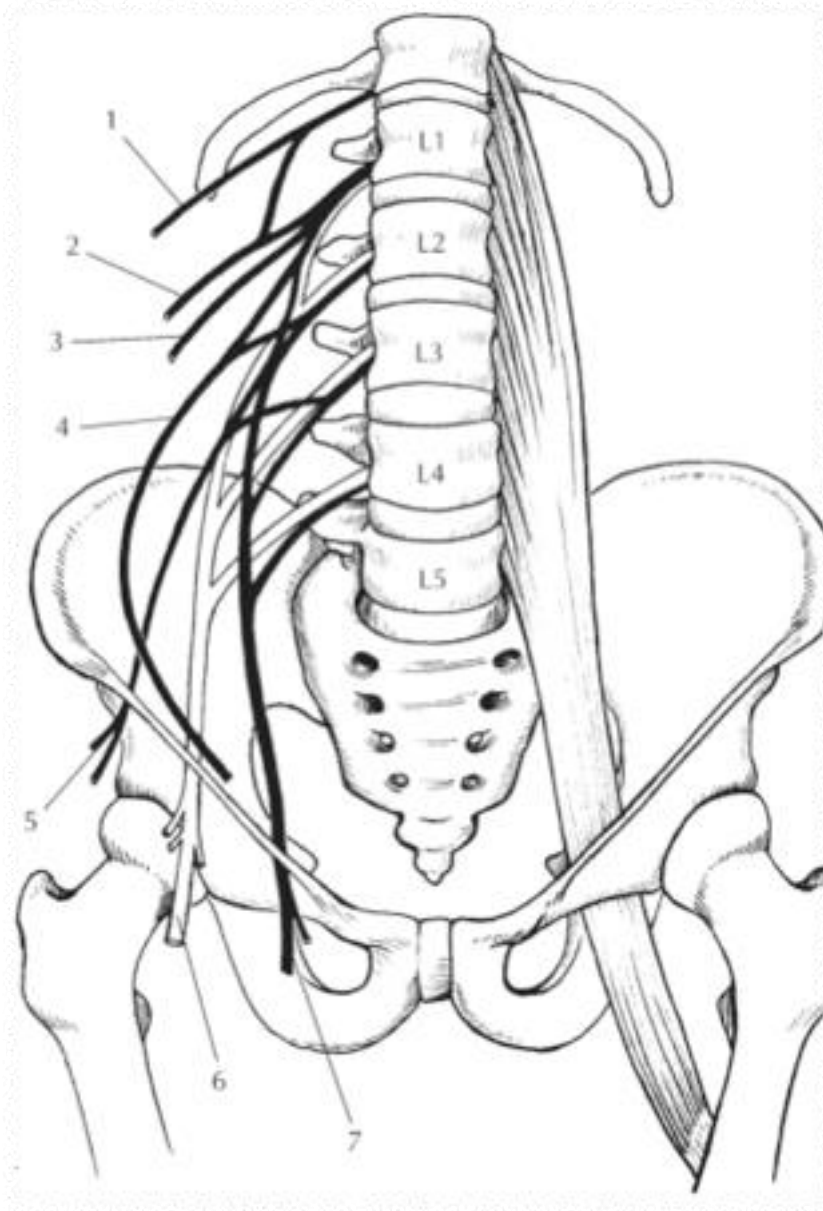
Local tumor affecting
the cervical plexus





Seroma compression of the brachial plexus

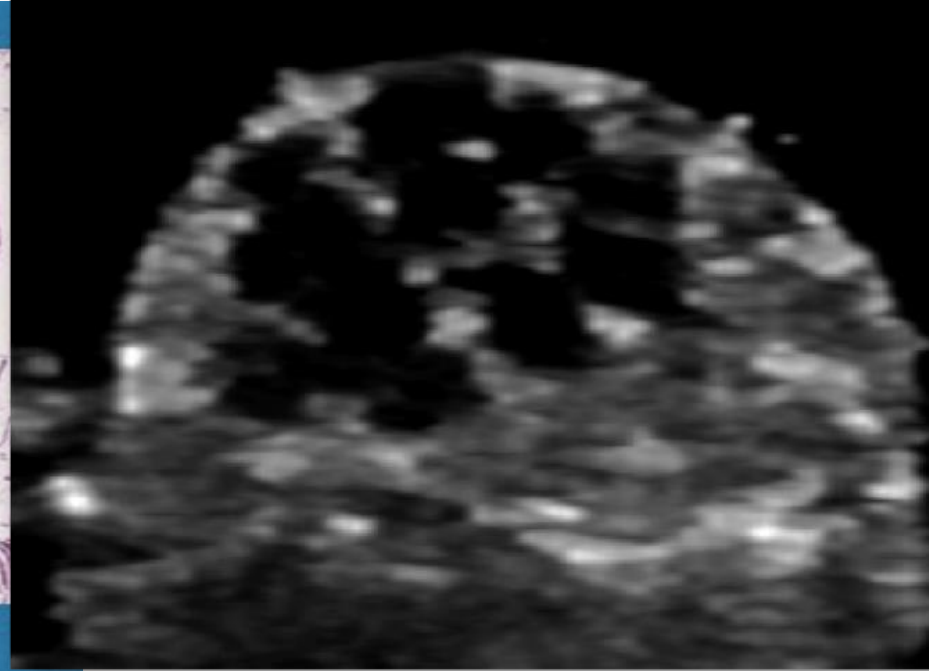
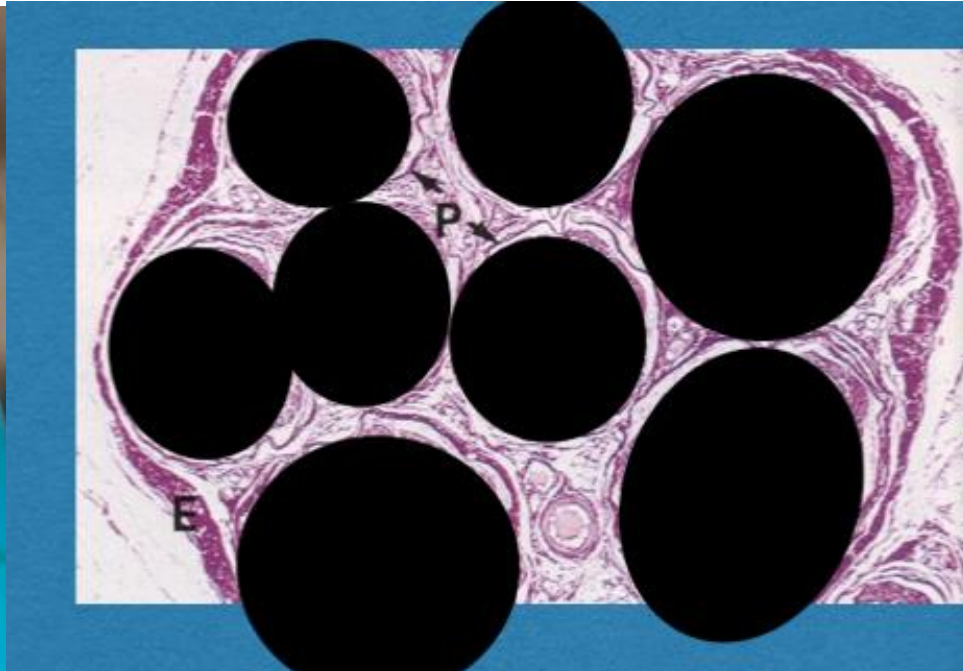
Lumbar, sacral - Anatomy



