

Parsonage -Turner syndrome Neuralgic amyotrophy

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No conflicts of interest

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Goal

- Understand Parsonage-Turner syndrome
 - Symptoms
 - Diagnosis
 - Management
 - Outcome
 - Pathophysiology

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Outline

- Definition of syndrome
- Symptoms
- Clinical findings
- ENMG
- Imaging
- Etiology
- Treatment
- Prognosis
- Cases

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ORIGINAL ARTICLES


NEURALGIC AMYOTROPHY THE SHOULDER-GIRDLE SYNDROME

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

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■ Maurice John Parsonage

- 1915 - 2009
- Neurologist
- Epilepsy, EEG



■ John William Aldren Turner

- 1911-1980
- Neurologist

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Precipitating causes

Precipitating Cause	Cases	Precipitating Cause	Cases
Operations	12	Infections	71
Herniotomy	8	Malaria	16
Appendectomy	1	Typhus and malaria	5
Varicocele	1	Typhus	4
Floundal cyst	1	Typhoid	1
Mastoid	1	Dysentery	5
Trauma	10	Smallpox	1
Gunshot wound of remote parts	5	Glandular fever	1
Minor local trauma	5	Rheumatic fever	1
Other conditions	5	Chest infections	9
Lumbar puncture	1	Septic infections	9
Air encephalogram	1	Minor fevers	11
Antisyphilitic treatment	2	Polioomyelitis	2
Severe exposure	1	Diphtheritic polyneuritis	6

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SUMMARY

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

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

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

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

The name "neuralgic amyotrophy" is suggested.

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Lancet 1957:273(6988):209-12

NEURALGIC AMYOTROPHY (PARALYTIC BRACHIAL NEURITIS)
WITH SPECIAL REFERENCE TO PROGNOSIS

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In 1948 Spillane described localised neuritis of the shoulder girdle, which fairly often affected soldiers in the Middle East. The essential clinical picture consisted of pain around the shoulder and upper arm followed within a few hours to several days by atrophic paralysis. The distribution varied from the muscles supplied by a single nerve—e.g., the long thoracic and the circumflex—to more extensive paralysis indicating involvement of several peripheral nerves or nerve roots. There was no constitutional disturbance at the onset, cutaneous sensory changes, particularly in the distribution of the circumflex nerve, were often present, and the cerebrospinal fluid was normal in the acute stages—all features which distinguished the condition from anterior poliomyelitis.

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Terminology

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

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Classical PTS - Symptoms

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

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Atypical PTS

- No pain
 - 5% without pain
- Onset with paresis followed by pain
 - 5%

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Affected nerves

- Spinal nerves (nerve roots)
- Plexus brachialis
 - Any part may be affected
- Nerves in the arm
 - Anterior interosseus nerve
 - Posterior interosseus nerve
 - Lateral cutaneous of the forearm
 - Radial nerve sensory branch
 - Individual palmar digital nerves
- Individual distal nerve branches
 - Individual muscles
 - Sensory branches

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Affected nerves

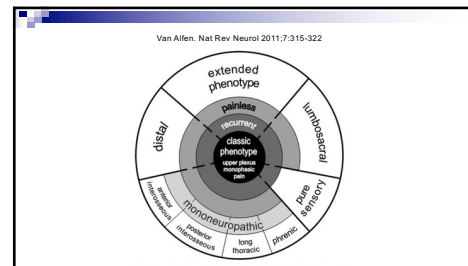
- Cervical plexus
 - Phrenic nerve (10%)
- Cranial nerves
 - IX (Glossopharyngeal nerve)
 - X (Vagus nerve or its branches)
 - XI (Accessory nerve)
 - XII (Hypoglossal nerve)
 - VII Facial nerve (Bell's palsy?)
- Individual leg nerves - sometimes, not often
 - Lumbar plexus
 - Proximal leg nerves

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PTS distribution

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

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

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

Hans van Alfen^{1,2}, Hester J. J. van Eijck³, Teunis Bouter⁴, Susan G. Pijpers⁵, Inge E. G. Meesters⁶, Jan T. Groenendaal⁷, Sigrid Hees⁸, Frank A. J. Swart^{9,10}

