DIAGNOSTICS

EMG

EMG - extracellular electrical activity recorded from muscle.

- EMG requires patient cooperation for full relaxation and maximal voluntary muscle contraction – EMG is less useful in pediatrics.

upward deflection indicates that active electrode is **negative** with respect to reference one

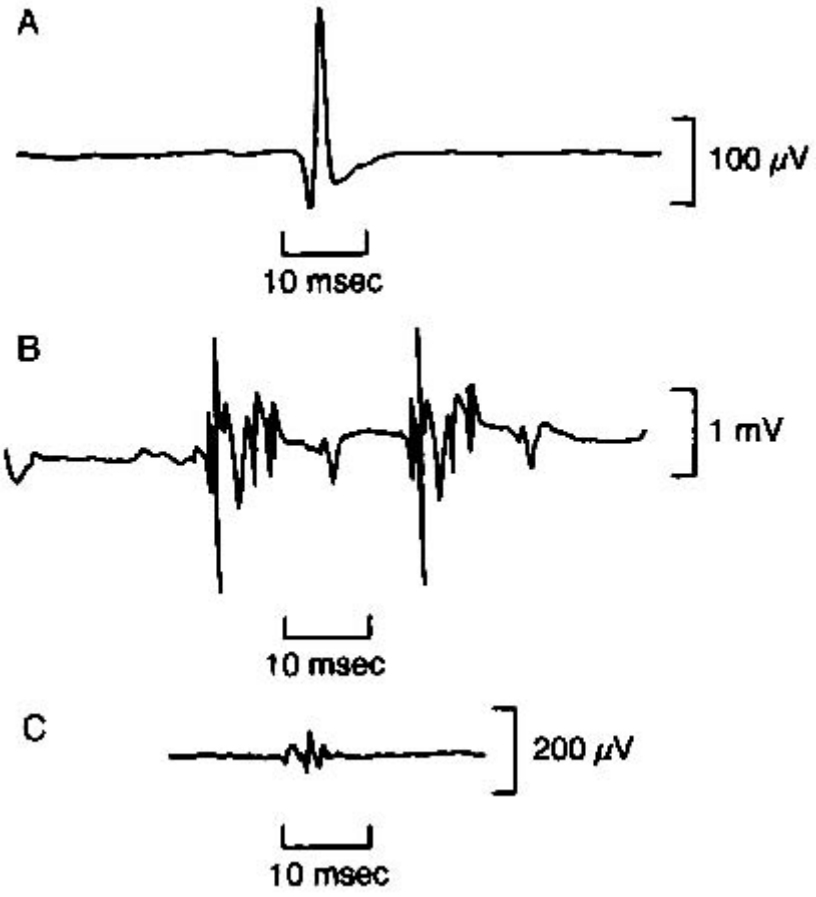
Abnormalities of MOTOR UNIT ACTION POTENTIALS

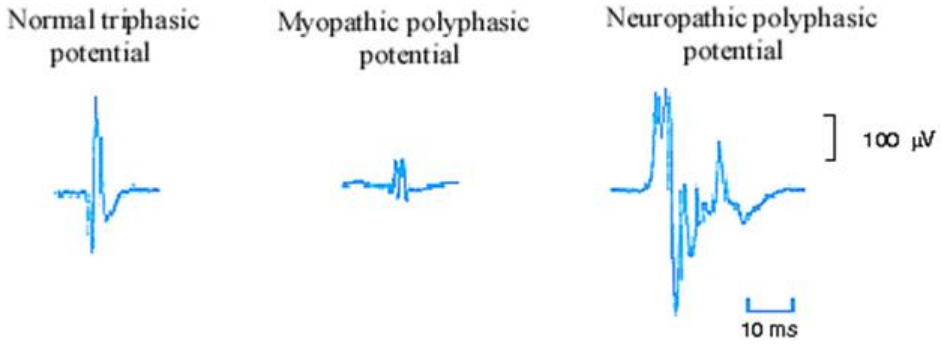
Motor unit action potentials (MAP).

A. Normal *biphasic* or *triphasic* potential.

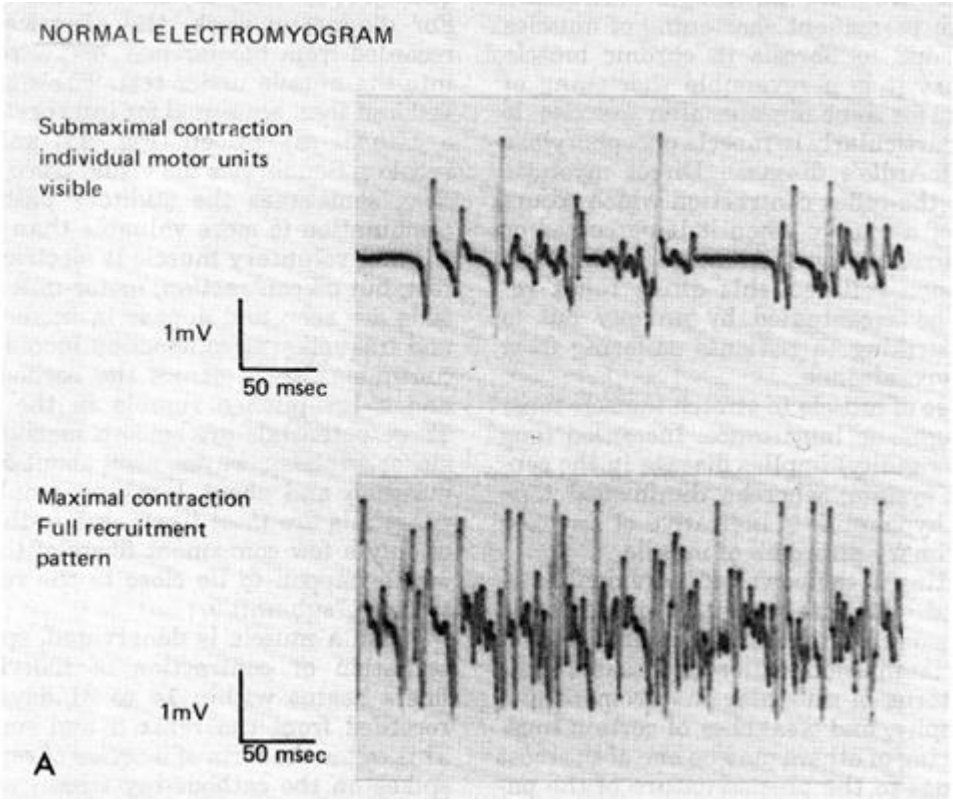
B. Long-duration, high amplitude *polyphasic* potential (shown twice) – *neuropathic potential*.

C. Short-duration, low-amplitude, *polyphasic* potential – *myopathic potential*.





Quantitative MAP analysis provides more reliable correlation with muscle and nerve disease than muscle biopsy!



Prolonged INSERTION ACTIVITY

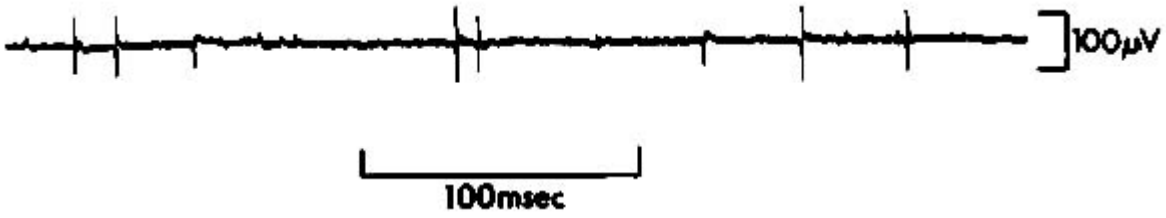
a) acute **denervation**
b) active **myopathy**

Normally: needle electrode is inserted → brief ***burst of activity*** for ≤ 2-3 seconds → ***no spontaneous activity***

Abnormal SPONTANEOUS ACTIVITY

Denervated muscle fibers discharge spontaneously!
→ **fibrillation potentials** and **positive sharp waves**.

Fibrillation potential - biphasic (or triphasic) discharge - action potential generated in ***single muscle fiber*** (so cannot be detected clinically):



Positive sharp waves - initial positive deflection → slow deflection in negative direction.

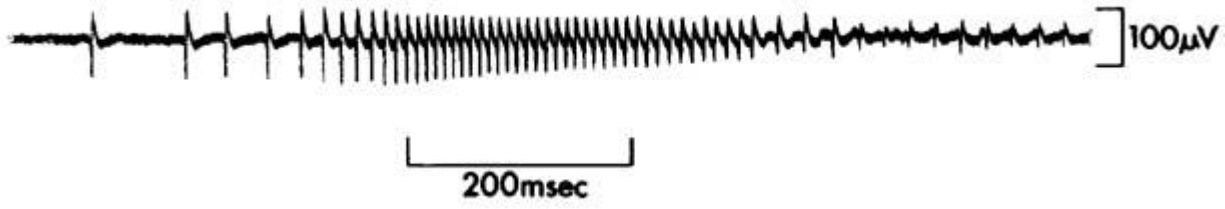
Spontaneous fibrillation potentials + positive sharp waves:

Fasciculation potential - spontaneous activation of ***all muscle fibers in motor unit***.

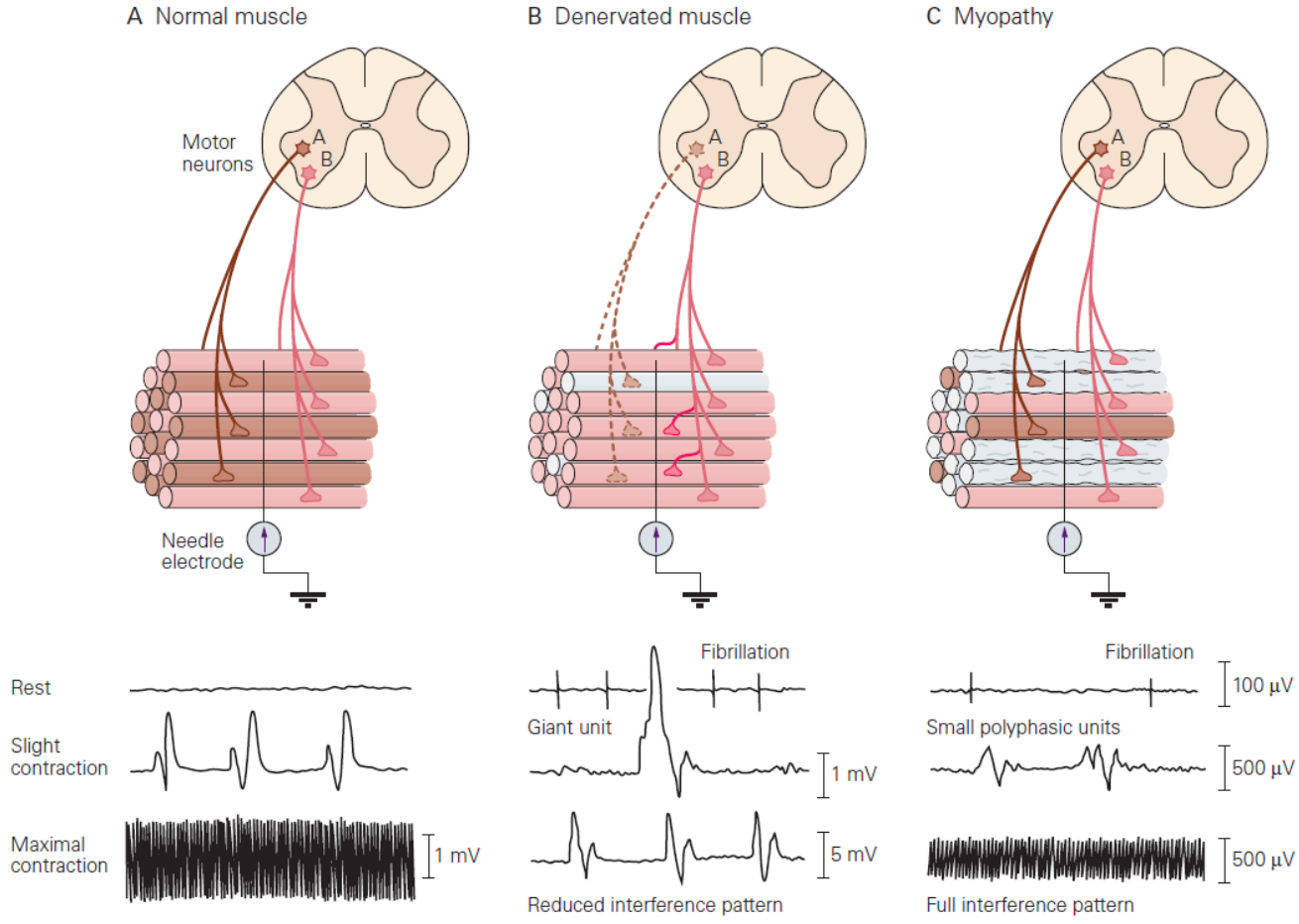
- indistinguishable from normal motor unit action potentials!
- amplitude & duration greater than fibrillation potential.
- etiology - **disease of anterior horn cells** (any site along motoneuron body - motor axon).

Myotonic discharges - spontaneous repetitive high-frequency **trains of action potentials** derived from ***single muscle fibers***; decreasing amplitude and frequency.

- etiology - **myotonic disorders**.
- sound myographically like "dive bomber".



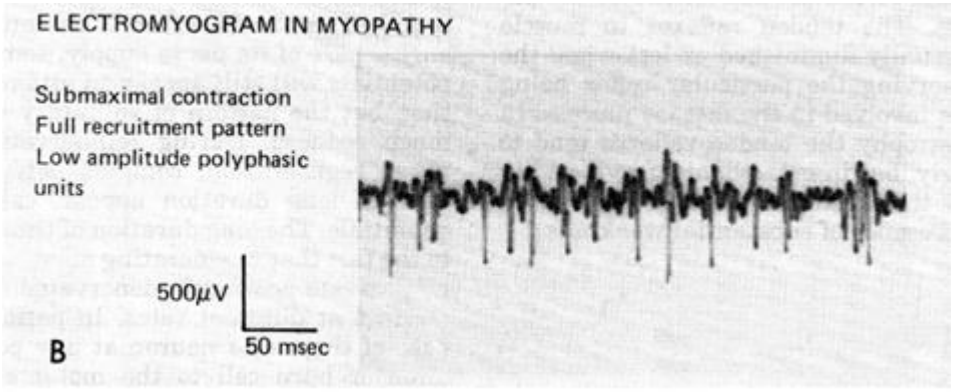
PATHOLOGIES



Myopathies (↓number of muscle fibers in individual motor units; number of motor units is normal):

1) INSERTIONAL ACTIVITY - **increased**.
2) SPONTANEOUS ACTIVITY - complex **polyphasic** motor unit potentials.
3) VOLUNTARY ACTIVITY - myopathic potentials (↓**duration & amplitude**); because individual motor units generate less tension than normal, ***increased number is recruited*** for any given degree of voluntary activity = **rapid recruitment** (**recruitment density is normal**, but envelope amplitude is reduced)

pathognomonic finding of myopathy: full recruitment in weak, wasted muscle.

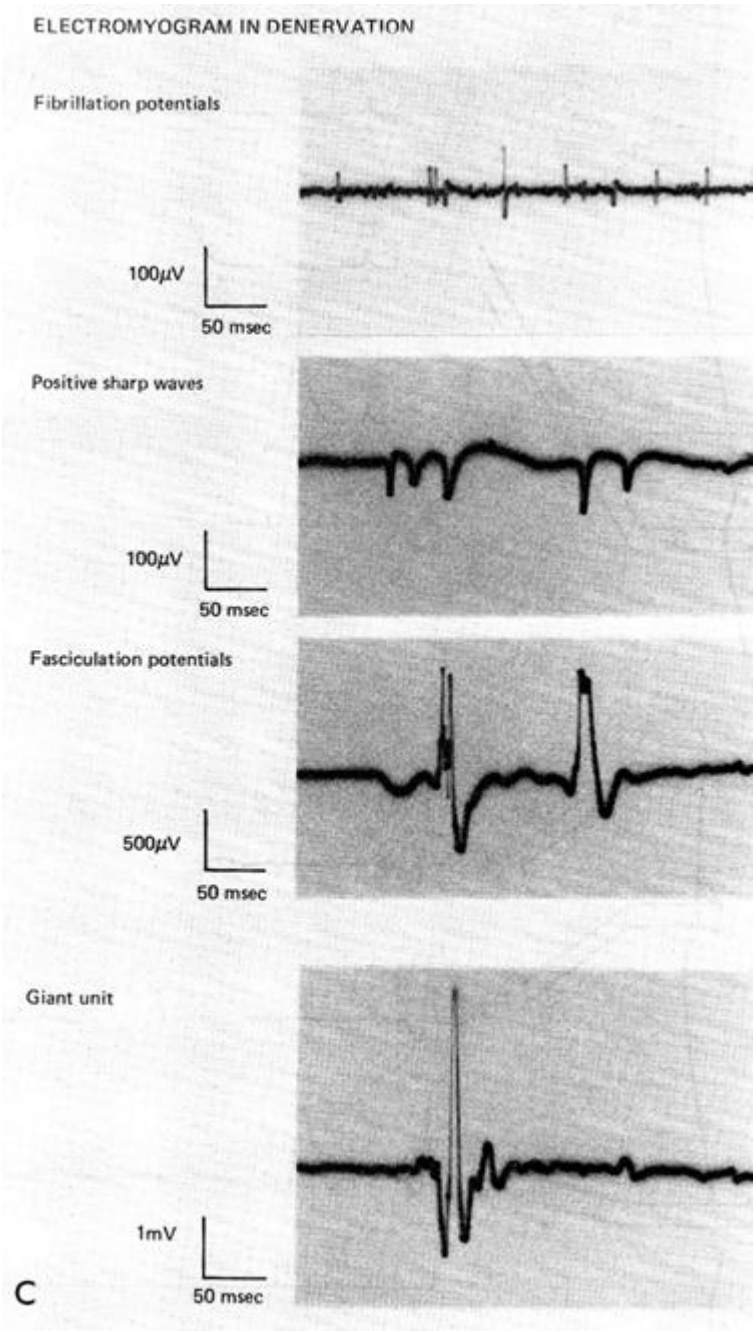


Denervation s. Neuropathies (↓number of motor units):

1) INSERTIONAL ACTIVITY - **increased**.
2) SPONTANEOUS ACTIVITY - **fibrillations**, **positive sharp waves**, fasciculations, complex repetitive discharges**;** increased **polyphasic** action potentials.
3) VOLUNTARY ACTIVITY - neuropathic potentials (↑**duration & amplitude**); ↓**recruitment density** (reduced interference pattern).
4) NCS - prolonged "*F response*", lost *H reflex*.

– after reinnervation, surviving axons branch to innervate adjacent muscle fibers, thus enlarging number of muscle fibers per unit → ↑**duration & amplitude** (> 10 msec, > 5 mV) of MAPs (neuropathic potentials) which frequently are **polyphasic**.

Radiculopathy – preserved SNAP (lesion before DRG) but abnormal EMG of paraspinal muscles.
Neuropathy – affected SNAP but normal paraspinals.



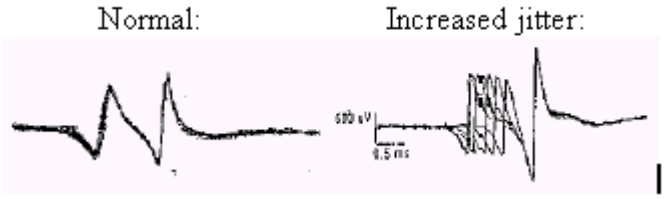
Diseases of neuromuscular transmission (reduced safety factor for neuromuscular transmission → variation in number of muscle fibers firing with each discharge of unit):

- MAPs **vary in amplitude & area**; excess of **small, short-duration potentials**.
- **abnormal** *repetitive motor nerve stimulation* results.
- *increased jitter, blockings* at single-fiber EMG.

SINGLE-FIBER EMG

- action potentials recorded from two or more muscle fibers belonging to same motor unit.

- special electrode.
- **temporal variability (jitter)** between two action potentials at consecutive discharges - reflects variation in neuromuscular transmission.
- main clinical application - **diseases of neuromuscular transmission** - ***increased jitter***, ***impulse blocking*** (failed muscle fiber activation = clinical muscle weakness):



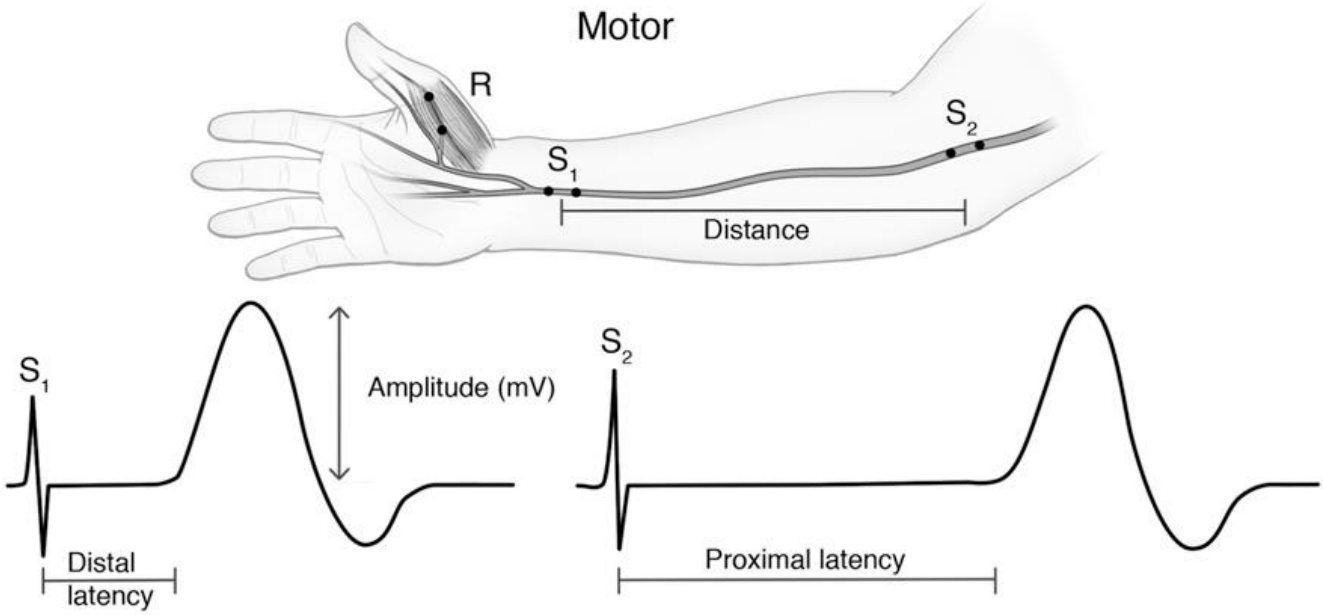
NERVE CONDUCTION STUDIES

- determine functional integrity of peripheral nerves.

Normal conduction velocity ≈ speed limit on highway (50-60)

MOTOR CONDUCTION STUDY

- performed *in conjunction with EMG*.
- ***nerve is stimulated*** at point along its course - electrical stimulus is applied to skin directly over nerve
- electrical ***response is recorded in one of muscles*** supplied by nerve.
  - surface or subcutaneous needle electrodes:
    - ACTIVE ELECTRODE is placed over **endplate** region (muscle belly);
    - REFERENCE ELECTRODE is placed over muscle **tendon**.
  - recorded response is sum of electrical activity of all activated muscle fibers - called **COMPOUND MUSCLE ACTION POTENTIAL (CMAP)**, or **M wave**.
  - stimulus intensity is increased until response no longer grows in amplitude (***supramaximal stimulus***), i.e. activated all nerve fibers.

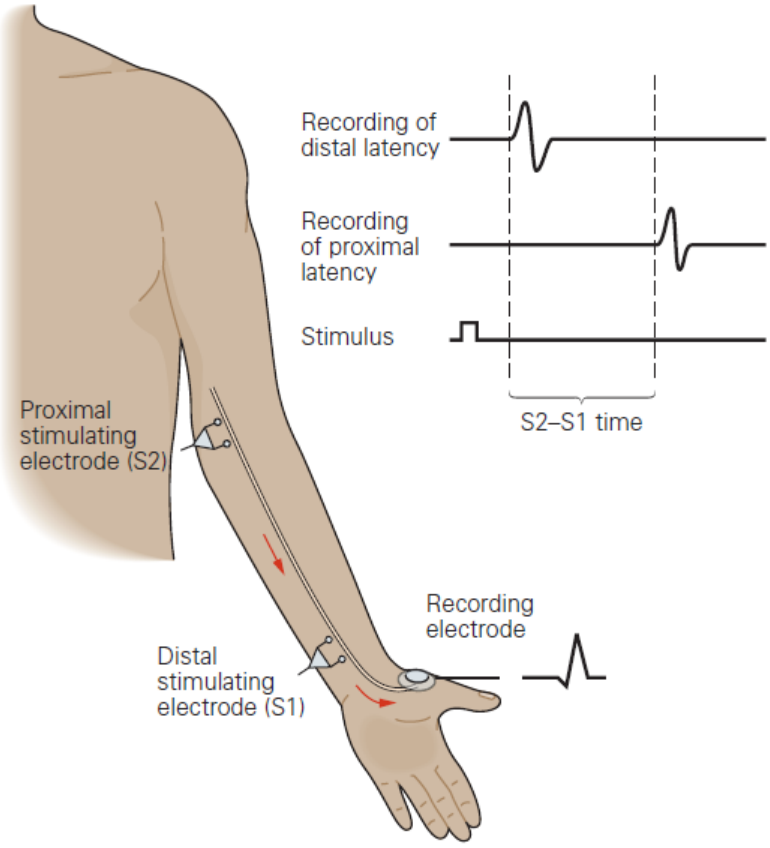


Nerve is stimulated at different sites – obtained responses are compared.

- formula to calculate conduction velocity in motor fibers:

motor conduction velocity* = *distance* between two stimulation sites / *time* difference in latencies.

*velocity is so measured only for fastest conducting fibers.



SENSORY CONDUCTION STUDY

- ***stimulating*** sensory nerve at one point → ***recording*** **SENSORY NERVE ACTION POTENTIAL (SNAP)** at another point along course of that nerve.

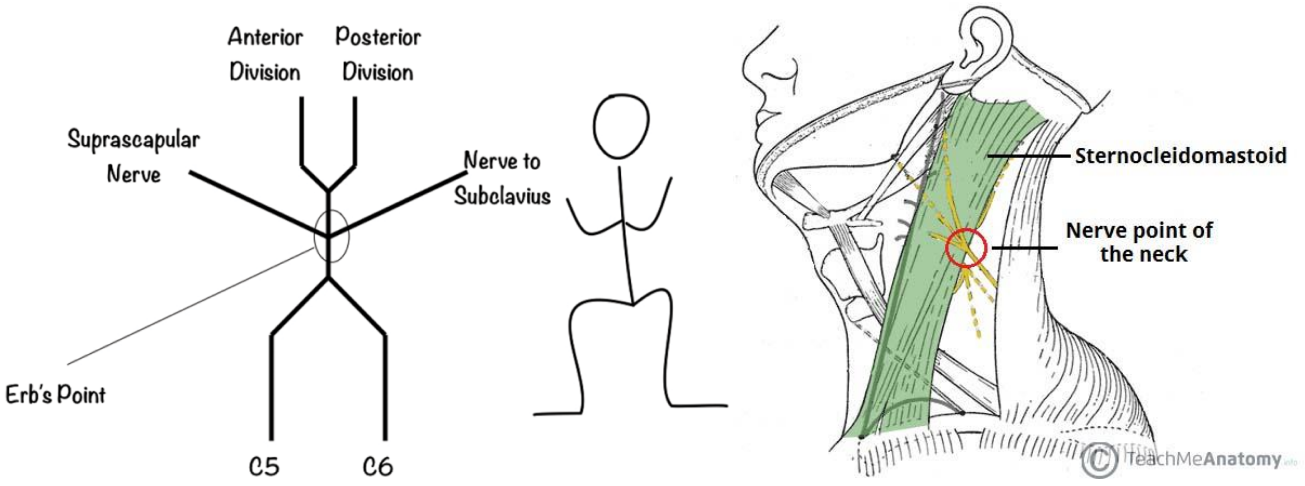
sensory conduction velocity = *distance* between stimulation and recording sites / *latency*

Lesions *distal* to dorsal root ganglion → **sensory nerve conduction studies**.

Lesions *proximal* to dorsal root ganglion → **SSEP** (most frequent site of stimulation: in upper extremity - **median nerve**; in lower extremity - **tibial nerve**).

Erb's point (s. **nerve point of the neck, punctum nervosum**) – point at upper trunk of brachial plexus; six nerves meet at this point:

1. C5 root
2. C6 root
3. Suprascapular nerve
4. Nerve to subclavius
5. Anterior division
6. Posterior division



RADICULOPATHY

- most valuable in advanced motor deficits (> 50% of motor axons affected in a nerve trunk), muscle stretch reflex asymmetry - NCS can provide information regarding physiology (axon loss or conduction block), age, activity, and severity of the process.
- in true radiculopathy, most have only radicular pain and sensory symptoms, which do not have electrophysiologic correlates measurable with standard nerve conduction studies (NCS)
  - **sensory** nerve (SNAP) amplitude, distal latency, and nerve conduction velocity should not be affected in radiculopathy!!!! SNAP is affected only if ***DRG or fibers distal to it*** are affected:
    a) pathologic processes that infiltrate or extend into neural foramen, such as malignancy, infection.
    b) if DRG reside in an intraspinal location they become vulnerable to compression by disk protrusion and spondylosis; e.g. L5 radiculopathy can uncommonly be associated with loss of the superficial peroneal SNAP; however, S1 radiculopathy is almost never associated with sural SNAP amplitude loss (S1 DRG is commonly intraspinal but intraspinal location is caudal to L5-S1 disk space).
- test paraspinal muscles: signs of denervation solidifies lesion location as nerve root!

AXON-LOSS NEUROPATHIES

- **focal conduction block** – CMAP and SNAP ↓**amplitude / completely lost**
- motor / sensory ***conduction velocities are normal***.
- EMG shows denervation!!!

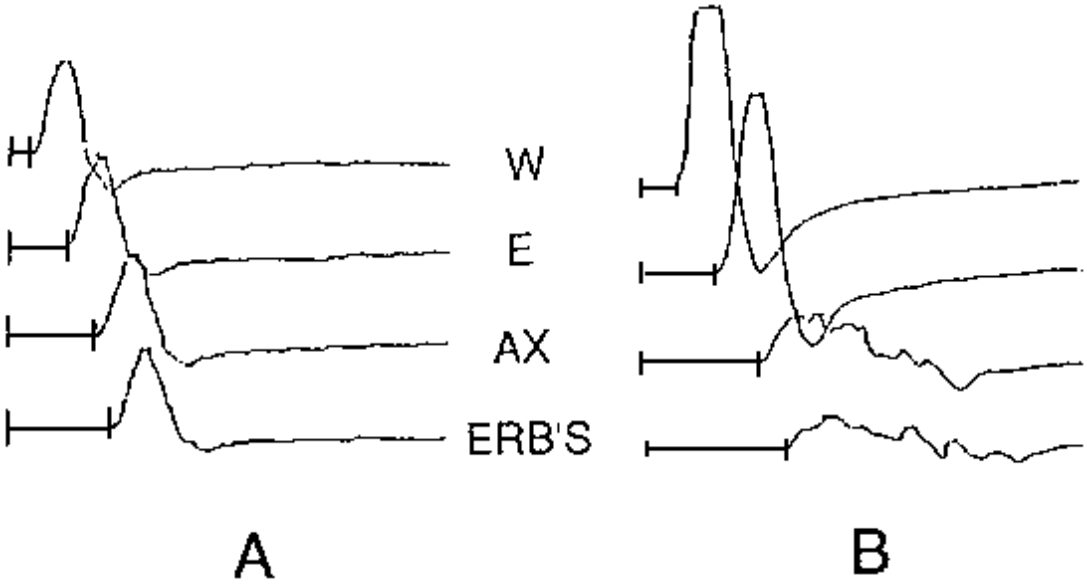
denervation >> conduction loss

Motor conduction block:

A. Normal. CMAP shows little change at all points of stimulation.

B. Conduction block with **sudden** amplitude reduction and temporal dispersion in nerve segment between axilla and elbow.

(W = wrist; E = elbow; Ax = axilla; Erb's = Erb's point)



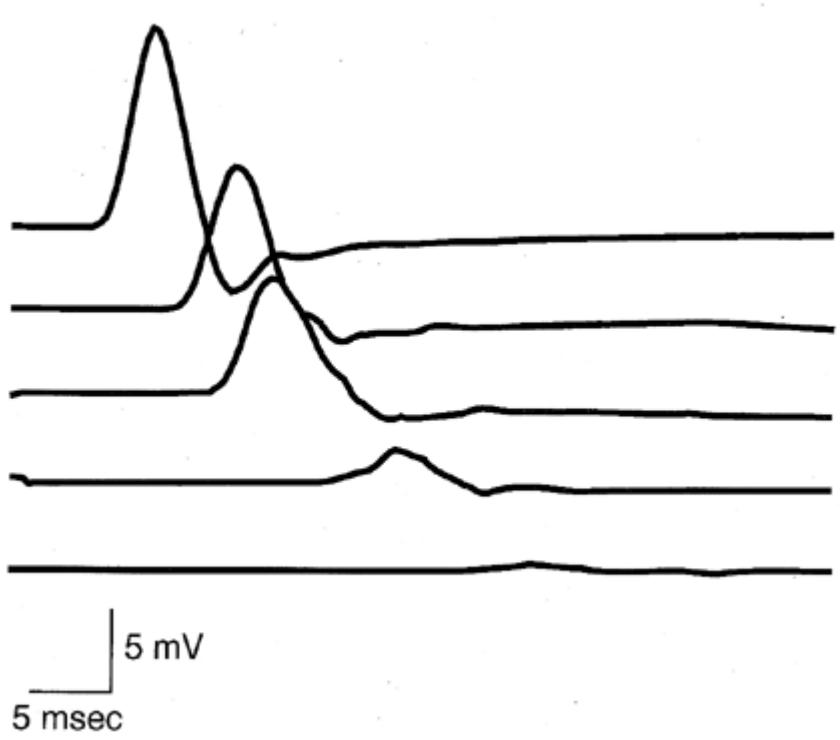
DEMYELINATING NEUROPATHIES

- **conduction slowing**! **prolongation of distal latencies**
- amplitudes and durations of CMAP:
    a) *all large myelinated fibers affected to same degree* - amplitudes and durations of CMAP are **unaltered**.
    b) *different fibers affected to different degrees* - **dispersion** of CMAPs + **↓amplitude** of CMAP (kuo didesnis atstumas between stimulating and recording electrodes, tuo vėliau "atvyksta" impulsai lėtesnėmis skaidulomis lyginant su greičiausiomis skaidulomis → motorinės skaidulos aktyvuojamos ne vienu metu – temporal dispersion)
- **SNAP** - markedly **attenuated / unrecordable** (because of dispersion).
- almost normal EMG!

denervation << conduction loss

N.B. conduction slowing alone is insufficient to produce weakness or significant sensory loss (although sensory modalities requiring timed volleys of impulse transmission along their pathways, such as vibration and proprioception, can be altered)

Progressively more proximal stimulation resulting in **progressive** dispersion of responses with conduction block:



N.B. AMPLITUDE REDUCTION may be due to:

a) **CONDUCTION SLOWING** (temporal dispersion)
b) **CONDUCTION BLOCK** (↓number of active fibers).

- to differentiate two, **area under negative phase** is measured (loss of > 50% area indicates both temporal dispersion and conduction block are present).

REPETITIVE NERVE STIMULATION

- amount of ACh released per impulse *normally* declines on repeated activity (**presynaptic rundown**).

Postsynaptic disorders (e.g. myasthenia gravis) - progressive *decrement* in response size.
Presynaptic disorders (e.g. Lambert-Eaton syndrome, botulism):

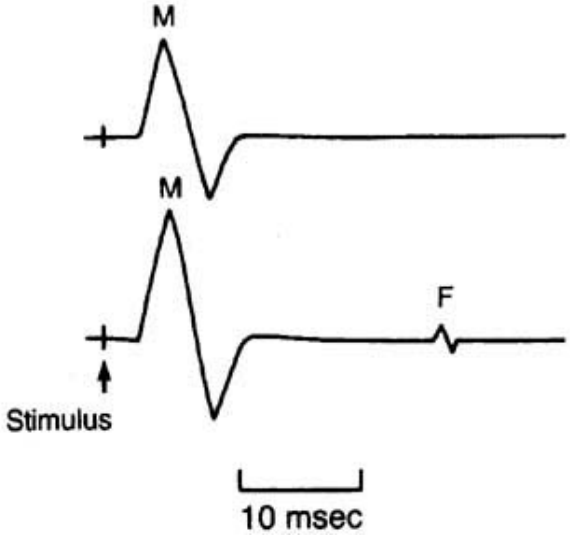
a) stimulation at ***slow rate*** → further *reduction* of already abnormally small response size;
b) stimulation at ***rapid rate*** → progressive *increase* in response size;

F-RESPONSE, H-REFLEX

- useful in **PROXIMAL peripheral neuropathies / radiculopathies** (when conventional nerve conduction studies fail to reveal abnormalities):

F response: electrical nerve stimulation (***motor fibers*** must be excited) → **antidromic (retrograde) activation of motoneuron soma** → orthograde conduction back to periphery → potential evoked from muscle (F response) – like a small echo from motor neuron that follows normal CMAP.

