

Post-polio syndrome

Clinical manifestations

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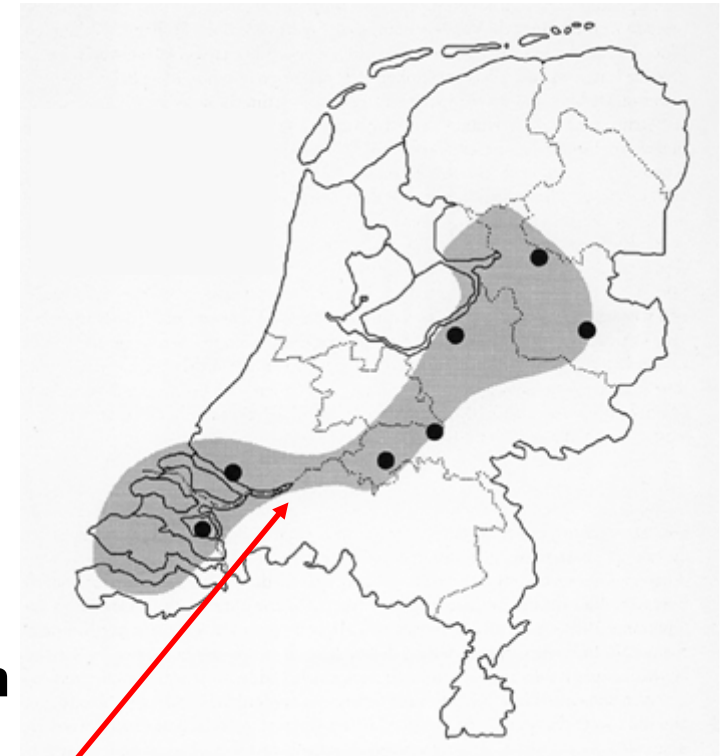
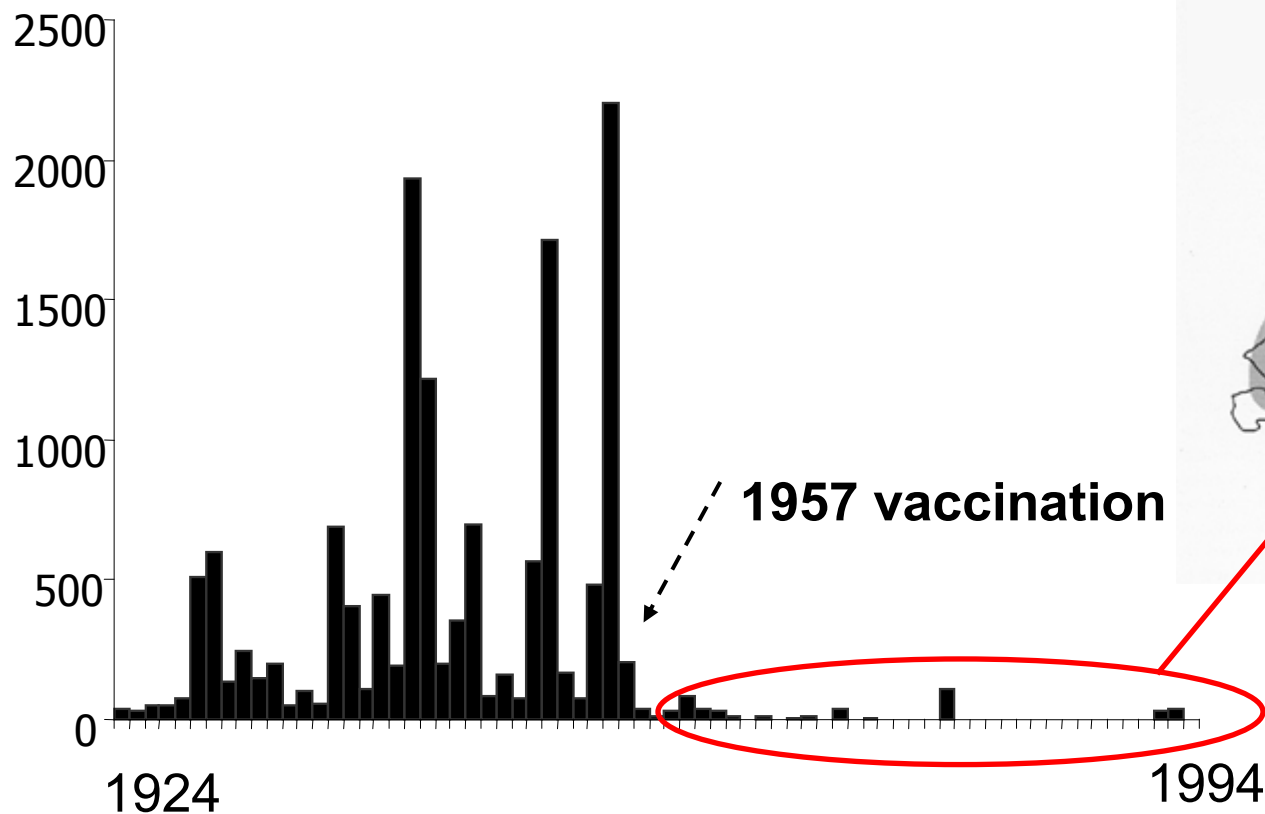
The Netherlands
16 million people



EU 27 countries
483 million people

Polio in the Netherlands

14.762 cases since 1924





Topics

- Polio and Postpolio Syndrome
 - Diagnosis
 - Prognosis
-

Polio Numbers

- ❑ About 20 million world wide
- ❑ About 600.000 in Europe
- ❑ New cases now < 1000 / year



WHO ICD-10

- A80 - Acute polio [viral infection CNS]

