

# Neuralgic Amyotrophy as a hallmark brachial plexus disorder



Jos IJspeert MSc. Physiotherapist/ clinical health scientist.

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**“Remember: you’ve not finished your job until you’ve taken care of the patient, not just the problem”**

NEJM 2010

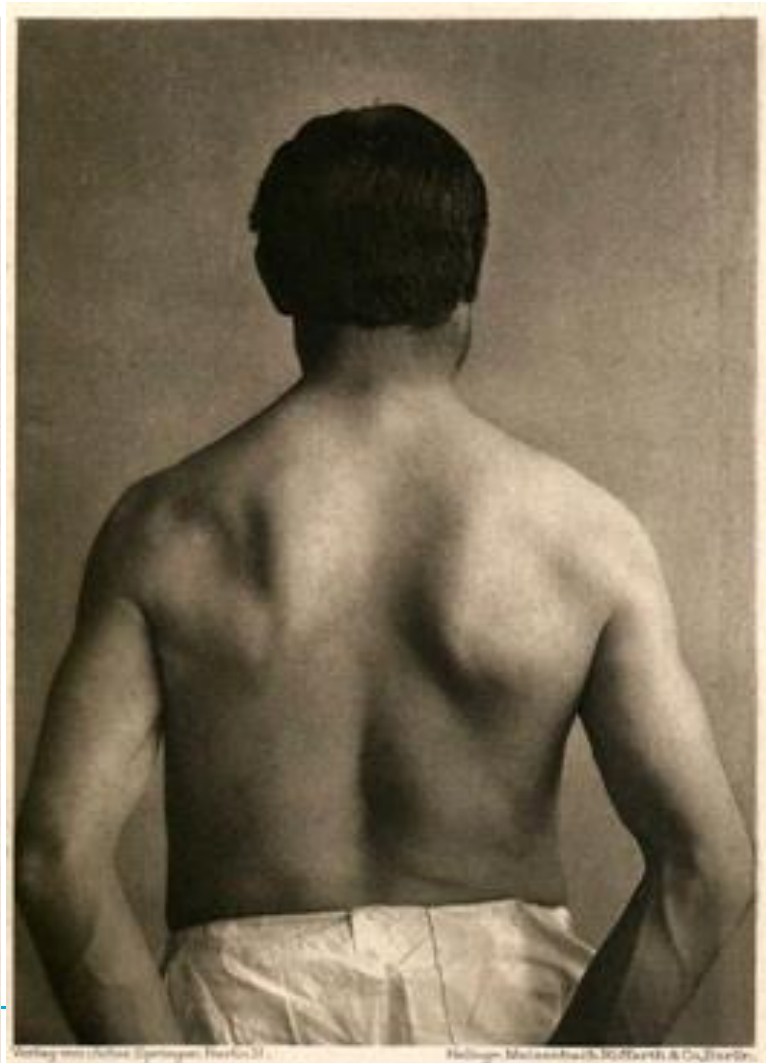
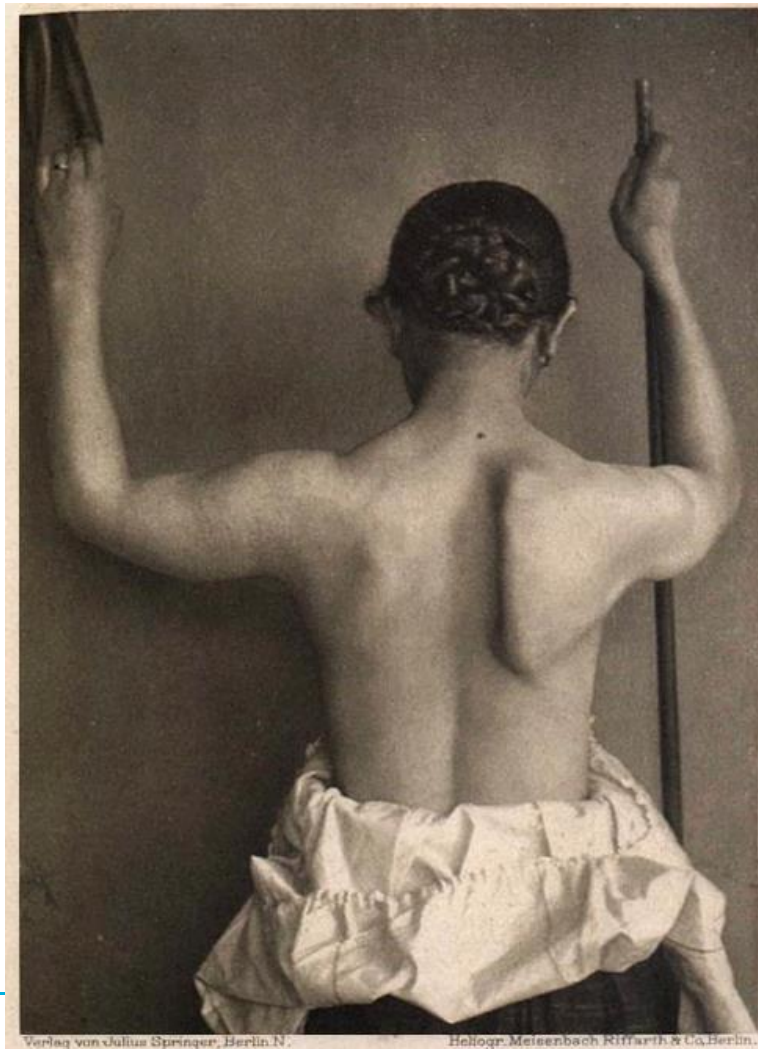
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# What is Neuralgic Amyotrophy?

