Brachial plexopathies and Parsonage-Turner syndrome

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Outline

- Brachial plexopathies
  - Anatomy
  - Causes of brachial plexopathies
  - EMG in brachial plexopathies
- Parsonage-Turner syndrome
  - Clinical picture
  - EDX and US
  - Treatment
- Cases

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Anatomy

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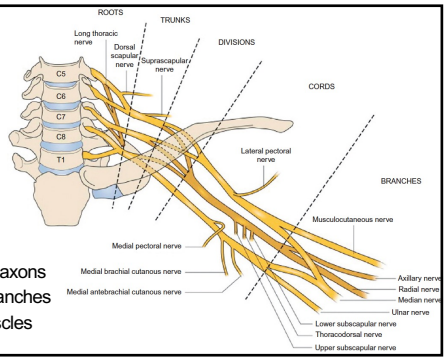

- 100 000 -150 000 axons
- 15 major nerve branches
- Innervates 50 muscles

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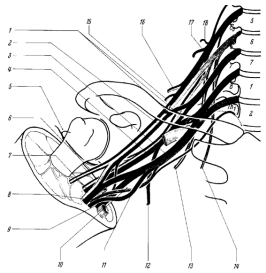
Plexus brachialis - clavicle



Mumenthaler&Schliack. Läsionen peripherer Nerven. Springer Verlag

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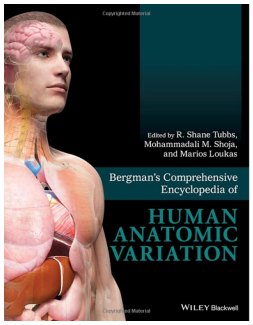
Tubbs, Shoija and Loukas
Bergman's Compendium of Human Anatomic Variation
Wiley Blackwell 2014



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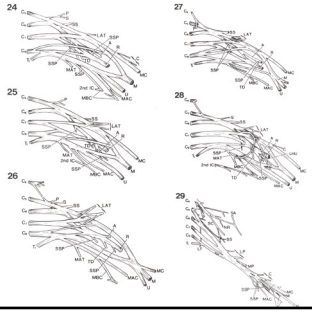
Kerr AT. The brachial plexus of nerves in man. Am J Anat 1918:23;285-395



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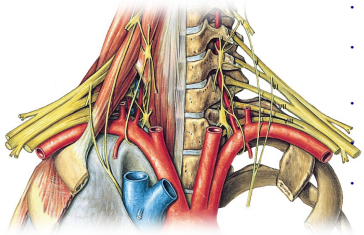
Surrounding structures

- Clavicle
  - Deformed fracture
- 1st rib
  - Anomalous cervical rib
- Arteries
  - Catheterization
  - Thrombosis
- Lymph nodes
  - Metastasis
- Lung
  - Cancer (Pancoast tumor)
- Scalene muscles
  - Scalenus anterior syndrome (?)



Putz and Pabst (ed) Sobotta. Atlas of Human Anatomy. 20 th edition. Urban & Scwarzenberg

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Causes of brachial plexopathies

- Parsonage -Turner sdr (plexus neuritis, neuralgic amyotrophy)
- Trauma
- Obstetric injuries (Erb's palsy)
- Temporary compression
- Iatrogenic
  - Surgery, especially surgery for TOS
  - Anesthesia, particularly local anesthesia in the plexus
  - Radiation therapy
  - Catheterization of the arteries
- Chronic entrapment (thoracic outlet syndrome, TOS)
- Cancer and metastatic infiltration

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Trauma

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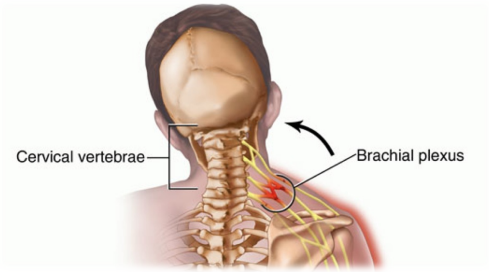
Traumatic mechanism



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Traumatic mechanisms

- Injury to the shoulder in motorcycle accidents
  - 95% men
  - Median age 28 years
  - Truncus superior
- Shoulder luxation
  - Timely repostion is important to minimize the injury
  - Often also axillary nerve involved
- Stabbing with knife
- Bullet wounds

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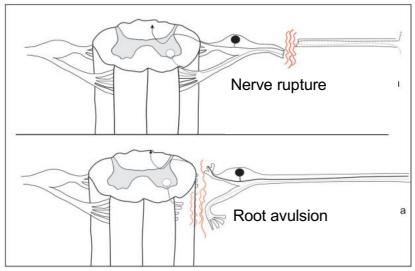
Nerve rupture/root avlusion



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Root avulsion

- Spinal nerves C5, C6, C7
  - Anchored extraforaminally to transverse processes
  - Less prone to avulsion
- Spinal nerves C8 and Th1
  - Anchored to the spinal cord
  - Tend to avulse more easily

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Erb's palsy

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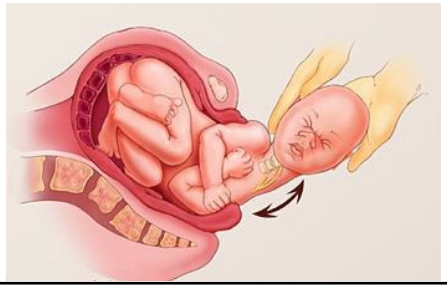
Erb's palsy – shoulder dystocia



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Incidence of Erb's palsy

- 0.5-2/1000 childbirths
- Number has decreased during the last decades

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Mechanisms

- Shoulder dystocia present in around 50%
- Traction forces applied externally
- 2-4% of Erb's palsies occur following caesarean section
  - During caesarian section similar traction forces

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Risk factors

- Large babies (>4500 g)
- Wide shoulders
- Mother has diabetes
- Previous Erb's palsy
- Twins
- If three risk factors - the risk for Erb's palsy is 8% (50 x times)
- NNT for Cesarean sections in mothers with 3 riskfactors to save one patient is 150

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Patterns

- C5 and C6
  - Classical Erb's palsy, 50%
- C5, C6 and C7
  - Erbs palsy +, 35 %
- C5-Th1 with sparing of finger flexion
- C5-Th1 and Horner's syndrome
- C8-Th1 and Horner's syndrome
  - Klumpkes palsy, very rare

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Prognosis

- 60% good
  - Return of elbow flexion at 2 months is a good sign
  - Usually, good recovery in 3-4 months
- 30% moderate
- 10% poor
  - Horner's syndrome
  - Total palsy without improvement in the first week
  - Paresis with no improvement in the first 6 months
  - Avulsion of spinal nerve
- Aberrant reinnervation
- Apraxia

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Surgery of Erb's palsy

- Significance of surgery not clear
- Usually at three months
  - Total palsy
  - Severe palsy with no improvement
- Repair of damaged nerves
- Transposition of nerves to denervated muscles
  - Intercostal nerve to musculocutaneous nerve
- Reattaching muscles to new sites

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Note

- Sometimes you will find in patients referred for EMG with inactive, old upper plexus lesions
  - May be sequalae of Erb's palsy the patient is not aware of
  - Have patient as mother about function of arm in neonatal period

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Temporary compression

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Temporary compression

- During coronary by-pass surgery
  - 5-7 % of patients have plexus lesion
  - Lower trunk is affected
- During general anesthesia
  - Arm is supinated and abducted more than 90 degrees
- Acute compression due to backpacks, especially lean soldiers

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Chronic compression - Entrapment

"Thoracic outlet syndrome" - TOS

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Thoracic outlet syndrome - TOS

- Second most published entrapment syndrome
- Cotroversial classification
- Diagnostic criteria unclear, no generally accpted criteria
- No controlled therapeutic studies

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Thoracic outlet syndrome - TOS

- True neurogenic
  - Objective EMG findings
  - Incidence 1/1000 000 per year
- Vascular
  - Venous
  - Arterial
- Disputed
  - Symptoms without obective findings

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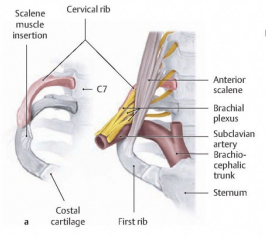
Cervical rib or ligament



- 0.3% of people
- 75% women

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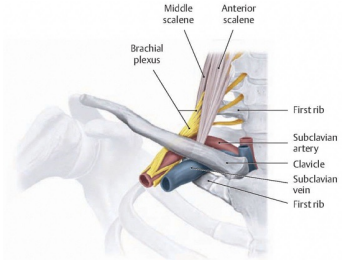
Costoclavicular compression



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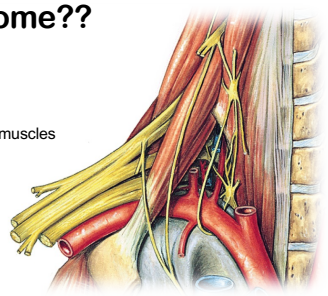
Scalenus syndrome??