Motor neuron



H reflex - monosynaptic reflex obtained by nerve stimulation (sensory ***proprioceptive fibers*** must be excited); limited clinical utility!

afferent pathway - **spindle afferent (Ia) fibers**;
efferent pathway - ***alpha motor axons***

H reflex is similar to tendon stretch reflex*, except neuromuscular spindles are bypassed

*i.e. evaluates both sensory and motor components (vs. F response – only motor)

Monosynaptic reflex

N.B. latencies of H & F depend on subject's height, limb length!

- it is helpful to *compare symmetry* (normal differences in latency < 2 msec).
- prolonged H / F latencies with normal conventional nerve conduction studies suggest ***proximal neuropathies / radiculopathies***.
- prolonged H reflex with normal F latency - ***dorsal root pathology***.

Main differential – peripheral nerve vs spine – use **provocative tests**:
Tinel, Phalen, thoracic outlet vs Spurling, Lasegue

1. **EDX**
2. **US / MRI** – image:
   1) surgical failures
   2) unusual sites of compression (e.g. localized with Tinel) to rule out a mass
   3) usual sites if strong clinical suspicion but EDX is (false)-negative
3. **Nerve blocks** – diagnostic, therapeutic
4. **Labs** – diabetes, etc

NEUROMUSCULAR JUNCTION

Neuromuscular junction - **fatigable weakness** (initially in **EXTRAOCULAR & BULBAR muscles**) - patients never complain of fatigue - myasthenic symptoms are *always due to WEAKNESS* not to rapid tiring!

Aminoglycosides are contraindicated in both presynaptic and postsynaptic disorders of neuromuscular transmission!

MYASTHENIA GRAVIS

Autoimmune acetylcholine receptor damage → postsynaptic destruction of neuromuscular junction (decreased numbers of muscle ACh receptors)

Curare also blocks muscle ACh receptors!

- diplopia + ptosis + dysphagia and dysarthria; limb & postural muscles are generally less affected; weakness becomes ***generalized***; examination of **neck flexors** is most sensitive (holding head up from surface of examining table while lying supine - gravity cannot be overcome for more than few seconds); weakness varies in *course of single day*
  Failure of respiratory muscles can be life threatening! (myasthenic crisis)

thymus is unequivocally involved in pathogenesis

Tests: **Tensilon (edrophonium) test, ACh receptor antibodies** (serum), **repetitive stimulation** (CMAP decrement), **single-fiber EMG** (increased "jitter", blockings!!!), **CXR, TSH**

Rx: Lifelong **Acetylcholinesterase inhibitors** (**NEOSTIGMINE**, **PYRIDOSTIGMINE**, **AMBENONIUM**); thymectomy → prednisone → cyclosporine / azathioprine
for **myasthenic crisis** – plasma exchange, IVIG

- currently - ***mortality is zero*** - most patients lead normal lives.

EATON-LAMBERT SYNDROME

- autoantibodies against **voltage-gated Ca^{2+}-channels** in presynaptic cholinergic cell → reduced acetylcholine release

N.B. botulism also affects Acch release!

- secondary to paraneoplastic or other autoimmune disorders; proximal muscles of lower limbs; ***repetitive / sustained*** *contraction can improve muscle strength*

respiratory, bulbar, ocular muscles spared

- **repetitive nerve stimulation** at > 10 Hz → CMAP increment

MUSCLES

MYOTONIA - ***painless*** muscle stiffness; specific EMG; with repeated exercise, myotonia improves ("warm-up").

PSEUDOMYOTONIA (S. PARAMYOTONIA) – *no* electrical evidence; exercise *makes* pseudomyotonia *worse*.

CONTRACTURE, CRAMPS – ***intensely painful*** muscle stiffness (contractures are *electrically silent* vs. cramps)

BIOPSY

- avoid clinically ***unaffected*** muscle (may not be involved pathologically).
- avoid ***severely affected*** muscle (may only show *endstage* features - atrophy, fat, fibrosis).

Muscles that are ***moderately weak*** should undergo biopsy - best is muscle with MRC grade 4/5 strength

N.B. pathologist will need to be informed about biopsy site - muscles vary in their normal ratio of type I to type II fibers making this information necessary

MUSCULAR DYSTROPHIES

– inherited progressive primary myopathies:

	Duchenne	***Facioscapulohumeral*** (s. Landouzy-Dejerine disease)	***Myotonic*** (trinucleotide repeat)
Age at onset	Childhood (< 5 yrs.); newborns normal	Adolescence (rarely childhood)	Adolescence or later (rarely congenital)
Inheritance	X-linked recessive	Dominant	
Sex	Male	Either	
Pseudohypertrophy	Common (calves)	Never	
Onset	**Pelvic girdle**	**Shoulder girdle**	**Distal limbs** difficulty in releasing hand grip; muscle atrophy & weakness dominate over myotony!!!
Weakness of face	Rare and mild (**bulbar & ocular muscles** are spared)	Always (**bulbar & ocular muscles** are spared)	Common!!!
Rate of progression	Relatively rapid (death at age 20)	Slow (compatible with long life)	
Contractures and deformity	Common	Rare	
Cardiac disorder	Late cardiomyopathy	None	Arrhythmias
EMG	myopathy		"Dive bomber"
Diagnosis	CK↑↑↑↑, **DNA** or **muscle biopsy** to demonstrate *dystrophin deficiency*		
Rx	Only medication to improve functioning - **alternate-day** PREDNISONE		PHENYTOIN, CARBAMAZEPINE reduce myotonia

CHANNELOPATHIES

- disorders of ion channels that result in **altered excitability** of cellular membranes:

A. Hyperexcitability → **MYOTONIAS** (i.e. nondystrophic myotonias - myotonia (not weakness) is most prominent symptom); normal [CK]; muscle biopsy - few abnormalities;
Rx - **membrane-stabilizing drugs**: PHENYTOIN, PROCAINAMIDE, QUININE, ACETAZOLAMIDE, MEXILETINE

B. Hypoexcitability → **PERIODIC PARALYSIS** (**hypokalemic**, **hyperkalemic**) - K^+ levels are checked q 15-30 min to determine direction of change when muscle strength is decreasing or improving; muscle biopsy – vacuoles;
Dx: ***hypokalemic & hyperkalemic challenges***
Rx:

Hyperkalemic periodic paralysis:

- attack termination – **glucose** + **insulin** ± i/v **Ca gluconate**
- attack prophylaxis: **urinary K^+ excretion promoters** – ACETAZOLAMIDE, **thiazides**, FLUDROCORTISONE; **inhaled β-adrenergics** (e.g. SALBUTAMOL).

Hypokalemic periodic paralysis

- attack termination – **KCl** ± **β-blockers**.
- attack prophylaxis – ACETAZOLAMIDE*, TRIAMTERENE, SPIRONOLACTONE.

*mechanism of action of acetazolamide is uncertain.

NEUROMYOTONIA (s. ISAACS' syndrome)

- ACQUIRED autoantibodies against **voltage-gated K^+ channels** on peripheral nerves→ hyperexcitable motor nerve → ***continuous muscle fiber activity***
Rx: PHENYTOIN, CARBAMAZEPINE

METABOLIC MYOPATHIES

DYNAMIC (exercise-induced) myopathies - symptoms appear during / after exercise (acute myalgias, stiffness → contractures, intermittent weakness → myoglobinuria).
STATIC (stable or slowly progressive) myopathies - chronic fixed progressive weakness (simulates muscular dystrophy; no exercise intolerance, no myoglobinuria).

Forearm (grip) exercise:

glycogenosis – [lactate] elevation does not occur; muscle develops painful contracture;
lipid metabolism disorders – normal profile;
myoadenylate deaminase deficiency – [ammonia] elevation does not occur;
mitochondrial disorders – excessive [lactate] elevation;
poor effort – neither [lactate] nor [ammonia] increase.

CONGENITAL MYOPATHIES

– rare **congenital nonprogressive** primary myopathies, not explained as *dystrophic* or *metabolic* abnormalities.
The commonest presentation - floppy baby with delayed motor milestones; muscle weakness → ***skeletal defects***
Dx: CK ≈ normal. EMG - myopathy
main diagnostic method - **muscle biopsy**:

Nemaline Myopathy: **rods** in muscle biopsy
Myotubular-Centronuclear Myopathy: Large **central nuclei**
Central Core Disease: **Central / eccentric cores**
Congenital Fiber-Type Disproportion: increased number of **small type I muscle fibers**

MITOCHONDRIAL DISORDERS

Clinical phenotype ranges from mild, slowly progressive **weakness of extraocular muscles** to severe, fatal infantile myopathies and **multisystem encephalomyopathies**.

CHRONIC PROGRESSIVE EXTERNAL OPHTHALMOPLEGIA is clinical hallmark of mitochondrial disease!

Calcification of basal ganglia is seen in all syndromes but in minority of patients with any syndrome!

Diagnosis:

1. Congenital **LACTIC ACIDOSIS**
2. **Muscle biopsy** - **RAGGED RED FIBERS**.
3. Myopathic **EMG**.
4. **PET** - reduced cerebral metabolic rates for *oxygen* but normal *glucose* utilization.
5. **CT / MRI** – **INFARCT-LIKE LESIONS**.
6. **IMPAIRED RESPIRATION** in **biochemical tests**.
7. **HETEROPLASMY** on **Southern blot analysis**.
8. Specific cause:
   1) ENZYME ANALYSIS in skin fibroblasts / muscle biopsy.
   2) DEMONSTRATION OF MUTATION in muscle biopsy / WBCs.

Rx: **cofactor supplementation** (UBIQUINONE, CARNITINE, VITAMINS) + DICHLOROACETATE (for lowering of lactic acidosis)

RHABDOMYOLYSIS, MYOGLOBINURIA

N.B. renal failure is likely if *hypotension (hypovolemia)* and *acidosis* coexist.

Rx: **bed rest**, **diuresis** (MANNITOL + BICARBONATE), control **hyperkalemia**

NEUROPATHIES (PNS)

radiculopathy – segmental distribution (dermatome, myotome, sclerotome).
plexopathy – distribution of > 1 peripheral/spinal nerve.
mononeuropathy – distribution of 1 peripheral nerve.
mononeuropathy multiplex – patchy distribution of ≥ 2 major named nerves in ≥ 2 limbs (either diabetes or vasculitis of vasa nervorum)
polyneuropathy – distribution of > 1 peripheral/spinal nerve.

N.B. **diabetes** can cause any type / category of neuropathy!!!

1. Diseases that affect primarily **Schwann cell** → ***segmental demyelination***.
   - axon and myocytes remain intact!
   - recurrent bouts → *PALPABLE "ONION BULBS"* nerve hypertrophy

2. Diseases that affect primarily **axon** → ***wallerian degeneration***.
   - with *secondary myelin disintegration*
   - myocytes undergo *DENERVATION ATROPHY*

     N.B. muscle atrophy is sign of axon-loss neuropathies!
   - atrophic muscle fibers may be reinnervated by normal neighboring axons ("**fiber type grouping**").
   - **axon regeneration** is slow process (vs. **remyelination** – quite rapid!).

3. Diseases that affect primarily **neuron body** → ***distal degeneration*** (s. distal axonopathy, "dying back").

First test – **EMG & nerve conduction studies**

small (unmyelinated and myelinated) ***fibers*** – *temperature* & *pain* sensation↓ + autonomic dysfunction + burning pain; ***large fibers*** – *position & vibratory* sensation↓ + somatomotor dysfunction.

axon-loss lesions - conduction **block** & amplitude **reduction**; EMG shows denervation *myelin-loss lesions* - conduction **slowing***; normal EMG

*severe demyelination may cause conduction **block**!

Pain can be most distressing part of disease!
Neuropathic pains often do not respond well to conventional analgesics!

PROGNOSIS

- **axon injury**: nearer injury to CNS, lower probability of regeneration (esp. cranial nerves); recovery is slow!
  Neuromas may form!
  Recovery may fail to occur at all!
- **myelin injury**: recovery is complete within few days or weeks.

POLYNEUROPATHY

- diffuse lesions of peripheral nerves:

Mostly **metabolic / toxic** causes (esp. diabetes mellitus, alcoholism, uremia).

Acute Axonal Polyneuropathy:
1) porphyric neuropathy
2) massive intoxications (e.g. arsenic)

Acute Demyelinating Polyneuropathy:
1) Guillain-Barré syndrome
2) buckthorn berry intoxication
3) diphtheritic polyneuritis.

symmetric legs > arms extensors > flexors distal → proceeds centripetally in graded "STOCKING" manner*

*nerve fibers are affected according to length (without regard to root or nerve trunk distribution).

Exceptions to distal distribution (i.e. predominantly proximal distribution):
1) lead neuropathy (tends to affect upper extremities first, esp. radial nerve)
2) Guillain-Barré syndrome

First symptoms tend to be **paresthesias** (tingling, burning, etc) → pansensory loss over both **feet**

- by time sensory disturbance has reached **upper shin**, dysesthesias are usually noticed in **tips of fingers**.
- in profound sensory loss → repeated traumatic injury* → *painless ulcers* on digits, *Charcot's joints*. *avoidable by proper care!

In some instances, process begins with feet weakness (without sensory symptoms).

- motor component begins as ***weakness*** and ***atrophy*** in **intrinsic feet muscles** →→→ quadriplegia, impaired ventilation, sphincteric dysfunction.

N.B. variations are common → diversity of clinical syndromes.

specific therapy for metal poisoning - D-PENICILLAMINE

MONONEUROPATHY

- most commonly due to local cause:
  a) **trauma** - most common cause!
  b) **entrapment**
  c) nerve **infarction** (diabetes, vasculitis).
- factors favoring surgical treatment (in absence of history of trauma):
  1) chronicity
  2) worsening neurologic deficit (particularly if motor)
  3) electrodiagnostic evidence of wallerian degeneration (i.e. axonal neuropathy)

RADICULOPATHY

– segmental distribution (dermatome, myotome, sclerotome).

Pain is ***precipitated*** by: moving spine, Valsalva maneuver, root stretching maneuvers (e.g. straight leg rising), root compression maneuvers (e.g. Spurling's test).

Pain location is most variable of clinical features!

- muscles are innervated by > 1 spinal root! (*actual motor innervation is multisegmental*)

Passive ***straight-leg raising*** (Lasègue sign) → ischiadic nerve traction → L_5-S_1 root traction → SCIATICA;

- *smaller angle of elevation required* to elicit pain, greater chance of root compression.
- characteristic pain on *opposite leg elevation* (crossed Lasègue sign) may be even stronger evidence of root compression.
- pain may also be elicited by having patient walk on heels; some patients avoid full weight bearing on heel of involved side (stand with knee flexed and heel off floor).

EMG* – denervation – *spontaneous activity* (positive waves, fibrillations), neuropathic potentials, and *decreased motor recruitment*

Nerve conduction velocities – normal (vs. in peripheral neuropathies) unless > 50% motor axons lost.

evoked potentials (H, F)* – abnormal

*localize level + determine severity of root lesions.

DORSAL ROOT GANGLION SYNDROMES (SENSORY GANGLIONITIS)

- inflammatory changes and loss of neurons in dorsal root ganglion (i.e. NEURONOPATHY).

1) **idiopathic**.
2) **neoplasia**!!! (most frequent associations - small cell carcinoma of lung)

Rapidly progressive sensory symptoms: ***numbness***, ***paresthesias***, lancinating ***pains***, loss of ***proprioception & vibration*** > loss of *pain & temperature*

Diagnosis
- sensory **nerve action potentials (SNAP)**↓↓↓
- normal **EMG**.
- sural **nerve biopsy** - loss of large myelinated fibers.

Consider surgery – **DRG stimulator**.

PLEXOPATHIES

1) ***trauma***
2) ***neoplastic*** compression / infiltration (early prominent pain is characteristic!).
3) ***radiation*** (painless progressive weakness).
4) ***immunologic attack*** (e.g. *brachial neuritis*, s. PARSONAGE-TURNER syndrome).
5) ***diabetes mellitus***.
6) ***neurofibromatosis***.

- *anatomy is complex* (difficult to recognize and localize) - different patterns of **motor** and **sensory** loss, **pain**.

DIAGNOSIS

- **imaging**: N.B. high-resolution **MRI + MRI neurography** is modality of choice!; **CXR** to rule out Pancoast tumor.
- **EMG** is fundamental in localizing lesion.
- **NCS**: in trauma

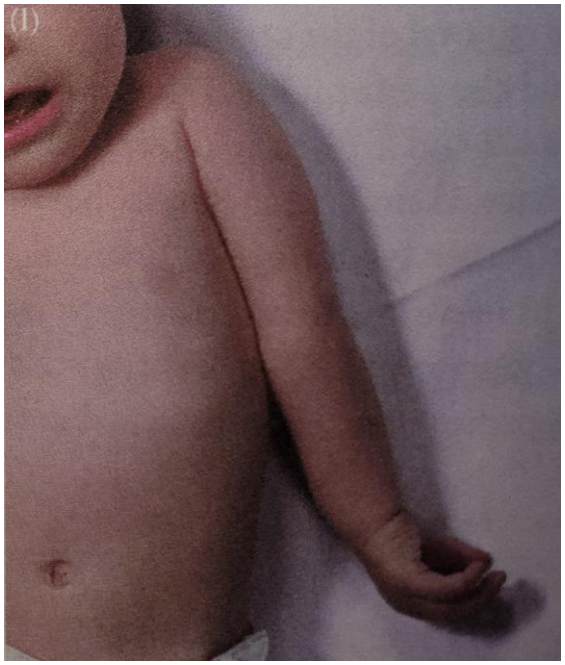
BRACHIAL PLEXUS TRAUMA

- acutely – ask for vascular evaluation (if penetrating trauma → tetanus shot)
- **MRI + MRI neurography** 4 weeks after injury - ***pseudomeningoceles***
- **EMG** is done 4-5 months after acute injury
- **NCS**: *plexus stretch* injuries vs. *avulsion* injuries
  - preganglionic (pure root) injuries leave dorsal root ganglion intact - distal **sensory nerve conduction velocities** are intact – bad sign – not recoverable (even cannot do Oberlin nerve transfer)
  - **evoked potential** (after median nerve stimulation) shows delay at Erb's point.
  - N9 dorsal root ganglion **evoked potential** is preserved in pure root avulsion.
- **CSF** may contain blood in trauma.

COMPLETE BRACHIAL PLEXUS lesion – **flail, anesthetic upper extremity** (except for medial strip along arm supplied by intercostobrachial branch of 2nd intercostal nerve).

DUCHENNE-ERB palsy (C_{5-6} roots or upper trunk lesion) - from ***downward arm displacement***: fall from horse or motorcycle, obstetrics (*shoulder dystocia*)

Clinical features – mainly **shoulder & upper arm** muscles - "waiter's tip" position:



- upper arm hangs adducted (*m. deltoideus* – n. axillaris, *m. supraspinatus* – n. suprascapularis) and internally rotated (*m. infraspinatus* – n. suprascapularis, overpull from intact m. pectoralis major)
- can't flex elbow; forearm is pronated (*m. biceps brachii* – n. musculocutaneus).
- can't reach with hand contralateral shoulder (*clavicular head of m. pectoralis major* – n. pectoralis lat.).
- triceps, wrist, hand – intact!
- *sensory loss* is incomplete (hypesthesia on outer surface of shoulder, arm and forearm).

"BURNERS" / "STINGERS" - symptoms following sudden shoulder depression in contact sports, usually football.

- burning dysesthesias going down ipsilateral upper extremity (often into thumb) ± weakness of biceps and shoulder girdle muscles.
- symptoms resolve within few minutes (occasional weeks).

DEJERINE-KLUMPKE palsy (C_8-Th_1 roots or lower trunk lesion) - from ***upward arm displacement***: obstetrics (*breech delivery*), shoulder dislocation, Pancoast tumor, ***thoracic outlet syndrome,*** CABG surgery (sternal retraction)

Clinical features – mainly **forearm & hand** muscles:

- *n. ulnaris* + *n. medianus* (flexor carpi ulnaris, flexor digitorum, interossei, thenar and hypothenar) – can't flex wrist, "claw hand", "simian (flattened) hand".
- *n. pectoralis med.* – can't adduct upper arm.
- lesion to communicating branch to inferior cervical ganglion → *Horner's syndrome*.

TREATMENT

Components:

1. Pain
2. Orthopedic
3. Reinnervation

Orthopedic

- flail or weak arm should be **supported** (immobilized across upper abdomen) against gravity to prevent additional damage!
- if no function return → shoulder fusion

Pain:

plexus avulsion pain (intractable debilitating sensory deprivation pain) → DREZ lesioning.
stretch injuries → SCS or DBS.
causalgia may respond to sympathectomy

Reinnervation

- **acute transections** by ***sharp object*** (lacerations with knife or glass):
  a) iatrogenic (scalpel) – immediate ***primary repair***.
  b) penetrating non-missile injuries → allow wounds to heal → ***primary repair*** without delay.
     – **lost neural tissue** (found during initial exploration for repair of other injuries) → early cable grafting.
- **GSW** (usually leave nerve in continuity; may use US to check for plexus continuity) → observation for up to 3 months (to help establish degree of neural injury); if serial examinations demonstrate 4-5° lesions → surgical intervention.
- **closed stretch injuries** → observation; severe axonal degeneration 3-5 months after injury → ***surgical exploration and repair***.
  – not improving **obstetric palsy** → ***surgery*** at 3-9 months of age.
  – **avulsion of roots (s. preganglionic injury)*** – ***untreatable*** vs. nerve transfer** to distal end of musculocutaneous nerve (gives useful elbow flexion).

Oberlin will not work!!! No options for cable grafting!!!

For ***brachial plexus stretch*** injuries, first NCS/EMG at 3-5 months, operate < 6 months from injury!

N.B. if deficit is progressive - suspect vascular cause (pseudoaneurysm, A-V fistula, or expansile clot) - explore immediately!

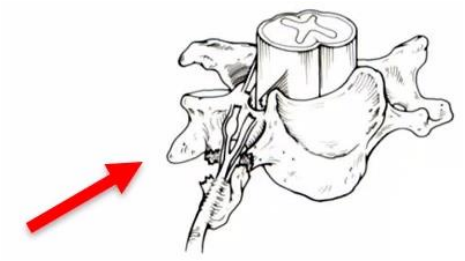
vs.

N.B. injuries to lower elements (Dejerine-Klumpke) are NOT operated at all! (i.e. no realistic hopes to regain hand function)

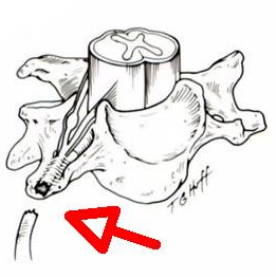
Summary of features when to suspect avulsion of roots (s. preganglionic injury)

1. Horner syndrome: pre-ganglionic injury interrupts white rami communicantes
2. Paralysis of proximal muscles (nerves): serratus anterior (long thoracic nerve) - winging of scapula; rhomboids (dorsal scapular nerve)
3. Early neuropathic pain.
4. Pseudomeningoceles on MRI (or CT myelogram).
5. No Tinel's sign at Erb's point (if present – it is a site of injury, i.e. postganglionic)
6. EMG (≥ 3 weeks from injury):
   a) denervation in paraspinal muscles (posterior ramus of spinal nerve originates just distal to dorsal root ganglion).
   b) normal sensory nerve action potential (SNAP): preganglionic injuries leave dorsal ganglion sensory cell body + distal axon intact = normal SNAP can be recorded proximally even in an anesthetic region.

Root avulsion (preganglionic injury) – Horner is present, sensory NCS (SNAP) is normal!



vs. postganglionic:



**<u>Donor nerves for elbow flexion neurotization:</u>

Total brachial plexus injury:

a) **intercostal nerves**
b) phrenic nerve
c) contralateral C7

Partial (upper trunk) brachial plexus injury:

a) ulnar nerve
b) median nerve
c) double fascicle (ulnar+median)
d) medial pectoral nerve

<u>For shoulder abduction neurotization</u>: CN11 → suprascapular

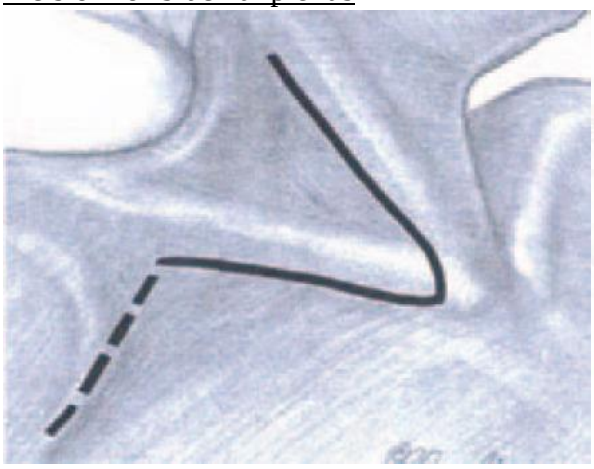
Priorities: elbow flexion > shoulder abduction

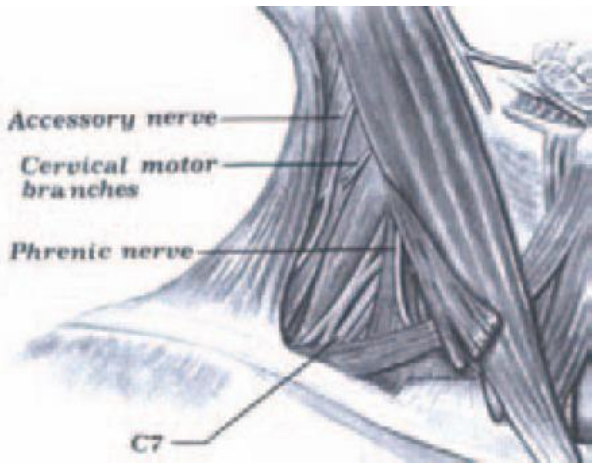
<u>Preparation for OR</u>

1. Stimulator (nerve action potentials*, EMG)
2. Cable graft donor site / Neurotization donor site

*not nerve conduction

<u>Incision for brachial plexus</u>





- incision starts at posterior border of **sternocleidomastoid muscle** → continues laterally above clavicle.
- clavicle osteotomy can be made if necessary.
- external jugular vein is the first important landmark - ***spinal accessory nerve*** lies posterior to this.
- transverse cervical artery is potential source of bleeding and good anastomosis for vascular graft.
- both external jugular vein and transverse cervical artery are divided along with **omohyoid muscle** in supraclavicular fossa - upper and middle trunks lie posterior to this muscle.
- **anterior and middle scalene muscles** come next - between these muscles, trunks of plexus emerge.
- ***phrenic nerve*** located anterior to anterior scalene muscle should be identified and protected.
- after identification of the neural element of C5 root, one can trace lower nerve roots and identify the beginning of the brachial plexus.

- if injuries extend infraclavicularly, at level of coracoid process, incision follows down deltopectoral groove (dotted line).
- **pectoralis minor muscle** detachment provides access to cords of plexus with lateral cord being most prominent.

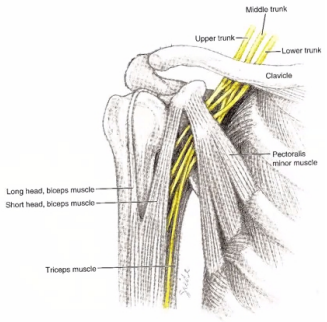
Infraclavicular Exposure

Incise along the delto-pectoral groove from the clavicle to the anterior axilla.

Separate and retract the deltoid a the pectoralis major.

Identify the coracoid process and pectoralis minor tendon.

Divide the tendon near the coracoid and reflect the muscle.



Postop: shoulder immobilization for several weeks → PT

See Case PN3 >>

NEURALGIC AMYOTROPHY (s. brachial plexitis, PARSONAGE-TURNER syndrome)

Similar disorder may affect LUMBOSACRAL plexus!

- unknown cause
- typically young men.
- often preceded by some ***antecedent event*** (e.g. general anesthesia, vaccination!!!).
- may be **bilateral** and asymmetric, may mimic peripheral mononeuropathy
  - upper trunk suffers most (actually, multiple proximal mononeuropathies but may mimic isolated mononeuropathy): sudden onset of **severe pain** in shoulder girdle → soon followed by **weakness & wasting**.
- **nerve conduction studies** - ***axonal*** neuropathies that do not fit peripheral nerve distributions (*demyelination* may play role in rare instances).
- establish diagnosis – **EMG** (MRI may show muscle denervation changes)
- corticosteroids have no proven benefit.
- clinical recovery takes 2 months ÷ 3 years (so don't rush to operate!!!); if no improvement by 18-24 months → **tendon transfer surgery**.

See Case PN4 >>

TRAUMA

Always check **peripheral pulses**! – vascular injury is emergency (PNS injury is not!)

Important quality of PNS (vs. CNS) is remarkable ability to recover after injury through axon regeneration and remyelination!

Mechanical nerve injuries are classified:

Seddon (1943)	Sunderland (1951)	Myelin	Axon	Endo-	Peri-	Epi-	Recovery
Neurapraxia	1°	±					Within days-weeks
Axonotmesis	2°	+	+				1 mm / day (s. 1 in / mo) *
Neurotmesis	3°	+	+	+			No spontaneous recovery**
	4°	+	+	+	+		
	5°	+	+	+	+	+	

*rate decreases with increasing distance from cell body:
above elbow/knee - 3 mm/d
between elbow and wrist or knee and ankle - 1.5 mm/d
below wrist or ankle - 0.5 to 1 mm/d
**after successful surgery, recovery proceeds as in axonotmesis, but *time to pass scar can be prolonged*!

SEDDON (1943) classification

A. ***NEURAPRAXIA*** - **myelin** damage, **axon** intact* - ***conduction block*** at site of lesion**; distal fibers do not degenerate (no denervation!); conduction block is fully & rapidly reversible.
*no histological abnormality or segmental demyelination
**but proximal & distal conduction is normal
N.B. proximal and distal recovery is simultaneous!

B. ***AXONOTMESIS*** - **axons** are interrupted but **Schwann cell tubes**, **endoneurium, connective tissue** are intact.
- NEURON CELL BODY undergoes either ***apoptosis*** or ***chromatolysis*** (preparation for regeneration).
- AXON:
  1. distal segment: ***wallerian degeneration***.
  2. proximal stump: ***retrograde degeneration, s. die-back*** (to at least next node of Ranvier) → ***regenerative response***
     a. axon invariably returns to end organ it originally innervated; recovery will proceed proximal → distal at 1 mm / day.

C. ***NEUROTMESIS*** - **axon**, **myelin**, and **connective tissue components** are damaged:
a) preserved continuity of epineurium (→ intraneural fibrosis, NEUROMA IN CONTINUITY).
b) nerve severed completely (→ STUMP NEUROMA).
- at site of injury – ***traumatic degeneration***; distally ***wallerian degeneration*** occurs; recovery occurs only if nerve ends are brought together!

SUNDERLAND (1951) classification

1° injury = ***NEURAPRAXIA***.
2° injury = ***AXONOTMESIS***.
3-5° injury = ***NEUROTMESIS***:
3° injury - loss of continuity of ENDONEURAL tubes (perineurium intact) - some regenerating axons are no longer confined to tubes they originally followed → new **anomalous**

patterns of innervation, **intrafascicular fibrosis** (→ recovery may be incomplete) – externally nerve may look normal!

4° injury - continuity of nerve trunk (epineurium) persists, but its internal structure (PERINEURIUM) is severely disrupted - *organized regeneration is unlikely* (involved segment is converted into tangled strand of connective tissue, Schwann cells, and regenerating axons → observable **neuroma in continuity**).

5° injury - EPINEURIUM disrupted → perineural scarring, observable **stump neuroma** – *spontaneous regeneration is impossible*.

N.B. macroscopically nerve starts to look abnormal with ≥ 4° injuries

Additions

0° injury = ***PHYSIOLOGICAL (METABOLIC / ISCHEMIC) CONDUCTION BLOCK*** – normal anatomy but local energy supply is interrupted.

6th category (MacKinnon and Dellon, 1988) - combination of above injuries.

All degrees of injury initially clinically appear the same!

- if neurological deficit is ***incomplete*** - injury is most likely *neurapraxic*.
- if neurological deficit is ***complete*** - injury may be *neurapraxic*, *axonotmetic*, or *neurotmetic*.

NEURAPRAXIA - complete recovery in hours ÷ weeks.

Nerves that do not regenerate well:

1) long nerves
2) nerves to fine muscles (high axon-to-myocyte rate)

- GSW to thigh – if sciatic nerve is damaged, typically it is peroneal distribution (tethered at fibular head) with tibial distribution spared.
- pure motor or sensory nerves recover better than mixed nerves.
- recovery is better in radial and musculocutaneous nerves (coarse muscles) than in median or ulnar nerves (fine muscles); tibial division fares better than peroneal division.
- high injury of sciatic nerve – muscles always degenerate before reinnervation – functional results are always bad!
- sign of recovery - distally migrating **TINEL's sign** (lightly tapping along nerve → paresthesias in sensory distribution of nerve)

ELECTROPHYSIOLOGIC TESTING

Both EMG and NCS earliest that can be helpful is **≥ 3 weeks** after injury!
EDX after trauma – at 3 weeks and 3 months

EMG

– the only clinically useful diagnostic test!

NEURAPRAXIA – EMG always normal!

- ***denervation*** changes (fibrillations and positive sharp waves) appear only after 2-5 weeks.

NERVE CONDUCTION STUDIES

Early – not helpful clinically! (initially, all injuries have conduction block and intact distal portion)

Proximal stimulation:
NEURAPRAXIA – slowing or conduction block.
≥ ***AXONOTMESIS*** – conduction block.
Distal stimulation – normal (intact axons distal to any injury site - normal CMAP)

Late – earliest that can be helpful is ≥ 3 weeks after injury
Proximal stimulation:
NEURAPRAXIA – normal (correspond to clinical recovery).
≥ ***AXONOTMESIS*** – conduction block.
Distal stimulation:
NEURAPRAXIA – normal (correspond to clinical recovery).
≥ ***AXONOTMESIS*** – conduction block.
– reduced evoked potential amplitude is observed by 7 days (wallerian degeneration).

MEDICAL TREATMENT

- initial treatment of choice in all cases except when nerve discontinuity is known (→ surgery).

SURGICAL TREATMENT

Allow sufficient time for spontaneous recovery without jeopardizing results of late repair

Earlier repair → better results

Surgery for nerve repair works best if done within 6 months (does not work well if > 9 months)
Surgical delays in excess of 5 months dramatically decrease rate of functional return!!!

Timing of Nerve Exploration (Repair) - **RULE OF 3'S + 1**	
Sharp clean lacerations	- (within) **3** days
Blunt or jagged / dirty lacerations	- **3** weeks
Closed, stretch, gunshot injuries in continuity	- **3** months
Tendon transfers, joint fusions	- **1** year

Surgery is indicated for ***NEUROTMESIS***! (i.e. anything > axonotmesis will need surgical repair)

A. **Sharp lacerations** → ***primary repair*** within first 48-72 h* (esp. **clean** lacerations made by sharp objects + obvious motor and sensory **deficits**); usually **end-to-end**.

- if injury is *several days* old, wait ≈ 2 weeks for edema to subside (US shows transected nerve better than MRI).

*or at any time you see the patient for the first time (sooner the better); exact timing depends on object:

a) iatrogenic (scalpel) – immediate ***primary repair***.
b) penetrating non-missile injuries → allow wounds to heal (tag ends of nerve under slight tension – "surgery #1") → ***primary repair*** without delay ("surgery #2 – approx. 3 weeks later).
N.B. some experts do definitive operation immediately after injury and decide on extent of injury based on intraop findings.