¹ Department of Neurology, Donders Centre for Neuroscience, Radboud university medical centre, Nijmegen, The Netherlands, ² Department of Neurology, Donders Research Institute, Donders Institute for Brain, Cognition and Behaviour, Donders Centre for Neuroscience, Radboud university medical centre, Nijmegen, The Netherlands, ³ Department of Neurology, Erasmus Medical Centre, Rotterdam, The Netherlands, ⁴ Royal College of Physicians, Nijmegen, Nijmegen, ⁵ Department of Neurology, Radboud University Medical Centre, Nijmegen, The Netherlands, ⁶ Department of Neurology, Donders Centre for Neuroscience, Radboud university medical centre, Nijmegen, The Netherlands, ⁷ Department of Neurology, Geesthuis Wilhelmina Hospital, Nijmegen, The Netherlands, ⁸ Department of Neurology, Geesthuis Wilhelmina Hospital, Nijmegen, The Netherlands, ⁹ Department of Neurology, Geesthuis Wilhelmina Hospital, Nijmegen, The Netherlands, ¹⁰ Department of Neurology, Geesthuis Wilhelmina Hospital, Nijmegen, The Netherlands

Incidence in primary care 1/1000/year

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PTS age and gender

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

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PTS constitutional risk factors

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

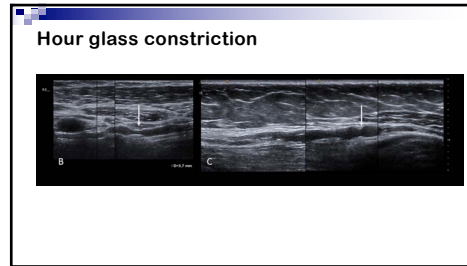
Constitutional risk factors for focal neuropathies in patients referred for electromyography

C. Martínez-Agüero^{1,2}, S. K. Jääskeläinen³, L. Pääkkö⁴, F. Reiche-Lortie⁵, P. Tornio-Poyatos⁶, J. Parrajas Sobol⁷ and B. Falck⁸

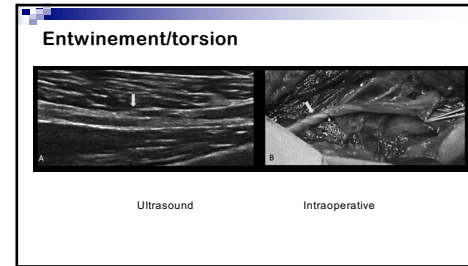
¹ Doctoral Programme of Medicine and Public Health, University of Granada, Granada; ² Department of Clinical Neurophysiology, Virgen Reyes de Mar Hospital, Almería, Spain; ³ Department of Clinical Neurophysiology, Turku University Hospital and University of Turku, Turku, Finland; ⁴ Department of Clinical Neurophysiology, University of Turku, Turku, Finland; ⁵ Department of Mathematics, University of Almería, Almería; ⁶ Faculty of Medicine, Hospital Universitario Clínico Granada; ⁷ Department of Clinical Neurophysiology, University Hospital Virgen de las Arceas, Granada, Spain; and ⁸ Department of Clinical Neurophysiology, University Hospital, Uppsala, Sweden

European Journal of Neurology 2020, 27: 529–535

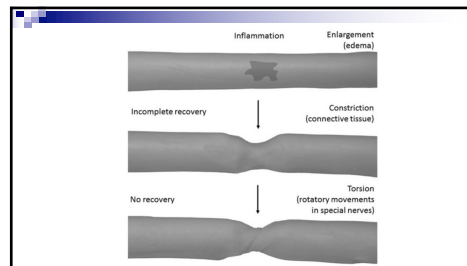
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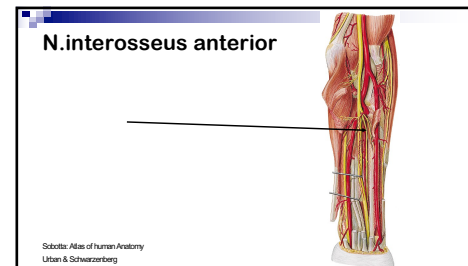
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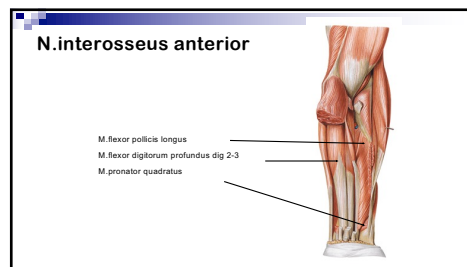
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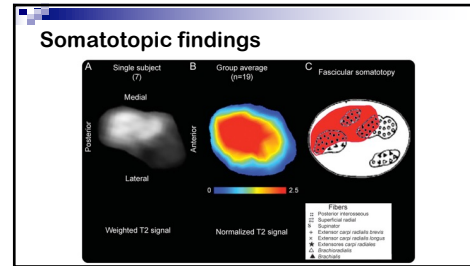