"Neuromythology"

Compression between m.scalene muscles



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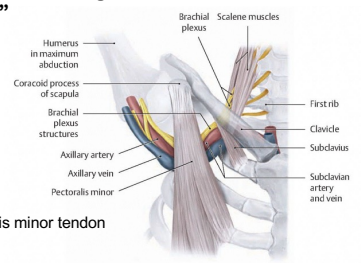
Pectoralis minor syndrome???

"Neuromythology"



Compression by m.pectoralis minor tendon

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Symptoms

- Shoulder pain
- Arm pain
- Paresthesia
- Weakness

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Tests used commonly

- Roos test – totally useless test
  - Arms above head in abduction and external rotation
  - 3 min extension and flexion of fingers
  - Non-specific and poorly validated
  - Abnormal in 47% of normal subjects and 77% of CTS patients
- Adson's maneuver – useless test
  - Feel the radial pulse bilaterally in the arms
  - Inhalation and rotation of head
  - Disappearance of pulse
  - Poor specificity

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Muscle and Nerve 1999:22

ISSUES & OPINIONS

THORACIC OUTLET SYNDROME

Thoracic Outlet Syndrome Is Underdiagnosed

DAVID B. ROOS, MD, FACS

Department of Surgery, University of Colorado Health Sciences Center, Denver, Colorado, USA

Thoracic Outlet Syndrome Is Overdiagnosed

ASA J. WILBOURN, MD

EMG Laboratory, Cleveland Clinic, Cleveland, Ohio, USA

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ISSUES & OPINIONS

THORACIC OUTLET SYNDROME

Thoracic Outlet Syndrome Is Underdiagnosed

DAVID B. ROOS, MD, FACS

Department of Surgery, University of Colorado Health Sciences Center, Denver, Colorado, USA

- Anomalies (cervical ribs and bands) are common
- Unusual activity may trigger anatomical alterations that cause neurovascular compression
- Roos test
- EMG has limited value in conformation of TOS
  - EMG sees only axonal damage
  - Thin fibers not tested

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Thoracic Outlet Syndrome Is Overdiagnosed

ASA J. WILBOURN, MD

EMG Laboratory, Cleveland Clinic, Cleveland, Ohio, USA

- Poorly defined entity
  - Neurogenic, vascular (arterial, venous)
- Symptomatology widespread
- How can a neurogenic process evade EDX?
- What does the Roos' test really test for?
- Problem could be settled only with controlled studies
  - Roos has refused to do this, considers it unethical

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Cochrane Library

Cochrane Database of Systematic Reviews

Treatment for thoracic outlet syndrome (Review)

Povlsen B, Hansson T, Povlsen SD

2015

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Cochrane - background

- TOS is one of the most controversial diagnoses in medicine.
- The term TOS represents three related syndromes
  - Brachial plexus is compressed
  - Major blood vessels of the upper chest are compressed
  - Painful non-specific or disputed TOS

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Cochrane - conclusions

- This review was complicated by a lack of generally accepted diagnostic criteria for the diagnosis of TOS.
- No evidence from RCTs for the use of other currently used treatments.
- Low-quality evidence that transaxillary first rib resection decreased pain more than supraclavicular neuroplasty,
  - No randomized evidence that either is better than no treatment

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Conlusions on TOS

- There is a need for an agreed definition
  - Diagnosis of TOS, especially the disputed form,
  - Outcome measures
  - Randomized trials that compare the outcome of interventions with no treatment and with each other.

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Neurological Sciences 2017:38:383-388

Neurol Sci (2017) 38:383–388
DOI 10.1007/s10072-016-2794-4

REVIEW ARTICLE

Thoracic outlet syndrome: wide literature for few cases. Status of the art

Pietro Emiliano Doneddu[1] · Daniele Coraci[2,3] · Paola De Franco[2] · Ilaria Paolasso[2] · Pietro Caliandro[1] · Luca Padua[1,2]

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Conclusions

- TOS subject to extensive litterature debate
  - Out of proportion to its actual incidence
- Neurophysiologists and neurologists
  - Sceptial, many deny its excitence
- Surgeons
  - Claim its common and responsive to surgery
- True neurogenic TOS
  - Very rare 1/1000000/year
  - Must be based on obejctive findings

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Own experience

- TOS over diagnosed in surgical specialities
- I have seen more complications following surgery for TOS than true neurogenic TOS patients
- Chronic compression of plexus brachialis is *very rare*
  - Following deformed clavicular fracture
  - Very rarely in association with cervical rib

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Iatrogenic plexus lesions

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Iatrogenic

- **Complication of plexus anesthesia**
- **Radiation therapy**
- Hematoma from transaxillary percutaneous angiograms
- Perioperative
  - Surgery in the neck region
  - Thoracotomy
  - **Complications of surgery for thoracic outlet syndrome**

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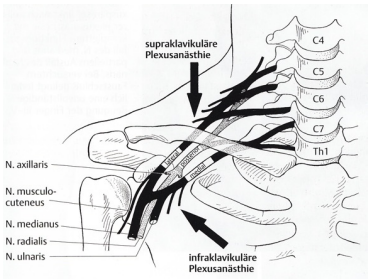
Plexus antesthesia



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Plexus antesthesia

- Localization of plexus
  - Ultrasound
  - Electrical stimulation
  - Mechanical paresthesia caused by needle
- 0,5% incidence of neuropathies

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Pathophysiology

- Direct laceration or minor trauma by the needle
- Toxic effect of the anesthetic
  - Type of drug, adrenaline, preservatives
- Intraneural injection of drugs
  - Intrafascicular - epineural injections

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Arteriography

- Direct trauma by catether
- Hematoma

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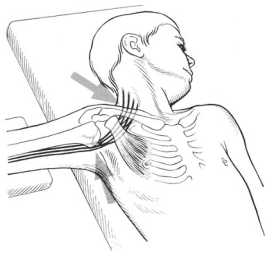
Perioperative plexus lesions



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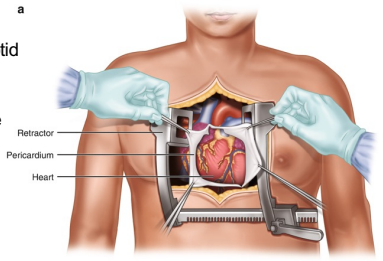
Sternotomy

- Occurs in 1-5% in carotid bypass sugery
- Inferior trunk
- Usually, good outcome



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Cancer

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Cancer

- Tumours
  - Lung cancer, Pancoast tumour
  - Metastasis of the lymph nodes
- Radiotherapy for cancer

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Brachial plexopathy after radiation therapy for breast cancer

- Dose related
  - TD_5 57-61 Gy
  - TD_{50} 68-73 Gy
- Onset 2,5 years to 20 years after radiation
- 4% at 5 years
- 25 % at 10 years
- 30 % at 15 years
- 55 % at 20 years
- Chemotherapy does not increase rate

Bajrovic A et al. Is there a life long risk of brachial plexopathy after radiotherapy of supraclavicular lymph nodes in breast cancer patients. Radiology and oncology. 2004:71:297-301

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Benign tumors

- Schwannnomas
- Neurofibromas

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Diagnosis of brachial plexopathies

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Diagnosis

- History
- Clinical findings
- EMG and ENG
- Imaging
  - MRI
  - Ultrasound
  - CT myelography

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Practical issues

- Brachial plexus lesions are rarely focal
  - Traction injuries extend diffusely along the nerves
  - Gunshot wound are diffuse
- Parsonage – Turner syndrome often affects individual nerves
  - May be multifocal
- Plexus anatomy varies
- EMG does not give etiology
  - History
  - Clinical findings
  - Imaging findings

