Neuralgic Amyotrophy as a hallmark brachial plexus disorder

Jos IJspeert MSc. Physiotherapist/clinical health scientist.
“Remember: you’ve not finished your job until you’ve taken care of the patient, not just the problem”
What is Neuralgic Amyotropy?

- 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...

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
Typical NA: Scapula alata (winged scapula)
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%
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
Incidence of NA: needle in a haystack?
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

- $\frac{16}{14118} \approx 1 \text{ per } 1000 \text{ per year}$
- **100 times more often** than assumed
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
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
Residual complaints after NA

Original Article

Residual Complaints After Neuralgic Amyotrophy

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From the aDepartment of Rehabilitation, Nijmegen Center for Evidence Based Practice and the bDepartment of Neurology, Donders Center for Neuroscience, Radboud University Nijmegen Medical Center, Nijmegen; and ‘Rehabilitation Center ‘De Trappenberg,’ Huizen, The Netherlands.
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
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

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

“But how?”
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.
PNS damage →

- Weakness
- ↓ endurance
- ↓ proprioception

Dyscoördination/compensated movements

- Pain / fatigueability

Strain affected and compensating muscles

- ↓ biomechanical efficiency
- Loss of movement selectivity

Hypertonia
Contractures
Impingement

‘En bloc’ movements

Persistent complaints
Therapeutic approach

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

- ‘Standard’ approach often no or aggravating effect
- Strength training when muscle < MRC 3 not useful
- Duration of “after-pain” should not > duration activity
- Needed is a combination of:
  - Information for patient and therapist
  - Optimising biomechanics
  - Relaxation and improving physical shape
  - Ergonomics
  - Balance in physical possibilities versus demand
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)¹
- Fatigue (63% CIS fatigue >35) ¹
- Paresis (60%) ¹

This leads to problems in

- ADL
- Work
- Sports
- Participation in general

A multifactorial approach

What do our patients look like?

1; Neuralgic amyotrophy >1 year
What do our patients look like?

2; Traumatic plexopathy >6 years
Normal scapular movement
Compensations Versus Disuse

Radboudumc
Misperceptions of scapular dyskinesia

Scapula alata ≠ Serratus weakness
Low MRC scores ≠ low muscle strength
Therapy approach

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

- Short outpatient treatment program
- "Home-exercise" for patients (implementing therapy in daily living)
- Treatment schedule
Exercise
Implementation

NO PAIN = GAIN
Implementation
Result 1
Result 2
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
Theory

Effect on functioning
The importance of good ergonomics

The importance of relaxation of the muscles!

Radboud UMC
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
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
# Stages of change; behavioral changes

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<tr>
<th>Stage</th>
<th>How to coach your patiënt to the next stage?</th>
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| Precontemplation | - Increase awareness of own behavior  
                    - Increases feeling about risks and problems of current behavior                                    |
| Contemplation    | Explore ambivalence: get behind reasons to change or not to change                                         |
| Preparation      | - Help patient determine the best strategy to achieve change  
                    - Increase confidence                                                                                           |
| Action           | - Help patient to take the first steps to implement their strategy                                         |
| Maintenance      | - Create insight into relapse  
                    - Make sure patient knows what to do when there is a relapse                                                      |
| Relapse          | - Help patient to get through this relapse                                                                  |
Motivational interviewing

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

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<th>Do’s</th>
<th>Don’ts</th>
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<tr>
<td>Ask permission to give information</td>
<td>Give an order</td>
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<tr>
<td>Support patient: give compliments</td>
<td>Try to convince the patient to change</td>
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<tr>
<td>Support patient: to discover their own strategies</td>
<td>Give suggestions or advice (only when the patient asks for it)</td>
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