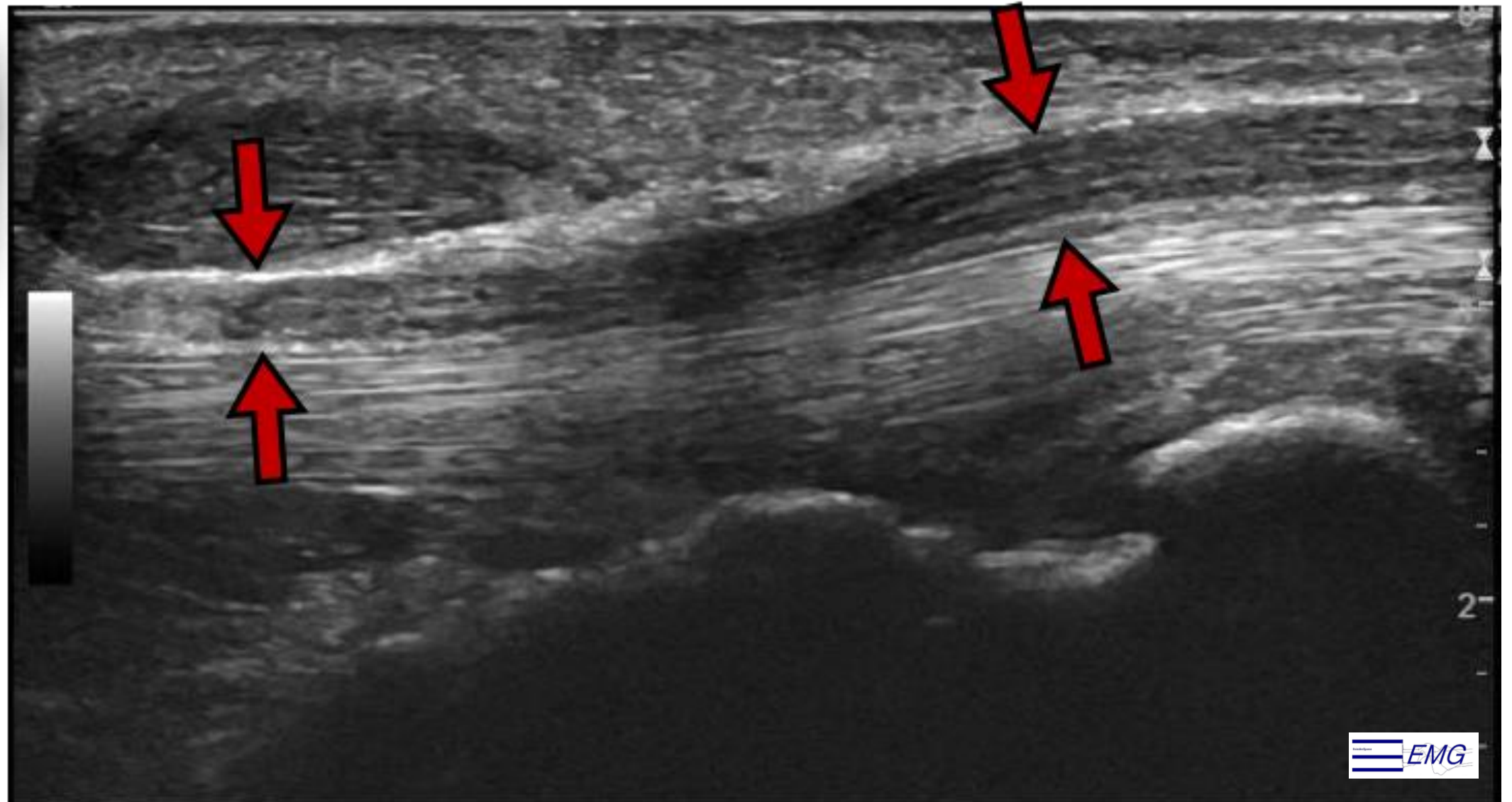
Sacral Plexus Vascular



Mononeuropathy

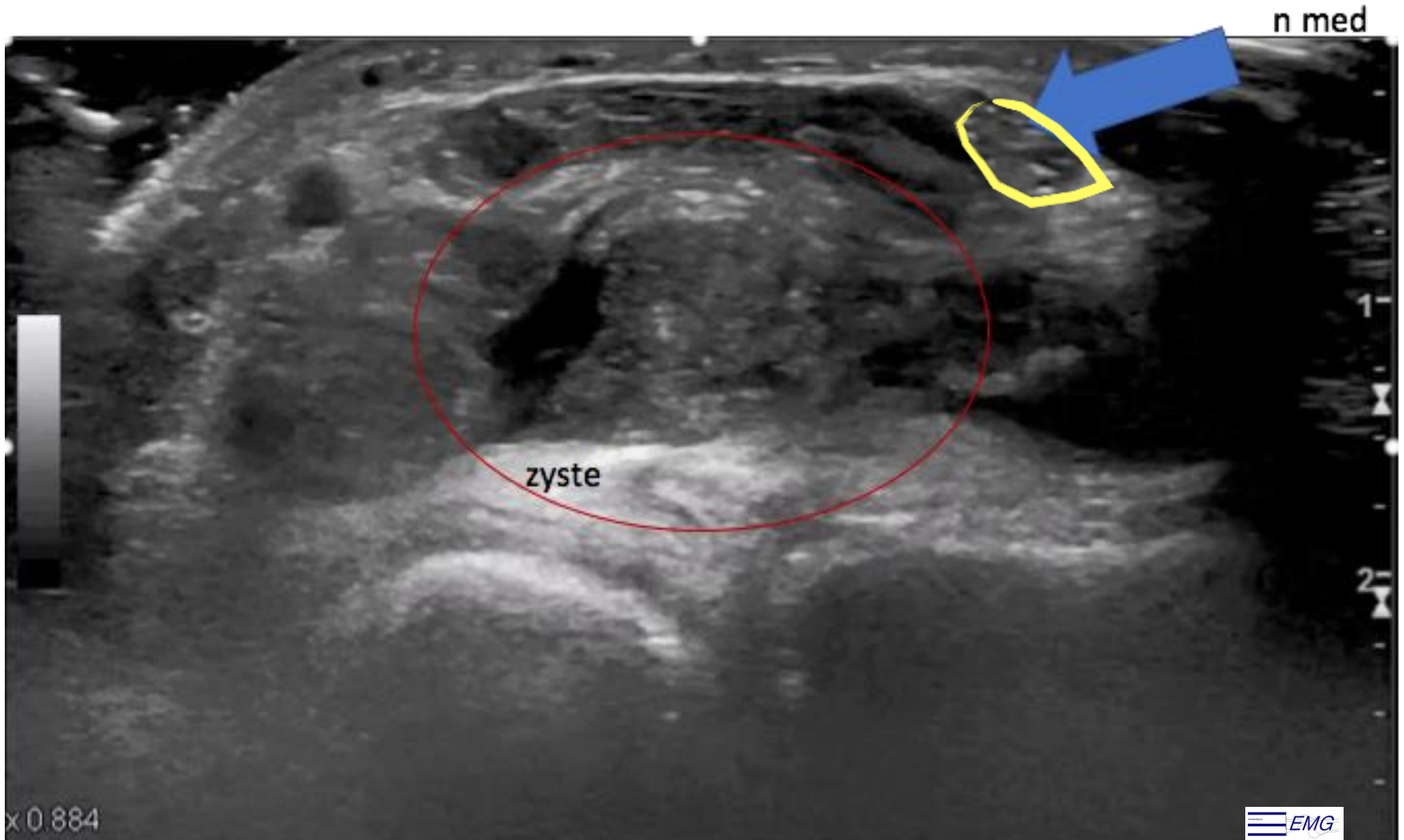


CTS

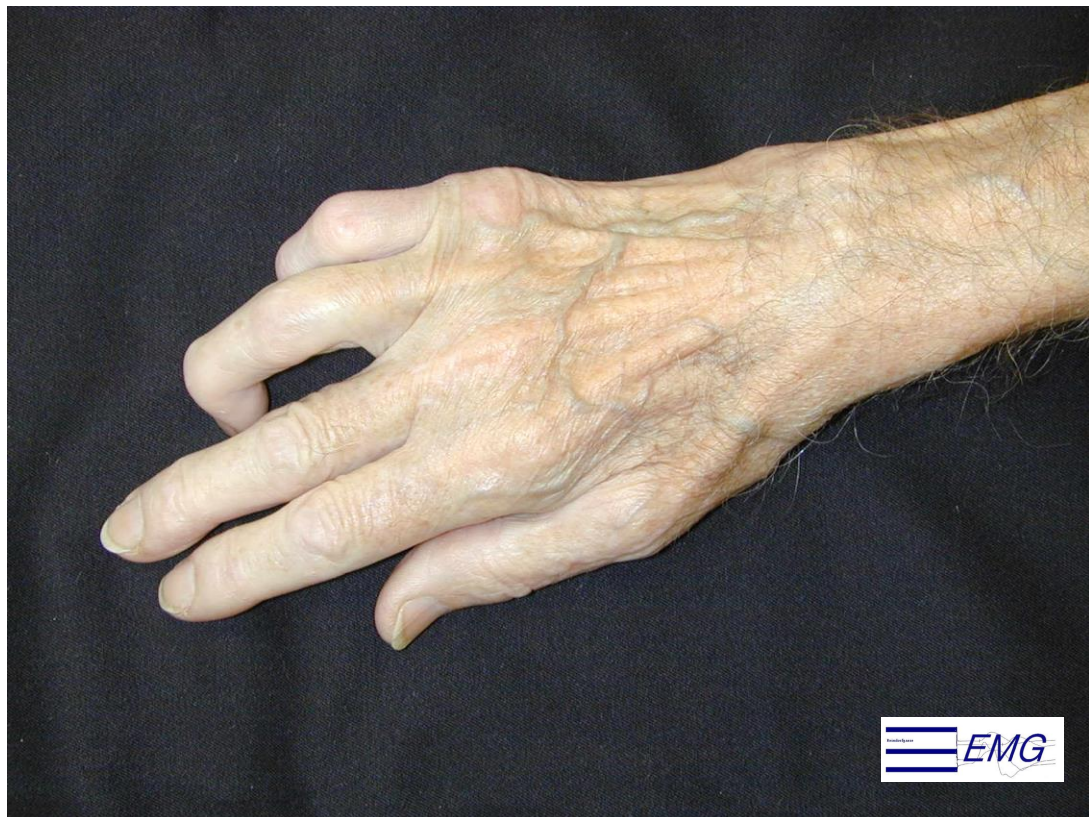




Acute Swelling and median nerve dysesthesia



**Acute Swelling and median nerve dysesthesia.
Ultrasound : synovial cyst, yellow: median nerve.**

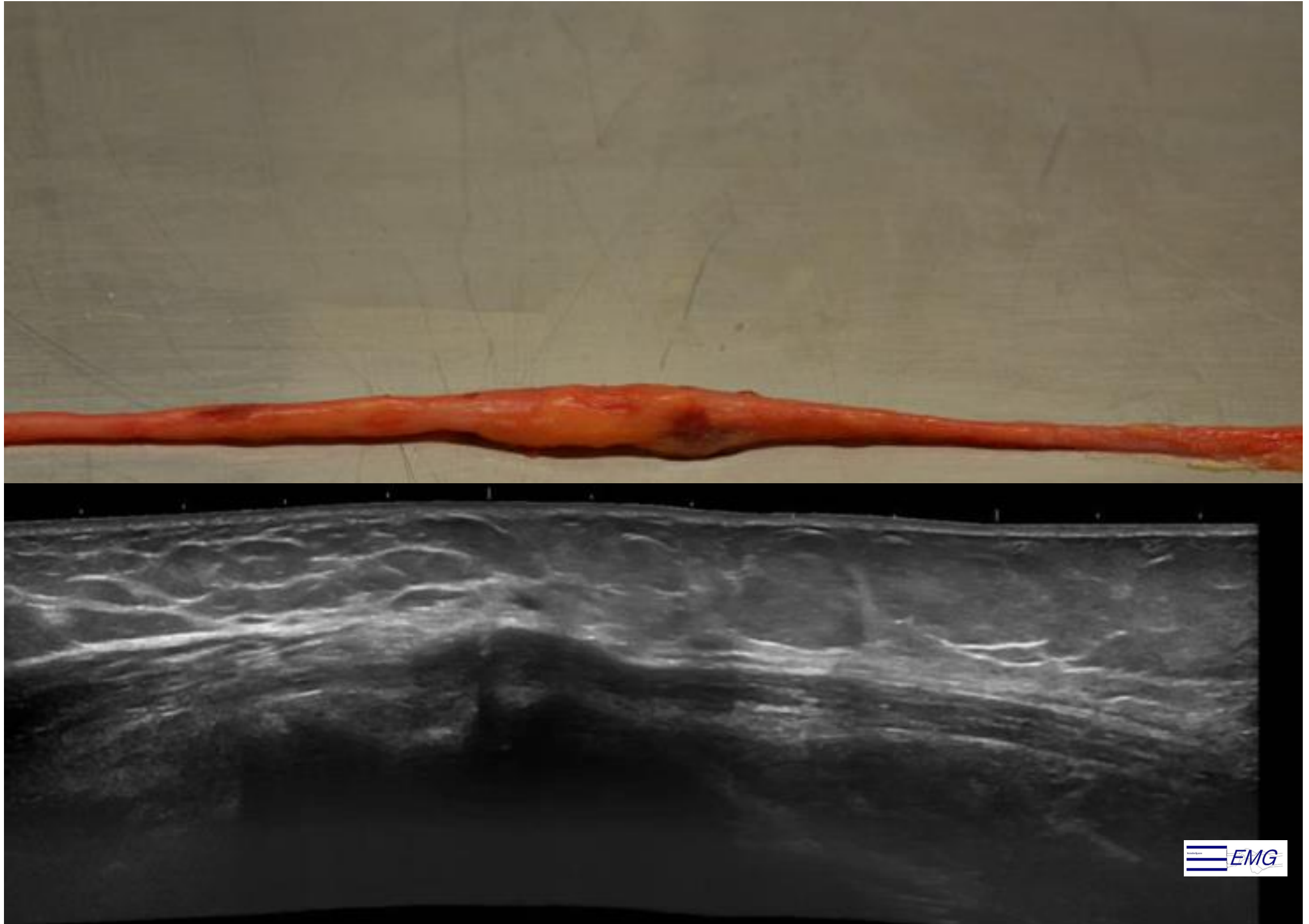


Ulnar nerve lesion



Median nerve: Thenar atrophy

SNUS

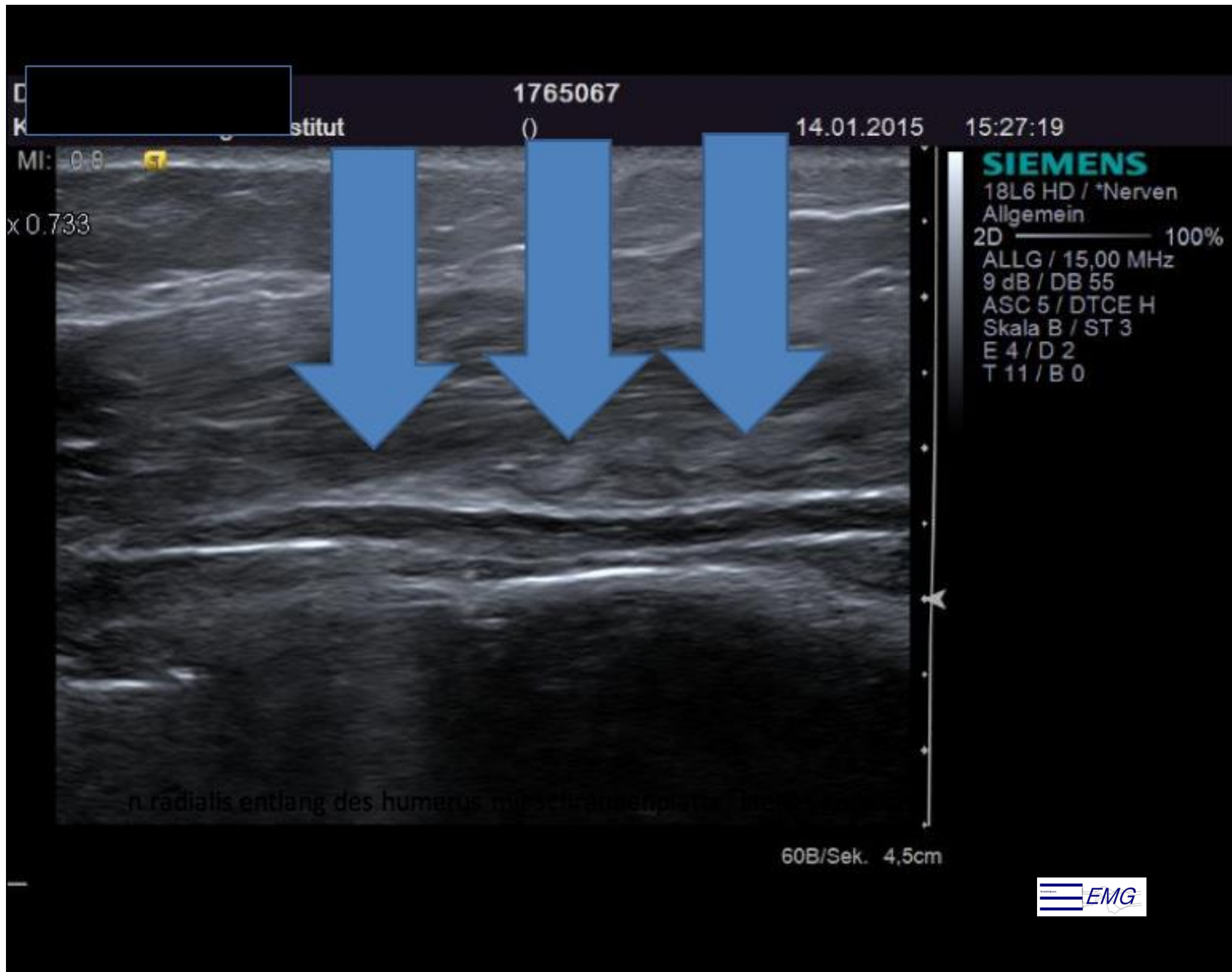




Chemotherapy paravasate, damage of cutaneous nerve