 - B91 - Sequelae of polio [residuals]

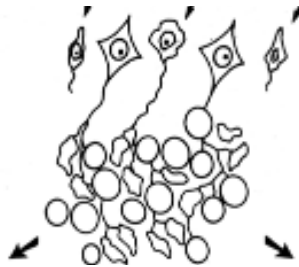
 - G14 - Post poliomyelitic syndrome
[late decline]
-

Pathophysiology of poliomyelitis

Normal



Acute polio



Full recovery



Partial



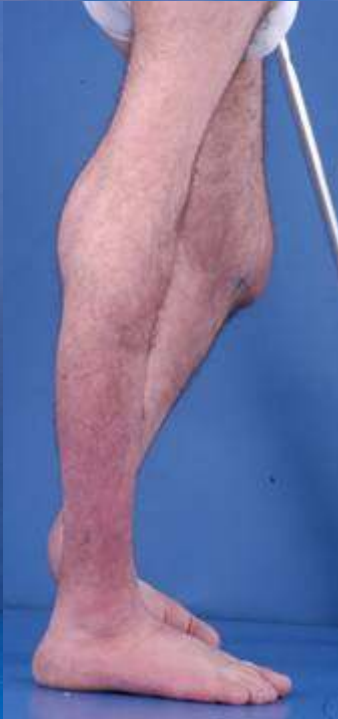
ACUTE POLIO – days/weeks
Destruction of motor neurons
in the spinal cord

RECOVERY PHASE – months/years

- 1 Recovery of motor neurons
- 2 Reinnervation of muscle fibers
- 3 Muscle fiber hypertrophy

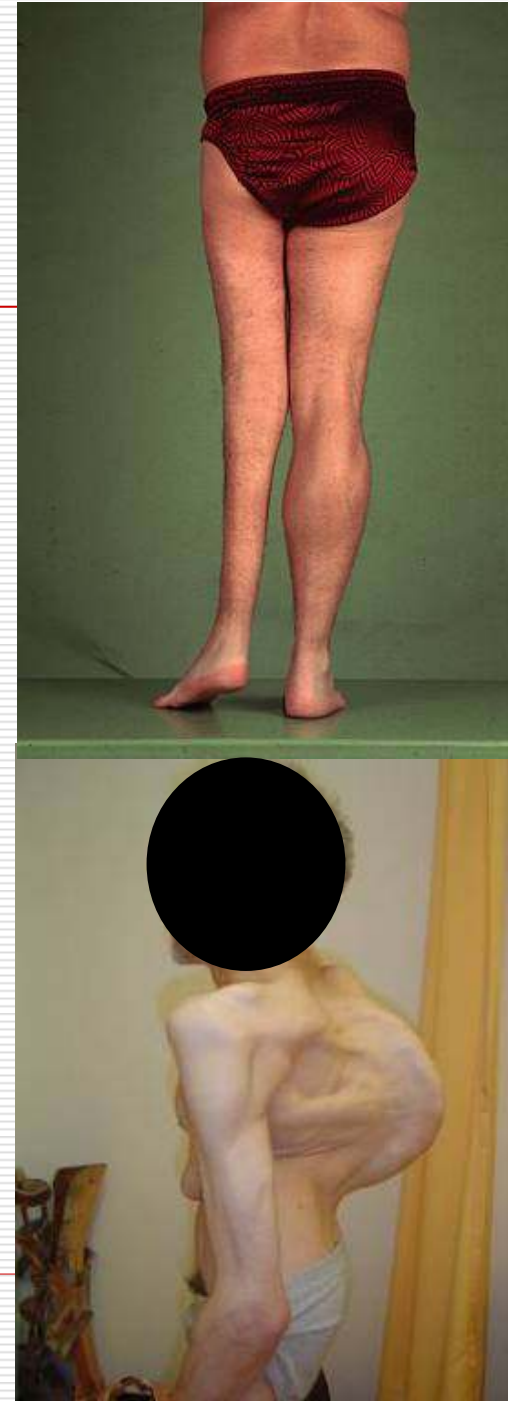
**GIANT MOTOR UNIT SIZES
UP TO 8-10x INCREASED**

→ **STABLE SITUATION FOR DECADES**



Stable Polio Residuals

- ❑ Paretic muscles
- ❑ Non-paretic muscles (!)
- ❑ Hypertrophic muscles (!)
- ❑ Bone and joint deformities (!)



Post-polio syndrome

- New late-onset neuromuscular symptoms
 - After a long period of stable functioning (usually 30-40 years after the acute polio)
 - Symptoms
 - New muscle weakness
 - Fatigue (general and/or local muscle fatigue)
 - Pain (muscle and/or joints) – 50%
 - Functional decline (physical mobility)
 - Cold intolerance,
 - Breathing and swallowing difficulties
 - 40-60% of polio survivors
-

