Salford Royal NHS
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Neurological disorders of the shoulder:- how to recognise them and the role of the neurologist.

Dr David Gow
Consultant Neurologist
Greater Manchester Neuroscience Centre

Neurological disorders that predispose to adhesive capsulitis

Neurological disorders that affect shoulder function

- Myopathy
  - FSH
  - Scapuloperoneal syndromes
  - Absent muscles
  - Immune myopathies
  - Myosin loss myopathy
  - Storage disorders
- Neuromuscular Junction
  - Myasthenia
- Neurogenic
  - Axillary nerve
  - Quadrilateral space syndrome
  - Radiculopathy
  - Brachial plexopathy
  - Motor neuron disease
  - Peripheral neuropathy

How to approach this talk?

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- Some of these disorders are characterised by Pain
  - Brachial plexopathy
    - Neuralgic amyotrophy
  - Radiculopathy
  - Entrapments

- Some of these disorders are characterised by Weakness and or Wasting
  - Anterior horn cell disease
  - Brachial plexopathy
    - Neuralgic amyotrophy
    - Post radiation
  - Myopathy
  - NMJ
  - Radiculopathy
  - Neuropathy

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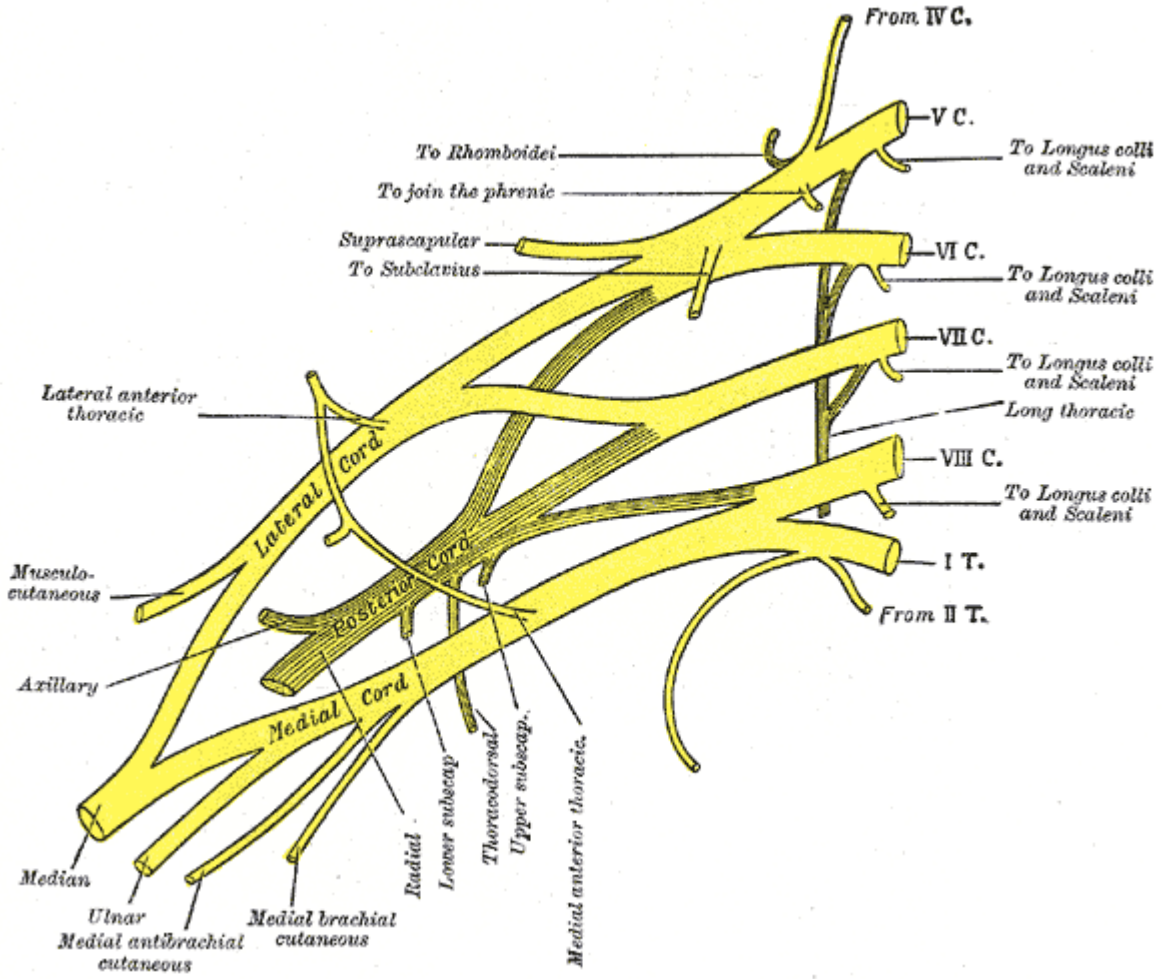