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N. anterior interosseus

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

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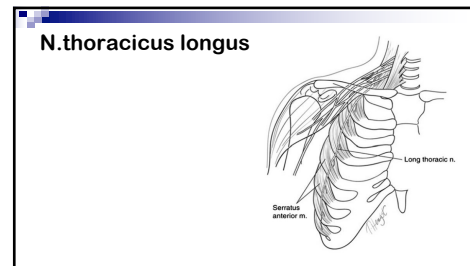


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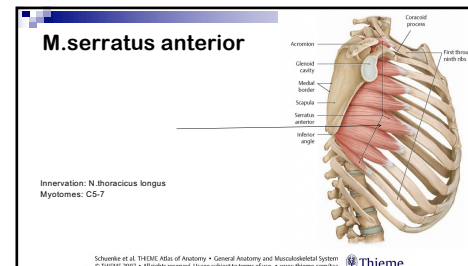
N.thoracicus longus

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

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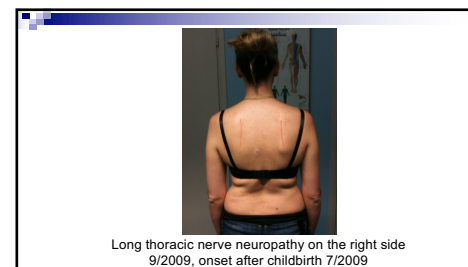
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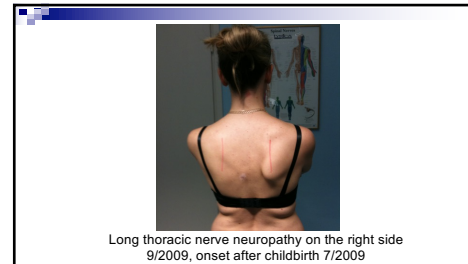
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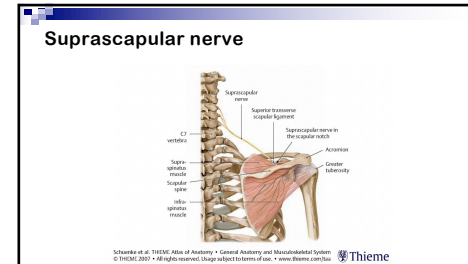
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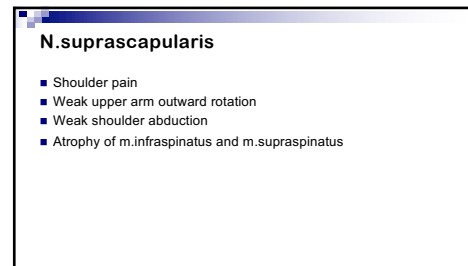
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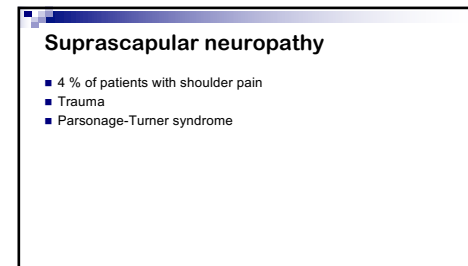
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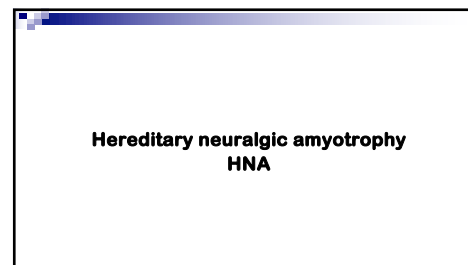
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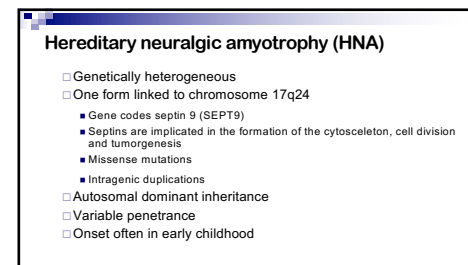
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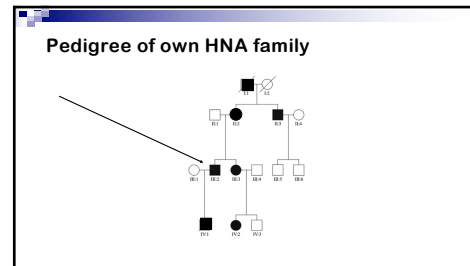
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HNA - Clinical features

