



POST POLIO SYNDROME – a challenge of today

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Post Polio Syndrome overview of current knowledge

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Content

Post Polio Syndrome

Cause

Therapy

Care in Europe

Aims of the Conference

Content

Post Polio Syndrome

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A typical story

- **Polio**
1961, 5 yrs old, legs
Residual paresis left leg
- **Functioning in stable period**
Walks with a limb, normal shoes
No activity limitations
Works full time, stands all day
Uses public transportation



33 years after polio

- **1994 - Post Polio Syndrome**
New muscle weakness and increased fatiguability left thigh
- **Functioning**
Walking difficulty affects his functioning with respect to work and mobility
- **Adaptation**
Gets an orthosis and later on a stick



50 years after polio, 17 years after diagnosis PPS

- **2011 – Further decline**

Right thigh becomes weaker

- **Functioning**

Needs personal help to walk outside, afraid of falling

Avoided consultation, afraid of consequences

- **Adaptations**

Changes housing, mobility aids, considers selling the company Responds emotionally

Late neuromuscular symptoms after polio

- Became recognized in the mid-nineteen eighties when survivors in the US of the large epidemics during the 1940's and 1950's became older and started to complain about:
- New weakness, fatigue, muscle and joint pain, atrophy, cold intolerance, sometimes breathing and swallowing difficulties, functional decline



POST POLIO SYNDROME

- Affects about 40-60% of polio survivors

Risk factors: time since polio, more severe initial polio, better recovery from polio, initial polio at older age, female gender, age

PPS diagnostic criteria

- Prior paralytic poliomyelitis
History of acute paralytic illness, residual weakness and muscle atrophy on examination, and signs of denervation on electromyography (EMG).
- Partial or complete functional recovery followed by 15 yrs or more of stable neurologic function.
- New muscle weakness or abnormal muscle fatiguability (decreased endurance) with or without generalized fatigue, atrophy, muscle and joint pain.
- Symptoms should persist for at least one year.
- No other causes.



March of Dimes, 2001

Progression of muscle weakness

- Systematic Review 2005
 - Strength decline is very slow 1-2%/yr (only in studies > 4 yrs f-up)
 - Few long term studies; No prognostic indicators !!
Stolwijk-Swüste J et al. Arch PMR 2005; 86;1693-701
- Mayo Clinic 15 yr follow-up 2005
 - Decline in 31/38 individuals with prior polio
 - Strength declined, not significant
 - No comparison with normal aging
Sorensen EJ et al. Neurology 2005;64:1070-2
- CARPA 5 year f-up study
 - 166 PPS patients, 45-81 years of age
 - Strength decline quadriceps 8%, functioning relatively stable
Stolwijk-Swüste JM et al. Arch Phys Med Rehabil 2010;91:523-8

Progression of functional decline

- Less muscle strength at baseline larger decline in functioning during follow up

Nollet F, et al. Arch Phys Med Rehabil 2003;84(7):1048-56.

Sorensen EJ et al. Neurology 2005;64:1070-2



Limited ‘spare capacity’

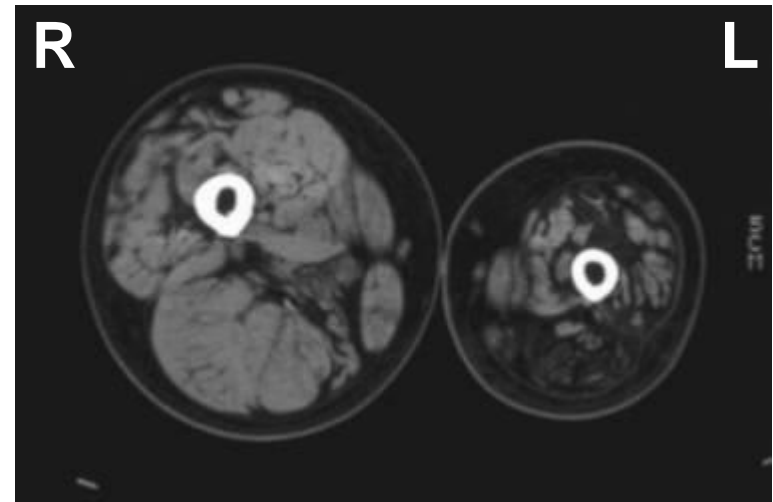
NOTE:

Progression is based on mean changes in a population, however, there is large interindividual variation in affection and location of new symptoms. Therefore individual evaluation is important.

Back to our typical case

- Strength decline right quadriceps*

2000	110.2 Nm
2010	72.5 Nm
Decline	37.7 Nm (34%) = 3,4%/yr



* Measured isometrically in fixed chair dynamometer

Polio paresis a continuum!

Beasley (1961):

‘A widespread view postulates the effects from poliomyelitis into two categories: paralytic and non-paralytic. If this were valid, quantitative muscle testing should result in two separate distributions: one at a ‘paralytic’ level far down the line from ‘normal’; the other, if truly non-paralytic, should remain within the bounds of a distribution for normal subjects.’