Largest Problems in PPS

- Fatigue (78%)
- Walking outdoors
- Climbing stairs
- Pain

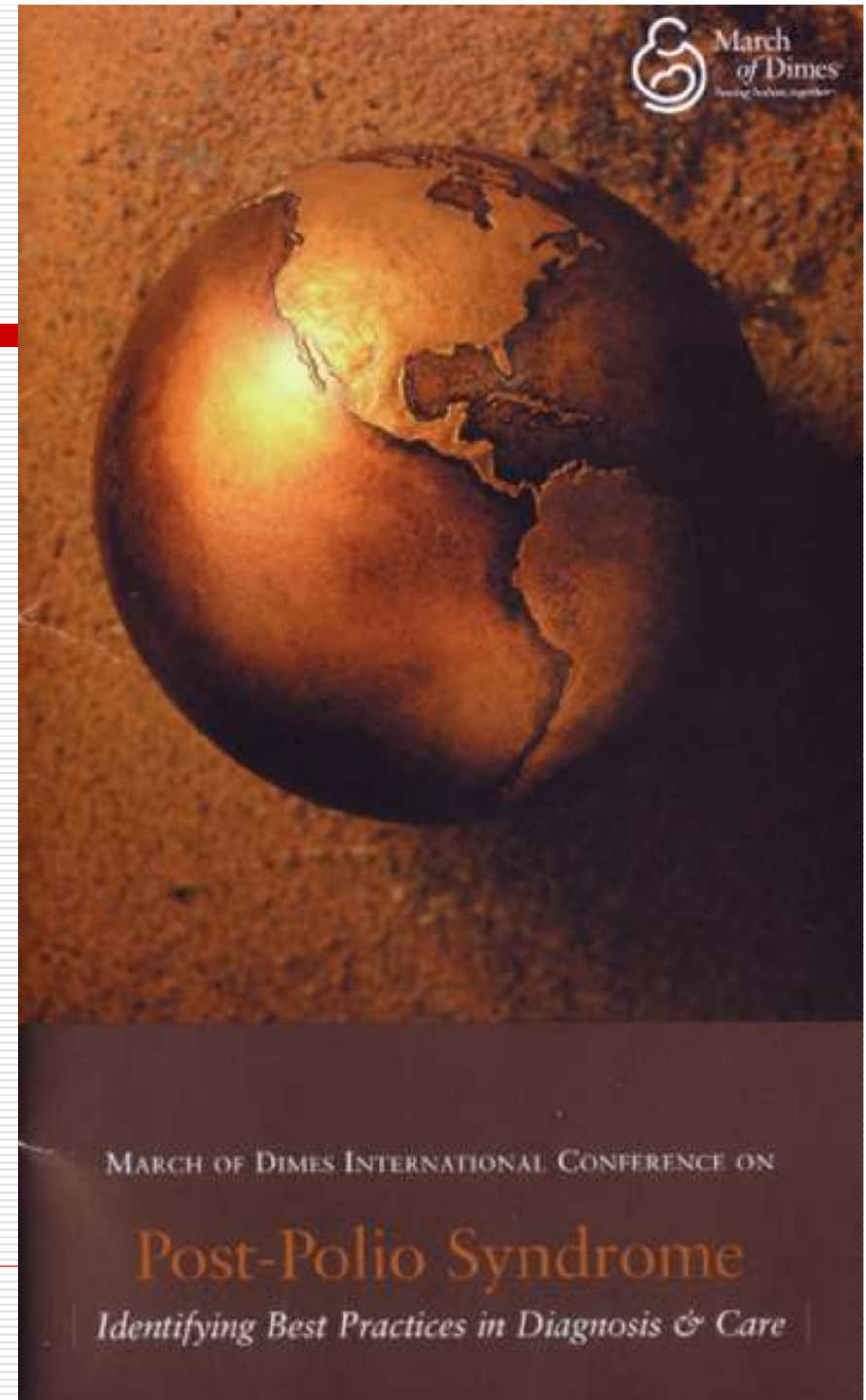
Nollet F et al. Arch Phys Med Rehabil 1999;80:136-43

- 63% severe fatigue in CARPA-study
 - Fatigue Severity Score > 5

Beelen A et al. ISPRM 09 – P-0268

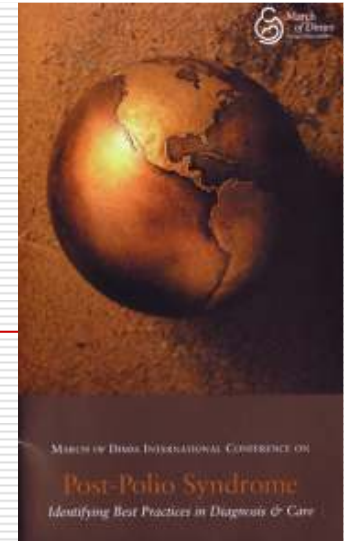
PPS diagnosis

- ❑ Criteria have stabilized since 2000



PPS Diagnosis - definition

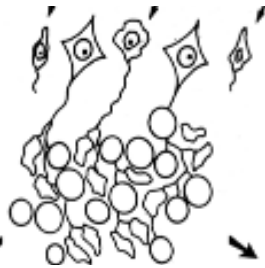
- ❑ New **muscle weakness** or abnormal **muscle fatiguability** (decreased endurance)
- ❑ With or without generalized fatigue, muscle atrophy, or muscle and joint pain
- ❑ Symptoms should **persist at least one year**
- ❑ **No other causes**
- ❑ Insufficient evidence to distinguish subgroups as post-polio progressive muscular atrophy



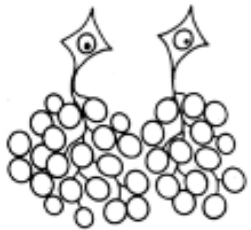
Normal



Acute polio



Full recovery



Partial



PPS



PPS Pathophysiology

ACUTE POLIO

Destruction of motor neurons
in the spinal cord

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 PPS Unknown

Leading hypothesis:

- premature metabolic exhaustion of chronically overloaded motor neurons which had to maintain enlarged motor units for many years

Alternative hypotheses:

- Immunological factors → IVIG
 - Virus persistence
-

PPS Diagnosis - Exclude Other Causes!

- Fatigue

- Anemia, hypothyroidism

- Other neurological conditions

- Spinal stenosis (lumbar, cervical)
- Nerve/root compression
- Anything.....