From IV C.
V C.
To Rhomboidei
To Longus colli and Scaleni
To join the phrenic
Suprascapular
VI C.
To Subclavius
To Longus colli and Scaleni
VII C.
To Longus colli and Scaleni
Lateral anterior thoracic
Long thoracic
Lateral Cord
VIII C.
To Longus colli and Scaleni
Posterior Cord
I T.
Musculo-cutaneous
From II T.
Axillary
Medial Cord
Radial
Lower subscap.
Thoracodorsal
Upper subscap.
Medial anterior thoracic
Median
Ulnar
Medial antibrachial cutaneous
Medial brachial cutaneous

The Parsonage Turner syndrome

Neuralgic Amyotrophy

Brachial Neuritis

Neuralgic Amyotrophy

- First described 19th century
- Severe upper limb pain followed by patchy wasting and weakness
- Sporadic or AD inherited trait secondary to mutation in the SEPT9 gene

doi:10.1093/brain/awh722 Brain (2006), 129, 438–450

The clinical spectrum of neuralgic amyotrophy in 246 cases

Nens van Alfen[1,2] and Baziel G. M. van Engelen[1]

[1]Department of Neurology, Neuromuscular Centre Nijmegen and [2]Clinical Neurophysiology, Radboud University Nijmegen Medical Centre, Nijmegen, The Netherlands

Correspondence to: Nens van Alfen, MD, Neuromuscular Centre Nijmegen, Department of Clinical Neurophysiology, 920 Radboud University Nijmegen Medical Centre, PO Box 9101, 6500 HB, Nijmegen, The Netherlands
E-mail: n.vanalfen@neuro.umcn.nl

Predisposing factors

Table 6 Antecedent events

Antecedent event	Percentage	Cases
Infection	43.5	50
Exercise	17.4	20
Surgery	13.9	16
Peripartal*	8.7	10
Vaccination	4.3	5
Stress (psychological)	4.3	5
Trauma	4.3	5
Other†	3.5	4

*6 INA and 4 HNA patients; nine during puerperium, one in third trimester of pregnancy. †Two patients, after sleeping with their arm in an unusual position (PMP22 unknown, but otherwise typical attacks and no signs of pressure palsies on EMG examination), one after CVA, one during bed rest for lumbar HNP.

Pain

- SEVERE
  - Sling sign
- Often starts at night
  - A&E sign
- Usually upper plexus location
- Average duration 40 days males, 20 days females

Weakness

- Proximal > distal
- Commonest infraspinatus and serratus anterior
- Very Variable
- *Anterior interosseous presentation
  - When to explore?

Investigation

- NCS/EMG
  - Abnormal in 96 % in largest series
  - Timing important
- SSEP’s
- CXR
  - Phrenic nerve palsy

Imaging

Clinical spectrum of neuralgic amyotrophy

Brain (2006), 129, 438–450

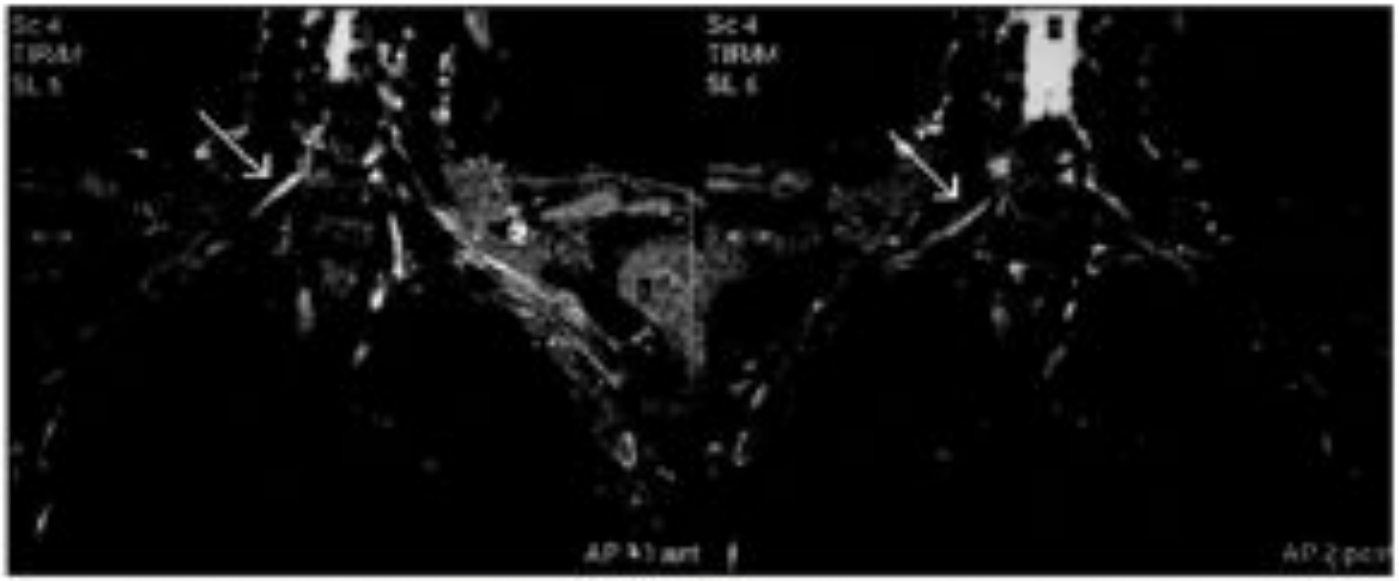


Fig. 1 T_2-weighted non-contrast MRI scan of the brachial plexus, showing a thickened and slightly hyperintense middle trunk on the right.