- NA, also known as..
  - Brachial plexus neuropathy
  - Brachial plexus neuritis
  - Parsonage Turner syndrome
  - .. Another 40 synonyms
- Usually very painful, acute inflammation of the nerves of the brachial plexus and other nerves (e.g. phrenic nerve, lumbosacral plexus, ...)



# NA is NOT a new disorder...



Photos by: H. Churschmann, 1894. [Http://www.artandmedicine.com](http://www.artandmedicine.com)

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# Clinical features

- Episodes or 'attacks'
- Extreme neuropathic pain in neck, shoulder and/or arm in 95%, lasts 3 weeks on average
- Fast multifocal pareses and atrophy of the upper limb, 1/3 bilateral (asymmetrical)
- 80-90% recovery within 6 months - 3 years
- Attack recurrence: 25% (hereditary cases: 75%)
- > 50% of patients have residual complaints; especially pain and decreased physical endurance

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## Typical NA: Scapula alata (winged scapula)



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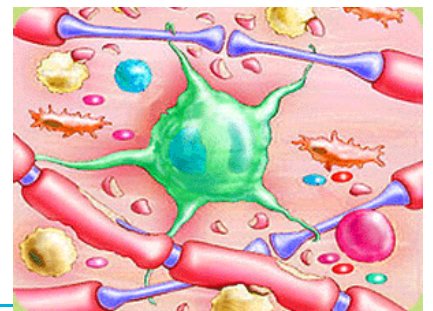
# More clinical features

- Patchy areas of vital hypaesthesia in 80%
- Scapular instability – not always winging- in 70%
- Men > Women (68% vs 32%)
- Painless attacks in 5%
- Lumbosacral involvement in 10%
- Phrenic nerve involvement in 5%

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# Etiology NA

- Exact pathophysiology unknown
- Generally assumed to be auto-immune
- Strong evidence for multifactorial cause:
  - underlying (hereditary) sensitivity
  - mechanical factors 'opening' blood-nerve barrier
  - immune trigger of the attacks



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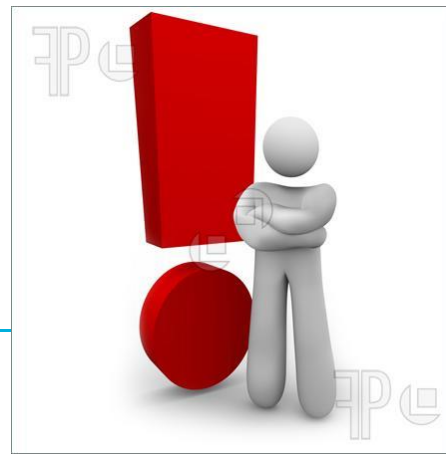
# Incidence of NA: needle in a haystack?



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## Incidence measurement: 1 year in GP practices

- In 2012: all patients with shoulder complaints (incl. NA) from 2 large GP practices (Lent and Oosterhout)
  - Total number of patients within the GP practices : 14118
  - 487 patients with new onset shoulder or arm complaints
  - 16 patients with definitive NA
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- $16 / 14118 \approx 1$  per 1000 per year
  - **100 times more often** than assumed



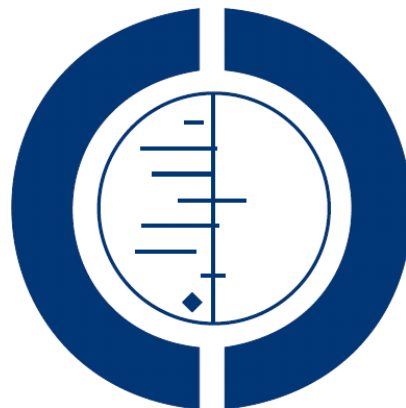
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# Treatment for NA?

- **No** studies were found that could provide **evidence** for a particular form of treatment in neuralgic amyotrophy.

Treatment for idiopathic and hereditary neuralgic  
amyotrophy (brachial neuritis) (Review)

van Alfen N, van Engelen BGM, Hughes RAC



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# Prognosis NA: 1890-1990

- "Good"
- "Nearly all patients recover within 3 years"
- However, clinical experience Radboudumc since 1995:
  - Many residual complaints
  - Many questions regarding therapy
  - Many questions regarding reintegration at work

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# Residual complaints after NA



**Archives of Physical Medicine and Rehabilitation**

journal homepage: [www.archives-pmr.org](http://www.archives-pmr.org)

Archives of Physical Medicine and Rehabilitation 2013;94:67-73



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## ORIGINAL ARTICLE

# Residual Complaints After Neuralgic Amyotrophy

Edith H. Cup, PhD,<sup>a</sup> Jos Ijspeert, BSc,<sup>a</sup> Renske J. Janssen, BSc,<sup>a</sup>  
Chaska Bussemaker-Beumer, MD,<sup>c</sup> Joost Jacobs, MSc,<sup>b</sup> Allan J. Pieterse, PhD,<sup>a</sup>  
Harmen van der Linde, MD, PhD,<sup>a</sup> Nens van Alfen, MD, PhD<sup>b</sup>

*From the <sup>a</sup>Department of Rehabilitation, Nijmegen Center for Evidence Based Practice and the <sup>b</sup>Department of Neurology, Donders Center for Neuroscience, Radboud University Nijmegen Medical Center, Nijmegen; and <sup>c</sup>Rehabilitation Center 'De Trappenberg,' Huizen, The Netherlands.*

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# Residual complaints after NA

- Cohort study 248 NA patients
- Follow-up: 6 months -3 years
- 50% still limited by pain
- 80% still difficulty reaching and overhead activities
- 63% still severe fatigue



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# Residual complaints: just “chronic pain?”