Radial nerve palsy after fracture

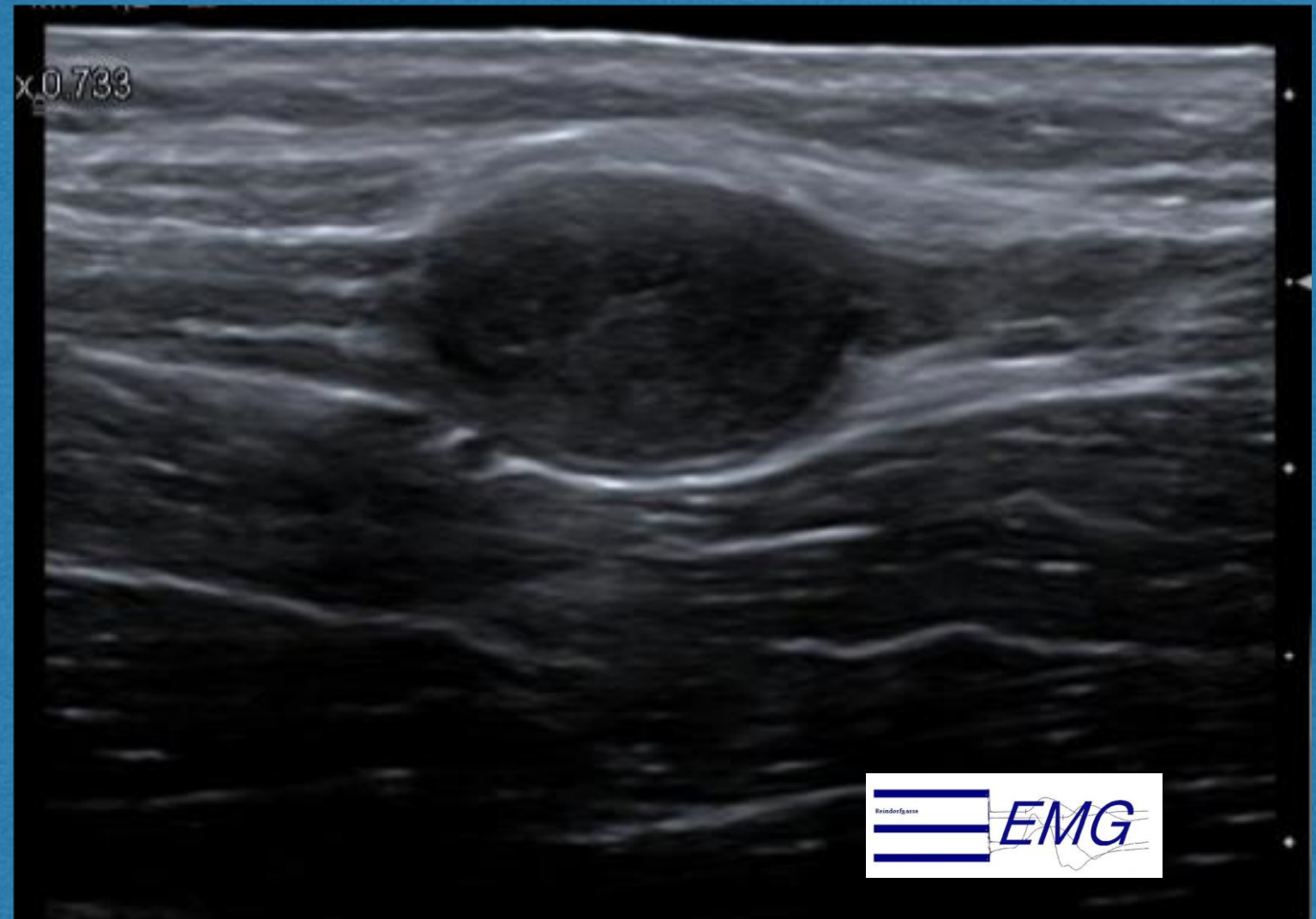


Ultrasound shows preserved continuity of the radial nerve

Diagnosis of nerve tumors



SCHWANNOM



Hip operations

- Hip operations
- Femoral nerve
- Sciatic nerve (traction)
- Cutaneous nerve

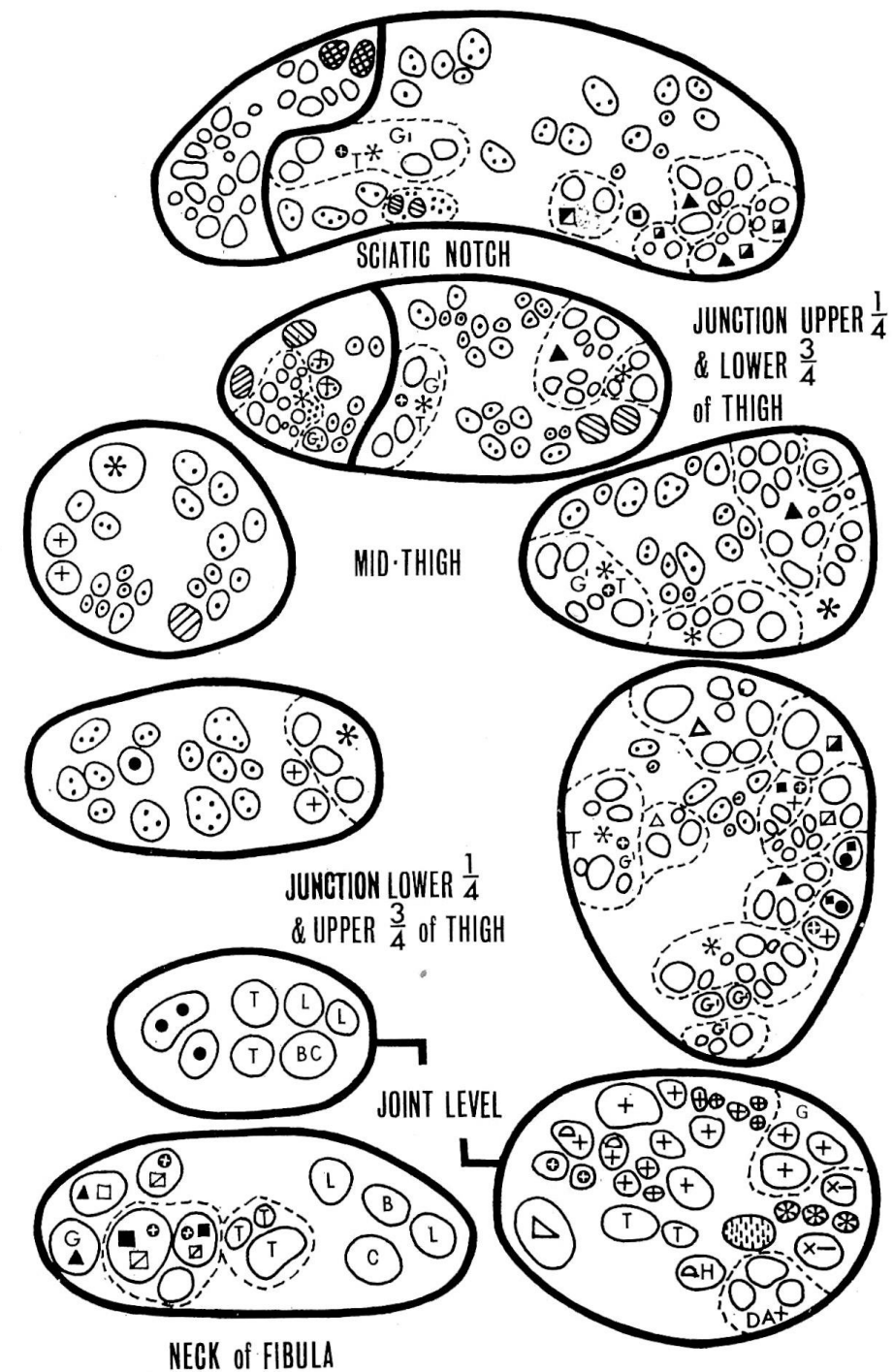
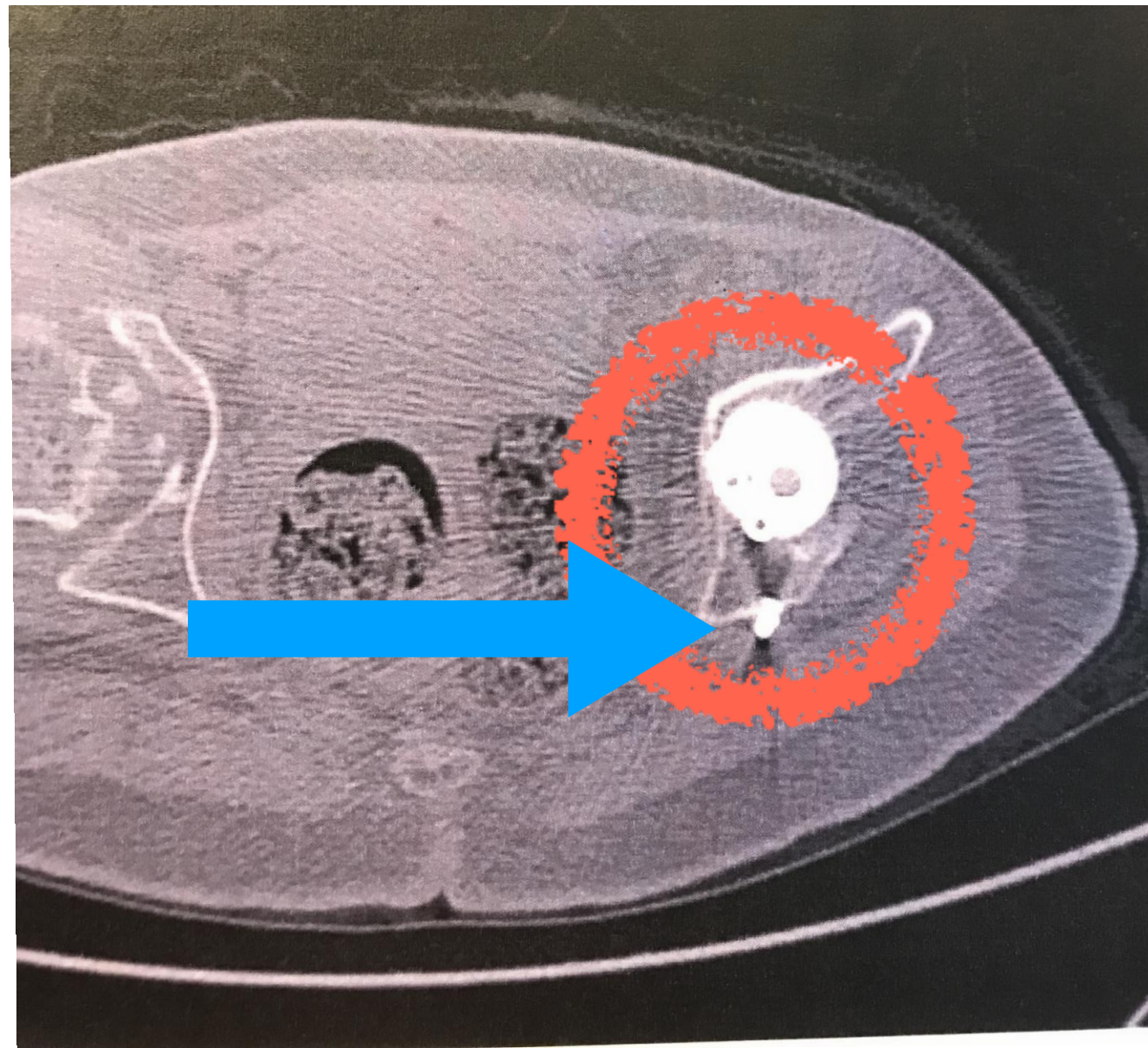


FIG. 191

The branch fibre composition of the funiculi of the sciatic nerve and its two main divisions. For key to symbols see Figures 192 and 193.