Attractive but rarely helpful

Treatment

- Corticosteroids and IVIG have been used
- No RCT evidence
- Analgesia
  - Most effective slow release NSAID and slow release Morphine
- Surgical management

Prognosis

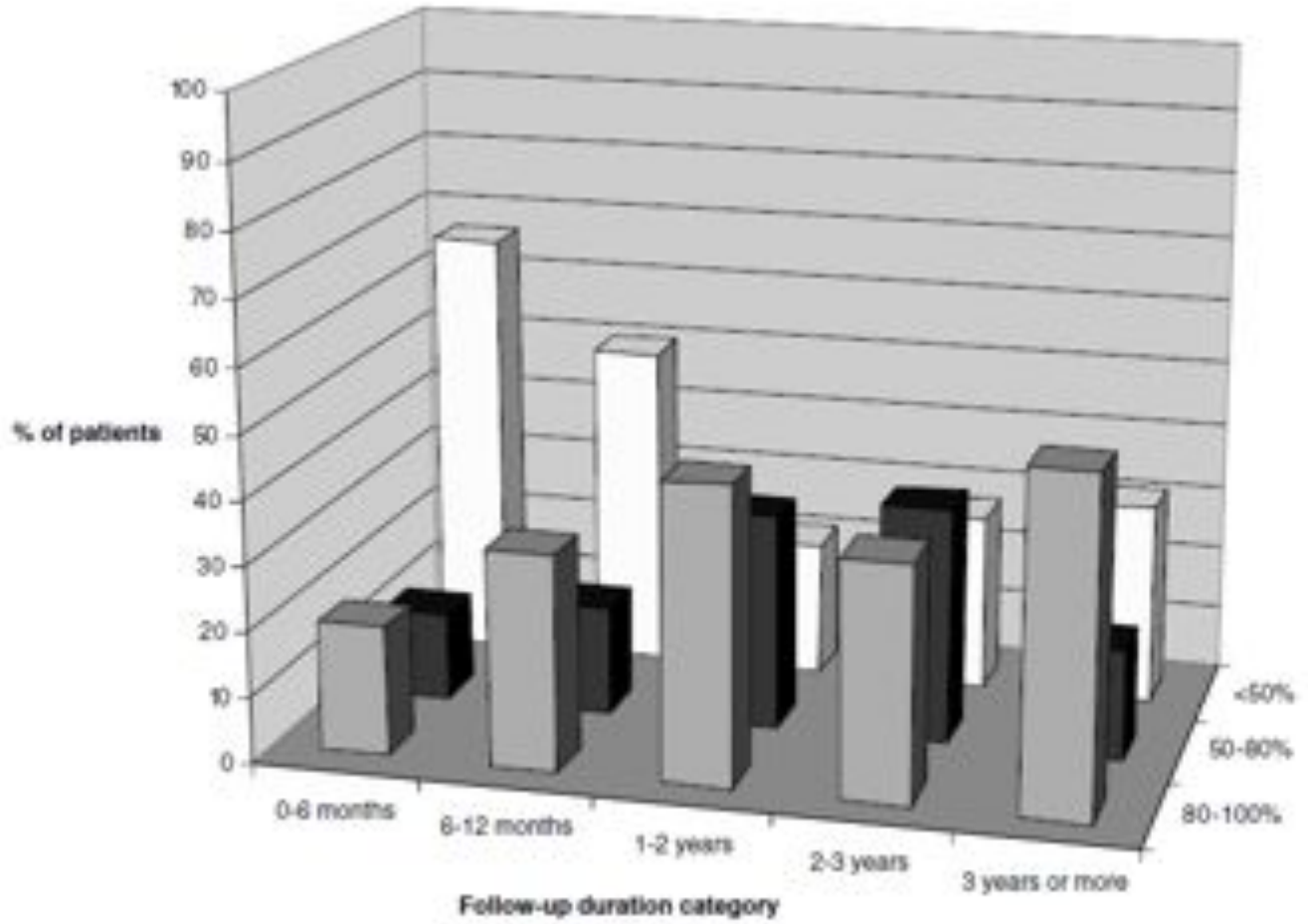


Fig. 4 Estimated subjective overall recovery percentages according to the patients, for each of the five follow-up periods.

Radiation plexopathy

- From the advent of therapeutic radiotherapy it was noted that neural tissue was vulnerable to injury

- Most of the literature pertains to brachial plexopathies and most of this to the treatment of breast malignancy.