ORIGINAL ARTICLE

## **Long-Term Pain, Fatigue, and Impairment in Neuralgic Amyotrophy**

*Nens van Alfen, MD, PhD, Sieberen P. van der Werf, MSc, PhD, Baziel G. van Engelen, MD, PhD*

Arch Phys Med Rehabil 2009;90:435-9.

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# Residual complaints: just “chronic pain?”

- Cross sectional cohort study 89 patients
- McGill (pain), CIS (fatigue), SCL-90 (psychologic distress), RAND SF-36 (QoL)
- 1/3 – 1/4 had significant long-term pain and fatigue
- 1/2 - 2/3 still had impairments in daily life
- > 1/3 suffered from severe fatigue
- No correlation of pain or fatigue with level of residual paresis
- The group did not meet the criteria of chronic fatigue or major psychologic distress

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# Residual complaints after NA

- High correlation between scapula instability, pain and fatigue
- Standard physical therapy\* was ineffective or even aggravated symptoms in 50% (!)

*\*Mostly resistance training of rotator cuff muscles and massage*



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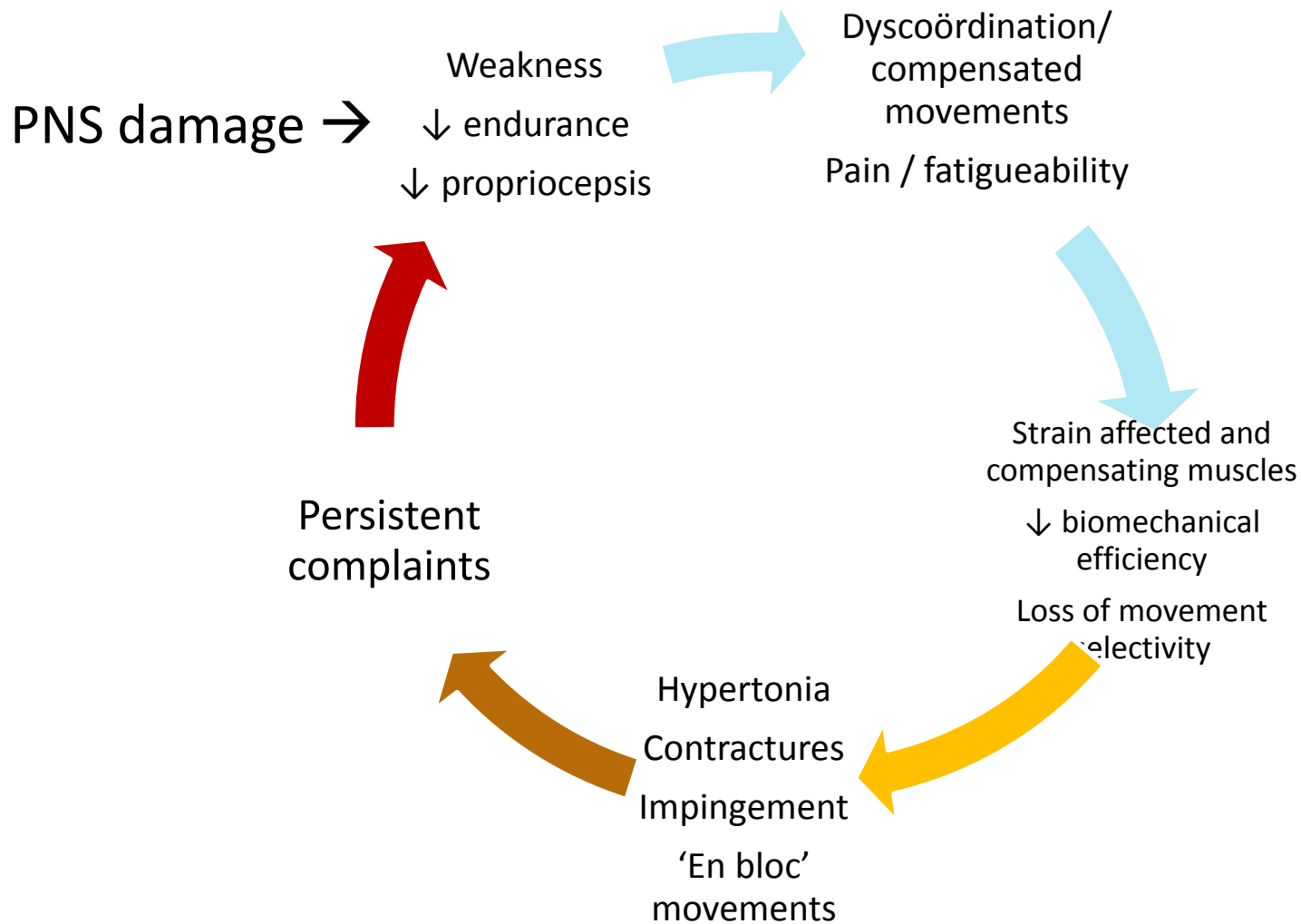
**“My doctor told me it’s NA and I need to learn to live with it.”**