{ Parkinson's disease
Multiple Sclerosis
Motor Neuron Disease
Malignancies

- Osteo-arthritis

- Psychological causes

- Depressive disorders
-

Joint problems



PPS and Aging

- Decline = multi causal
 - Gradual strength loss
 - Compensation ends
 - Degenerative joint disorders
 - Aging effects*
 - Co-morbidity*
 - Overweight
 - Giving up the fight
 - Inactivity



* Prospective cohort study focussing on effects of co-morbidity and aging on functioning. *Stolwijk-Swüste JM, et al. J Rehabil Med 2007, Archives Physical Med Rehabil 2010*

Time Course

- Systematic Review 2005
 - Clinical course strength and functioning
 - 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 patients
 - No comparison with normal aging

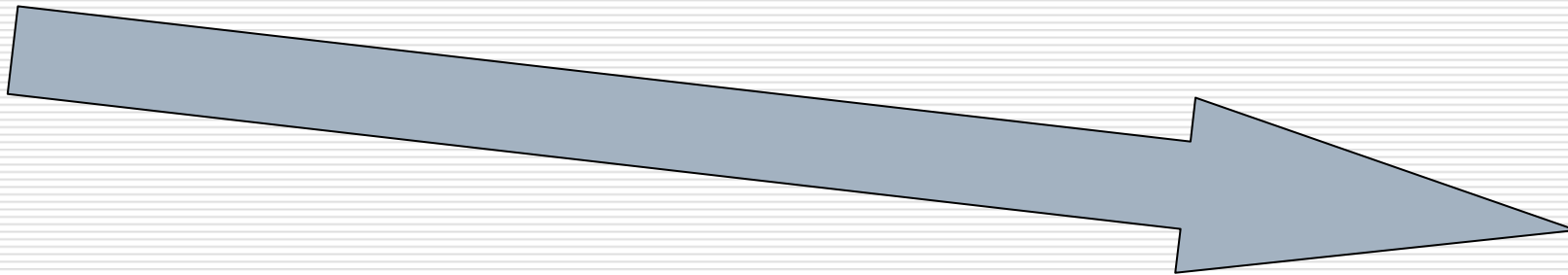
Sorensen EJ et al. Neurology 2005;64:1070-2

- CARPA 5 year f-up study
 - 166 patients, 45-81 years of age
 - Strength decline 8%, functioning relatively stable

Stolwijk-Swüste JM et al. Arch Phys Med Rehabil 2010;91:523-8

PPS Course

- Slow decline in strength



- Less muscle strength at baseline larger decline 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

Slow Decline... Large Consequences



50 years later



Summary I - What is PPS?

- Frequently occurring late decline in muscle function due to the loss of isolated muscle fibers of enlarged motor units, of which the precise cause is unknown
 - With fatigue and reduced physical functioning as the main functional consequences
 - Symptoms due to increasing overuse of slowly diminishing muscle capacity
-

Diagnosing PPS

- How to make the diagnosis PPS?

 - No diagnostic test
 - EMG does not discriminate
 - To confirm polio
 - To detect (subclinical) affection

 - Strength decline is slow
-

PPS

- Can we rely on what our patients tell us?
 - Severity of the acute polio?
 - Extent of residuals?
 - Rate and severity of strength decline? } No!

 - Why not?
 - Severity of the acute polio often unknown
 - The best leg is the 'good' leg
 - Onset of decline often vague, no external reference
-

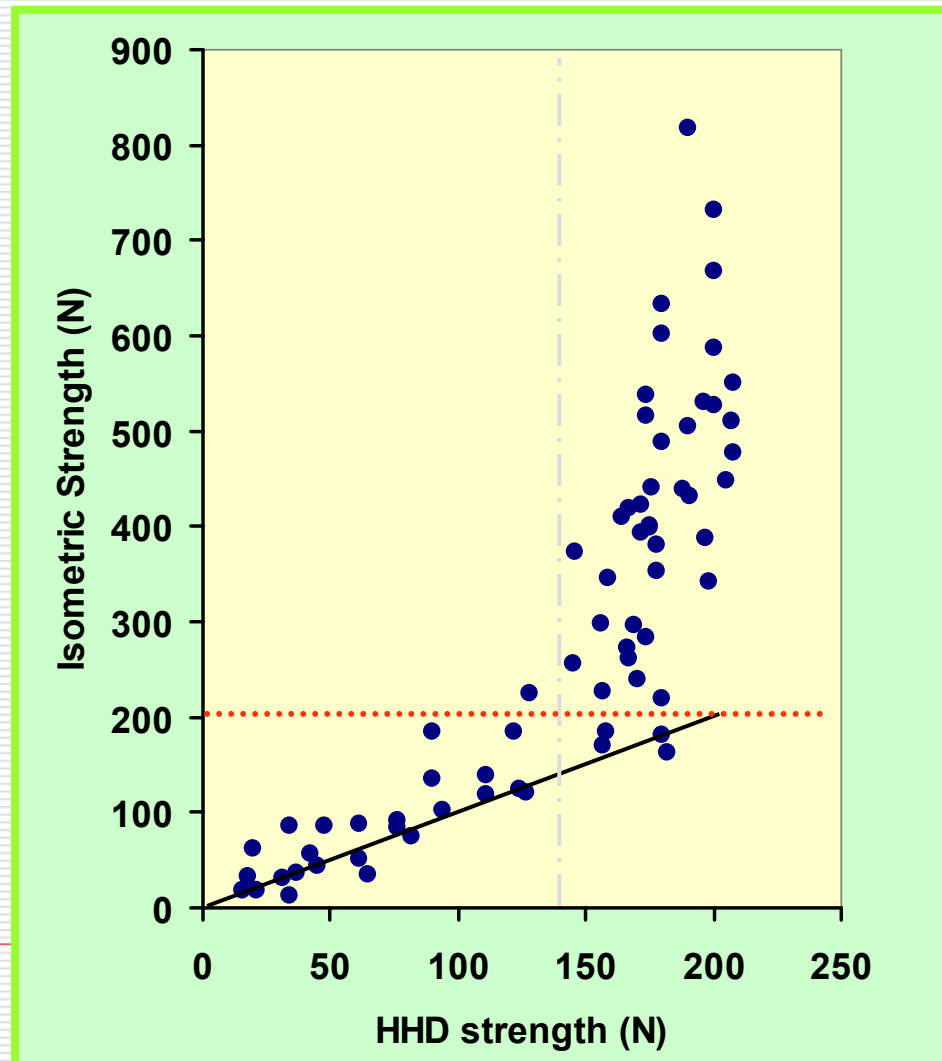
PPS

- Can we rely on our own medical examination?
 - To assess muscle weakness?
 - To assess change in strength? } No!