B. **Blunt (closed) injury** (degree of injury unknown) → ***observation*** for at least 6 weeks (up to 6 months) for recovery from possible ***NEURAPRAXIC-AXONOTMETIC*** injury (Sunderland's 1-2°)

- continued observation: clinical + EMG/NCS:
  - clinical recovery evident → continue observation.
  - if no clinical recovery → EMG - if some potential recruitment is seen (i.e. nerve action potential distal to injury regardless of amplitude or latency) → observe another month (clinical recovery may follow).
  - if electrodiagnostic nerve function is progressively deteriorating → ***delayed repair*** may be indicated because status of connective tissue cannot be assessed without direct exploration and stimulation across neuroma.
- interposition cable **graft** is often needed (stump shrinkage)

C. **Gunshot wounds / open fractures / contaminated lacerations** are treated as "closed nerve injury" with ***delayed repair*** (at 3-6 months) if going to OR for exploring / debriding / ortho or vascular repair, then identify nerves → make later delayed re-exploration easier:

a. **tie** divided nerve ends together to loosely approximate with inert sutures (to prevent retraction) and suture to surrounding fascial tissue

b. **tag** divided nerve ends

N.B. in vast majority of gunshot wounds, nerve is not divided (only contused) – very important for management strategy!

Recommended timing to obtain NCS/EMG

A) to identify an area of conduction block (regardless if lesion is from neuropraxia, axonotmesis, or neurotmesis) – acutely
B) partial injuries & entrapments, compressive lesions and tumors – any time
C) relatively focal contusions - 2–4 months
D) stretch injuries (esp. brachial plexus) - 4–5 months

Delayed surgery must be planned according to facts that:

– axon regenerates 1 mm/day or 1 inch / month;
– most delayed surgeries are done at 5-6 months.
– **24 month rule**: after 24 months of denervation, most muscles atrophy - cannot recover useful function even with reinnervation. Exceptions: facial muscles, large bulky muscles.
  e.g. not worthwhile to suture some nerves if injury is rather proximal.
  sensory aspect: restitution may still be possible – crude sense of touch returns ± temperature (other senses do not return)
  e.g. worthwhile to suture some nerves to effect return of at least protective sensation.

Humerus fracture with wrist drop – document prior to ortho surgery (if ortho asks to take a look – fine: if nerve is transected [very rare] may repair; else, if just contusion, expectant management)

Surgery windows according to **nerve**:

- **brachial plexus** stretches / contusions are observed for 4 months; operate < 6 months from injury!
- **ulnar nerve**: adults > 40 years rarely achieve functional result from ulnar nerve repairs proximal to elbow; i.e. not worthwhile to suture ulnar nerve near axilla.
- **median nerve**: worthwhile to suture median nerve near axilla to effect return of at least protective sensation; one can observe distal median nerve injury for 3-4 months and still obtain good surgical result.
- time frame for **radial nerve** exploration ranges 2-5 months.
- **sciatic nerve**: with blunt injury, surgical intervention should occur as soon as possible if no clinical recovery within 3-8 weeks.

- **tibial nerve**: worthwhile to suture tibial nerve above midthigh to effect return of at least protective sensation.
- **peroneal nerve**: not worthwhile to suture peroneal nerve above midthigh.

Release regenerating nerve fibers (**neurolysis**) or **excise** damaged segment ± **cable graft** (between proximal and distal nerve ends) → anastomose cleanly cut ends (**neurorrhaphy**).

Clinically nonfunctioning nerve in continuity (≥ 4° lesion, s. *neuroma in continuity*) → OR (have stimulator ready, prep for graft donor site):

A. **Bad scar** → ***excise*** → cable ***graft***

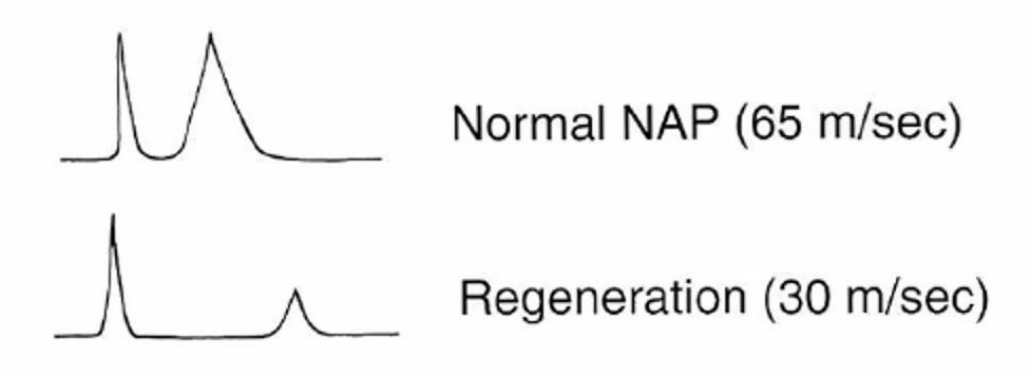
B. **Modest scar** → electrically stimulate nerve proximal to injury and look distally for evidence of muscle contraction or transmission of nerve action potentials (***intraoperative action potentials***):

N.B. do not use *local anesthetic* if planning to use nerve stimulation intraop!

a) no transmission across area of injury → ***excise*** neuroma → cable ***graft***

b) transmission across area of injury → ***external neurolysis***; sometimes ***internal neurolysis*** is performed to test more selectively for nonfunctional fascicles that need excision repair.

N.B. neurolysis benefit hard to demonstrate (unless fibrosis)



EPINEURAL vs. **FASCICULAR** repair - neither technique is superior!

A. EPINEURAL repair appears appropriate for most cases!!!

B. FASCICULAR (PERINEURAL) repair is technically more challenging and more traumatic to nerve (extra sutures add to scar tissue production); conditions for fascicular repair:

1) done in first 48-72 h.
2) nerve is cut distally (clear distinction can be made between sensory and motor divisions of nerve).

How to improve proper fascicular alignment:

1) epineurial vessel alignment
2) serial cross-section topography - gross fascicular matching
3) stimulation: "awake surgery"; still must trace out distal components

Technique

- ***clean adventitial*** connective tissue.
- dissect back to healthy fascicles.
- place nerve ends on blue background and align
- ***ends are trimmed*** to healthy nondamaged axons (use blade breaker or razor blade chip against cutting surface).
- keep nerve ends moist.
- precise suturing - use *magnification* (up to operating microscope).
- suture:

size (suture gauge & needle fineness must be consistent with nerve size):

8-0 for epineurium
10-0 for perineurium

material NYLON, POLYESTER, POLYPROPYLENE.
suture **strength** is less of consideration than degree of **inflammatory & fibroplastic reaction**.
Use minimum number of microsutures!!! (consider use of **fibrin glue** - itself or to reinforce)

- *deep side of anastomosis* is performed first (after two sutures are placed at each side of line bisecting horizontal axis for orientation; this also aids in nerve rotation), *superficial repair* is accomplished last.
- suture has to ***closely approximate epineurium*** (to prevent regenerating axons from escaping, to prevent separation), but avoid constriction.
- recovery is better if anastomosis is ***tension-free*** + ***without damage to blood supply***; within 3 weeks after injury, nerve may lose 8% of its length (10% stretch is generally considered OK); measures to reduce tension:
    a) nerve mobilization
    b) release bands, e.g. lacertus, PT, FDS/carpal tunnel
    c) interposition graft
    d) joint flexion
    e) rerouting nerve across joint (e.g. anterior ulnar transposition – useful in elbow and proximal, but not in forearm)
    f) bone shortening
- how to know if it is still too tight: one or two 9-0 stitches in digital nerve and put through a full ROM.

NEUROMA IN CONTINUITY:

A) complete loss of motor function of 3-12 mo duration + intraoperative nerve action potentials show ***no regeneration*** across site of injury: neuroma ***excised*** → primary ***neurorrhaphy*** (or cable grafting).
B) intraoperative nerve action potential show ***recovery of function***: external or internal (interfascicular) ***neurolysis***.

CABLE GRAFTS

Graft undergoes Wallerian degeneration and provides ***mechanical*** NO TENSION ***guidance*** for ingrowing axons!

- indicated when destroyed length of nerve gap > 2-3 cm
  N.B. whenever going to OR – have **stimulator** ready + prep for nerve **graft donor** site!
- use graft that is 10% longer than the defect.
- reinforce anastomosis sites with fibrin glue or tube.

Sources for grafts:

1. **Autografts** – golden standard!
    1) *sural* nerve
    2) *antebrachial cutaneous* nerve (*medial* or *lateral*)
    3) *lateral femoral cutaneous* nerve
    4) *superficial radial* nerve
    5) any ***amputated extremity***.
2. **Allograft** - requires immunosuppression for ≈ 1 year (exception - ***cryopreserved allografts***, ***acellular allografts***)

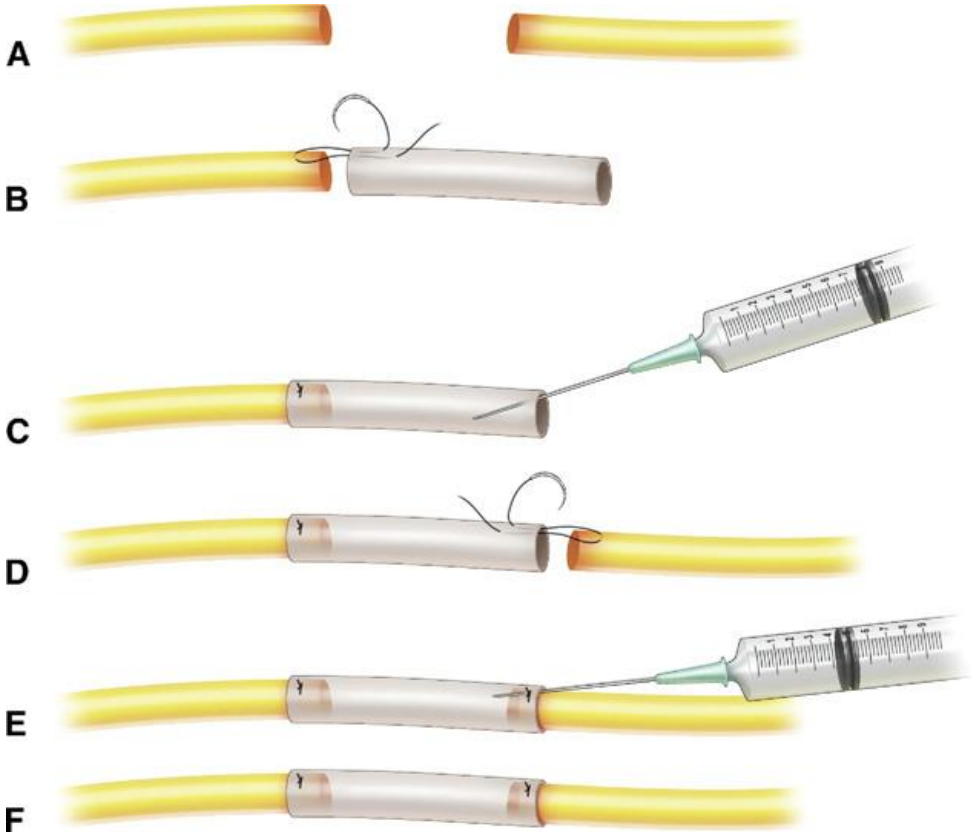
Do not use allografts on Boards!

CONDUITS (TUBES)

Do not use tubes on Boards!

1. **Biological conduits**:
    1) ***predegenerated or fresh skeletal muscle***.
    2) ***arteries, veins***
    3) ***mesothelial chambers***
    4) ***epineural sheath***
2. **Artificial conduits** (e.g. type I collagen, silicone, polyglycolic acid (PGA), polycaprolactone)

May be used as standalone without neurorrhaphy if gap is ≤ 5 mm



AMPUTATED STUMP NEUROMA

1) daily ultrasound for 5-10 sessions
2) injection of corticosteroids
3) ***sharply sectioning*** nerve proximal to neuroma → ***embedding*** freshly sectioned nerve end in adjacent deep soft tissue (surrounded by muscle).

N.B. most common cause of stump pain is poorly fitted prosthetic socket;
other common cause is spur formation at amputated end of bone; diagnosed by palpation and x-ray; H: surgical resection

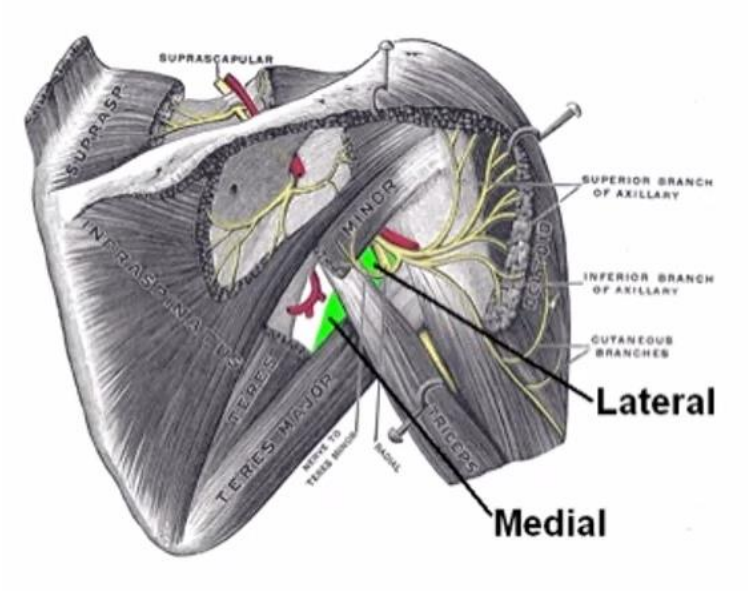
UNRESTORABLE / UNSUCCESSFUL NERVE REPAIR

1. **Tendon transfers** - to increase extremity function.
    e.g. posterior tibialis tendon passing through interosseous membrane → added power to foot with peroneal deficiency.
2. **Nerve transfers (neurotization)**

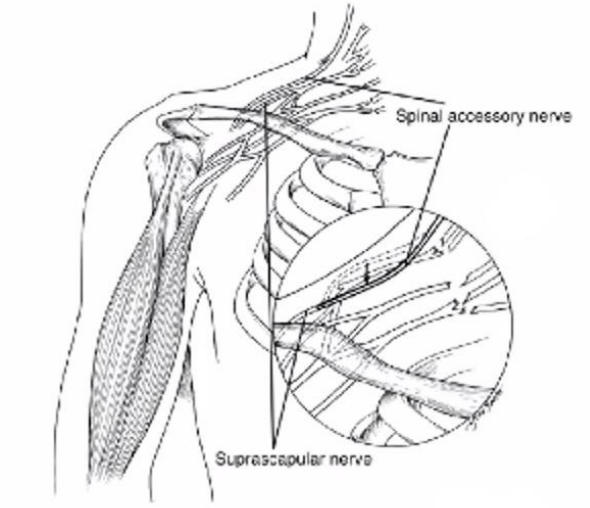
- better to sacrifice a branch or fascicles closer to end plates.
- most commonly to musculocutaneous, axillary, suprascapular nerves.

- neurotization procedures of median, ulnar, and radial nerve do poorly.

Donors to **axillary**
Radial nerve (its branch to long head of triceps)

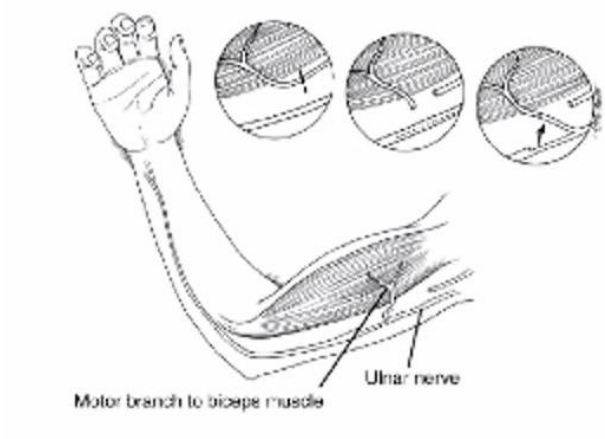


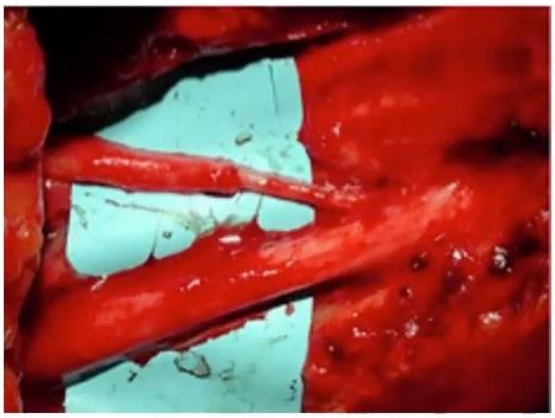
Donors to **suprascapular**
Spinal accessory nerve



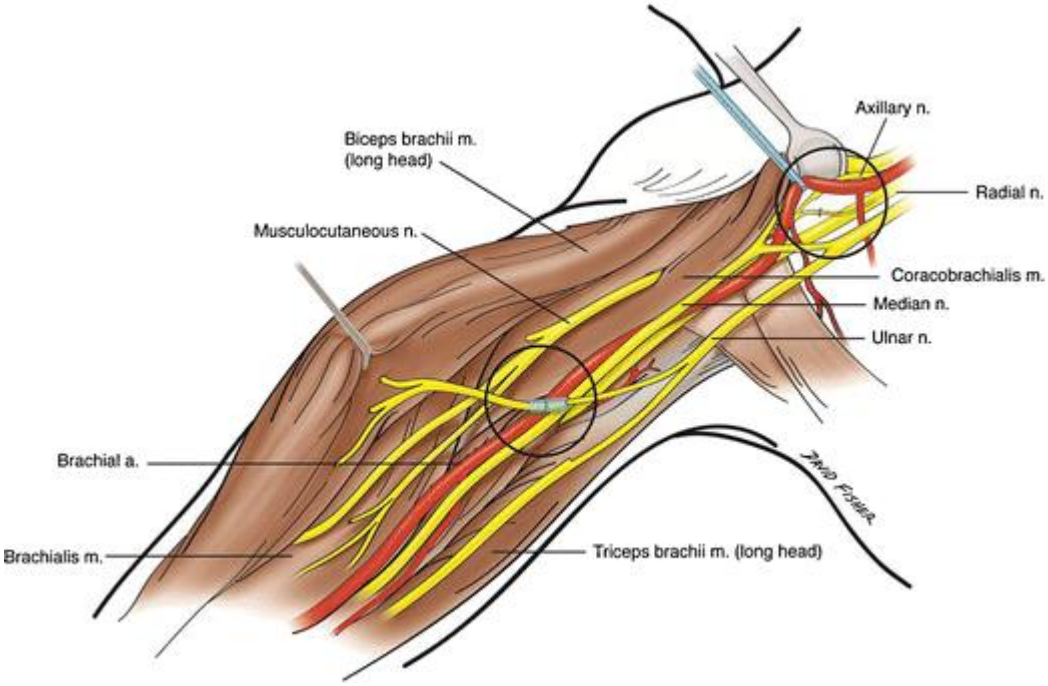
Donors to **musculocutaneous**
Intercostal nerves - usually 3rd, 4th and 5th intercostal nerves are used
Ulnar nerve (its fascicle to flexor carpi ulnaris) (**Oberlin procedure**):

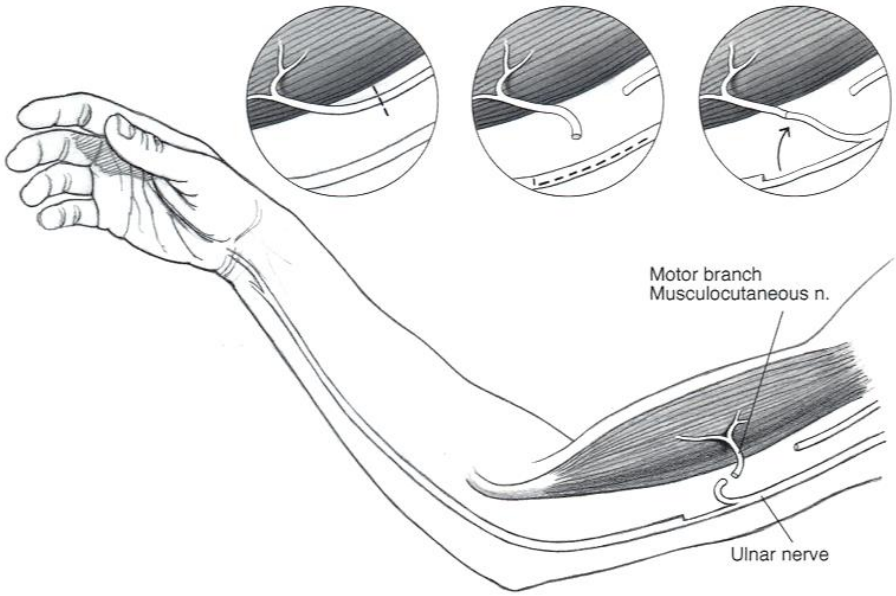
- rapid recovery rate in 90% of patients with M4 results.
- donor fascicles should be assessed intraoperatively with a nerve stimulator - branch going to FCU is selected for transfer (no obvious donor deficits postop).





picture also shows partial radial to deltoid fascicle of axillary nerve neurotization:





POSTOPERATIVE

- protect repairs by **relaxed joint posturing** for ≈ 3 weeks.
- begin **PT** 3 weeks after operation

See Case PN2 >>

HEREDITARY

Hereditary SENSORY and MOTOR neuropathies ≈ 90% of all hereditary neuropathies!

Charcot-Marie-Tooth disease 1 (peroneal muscular atrophy) - ***intrinsic foot*** & ***peroneal*** muscles – "stork" legs

Tomaculous neuropathy - recurring sensory and motor demyelinations brought on by mild pressure or trauma

Hereditary SENSORY and AUTONOMIC Neuropathies - **INSENSITIVITY TO PAIN!!!**

Familial dysautonomia - LACK OF FUNGIFORM PAPILLAE on tongue (→ hypogeusia) + prominent **dysautonomia**

Hereditary SENSORY Neuropathies

COMPRESSIVE

Pressure-induced injury to segment of peripheral nerve secondary to anatomic / pathologic structures

Most frequent:
1. Carpal tunnel syndrome
2. Ulnar nerve compression at elbow.

Entrapment neuropathies may be associated with:
1. Diabetes mellitus
2. Hypothyroidism: due to glycogen deposition in schwann cells
3. Acromegaly
4. Amyloidosis: primary or secondary (as in multiple myeloma)
5. Carcinomatosis
6. Polymyalgia rheumatica
7. Rheumatoid arthritis: 45% incidence of 1 or more entrapment neuropathies
8. Gout

- patients with *any polyneuropathy* are more vulnerable to mechanical injury of nerves!!!
- nerve compression affects ***myelinated fibers*** first (A type > B type > C type)
  N.B. larger fibers are more susceptible than small fibers
  - **brief** compression primarily affects myelinated fibers, and classically spares unmyelinated fibers
  - **chronic** compression affects both myelinated and unmyelinated fibers → demyelination, axon loss, fibrosis-neuroma.

DOUBLE CRUSH SYNDROME

- coexistence of compressive lesions in series along course of peripheral nerve, with one lesion rendering nerve susceptible to distal or proximal compression.

CLINICAL FEATURES

- temporal sequence: **irritative sensory symptoms** (pain, paresthesia) → **ablative sensory symptom** (numbness) → **ablative motor signs** (weakness and atrophy).
  sensory loss is less extensive than anatomic distribution of nerve!
- in major mixed nerve (e.g. sciatic, median) **sympathetic dystrophy** may be prominent.
- ***palpate*** entire length of nerve to check for masses, points of tenderness, bony abnormalities.

N.B. referred pain with entrapment neuropathy can radiate proximally (mimics radiculopathy)!!!
Referred pain is so common that Frank Mayfield once said that *patients with nerve entrapment don't know where the problem is located*

DIAGNOSIS

Diagnosis is **clinical**!

For compressive neuropathies:

Always check Tinel sign!

Always order EDX

May add US (not pricy MRI)*

*uncommon entrapments (e.g. Guyon, PIN) – order MRI!

- **nerve conduction** abnormalities across entrapment tunnel.
- **EMG** - signs of *denervation* (but only after > 3 weeks)
- **ultrasound** – swollen nerve
- **MRI** using short inversion imaging recovery technique (**STIR**) - *high signal in thickened nerve* at site of compression (edema).
- **MR neurography** - only large nerves (ulnar, median, sciatic).

Laboratory tests - recommended only if underlying peripheral neuropathy is suspected (i.e. unclear etiology in a young individual with no risk factors such as repetitive hand use):

1. HgA1c (DM)
2. BMP (uremic neuropathy)
3. Thyroid hormone levels (myxedema).
4. Vit. B12 levels
5. Multiple myeloma: anemia, 24 hour urine for kappa Bence-Jones protein, SPEP, skeletal radiologic survey.

TREATMENT

Conservative therapy should be tried first.

- adopt ***avoidance behaviors***.
- various ***splints*** and ***paddings***
- steroid injections.
- PT, TENS
- Botox injections
- local measures – lidocaine patch, capsaicin cream, ice applications.
- NSAIDs, antiepileptic, antidepressant, and narcotic pain medications.

Surgical:

1) decompressions, s. **external neurolysis** (incl. endoscopic techniques).
2) **neurectomy**

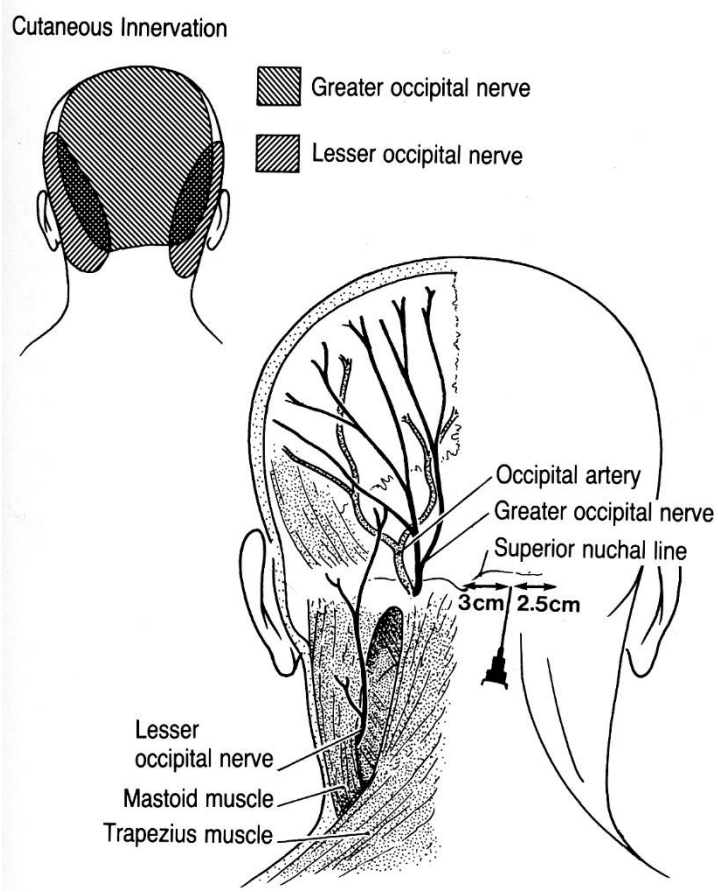
3) **stimulators**

N. OCCIPITALIS

- greater occipital nerve (nerve of Arnold) - sensory branch of C2.
- occipital neuralgia: pain in occiput + trigger point near superior nuchal line.
- traumatic cervical extension may crush C2 root and ganglion between C1 arch and C2 lamina.

TREATMENT

- PT, TENS
- Botox injections
- **trigger point injection**
- surgical nerve **decompression or neurectomy**:
    - occipital nerve pierces cervical muscles ≈ 2.5 cm lateral to midline, just below inion.
    - Doppler localization of accompanying greater occipital artery helps to locate the nerve
- decompression of C2 nerve root if compressed between C1 and C2 ± atlanto-axial fusion.
- occipital nerve **stimulators**.

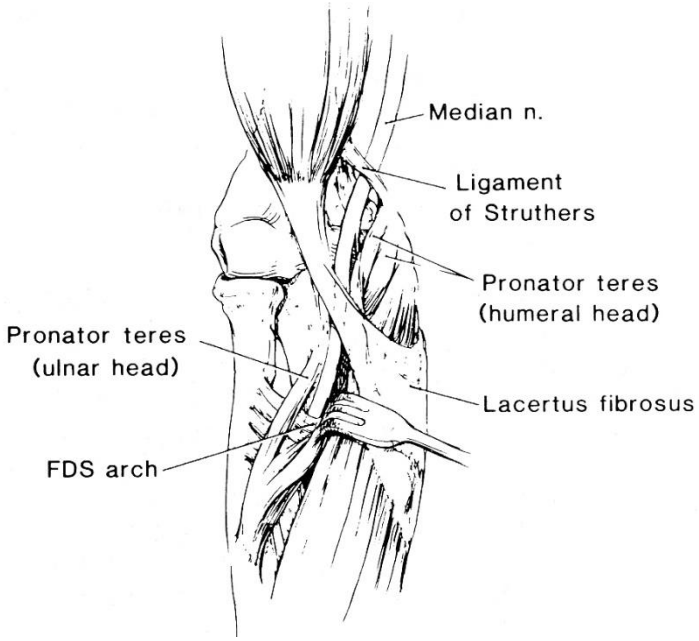


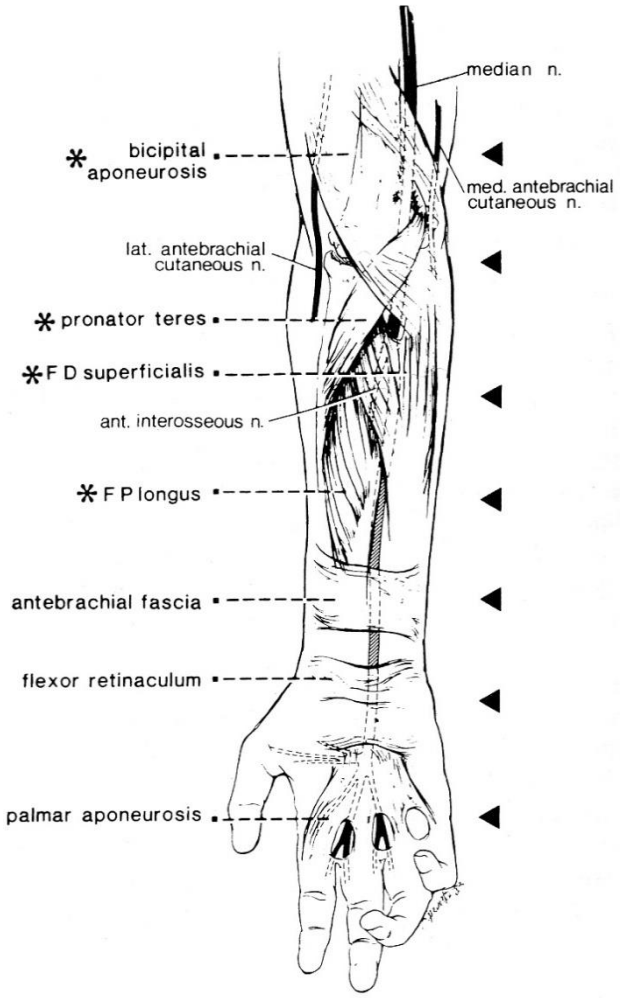
N. MEDIANUS

PLACES OF COMPRESSION

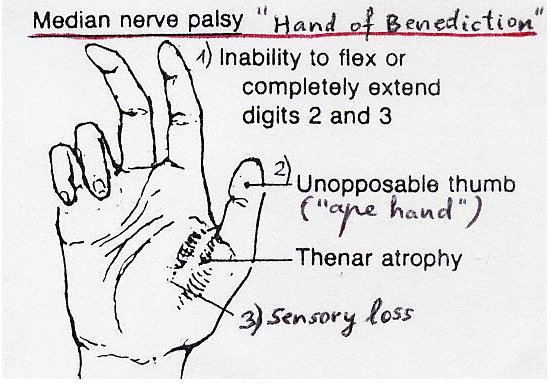
1. ***Within carpal canal*** (**carpal tunnel syndrome**)

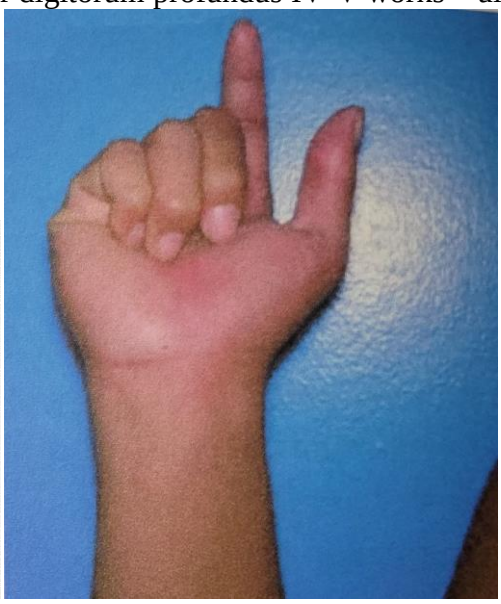
2. ***Near elbow*** (**proximal median neuropathy**):
   1) ligament of Struthers / supracondylar process of humerus – those are abnormal structures (vs. Struthers arcade)
   2) lacertus fibrosus (bicipital aponeurosis)
   3) between two heads of hypertrophied pronator teres (pronator teres syndrome)
   4) flexor digitorum profundus fascial arch (sublimis bridge)
   5) direct external compression ("honeymoon palsy"), needle injury during cubital phlebotomy





- upon attempt to make a fist (only flexor digitorum profundus IV-V works – ulnar nerve):





Causalgia is most commonly associated with lesions of median nerve!
N.B. "benediction" vs "claw" – depends what you are asking patient to do!

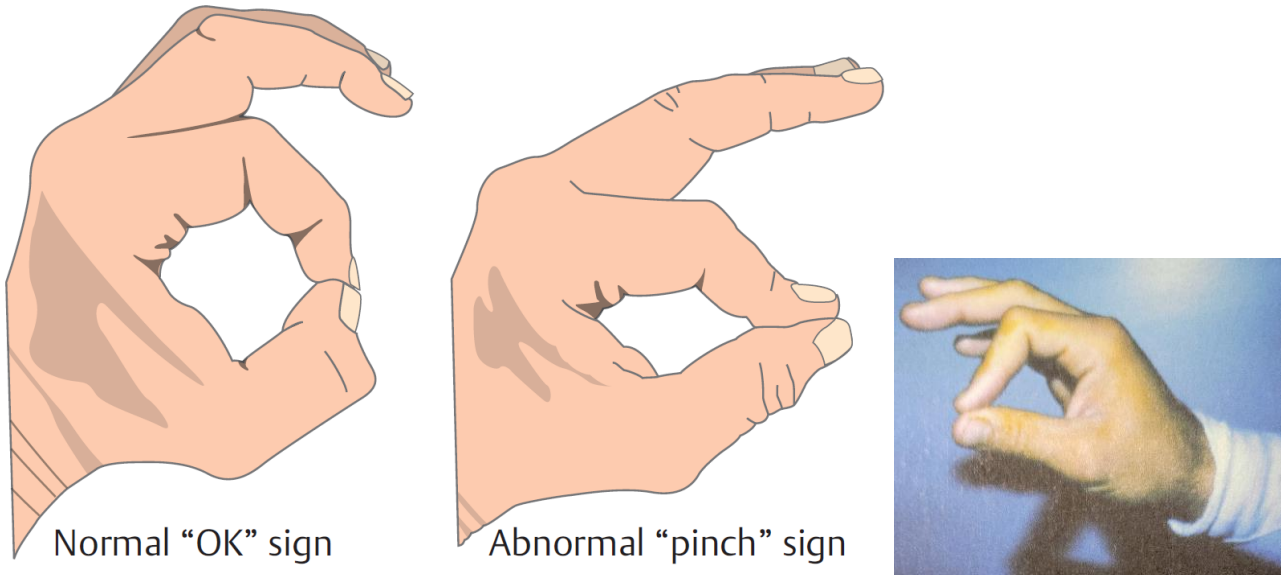
ANTERIOR INTEROSSEOUS NEUROPATHY

- **purely motor branch** of median nerve that arises in upper forearm (same as PIN of radial nerve)

No sensory loss + weakness of 3 forearm muscles:

1) flexor digitorum profundus (FDP) - flexion of distal phalanx of digits 2 & 3
2) flexor pollicis longus (FPL) - flexion of distal phalanx of thumb
3) pronator quadratus (in the distal forearm): difficult to isolate clinically (H: EMG)

Summary – distal phalanx of I-III digits → abnormal "OK" sign:



Important to evaluate pronator teres (abnormalities suggest involvement more proximal than forearm)!