**“But how?”**

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# Persistent pain after shoulder nerve damage

- Most patients experience significant but not complete “hardware” recovery because of decreased endurance
- Serratus paresis leads to automatic compensatory movements which are less efficient (cost more energy)
- Decreased shoulder coordination leads to an increased risk for glenohumeral / rotator cuff pathology
- Recovery takes a long time, most patients resume normal activity level (100%) while they are still only capable of less



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# Therapeutic approach

- Passive glenohumeral mobilisation
- Strength training rotator cuff muscles
- Massage therapy or manual therapy for hypertonic cervical spine muscles

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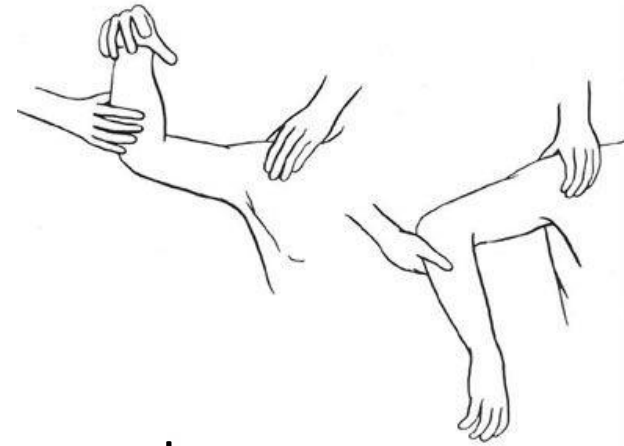




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# Physical therapy and rehabilitation

- 'Standard' approach often no or aggravating effect
- Strength training when muscle < MRC 3 not usefull
- Duration of "after-pain" should not > duration activity
- Needed is a combination of:
  - Information for patient and therapist
  - Optimising biomechanics
  - Relaxation and improvig physical shape
  - Ergonomics
  - Balance in physical possibilities versus demand



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# The reasons for a multifactorial approach

## Complex disability and complaints

Patients do not recover as well as they are supposed to do.

After 6 months to 3 years:

- Pain (53% during ADL)<sup>1</sup>
- Fatigue (63% CIS fatigue >35) <sup>1</sup>
- Paresis (60%) <sup>1</sup>

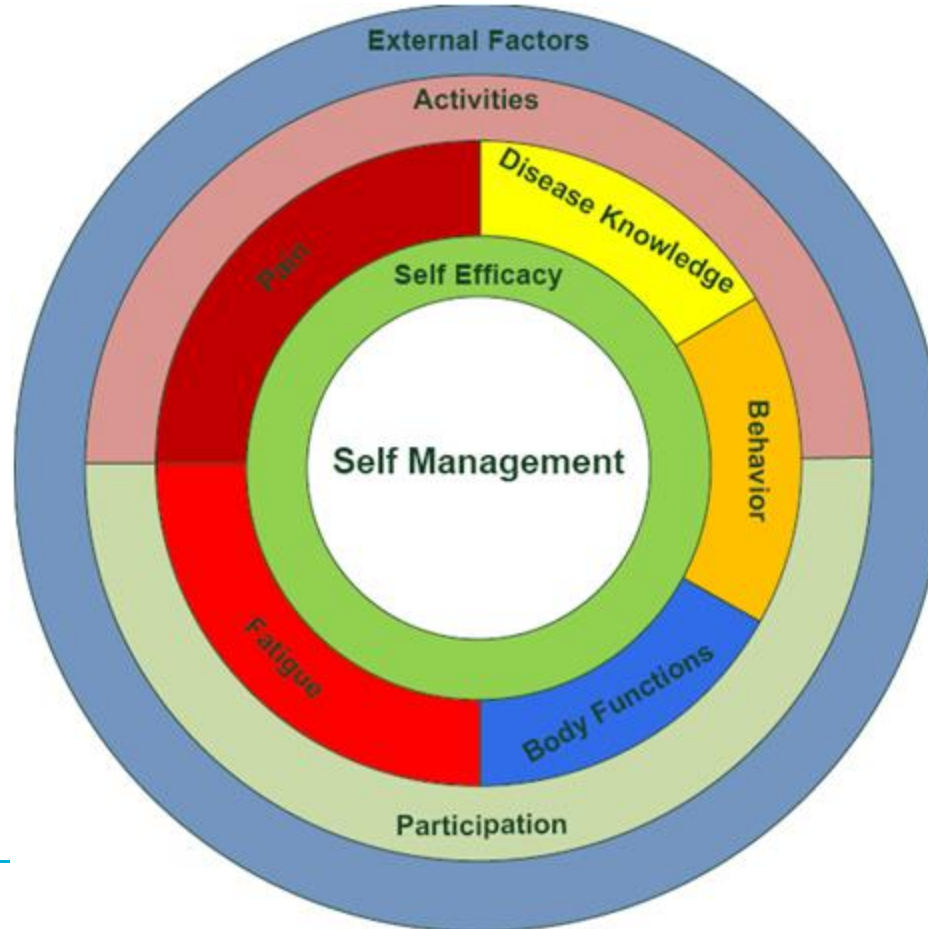
This leads to problems in

- ADL
- Work
- Sports
- Participation in general

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1: Cup EH, Ijspeert J, Janssen RJ, Bussemaker-Beumer C, Jacobs J, Pieterse AJ, et al. Residual Complaints After Neuralgic Amyotrophy. Arch Phys Med Rehabil. 2012. Epub 2012/08/02.