- Onset usually in second to third decades
- Sometimes in the first decade
- Painful episodes of local nerve lesions
- Pain lasting a few days to a few weeks
- Often satisfactory recovery
- Repeated or severe attacks leave residual symptoms
- Penetrance high - 80%

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EMG findings in HNA

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

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Diagnosis

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Diagnosis

- Clinical picture
- Symptoms
- Laboratory tests
- Imaging
- ENMG

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Laboratory tests

- SR, CRP normal
- Autoimmune antibodies normal
- Spinal fluid
 - Sometimes elevated proteins or lymphocyte counts
- If risk factors present
 - Borrelia
 - HIV

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EMG

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

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Imaging

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

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Muscle & Nerve. 2022;66:709–714
CLINICAL RESEARCH ARTICLE **MUSCLE & NERVE** WILEY

Imaging of neuralgic amyotrophy in the acute phase

Paolo Ripellino MD, MSc¹ | Zsuzsanna Arányi MD, PhD² |
 Nens van Alfen MD, PhD³ | Elia Ventura MD⁴ | Anne-Kathrin Peyer MD, PhD⁵ |
 Alessandro Cianfoni MD, PhD^{6,8} | Claudio Gobbi MD¹ | Emily Pedrick BA⁷ |
 Darryl Brett Sneag MD⁹

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

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Muscle Nerve 56: 1054–1062, 2017

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

ZSUSZSANNA ARÁNYI, MD, PhD² ANITA CSILLIK, MD¹ KATALIN DEVAY, MD¹ MAJKA ROSERO, MD,³
 PÉTER BARSZ, MD, PhD⁴ JOSEF BÖHM, MD, PhD⁵ and THOMAS SCHELLE, MD⁶

¹MTA-SE NAP B Peripheral Nervous System Research Group, Department of Neurology, Semmelweis University, Balassa u. 6, Budapest 1058, Hungary
²Department of Traumatology, St. Imre and László Hospital, Budapest, Hungary
³MRB Research Center, Semmelweis University, Budapest, Hungary
⁴Neurologische Praxis, Dr. Friedrich Behse/Dr. Josef Böhm, Berlin, Germany
⁵Department of Neurology, Städtisches Klinikum Dessau, Dessau, Germany
 Accepted 23 May 2017

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US findings

Table 1. Summary of affected nerves with respective ultrasonographic findings

Affected nerves	Number (total)	Ultrasonographic findings			
		None	Swelling without constriction	Incomplete constriction	Complete constriction
AIN	16 (71)	2	10	3	1
Radial nerve / PIN	12 (11)	1	2	3	6 (3)
Long thoracic nerve	12	7	5	—	—
Suprascapular nerve	8	2	2	4	—
Accessory nerve	6	1	5	—	—
Axillary nerve	5	3	1	—	1
Musculocutaneous nerve	3	—	1	1	1 (1)
Median nerve	2	—	2	—	—
Superficial radial nerve	2	—	1	—	1
LAICN	1	—	—	1 (1)	—
Thoracodorsal nerve	1	1	—	—	—
Dorsal scapular nerve	1	1	—	—	—
Upper trunk	1	—	1	—	—
Total	70 (100%)	18 (26%)	30 (43%)	12 (17%)	10 (14%)