MANAGEMENT

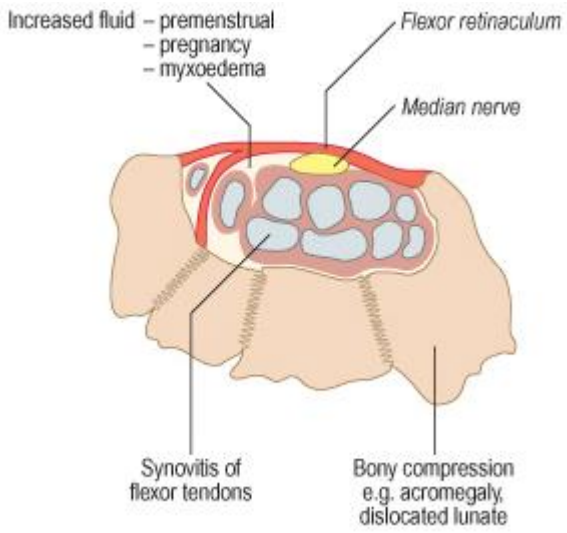
- wait 8–12 weeks → exploration (e.g. constricting band near the origin)

CARPAL TUNNEL SYNDROME (CTS)

- most common compressive neuropathy!
- 50% ***bilateral***, dominant side being affected more severely.

PRECIPITATING FACTORS

1) **overuse** - repetitive motion of fingers (frequent prolonged wrist flexion, especially with force) - often ***occupational***.
2) **pregnancy** (esp. fluid retention in 3rd trimester; resolves spontaneously after birth!)
3) **tenosynovitis**, **rheumatoid arthritis** (synovial hypertrophy), osteoarthritis, gout
   N.B. arthritis per se may cause thenar pain but no numbness (numbness is a must for CTS)
4) **trauma**: wrist fractures, lunate dislocation
5) ganglionic cysts
6) nerve sheath **tumor**
7) **hypothyroidism**, mucopolysaccharidosis, acromegaly, sarcoidosis
8) **diabetes mellitus** (microvascular injury)
9) **amyloidosis** (esp. **hemodialysis** - deposition of β-microglobulin derived amyloid, vascular steal from AV fistula)



CLINICAL FEATURES

Referred pain can ***radiate proximally*** - to the arm and even neck! (mimics C6-7 radiculopathy)

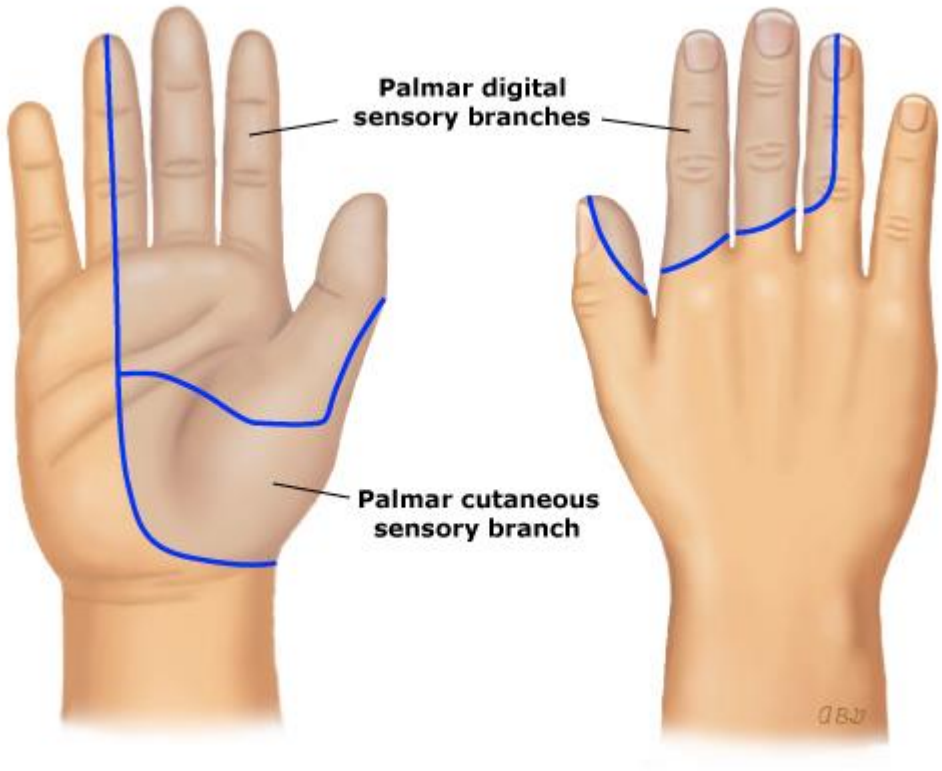
Mild disease: **paresthesias** & **pain** in median nerve distribution (after strenuous wrist movements or nocturnal*).

*because of ***venous stasis*** or because ***wrist falls into flexion*** with sleep

- pain is burning and may be severe (awakening from sleep with painful "hand falling asleep"); exacerbated by hand elevation.
    - patients often seek relief by: shaking or dangling or swinging the hand, opening and closing or rubbing the fingers, running hot or cold water over the hand, or pacing the floor
- grasping objects is painful and patients may report dropping cups and glasses.
- sensation in **thenar eminence** is not affected (*palmar cutaneous nerve* emerges from median nerve before carpal tunnel).

Median palmar cutaneous nerve arises from radial side of median nerve approximately 5 cm proximal to TCL and travels superficial* to carpal tunnel to provide sensory innervation to thenar eminence.

*thus, preserved in carpal tunnel syndrome!

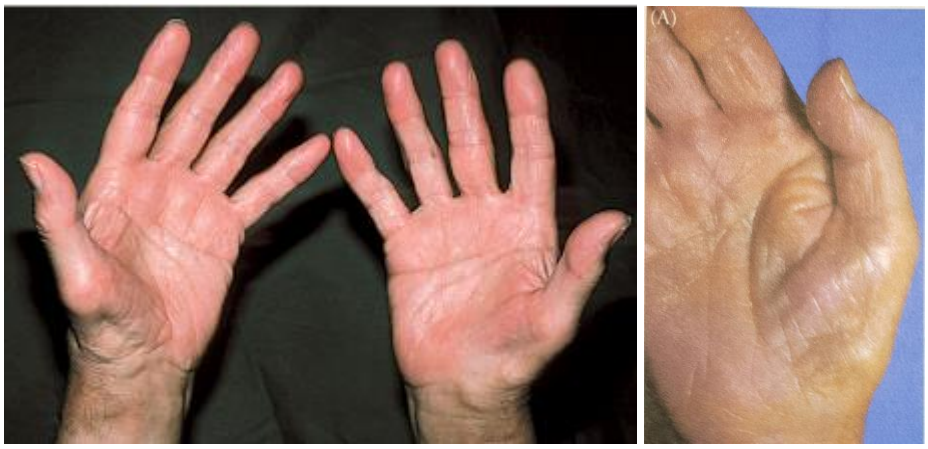


More severe disease: **sensory loss & weakness** (with THENAR **atrophy***).
*may be absent in patients with Riche-Cannieu anastomosis.

- hand grip weakness, especially opening the jar.
- hand clumsiness is more related to numbness than motor deficit

N.B. most reliably affected muscles:

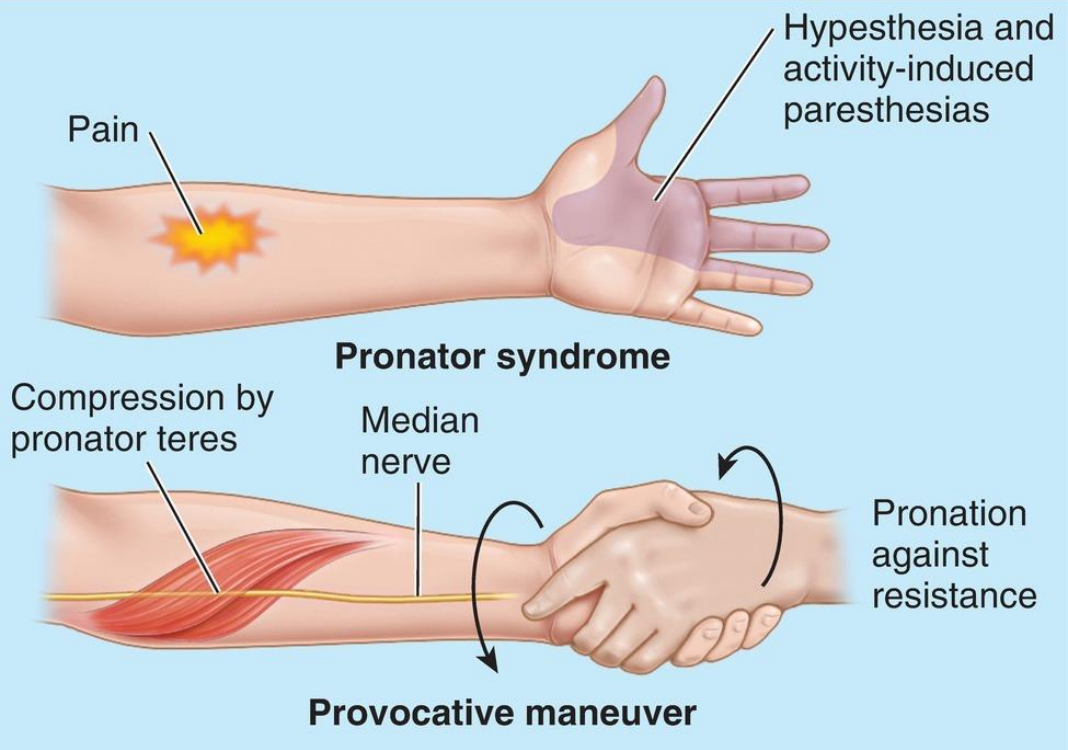
1) **opponens pollicis** – ability of thumb to move toward little finger against resistance
2) **abductor pollicis brevis (APB)**! – ability to lift thumb proximal phalanx against resistance



No weakness in more proximal muscles (difference from radiculopathy)

vs.

Proximal median neuropathy – tenderness along nerve course, motor deficit more widespread:
below elbow, only **AIN** branch (test distal I-II finger flexion - "OK" sign).
at elbow, **entire median nerve** - test **pronator teres**:



DIAGNOSIS

1) **TINEL sign** – only ≈ 50-60%
2) **PHALEN sign** (hold forcedly patient's wrist in acute 90 degree flexion for 30-60 seconds → paresthesias; sensitivity 80%):

3) **DURKAN compression test** - performed by examiner placing thumb over carpal tunnel and exerting downward pressure for 30 seconds - best sensitivity (82-89%) and specificity (90-99%)
4) **sensory nerve conduction slowing** (> 3.7 msec) across carpal tunnel (focal ***demyelination***; rarely can progress to axonal loss)

   Distal motor latency may be normal in 25% of patients!!!, i.e. sensory latencies are more sensitive than motor.

   - electrodiagnostic studies are also helpful in ***grading severity*** of CTS:
     mild: prolonged sensory latencies.
     moderate: + prolongation of motor distal latency.
     severe: + axonal loss
   - for uncertain cases compare median nerve sensory conduction velocity to ulnar nerve (or radial nerve): normal median nerve should be at least 4 m/sec faster than the ulnar.

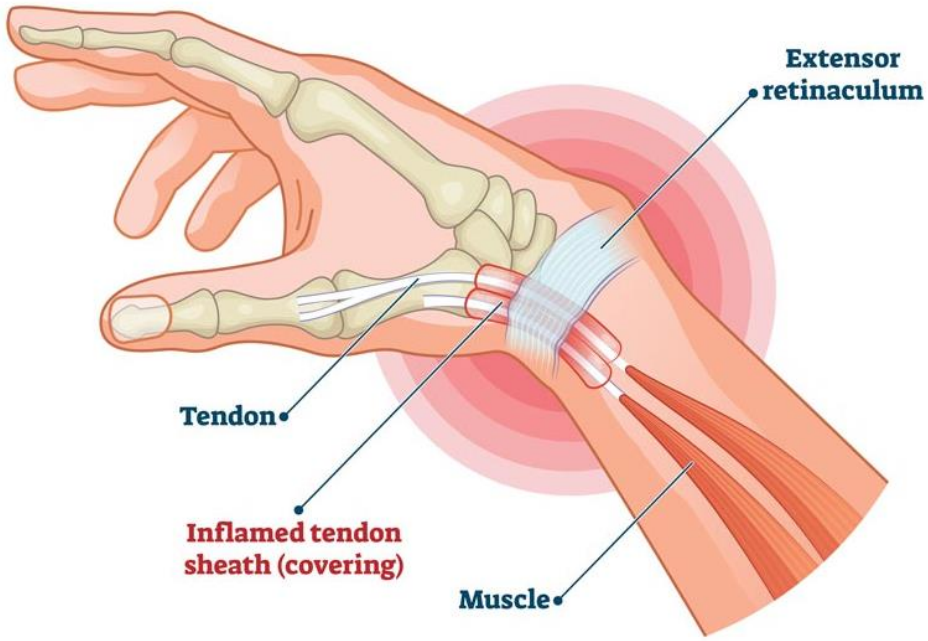
5) **EMG** (abductor pollicis brevis or opponens pollicis).
6) **MRI** of wrist (only indicated if mass is suspected).
7) **US** with 18 MHz probe.
8) lab tests (thyroid, DM, uremia, multiple myeloma) – if systemic disease suspected

DIFFERENTIAL

1) Main mimicker – C6 radiculopathy! CTS pain sometimes radiates *proximally* to forearm or even shoulder

   Double-crush syndrome – C6 root compression may interrupt axoplasmic flow and predispose nerve to compressive injury at carpal tunnel

2) **de Quervain's syndrome** - tenosynovitis of the abductor pollicis longus and extensor pollicis brevis tendons
   - NCVs normal
   - pain and tenderness in wrist near the thumb.
   - **Finkelstein's test**: thumb is passively abducted while thumb abductors are palpated, positive if this aggravates pain.
   - H: wrist splints and/or steroid injections.



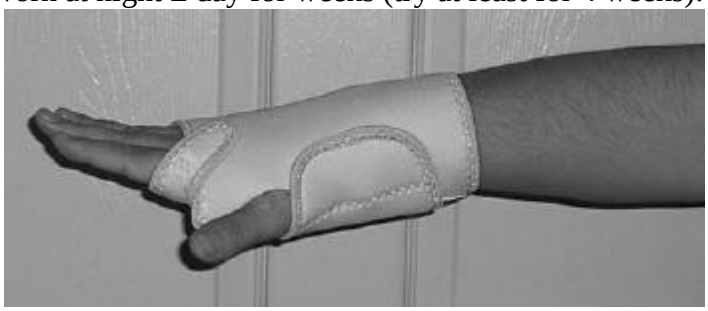
TREATMENT

American Association of Orthopedic Surgeons (AAOS) Clinical Practice Guideline (2010) endorsed by AANS, CNS, American Society of Plastic Surgeons, American Academy of PM&R and AANEM
Early surgery is an option when there is clinical evidence of median nerve denervation or the patient elects to proceed directly to surgical treatment.
Grade B, Level I and II: another non-operative treatment or surgery is suggested when the current treatment fails to resolve the symptoms within 2–7 weeks.
Insufficient evidence to provide specific treatment recommendations for CTS when found in association with diabetes*, coexisting cervical radiculopathy, hypothyroidism, polyneuropathy, pregnancy, rheumatoid arthritis, and CTS in the workplace.
Management specifics:

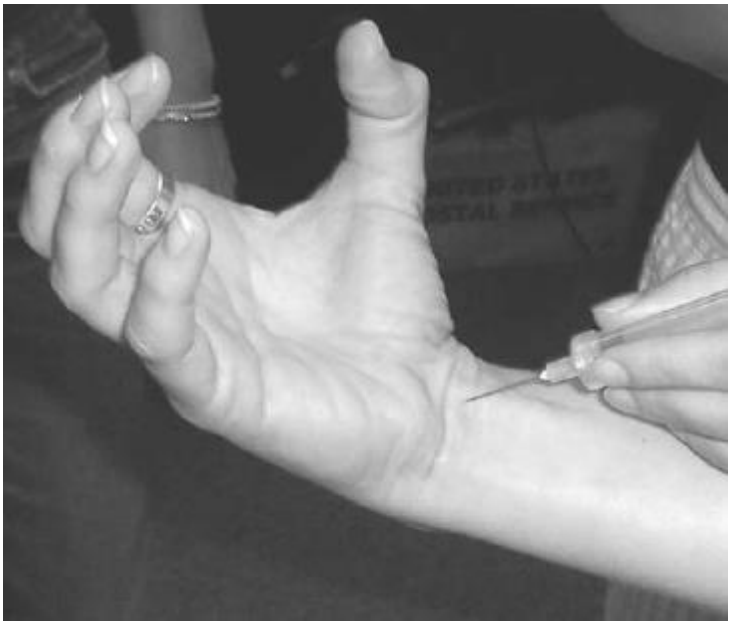
- local steroid injection or splinting is suggested before considering surgery (***Grade B, Level I and II***).
- oral steroids or ultrasound are options (***Grade C, Level II***).
- carpal tunnel release is recommended (***Grade A, level I***)
  *multiple studies report that the results of CTR in diabetics are good even when polyneuropathy is present.

CTS is usually progressive condition, but course of conservative therapy should be completed before surgical intervention:

1. **Ergonomic** corrections (do not return to heavy manual labor) and **rest**
2. **Splinting** of wrist in neutral / slight dorsiflexion (cross-sectional area↑ of carpal tunnel) - splint should be worn at night ± day for weeks (try at least for 4 weeks):



3. **Ultrasound** therapy
4. Injection of depot **corticosteroids** into carpal tunnel (***medial to m. palmaris longus tendon, just proximal to distal wrist crease***) - significant, but temporary improvement:
   N.B. aim to inject **tendon sheaths**; injection adjacent or into nerve is to be avoided! – all *steroids are neurotoxic* upon intrafascicular injection, and so are some of the carrier agents!
   - 10–25 mg HYDROCORTISONE.
   - *avoid local anesthetics* (may mask symptoms of intra-neural injection)
   - 25G needle.



5. NSAIDs, diuretics, vit. B_6 – ineffective!

SURGERY

Treat other causes for neurologic symptoms (e.g. cervical radiculopathy, C7 or middle trunk compression in thoracic outlet, proximal median nerve compression) first!

- CTR is indicated for recalcitrant, severe CTS cases (neurological deficits, duration > 1 year).

BILATERAL CTS

a) **simultaneous treatment** - reduction in total disability time and reduced surgical cost but compromised ability of patient to perform self-care.
b) **staged treatment** - treat more affected hand first, followed 2-6 weeks later by other hand.

ANATOMY

- carpal tunnel is 4-6 cm in length.
- carpal tunnel contains 9 structures:
    1) four tendons of flexor digitorum superficialis (FDS)
    2) four tendons of flexor digitorum profundus (FDP)
    3) tendon of flexor pollicis longus
    4) median nerve - travels between transverse carpal ligament and second and third FDS tendons; nerve is positioned slightly to the radial side in carpal tunnel.

Roof of carpal tunnel - **FLEXOR RETINACULUM**

- transverse carpal ligament (TCL) arches between **pisiform** and **hook of hamate** *ulnarly*, and **scaphoid tuberosity** and **crest of trapezium** *radially*
- TCL is 3-4 cm in width and 2.5-3.5 mm in thickness

N.B. TCL extends distally into palm ≈ 3 cm beyond distal wrist crease!

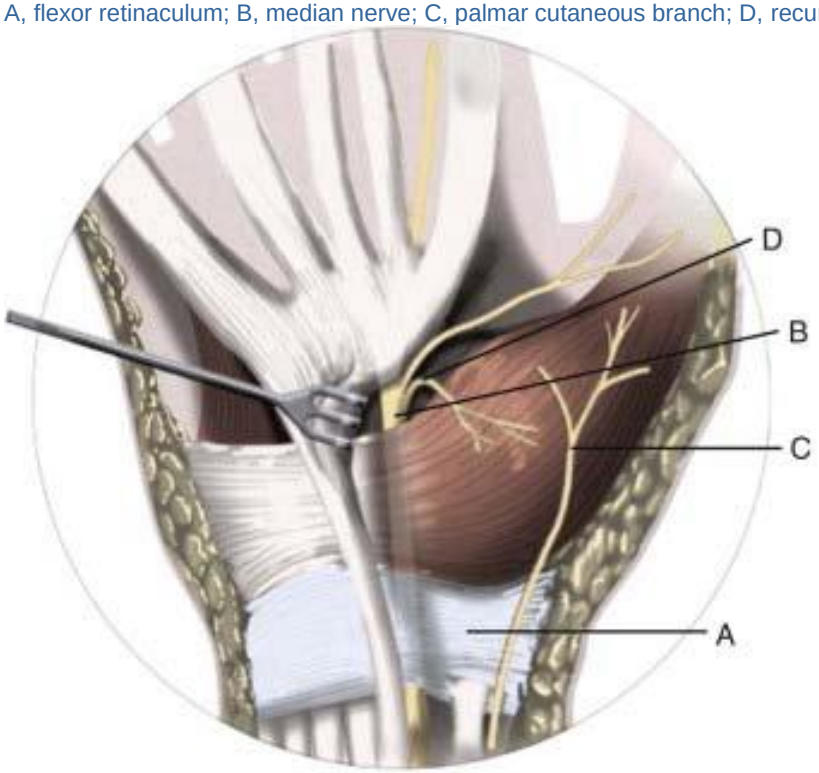
- palmar fascia is fused to TCL proximally and then fans out to soft tissue of palmar skin as *palmar aponeurosis*

FLEXOR RETINACULUM = TCL + proximal palmar fascia

Surgeon cuts TCL (transverse fibers); flexor retinaculum fibers are longitudinal

- ulnar nerve & artery run superficially on ulnar side of the TCL.

A, flexor retinaculum; B, median nerve; C, palmar cutaneous branch; D, recurrent motor branch:



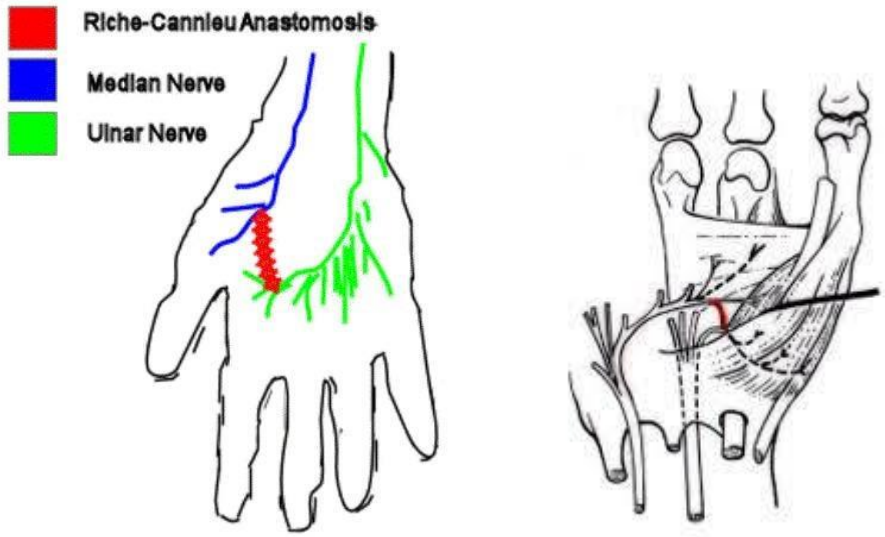
Recurrent motor branch - innervation to "LOAF muscles" (Lumbricals 1 & 2, Opponens, Abductor and Flexor pollicis brevis) - arises from median nerve distal to TCL.

transligamentous (20%) – maybe injured during carpal tunnel release.

- called "million dollar nerve" - loss of thumb function → possible malpractice lawsuit.

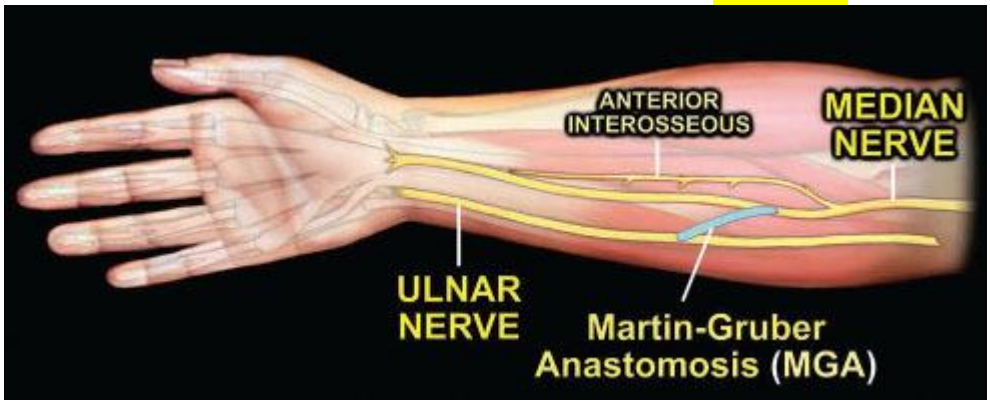
Riche-Cannieu anastomosis - anatomic variant: communication between recurrent branch of median nerve and deep branch of ulnar nerve in hand - innervation of thenar muscles is provided by ulnar nerve

Riche-Cannieu Anastomosis



- present in 70-77% of hands.
- examples of confusion this might cause:
  (1) Median lesion could cause denervation in typically ulnar muscle, such as adductor digiti minimi or first dorsal interosseous muscle.
  (2) Ulnar lesion could cause denervation in typically median muscle, such as flexor pollicis brevis or abductor pollicis brevis; most extreme version is so-called **all-ulnar hand** (very rare).

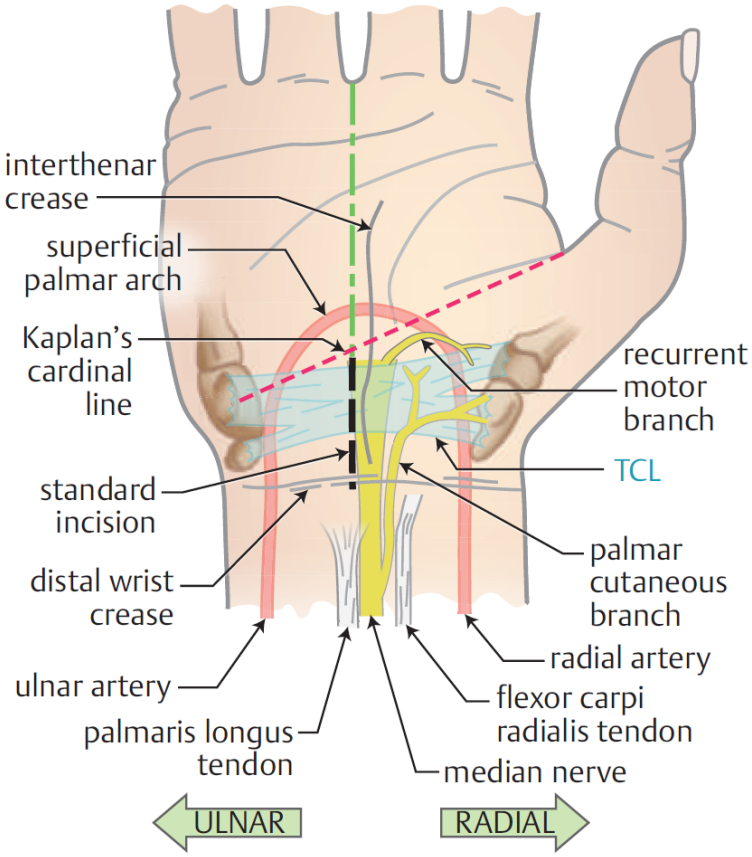
Martin-Gruber anastomosis - connection from median nerve to ulnar nerve in forearm:



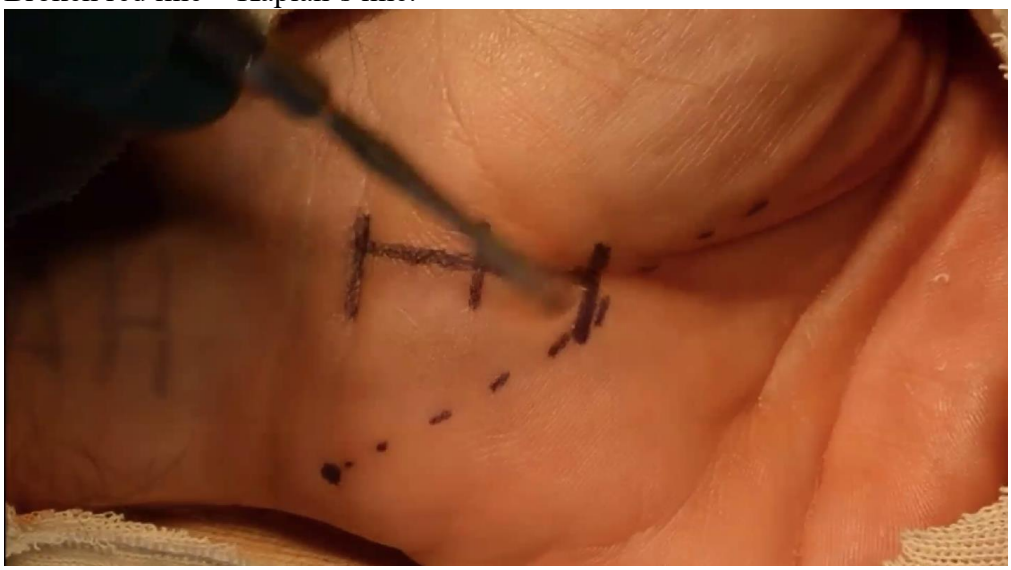
- in the setting of proximal ulnar nerve injury, anastomosis can prevent complete paralysis of intrinsic hand muscles

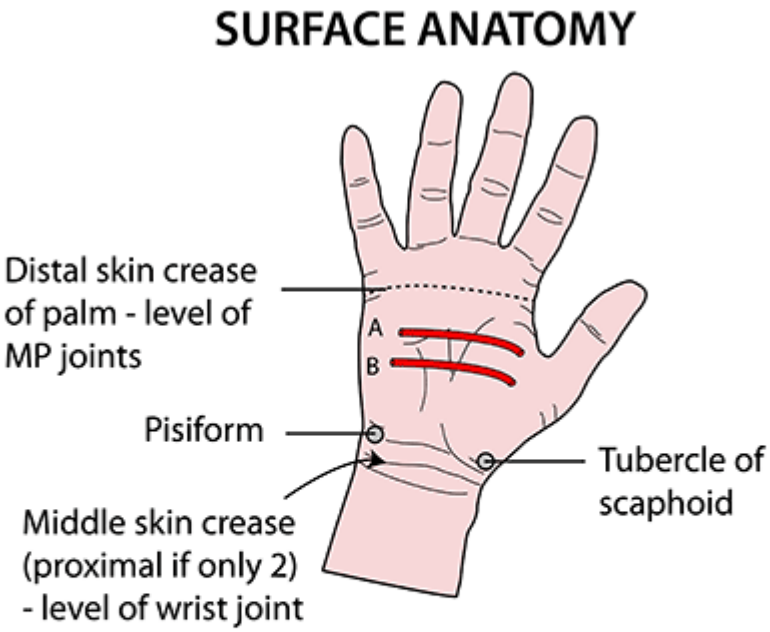
ARTERIAL ARCS IN PALM

Kaplan's cardinal line: runs from base of thumb web space to hook of hamate; superficial palmar arch (from ulnar artery; vs. deep arch is even more proximal – from radial artery), which is vulnerable during carpal tunnel surgery, is distal to this line:



Broken green line = trajectory of incision (parallel to interspace between digits #3 & 4).
Broken red line = Kaplan's line.





A Superficial palmar arch
Level with outstretched thumb.
From ulnar artery.
1/2 way between distal palmar crease and distal wrist crease

B Deep palmar arch
From radial artery.
1cm proximal to superficial arch

TECHNIQUE

Surgical release of transverse carpal ligament (TCL) – successful in 70-90% cases.

- RA patients – also add ***tenosynovectomy***.
- most reliable guide to nerve at operation - **m. palmaris longus tendon** - inserts in palmar aponeurosis and lies directly over median nerve just proximal to TCL.
  m. palmaris longus is absent in 10-25% individuals.
- 10% ulnar nerves and 4% ulnar arteries lie radial to hook of hamate outside of Guyon canal - risk for injury during carpal tunnel surgery.

- supine with arm abducted 60 degrees and forearm supinated on arm board.
- tourniquet is optional.
- wrist is often placed on a roll to provide wrist extension.

a) local nerve block (0.25% bupivacaine containing 1:200,000 epinephrine) – infiltrate along incision
b) axillary block
c) Bier block.

Endoscopic – earlier return to work but more neurapraxia.

SKIN INCISION

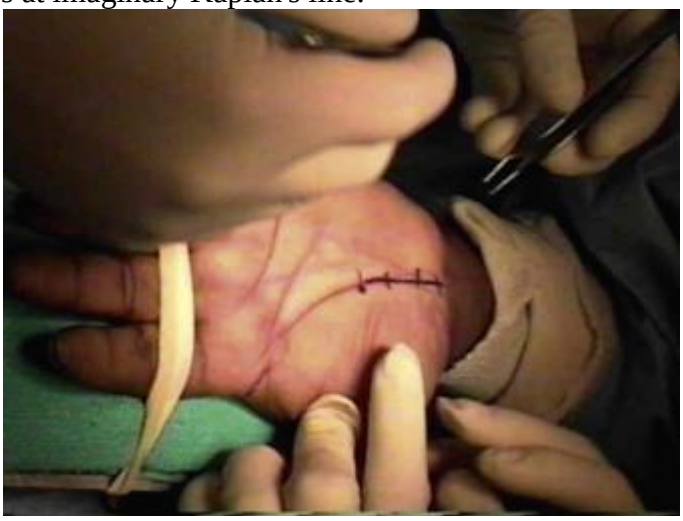
- straight or slightly curvilinear, 3-4 cm
  Incision cannot be too short but *postoperative pain is proportional to length of incision*.

a) just ulnar to **m. palmaris longus tendon** (≈ longitudinal midpalmar crease or 6 mm to ulnar side of thenar crease)

- proximal end - at *proximal wrist crease* (exposes median nerve just before it dips under flexor retinaculum);
- *zigzag* across wrist (to avoid scar which restricts motion).
- distal end stops ≈ 1.5 cm beyond *distal wrist crease*

b) along (or just ulnar to) **middle palmar crease**

- proximal end - at *distal wrist crease*
- distal end stops at imaginary Kaplan's line.



- if tendons of flexor digitorum superficialis are encountered = you are too deep and need to back out and look more radially (toward the thumb) to find the nerve.

RETINACULUM CUT

- cut over Penfield 4 to protect nerve!
- accurately over midpoint of nerve surface (so as to avoid sliding down dangerous radial side of nerve
- motor branch to thenar)

Do not to injure ulnar neurovascular bundle by retractor blade!
Also recurrent motor branch of median nerve may be transligamentous or subligamentous! Recurrent_motor_branch

- proceed distally until deep palmar fat around superficial palmar arch is encountered.
- completeness of distal cut is confirmed by checking for remaining cross bands while dragging curve-tipped clamp backward toward opening.
- check completeness of proximal cut: skin is elevated to permit visualization 2-3 cm into forearm
- *neurolysis* of median nerve is not generally recommended.

CLOSURE

- meticulous hemostasis (if used, tourniquet should be released at this point).
- 2-0 or 4-0 nylon (vertical mattress) on skin
- bulky fluff gauze dressing is applied to volar surface, along with elastic wrap.

COMPLICATIONS

1. palmar **pain**
2. pillar **pain** along thenar & hypothenar eminences (related to adjustment of carpal bone alignments);
3. temporary **loss of grip** strength (secondary to relocation of origin of hypothenar and thenar muscles and bowing of flexor tendons through TCL incision).
4. **median nerve injury** → repair with epineurial sutures; may lead to CRPS (most cases are self-limited within 2 weeks).
5. **ulnar nerve injury** by retractor → observe

6. **hematoma** from "blind" distal division of TCL and injury to superficial palmar arterial arch → apply pressure, stitch repair, emergency evacuation of postop hematoma
7. **incomplete** TCL sectioning;
8. cutting *ulnar-median anastomotic branch Riche-Cannieu* (runs parallel to and ≈ 1 cm beyond distal TCL edge)
9. injury to *recurrent motor branch* (stay on ulnar side!) → explore ASAP and repair!
10. injury to *palmar cutaneous branch*
11. **hypertrophic scar** causing compression of median nerve - caused by incision crossing wrist perpendicular to flexion crease; H: avoid crossing flexion wrist crease, or in cases where necessary by crossing wrist obliquely at 45° angle directed toward ulnar side.

POSTOPERATIVE

- ACETAMINOPHEN WITH CODEINE for 3–4 days.
- remove elastic wrap after 6 hours; keep dressing for 2 days.
- keep hand elevated for several days.
- encouraged to perform gentle finger ROM exercises ASAP – ***move fingers frequently***
  - suture removal at 7-10 days.
  - return to work 3-4 weeks after surgery.
- ***postoperative splinting*** is not recommended.