# A multifactorial approach



1: IJspeert J, Janssen RM, Murgia A, Pisters MF, Cup EH, Groothuis JT, et al. Efficacy of a combined physical and occupational therapy intervention in patients with subacute neuralgic amyotrophy: A pilot study. *NeuroRehabilitation*. 2013.

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# What do our patients look like?

1; Neuralgic amyotrophy >1 year



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# What do our patients look like?

2; Traumatic plexopathy >6 years

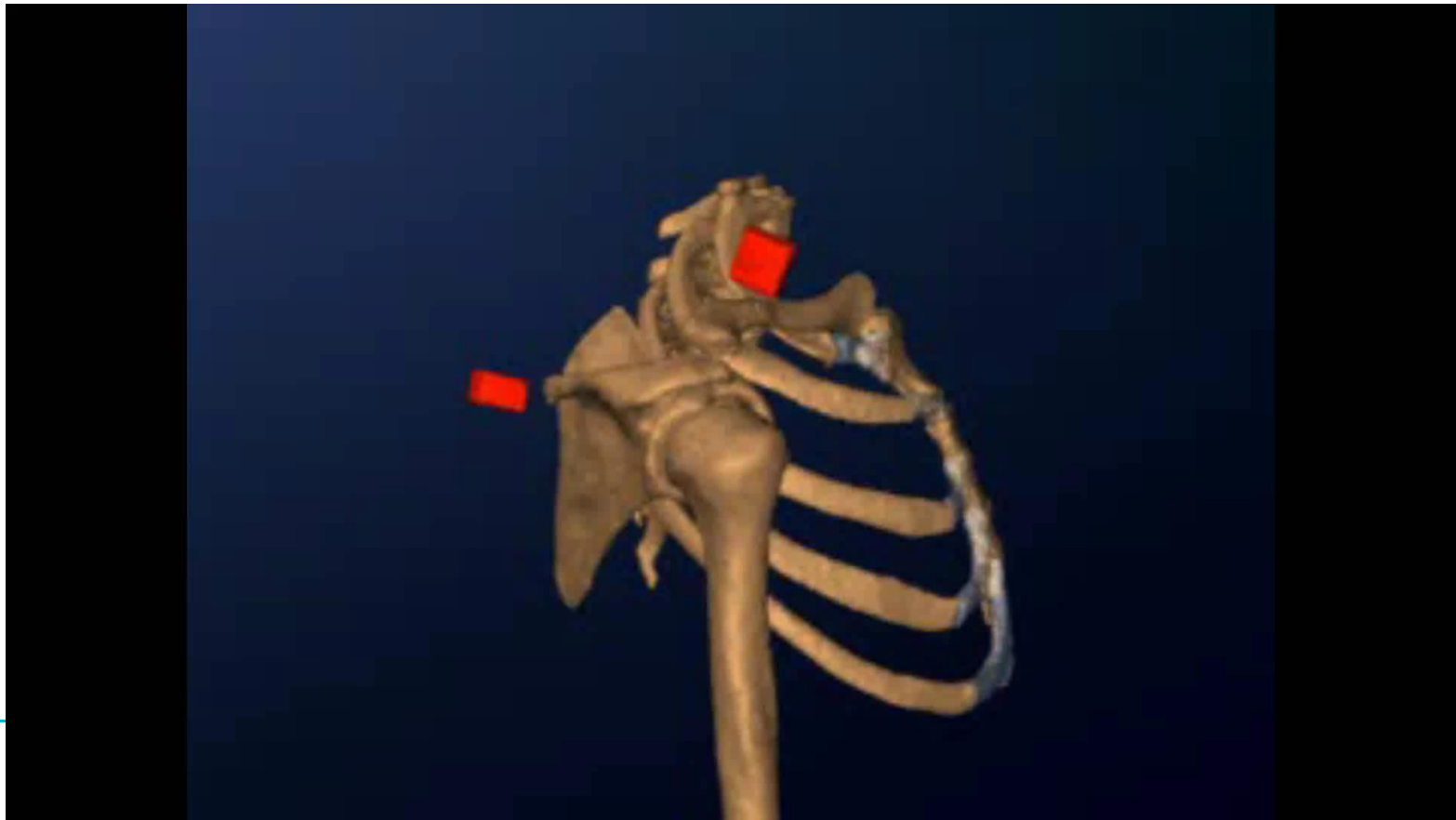


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# Normal scapular movement



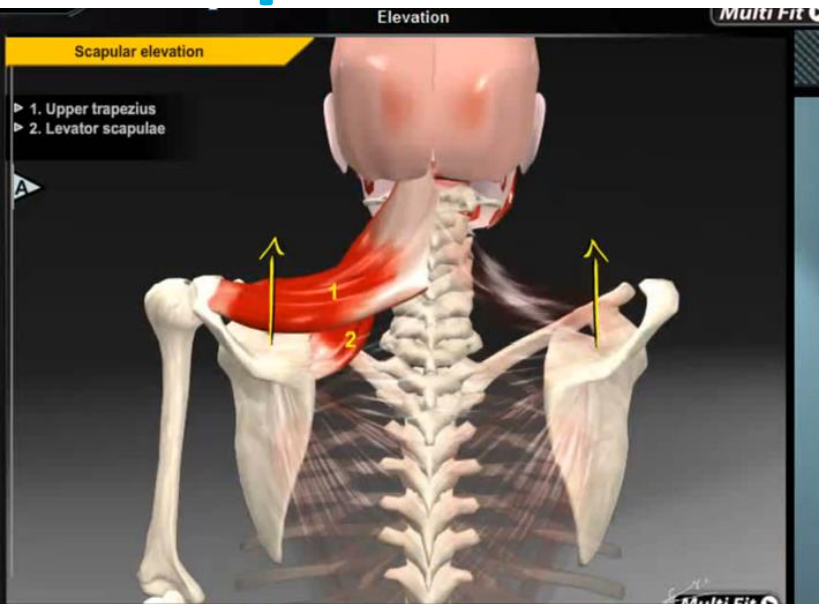
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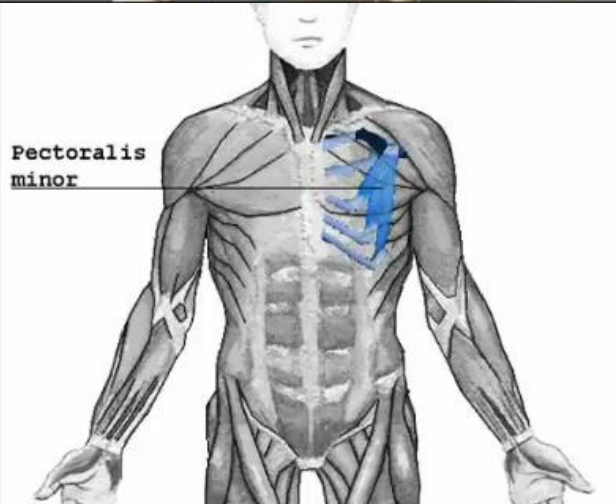