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History

- Time course
  - When did it start?
  - Acute onset?
  - Progression?
- Sensory symptoms
  - Paresthesias
  - Pain
- Weakness
- Predisposing factors
  - Diabetes, rheumatoid arthritis, cancer
  - Clavicular factures
  - Trauma

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Clinical examination

- Testing muscle strength
- Tendon reflexes
- Horner's sign (severe lesions of C8-Th1 spinal nerves)
  - Miosis, ptosis and anhidrosis

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Demonstrate

- Neurogenic EMG findings in affected muscles
- Abnormal neurography
  - Reduced M wave amplitude
  - F wave latency and number
  - Reduced sensory nerve responses
  - Demonstrate local nerve conduction abnormality
- Differentiate from radiculopathy
  - Differential diagnosis is not always simple

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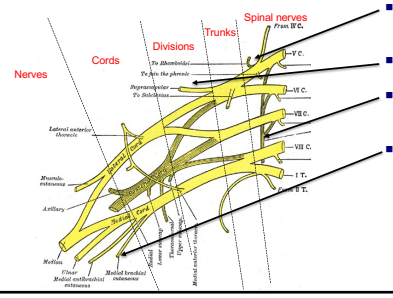
Plexus brachialis



- Dorsal scapular nerve to rhomboid muscles leaves from spinal nerve C5
- Suprascapular nerve leaves from upper trunk
- Long thoracic nerve leaves from spinal nerves C5, C6 and C7 proximally
- Medial antebrachial cutaneous nerve leaves from medial cord

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EMG

- M.rhomboideus (spinal nerve C5)
- M.infraspinatus/m.supraspinatus (upper trunk)
- M.deltoideus (upper trunk - posterior cord)
- M.biceps brachii (upper trunk - lateral cord)
- M.triceps (middle trunk – posterior cord)
- M.flexor carpi radialis (middle trunk – lateral cord)
- M.extensor indicis (lower trunk – posterior cord)
- M.interosseus dorsalis (lower trunk – medial cord)
- M.opponens pollicis (lower trunk – lateral+medial cord)
- M.serratus anterior (spinal nerves C5-C7)
- Paravertebral muscles C5-Th1

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Neurography

- Sensory
  - Median nerve digits 1-4
  - Ulnar nerve digits 4-5
  - Radial nerve
  - Lateral cutaneous of the forearm
  - Medial cutaneus of the forearm
- Motor (F-waves included)
  - Median
  - Ulnar

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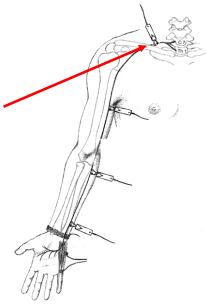
Ulnar motor neurography

High stimulus intensity 80-100 mA, 0.2 ms duration



Stöhr & Bluthart

"Atlas der klinischen Elektromyographie und Neurographie" Kohlhammer 1983

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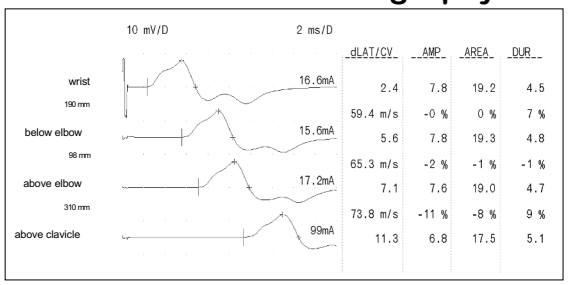
Ulnar nerve motor neurography



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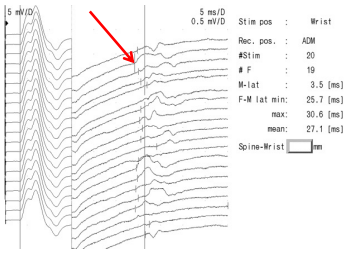
F waves



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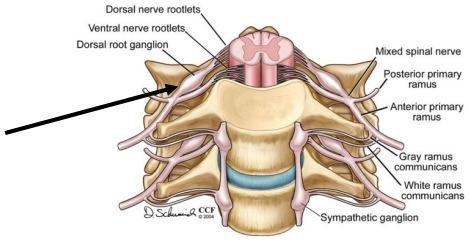
Sensory ganglion



Ferrante MA. Brachial plexopathies: Classification, causes and consequences. Muscle and Nerve 2004;30:547-568

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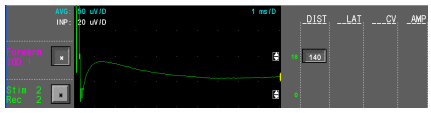
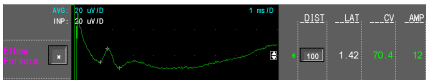
Sensory nerve action potential

- Low amplitude in plexopathies and peripheral neuropathies



- Normal/slightly reduced amplitude in radiculopathies



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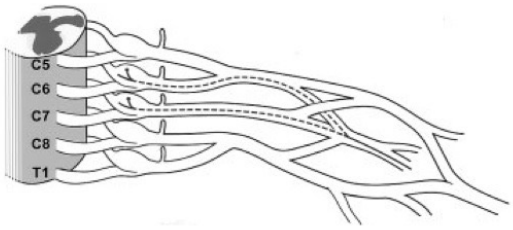
Radial nerve



Ferrante MA. Brachial plexopathies: Classification, causes and consequences. Muscle Nerve 2004;30:547

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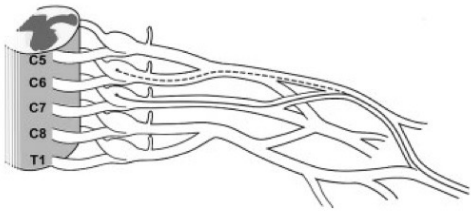
Median nerve to index finger



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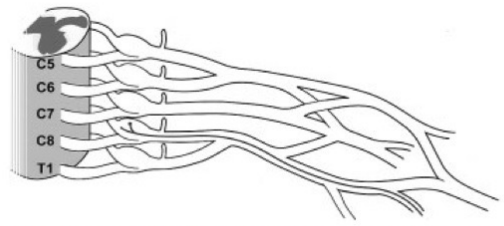
Ulnar nerve to little finger



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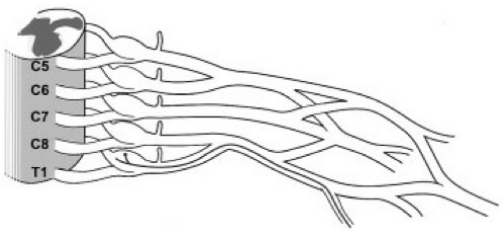
Medial cutaneous of forearm



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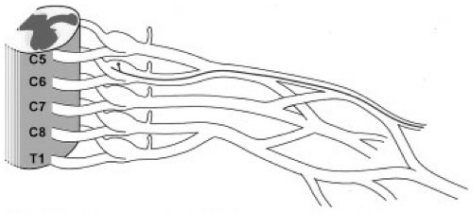
Lateral cutaneous of forearm



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ELECTRODIAGNOSTIC FEATURES OF TRUE NEUROGENIC THORACIC OUTLET SYNDROME

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Accepted 19 August 2013

ABSTRACT: *Introduction*: We report the electrodiagnostic (EDX) features of 32 patients with surgically verified true neurogenic thoracic outlet syndrome (TN-TOS). *Methods*: Retrospective record review. *Results*: We found uniform EDX evidence of a chronic axon loss process that affected the lower portion of the brachial plexus and disproportionately involved the T1 more than the C8 sensory and motor fibers. Because of this relationship, the medial antebrachial cutaneous sensory nerve (T1) and median motor (T1 > C8) study combination was abnormal in 89%, whereas response combinations that primarily assessed the C8 fibers were less frequently affected. *Conclusions*: The characteristic EDX features of TN-TOS are T1 > C8 nerve fiber involvement. A comprehensive EDX examination of the lower plexus with contralateral comparison studies is imperative to diagnose this disorder accurately.