Three main syndromes

- Acute
  - Shortly after treatment
  - Ischaemic plexopathy secondary to vascular compromise
  - Painless weakness and sensory loss
  - Prognosis for recovery is poor
  - Rare
- Reversible
  - Mainly sensory syndrome
  - Early
  - Recovery 6-12 months
  - Rare

Delayed Radiation Plexopathy

- Commonest
- Can be relatively acute but often very delayed up to 30 years
- Is this setting major differential is disease recurrence or alternate musculoskeletal disorders
- Pathologically there is extensive vascular obliteration, loss of myelin and fibrosis

Clinical features

- Usually upper trunk sensory pattern
  - Lower trunk more common in infiltrative
- Intrinsic hand weakness common
- Pain occurs in about 50%

Predisposing factors

- Total dose given
  - <50 Gy 1.3%
  - >50 Gy 5.6%
- Simultaneous chemotherapy
- Overlapping fields creating “hot spots”
- Hypo fractionation
  - 6% 45 Gy/15
  - 1% 54 Gy/30

Following factors favour recurrence

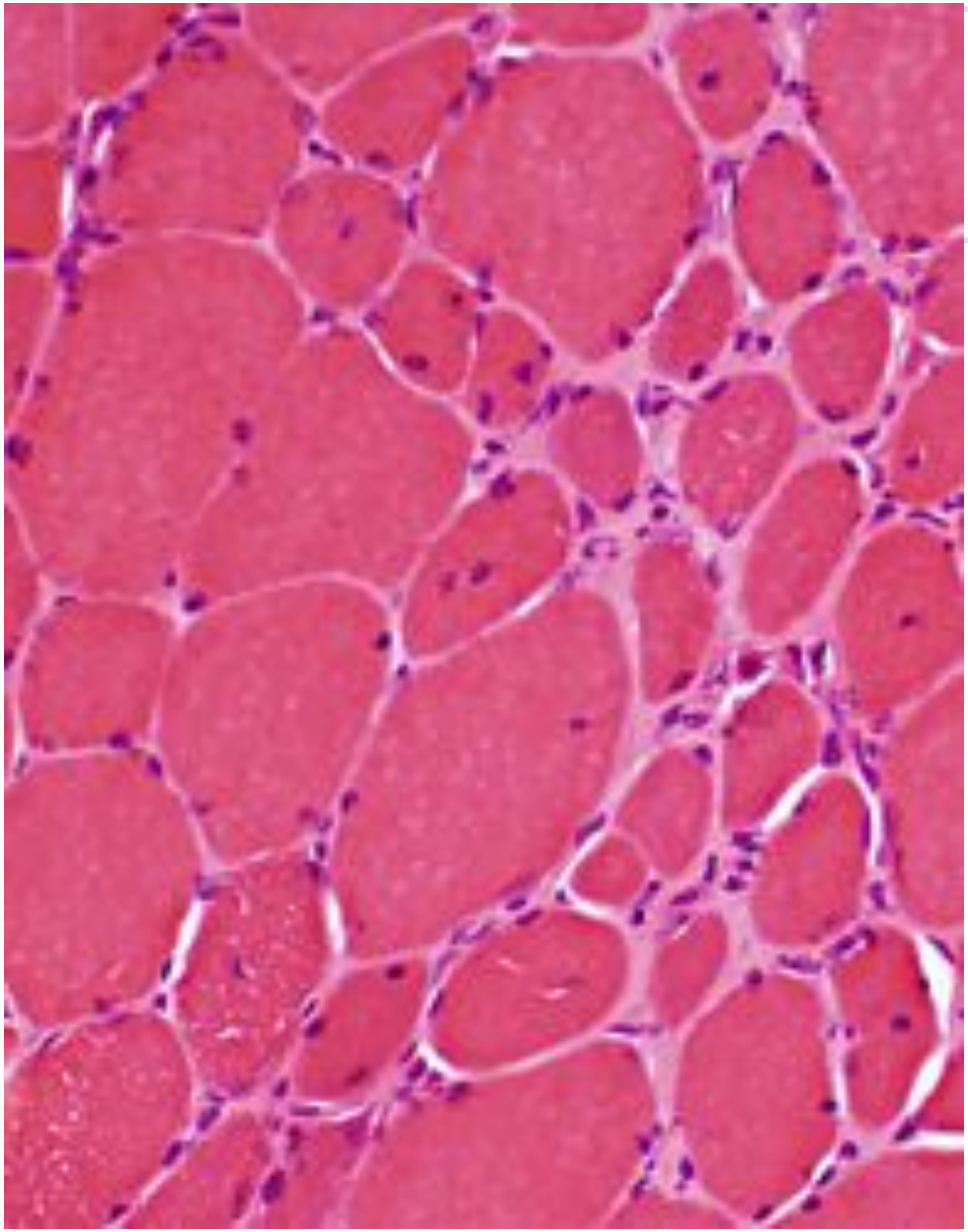
- Horner's syndrome
- Severe pain
- Involvement of the lower brachial plexus (C7, C8, T1)
- Radiation dose <60 Gy

Investigation

- NCS/EMG
  - Excellent at confirming plexopathy
  - Can't reliably differentiate from recurrence
- MRI/PET
- Biopsy