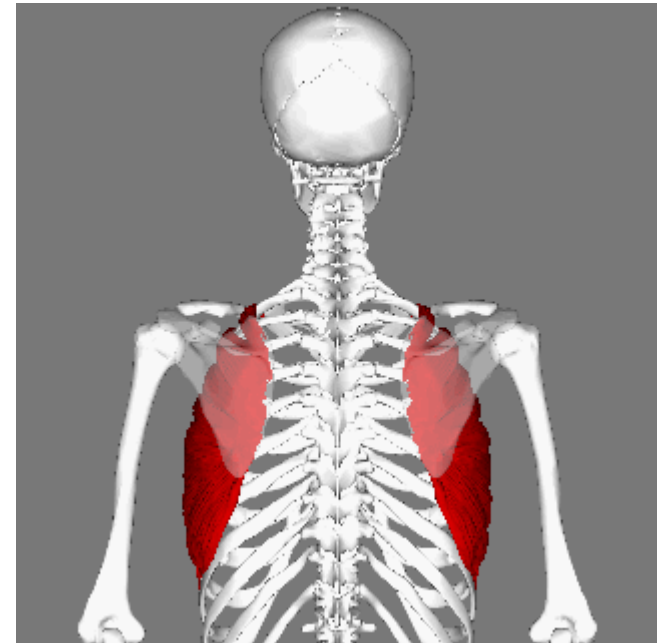
# Compensations

# Versus

# Disuse



# Versus



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# Misperceptions of scapular dyskinesia

Scapula alata  $\neq$  Serratus weakness

Low MRC scores  $\neq$  low muscle strength

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# Therapy approach

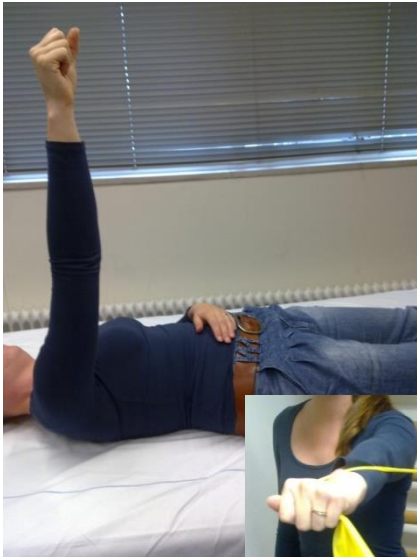
- Reduce pain
- Teach energy conservation strategies (mainly OT)
- Adapt environment (mainly OT)
- Behavioral change (OT+ PT)
- Training
  - Scapular kinesiia
  - Cervical stability
  - Muscle lenght
  - Neural mobility plexus

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# Therapy program

- Short outpatient treatment program
- “Home-exercise” for patients (implementing therapy in daily living)
- Treatment schedule