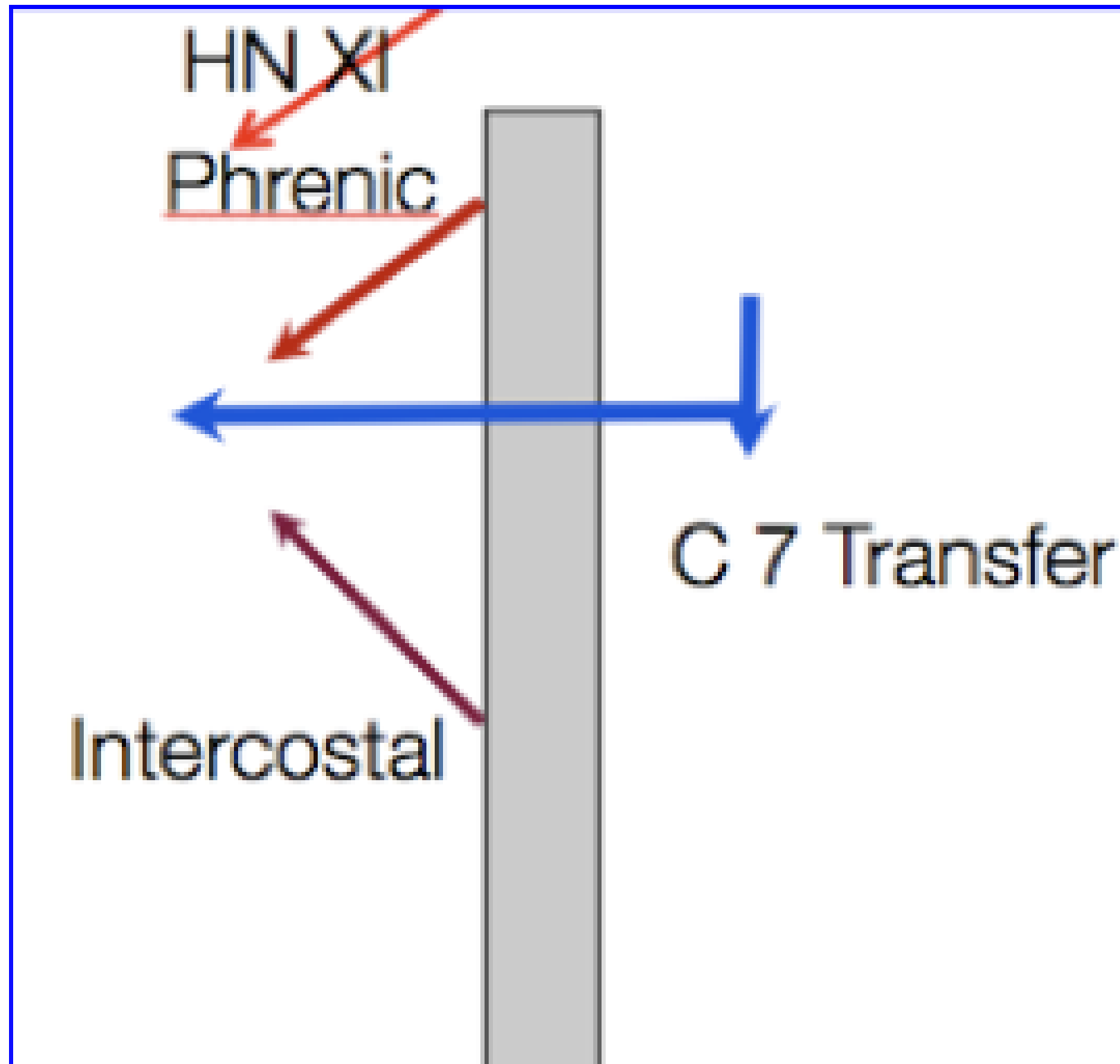
From: Sunderland



Arrow points towards screw from the acetabulum in contact with the sciatic nerve

Nerve reconstruction

- Neuroplasticity:
- Brachial plexus surgery:
 - Phrenic nerve and
 - intercostal
 - Contralateral C 7 transfer
- Hand transplant



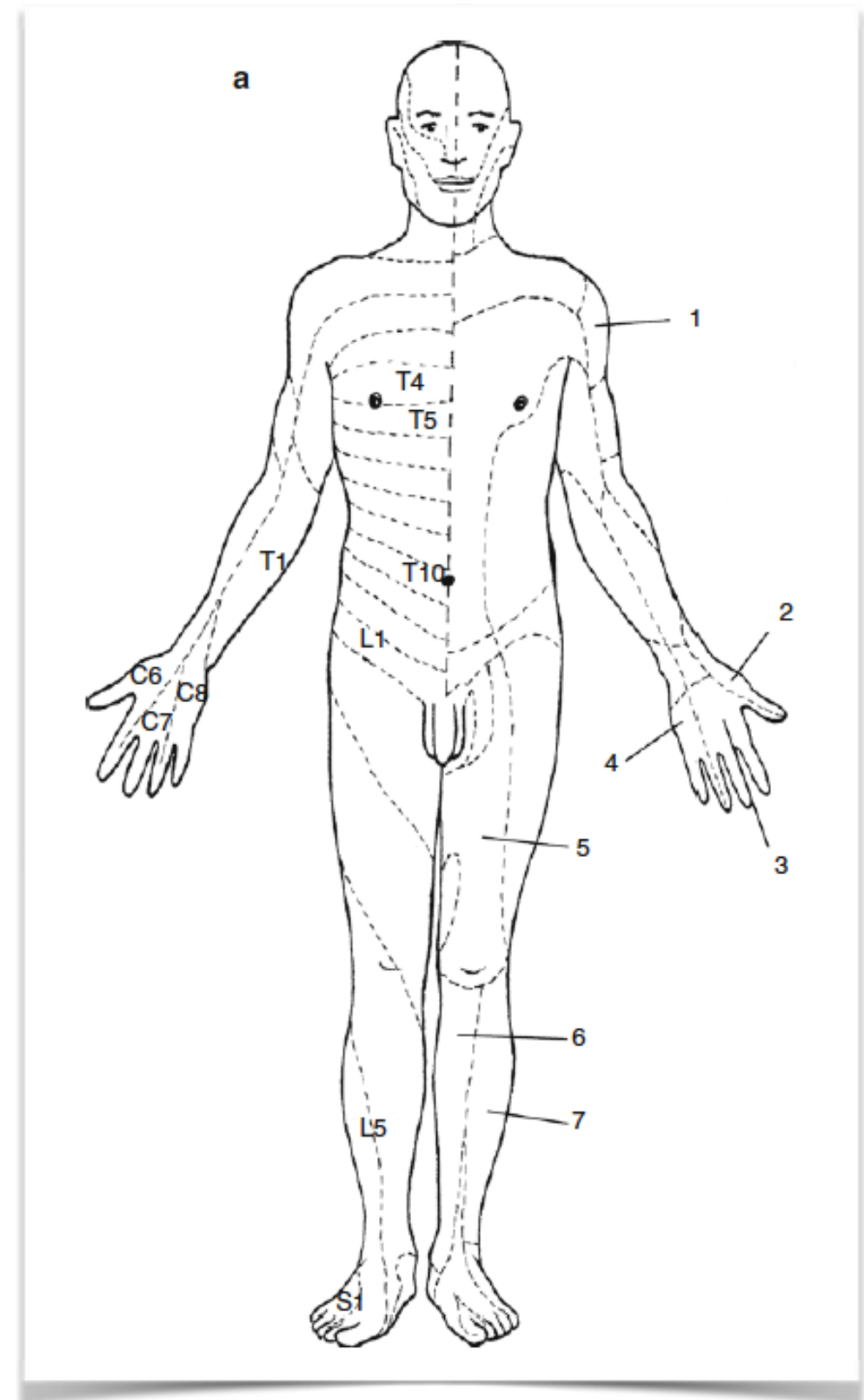
M. Tibialis posterior transfer

Ersatzoperationen



Polyneuropathy

- Distribution
- Statistics
- symmetric
- Asymmetric



Frequency

Diabetes

Toxic neuropathies

HIV

Immune mediated

Infection

Hereditary

Small fiber

**Painful
neuropathies**

Neoplastic

Immune mediated

Paraneoplastic

Frequency of neuropathy

- Most frequent: Diabetes, cryptogenic and toxic.
- Inflammatory, genetic, etc less frequent.

Visser NA, Notermans NC, Linssen RS, van den Berg LH, Vrancken AF. Incidence of polyneuropathy in Utrecht, the Netherlands. *Neurology* 2015;84:259-264.

Diabetes

- Symmetric
- **Distal symmetric neuropathy**
- Diabetic autonomic neuropathy
- „Pseudotabac“ type
- CIDP like

Diabetes

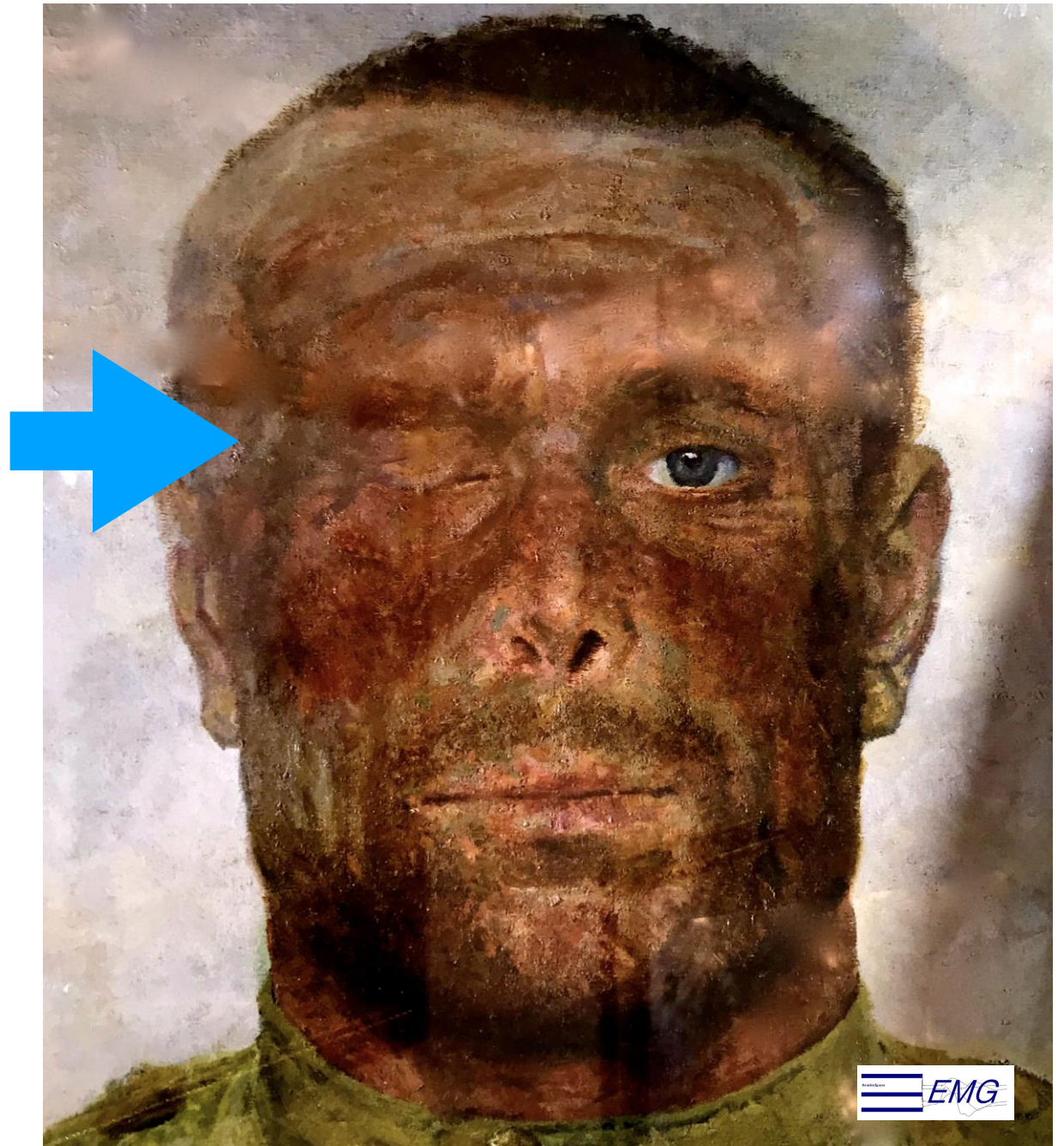
- Asymmetric forms
- Multiplex
- Truncal neuropathy
- Diabetic amyotrophy