 - Why not?
 - Limited measurement range
 - Poor reproducibility
-

Limited Measurement Range

manual testing and handheld dynamometers



Poor Ability of Strength Tests to Detect Change

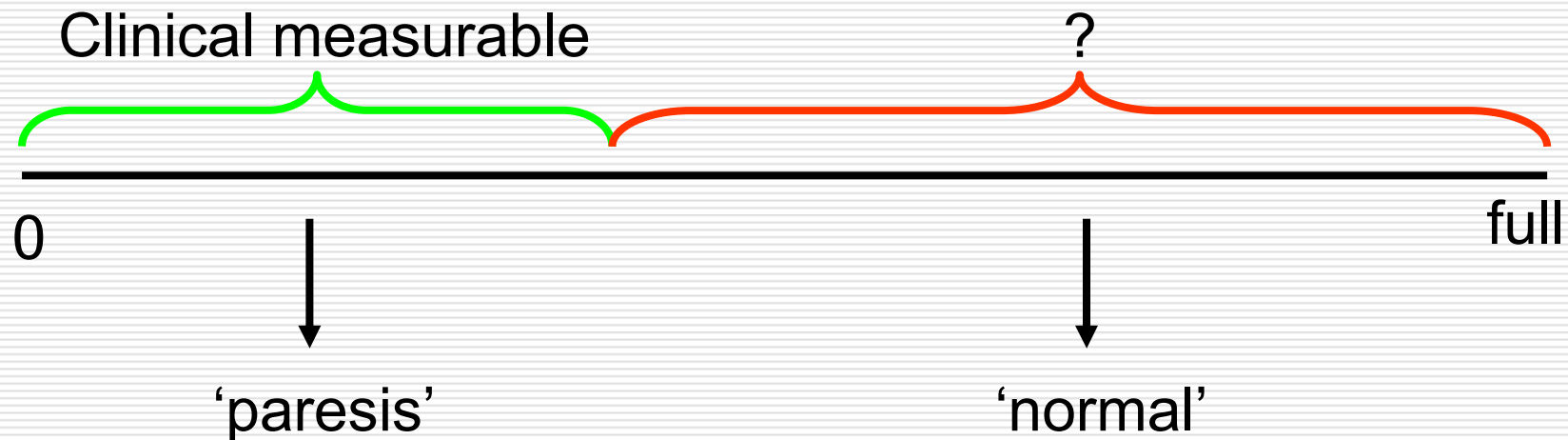
Method	Range	Smallest detectable change
Manual muscle testing	Ceiling effect Grade '5' (250 N)	Not sensitive all grade 4
Hand-held dynamometer ¹	Ceiling effect 250 N	30 - 50%
Fixed dynamometer ²	Unlimited Practical use?	25%

¹ Nolle F, et al. *Arch Phys Med Rehabil* 1999;80:1316-23.

² Horemans HLD, et al. *Arch Phys Med Rehabil* 2004;85: 1273-8

Polio Residuals

- Manual muscle testing
(especially in the lower extremities)