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

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Outcome with conservative therapy

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

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Treatment

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

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J Neurol Neurosurg Psychiatry 2009;80:1120-1124

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

J J J van Eijk,¹ N van Alfen,^{1,2} M Berrevoets,¹ G J van der Wilt,³ S Pillein,² B G M van Engelen¹

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J Neurol Neurosurg Psychiatry 2009;80:1120-1124

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

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

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

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

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Gstoettner C, et al. J Neurol Neurosurg Psychiatry 2020;91:879-887

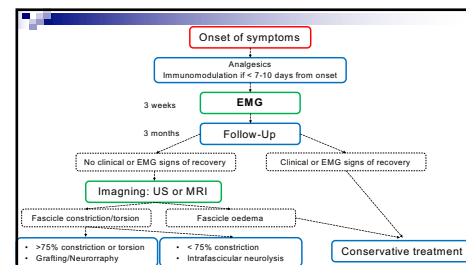
Neurosurgery

REVIEW

Neuralgic amyotrophy: a paradigm shift in diagnosis and treatment

Clemens Gstoettner¹, Johannes A Mayer,^{1,2} Stephanie Rassam,^{1,3} Laura A Hruby,^{1,4} Stefan Salminger,^{1,5} Agnes Sturm,^{1,6} Martin Aman,^{1,7} Leila Harhaus,⁷ Hannes Platzgummer,⁸ Oskar C Aszmann^{1,5}

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J Hand Surg Am 2021;46:43-53

EDITOR'S CHOICE

Outcomes of Microneurolysis of Hourglass Constrictions in Chronic Neuralgic Amyotrophy

Karthik R. Krishnan, MS¹; Darryl B. Szeag, MD^{2††}
 Joseph H. Feinberg, MD³; Ogonna K. Nwawaka, MD^{2††}
 Steve K. Lee, MD⁴; Zausanna Arinyi, MD, PhD⁵; Scott W. Wolfe, MD⁴

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J Hand Surg Am 2021;46:43-53

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

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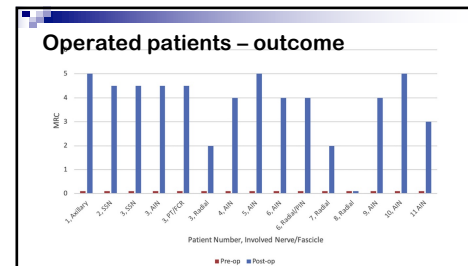
J Hand Surg Am 2021;46:43-53

TABLE 3. Changes in Electrodiagnostic Outcomes

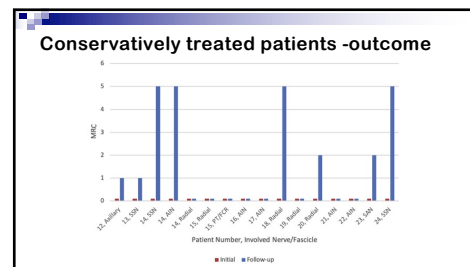
Outcome	Classification Category	Before Surgery	After Surgery
Motor unit recruitment	None	30	6
	Low discrete	2	4
	High discrete	0	5
	Decreased	0	14
	Full	0	3
Motor unit configuration	None	30	6
	Nascent	2	17
	Increased polyphasic	0	0
	Disruptive	0	9

Preoperative and postoperative counts of the number of muscles demonstrating each pattern of motor unit recruitment or configuration across all operative patients.

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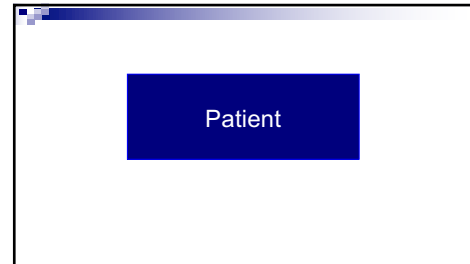


