

# EFNS Guidelines on Post-polio syndrome

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# EFNS: European Federation of Neurological Societies

- Aim of developing, publishing and disseminating guidelines:
  - Improve neurological practice
- Task forces established
  - Mix of experts from different countries, patients representative, optional external experts



# Task force on PPS

- Established 2004
- First author E. Farbu
- Discussions and consensus by e-mail
- Extensive literature search
- Evaluation of scientific evidence
- Other members:
  - N.E. Gilhus
  - M.P. Barnes
  - K.Borg
  - M. de Visser
  - A. Driessen
  - R. Howard
  - F. Nollet
  - J. Opara
  - E. Stalberg

# Scientific evidence\*

- **Evidence classification**

- **Class I:** Well-designed RCTs
- **Class II:** Matched group-cohort study or less well designed RCTs
- **Class III:** Controlled studies, outcome assessment independent of treatment
- **Class IV:** Uncontrolled studies, case series, expert opinions

- **Rating recommendation**

- **Level A:** Class I or at least two Class II-studies
- **Level B:** Class II or overwhelming Class III
- **Level C:** Two Class III

\*Brainin et al. 2001

# What is PPS?

- Clinically described in 1875
- Recognised during the 1980's
- Halstead, Dalakas, Borg
- March of Dimes:
  - Prior paralytic polio
  - Period of partial or complete recovery
  - Onset of progressive and persistent muscle weakness
  - Symptoms persist for one year
  - Exclusion of other causes

# First version

- National surveys
- Literature search:
  - Medline via PubMed, EMBASE, ISI, Cochrane library: 1966-2004
  - Key words: post-poliosyndrome/  
postpoliomyelitis/PPMA/PPMD/poliomyelitis  
*and* management, therapy, treatment,  
medicaments, physiotherapy, intervention

# Role of clinical neurophysiology

- Establish previous polio affection
  - Neurogenic EMG-findings, normal sensory findings
- Exclude differential diagnoses
- Exclude concomitant findings
  - Radiculopathies, peripheral nerve entrapments
- Assess degree of motor neuron loss

# Conclusions 2006:

- Level A:
  - No effect of pyridostigmine, steroids, amantadine
- Level B:
  - Supervised muscular training is beneficial
    - Prevent further decline, reduce symptoms
- Level C:
  - Recognition of respiratory failure
  - Group training, regular follow-ups
    - Useful for well-being
- GPP:
  - Weight loss, assistive devices

# Revision 2010

- No new national surveys
- Same authors
- Literature search 2004-2009
  
- No major changes, but studies on IVIG have emerged

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## CHAPTER 18

# Post-polio syndrome

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### Objectives

The aim was to revise the existing EFNS task force document, with regard to a common definition of PPS, and evaluation of the existing evidence for the effectiveness and safety of therapeutic interventions. By this revision, clinical guidelines for management of PPS are provided.

### Background

Many previous polio patients experience new muscle weakness, fatigue, myalgia and joint pain, and cold intolerance, and develop new atrophy several years after acute paralytic poliomyelitis. The first case of new atrophy and

1 Prior paralytic poliomyelitis with evidence of motor neuron loss, as confirmed by history of the acute paralytic illness, signs of residual weakness, and atrophy of muscles on neurological examination, and signs of denervation on electromyography (EMG).

2 A period of partial or complete functional recovery after acute paralytic poliomyelitis, followed by an interval (usually 15 years or more) of stable neurologic function.

3 Gradual or sudden onset of progressive and persistent muscle weakness or abnormal muscle fatigability (decreased endurance), with or without generalized fatigue, muscle atrophy, or muscle and joint pain. (Sudden onset may follow a period of inactivity, or trauma, or surgery.) Less commonly, symptoms attrib-

# Physical exercise and muscular training



- Basis of PPS-care
- Relieves symptoms
- Probably safe, but long-term studies and studies on severe weakened muscles are lacking
- Water training particularly useful

# Bulbar symptoms

- Spine deformities increases risk of nocturnal hypoventilation – NIV is useful
- Speech therapist
- General precautions such as no smoking, influenza- and pneumococcus vaccine



# Medications

- No effects of:
  - Steroids
  - Amantadine
  - Pyridostigmine
  - Coenzyme Q10
- Lamotrigine needs further studies
  - Possible beneficial effect on QoL



- And IVIG?

# IVIIG in PPS

- Rationale: inflammatory changes in the CSF
- Two randomized and one open study were reviewed
- Gonzales et al:
  - Effect on muscular power
- Farbu et al:
  - Effect on pain
- Kaponides et al:
  - Effect on vitality

*Task force: diverging results and treatment regimens, although a modest beneficial effect is possible, IVIG is not recommended as standard treatment*

# Summary

- The EFNS Task force has implemented PPS as a neurological disorder
- Today's knowledge has been evaluated
  - Some particular medications are not useful
- RCTs for interventions in PPS are difficult to run
  - None of the authors have reported conflicts of interests