OUTCOME

- 70-89% patient satisfaction
- in severe cases, nerve recovery may not occur; symptoms persist in 6% of patients.
  - if repeat NCS are worse or if the EMG needle exam has findings of denervation not previously present, then repeat surgery is indicated.
  - if preoperative study is not available, repeating study with comparison at 2 points in time to evaluate for improvement or worsening.

  N.B. prolonged latencies alone are not an indication for reoperation!!! (latencies continue to improve after successful CTR up to 2 years)

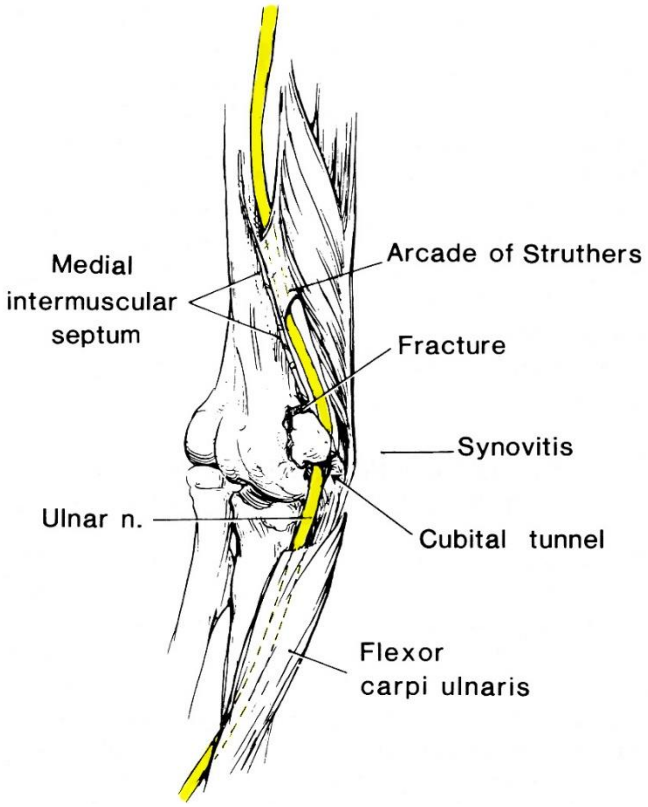
Recurrent CTS → image (US, MRI)
Progressive thenar atrophy (after CTR) → explore (incl. recurrent motor branch)

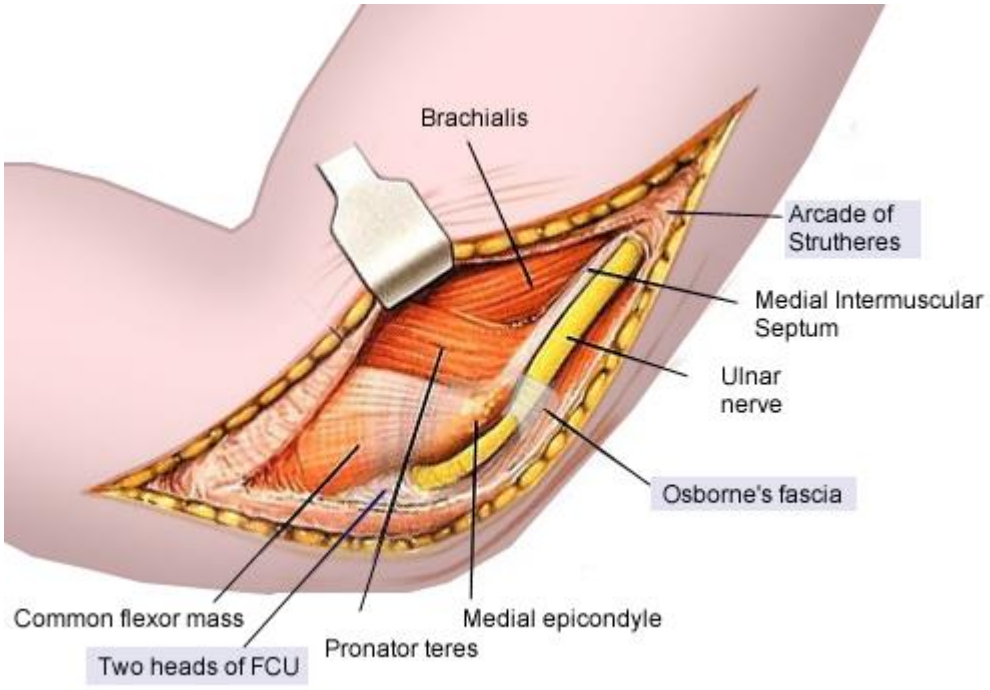
See Case PN6 >>

N. ULNARIS AT ELBOW

Places of compression (proximal to distal):

1. **Medial intermuscular septum** - sharp edge that can indent nerve (esp. after anterior transposition where nerve may be kinked).
2. **Arcade of STRUTHERS** (hiatus in medial intermuscular septum; tense sheet of fascia stretching from medial head of triceps to insert into medial intermuscular septum) 6-8 cm above cubital tunnel.
3. **ULNAR GROOVE** - between the medial epicondyle and olecranon process (most common *anesthesia-related* compressive neuropathy!!!).
4. **CUBITAL TUNNEL** just distal to the ulnar groove – compression between cubital tunnel retinaculum (**OSBORNE'S ligament**) and ***medial collateral ligament (MCL)***.
5. **Between two heads of flexor carpi ulnaris** (aponeurosis of flexor carpi ulnaris also referred to as **OSBORNE'S fascia**; 3-5 cm distal to cubital tunnel).





N.B. *elbow flexion* narrows cubital tunnel (flexion can cause *anterior subluxation* of nerve).

CLINICAL FEATURES

1) **paresthesias**, **pain**, **sensory loss** - little finger and ulnar half of ring finger; exacerbating activities include: N.B. sensory only IV-V fingers (vs. C8 – also ulnar forearm)

- peripheral nerves have much more precise sensory and motor borders (vs. radiculopathies): sensory loss at V and ulnar half of IV digit – ulnar neuropathy (not C8 radiculopathy)
- cell phone use (excessive flexion)
- sleeping with elbow in flexion → nocturnal paresthesia and pain.

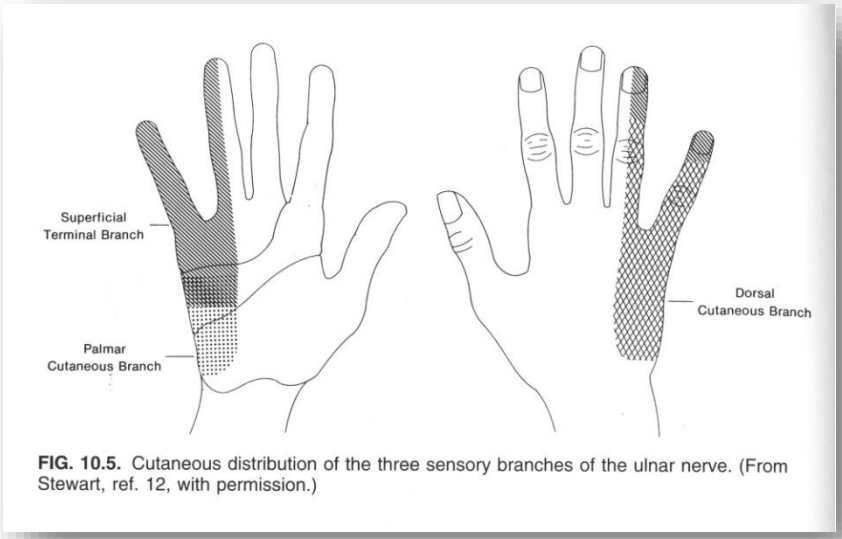
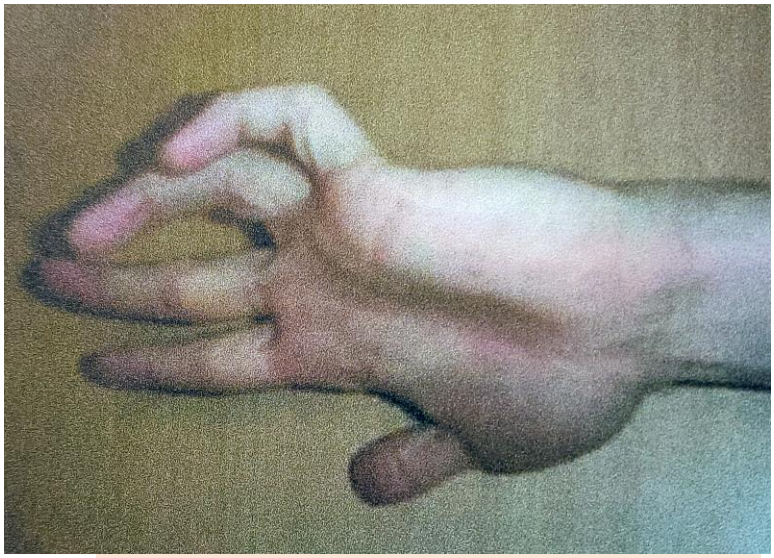


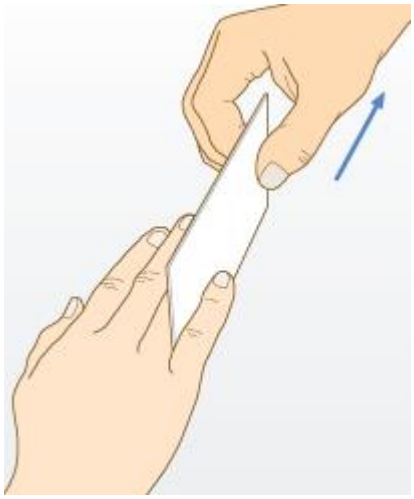
FIG. 10.5. Cutaneous distribution of the three sensory branches of the ulnar nerve. (From Stewart, ref. 12, with permission.)

2) attempt to extend fingers → only MCP IV-V joints extend, IP joints do not extend - **"claw hand" (main en griffe)**; hand clumsiness, dropping objects; hypothenar + interossei **weakness** and **atrophy**:



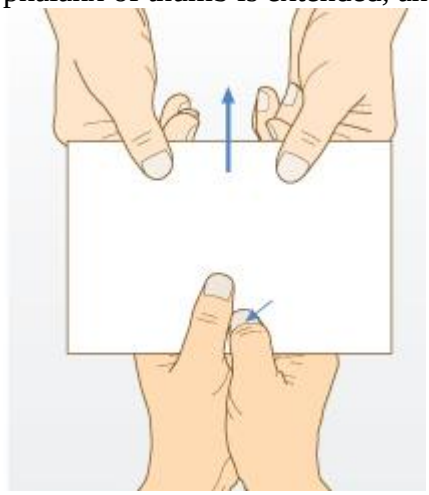
N.B. “benediction” (median) vs “claw” (ulnar) – depends what you are asking patient to do!

- ulnar neuropathy may start with motor signs before sensory (opposite to CTS) - because of predominance of motor fibers within UN!
- atrophy is most evident in the first dorsal interosseous (in thumb web space).
- test *interossei*: ask patient to hold sheet of light card between fully extended little and ring fingers:



- fifth finger may be abducted away from other fingers at rest (**Wartenberg sign**); patients complain of catching pinky finger when placing hand in pocket

– *m. adductor pollicis* weakness **FROMENT prehensile thumb sign (signe du journal)** - when sheet of paper, grasped between thumb and index finger, is pulled → proximal phalanx of thumb is extended, and distal phalanx is flexed:



Motor (differential from C8):

ulnar nerve innervates all intrinsic hand muscles, except LOAF (5 muscles): *abductor - opponens - flexor pollicis brevis, and lateral two lumbricals* ← innervated by C8 and T1 (recurrent motor branch of median nerve);

ulnar nerve does not innervate *flexor digitorum superficialis* and *first two flexor digitorum profundus muscles* ← innervated by C8 (median nerve)

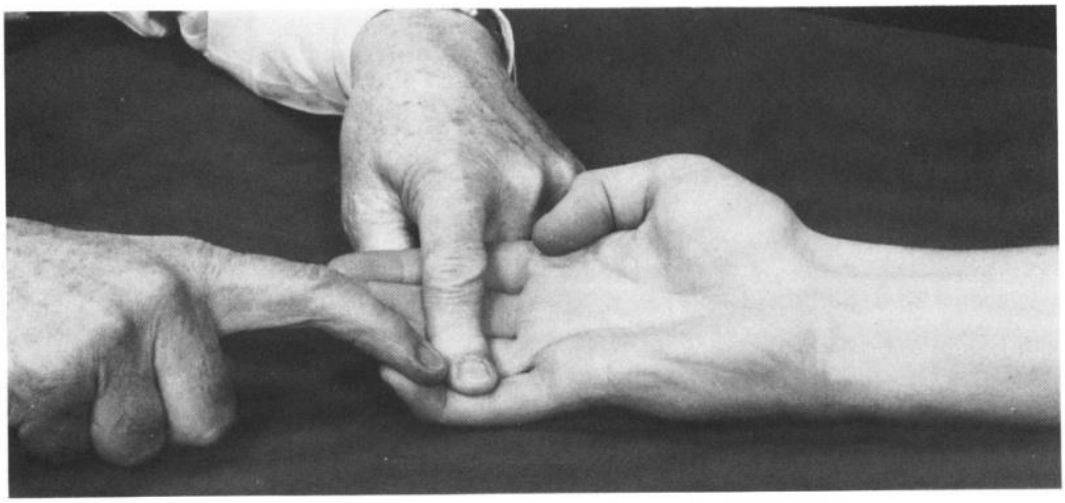
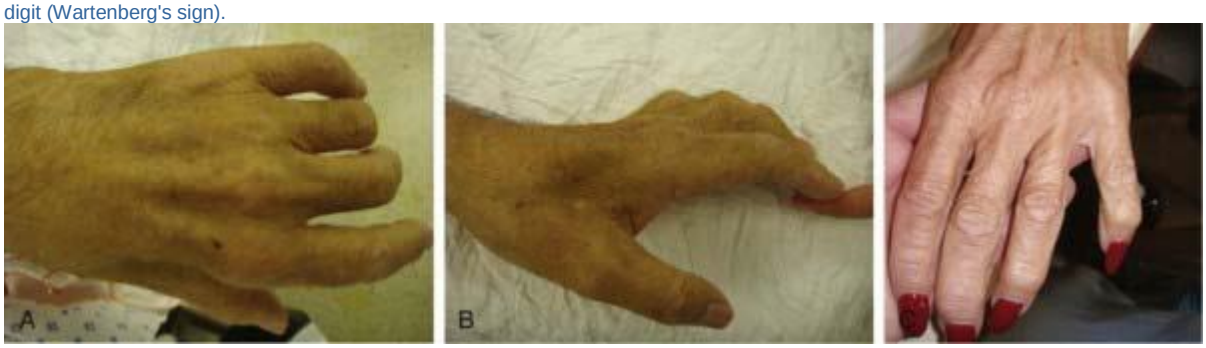


Fig. 39 Flexor Digitorum Profundus III and IV (Ulnar nerve; C7, C8)
The patient is flexing the distal interphalangeal joint against resistance while the middle phalanx is fixed.

A, Interosseous atrophy resulting in prominent metacarpal bones. **B,** Atrophy of the first dorsal interosseous muscle. **C,** Abduction at rest of the fifth digit (Wartenberg's sign).



- course can be prolonged – e.g. due to asymmetric bone growth after childhood fracture (**tardy ulnar palsy**).
- old, "burnt out" neuropathic hand is atrophic, thin-skinned but, surprisingly, painless and free of other sensory phenomena.

Proximal Ulnar nerve

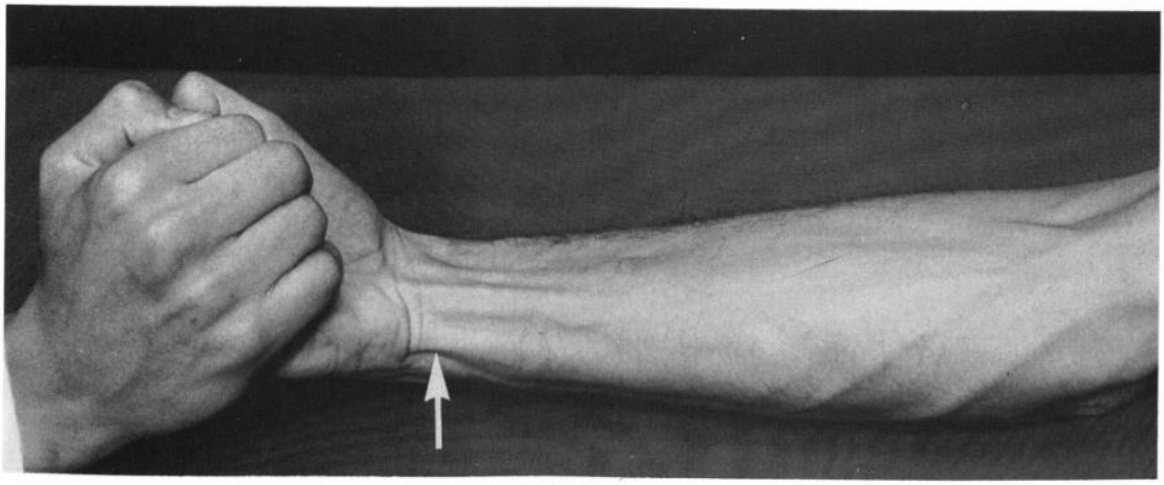


Fig. 38 Flexor Carpi Ulnaris (Ulnar nerve; C7, **C8,** T1)
The patient is flexing and adducting the hand at the wrist against resistance. *Arrow:* the tendon can be seen and felt.

DIAGNOSIS

1. **Nerve percussion** (TINEL sign) → paresthesias
2. **Elbow flexion test** - positive when flexion elbow for > 60 seconds → paresthesias
3. **Elbow pressure-flexion test** (sensitivity 91%) - elbow is flexed and pressure applied over cubital tunnel for 30 seconds → paresthesias
4. **Nerve conduction studies**
5. **EMG** - signs of denervation
   N.B. in contrast to CTS, which is predominantly demyelinating, UNE has more ***axonal loss***! – surgery results worse than with CTR
   N.B. in contrast to CTS, ***motor NCS findings*** are more useful for localization for site of entrapment than sensory abnormalities!
6. **Plain radiographs** of elbow - search for fracture / deformity when there is history of trauma.
7. **MRI** - increased T2 nerve signal; nerve subluxation / dislocation can be seen on axial images acquired during elbow flexion

DIFFERENTIAL

Referred pain with entrapment neuropathy can ***radiate proximally*** (mimics C8 radiculopathy)

N.B. *sensory testing of **dorsal** ulnar hand is important* – preserved sensation in this area with sensory deficits in ulnar distribution of fingers suggests entrapment at Guyon's canal (spared dorsal cutaneous branch distribution).

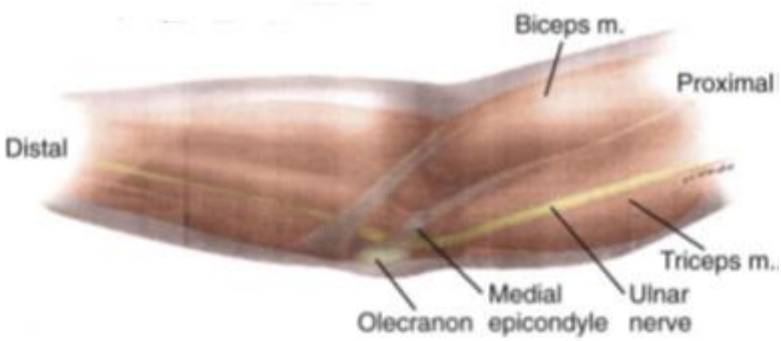
TREATMENT

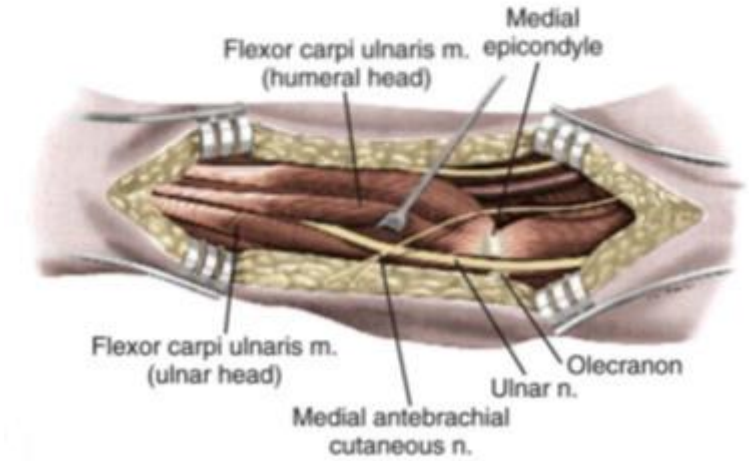
No guidelines or consensus!

1. **Half-splint with elbow pad** (elbow in gentle extension) at nighttime ± daytime.
2. **NSAIDs**

N.B. *steroid injections* have no role in treatment!
If treating conservatively, follow patient at 1-2 month intervals as long as stable or improving; if worsening → surgery

SURGERY





- anesthesia: MAC + local.
- **SUPINE** with arm abducted on armrest table

Neurolysis (In situ decompression)

- simple cubital tunnel release (sectioning taut aponeurosis).

- "lazy omega" incision over medial epicondyle of humerus
  a) *convex portion of incision faces anteriorly* – thus, away from tension when elbow is flexed, plus, not overlying nerve).
  b) if planning transposition - *convex portion of incision faces posteriorly* – this way transposed nerve will be protected under flap.

  N.B. *sensory nerve (medial antebrachial) runs across incision* in subcutaneous layer – try to preserve it (or annoying anesthesia will result)!
- division of the **cubital tunnel retinaculum**
- carefully dissect almost circumferentially (but < 360 degrees)* with care taken to preserve branches of feeding vessels.
  *ideally, nerve is not circumferentially dissected out! (devascularization, damage to slender branches to flexor carpi ulnaris; may make nerve to subluxate with elbow flexion).
- neurolysis is extended ***distally*** to point of nerve entry between heads of flexor carpi ulnaris (split FCU along fibers – rather far distally!).

- neurolysis is extended ***proximally*** to arcade of Struthers (divide it), medial intermuscular septum (between distal biceps and triceps muscles) must be cut in distal arm to prevent nerve from being kinked over it.
- once decompression is completed, *elbow is flexed and extended to look for nerve subluxation* - if significant subluxation is present, some surgeons believe that a transposition procedure is warranted.
- early mobilization of the arm starting on POD#2

Surgical failure usually is due to inadequate release:

a) at intermuscular septum
b) between heads of flexor carpi ulnaris.

- failure of ***in situ decompression*** → ***subcutaneous transposition***.
- failure of ***subcutaneous transposition*** → ***remove kinking*** of nerve at both ends of transposition.
- exquisite nerve tenderness in palpable subcutaneous course → ***submuscular transposition***.

Boards: recurrence after neurolysis → submuscular transposition (bigger incision: 10 cm above, 10 cm below epicondyle)

IN SITU DECOMPRESSION VS. NERVE TRANSLOCATION

Currently, there is no consensus whether in situ decompression or nerve translocation is the optimal surgical procedure.
Cochrane systematic review (430 patients) failed to find a significant difference in outcomes between the 2 procedures.

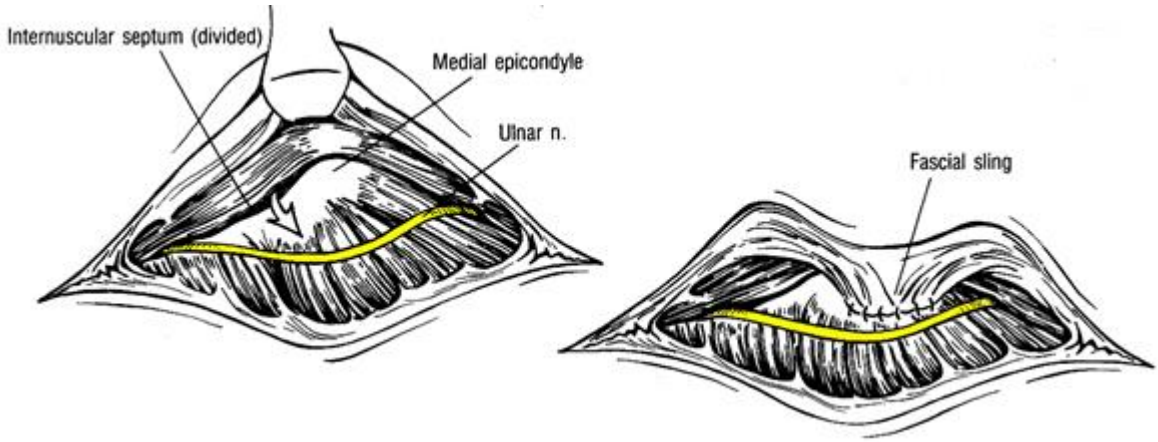
In situ decompression with Medial epicondylectomy

- removal of deformed medial epicondyle (medial wall of ulnar groove)

Subcutaneous transposition

- nerve mobilization and moving anterior to epicondyle

- ***avoid kinking nerve*** at both ends of transposition.
- fascial sling is created to hold nerve from slipping backward by row of absorbable sutures (between flap and surface of *m. pronator teres* just in front of medial epicondyle).



Intramuscular transposition

- postoperative scarring within intramuscular bed!

Submuscular transposition

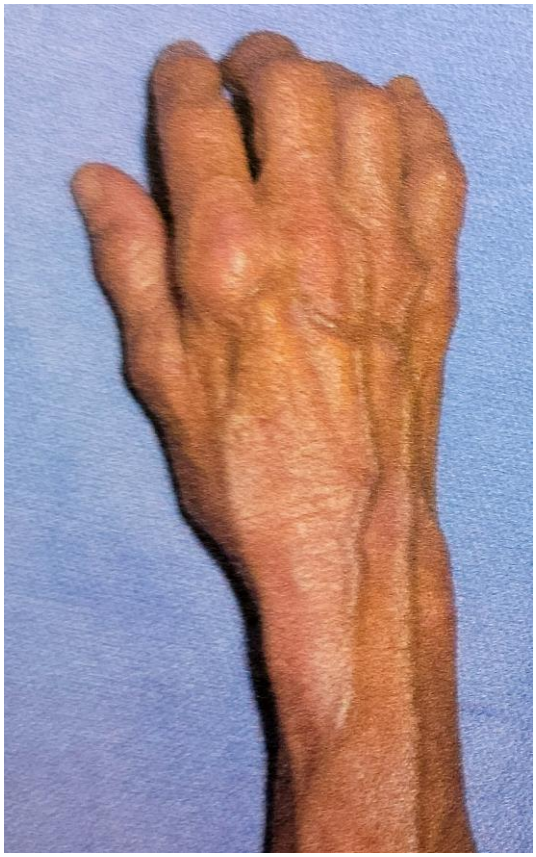
- placing nerve in submuscular plane - ***nerve can glide*** without being "stuck down" (as in intramuscular or subcutaneous locations).

- under *m. pronator teres* and *m. flexor carpi ulnaris*
- anteriorly transposed ulnar nerve is placed over *m. flexor digitorum superficialis* and *m. brachialis*.
- elbow is cast at 45° flexion for 3-4 weeks (avoid pronation exercises).

N. ULNARIS AT WRIST

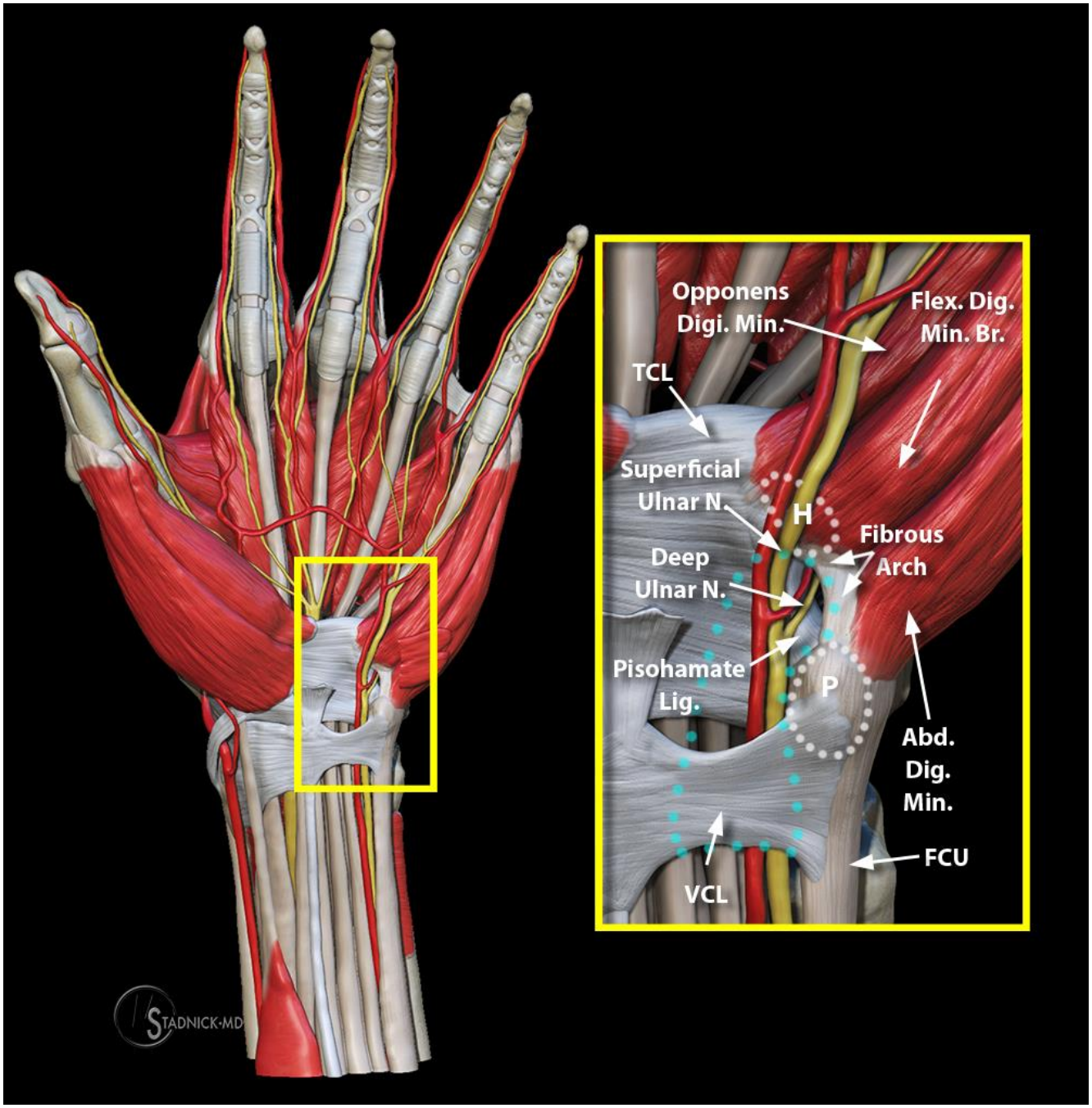
- compression at ulnar **GUYON canal** (only 1% of all ulnar neuropathies)
- ulnar nerve runs above flexor retinaculum (lateral to flexor carpi ulnaris tendon and medial to a. ulnaris).

Wasted 1st dorsal interosseus:

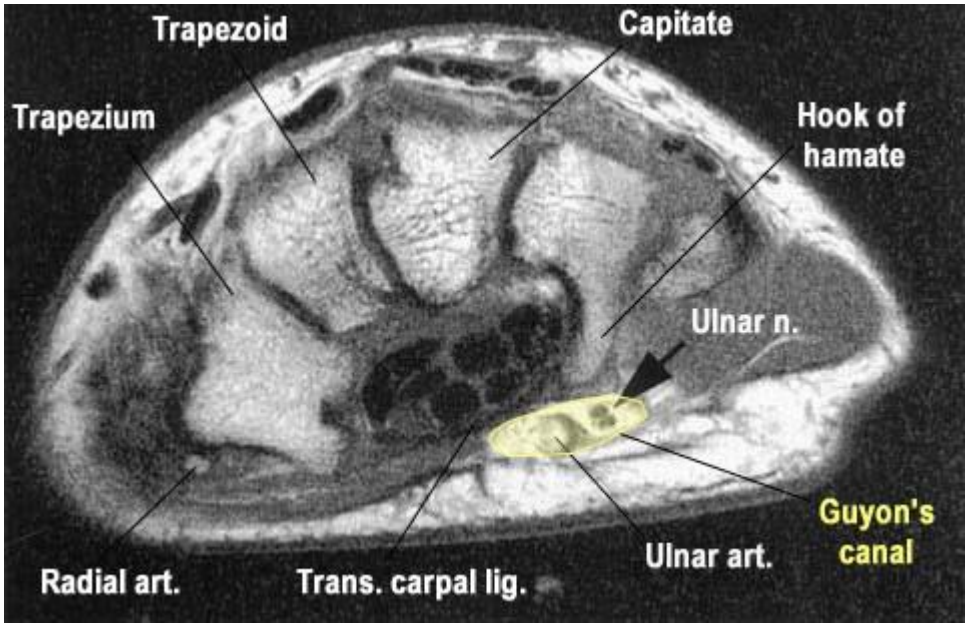


N.B. no sensory loss in dorsal ulnar side of hand!!!!
If no sensory loss at all – either ALS or only deep branch!

GUYON canal – only artery and nerve (nerve is on ulnar side of artery!)

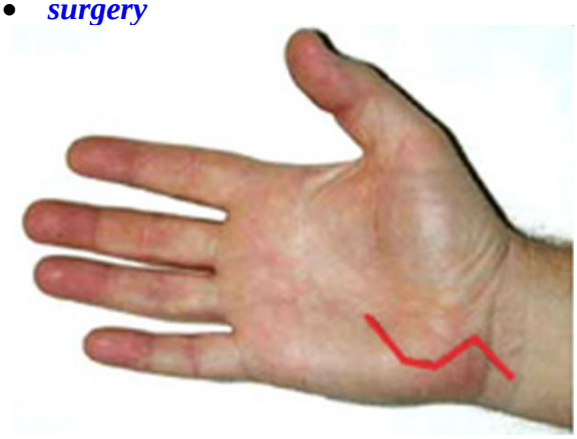


Diagnosis – EDX and MRI:



TREATMENT

- ***avoidance*** & use of ***palmar padding***.
- preop order MRI! (ganglion cyst?)
- ***surgery***

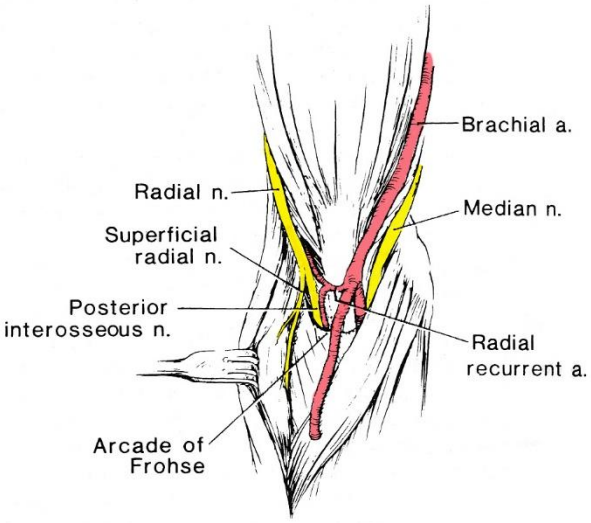


  - fibrotendinous bands over deep branch are cut.
  - nerve is traced straight through, past hamate, and extraneurally decompressed all along.
  - particularly sharp and downward pinching hook may be resected by Kerrison punch.

N. RADIALIS

Places of compression:

1. ***Distal brachial plexus*** - when patient falls asleep with arm draped over chair - nerve is acutely compressed against humerus - SATURDAY NIGHT PALSY.
2. HUMERUS SHAFT FRACTURES (***spiral groove*** between medial and lateral heads of triceps).
3. Underneath **arcade of FRÖHSE** (musculotendinous arcade, formed by upper free border of superficial head of m. supinator) → ***radial tunnel*** - RADIAL TUNNEL (S. POSTERIOR INTEROSSEUS NERVE, PIN) SYNDROME; no sensory loss!



4. ***Wrist*** (sensory superficial radial branch).

leash of *arterial branches (of Henry)* from radial recurrent artery cross over nerve just before arcade of Frohse!

CLINICAL FEATURES

1. **Sensory** – screen **dorsal aspect of skin web between 1st and 2nd fingers; pain** (exacerbated by wrist extension)

2. **Motor**: **WRIST DROP*** with **finger drop at MCP joints** (IP joints extension – action of mm. lumbricales – ulnar and median nerves).