Treatment DRP

- Pentoxifylline and Vitamin E
- Personal experience no demonstrable effect
- Surgical
  - External neurolysis
  - Nerve transfer
  - Tendon transfer
- OT/PT etc

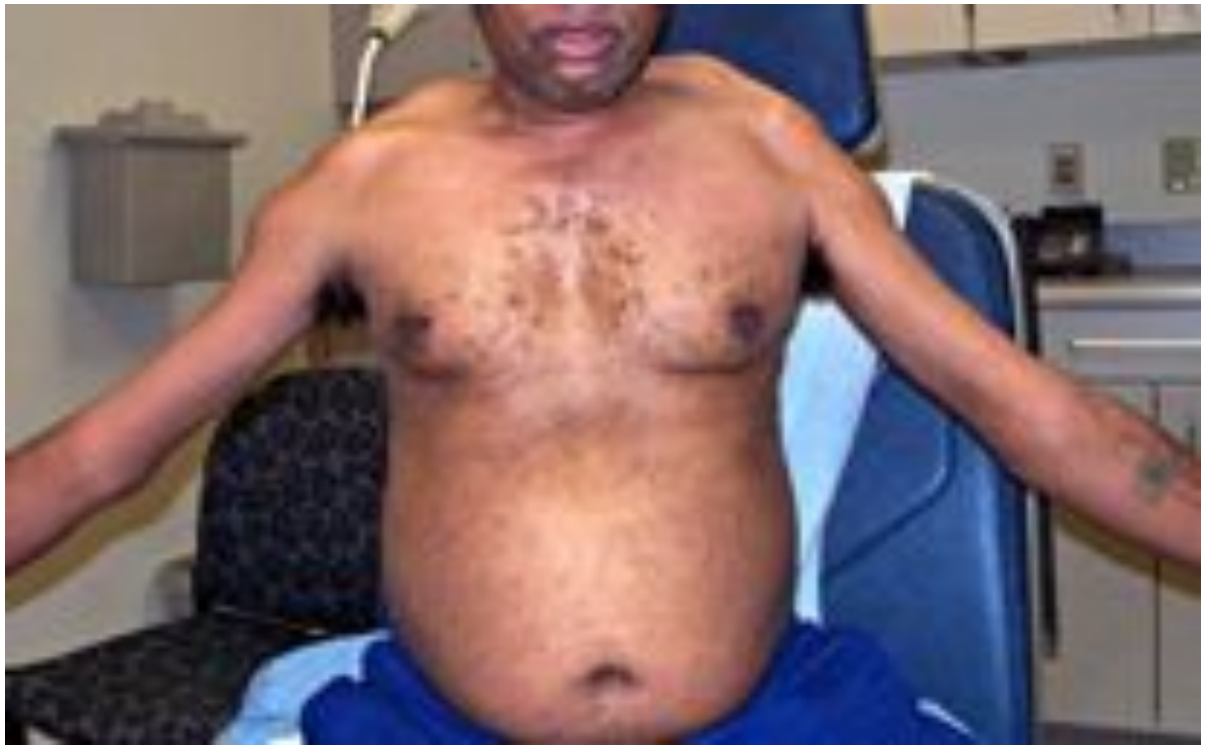


Neuromuscular homepage Brachio-Cervical Inflammatory Myopathy

Myopathies that affect the shoulder

- Myopathy is a vast subject.
- Muscle disease can be acquired or inherited.
- The shoulder girdle can be affected in a number of these conditions.
- Predominantly the problem is weakness.
- Due to this weakness joint disruption and pain are common.