Muscle Nerve **49**: 724–727, 2014

- Inferior trunk
- C8 and Th1 muscles
  - Interosseus dorsalis
  - Thenar muscles
  - Hypothenar muscles
  - Extensor indicis
- Sensory neurography
  - Ulnar
  - Medial antebrachial

77

Root avulsion

- Proximal to sensory ganglion
- Loss of sensation
- Preserved sensory nerve action potential
- Severe denervation of muscles
- Paraspinal muscles denervated
- No recovery expected

78

Twenty patients with thoracic outlet syndrome (TOS) seen at the Mayo Clinic between October 1984 and November 1985 were studied prospectively with routine nerve conduction studies, concentric needle examination, and bilateral median and ulnar somatosensory evoked potentials (SEPs). Results of nerve conduction studies and needle examination were abnormal in 30% of the patients, one patient having a reduced ulnar sensory nerve action potential amplitude and five others having neurogenic motor unit potential changes in the hand muscles. Ulnar SEPs were abnormal in three patients (15%), and median SEPs were abnormal in one patient, who also had abnormalities in ulnar SEPs. In patients with TOS, routine nerve conduction studies and needle examination were the most helpful electrophysiologic studies in excluding more common conditions. The routine use of ulnar SEPs in the evaluation of patients with TOS is probably not worthwhile.

MUSCLE & NERVE 11:571–575 1988

SOMATOSENSORY EVOKED POTENTIALS: LACK OF VALUE FOR DIAGNOSIS OF THORACIC OUTLET SYNDROME

MARTIN VEILLEUX, MD, J. CLARKE STEVENS, MD, and J. KEITH CAMPBELL, MD

SEPs are not helpful in the diagnosis of plexopathies

79

MRI and US in the study of plexus brachialis

- Differential diagnosis with radiculopathies
- Root avulsions in traumatic plexopathies
- Tumors of brachial plexus
- Pancoast tumor
- Often focal changes in Parsonage-Turner syndrome

80

Differential diagnosis

- Acute cervical radiculopathy due to disc herniation
- Borreliosis related polyradiculopathy
  - Often combined with facial palsy
- Mononeuritis multiplex
- Immune mediated neuropathies
  - Lewis-Sumner
  - Multifocal motor neuropathy with conduction blocks
- Infectious neuro(no)pathies
  - Herpes zoster
  - Tick borne encephalitis

81

Patient

82

History

- 72 year old woman
- 18 years ago surgery for left breast cancer
- 5 years ago surgery for lymph node metastasis
- Twice radiation therapy left axilla
- For 6-7 months progressive weakness and pain in left arm

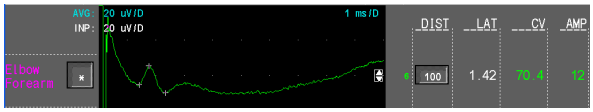
83

Clinical findings

- Atrophy of proximal shoulder muscles
- Weakness
  - Upper arm abduction
  - Elbow extension
  - Finger extension
- Reflexes
  - Biceps, brachioradialis normal
  - Triceps missing
- No obvious sensory abnormalities

84

Lateral cutaneous of the forearm



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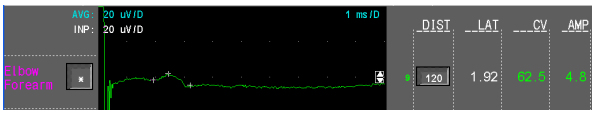
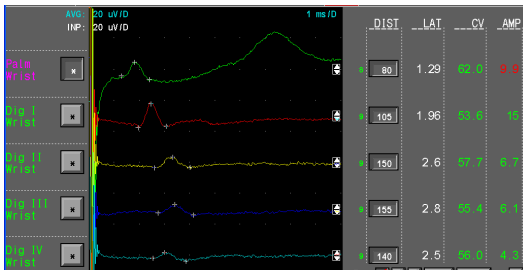
Medial cutaneous of forearm



86

Median nerve



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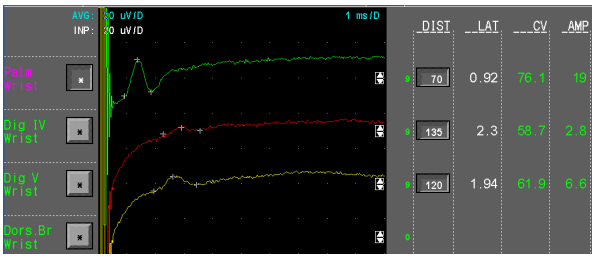
Ulnar nerve



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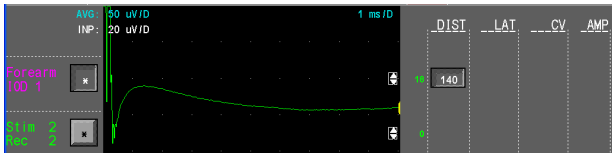
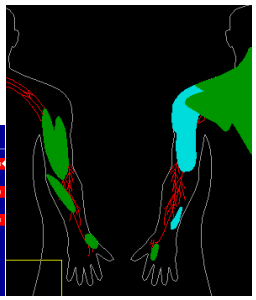
Radial sensory



89

EMG findings

Investigated muscles	
Left Deltoideus	: Uttal.kronisk neurop
Left Biceps	: Normal
Left Triceps	: Uttal.kronisk neurop
Left Flex carpi rad	: Normal
Left Ext indicis	: Uttal.kronisk neurop
Left Inteross dors I	: Normal
Left Opponens pollicis	: Normal
Left Infraspinatus	: Normal
Left Supraspinatus	: Normal
Left Trapezius	: Normal



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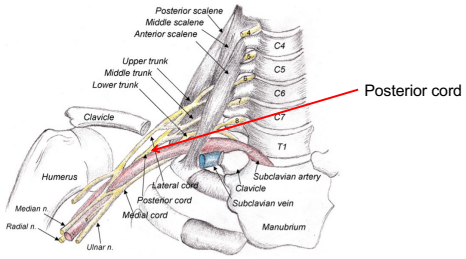
Where is the problem?

91

Plexus brachialis



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Conclusion

- Focal, severe, chronic lesion in the posterior cord
- Etiology
  - Metastasis likely
  - Parsonage-Turner syndrome unlikely, chronic progression
  - Radiation complication unlikely because the lesion is focal

93

Follow-up

- MRI of the brachial plexus showed tumor below the clavicle

94

Parsonage -Turner syndrome
Neuralgic amyotrophy

95

History

- Spillane JD. Localised neuritis of the shoulder girdle. Lancet 1943;ii:532–5
  - 46 patients with acute brachial plexopathy
    - "There is acute onset of pain in the shoulder and side of the neck, over the scapula and down the affected arm usually not further than the elbow…The pain is commonly worse at night, disturbing or preventing sleep…and usually persists for a week or 10 days…little or no general upset, no stiffness of the neck and upper limb reflexes may remain unaltered. After a few days or when the pain is subsiding, paralysis supervenes. The muscles most commonly affected are the serratus, the spinati, the deltoid and the trapezii in that order. The neuritis may be bilateral…Sensory loss is as a rule slight or absent; a small zone of hypo-aesthesia over an affected deltoid muscle…The prognosis is good but affected muscles often waste rapidly and recovery is slow…a year or more. Rarely there may be a recurrence in the same or opposite shoulder."
- Turner JWA. Acute brachial radiculitis. BMJ 1944;2:592–4
- **Parsonage MJ, Turner JWA. Neuralgic amyotrophy: the shoulder-girdle syndrome. Lancet 1948;1:973–8.**

96

Lancet 1948:251:973-978

NEURALGIC AMYOTROPHY

THE SHOULDER-GIRDLE SYNDROME

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A SYNDROME comprising pain and flaccid paralysis of the muscles round the shoulder girdle occurred fairly often during the war years 1941–45, though previously it had been rare. We observed 136 cases during neurological work in the Army in the United Kingdom and in India Command.

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SUMMARY

A syndrome consisting in severe pain across the shoulder and upper arm, followed by atrophic paralysis of muscles round the shoulder girdle, is described.

On clinical grounds it is thought that the pathological process can involve one or more peripheral nerves, two or more spinal roots, or the spinal cord.

The condition appears to be a distinct clinical entity which became increasingly common during the war years.

A similar syndrome may occur some days after the injection of serum, and the two conditions are probably identical, though the ætiology is unknown.

The name "neuralgic amyotrophy" is suggested.