Severity of Polio a Continuum

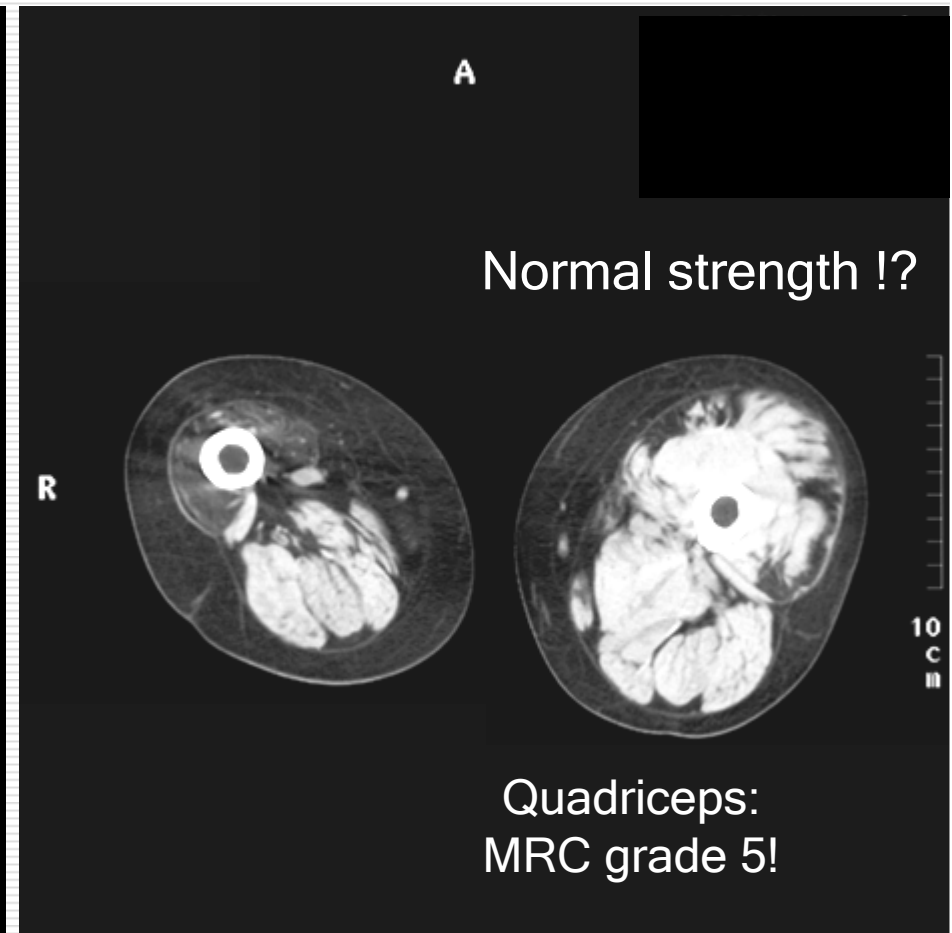
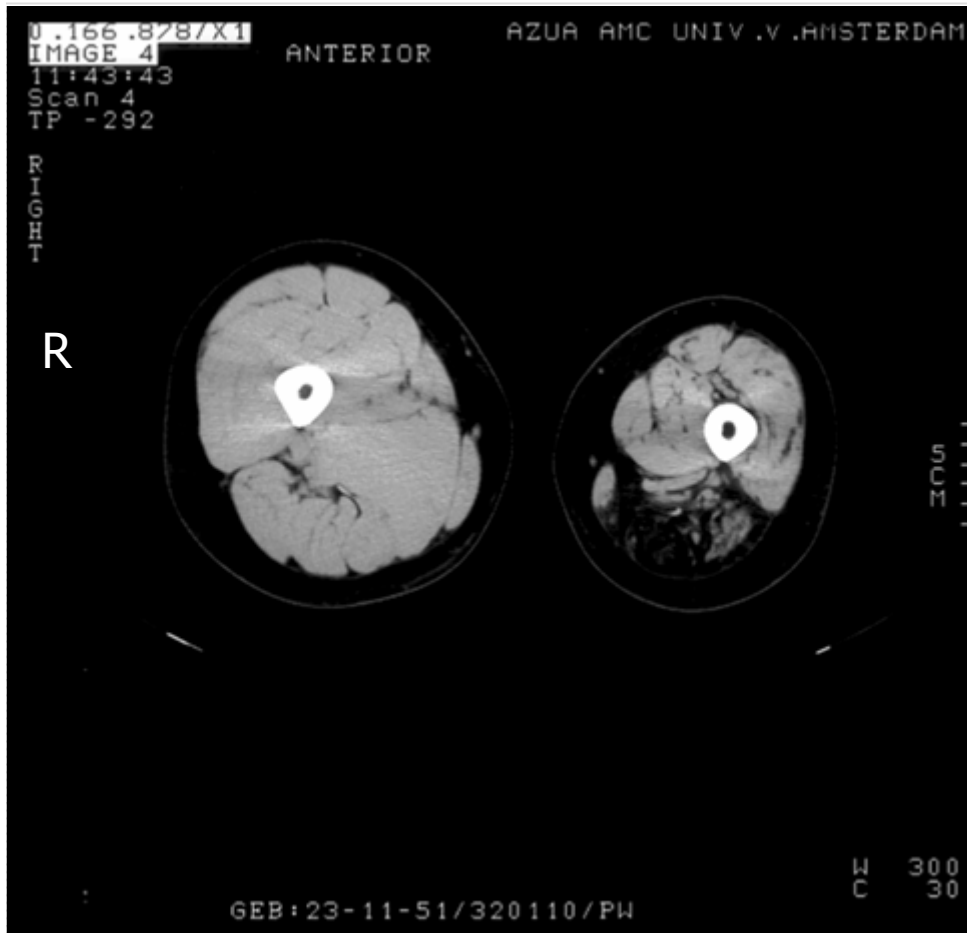
Beasley (1961):

'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.'

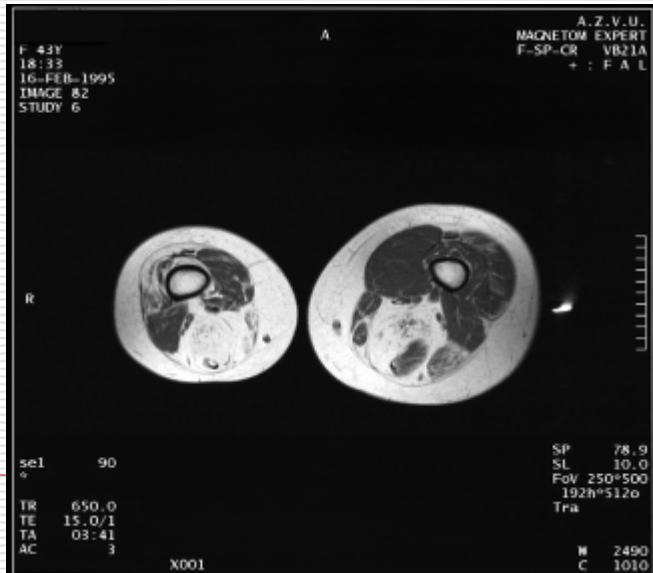
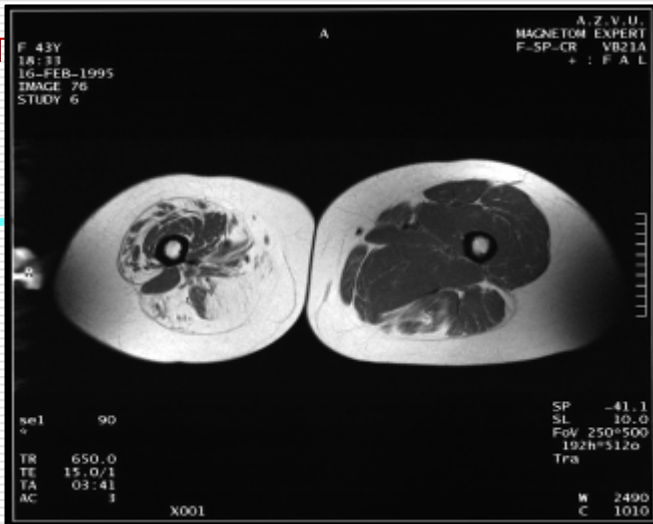
'Rather, there is a continuum in degree of paresis, with moderate amounts being more frequent than truly severe amounts.'