‘Rather, there is a continuum in degree of paresis, with moderate amounts being more frequent than truly severe amounts.’

Beasley WC. Arch Phys Med Rehabil 1961;42:398-425

Individual differences in decline

- Polio in 1956, 6 yrs old, full paralysis, respirated



50 years later



Additional factors negatively affecting functioning

- Ageing
- Life style (healthy behavior)
 - Activity, weight gain
- Comorbidity (other diseases)
 - Degenerative disorders such as osteo-arthritis
 - Cardiovascular diseases
 - Diabetes and so on



Content

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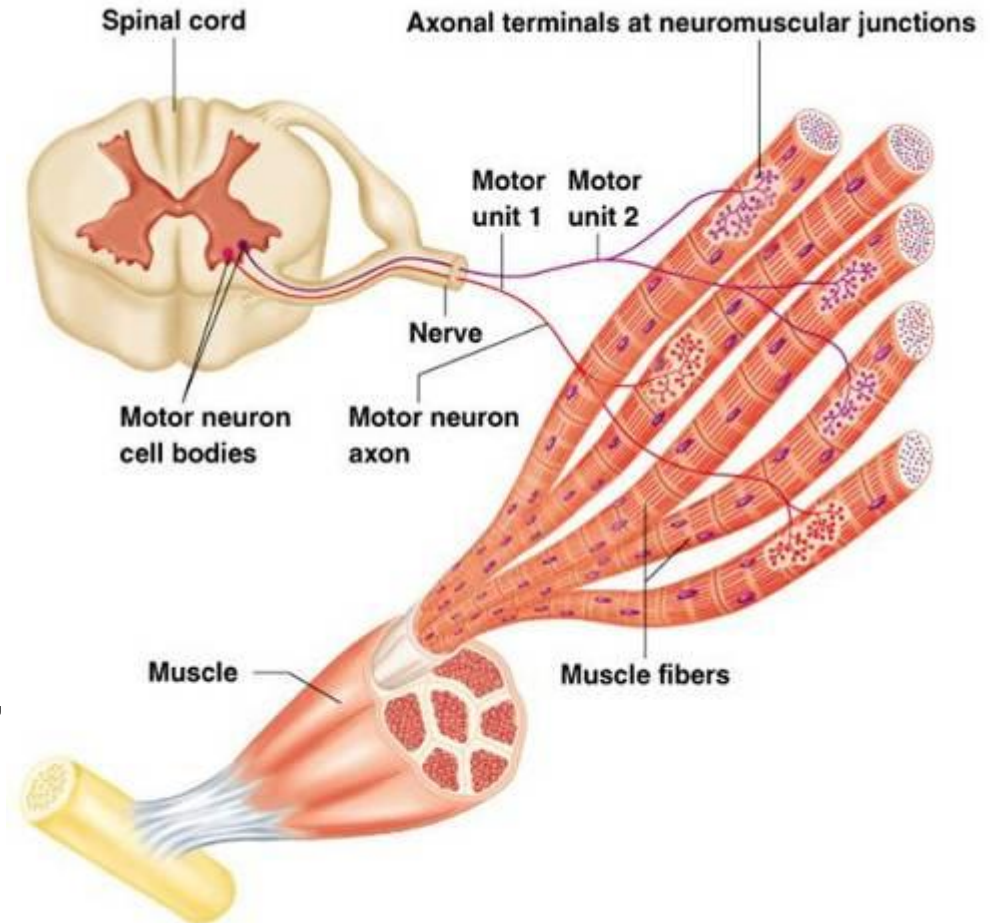
Care in Europe

Aims of the Conference

Cause of post polio syndrome

Motor unit

- Nerve cells called ‘motor neurons’ in the spinal cord form nerve fibers which are grouped in nerves and innervate the voluntary muscles
- Each nerve fiber innervates a certain number of muscle fibers
- A motor unit is one motor neuron, the nerve fiber and the muscle fibers it innervates



(a)

Cause of post polio syndrome

MOTOR UNITS

Normal situation

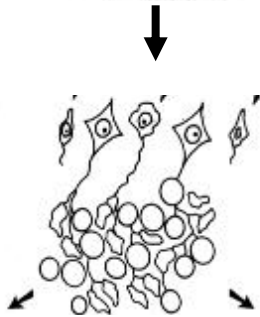


ACUTE POLIO

Destruction of motor neurons

Denervation of muscle fibers

Flaccid paralysis



Full recovery

Partial

RECOVERY PHASE

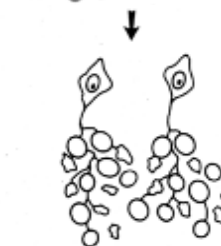
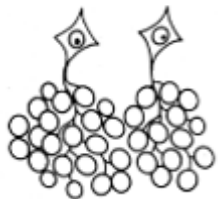
1 Recovery of motor neurons

2 Reinnervation of muscle fibers

3 Muscle fiber hypertrophy

POST POLIO SYNDROME

Loss of isolated muscle fibers
of enlarged motor units



Cause of post polio syndrome

Leading hypothesis:

- Loss of unstable newly formed axonal sprouts due to premature metabolic exhaustion of chronically overloaded motor neurons which had to maintain enlarged motor units for many years.