Diabetes

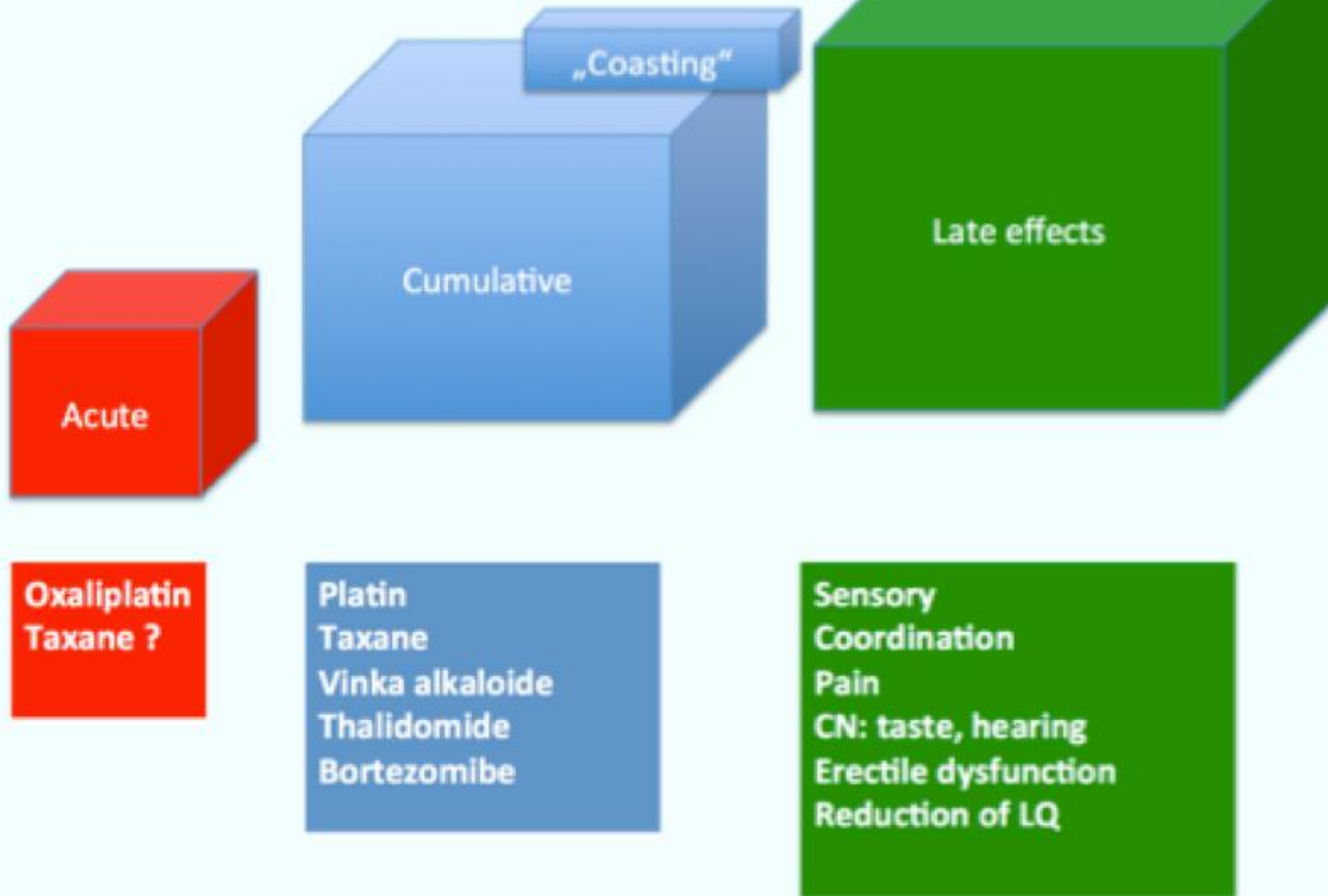
- Prediabetic neuropathy
- „Insulin neuritis“

Diabetes

- CN: III, (VII)
- Mononeuropathy: CTS
- Diabetic muscle infarct
- Diabetic cachexia

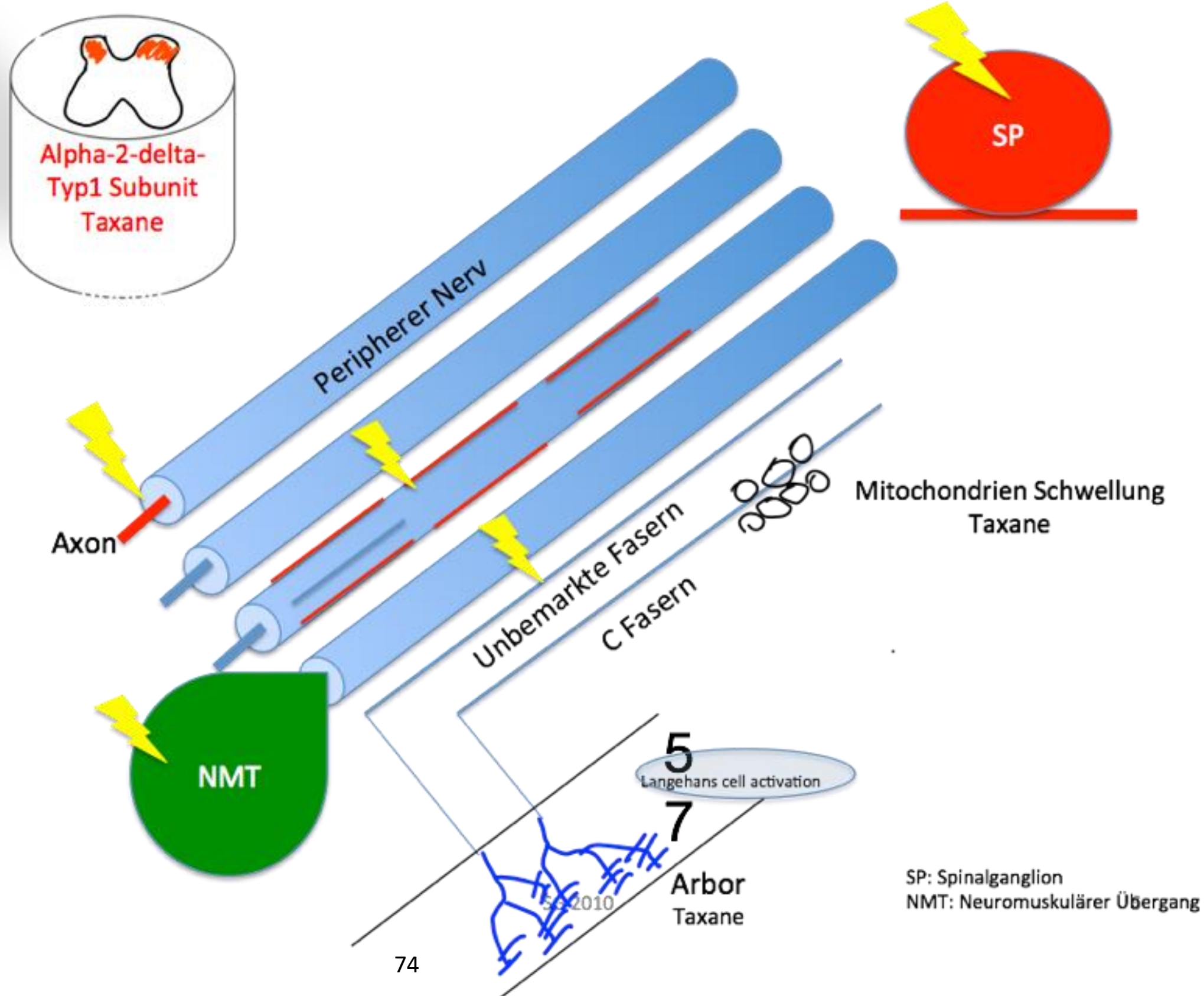


Chemotherapy induced neuropathy



Chemotherapy induced neuropathy

Different locations of pathology



CIPN-PNP

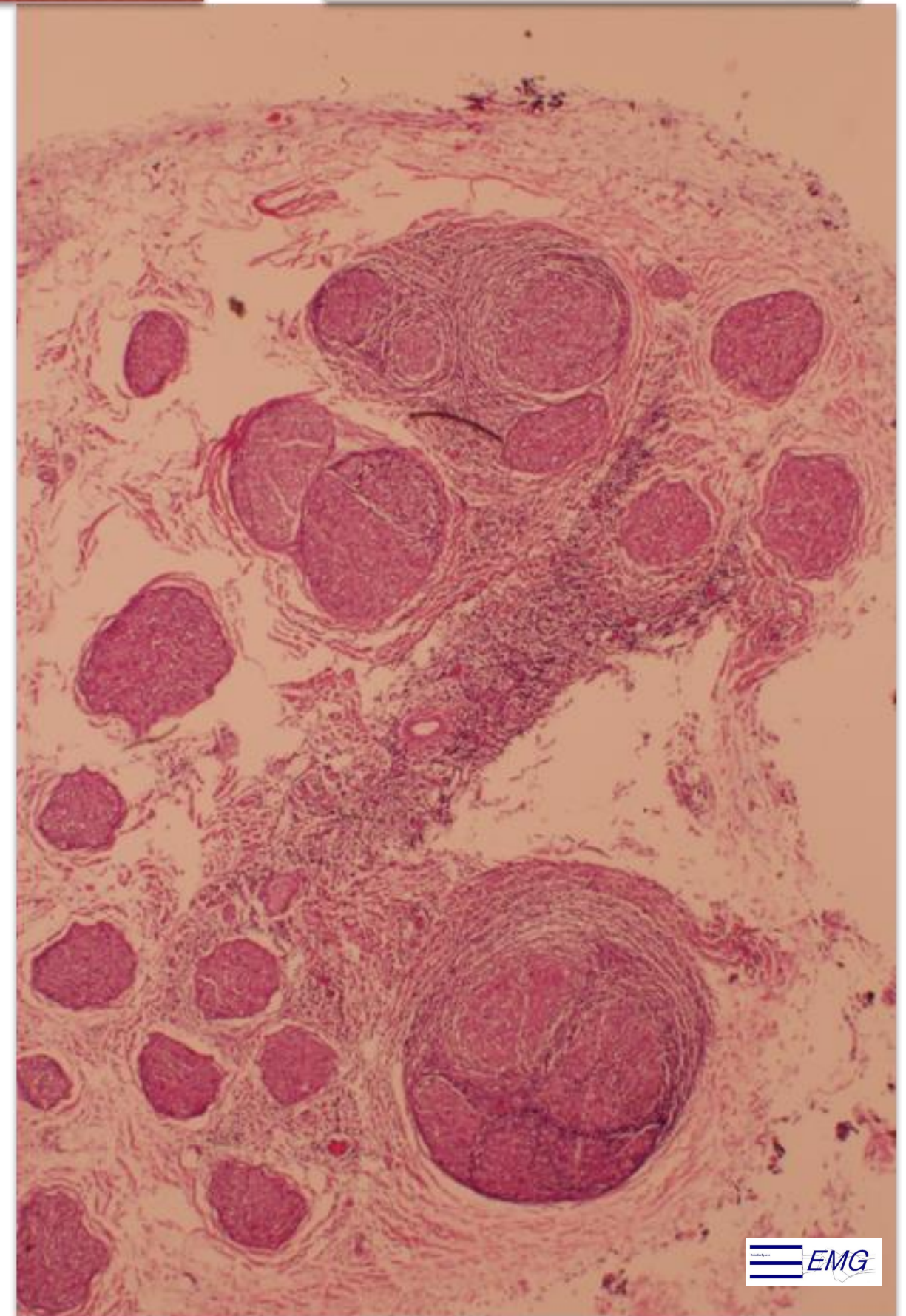
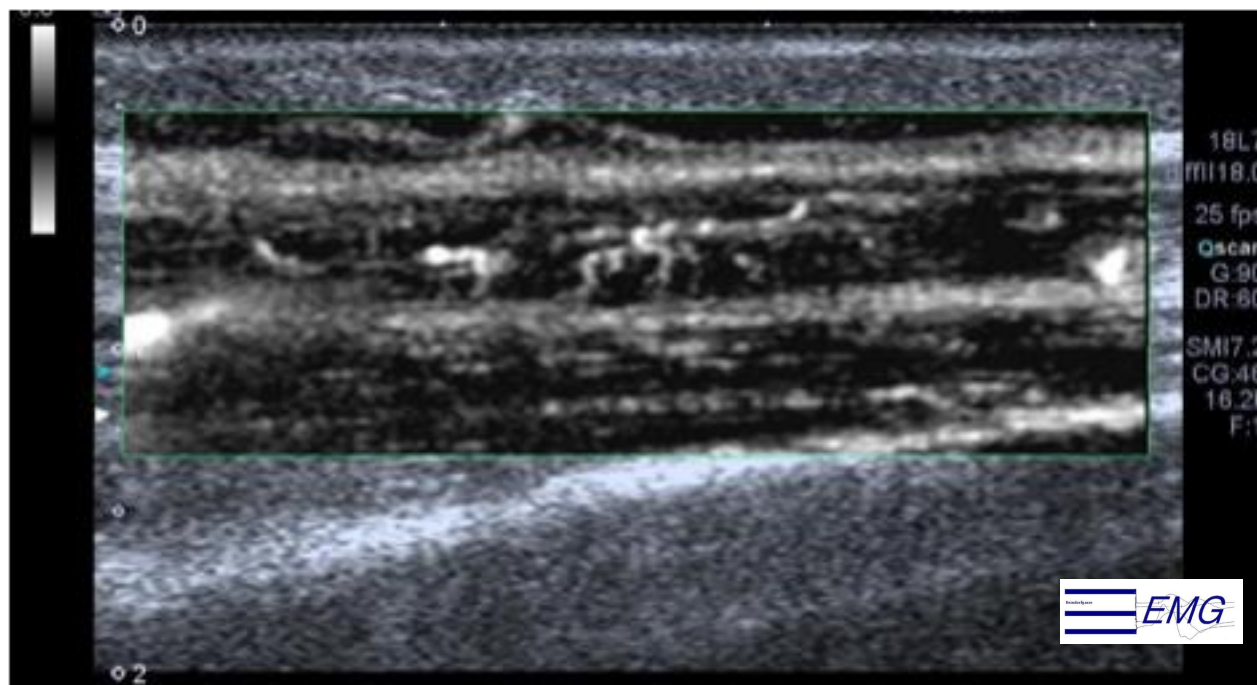
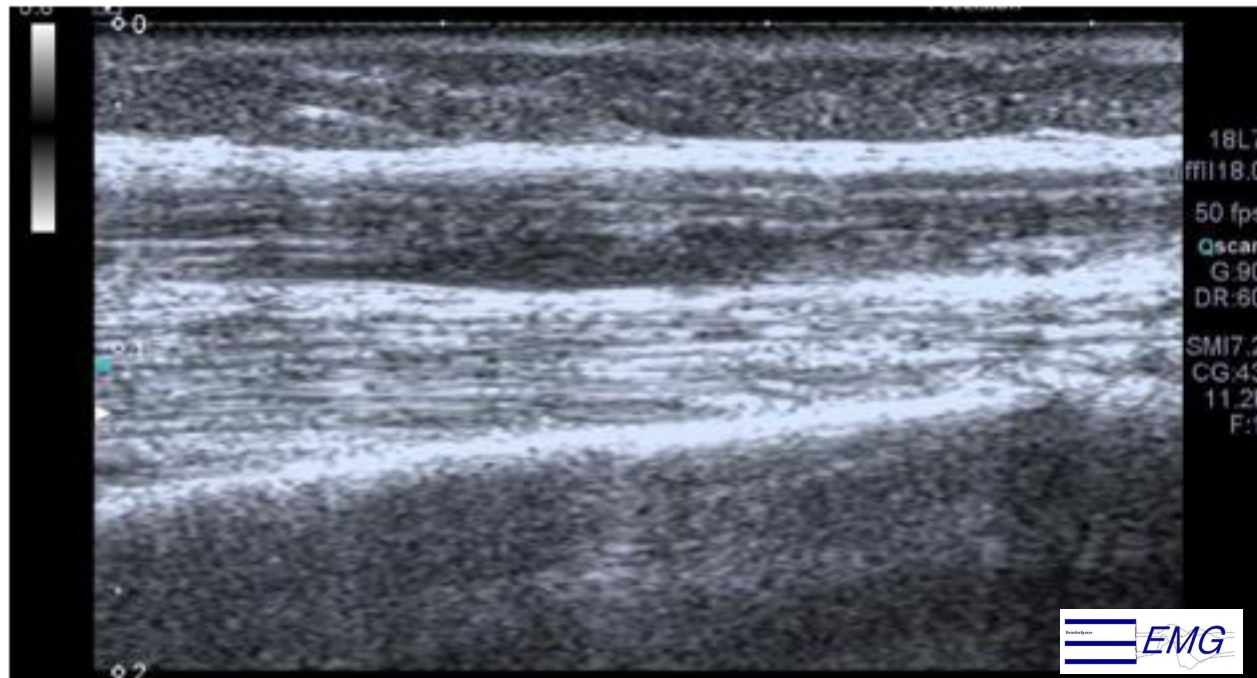
- Acute (Oxaliplatinum)
- Chronic and cumulative
- Coasting
- Late CIPN, RT, Muscle
- Induced (PD)