Beasley WC. Quantitative muscle testing: Principles and applications to research and clinical services. Arch Phys Med Rehabil 1961;42:398-425

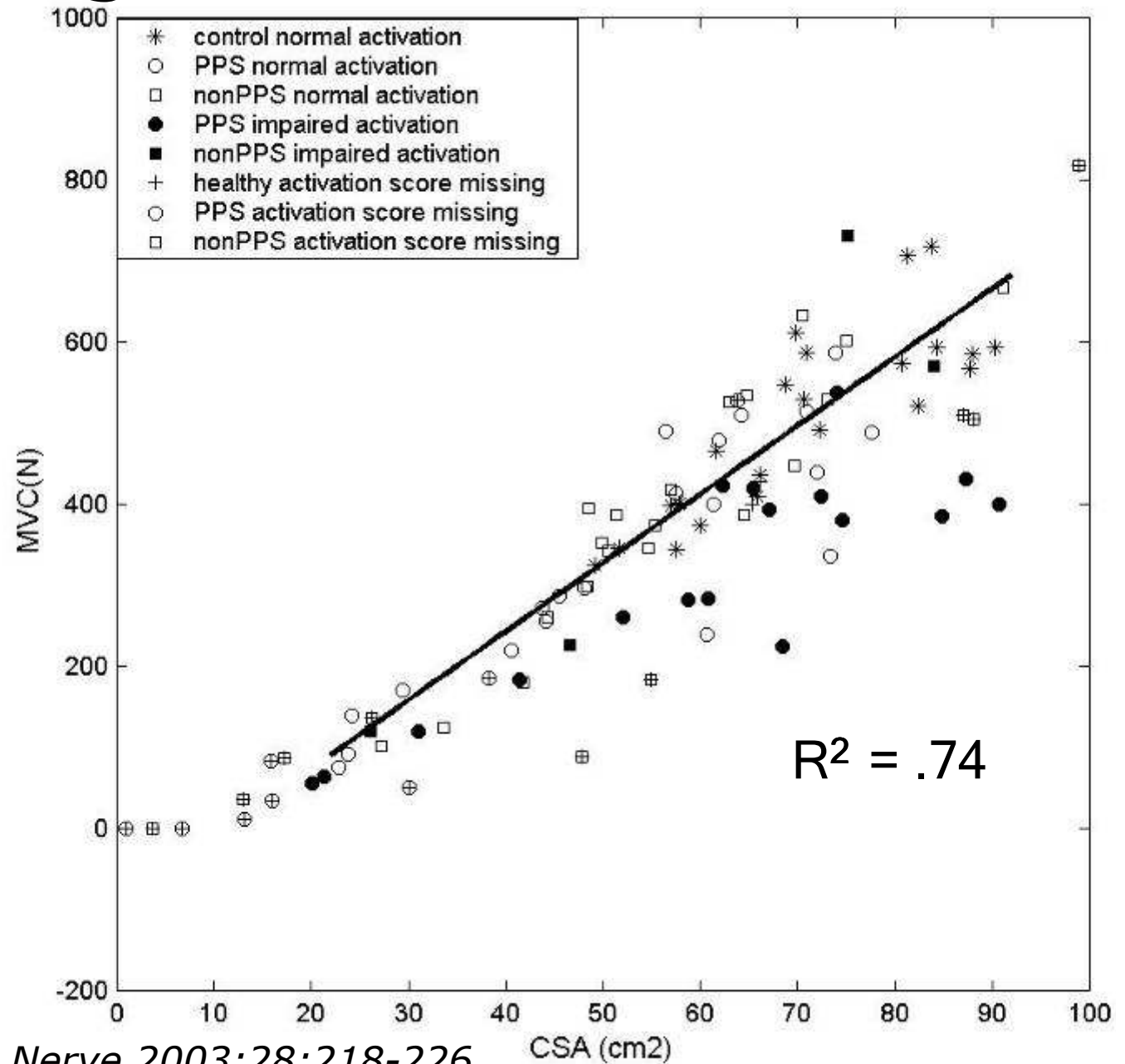
Imaging to Evaluate Muscle Status



Imaging to Evaluate Muscle Status

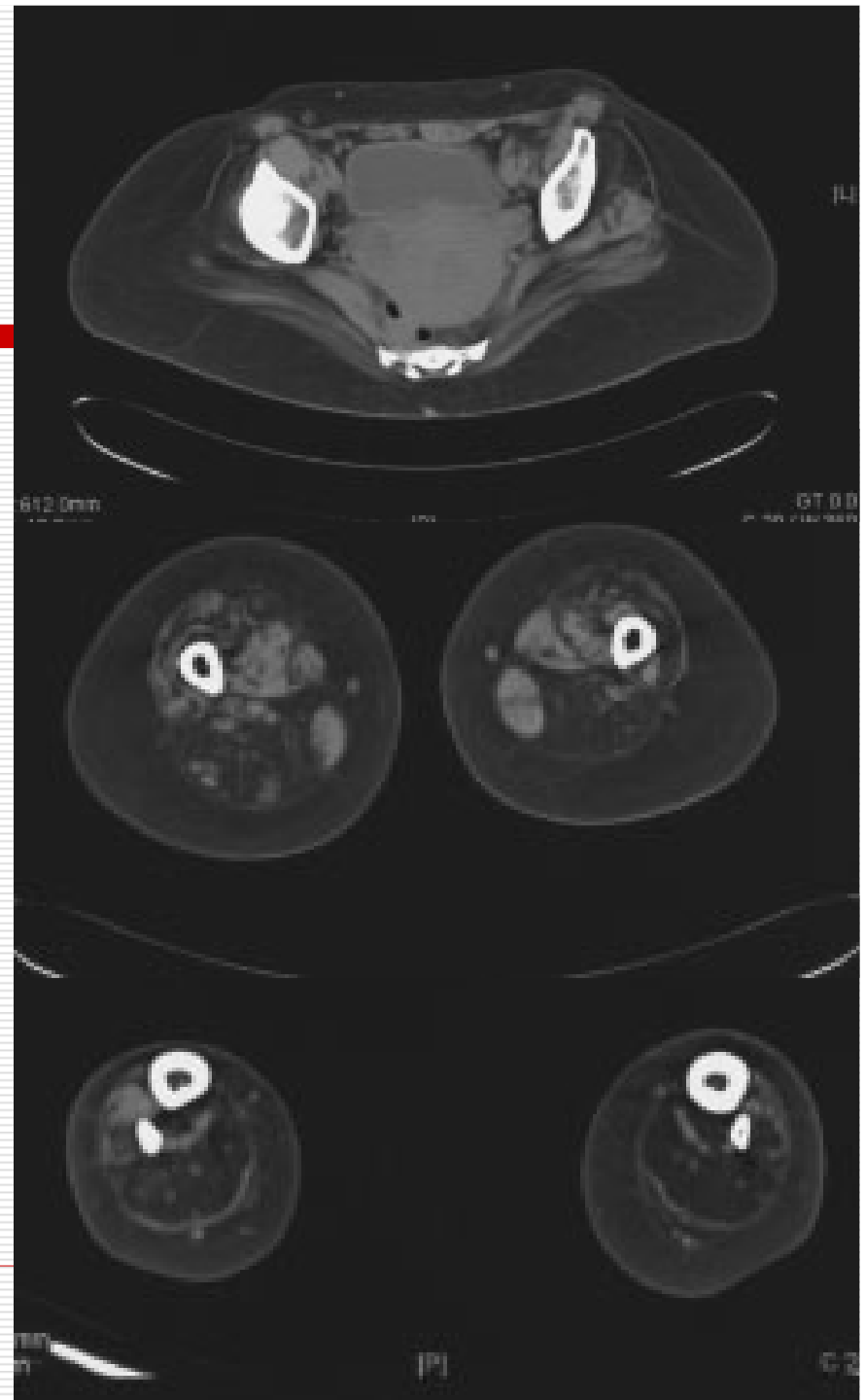
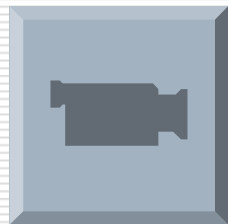


Muscle Strength and CSA



□ Beelen et al. *Muscle Nerve* 2003;28:218-226

'I could always walk very good but now it is getting difficult, I fall a lot'



Respiratory dysfunction

- CO₂ retention
 - Sleep apnea syndrome
-

PPS is not likely in case of

- Fast decline in strength (observed)
 - No (partial) relief on rest
 - Sensory symptoms
-

Summary II- When diagnosing new muscle weakness realize:

- ❑ There is a **continuum** in residual paresis
 - ❑ Clinical muscle tests may **overestimate** strength
 - ❑ A perceived fast decline in strength may be **small in absolute sense** but relatively large compared to the available muscle mass
 - ❑ Objectifying strength decline requires **long follow-up**
-

Summary II – Additional investigations to be used

- Use **EMG** if you need confirmation of prior polio or to exclude other causes
 - Consider **muscle imaging** (CT or MRI) to gain better insight in severity of polio residuals
 - Make **X-rays** to identify secondary joint degeneration