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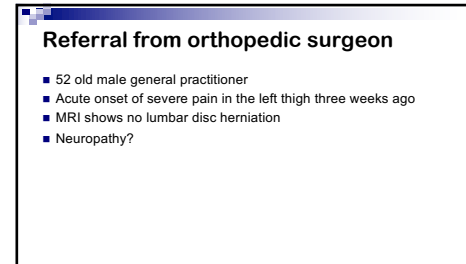
Summary

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

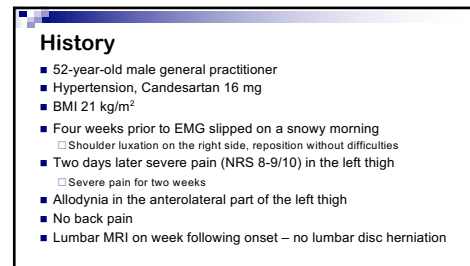
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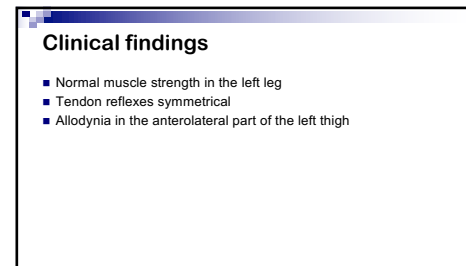
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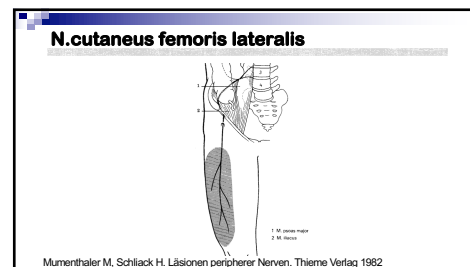
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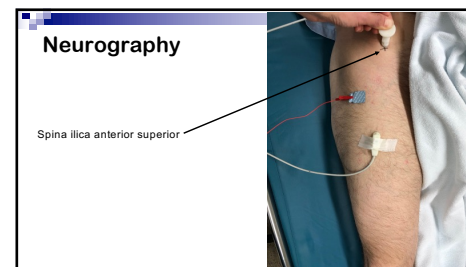
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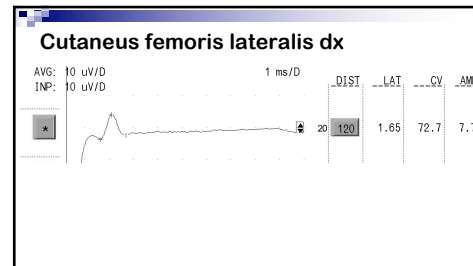
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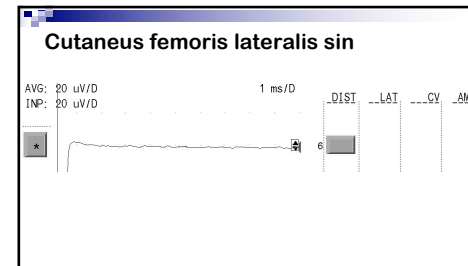
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EMG – normal findings

- m.vastus lateralis
- m.liopsoas
- m.adductor magnus
- m.tensor fascia latae
- m.biceps femoris
- m.gastrocnemius
- m.extensor hallucis longus:

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Neurography

- N.cutaneus femoris lateralis
 - Normal on right side
 - No response on the left side
- Left n.suralis sensory neurography normal
- Left n.peroneus profundus motor neurography normal finding

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Conclusion

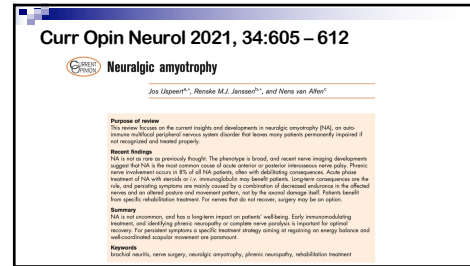
- N.cutaneus femoris lateralis neuropathy on the left side
- Etiology?
 - Acute onset and severe pain not compatible with "meralgia paresthetica"
 - Normal BMI not compatible with "meralgia paresthetica"
- Most likely cause is Parsonage-Turner syndrome
 - Triggered by the shoulder luxation

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Outcome one year later

- Pain subsided completely in 6 weeks
- Slight dysesthesia and hypoesthesia over anterolateral part of the left thigh

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