NCV/EMG/US

Neuropathy/muscle

Mononeuropathy

Neoplastic neuropathy



Toxic neuropathies

- Alcohol
- Industrial agents
- Historical:
 - Spanish toxic oil syndrome
 - U boat oil TCE neuropathy

Infection

- HIV
- Leprosy
- CMV
- Herpes
 - VII
 - Mononeuropathy
 - Motor involvement
 - Myelopathy



HIV and the PNS

ALS-variant syndrome
Autonomic

Toxic Drug treatment

CIDP
Cytomegalovirus
DILS
Drug-induced
Guillain-Barré
Mononeuritis multiplex
Neuromyotonia
Sensory motor polyneuropathy
VII nerve palsy
Mononeuropathy
Brachial plexopathy

Hereditary neuropathies

- CMT
- Fabry
- Amyloid
- Porphyria

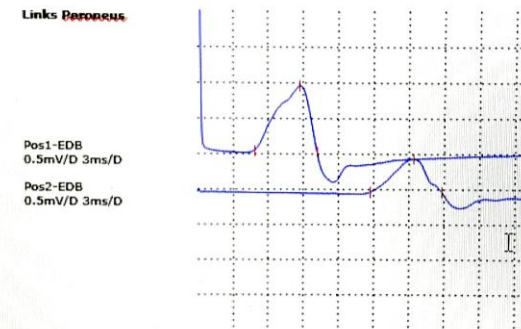
CMT detected in an asymptomatic patient.

Left: foot deformity.



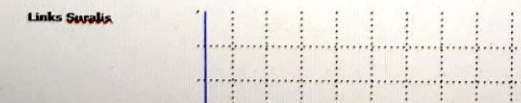
Links Peroneus Motor				
	Lat	Amp	CV	Amp%
	ms	mV	m/s	%
Pos1 - EDB	4.81	0.91		
Pos2-Pos1	14.7	0.47	32.4	-48.4

Rechts Peroneus Motor				
	Lat	Amp	CV	Amp%
	ms	mV	m/s	%
Pos1 - EDB	5.30	0.41		
Pos2-Pos1	17.3	0.37	28.3	-9.8



Sensible Daten und Kurven

Links Suralis Sensor			
	PeakLat.	Amp.	CV
	ms	µV	m/s
Pos1 - Lat. Malleolus	--	--	--



EMG

EMG

NMT

MG (AChR pos)

**MusK
LRP4**

„Seronegative“

LEMS

Neuromyotonia

**Other NMT
disorders**



Ocular myasthenia

- Triple furrow tongue

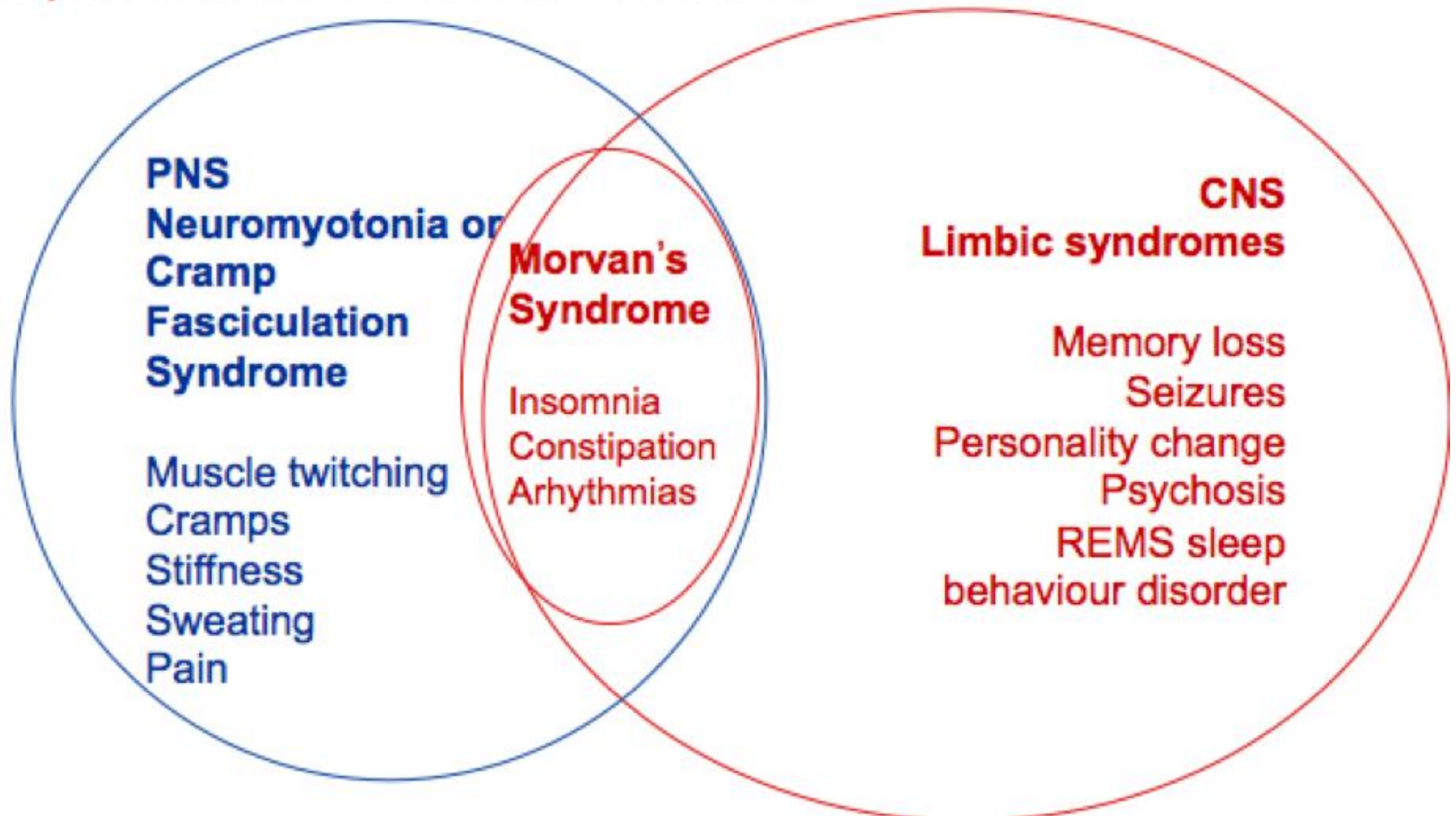


Lambert Eaton myasthenic syndrome

- Autoimmune presynaptic disorder
- 50 % paraneoplastic (SCLC)
- Electrophysiology: Increment
- Clinical: proximal weakness, facilitation of reflexes