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Terminology

- **Neuralgic amyotrophy**
  - Most commonly used term today
  - 5-10% do not have pain
  - Purely sensory nerves may be affected
- Plexus neuritis
  - Traditionally used previously
  - Affects also other structures
- Multifocal multifascicular inflammatory and constrictive brachial neuritis
  - Often very focal, limited to a few fascicles of a nerve
  - Fascicular entrapment (Vastamäki)
- Wartenberg's migrant sensory neuritis
  - Purely sensory neuropathy
- **Parsonage – Turner syndrome**

99

Classical PTS - Symptoms

- Acute onset with pain
- 95% onset with severe pain
- Often onset at night
- Initial pain usually a few weeks (1 day - 2 months)
  - 5% 24 hours
  - 10% > 2 months
- Pain is constant, not related to position
- Pain is often not in the same region as the neurological deficit
- Neurologic deficits become evident some time after onset of pain
  - Weakness of muscles innervated by affected nerve
  - Sensory abnormalities

100

Atypical PTS

- No pain
  - 5-10% without pain
- Onset with paresis followed by pain
  - 5 %
- Pure sensory neuropathy
- Involvement of lower extremity nerves
  - Mostly proximal

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Affected nerves

- Spinal nerves (nerve roots)
- Plexus brachialis
  - Any part may be affected
- Nerves in the shoulder
  - Long thoracic nerve
  - Suprascapular nerve
- Nerves in the arm
  - Anterior interosseus nerve
  - Posterior interosseus nerve
  - Lateral cutaneous of the forearm
  - Radial nerve sensory branch
  - Individual palmar digital nerves

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Affected nerves

- Individual distal nerve branches
  - Individual muscles or sensory nerve branches
- Cervical plexus
  - Phrenic nerve (10%)
- Cranial nerves
  - VII (Facial nerve)
  - IX (Glossopharyngeal nerve)
  - X (Vagus nerve or its branches)
  - XI (Accessory nerve)
  - XII (Hypoglossal nerve)
- Individual leg nerves - sometimes, not often
  - Lumbar plexus
  - Proximal leg nerves

103

PTS distribution

- Mononeuropathies
- Often multiple mononeuropathies
  - *Lesions appear sequentially at different times*
- Unilateral 75%
- Bilateral 25%
- Dominant > non-dominant arm

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PTS incidence: PLOS ONE 2015; May:1-9

RESEARCH ARTICLE

Incidence of Neuralgic Amyotrophy (Parsonage Turner Syndrome) in a Primary Care Setting - A Prospective Cohort Study

Nens van Alfen[1]*, Jeroen J. J. van Eijk[2], Tessa Ennik[2], Sean O. Flynn[4], Inge E. G. Nobacht[5], Jan T. Groothuis[6], Sigrid Pillen[7], Floris A. van de Laar[8]

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Incidence in primary care 1/1000/year

105

PTS age and gender

- Children and adults
  - Range 0 - 90 years
  - Mean age of onset 40 years
- 10% have a family history

106

PTS constitutional risk factors

- Lean > obese
- Male : female = 2:1

Constitutional risk factors for focal neuropathies in patients referred for electromyography

C. Martínez-Aparicio[a,b], S. K. Jääskeläinen[c], L. Puksa[d], F. Reche-Lorite[e], P. Torné-Poyatos[f], J. Paniagua Soto[g] and B. Falck[h]

[a]Doctoral Programme of Medicine and Public Health, University of Granada, Granada; [b]Department of Clinical Neurophysiology, Vithas Virgen del Mar Hospital, Almería, Spain; [c]Department of Clinical Neurophysiology, Turku University Hospital and University of Turku, Turku, Finland; [d]Department of Clinical Neurophysiology, University of Tartu, Tartu, Estonia; [e]Department of Mathematics, University of Almería, Almería; [f]Faculty of Medicine, University of Granada, Hospital Universitario Clínico, Granada; [g]Department of Clinical Neurophysiology, University Hospital Virgen de las Nieves, Granada, Spain; and [h]Department of Clinical Neurophysiology, University Hospital, Uppsala, Sweden

European Journal of Neurology 2020, 27: 529–535

107

Recurrence

- 20% one recurrence
- 5 % two recurrences
- 4 % three recurrences
- 1 % four or more recurrences

30% will recur!

Van Alfen N, van Engelen BGM. The clinical spectrum of neuralgic amyotrophy in 246 cases. Brain 2006;129:438-450

108

Antecedent events to PTS

- Infection
  - 10% have hepatitis E
  - Coxacie A2, Influenza
  - Covid 19 not more prone than other virus infections
- Surgery
- Childbirth
- Unusual physical activity
- Vaccination
  - Covid 19 vaccines not more prone than other vaccines
- Trauma
- No identified antecedent event 30-40%

- May start within hours of antecedent events
- Onset usually within 2 weeks of predisposing event

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Nerve biopsy findings

- Epineural perivascular mononuclear T-cell infiltration
- Active multifocal axonal degeneration without blood vessel wall inflammation or necrosis
- Perineural thickening
- T-Cell mediated and/or B-Cell mediated(Postinfectious nature)

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Pathophysiology of PTS

- Immune triggers
- Genetic predisposition
  - Hereditary neuralgic amyotrophy (HNA – SEPT9 mutation)
  - Other yet unknown mutations
  - Tendency for recurrent episodes
- Mechanical factors
  - Lesions often around joints
  - Preceding repetitive physical activity, dominant arm > non dominant arm
- *Organ specific immune mediated disorder*

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ULTRASONOGRAPHIC IDENTIFICATION OF NERVE PATHOLOGY IN NEURALGIC AMYOTROPHY: ENLARGEMENT, CONSTRICTION, FASCICULAR ENTWINEMENT, AND TORSION

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Accepted 10 February 2015

ABSTRACT: *Introduction:* The aim of this study was to characterize the ultrasonographic findings on nerves in neuralgic amyotrophy. *Methods:* Fourteen patients with neuralgic amyotrophy were examined using high-resolution ultrasound. *Results:* Four types of abnormalities were found: (1) focal or diffuse nerve/fascicle enlargement (57%); (2) incomplete nerve constriction (36%); (3) complete nerve constriction with torsion (50%: hourglass-like appearance); and (4) fascicular entwinement (28%). Torsions were confirmed intraoperatively and were seen on the radial nerve in 85% of patients. A significant correlation was found between no spontaneous recovery of nerve function and constriction/torsion/fascicular entwinement ($P = 0.007$). *Conclusion:* Ultrasonographic nerve pathology in neuralgic amyotrophy varies in order of severity from nerve enlargement to constriction to nerve torsion, with treatment ranging from conservative to surgical. We postulate that the constriction caused by inflammation is the precursor of torsion and that development of nerve torsion is facilitated by the rotational movements of limbs.

Muscle Nerve **52**: 503–511, 2015

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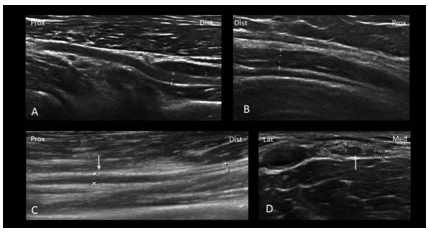
Nerves with edema



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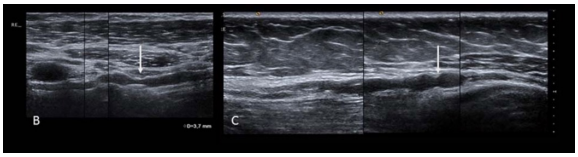
Hour glass constriction



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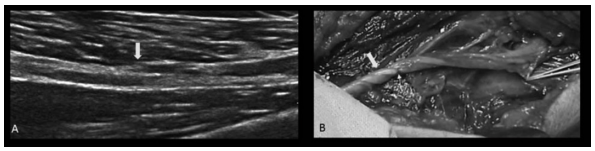
Entwinement/torsion



Ultrasound Intraoperative

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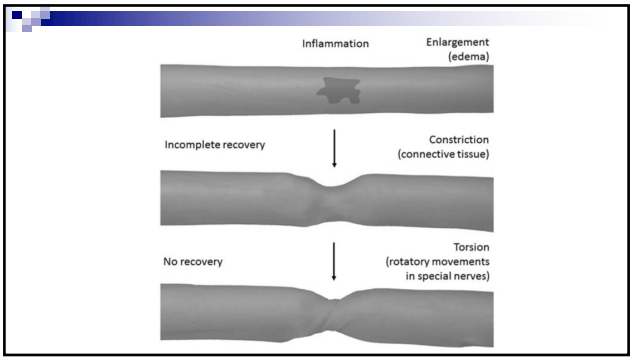