*extensor carpi ulnaris – PIN; extensor carpi radialis – proximal radial nerve

RADIAL TUNNEL (S. POSTERIOR INTEROSSEUS NERVE, PIN) SYNDROME:

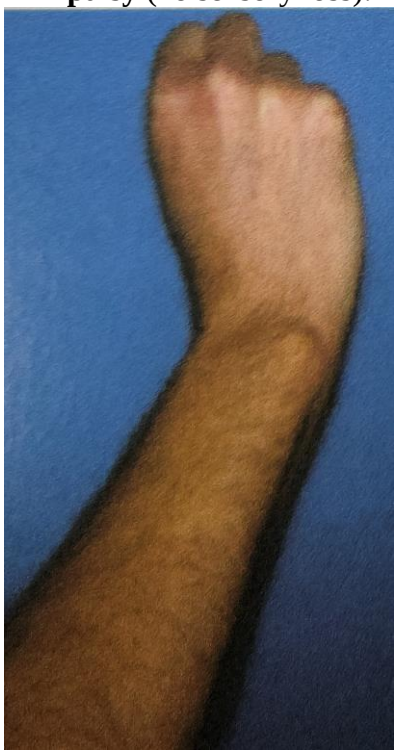
1) mm. extensor digitorum → ↓finger extension at MCP joints

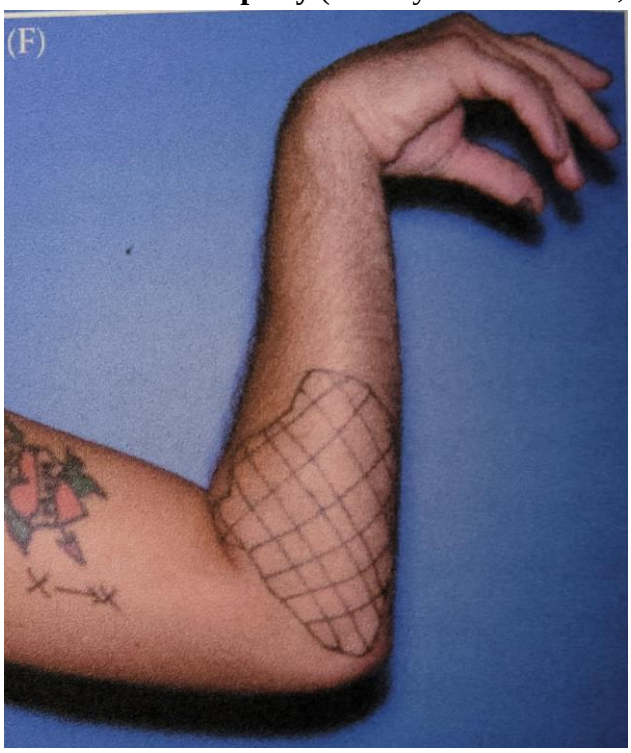
2) m. extensor carpi ulnaris → wrist radial deviation (no wrist drop!) – wrist extension weakness in neutral position (but normal wrist extension in radial deviation – no need for extensor carpi ulnaris!)

PROXIMAL RADIAL NERVE – add wrist drop (at spiral groove), triceps weakness (proximal to spiral groove)

Attempt to extend wrist and fingers:

PIN palsy (no sensory loss): **Proximal Radial palsy** (sensory loss – hatched; x-x – Tinel area):





DIAGNOSIS

1) ***TINEL sign*** at radial tunnel.
2) **nerve conduction studies** - conduction block (locating exact site of compression).
3) **EMG**

DIFFERENTIAL

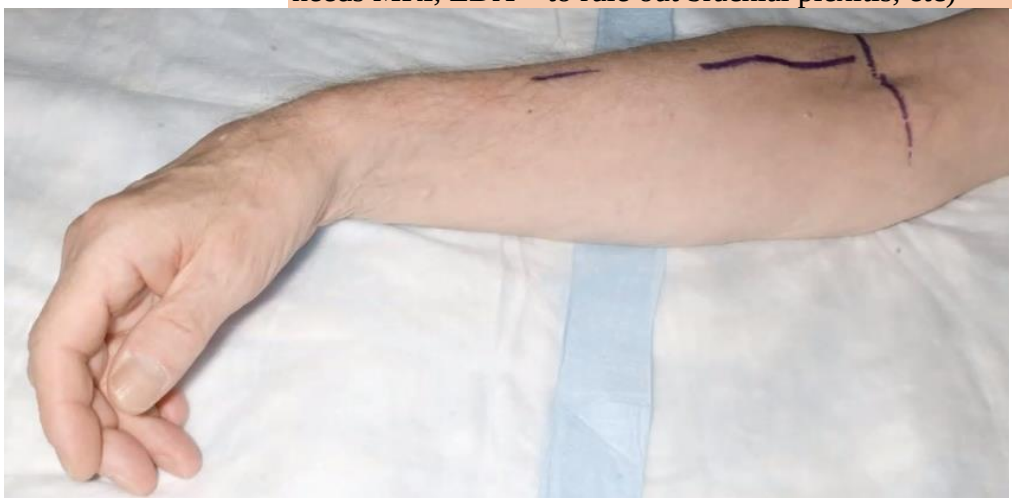
1) lead poisoning - isolated wrist and finger extensor weakness (usually bilateral)
2) C7 radiculopathy: triceps will be weak
   N.B. takeoff of nerve to triceps is proximal to spiral groove!

TREATMENT

- ***spring-loaded brace*** for finger and wrist extension.
- acute radial palsy patients usually recover completely within 4-6 weeks; even after severe injury full late recovery can occur.
- no improvement within 3-4 months following humeral fracture → ***surgical exploration***.

SURGICAL EXPLORATION - for RADIAL TUNNEL (PIN) SYNDROME (excellent outcome in 90-95% cases)

RADIAL TUNNEL SYNDROME is motor neuropathy - diagnosis mandates **surgical decompression**; conservative treatment has no place! (preop needs MRI, EDX – to rule out brachial plexitis, etc)



THORACIC OUTLET SYNDROME (TOS)

- **compression** of BRACHIAL PLEXUS or SUBCLAVIAN VESSELS in their passage from cervical and upper thoracic area toward axilla and proximal arm – between clavicle and 1st rib.

CLASSIFICATION & CAUSES

VASCULAR TOS

- affect subclavian artery or vein → neurological symptoms by ischemia of nerves / muscles.

N.B. brachial plexus is not directly affected!

Neurogenic and vascular TOSs do not coexist!

NEUROGENIC TOS

1. **TRUE (CLASSIC) NEUROGENIC TOS** - caused by structural anomalies: congenital aberrant band between **prominent C7 transverse process** (or rudimentary cervical rib) and **1st rib** (behind tubercle of scalenus ant.)

   Syndrome is very rare!

   - compresses / irritates lower trunk of brachial plexus (C_8-T_1).

2. **SYMPTOMATIC (COMMON, SECONDARY, DISPUTED) NEUROGENIC TOS** - no identifiable anatomical structure causing nerve compression! ("wastebasket" diagnostic group)

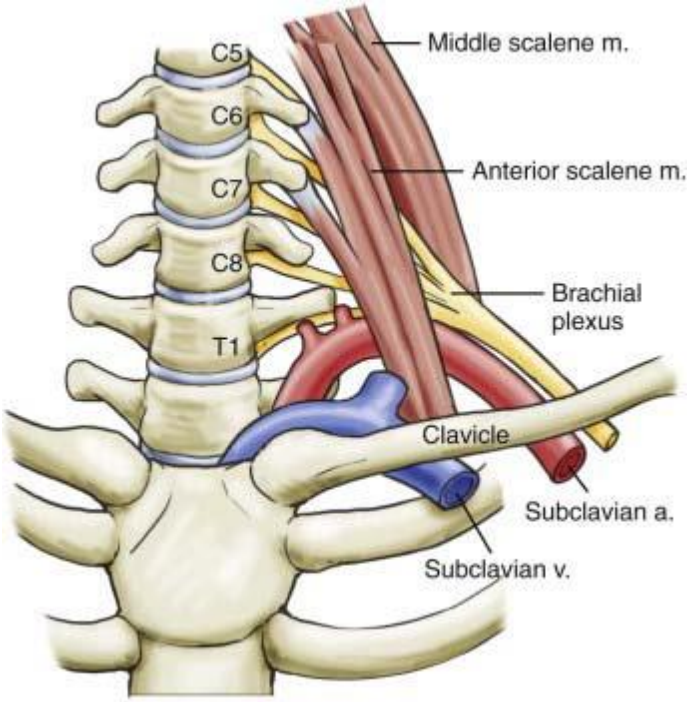
   Precipitating factors:

   1) ***scalenus muscle spasm*** (*scalenus anticus syndrome*).
   2) ***abnormal shoulder posture***:
      a) "*droopy shoulder syndrome*" - tall, slender, and round-shouldered person.
      b) occupational arms *above head*.

Three sites within thoracic outlet where neurovascular compression may occur – going from proximal to distal:

1. **INTERSCALENE TRIANGLE** (anterior scalene muscle anteriorly, middle scalene muscle posteriorly, and medial surface of first rib inferiorly) contains trunks of brachial plexus and subclavian artery (subclavian vein runs anterior to anterior scalene muscle) - *vast majority of neurogenic TOS cases*!!!

2. **COSTOCLAVICULAR SPACE** (middle third of clavicle anteriorly, first rib posteromedially, upper border of scapula posterolaterally) - immediately distal to interscalene triangle.
3. **SUBPECTORAL TUNNEL** (deep to pectoralis minor tendon).



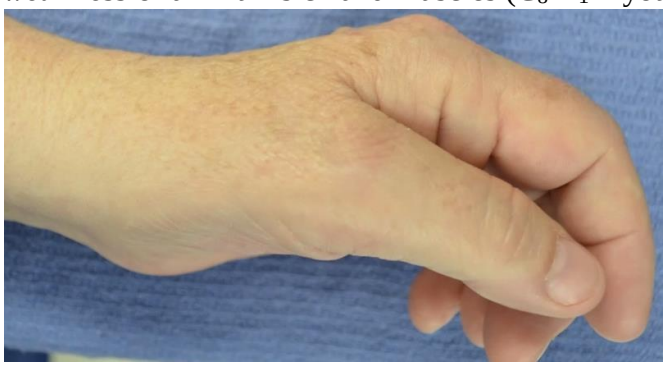
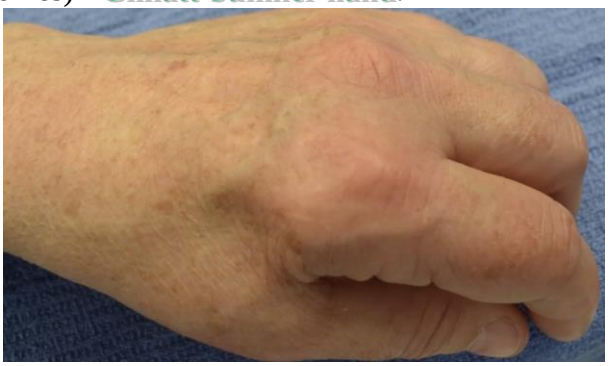
CLINICAL FEATURES

NEUROGENIC TOS

- wide variety of clinical manifestations; two extremes:
    a) **painless form** - neurological and electrodiagnostic findings are quite dramatic.
    b) **chronic pain syndrome** - few, if any neurological and electrophysiologic abnormalities.

TRUE (CLASSIC) NEUROGENIC TOS - stereotyped clinical picture in ***C_8-T_1 distribution***:
N.B. ***motor findings*** include both median and ulnar nerve distributions whereas ***sensory findings*** are confined to ulnar nerve distribution!

1) ***weakness*** of all intrinsic hand muscles (C_8-T_1 myotomes) - **Gilliatt-Sumner hand**:

2) ***numbness, pain, sensory loss*** (lateral neck, shoulder, axilla, parascapular region, *ulnar side* of hand and forearm)
    - pain is aggravated by pulling arm down or repetitive overhead arm use; arm "fatigue" is often prominent.
3) ***vasomotor disturbances*** (changes in skin color and temperature) - in advanced cases related to compression of sympathetic fibers.

Various provocative maneuvers two best tests (best predictive value):
    1. 90-degree shoulder abduction and external rotation

2. Tinel sign over supraclavicular brachial plexus

SYMPTOMATIC (SECONDARY) NEUROGENIC TOS - ***chronic pain / positional numbness*** that may or may not follow dermatomal pattern.

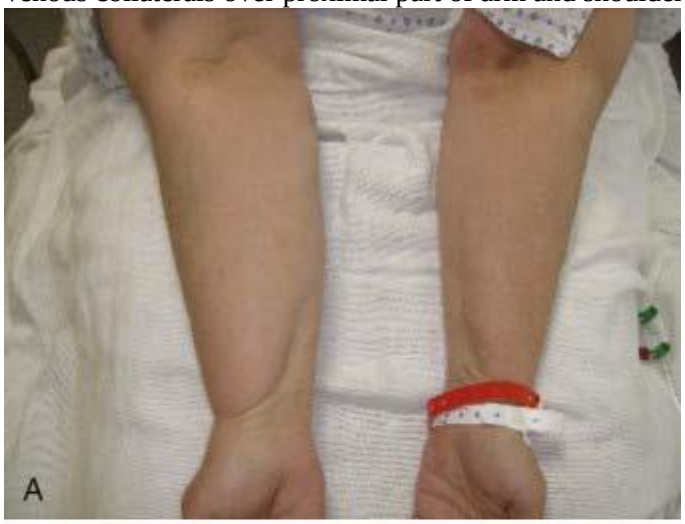
- no neurological deficit! (but due to pain patient may demonstrate give-way type of weakness)
- radial pulse may diminish with arm abduction (it is present in 15% of normals!).

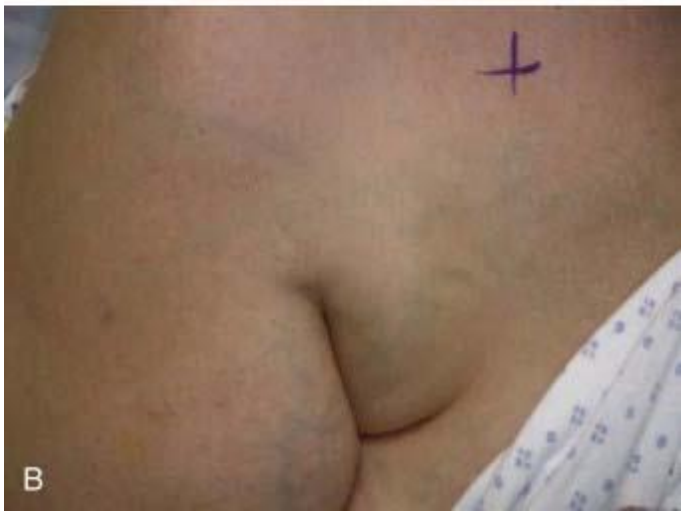
VASCULAR TOS

– ***ischemic symptoms*** in young adults with history of vigorous arm activity:

1) *ischemic muscular pain* - cold, pale, diffusely painful arm that is easily fatigued with activity.
2) *distal pulse*↓ (pulse may even disappear on arm elevation and turning head toward affected side).

Subclavian vein occlusion in venous thoracic outlet syndrome - upper extremity edema (A) and superficial venous collaterals over proximal part of arm and shoulder (B):





DIAGNOSIS

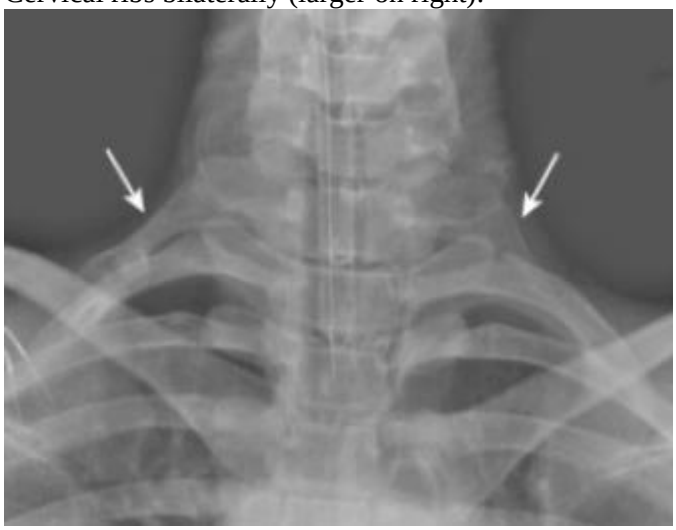
NEUROGENIC TOS

In **TRUE (CLASSIC) NEUROGENIC TOS** injury is ***axonal***:

1) **nerve conduction studies**
2) **EMG** findings in C8-T1 myotomes (i.e. beyond ulnar distribution)

- **MRI** (cervical spine, brachial plexus) + **MR neurography** - compression site and cause.
- **chest XR** – cervical rib + to rule out Pancoast tumor.

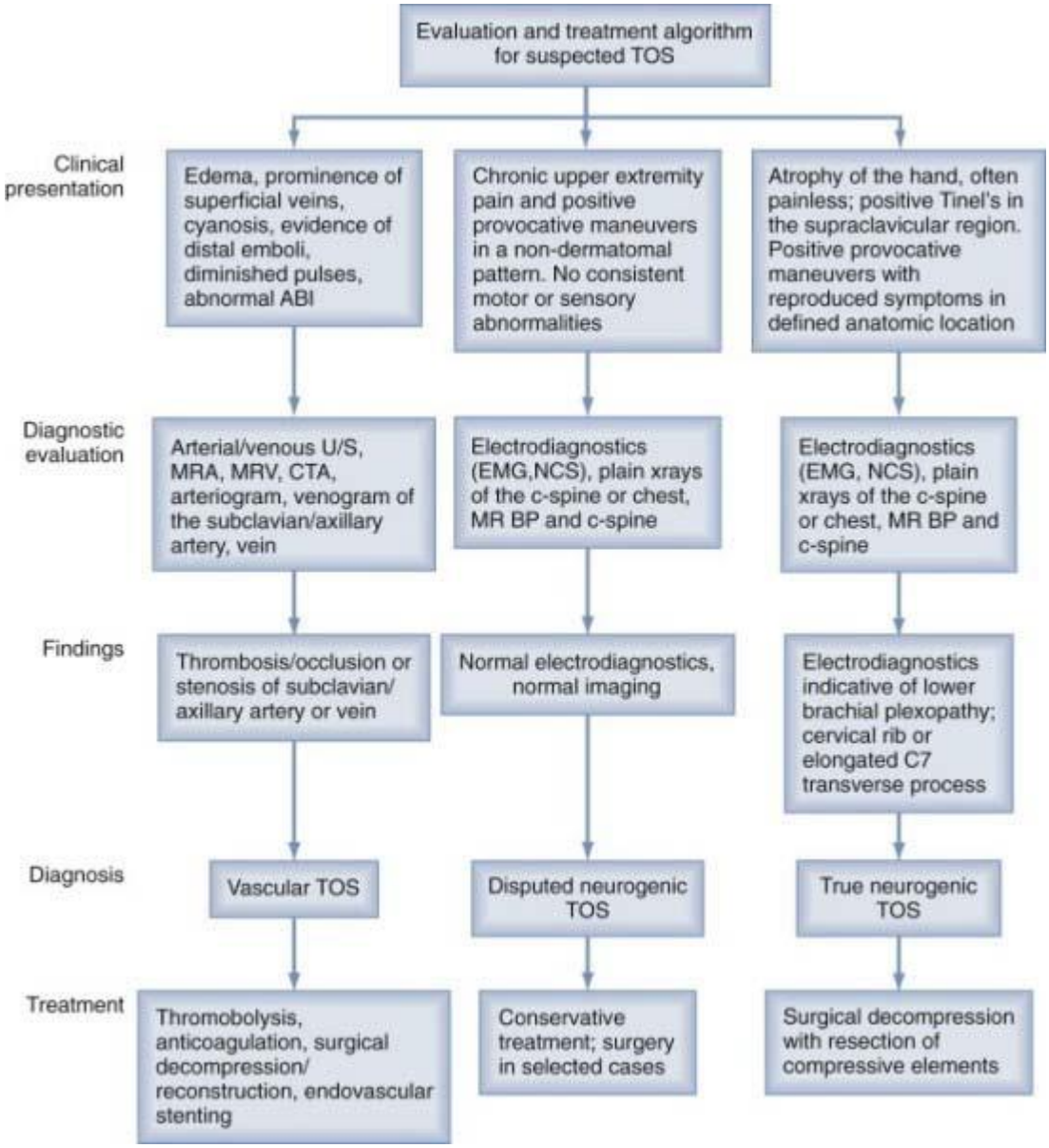
Cervical ribs bilaterally (larger on right):



SYMPTOMATIC (SECONDARY) NEUROGENIC TOS - electrophysiologic studies are normal.

VASCULAR TOS

- usually easy to detect on **clinical examination** or **vascular imaging** (US, MRA – with/without arm elevation)



TREATMENT

NEUROGENIC TOS

Most patients deserve trial of (and only need) conservative therapy:
Lifestyle modification - avoidance of overhead activities, carrying of heavy bags over shoulder, sleeping in positions with arms overhead.
Physical therapy directed at strength of shoulder girdle **(PEET's exercises)**.

SYMPTOMATIC (SECONDARY) NEUROGENIC TOS – maximal PT for at least 3-6 months is mainstay (no risk involved - syndrome does not transform into or progress to true neurogenic TOS)

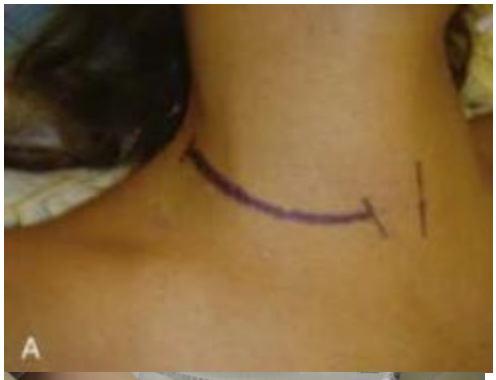
- ***scalene muscle denervation*** (injection of botulinum toxin)
- ***surgery*** is often offered only as a last resort (patients who respond to ***scalene muscle blocks*** are more likely to respond to surgery) - significant chance that the patient will not improve!!!

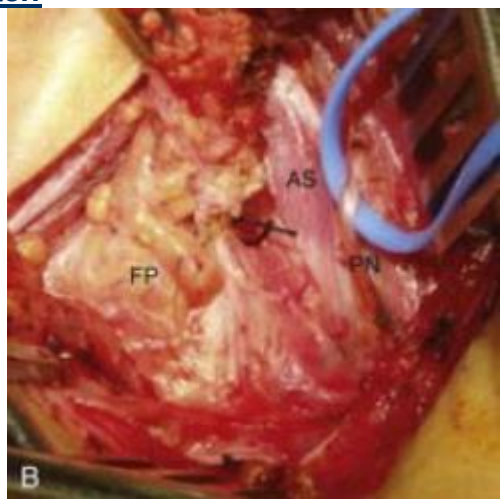
TRUE (CLASSIC) NEUROGENIC TOS: PT + scalene Botox → surgical release (transection of aberrant bundle, removal of cervical rib / 1st rib, scalenotomy at insertion):

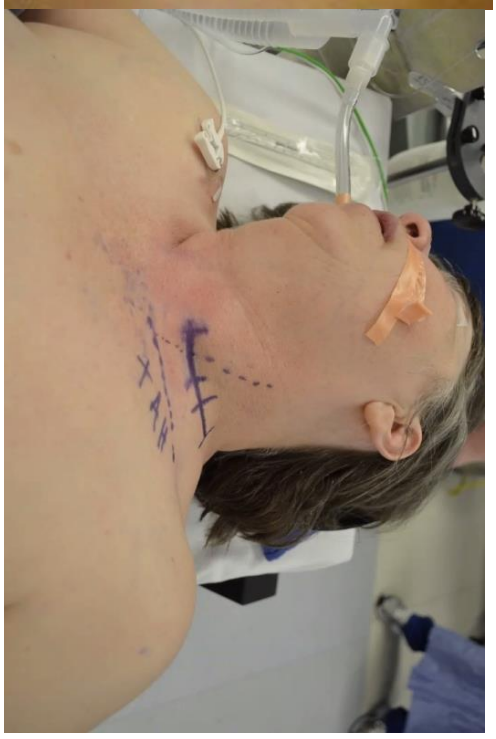
a) *anterior **supraclavicular** approach* - favored by most neurosurgeons!

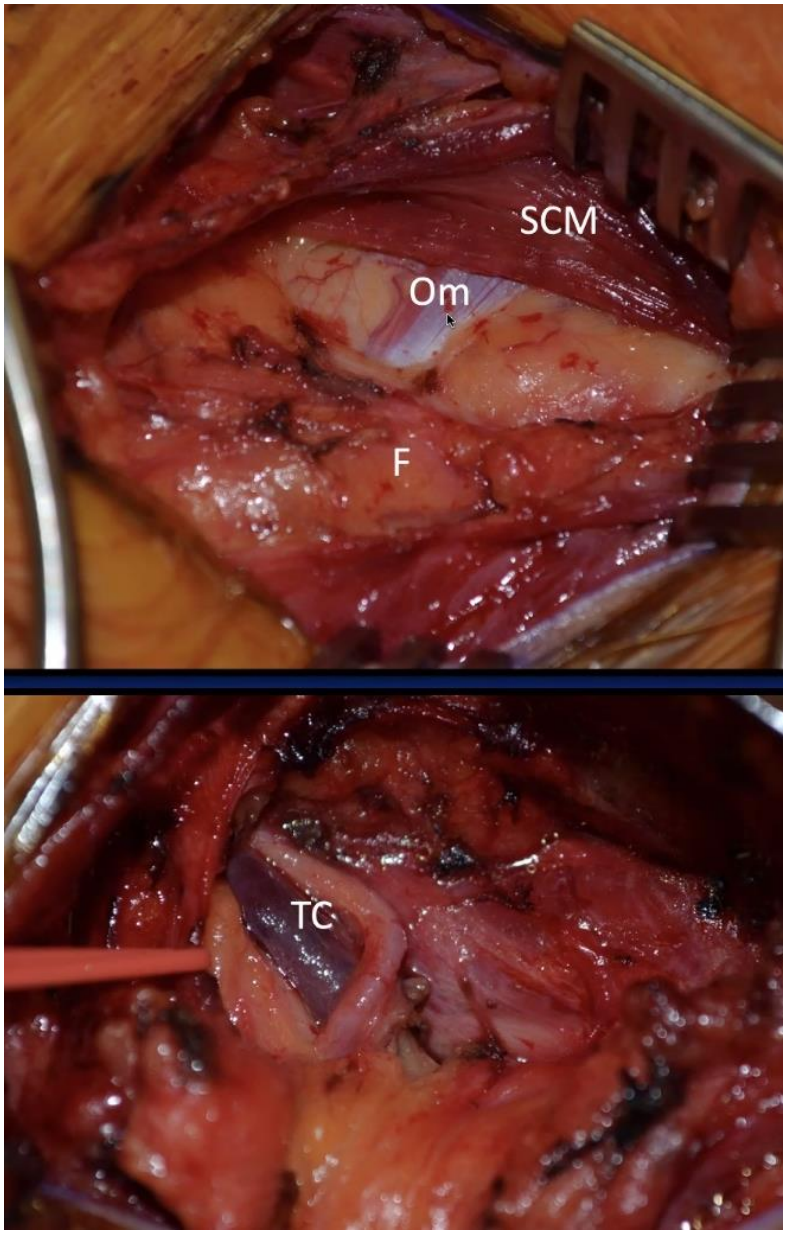
b) *transaxillary approach* (with first rib removal) - has many complications (neurovascular injures).
c) *posterior subscapular approach*

Anterior Supraclavicular Approach



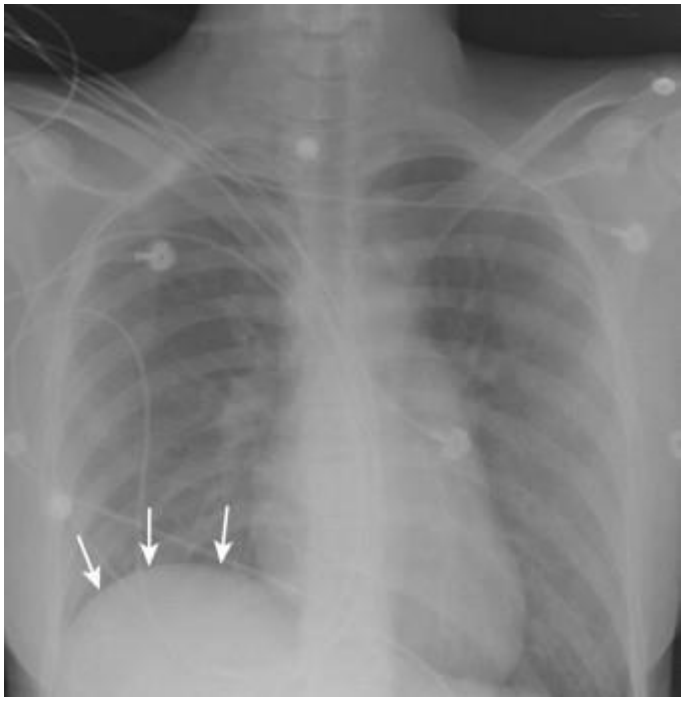






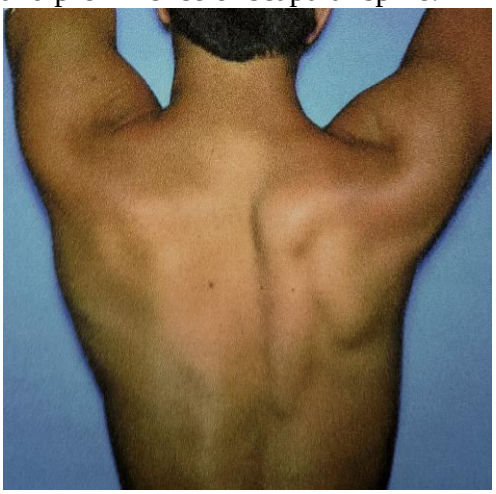
- **phrenic nerve** has a unique course; *it runs superolaterally to inferomedially on the anterior surface of the anterior scalene muscle*, beneath its investing fascia; identity of the phrenic nerve is confirmed by stimulating it and feeling contraction of the ipsilateral hemidiaphragm.
- anterior scalene is transected while carefully protecting phrenic nerve.
- upper, middle, and lower trunks of the brachial plexus are identified.
- subclavian artery is found by palpation and visual inspection running inferiorly in the plane of the brachial plexus and is controlled with a vessel loop.
- neural elements are inspected in circumferential fashion, and any compressive bands or anomalous structures are resected.

Phrenic nerve injury (takes long to regenerate! – dedicated respiratory PT → diaphragm plication):



N. SUPRASCAPULARIS

- motor nerve (C_{5-6}) → weakness of:
    1) **m. supraspinatus** (initiation of shoulder abduction); atrophy is not obvious due to overlying m. trapezius.
    2) **m. infraspinatus** (only muscle for external rotation of humerus) → hollowing of infraspinous fossa and prominence of scapular spine:



Etiology – athletes – compression at *suprascapular notch of scapula* (stout, strong suprascapular ligament).

Clinical Features

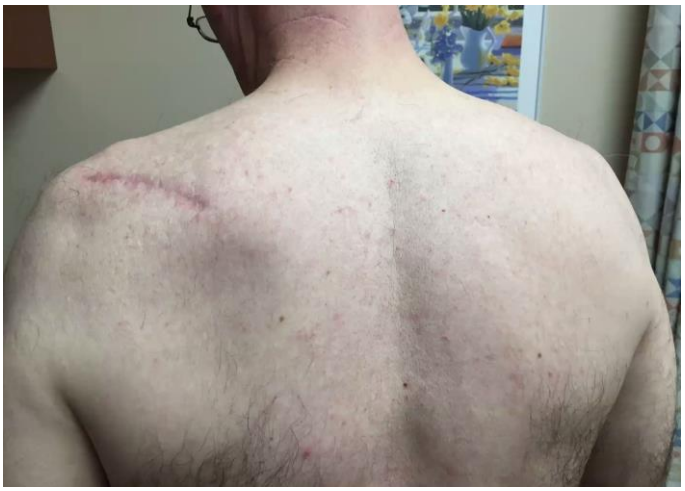
- posterior aspect of shoulder → dull aching ***pain***.
- deep pressure over midpoint of superior scapular border may produce discomfort.

Best diagnosis - **EMG** evidence of denervation of supraspinatus and infraspinatus muscles.
MRI may show ganglion cyst.

TREATMENT

a) **CONSERVATIVE MANAGEMENT**: cessation of athletic activities, conditioning exercises of upper girdle, periodic injection of nerve (bupivacaine and dexamethasone).

b) *failure of pain control / severe weakness* → **SURGICAL DECOMPRESSION** (some patients never regain full strength due to atrophy - early detection is most important predictor of outcome!):
   - **incision** - 2 cm above and parallel to scapular spine.
   - horizontal trapezial fibers are atraumatically split to expose constant fat pad separating trapezius from supraspinatus muscle.
   - digital palpation along superior scapular border detects abrupt change into rubbery springiness of suprascapular ligament.
   - suprascapular artery, which crosses above ligament, is swept aside.
   - ligament is cut and bony notches enlarged with rongeur, if necessary.



ILIOHYPOGASTRIC NERVE

- may cause lower abdominal musculature weakness with bulging ("pseudohernia syndrome")

GENITOFEMORAL NERVE

- may be injured during psoas muscle retraction during LLIF surgery – nerve exits at medial edge of psoas (other nerves – at lateral edge) → burning pain in genitalia

OBTURATOR NERVE

- may be compressed by pelvic tumors, fetal head or forceps.
- sensation to inner thigh, and motor to thigh adductors.

FEMORAL NERVE

- exits into thigh 1 cm lateraliau a. femoralis just below lig. inguinale.

ETIOLOGY

1. **Diabetes** - most frequent cause! (e.g. plexopathy)
2. Entrapment (rare) - secondary to hernia or its repair (deep sutures placed during herniorrhaphy), prolonged pelvic surgery (retractor compression)
3. DSA with femoral arterial catheterization

4. Intraabdominal tumor, retroperitoneal hematoma
5. Pelvic fracture

CLINICAL FEATURES

- motor deficits - quadriceps femoris (knee extension)
  N.B. ***weakness of iliopsoas (hip flexion) indicates very proximal pathology*** (lumbar root or plexus lesion) as branches to iliopsoas arise just distal to neural foramina!
- patellar (knee jerk) reflex↓
- sensory loss and pain over anterior thigh and medial calf (saphenous nerve)
- positive femoral stretch test.

MERALGIA PARESTHETICA

(Greek: *meros* – thigh, *algos* – pain)
- entrapment of purely sensory *lateral femoral cutaneous nerve* (L_{2-3}) where it passes ***beneath inguinal ligament*** at its attachment to the anterior superior iliac spine.

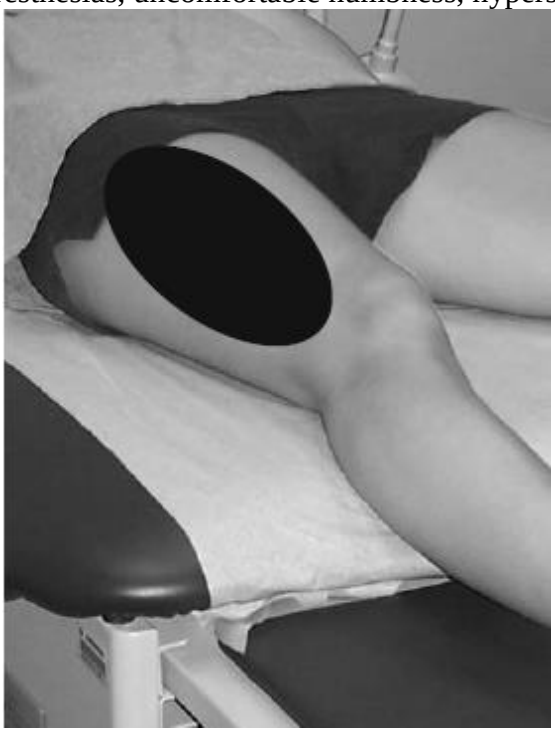
ETIOLOGY

protruding, pendulous abdomen (pregnancy, obesity, ascites), tight belt or corset, excessive walking or marathon running*; also may be initial manifestation of diabetic neuropathy; may also occur post-op in slender patients positioned prone.

*nerve angulation is exaggerated with thigh extension.

CLINICAL

- burning paresthesias, uncomfortable numbness, hypersensitivity, hyperpathia:



- patient learns to relieve symptoms by:
  - placing pillow behind thighs;
  - sitting or lying prone helps;
  - assuming slightly hunched posture while standing.
  - spontaneous rubbing the area in order to obtain relief is very characteristic!

- deep digital pressure 1 cm medial to ASIS may set off shooting paresthesia.

DIAGNOSIS

- diagnosis is confirmed with **nerve block** - 0.5% BUPIVACAINE injected finger's breadth medial to ASIS → anesthesia + complete cessation of pain and tingling (may be long lasting).
- imaging (18 MHz US, MRI) – only for select cases.