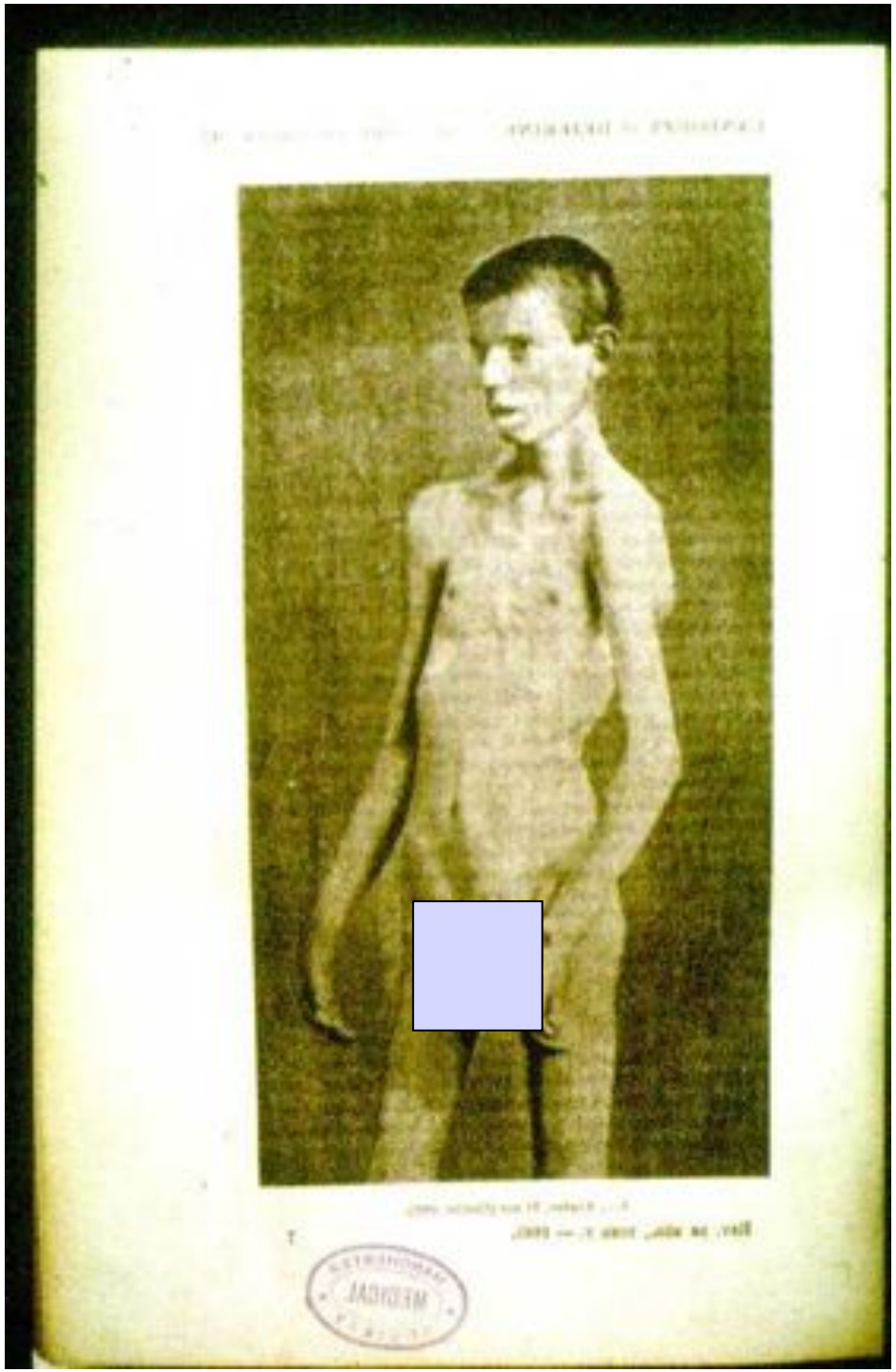
Fascioscapulohumeral dystrophy AD



Neuromuscular homepage

FSHD: Nomenclature

- Weakness and atrophy of facial, shoulder, humeral muscles
- (Erb) Landouzy-Dejerine Syndrome (1884-6 4 gen. family)
- Utah Family (1952 Tyler and Stephens Core features, 1249 descendants of a man who emigrated to Utah in 1840)
- Scapulo-peroneal Syndrome, Atypical “Faceless” FSH



FSHD

1 -5 per 100,000 i.e. 1 in 20,000

Around 3000 cases in UK (MDC),

490, 000 worldwide

Autosomal Dominant

FSHD: Natural History and Genetics

- 1/3 arise de novo, new mutation
- 2/3 patients in known families, most symptomatic by 20-30s, males often symptomatic earlier, females present later or may be asymptomatic
- Progressive
- High penetrance on examination (♂90% by 30y, ♀70%)
- Intergenerational variability common
- No clear anticipation with successive generations
- Infantile and child presentation are rare (severe)

FSHD: Clinical Features

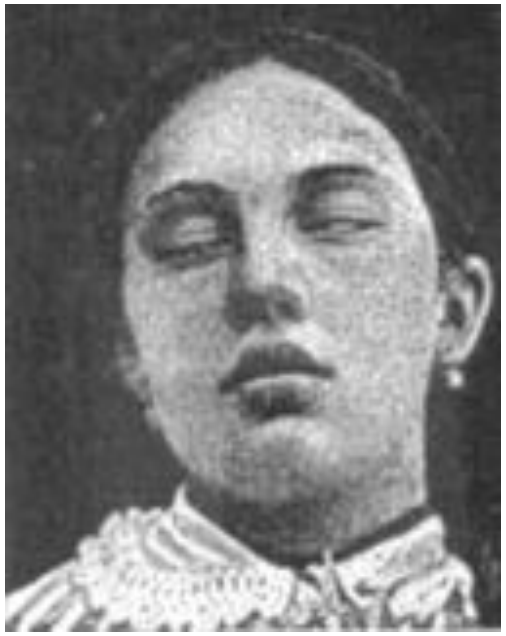
Descending Involvement

Facial

Scapular /humeral

Pelvic /leg weakness

Peri-oral & peri-orbital weakness in FSHD



Neuromuscular Home Page

Symptomatic Facial weakness in >50% in a family (95% at age 30 with examination), often asymmetric.