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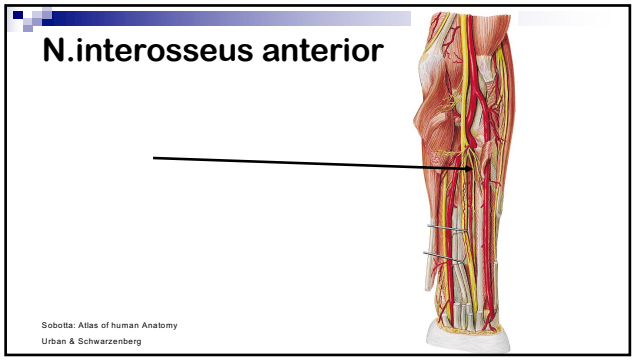

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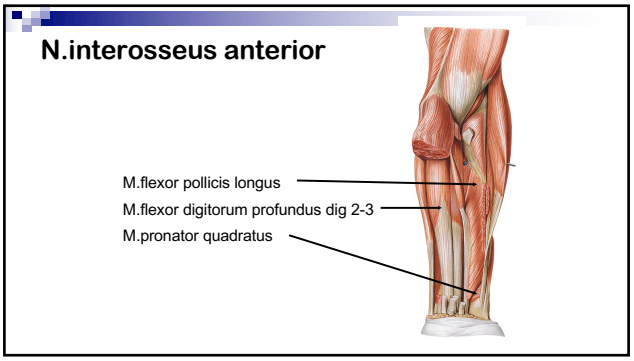

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N.interosseus anterior

- Severe pain in the forearm/upper arm/shoulder
  - 2 days to 2 months
- Distal phalanx of thumb and forefinger flexion weak
- Variable recovery
- Parsonage Turner syndrome (neuralgic amyotrophy)
- ***Anterior interosseus syndrome is not an entrapment***

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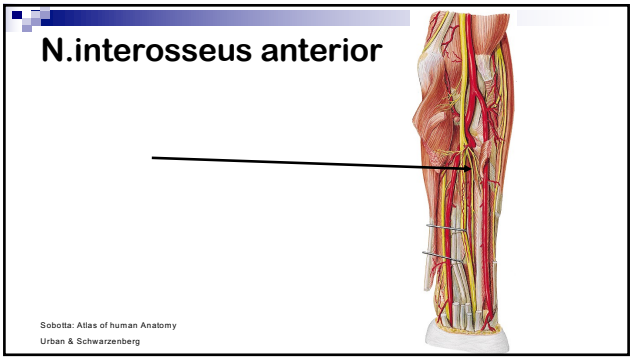

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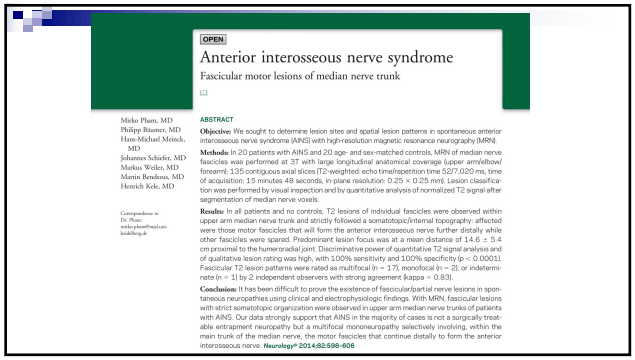

OPEN

Anterior interosseous nerve syndrome

Fascicular motor lesions of median nerve trunk

Mirko Pham, MD
Philipp Bäumer, MD
Hans-Michael Meinck, MD
Johannes Schiefer, MD
Markus Weiler, MD
Martin Bendszus, MD
Henrich Kele, MD

ABSTRACT

Objective: We sought to determine lesion sites and spatial lesion patterns in spontaneous anterior interosseous nerve syndrome (AINS) with high-resolution magnetic resonance neurography (MRN).

Methods: In 20 patients with AINS and 20 age- and sex-matched controls, MRN of median nerve fascicles was performed at 3T with large longitudinal anatomical coverage (upper arm/elbow/forearm): 135 contiguous axial slices (T2-weighted: echo time/repetition time 52/7,020 ms, time of acquisition: 15 minutes 48 seconds, in-plane resolution: 0.25 × 0.25 mm). Lesion classification was performed by visual inspection and by quantitative analysis of normalized T2 signal after segmentation of median nerve voxels.

Results: In all patients and no controls, T2 lesions of individual fascicles were observed within upper arm median nerve trunk and strictly followed a somatotopic/internal topography: affected were those motor fascicles that will form the anterior interosseous nerve further distally while other fascicles were spared. Predominant lesion focus was at a mean distance of 14.6 ± 5.4 cm proximal to the humeroradial joint. Discriminative power of quantitative T2 signal analysis and of qualitative lesion rating was high, with 100% sensitivity and 100% specificity ($p < 0.0001$). Fascicular T2 lesion patterns were rated as multifocal ($n = 17$), monofocal ($n = 2$), or indeterminate ($n = 1$) by 2 independent observers with strong agreement (kappa = 0.83).

Conclusions: It has been difficult to prove the existence of fascicular/partial nerve lesions in spontaneous neuropathies using clinical and electrophysiologic findings. With MRN, fascicular lesions with strict somatotopic organization were observed in upper arm median nerve trunks of patients with AINS. Our data strongly support that AINS in the majority of cases is not a surgically treatable entrapment neuropathy but a multifocal mononeuropathy selectively involving, within the main trunk of the median nerve, the motor fascicles that continue distally to form the anterior interosseous nerve. **Neurology® 2014;82:598–606**

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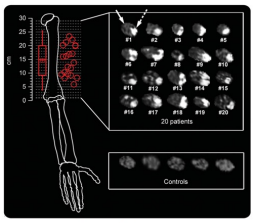
Anterior interosseus neuropathy



Pham et al. Anterior interosseous nerve syndrome. Neurology® 2014;82:598–606

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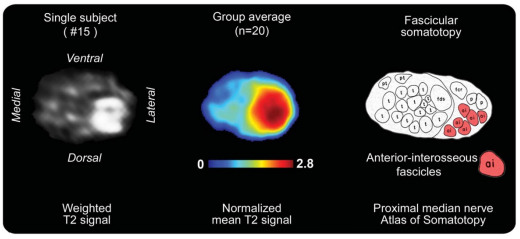
Anterior interosseus neuropathy



Pham et al. Anterior interosseous nerve syndrome. Neurology® 2014;82:598–606

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N.thoracicus longus

- Winging of the scapula
- Difficulty of abduction of the arm above the shoulder
- Slow recovery
  - axonal reinnervation starts at 6-8 months after onset
  - recovery completed at two years after onset

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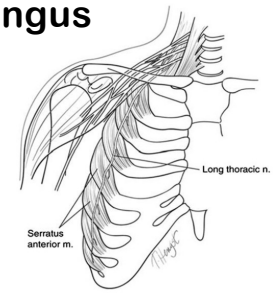
N.thoracicus longus



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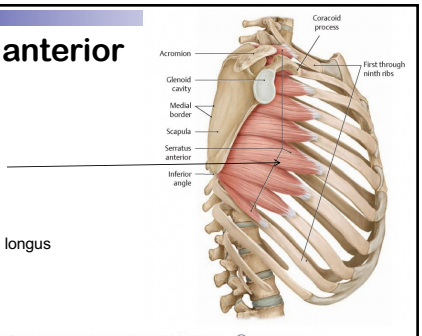
M.serratus anterior



Innervation: N.thoracicus longus
Myotomes: C5-7

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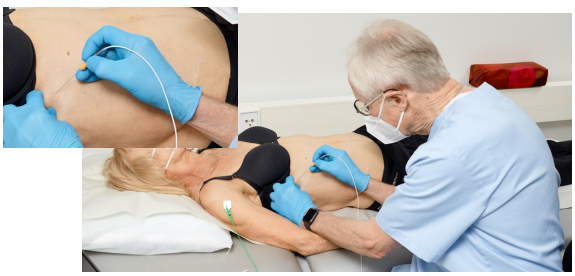
M.serratus anterior



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M.serratus anterior - activation

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Long thoracic nerve neuropathy on the right side
9/2009, onset after childbirth 7/2009

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Long thoracic nerve neuropathy on the right side
9/2009, onset after childbirth 7/2009

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Long thoracic nerve neuropathy