DIFFERENTIAL

1. Femoral neuropathy: sensory changes more anteromedial, extend to anteromedial lower leg (saphenous nerve!)
2. L2 or L3 radiculopathy: motor weakness (thigh flexion or knee extension)
3. Nerve compression by abdominal or pelvic tumor (concomitant GI or GU symptoms)

TREATMENT

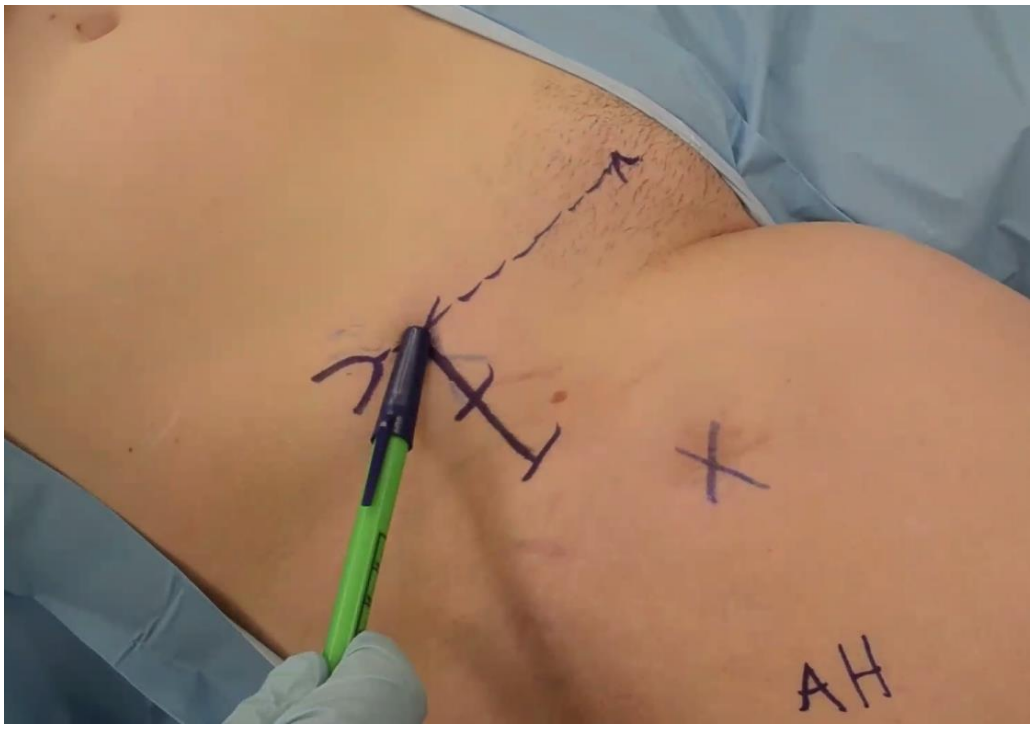
1) weight loss, avoidance of all constrictive garments, and postural modification (avoiding hip extension).
2) serial injections of **local anesthesia** and **steroid**.
3) **local measures** – ice applications, capsaicin ointment, lidocaine patches.

N.B. anatomic variation is common – LFCN may actually pass through the ligament, and as many as four branches may be found.

Surgical decompression

- **incision** - along medial border of sartorius, 2 cm below ASIS; extends 6-7 cm.
- nerve is located at medial muscle border or just behind it.

  Use US to find nerve!



- nerve is *traced proximally* - toward exit site just medial to ASIS.
- bands of inguinal ligament over nerve are divided (hernia is extremely rare after this procedure!).
- nerve is followed into pelvis for 2-3 cm to ensure clearance of other iliacus fascial bands.

N.B. operation is exploratory in nature - generous exposure is required; if nerve can't be located, it is usually because exposure is too superficial; if nerve still cannot be found → small abdominal muscle incision and nerve located in the retroperitoneal area.

15-20% cases recur → **nerve transection (neurectomy)** – more effective than neurolysis:

- after freeing nerve at ASIS and proximally toward pelvis, ligature is tied tightly around nerve.
- nerve is ***firmly tugged downward*** → cut is made just proximal to tie.
- upper cut end of nerve springs back and disappears into pelvic cavity - this prevents painful neuroma formation on surface of thigh.
- pain is gone, and patient usually adjusts well to numbness (but risk of denervation pain).

CAUTION: cases have occurred where the femoral nerve has erroneously been divided! – always stimulate before cutting!

See Case PN5 >>

SCIATIC NERVE (N. ISCHIADICUS)

There is no consistent area in lower extremity where entrapment occurs!

1) ***retroperitoneal bleeding***
2) course of sciatic nerve between parts of ***piriformis muscle*** **(PIRIFORMIS SYNDROME)**
3) ***myofascial band*** in distal portion of thigh (between biceps femoris and adductor magnus)
4) ***trauma*** (fractures of hip, surgical trauma from hip replacement).

Piriformis syndrome

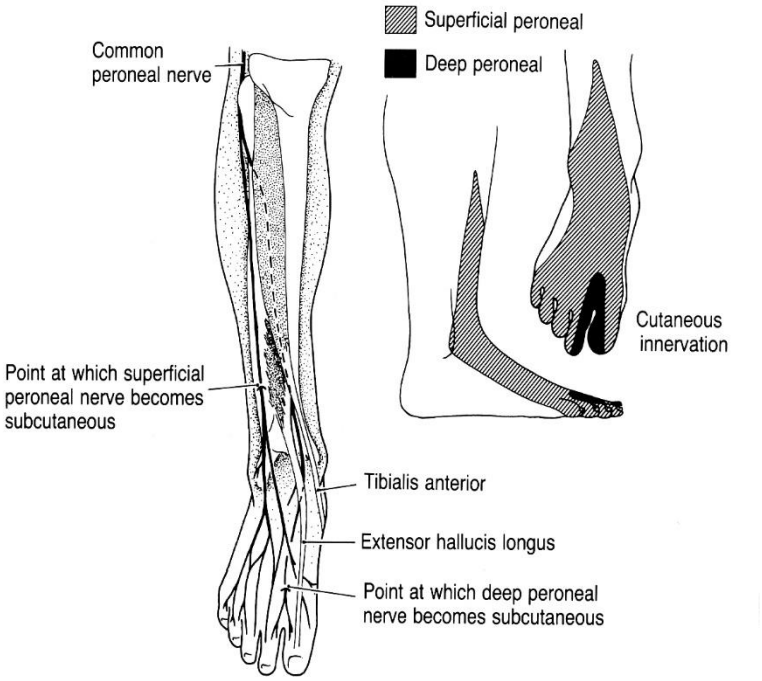
- main symptom - pain aggravation by sitting on hard surface.
- diagnostic provocative maneuvers – FAIR test
- treatment:
    1) stop aggravating activity
    2) stretching exercises
    3) corticosteroid injection (decreases fat amount around m. piriformis – more room for nerve).

N. PERONEUS

GSW in thigh almost as a rule injures peroneal but spares tibial divisions of sciatic nerve!

Just distal to fibular head, CPN divides into:

1. **Deep peroneal nerve** (AKA anterior tibial nerve) - primarily motor:
    - motor: foot and toe extension (anterior tibialis, extensor hallucis longus, extensor digitorum longus).
    - sensory: very small area between great toe and second toe.
2. **Superficial peroneal nerve**:
    - motor: foot eversion (peroneus longus and brevis).
    - sensory: lateral distal leg and dorsum of foot.



Mechanism:
1) damage at fibular head (fractures, bandages, stockings, crossing knees while sitting).
2) forcible foot inversion (nerve stretching).

Etiology:
1) thin individuals who *habitually cross* their legs
2) patients who *lose significant amount of weight* (slimmer's palsy)
3) certain professions that require *frequent sitting, squatting, or kneeling* (e.g. roofers, carpet layers, strawberry pickers).
4) *prolonged squatting* during childbirth
5) asleep while intoxicated
6) *iatrogenic injury* - improper cushioning or positioning of leg under anesthetic (esp. in dorsal lithotomy or lateral decubitus positions), improperly applied casts
7) any *contact sport*.
8) ganglion cysts

Clinically – ***foot drop*** (analogous to wrist drop with n. radialis damage; patients compensate for footdrop by lifting leg higher – **steppage gait** with exaggerated thigh & knee flexion) ± ***pain*** laterally in leg and foot.
- ask to heel-walk.
- Tinel sign is frequently present at site of compression.
- coexistent *foot inversion* weakness may suggest either L5 radiculopathy or sciatic nerve injury.
- *biceps femoris* weakness - CPN injury above knee.
- chronic foot drop may produce Achilles tendon contracture (**talipes equinus**).

Diagnosis
1. **Electrophysiologic evaluation** (after > 3 weeks of symptoms)
   - EMG - on both peroneal-innervated muscles and non–peroneal, L5-innervated muscles.
     N.B. short head of biceps femoris is the only peroneal-innervated muscle proximal to peroneal tunnel!
2. **Imaging** - plain films, MRI, ultrasound.

DIFFERENTIAL OF FOOT DROP

- ***deep peroneal nerve*** → weak **anterior tibialis** (L4 > L5), **EHL & extensor digitorum longus** (L5)
  N.B. foot drop is L5 > L4

1) **L4/L5 radiculopathy** – also affects posterior tibialis (foot inversion) and gluteus medius (internal rotation of flexed hip); pain!
2) **sciatic nerve** palsy (hip fracture-dislocation, IM injection) - **flail foot** (paralysis of dorsiflexors + plantarflexors)
3) **common peroneal nerve** palsy
4) **Charcot-Marie-Tooth**
5) **heavy metal** poisoning (esp. *lead*)
6) **diabetic** neuropathy
7) **Hansen's disease** (leprosy)
8) lesion anywhere along **pyramidal tract**, **motor neuron disease** - **spastic foot drop** (Babinski sign, hyperactive Achilles reflex).
9) anterior compartment syndrome, severe ankle inversion sprains
10) muscular dystrophy
11) popliteal fossa cysts (Baker cyst)
12) anterior tibial artery aneurysm

Foot drop → get **MRI** to rule out mass effect – either **L-spine** or **peroneal** ← decide clinically:
N.B. painless foot drop is unlikely to be due to radiculopathy!
N.B. L5 affects both foot inversion and eversion (deep peroneal nerve – only partial inversion; superficial peroneal nerve – only eversion)

TREATMENT

CONSERVATIVE THERAPY

- effective for most cases of CPN entrapment:

1. **PT** to prevent Achilles contractures (heel cord), which would impair ankle dorsiflexion if nerve function returns.
2. **Ankle-foot orthosis (AFO)**.

SURGERY

Peroneal Nerve Decompression – for patients who show little or no improvement after 3 months.
N.B. operate for foot drop early (maximum wait – 3 months)

- **skin incision** just medial to the tendon of the short head of the biceps femoris (lateral hamstring) as the peroneal nerve is best located deep to or slightly medial to this tendon → incision is carried distally slightly laterally along the surgical neck of fibula; biceps femoris is retracted laterally and the nerve is isolated and tagged with a Penrose drain:

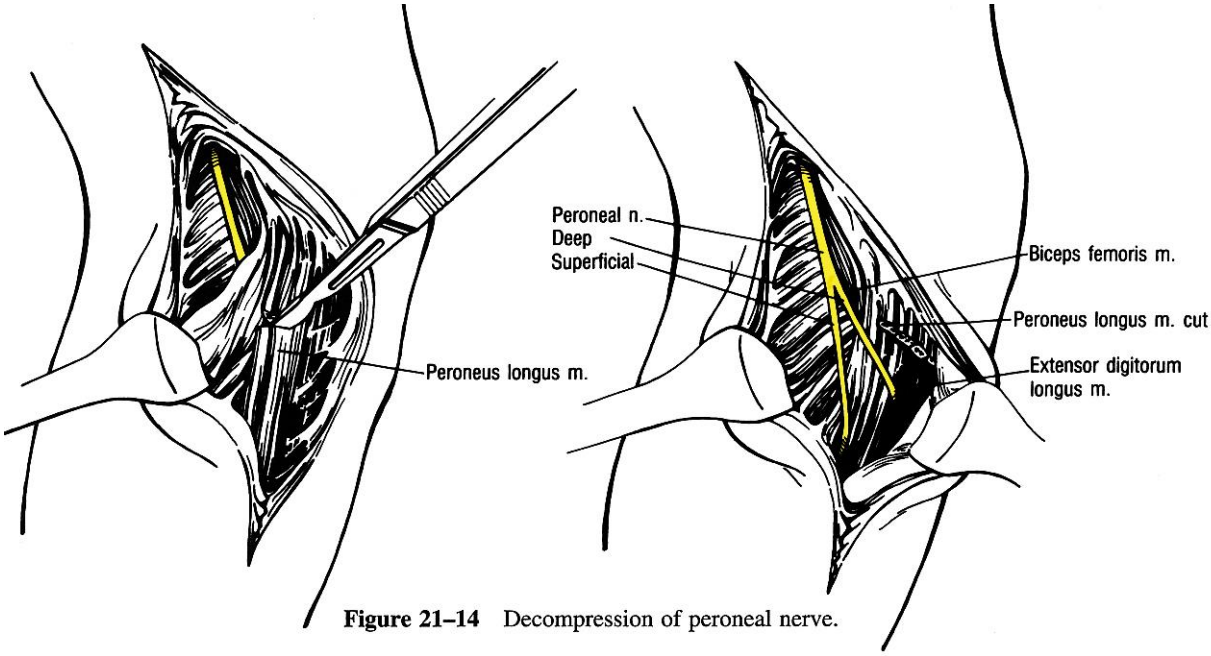


Figure 21–14 Decompression of peroneal nerve.

N.B. peroneal injuries above knee usually do not regenerate enough!

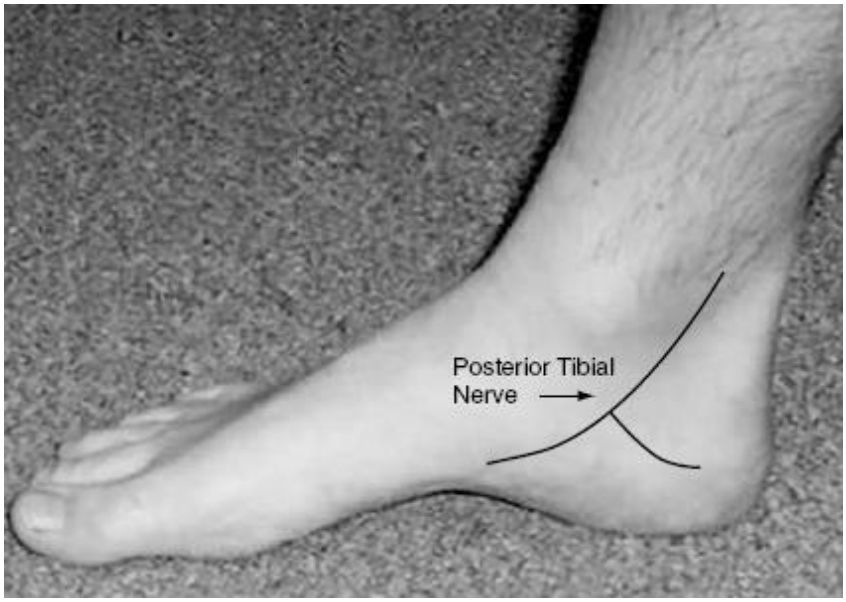
Persistent footdrop after surgery → **TP tendon transfer** - highly effective for footdrop caused by CPN injury, particularly in men < 30 years.

N. TIBIALIS POSTERIOR / TARSAL TUNNEL SYNDROME

- *posterior tibial nerve* entrapment posterior-inferior to medial malleolus at flexor retinaculum or more distally.

Tarsal tunnel (TT) anatomy

- TT is covered by **flexor retinaculum (laciniate ligament)** which extends downward from the medial malleolus to the tubercle of the calcaneus.



- clinical features:

1) burning, unpleasant poorly localized pain and paresthesias in medial heel* + sole (down to first, second, and third toes)
   *__*calcaneal branch* (sensation to heel)__ often is spared because of its proximal takeoff.
   – pain *reminds plantar fasciitis*, but positive **Tinel sign** is present.
2) intrinsic toe flexors are weak - **toe clawing**.
3) provocative testing: foot dorsiflexion-eversion – examiner maximally everts and dorsiflexes the ankle while dorsiflexing the toes at the MTP joints for 5–10 seconds - positive test reproduces pain.

Diagnosis
1. **EDX**
2. **Imaging** - plain films, MRI, ultrasound.

TREATMENT

Period of **conservative therapy**.
- **lifestyle modification** (weight loss and avoidance of ill-fitting shoes or high heels).
- trial of **immobilization**
- **orthotics** (medial arch support)
- **corticosteroid** injections, **nerve blocks**
- antiepileptic, antidepressant, and narcotic pain medications.

Surgical Decompression
- **incision** begins 2 cm proximal to medial malleolus to pick up neurovascular bundle above flexor retinaculum.
- nerve is followed distally with **release of retinacular fibers**.

MORTON'S NEUROMA

Benign perineurium thickening (fibrosis, not true neuroma!) of *3rd interdigital nerve* due to pinching between heads of *3rd and 4th metatarsals*.
- causes:
  1) tight shoes (compress toes)
  2) loss of fat-pad of ball
- clinical features:
  – pain (metatarsalgia), tenderness, paresthesias along nerve (sometimes patient takes off shoe to decrease pain)
  – decreased sensation in web space.
- diagnosis: tenderness between 3rd and 4th metatarsal heads; compressing metatarsal heads between examiner's thumb and fifth digit will accentuate pain:

- treatment:
    1) comfortable shoes, orthotics (metatarsal pad).
    2) lidocaine + corticosteroid infiltration - given dorsally (top of foot) so that it is less painful.
    3) surgical excision

PERIPHERAL NERVE STIMULATORS

Pain > 3 mos duration: Occipital neuralgia, PNS injury / post-surgical pain, treatment-resistant migraines
Neuropsych clearance + trial > 50% pain relief
Percutaneous or open implantation

NERVE BIOPSY

a) relatively expendable **SENSORY** nerves (e.g. ***sural*** behind lateral malleolus, ***superficial radial*** at wrist, ***greater auricular***)
b) portions of peripheral **MOTOR** nerve twigs (as part of muscle biopsy).
    – choose superfluous or accessory muscle (such as ***gracilis*** muscle).

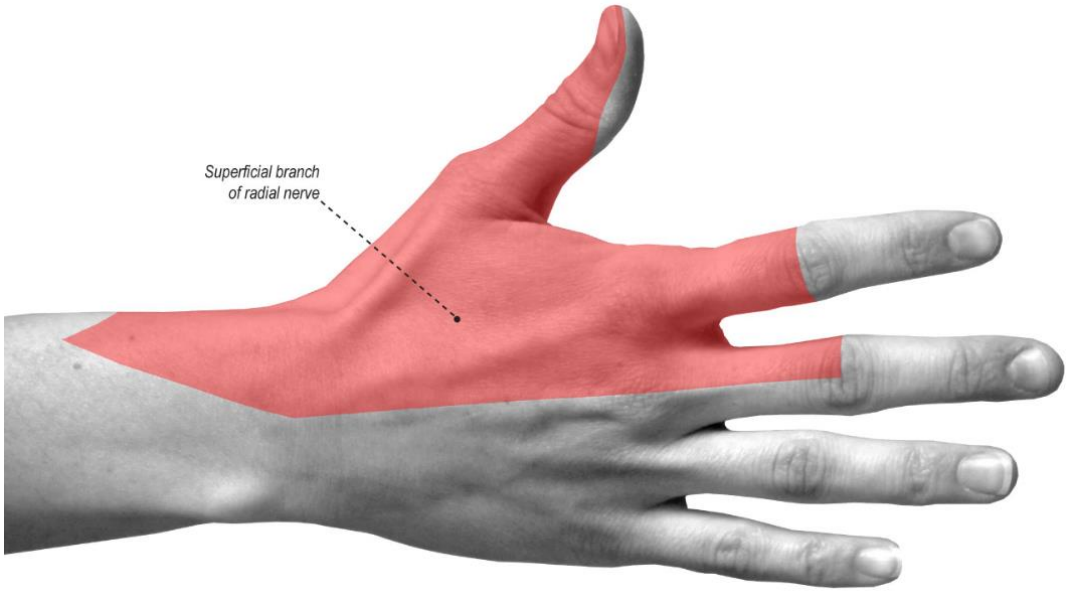
Divide nerve sharply beyond incision margins (to minimize subsequent painful neuroma formation)

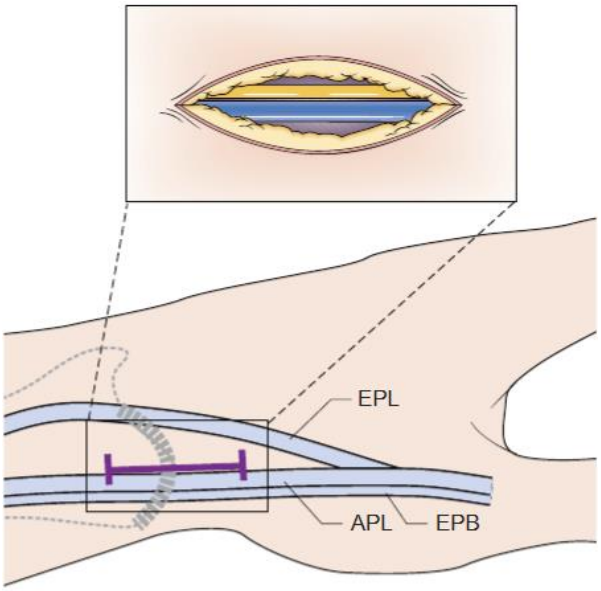
Sural nerve (most commonly sampled nerve):
- pure sensory nerve that supplies *small area* of skin.
- often involved in clinically predominantly *motor neuropathies*!
- nerve *regeneration* occurs in > 90% cases.
- check if **sensory nerve action potential (SNAP)** is recordable for the nerve that ultimately needs biopsy – perform nerve biopsy with a preserved SNAP, although reduced in amplitude; if SNAP is not recordable, less chances of postop neuropathic pain.
- sural nerve is deep to lesser saphenous vein

Complications of sural nerve biopsy:

1) annoying **causalgia** (5%), esp. when bending forward (stretching of nerve).
2) all patients should anticipate permanent **loss of discriminative sensation** in lateral border of foot (extending to 5th toe, heel, and lateral malleolus).
3) poor healing - ankle is a notorious region for poor circulation

Superficial Radial Nerve (next to cephalic veins) – over distal radius where enters anatomical snuff box





BLOCKADES OF PERIPHERAL NERVES

- rezorbcijai sulėtinti, kraujavimui sumažinti pridedama EPINEPHRINE;

  EPINEPHRINE draudžiama naudoti:

  1) **rankose** (ypač nn. digitales - galima pirštų nekrozė)
  2) **kojose** (ypač nn. digitales)
  3) **varpoje**

1. **Toxic reactions**:

1) ***seizures*** H: airway, O_2, diazepam / thiopental i/v.
2) ***cardiorespiratory collapse*** H: i/v fluids, vasopressors.

- priežastys: a) *masyvi rezorbcija* (profilaktika - EPINEPHRINE).
  b) *perdozavimas* (naudok mažesnių koncentracijų anestetiko tirpalus).
  c) *intravaskulinė injekcija* (keep needle in motion, inject only after negative aspiration).

2. True **anaphylaxis** is rare - dažnesnė su ester-type anestetikais (e.g. PROCAINE), bet reta su amide-type anestetikais (e.g. LIDOCAINE).
3. **Residual neurologic deficit** (gali likti iki kelių mėnesių ar net permanent) - profilaktika: **neinjekuok tiesiai į nervą** (kai ligonis pajunta paresteziją, adatą šiek tiek atitrauk ir tada suleisk anestetiką).

CRANIAL NERVES

Cranial nerve	Syndrome	Typical offending vessel
V	trigeminal neuralgia	superior cerebellar artery (SCA)
VII (facial nerve proper)	hemifacial spasm (p. 1651)	anterior inferior cerebellar artery (AICA)
VII (nervus intermedius)	geniculate neuralgia (p. 1653)	
VIII	disabling positional vertigo (p. 1654)	
IX	glossopharyngeal neuralgia (p. 1654)	posterior inferior cerebellar artery (PICA)
X	superior laryngeal neuralgia (p. 1654)	PICA or VA
XI	torticollis of XI nerve origin (p. 1630)	VA or (rarely) PICA

Extraparenchymal

Syndrome	**Cranial nerves**											
	I	II	III	IV	V	VI	VII	VIII	IX	X	XI	XII
Foix (superior orbital fissure)			+	+	V_1	+						
Tolosa-Hunt (lateral wall of cavernous sinus)			+	+	V_1	+						
Gradenigo (apex of petrous bone)					+	+	±	±				
internal auditory meatus							+	+				
pontocerebellar angle					+		+	+	±	±		
Vernet (jugular foramen)									+	+	+	
Collet-Sicard (retropharyngeal, posterior laterocondylar space)									+	+	+	+

Syndrome	Cranial nerves												Associa
	I	II	III	IV	V	VI	VII	VIII	IX	X	XI	XII	
INTRAPARENCHYMAL													
Weber (ventral midbrain syndrome)			+				+ [3]						*cerebral p*
Claude			+										*red nucle rubro-th*
Benedikt			+										*red cerebral p*
Nothnagel			+										ipsilateral c dizziness, rolling nys
central midbrain syndrome			+										*red nucleu medial*
Foville						+							C
Millard-Gubler							+						C
medial medullary syndrome												+	CHP, *med*
medial pontine syndrome						+							CHP, *med MLF* (i ophthal *cerebella*
Wallenberg (lateral medullary syndrome)					+ [2]		+ [1]		+	+			lateral me stru *tr. spino tr. retic* (sympath *vestibular inf. cerebe*
Marie-Foix (lateral inferior pontine syndrome)							+	+					
lateral superior pontine syndrome													
pseudobulbar paralysis								+	+	+		+	
bulbar paralysis									+	+		+	

INTRAPARENCHYMAL lesions - ***crossed*** sensory or motor paralysis (cranial nerve signs on one side of body and tract signs on opposite side).

IDIOPATHIC MULTIPLE CRANIAL NERVE INVOLVEMENT

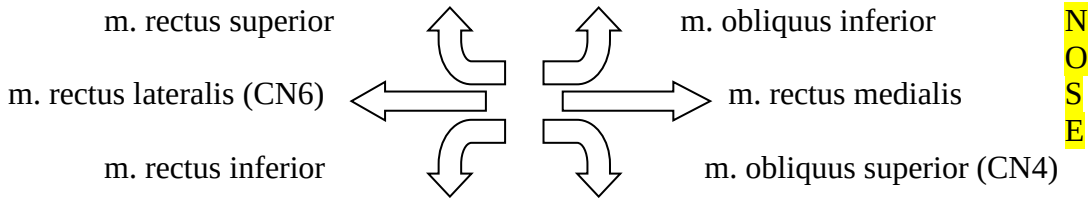
- on one or both sides of face.
- subacute onset of boring facial ***pain*** → ***paralysis*** of motor cranial nerves.
- clinical features overlap those of *Tolosa-Hunt syndrome*.
- frequently responsive to **steroids**.

Vascular compression syndromes:

1) trigeminal neuralgia
2) hemifacial spasm
3) CN9 neuralgia
4) torticollis

Ekstraokulinių raumenų testavimo kryptys

N.B. tai ne raumenų veikimo kryptys, bet būdas izoliuotai testuoti kiekvieną raumenį



CN1

Whole-mouth **taste function** is much more resilient to alterations than is **olfactory function**, in large part because taste buds have redundant innervation (i.e. CN VII, IX, X).

Complaint of taste loss usually reflects olfactory disorder!

DYSOSMIA - perverted smell perception:

PAROSMIA (CACOSMIA) – "rose smells more like garbage" (e.g. in "uncal fits").
PHANTOSMIA (OLFACTORY HALLUCINATION) - medicine-like smell in absence of odor stimulation.

- frequent during olfactory epithelial *degeneration / regeneration*.
  N.B. differentiate from foul odors produced within nasal cavity (e.g. infections) or within body proper (e.g. altered metabolism).

- *problem duration* is important - spontaneous recovery is unlikely after 6 months if damage to olfactory epithelium has occurred.

1. Alterations in ability to smell - first signs of ***Alzheimer's disease, idiopathic Parkinson's disease*** (but patients are unaware!)
2. ***Head trauma*** (anosmia / hyposmia is frequently the only residual neurological impairment)
3. ***CNS tumors***
   e.g. tumors in olfactory groove or sphenoid ridge (e.g. meningiomas) can cause *Foster Kennedy syndrome* (ipsilateral anosmia, ipsilateral optic atrophy, contralateral papilledema).

TRIGEMINAL NEURALGIA (TIC DOULOUREUX)

- most common neuralgia!!!
- may drive sufferers to suicide
- N.B. appearance in young patient - suspicion of *demyelinating disease*!

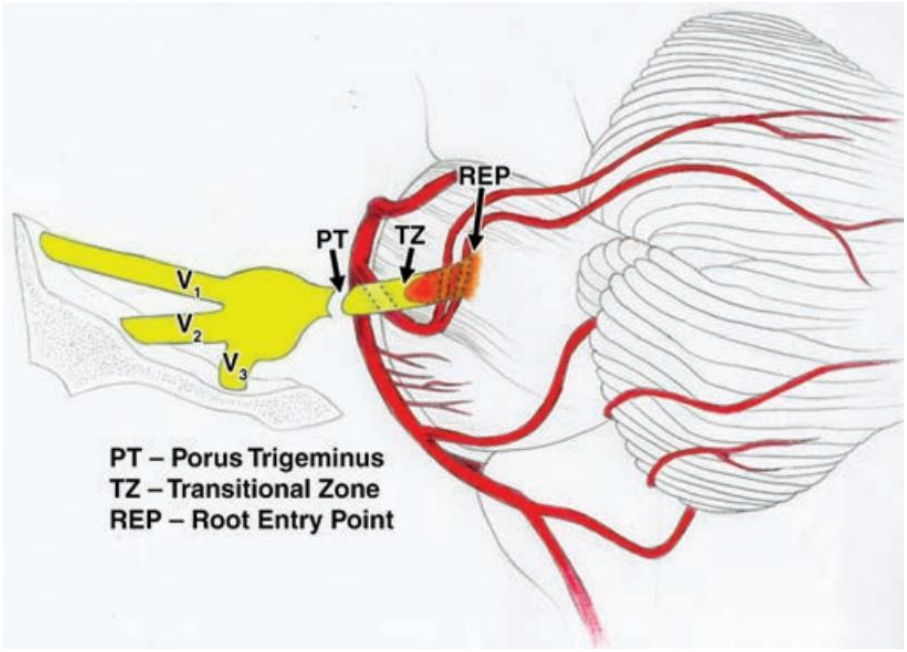
ETIOLOGY

a) **SECONDARY** - ***tumors*** near gasserian ganglia, ***multiple sclerosis****, ***postherpetic***, syringomyelia, infarction, aneurysm, (iatrogenic) trigeminal deafferentation

*denuded axons promote *ephaptic transmission*

Trigeminal neuralgia related to MS is more difficult to manage pharmacologically and surgically!

b) **IDIOPATHIC** (patients are > 40 yo)
N.B. most idiopathic cases are due to *pulsations of aberrant vascular loop* compressing root between entry point (old term – "root entry zone") and transition zone! – **NVC (neurovascular contact)**



- most commonly - **SCA** (75%) or AICA; less commonly – vein.
- with aging, blood vessels can become ectatic.

- BIOPSY - ***focal demyelination*** but no inflammatory cells.

CLINICAL

- brief excruciating **lightning-like series of jabs (tics!)**; jab lasts **fraction of second**, episode lasts **seconds ÷ few minutes**.
- **unilateral** (in ≈ 5% bilateral, but simultaneous bilateral spasms are quite atypical).
- strictly affects divisions of CN V (in 15% all three divisions):

3^{rd} (70%) > 2^{nd} > 1^{st} (5%)

vs. postherpetic neuralgia most frequently affects CNV_1

- > 90% have demonstrable **sensory trigger point**.
- *between attacks, there are no symptoms*.

N.B. it is neuralgic pain (vs. neuropathic pain – constant burning pain associated with numbness)

- *no neurologic deficits*!!! (only exception: mild sensory loss).

Significant sensory loss suggests secondary process!!!!!

- disease lasts indefinite years (severity steadily increases).

BURCHIEL (2003)

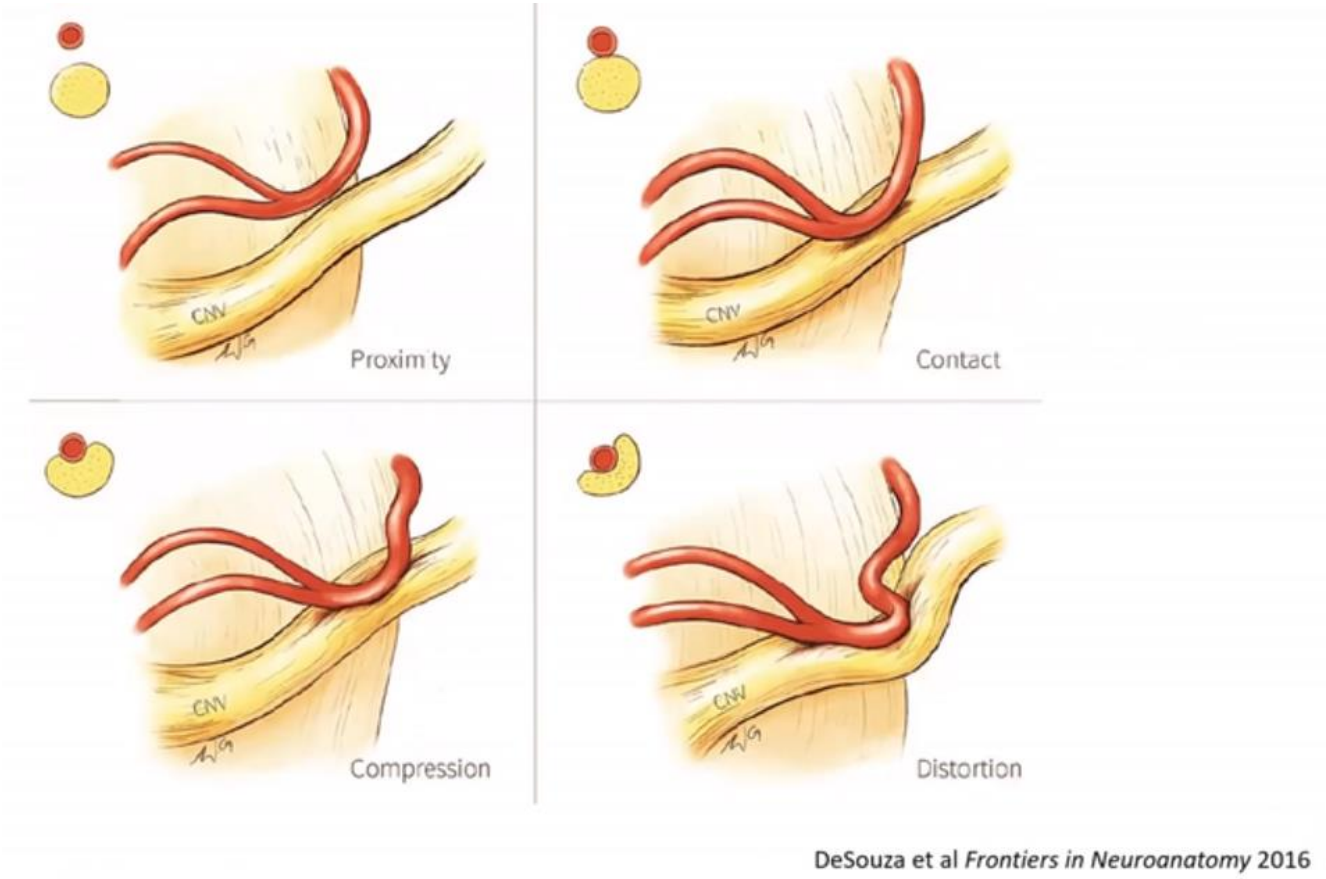
type 1 (> 50% episodic pain)
type 2a (> 50% constant pain with history of episodic pain)
type 2b (constant pain with no history of episodic pain)

- natural history is gradual transition from Type 1 to Type 2

DIAGNOSIS

- diagnosis can usually be made by history alone.