Wiechers & Hubbell, 1981

Alternative hypotheses:

- Chronic inflammation in the CNS
- Virus (fragments) persistence

Gonzalez, Borg 2002

Toniolo, 2007

Content

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Therapies for post polio syndrome

Review. Gonzalez et al. Lancet Neurol 2010

➔ Insufficient scientific evidence for effectiveness of any intervention

Cochrane Review. Koopman et al, 2011

Medication

- IVIG (Immunoglobulines)?

EFNS Guideline. Elovaara et al, 2008

Rehabilitation

- Exercise? (not harmful)

Systematic Review. Cup et al, 2007

Need further studies!

Some trials are running or in preparation



How to manage post polio syndrome?



Therapy guidelines

Exercise

- Aerobic exercise to maintain muscle status

Behavior

- Patient education to improve mental status and well being
- Psychosocial support to change behavior

Other

- Properly fitted assistive devices
- Weight loss
- Mobility aids and environmental adaptations

EFNS Guideline, Farbu et al. 2006

Content

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Care in Europe?

- European Polio Union founded in 2003
- 19 polio unions from 13 countries



European Union meeting 17-18 October 2007

To get political commitment



Care in Europe? Is it 'bad'?

- Enquiry among member unions in 2008
 - EU + Norway an Suisse 505 million inhabitants
- Mailing to:
 - 16 countries 421 million people (83%)
 - 12 countries (75%) replied 367 million people (73%)
 - 3 lists from 1 country, 2 lists from 2 countries
- Response from:
 - Finland, Denmark, Ireland, Germany, Switzerland, Netherlands, Belgium, France, Spain, Italy, UK, Sweden
 - Not from East European countries

Questions

1. Numbers?
2. PPS recognition and knowledge among health care providers?
3. Organization of post-polio care and care facilities?
4. Availability of therapy, aids, adaptations and services?
5. Scientific topics to be prioritized by the patients?

Estimated numbers of polio cases

- Lowest nr 397.500 or 0.8/1000 (range 0.4 – 2.3)
- Highest nr 1.114.752 or 2.2/1000 (range 0.8 – 6.9)

- Likely 5-600.000 alive 60% PPS / late effects = 300.000



Care for post polio syndrome in EU

1. PPS recognized but knowledge is low and interest questionable.
2. Specialized care is scarce.
3. Access to 'help' is bad in 50% or more of the countries for social work, psychologists, adaptations at home and work, transportation and housing.
4. Research should focus in the first place on symptom management (pain, fatigue) and therapies (medication and rehabilitation).

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Therapy

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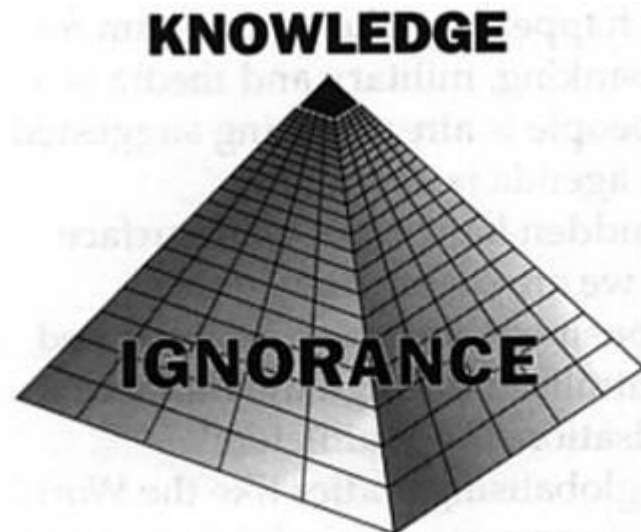
Aims of the Conference

Conclusions

- Research is needed regarding many aspects of post polio syndrome, including cause, course, and effective interventions to maintain muscle function and to reduce symptoms, and also to best aging with polio residuals itself.

Commentary. Nollet, Lancet Neurol 2010

- Care is not optimal in many countries and in general knowledge and interest is low.



Background of the conference

- EPU initiative to bring together polio survivors and researchers
- Inspired by PHI, USA, organizes e-conferences for polio survivors
- Clinical researchers also interested to stimulate care and research



Eventually made possible by



Aim of the conference

- Bring together polio individuals and health providers including all relevant professions and researchers.
- To increase awareness, knowledge and (research) interest and to promote European cooperation among parties,
- With the aim to improve the care for polio survivors with (and without) post polio syndrome to preserve their desired functioning and well-being.



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