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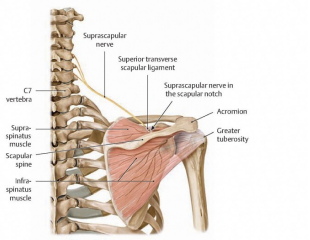
Suprascapular nerve



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N.suprascapularis

- Shoulder pain
- Weak upper arm outward rotation
- Weak shoulder abduction
- Atrophy of m.infraspinatus and m.supraspinatus

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Suprascapular neuropathy

- 4 % of patients with shoulder pain
- Trauma
- Parsonage-Turner syndrome

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Hereditary neuralgic amyotrophy (HNA)

- Genetically heterogeneous
- One form linked to chromosome 17q24
  - Gene codes septin 9 (SEPT9)
  - Septins are implicated in the formation of the cytosceleton, cell division and tumorgenesis
  - Missense mutations
  - Intragenic duplications
- Autosomal dominant inheritance
- Variable penetrance
- Onset often in early childhood

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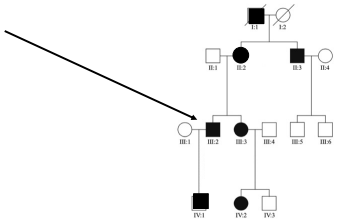
Pedigree of own HNA family



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EMG findings in HNA

- In unaffected parts normal EMG and neurography
- Abnormalities only in affected nerves
- Different from HNPP (hereditary liability to pressure palsies)

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Diagnosis of PTS

- Symptoms
- Clinical findings
- ENMG
- Imaging
  - US
  - MRI

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EMG

- Optimal timing 3 weeks following onset
  - Denervation in muscles is seen > 2-3 weeks from onset
- Many clinical neurophysiologists do US

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Imaging

- MRI of the plexus
  - Abnormalities often seen after 2-4 weeks
  - T2-weighted Dixon fat suppression/inversion recovery sequences, multiple planes
  - Intravenous gadolinium contrast at the discretion of the radiologist.
- Ultrasound
  - 6-18 MHz linear transducer
  - Very good in experienced hands
- Imaging not only the affected nerve, but also the parent nerve
- Choice of imaging method depends on availability/experience
- MRI better in deeply located nerves

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Muscle & Nerve. 2022;66:709–714

CLINICAL RESEARCH ARTICLE

MUSCLE&NERVE WILEY

Imaging of neuralgic amyotrophy in the acute phase

Paolo Ripellino MD, MSc[1] | Zsuzsanna Arányi MD, PhD[2] | Nens van Alfen MD, PhD[3] | Elisa Ventura MD[4] | Anne-Kathrin Peyer MD, PhD[5] | Alessandro Cianfoni MD, PhD[4,6] | Claudio Gobbi MD[1] | Emily Pedrick BA[7] | Darryl Brett Sneag MD[7]

- US/MRI showed oedema or hourglass constriction (HGC) in 90% within one month
- Earliest change with US in 12 hours with MRI 8 days
- HGC 4 patients in 1 week, 8 in 2 weeks, 5 within 3 weeks, 12 within 4 weeks

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Muscle Nerve 56: 1054–1062, 2017

ULTRASONOGRAPHY IN NEURALGIC AMYOTROPHY: SENSITIVITY, SPECTRUM OF FINDINGS, AND CLINICAL CORRELATIONS

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US findings

Table 1. Summary of affected nerves with respective ultrasonographic findings

		Ultrasonographic findings			
Affected nerves	Number (total)	None	Swelling without constriction	Incomplete constriction	Complete constriction
AIN	16 (7*)	2	10	3	1
Radial nerve / PIN	12 (1*)	1	2	3	6 (3†)
Long thoracic nerve	12	7	5	—	—
Suprascapular nerve	8	2	2	4	—
Accessory nerve	6	1	5	—	—
Axillary nerve	5	3	1	—	1
Musculocutaneous nerve	3	—	1	1	1 (1†)
Median nerve	2	—	2	—	—
Superficial radial nerve	2	—	1	—	1
LABCN	1	—	—	1 (1†)	—
Thoracodorsal nerve	1	1	—	—	—
Dorsal scapular nerve	1	1	—	—	—
Upper trunk	1	—	1	—	—
Total	70 (100%)	18 (26%)	30 (43%)	12 (1†) (17%)	10 (4†) (14%)

AIN, anterior interosseous nerve; PIN, posterior interosseous nerve; LABCN, lateral antebrachial cutaneous nerve.

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Treatment

- Analgesia
  - NSAID, opioids if necessary
- Immunomodulation
  - No controlled studies
  - Should be started acutely at the onset! Within 30 days from onset
    - Prednislone high dosage! 100 mg – 80 mg – 60 mg mg – 40 mg – 20 mg
    - IVIg
  - Shorter and better recovery times suggested in some uncontrolled studies
- Surgery
  - In severe cases with complete axonal involvement and torsion?
  - Future studies will soon show

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Outcome with conservative therapy

- Nens Van Alfen
  - 10% full recovery
  - 60% will have some pain
  - 50% fatigue
  - 25% unable to work
- Cruz-Martinez A et al J Peripheral Nervous System 2002:7:198-204
  - Good outcome in the 22 out of 40 patients that were followed for 2 years
  - In 41 of 43 affected nerves good outcome

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J Neurol Neurosurg Psychiatry 2009;80:1120–1124

Evaluation of prednisolone treatment in the acute phase of neuralgic amyotrophy: an observational study

J J J van Eijk,[1] N van Alfen,[1,2] M Berrevoets,[1] G J van der Wilt,[3] S Pillen,[2] B G M van Engelen[1]

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J Neurol Neurosurg Psychiatry 2009;80:1120–1124

- Observational study
- 50 patients treated, 203 historical controls
- Within 1 month from onset
- Prednisolone for 2 weeks
  - 60 mg/day for one week tapering 10 mg per day

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Table 2 Outcomes for the study (prednisolone) group (SG) and the historical controls (HC)

	Study group	Historical controls	
Median time (days) until initial pain relief (mean)	12.5 (17.1)	20.5 (37.2)	Not significant, p = 0.13
Recovery of strength within 1 month	9/50 (18.0%)	11/174 (6.3%)	p = 0.011
Full functional recovery within the first year	6/50 (12.0%)	2/189 (1.0%)	p<0.001
Good (but not full) self-reported recovery within			
6 months	16/50 (32%)	3/103 (2.9%)	p<0.001
12 months	22/50 (44.0%)	11/103 (10.7%)	p<0.001

- Shorter duration of pain
- Functional recovery of weakness earlier
- Better final outcome

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J Hand Surg Am 2021:46:43-53

EDITOR'S CHOICE

Outcomes of Microneurolysis of Hourglass Constrictions in Chronic Neuralgic Amyotrophy

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Joseph H. Feinberg, MD,* Ogonna K. Nwawka, MD,*††
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J Hand Surg Am 2021:46:43-53

- 24 patients with PTS with hour-glass constriction
- 11 treated with microsurgical epineurolysis or perineurolysis
  - 9 patients significant recovery
  - Time from onset to surgery 6-18 months
- 13 treated non-surgically
  - 3 patients significant recovery

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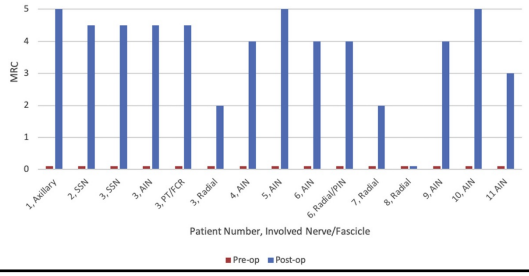
Operated patients – outcome



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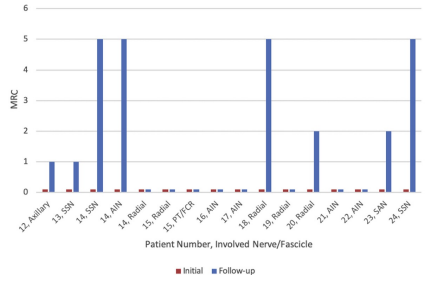
Conservatively treated patients -outcome



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Gstoettner C, et al. J Neurol Neurosurg Psychiatry 2020;91:879-