Neuromyotonia

Peripheral and CNS disorders with VGKC antibodies



Muscle

- Distribution
- Onset/Course
- Acquired/genetic

Muscle

Hereditary

Inflammatory

Endocrine

Metabolic

Deposition

Toxic

Muscle

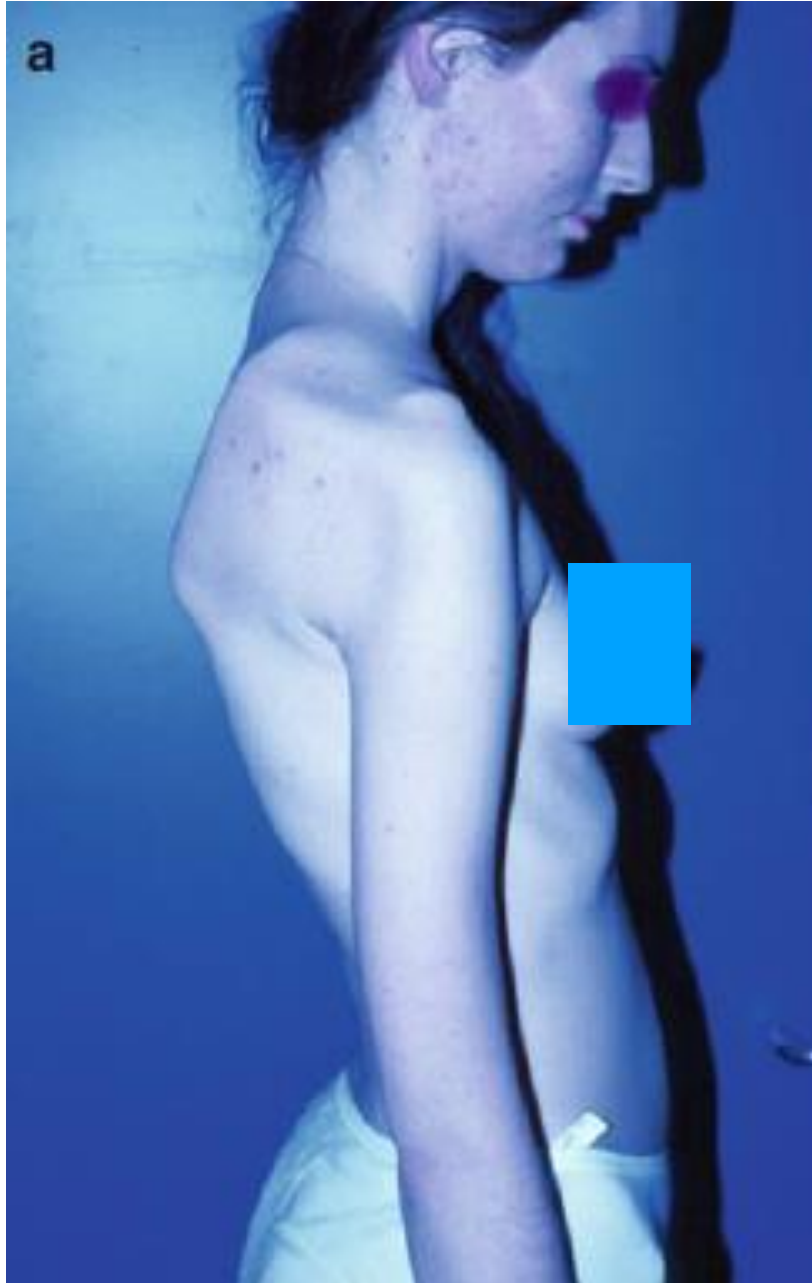
Infection

Systemic disorders

Myotonia

Critical illness

**Sarcopenia
Cachexia**



Hyperlordosis

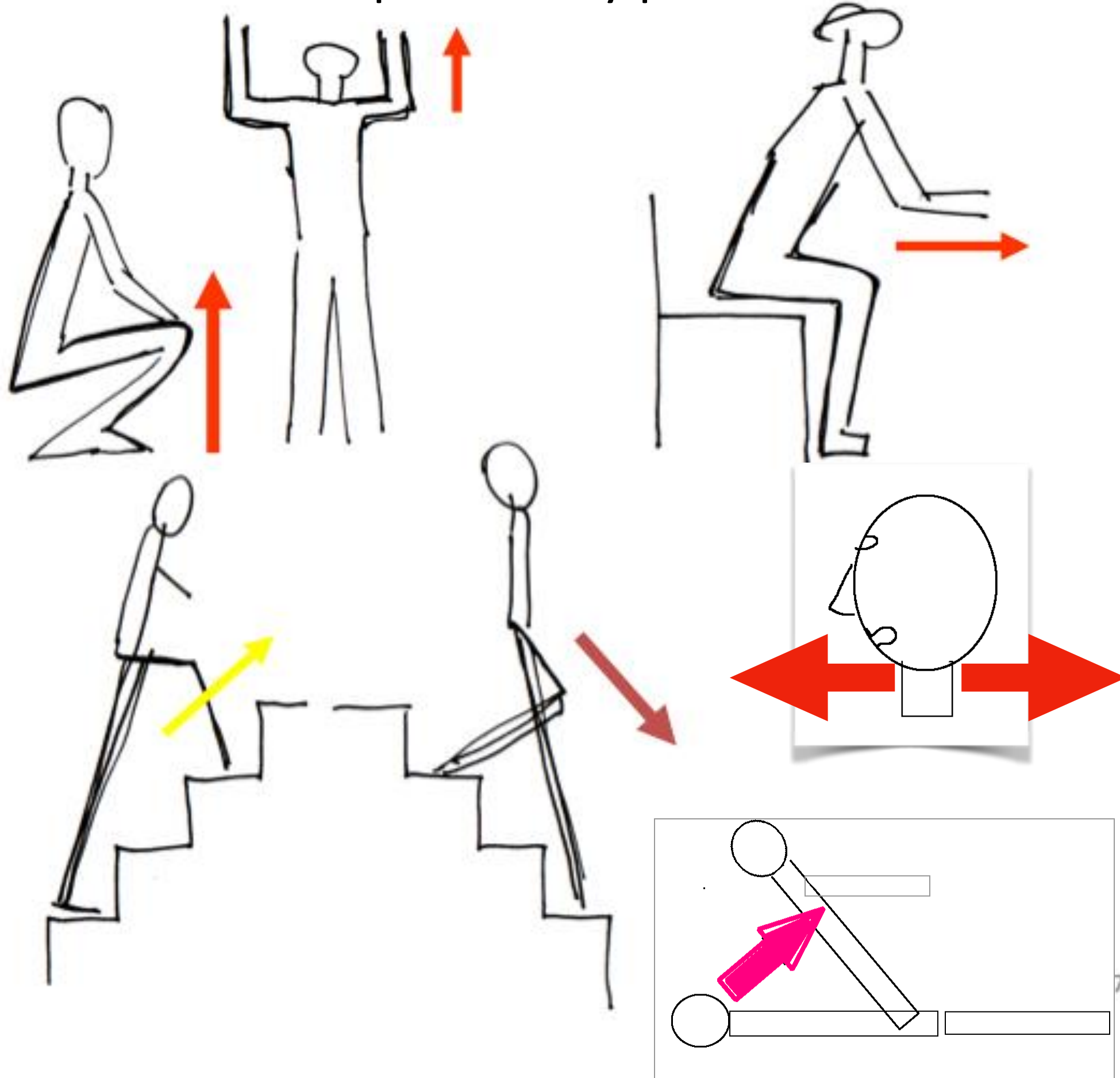


Shoulder girdle weakness



Shortened achilles tendons

A quick test for myopathic weakness



Mitochondrial myopathy

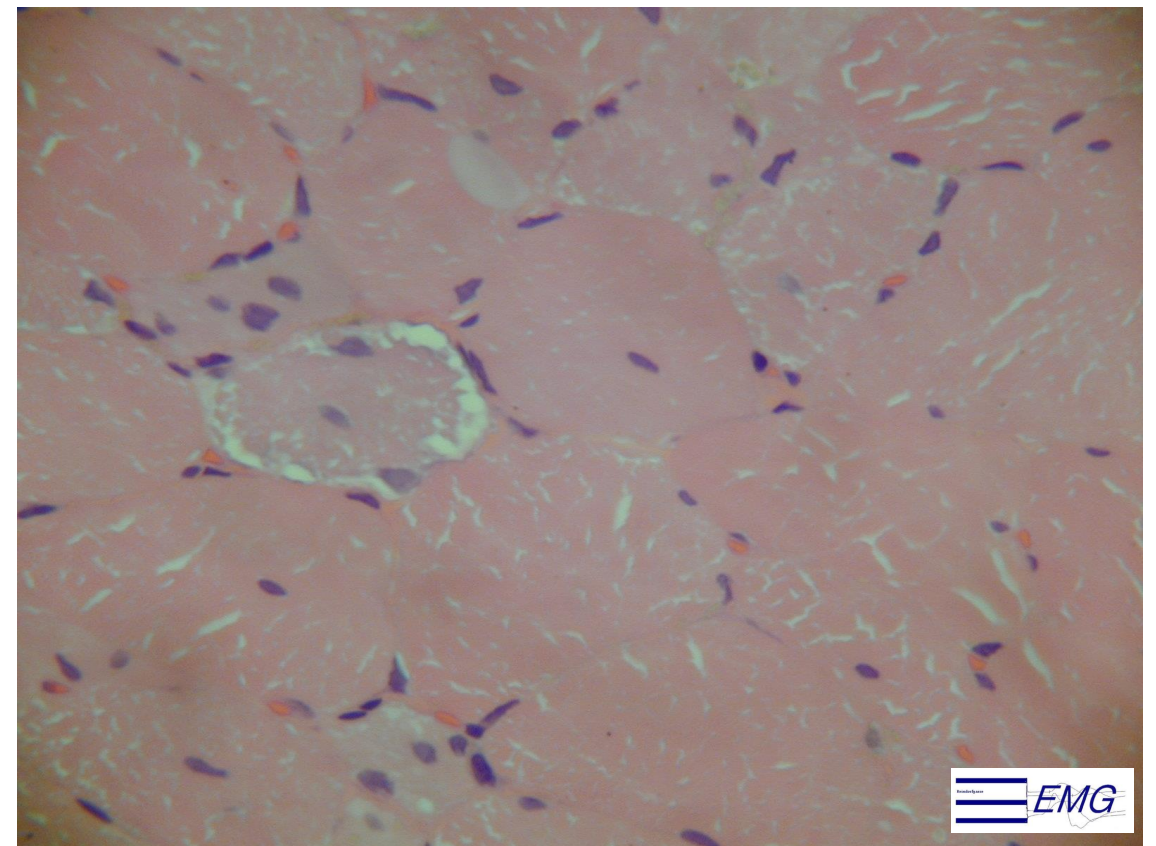


Dermatomyositis



Myositis

- DM (25 %) (Anti-TiF1 γ)
- PM (10 %)
- Necrotizing myopathy (NAM)

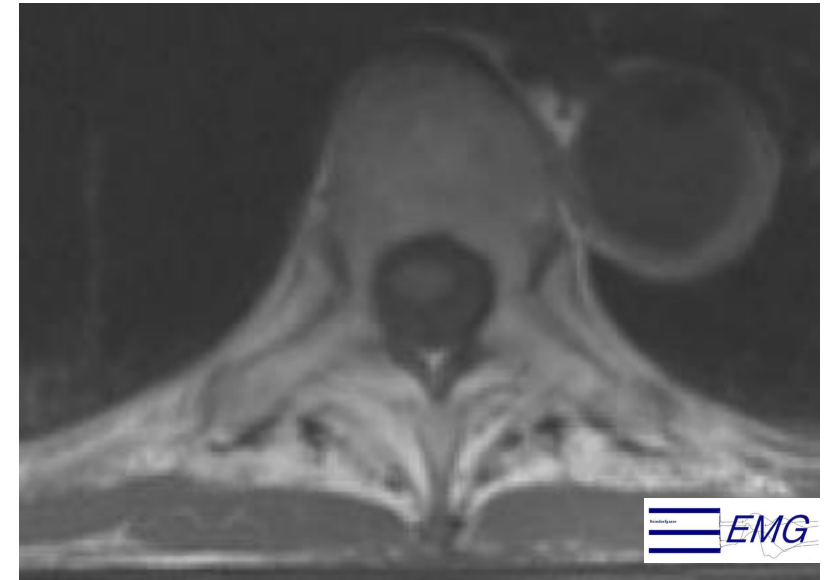




Volar atrophy

IBM

Axial myopathy



Toxic myopathies, Hyper CK emir

- Toxic myopathies: see: <http://neuromuscular.wustl.edu>
- Hyper CK emia: EFNS guidelines on the diagnostic approach to pauci- or asymptomatic hyperCKemia *European Journal of Neurology* 2010, 17: 767–773 (download)