# Exercise



# Implementation



NO PAIN = GAIN



# Implementation



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# Result 1



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## Result 2



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# Occupational Therapy

*Aim: For the patient to achieve control over*

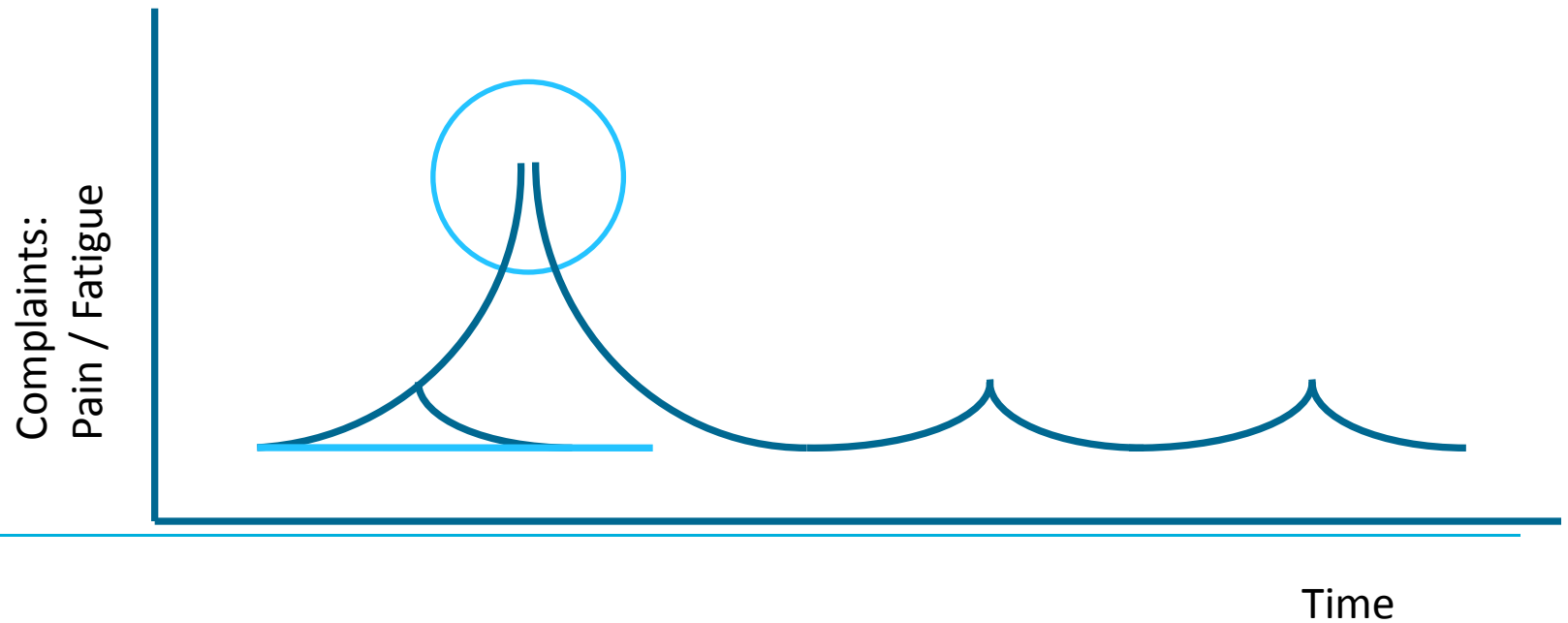
- *their complaints*
- *in their live*

- Disease specific information
  - The importance of good ergonomics
  - Practical situations / activity analysis
  - Balanced distribution between activities and relaxation
    - Overuse, underuse
    - “Activiteitenweger”
    - Stages of change; behavioral changes
- 
- Motivational Interviewing

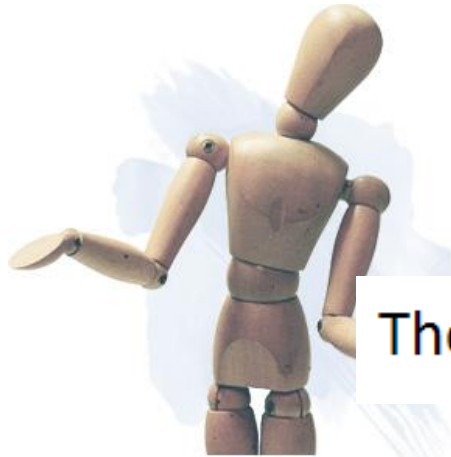
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# Theory

Effect on functioning



# The importance of good ergonomics



The importance of relaxation of the muscles!



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# Posture of the body

Ergonomics within activities

It is about **the way** you perform activities

Implement physical therapists' advices into daily activities

Main problems:

- Activities above shoulder height
- Reaching
- Repetitive movements
- (Heavy) lifting
- Static, strenuous posture

# Practical situations

Performance of activities: How??