Neurosurgery

REVIEW

Neuralgic amyotrophy: a paradigm shift in diagnosis and treatment

Clemens Gstoettner [1], Johannes A Mayer,[1,2] Stephanie Rassam,[1,3] Laura A Hruby,[1,4] Stefan Salminger,[1,5] Agnes Sturma,[1,6] Martin Aman,[1,7] Leila Harhaus,[7] Hannes Platzgummer,[8] Oskar C Aszmann[1,5]

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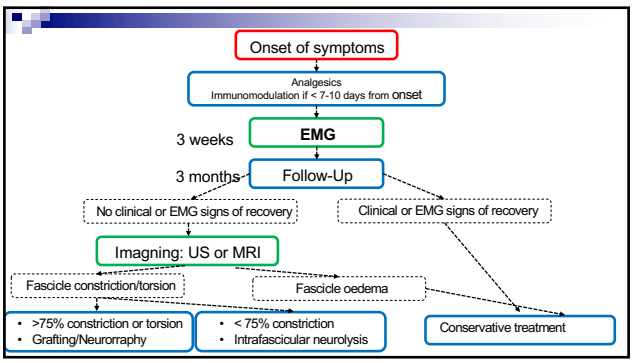

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Summary

- PTS is a common focal peripheral neuropathy
- Immune mediated focal neuropathy
- Most common neurological disorder causing shoulder pain
- Onset acute with severe pain
- May affect any peripheral nerve
  - Most often single nerves emerging from plexus brachialis
  - Rarely affects leg nerves
- 60-70% preceded by infection, trauma, surgery, etc
- Diagnosed with EMG, US/MRI
- Treatment in mild/moderate cases conservate, in severe cases surgery

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Patient

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50 year old male

- Type 2 diabetes for 7 years
- Ten years ago, an episode of pain first in in the right shoulder and then in a few days later in the left shoulder lasting for a few weeks. Not investigated at that time.
- Two months prior to EMG TBE vaccination
- One-week after TBE vaccination pain in the right shoulder at night (VAS7/10)
  - Weakness of right arm and shortness of breath
- One-week later pain in the left shoulder
  - Weakness of external rotation and abduction of upper arm

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50 year old male

- Consulted a private neurologist
  - No breathing sounds on right ride
  - Chest X-ray showed elevation of right diaphragm
  - Refers patient to department of lung medicine

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Investigations

- Cervical MRI
  - Slight age-related degenerative changes
- Chest X-ray
  - Elevated diaphragm on the right side

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50 year old male

- Examined at the department of lung medicine
- No cause for the problem found
- Referred for neurological consultation
- Before consultation neurologist refers patient for EMG
  - Neuromuscular disorder?

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Clinical findings

- Obese patient, BMI 34
- Right triceps reflex -, other tendon reflexes normal bilaterally
- Weakness of right elbow extension
- Weakness of left upper arm external rotation and abduction

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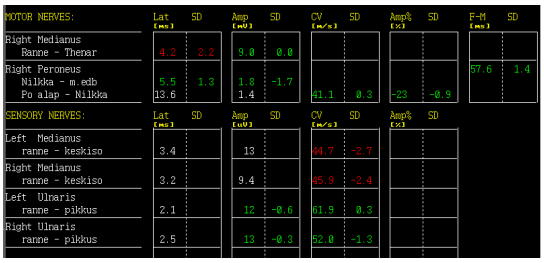
Neurography



MOTOR NERVES:	Lat (ms)	SD	Amp (mV)	SD	CV (m/s)	SD	Amp% (%)	SD	F-M (ms)	SD
Right Medianus										
Ranne - Thenar	4.2	2.2	9.0	0.0						
Right Peroneus									57.6	1.4
Nilkka - m.edb	5.5	1.3	1.8	-1.7						
Fo alap - Nilkka	13.6		1.4		41.1	0.3	-23	-0.9		

SENSORY NERVES:	Lat (ms)	SD	Amp (uV)	SD	CV (m/s)	SD	Amp% (%)	SD
Left Medianus								
ranne - keskiso	3.4		13		44.7	-2.7		
Right Medianus								
ranne - keskiso	3.2		9.4		45.9	-2.4		
Left Ulnaris								
ranne - pikkus	2.1		12	-0.6	61.9	0.3		
Right Ulnaris								
ranne - pikkus	2.5		13	-0.3	52.0	-1.3		

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Neurography

SENSORY NERVES:	Lat (ms)	SD	Amp (uV)	SD	CV (m/s)	SD	Amp% (%)	SD
Right Radialis								
kyynärv - ranne	2.5		13	0.1	56.0	-0.7		
Right Peroneus super								
sääri - metat 1	3.5		3.8	0.1	41.4	1.3		
Right Cut antebr lat								
kyynärt - kyynärv	--		--		--			
Right Cut antebr med								
kyynärt - kyynärv	1.92		1.5	-2.7	69.8	0.8		

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EMG findings – left side

Muscle	Fibrillations	MUP amplitude	Interference pattern	Interpretation
Trapezius				Normal
Deltoideus		⬆	⬇	Slight inactive neurogenic
Biceps brachii		⬆	⬇	Slight inactive neurogenic
Triceps				Normal
Extensor indics				Normal
Flexor carpi radialis				Normal
Interosseus dors 1				Normal
Pectoralis major				Normal
Supraspintus	10/10		⬇⬇⬇	Severe acute neurogenic
Infraspinatus	10/10		⬇⬇⬇	Severe acute neurogenic

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EMG findings – right side

Muscle	Fibrillations	MUP amplitude	Interference pattern	Interpretation
Trapezius				Normal
Deltoideus		⬆	⬇	Slight inactive neurogenic
Biceps brachii		⬆	⬇	Slight inactive neurogenic
Triceps	6/10		⬇⬇	Moderate acute
Extensor indics				Normal
Flexor carpi radialis	8/10		⬇⬇	Moderate acute
Interosseus dors 1				Normal
Diaphragm	10/10		0	Severe acute neurogenic
Vastus lateralis				Normal
Tibialis anterior				Normal

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EMG summary

- Moderate acute right C7 spinal nerve neuropathy
- Severe involvement of the right phrenic nerve
- Severe acute left suprascapular nerve neuropathy
- Bilaterally a mild old upper plexus lesion
- Bilateral findings of CTS, no subjective symptoms
- No diabetic polyneuropathy

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Diagnosis

- Recurrent Parsonage-Turner syndrome

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Outcome two years later

- Good recovery of muscle strength of right triceps
- Mild residual weakness of left infraspinatus
- Total paralysis of the right diaphragm
  - Able to walk without shortness of breath on even ground
  - Difficult to lie flat on the back

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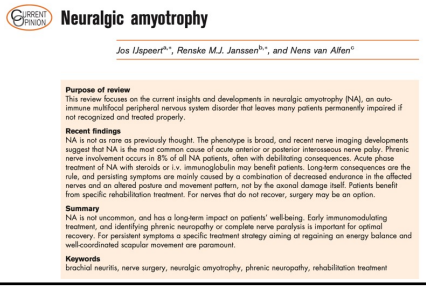
Curr Opin Neurol 2021, 34:605 – 612



Neuralgic amyotrophy

Jos IJspeert[a,*], Renske M.J. Janssen[b,*], and Nens van Alfen[c]

Purpose of review
This review focuses on the current insights and developments in neuralgic amyotrophy (NA), an autoimmune multifocal peripheral nervous system disorder that leaves many patients permanently impaired if not recognized and treated properly.

Recent findings
NA is not as rare as previously thought. The phenotype is broad, and recent nerve imaging developments suggest that NA is the most common cause of acute anterior or posterior interosseous nerve palsy. Phrenic nerve involvement occurs in 8% of all NA patients, often with debilitating consequences. Acute phase treatment of NA with steroids or i.v. immunoglobulin may benefit patients. Long-term consequences are the rule, and persisting symptoms are mainly caused by a combination of decreased endurance in the affected nerves and an altered posture and movement pattern, not by the axonal damage itself. Patients benefit from specific rehabilitation treatment. For nerves that do not recover, surgery may be an option.

Summary
NA is not uncommon, and has a long-term impact on patients' well-being. Early immunomodulating treatment, and identifying phrenic neuropathy or complete nerve paralysis is important for optimal recovery. For persistent symptoms a specific treatment strategy aiming at regaining an energy balance and well-coordinated scapular movement are paramount.

Keywords
brachial neuritis, nerve surgery, neuralgic amyotrophy, phrenic neuropathy, rehabilitation treatment

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Game over

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