Lagophthalmos

No ptosis

Poor/inability or idiosyncratic whistling

Smooth forehead

Facial wasting rare

"Bouche de Tapir" Involuntary protrusion of upper lip
Westphal 1886

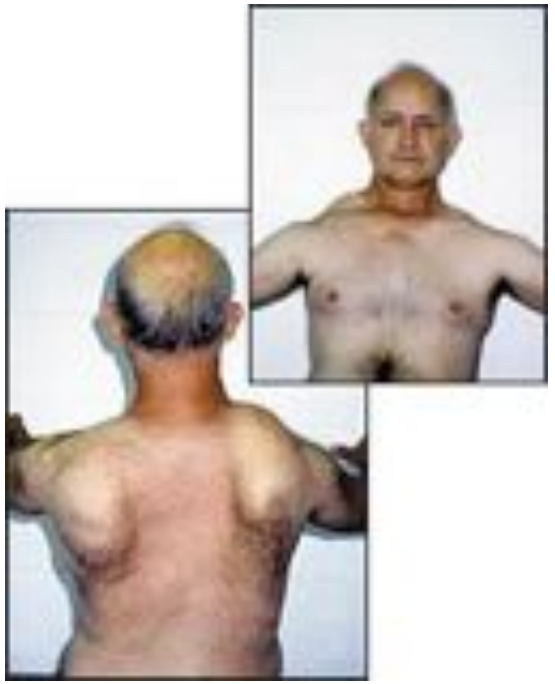
FSHD: Shoulder Involvement

Winging often asymmetric, typically worse on dominant side

Latissimus dorsi; Trapezius; Rhomboids; Serratus anterior

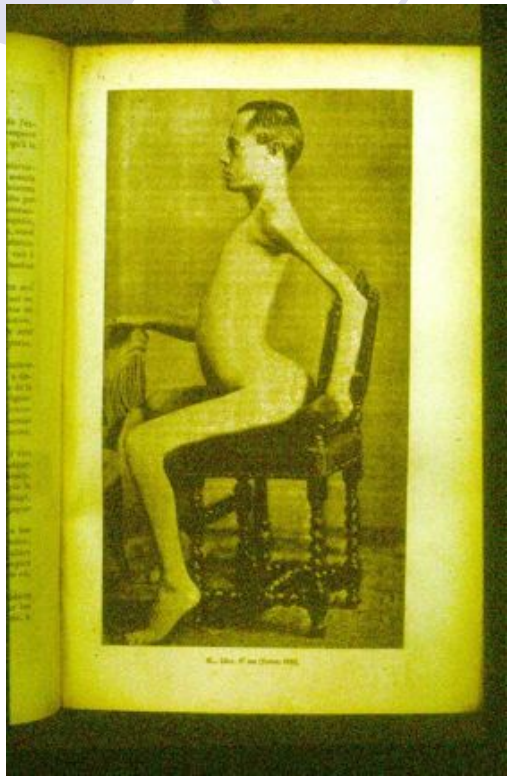
Deltoids often relatively spared

“Deltoid Dip” sign



ISNO Dutch NMD Centre

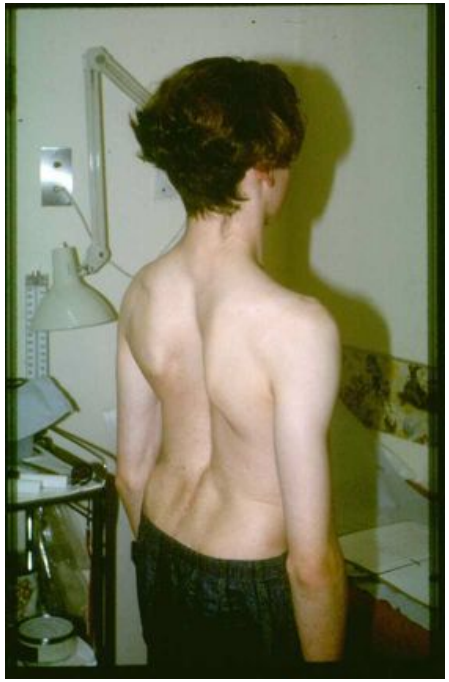
Lower limb involvement

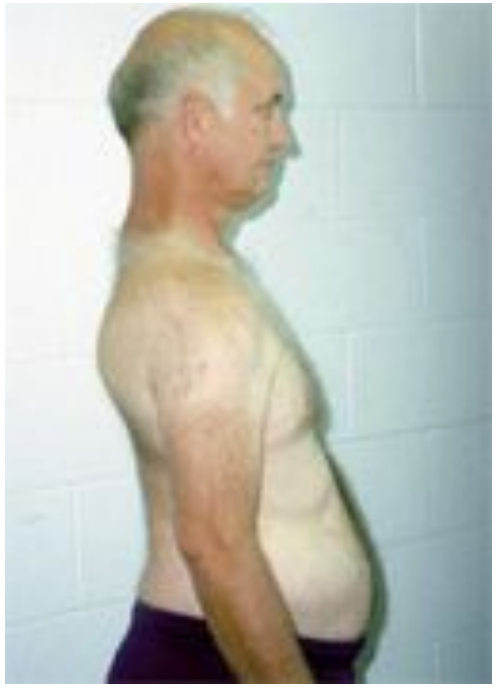


20% require wheelchair by 40s

Ankle>toe DF, rare without facial weakness, males

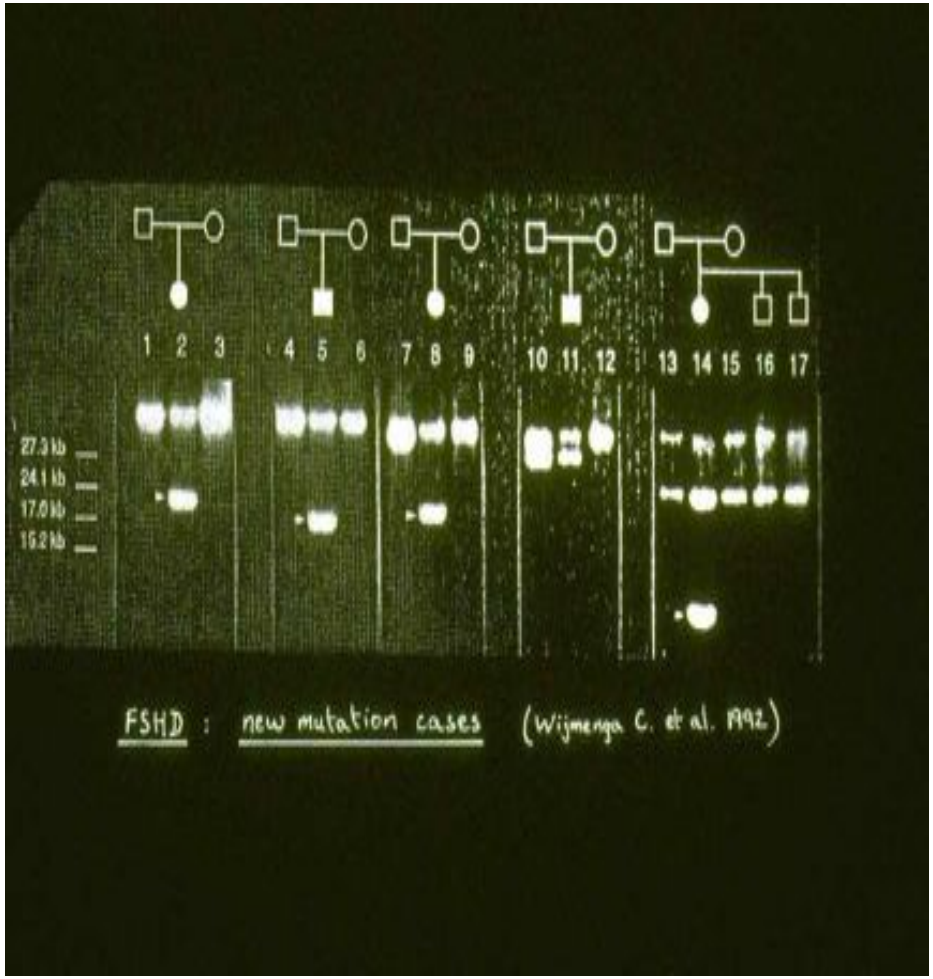
Lordosis is common ..





Bent Spine, and scoliosis are relatively rare (1% require NIV, Wohlgemuth 2004)

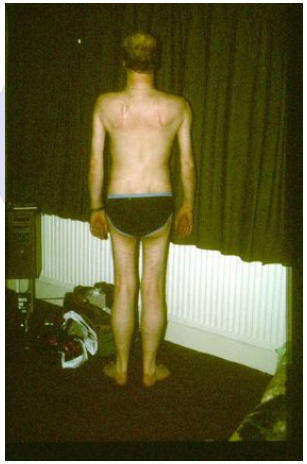
De novo DNA rearrangement at 4q35



Generic management

- Inflammatory disease can be managed with immunosuppression.
- Physiotherapy is very helpful at maximising function and preventing secondary adhesive capsulitis.

FSHD: Treatment



Scapular fixation

“Operative interventions appear to produce significant benefits, though these have to be balanced against postoperative immobilisation, need for physiotherapy and potential complications. We conclude that a randomised trial would be difficult, but a register of cases and the use of a standardised assessment protocol would allow more accurate comparison of the disparate techniques”

Mumery et al (2003) Orrell et al 2010 Cochrane Reviews


YORK

Anterior horn cell disease A.K.A MND

- Not infrequently presents regionally with wasting and weakness of an arm or hand.

- Can present to a number of different specialist
  - Orthopaedics
  - ENT
  - Stroke services

Main sub-categories

- Progressive bulbar palsy
- Progressive muscular atrophy
- Amyotrophic lateral sclerosis
- Primary lateral sclerosis

Major feature

It gets worse

Investigation

- Clinical assessment
- Regional imaging
- NCS/EMG
  - Directed

Management

- Care Centre
- Riluzole slows progression
- Therapy input
  - Shoulder bracing
- NIV


WAKE UP
& smell the
COFFEE

When to involve the neurologist?

- When you are not sure.
- When it is not behaving as you had expected.
- Regional peripheral nerve and myopathy service happy to help.