- Posture
- Use / disuse of the affected arm(s)
- Movement pattern
- Compensation



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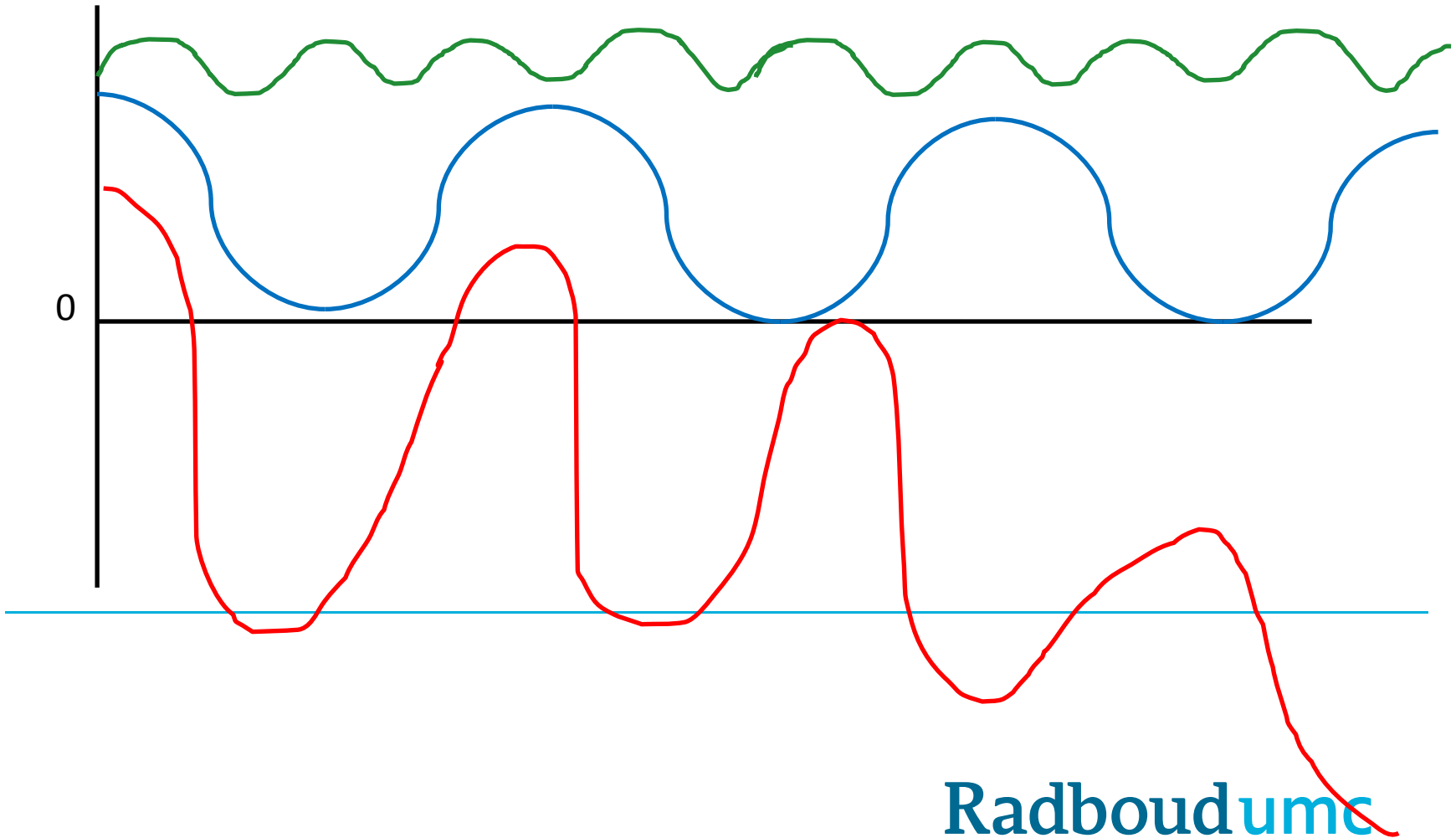
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# Overuse / Onderuse

## Fatigue

- Most energy of the body is used to 'repair' the problems in the nerves
- Affected muscles receive no/less information from the nerves
  - strain on the affected muscles
- Non-affected muscles are (over)compensating
  - strain on the non-affected muscles
- Pain is exhausting
- Overburdening: most energy that is available, is already used

# Energy distribution



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# Stages of change; behavioral changes

| Stage            | How to coach your patiënt to the next stage?  |
|------------------|---|
| Precontemplation | <ul style="list-style-type: none"><li>- Increase awareness of own behavior</li><li>- Increases feeling about risks and problems of current behavior</li></ul> |
| Contemplation    | Explore ambivalence: get behind reasons to change or not to change  |
| Preparation      | <ul style="list-style-type: none"><li>- Help patient determine the best strategy to achieve change</li><li>- Increase confidence</li></ul>                    |
| Action           | <ul style="list-style-type: none"><li>- Help patient to take the first steps to implement their strategy</li></ul>  |
| Maintenance      | <ul style="list-style-type: none"><li>- Create insight into relapse</li><li>- Make sure patient knows what to do when there is a relapse</li></ul>            |
| Relapse          | <ul style="list-style-type: none"><li>- Help patient to get through this relapse</li></ul>  |

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# Motivational interviewing

## Conversation techniques

- Suppress the instinct to repair/give unasked advice
- Invite patient to talk about change (possibilities, needs, reasons etc.)

| Do's  | Don'ts   |
|---|--|
| Ask permission to give information                | Give an order  |
| Support patient: give compliments                 | Try to convince the patient to change                          |
| Support patient: to discover their own strategies | Give suggestions or advice (only when the patient asks for it) |