- **MRI** is only test always indicated (in 0.8-15% cases tumor is found!) – CISS or FIESTA



DeSouza et al *Frontiers in Neuroanatomy* 2016

MEDICAL THERAPY

(many require lifelong medication!):

1) **CARBAMAZEPINE**!!!* – first-line & most effective
   - started 100 mg BID; max daily dose 1200 mg;
   - side effects: drowsiness, rash (possible Stevens-Johnson syndrome), relative leukopenia is common
   - follow serum levels, LFT and WBC to avoid toxicity.
   - dose may be tapered once pain is controlled, since remission may occur.
2) **OXCARBAZEPINE**!!* – alternative.
3) **BACLOFEN**!! – second best choice
4) **LAMOTRIGINE**
5) GABAPENTIN PREGABALIN

- intravenous fosphenytoin (250 mg) - for acute severe attack (**status trigeminus**), e.g. in too much pain to open their mouths to take carbamazepine orally
- 50-75% patients eventually have some kind of surgical procedure!

  many experts believe that patients failing to respond to first-line therapy are unlikely to respond to alternative medications and suggest early surgical referral.

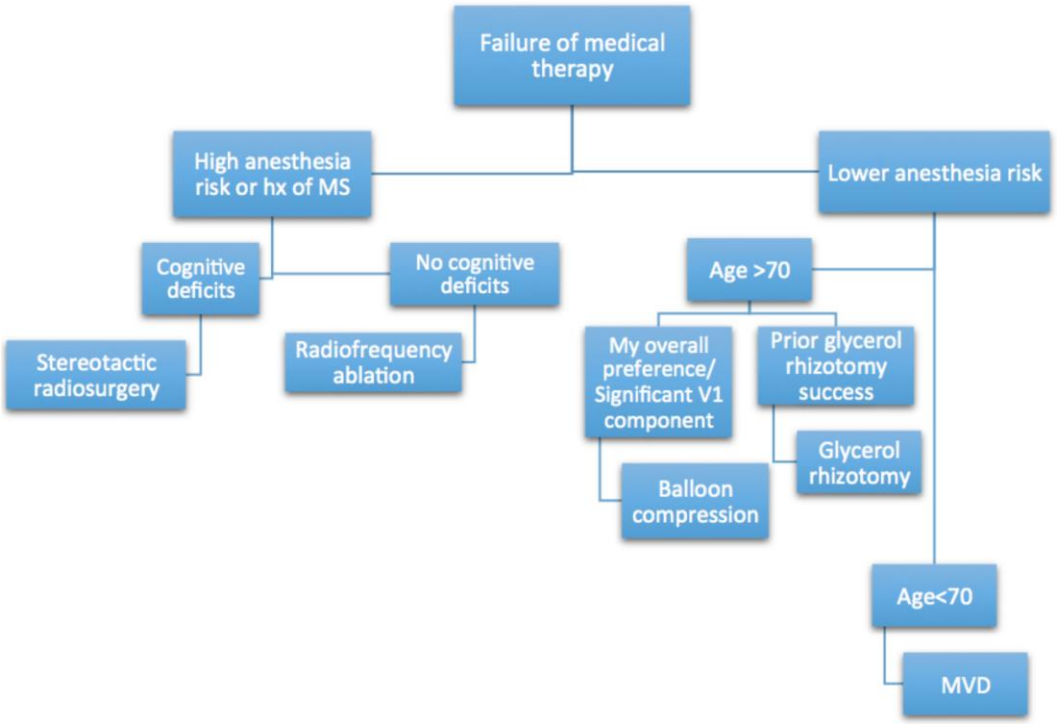
*test Asians for HLA-B*152 allele

SURGICAL THERAPY

Many options available – ***patient characteristics*** are important.
N.B. no treatment exist for anesthesia dolorosa (CN5 deafferentation pain) H: **trigeminal tractotomy, motor cortex stimulation.**
N.B. atypical pain (constant, burning) worsens with MVD!

- treatment failure occurs in most of **MS-related** cases independently of type of treatment (maybe balloon is the best)

Fit for surgery (even if MRI shows no vascular conflict) → MVD
Unfit, secondary cases, on "blood thinners" → SRS (if rapid result is needed, then RF*/balloon**/glycerol***)
For all recurrences → SRS
*preferred for V3
**preferred for V1-2 (less effect on corneal protective reflex)
***preferred for bilateral pain



- after any procedure, check for:
    1) *corneal reflex* (usually just mild decrease) – if impaired, needs eye protection.
    2) facial numbness
    3) jaw opening weakness / deviation.

Outcomes:

	RF	Glycerol	Balloon	MVD
Initial success	91-99%	91%	93%	85-98%
Recurrence 2-6 yrs	19% 6 yrs	54% 4 yrs	21% 2 yrs	15% 5 yrs
Recurrence >10 yrs	80% 12 yrs			30% 10 yrs
Facial numbness	98%	60%	72%	2%

1. DISTAL (PERIPHERAL) PROCEDURES

- generally not recommended - high incidence of early recurrence.

2. PERCUTANEOUS GANGLIOLYSIS

Idea: to *selectively destroy A-delta and C fibers (nociceptive)* while preserving A-alpha and beta fibers (touch).

- recurrence rates and incidence of dysesthesias are comparable among various lesioning techniques.
- numbness is expected in successful cases (numbness is not complication!; beware neuropathic keratitis! H: artificial tears 2 gtt q 2 hrs while awake, Lacrilube® & tape eye shut at bedtime).
- bradycardia and hypotension: 1% with RF, 15% with glycerol, very common with balloon.
- intracranial hemorrhage (6 fatal cases in > 14,000 procedures) - due to transient HTN (SBP up to 300)

RF thermocoagulation

- under MAC (other procedure – under general)
- **patient has to be awake** (N.B. may cause HTN!)- electrode position is manipulated until *paresthesias (upon stimulation) are confined to distribution in which pain is located* – can ablate selective branch (V1 or 2 or 3).
- 60º x 60 sec
- must produce *HYPOESTHESIA* in pain distribution (if complete anesthesia - risk of postoperative anesthesia dolorosa) - *continuous sensory testing is ideal*.
- lowest recurrence rates of all percutaneous procedures!

Glycerol rhizotomy

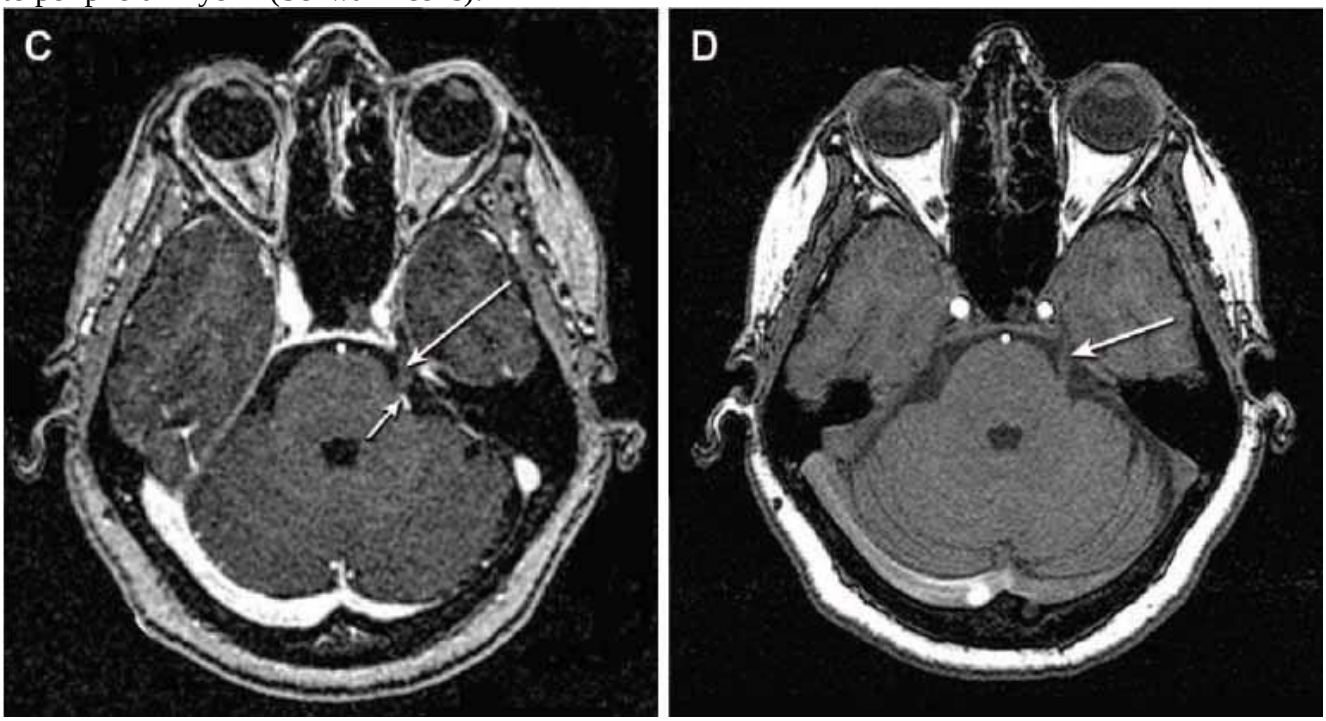
- cannulate trigeminal cistern with18G ***foramen ovale needle***
- needle is left in place → patient is seated upright with head flexed.
- empty Meckel's cave by letting CSF drip.
- sterile anhydrous **GLYCEROL** injection with tuberculin syringe; volume – glycerol fills Meckel's cave from bottom up: if treating V3 – enough 0.2 cm^3, for V1 – need 0.4 cm^3 (Meckel's cave volume 0.4 cm^3); Dr. Broaddus injects 0.5 cm^3 in all cases.
- extubated sitting upright with head flexed for **2** hours after procedure.
  N.B. if neck is extended at any time, glycerol is lost – procedure is in vain
- neuralgia relief is immediate; if onset is delayed for > 7 days, likely result will be poor.

Balloon (percutaneous balloon compression, PBC)

- 13G needle with a semisharp stylet inserted through a stab incision 2 to 3 cm lateral to the angle of the mouth, directed into the oval foramen
- **4F Fogarty balloon** catheter inserted 17-19 mm beyond the needle tip → balloon inflated with 0.3-0.8 mL Omnipaque – "pear-shaped" configuration (reflects shape of Meckel's cave) → 1.4 atm pressure held for 1-2 minutes → contrast is aspirated.
- bradycardia occurs regularly! (have atropine ready)
- instant pain relief (with associated sensory loss; *temporary masseter weakness* is common).
- lowest risk of corneal anesthesia.

3. POSTERIOR FOSSA (ROOT ENTRY POINT) PROCEDURES

Transition zone (s. **Obersteiner-Redlich zone**) - where central myelin (oligodendroglial cells) changes to peripheral myelin (Schwann cells):



Microvascular decompression (MVD), s. Jannetta procedure

– classic, most effective procedure! (addresses etiology!); durable and nondestructive; risks associated with craniotomy and general anesthesia

- indicated for *younger, healthier patients* (without MS ← low response rate) with life expectancy > 5 years.

  Gold standard treatment for most TN patients unless they have significant comorbidities!

  N.B. results are long lasting, facial anesthesia is uncommon - anesthesia dolorosa does not occur!
- **otovestibular testing** is performed in all preop - hearing loss is occasional complication of MVD.

Retrosigmoid craniotomy in park-bench position
see p. Op300 >> then see p. Op350 >>

- facial numbness is common after MVD? (check corneal sensation!)
- 20% patients develop *chemical meningitis* (still do LP) from Teflon at 1-2 weeks postop; H: steroids.

Risk factors for recurrence: lack of immediate post-operative relief, female sex, venous compression, symptoms of > 8 years
Factors that favor the success: typical trigeminal pain, objective evidence of vascular compression, short history prior to surgery, no previously failed procedure.

Predictors of pain freedom after MVD on meta-analysis:
(1) type 1 pain Burchiel classification (OR = 2.49, 95% CI = 1.32-4.67).
(2) disease duration ≤ 5 yr (OR=2.06, 95% CI=1.08- 3.95)
   — demyelination after a certain disease duration may be severe enough to induce an irreversible central sensitization even after decompression
(3) arterial compression over venous or other (OR = 3.35, 95% CI = 1.91-5.88)

TABLE 2. Derivation of TN Grading System Score from Composite Clinical Characteristics

Composite characteristics	Assigned point value
TN symptom type[a]	
Nonclassical	0
Classical	1
Response to medication[b]	
No	0
Yes	1
Neurovascular contact[c]	
Absent or venous only	1
Arterial contact	2
Arterial deformity	3
Total TN grading system score[d]	1 to 5

TABLE 6. Probability of Long-Term Pain Relief Without Medication by TN Grading System Score

TN grading system score[a]	Probability of long-term pain-free status, % (95% CI)[b]
1	4% (0.03-0.06)
2	16% (0.12-0.21)
3	44% (0.40-0.47)
4	76% (0.74-0.78)
5	93% (0.92-0.93)

[a]TN grading system score derived from adding points from 3 characteristic categories, points were assigned as follows: TN type (1 point for classical, 0 for nonclassical), response to medication (1 point for yes, 0 for no), and neurovascular contact (3 points for arterial deformation, 2 point for arterial contact, 1 points for absent or venous contact).
[b]Estimated probability and 95% CI (95% CI).

Vasculopexy

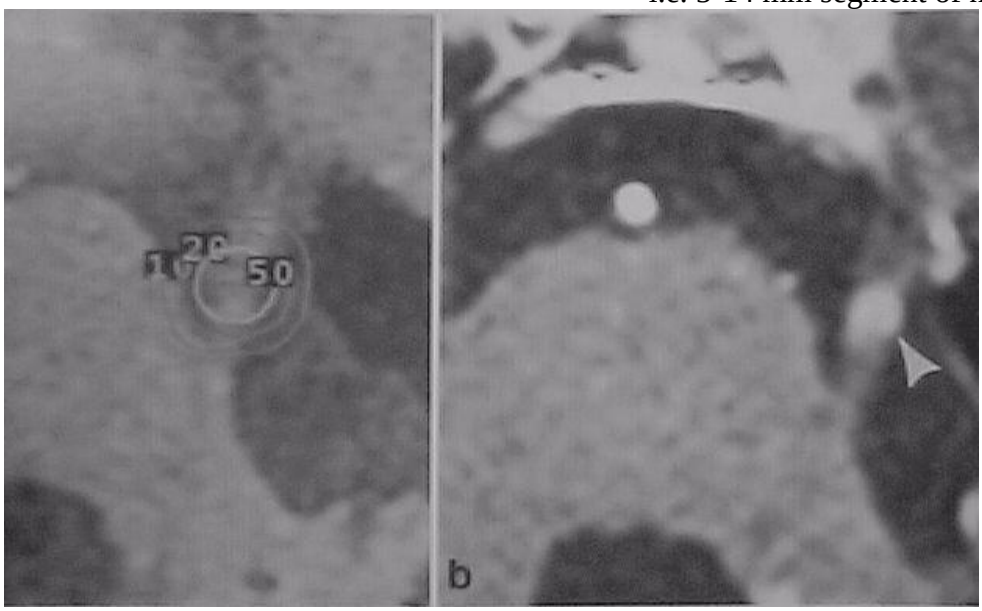
- if compression caused by a *severely ectatic and tortuous basilar artery* – BA **vasculopexy** to the tentorium.
  - — subtemporal transtentorial approach
  - — basilar artery mobilized away from CN5.
  - — suture is passed through the wall of the basilar artery (tunica media) and secured to the tentorial edge, to keep artery away from nerve.

Radiosurgery rhizolysis

- least invasive safe procedure with low morbidity.
  Avoid for patients with atypical features!

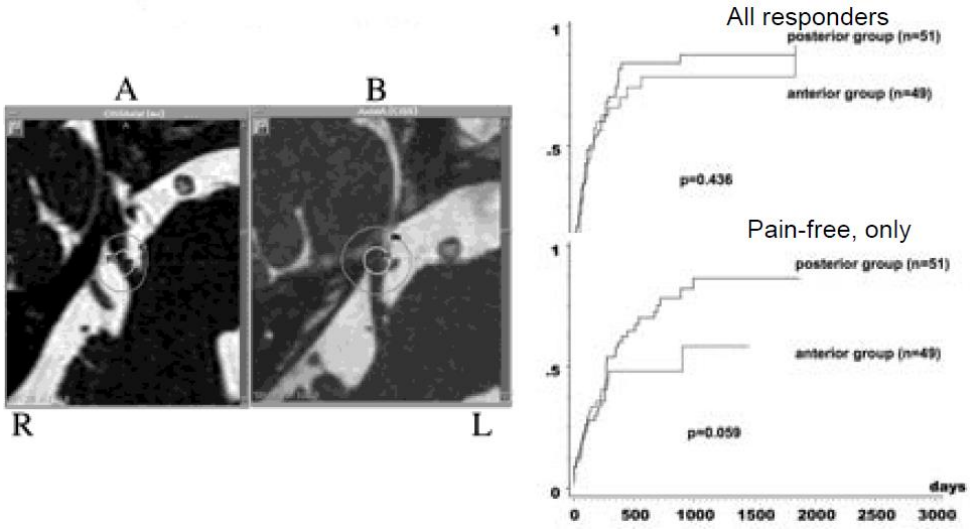
- pre-radiosurgery pain medications are continued at the same doses until pain relief is obtained → medications gradually tapered off if the patient remains pain free.
- standard (posterior) approach: single dose of 86 Gy at 100% isodose (or 43 Gy at 50% isodose) to trigeminal root (single 4-mm isocenter at 5-14 mm distance anterior to emergence of nerve*, so 50% isodose is next to but does not touch brainstem).

*i.e. 5-14 mm segment of nerve treated



- alternative (anterior) approach: target distal cisternal portion.

Comparison of two targeting methods:



Complications: hypesthesia, troubling dysesthesias.
Increasing volume (to include more of nerve root) increases complications but does not provide better pain relief!

Outcome

- takes time, mean 4 weeks to reach effect.
- 55% excellent relief
- 1 month follow up - **acute toxicity**:
  Facial numbness < 10%
  Neuropathic pain < 1%
  Motor weakness < 1%
  Corneal keratitis (5-7%)
- at 2 years failure rate is ≈ 35-40% → repeat SRS.

Repeat SRS:

- target is placed anterior to the first target so that the volumes overlap by 50%.
- use lesser dose (50–70 Gy) – to avoid facial sensory symptoms.
- safe interval is 6 months.

MVD vs. Gamma Knife

	MVD	Gamma Knife
Initial success	89-100%	57-77%
5 years	61-80%	33-56%
Cost	slightly more costly	
Numbness and dysesthetic pain		much more common

N.B. some treatment failures are not persistent TN, but rather represent trigeminal neuropathic pain (trigeminal deafferentation pain).

See N7 case >>
See Case F3 >>

GRADENIGO SYNDROME

– ***apical petrositis*** (osteomyelitis) with localized meningitis involving **CN5 & CN6**:

- may also involve CN7 (facial palsy), CN8 (deafness)
- multiple approaches to infected petrous cells are possible:

a) if it is complication of otitis media or mastoiditis: ***simple mastoidectomy*** → air cell track containing granulation tissue can be followed into petrous apex and adequate drainage can be obtained.
b) ***middle cranial fossa approach***.

FACIAL PALSY

1. **Trauma** (fracture of temporal bone, birth trauma)

improvement is rule (when palsy is associated with head trauma), but recovery may not be complete

2. **Surgery**:
   a) palsy immediately after surgery - ***nerve transection*** → prompt surgical evaluation
   b) delayed onset - ***nerve edema*** - improves with time.

Diagnosis

1. **ELECTROPHYSIOLOGY**:
   1) **ENoG (electroneurography)**.
      – even in complete transection, nerve conducts impulses distal to injury for 72 hours (until wallerian degeneration occurs) - ENoG is not performed until more than 3 days after total paralysis!
      – ENoG is not employed in paresis - even minimal voluntary motion after 3 days indicates minor injury, with full recovery to be expected.
   2) **trigeminofacial reflex** (electrodes record blink reflex after percutaneous stimulation of supraorbital nerve) - conduction of reflex arc between CN5 (afferent) and CN7 (efferent) – the only direct test able to measure intracranial CN7 pathology!!!
2. If facial reanimation procedure is considered (in longstanding or congenital facial paralysis) → facial **MUSCLE BIOPSY** (whether viable muscle fibers are present).

HOUSE-BRACKMANN GRADING SCALE

Grade	Definition
I	Normal symmetrical function in all areas
II	Slight weakness noticeable only on close inspection Complete eye closure with minimal effort Slight asymmetry of smile with maximal effort Synkinesis barely noticeable, contracture, or spasm absent
III	Obvious weakness, but not disfiguring May not be able to lift eyebrow Complete eye closure and strong but asymmetrical mouth movement with maximal effort Obvious, but not disfiguring synkinesis, mass movement or spasm
IV	Obvious disfiguring weakness Inability to lift brow Incomplete eye closure and asymmetry of mouth with maximal effort Severe synkinesis, mass movement, spasm
V	Motion barely perceptible Incomplete eye closure, slight movement corner mouth Synkinesis, contracture, and spasm usually absent
VI	No movement, loss of tone, no synkinesis, contracture, or spasm

CEREBELLOPONTINE ANGLE SYNDROME

- ipsilateral **full (upper & lower) face** paralysis.

- stapedius paralysis (HYPERACUSIS - painful sensitivity to loud sounds), tearing↓ (XEROPHTHALMIA), salivation↓ (XEROSTOMIA), taste↓.
- CN8 is often involved; cerebellum and CN5 can also be involved.

FACIAL CANAL SYNDROME

a) LABYRINTHINE SEGMENT – most full facial palsy (SVE, SVA, GVE, GVA, GSA)
b) TYMPANIC SEGMENT – lacrimation spared.
c) MASTOID SEGMENT distal to chorda tympani – only motor facial palsy.

STYLOMASTOID FORAMEN SYNDROME

- only motor facial palsy.
- some motor branches may be spared.

Treatment

If you cannot keep contact lens moist, do not wear it!

Any corneal abrasion / infection should be treated immediately!

- ELECTRICAL STIMULATION continues to be widely used although there is mounting evidence that it may be contraindicated:
  - *facial muscles resist degeneration* post denervation for longer periods of time than other skeletal muscle - facial muscles may remain viable for 3 or more years!
  - electrical stimulation may interfere with neural regeneration!
  - patients who undergo electrical stimulation acutely may demonstrate more synkinesis and mass action

N.B. one of first signs of regenerating nerve may be pain; in case of CN7 – pain in ear canal!

- *wait minimum 6 months* for spontaneous CN7 function return (exception - *nerve cut during surgery* – restore nerve continuity ASAP!)
- extracranial CN12→CN7 or CN11→CN7 ***anastomosis***

All complete TRAUMATIC cases must be considered for surgical repair!

BELL'S PALSY (s. idiopathic acute facial palsy)

- 60-70% had **viral prodrome** 7-10 days before.
- most commonly accepted cause is **HERPES SIMPLEX-1** infection.
- sudden onset; maximal facial weakness is reached within 48-72 hours (vs. CN7 schwannoma – progresses over > 3 weeks).
- pain is absent (if present – consider Ramsay-Hunt syndrome).
- diagnosis of exclusion!!!
- no imaging is required!
  N.B. perform MRI if tumor is suspected!
- in endemic areas, *Lyme disease* is consideration.
- **ENoG (electroneurography)** during days 4-21 – for prognostic purposes: denervation indicates axonal degeneration - will be long delay (3 months, as rule) before regeneration occurs (may be incomplete).
- N.B. *all patients should show some improvement by 6 months*! (vs. CN7 schwannoma – gradually worsening paralysis, steroid responsiveness eventually disappears)
- mainstay of treatment - 10-day course (must be started early!) of PREDNISONE (60 mg/d for 5 days → tapered down by 10 mg/day for 5 days) – level A recommendation!
- adding ACYCLOVIR is level C recommendation;

ACYCLOVIR alone is not effective in facial recovery!!! (must be used only in COMBINATION WITH PREDNISONE).

According to newest studies *surgical decompression of CN7 is not beneficial* in Bell palsy!

RAMSAY-HUNT syndrome (s. herpes zoster oticus)

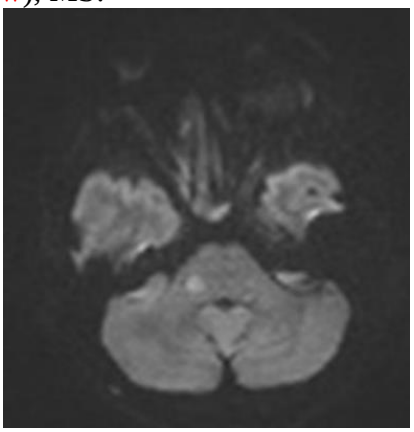
- **herpes zoster** of geniculate ganglion:
  1) very painful vesicular eruption on pinna, external auditory canal, pharynx
  2) severe CN7 palsy
  3) often CN8 is affected (vertigo, high-tone deafness)

Treatment – VALACYCLOVIR.

HEMIFACIAL SPASM

- involuntary, unilateral, painless, episodic **tonic & clonic** contraction of CN7 muscles.

- begins as *twitches around eye* (always starts in orbicularis oculi with upgoing eyebrow – "reverse Babinski") → progresses (during few months) to involve remaining ipsilateral facial muscles (99% **unilateral**)
- only movement disorder that persists in sleep! (also palatal myoclonus)
- etiology (generally, only cosmetic problem but ability to see with involved eye is impaired!)
  a) compression of motor nerve root at brain stem by aberrant / ectopic **vascular loop** e.g. **AICA** loop in internal auditory canal.
  b) recovery from Bell's palsy, tumor in CP angle, MS, bony abnormalities.
- differentiate from *blepharospasm* (involuntary spasm of **bilateral** orbicularis oculi muscles with downgoing eyebrow), MS:



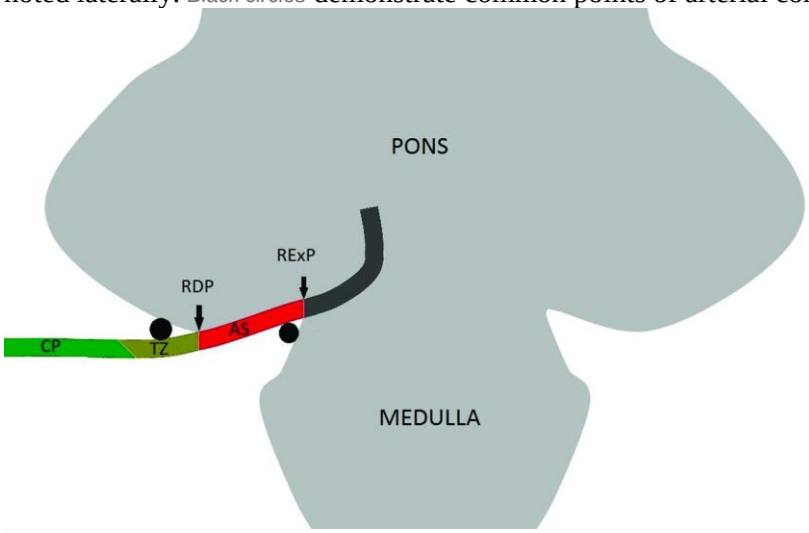
- diagnosis:
  MRI (vascular loop).
  EMG – 5-20 Hz bursts of muscle action potentials.
- treatment:
  1) BOTULINUM TOXIN – treatment of choice for mild cases!!!
     - carbamazepine, clonazepam, baclofen – not effective
  2) procedure of choice - **MVD** between root exit point and transition zone, i.e. attached segment*

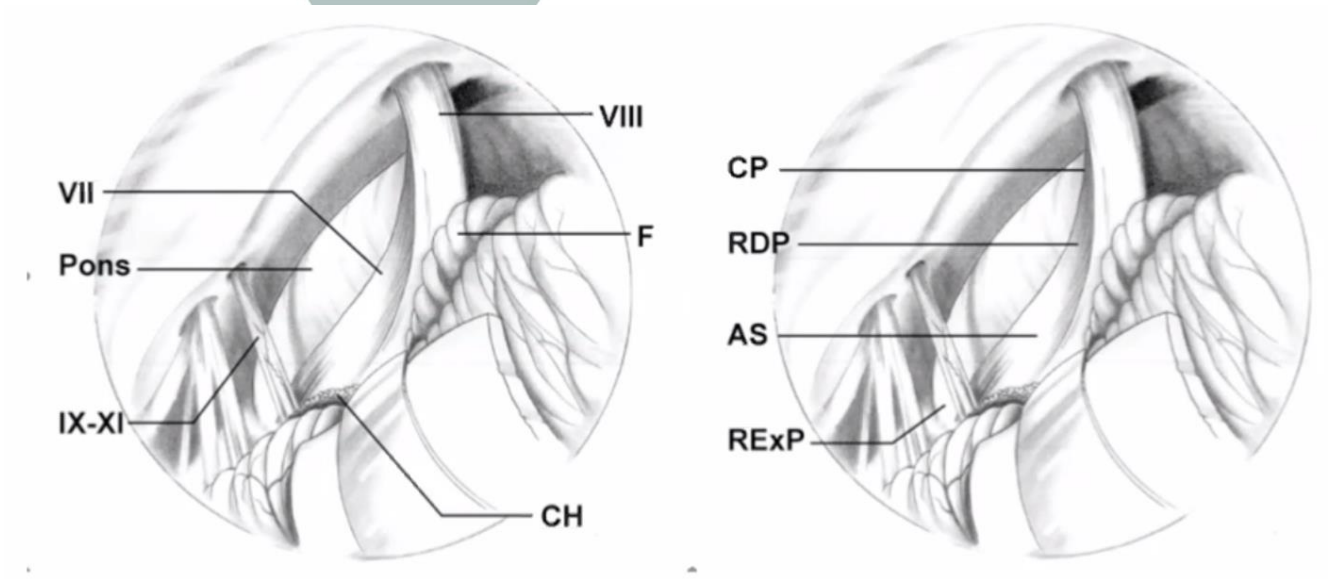
     *more distal decompressions are ineffective!

     - facial nerve should not be manipulated!
     - vessels must be preserved, esp. cochlear artery and small perforators.

     N.B. risk of hearing loss is ≈ 20%
     Failure of relief → reoperate

Coronal drawing: facial nerve exiting the brain stem at the root exit point (RExP) in the pontomedullary sulcus. The attached segment (AS) of the nerve then runs along the pons until the nerve separates from the brain stem at the root detachment point (RDP). The transition zone (TZ) is the 3- to 4-mm segment of the facial nerve where the central glial myelin transitions into the peripheral myelin. The cisternal portion (CP) of the facial nerve is noted laterally. *Black circles* demonstrate common points of arterial compression of the facial nerve





MYOKYMIA

- progressive, irregular **fibrillation** of individual facial muscle fibers.

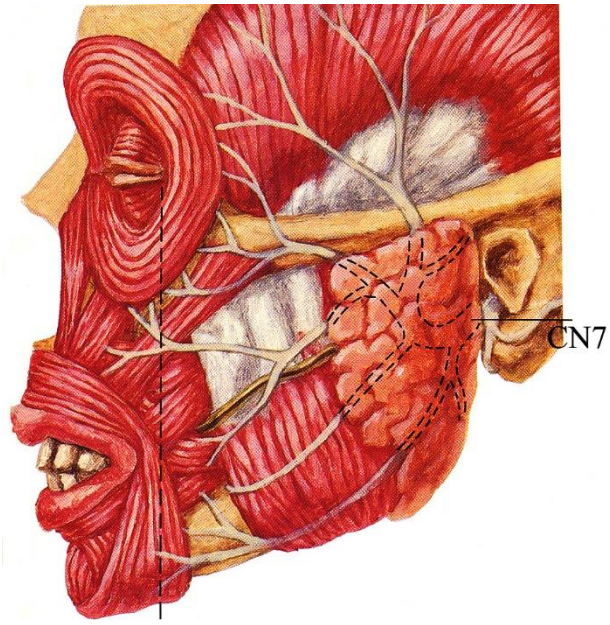
- begins in frontalis → extends to involve all ipsilateral facial muscles.
- etiology (grave! vs. hemifacial spasm) - pontine gliomas, multiple sclerosis!

FACIAL NERVE TRAUMA

CN7 injuries should be repaired surgically if they occur ***posterior to vertical line drawn from LATERAL CANTHUS of eye***.

- injuries ***anterior to this line*** do not need to be repaired (with microsurgical techniques, some branches can be rejoined).

- if nerve is *cut by sharp object* → repair primarily if wound site is clean.
- *destructive injuries* → nerve repair is usually secondary using graft.



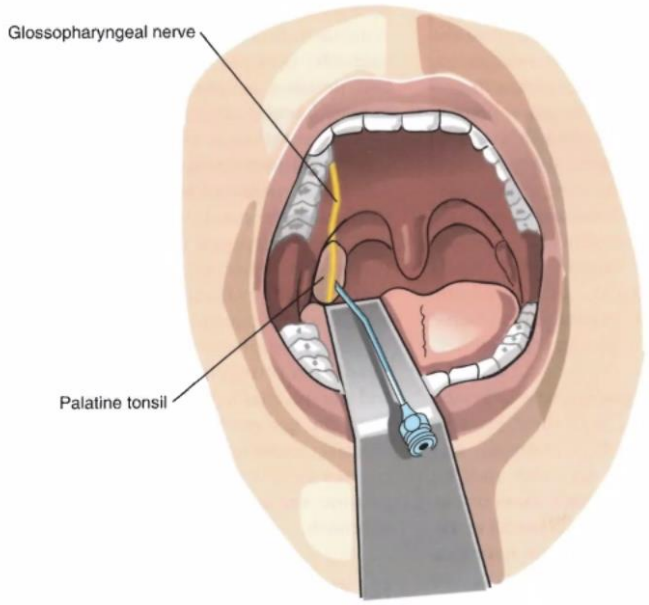
GLOSSOPHARYNGEAL NEURALGIA (s. TIC DOULOUREUX of CN9)

N.B. no association with MS (vs. trigeminal neuralgia)
PAIN - similar to ***trigeminal neuralgia*** - in middle ear, tonsil, base of tongue, posterior pharynx, angle of jaw, larynx.
May have diminished gag (on affected side)
anesthetizing throat (with LIDOCAINE on applicator or spray) may temporarily **relieve** pain - patient can swallow food and talk without discomfort.

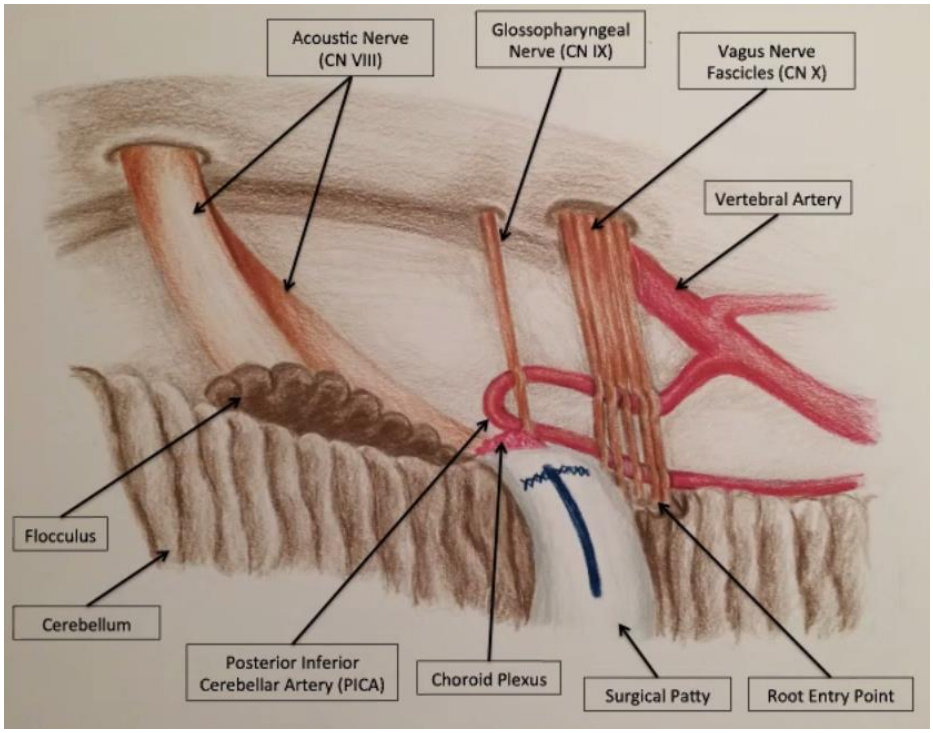
Medical - identical to ***trigeminal neuralgia*** (CARBAMAZEPINE is drug of choice).

Surgical:

a) ***rhizotomy*** of glossopharyngeal nerve (definitely terminates symptoms but sacrifices nerve) – test with block:



b) ***microvascular decompression*** (most common offending blood vessel is **PICA**).


Acoustic Nerve (CN VIII)
Glossopharyngeal Nerve (CN IX)
Vagus Nerve Fascicles (CN X)
Vertebral Artery
Flocculus
Cerebellum
Posterior Inferior Cerebellar Artery (PICA)
Choroid Plexus
Surgical Patty
Root Entry Point