 - Test **Hb, TSH** to exclude anemia, hypothyroidism
 - Do **spirometry, blood gasses**, and/or **somnography** if you consider respiratory problems
-

Thank you for your attention



FIRST ANNOUNCEMENT // EUROPEAN CONFERENCE
COPENHAGEN DENMARK AUGUST 31 – SEPTEMBER 2, 2011

POST POLIO SYNDROME

– a challenge of today

VENUE

Hotel Crowne Plaza Copenhagen Towers,
Østtårns Boulevard 114 – 118,
DK 2300 København S, Denmark,
+45 88 77 88 55
www.cpcopenhagertowers.dk



REGISTRATION FEE //

Registration fee for professionals before
April 1st 2011 – 3000 DKK / 400 Euro
Registration fee for polio survivors before
April 1st 2011 – 1465 DKK / 195 Euro
Registration fee for professionals after
April 1st 2011 – 3500 DKK / 470 Euro
Registration fee for polio survivors after
April 1st 2011 – 1950 DKK / 260 Euro

Registration fee includes participation of the sessions and catering during the conference & free admission for the Welcome reception. Conference dinner must be paid separately.

Online registration will be open via the congress website from December 2010

FURTHER INFORMATION //

The progress of the conference can be followed on the congress website
www.polioconference.com
You are also welcome to contact us at:
mbe@ptu.dk



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LIVET EFTER ULYKKEN