Motor Neurone Disease

- MND
- ALS
 - Variations
 - Genetic forms
- Kennedy syndrom

- ALS
- Variants
- Genetic types



Flail arm syndrome

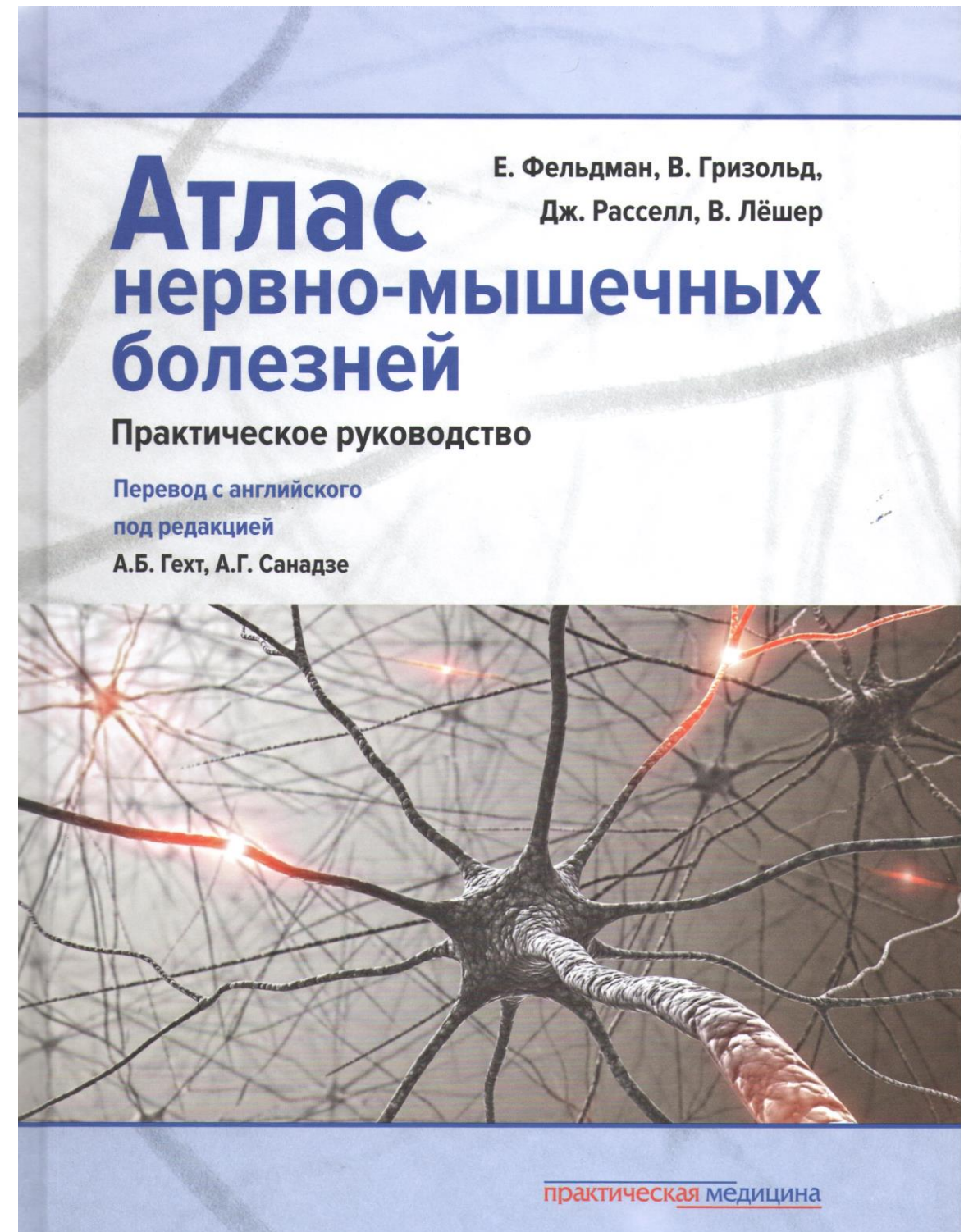
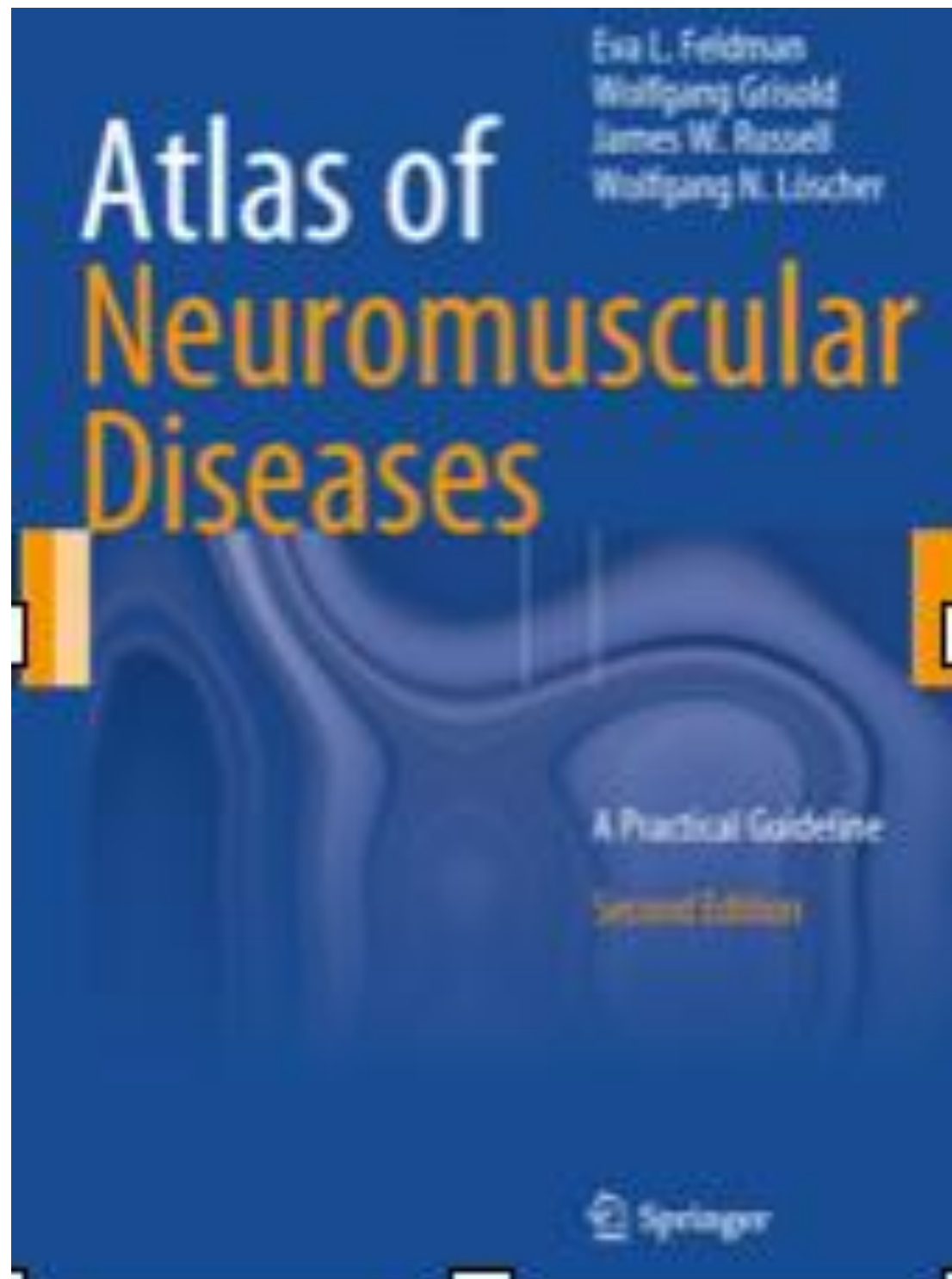


Atrophy of shoulder girdle (flail arm)



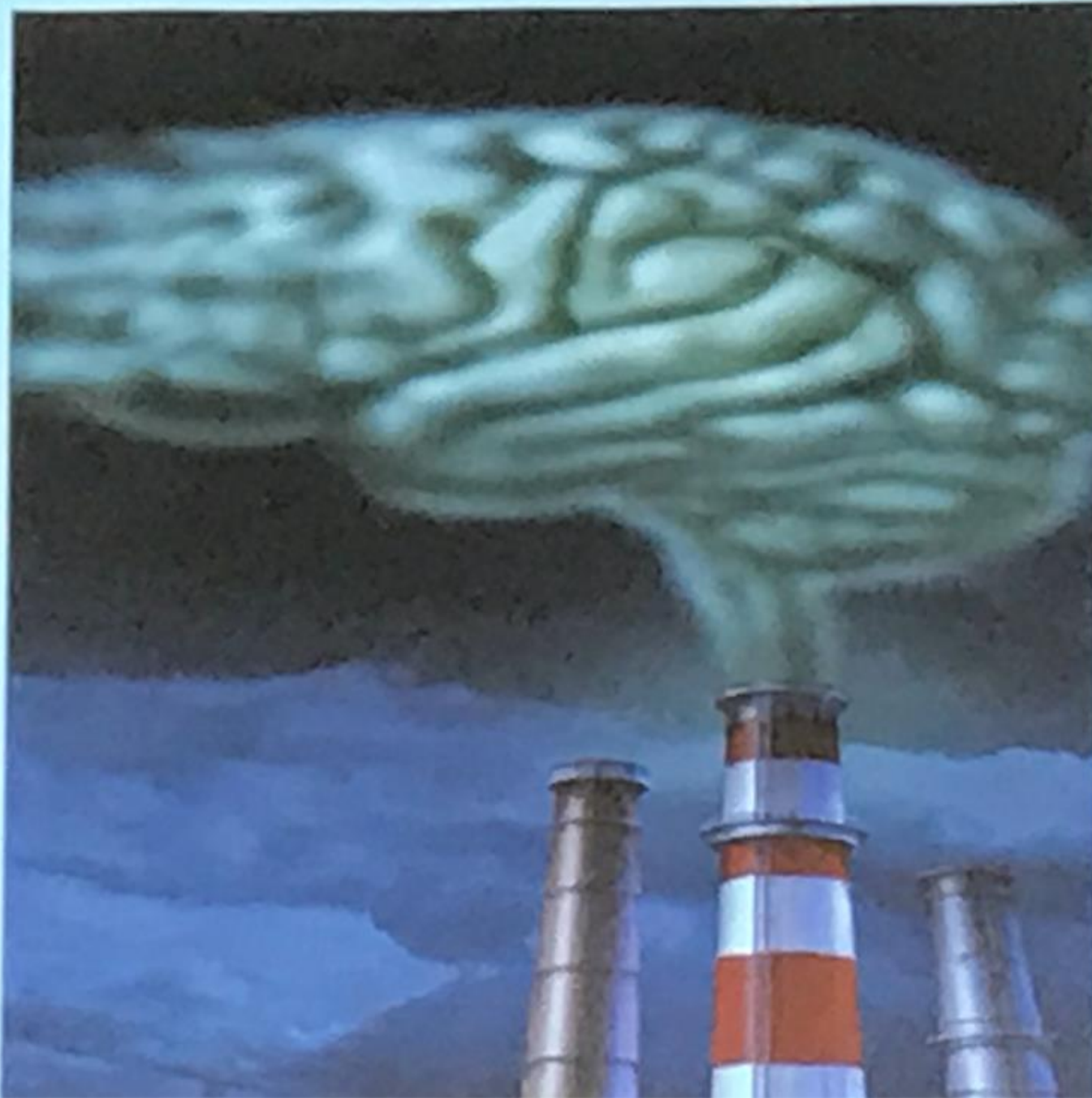
Thigh and leg muscles preserved

Sources:



Join the World Brain Day 2018 !

Brain impacted by air pollution



Check the WFN website for information.



Save the Date

JULY 6 - 10, 2018

ICNMD
2018



**15TH INTERNATIONAL CONGRESS
ON NEUROMUSCULAR DISEASES**

July 6 - 10, 2018 Vienna, Austria

www.icnmd2018.org

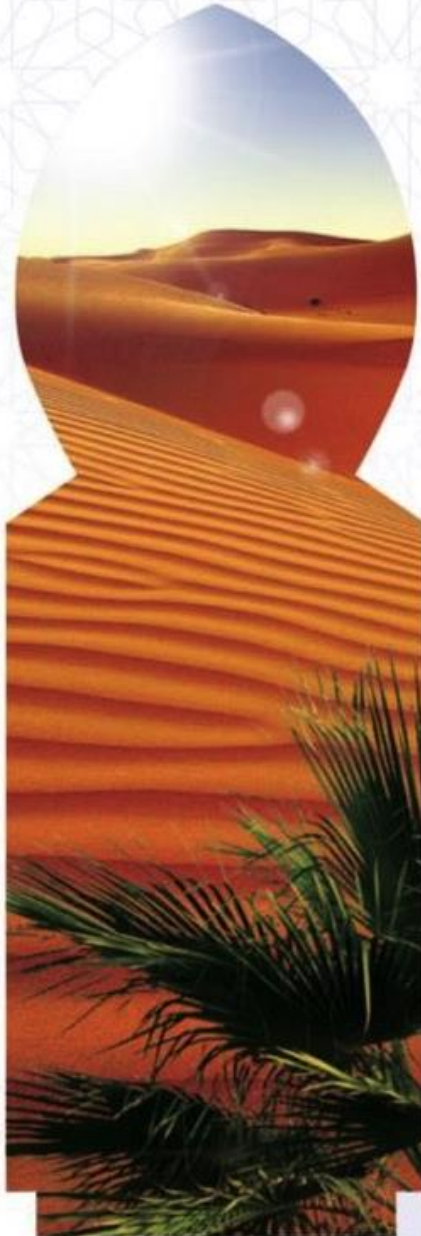


Ludwig Boltzmann Institut
für experimentelle und klinische Neurologie



دبي

ACCELERATING THE PACE OF CHANGE



DUBAI
WCN 2019