

Management of Abnormal Tone

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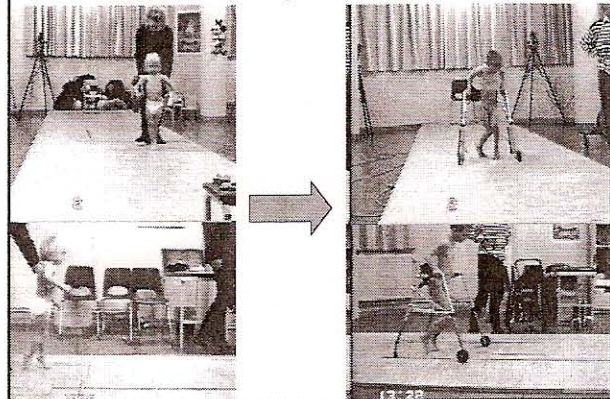
Gait Lab Pellenberg

CP Team Pellenberg

University Hospital Pellenberg,
Leuven



The natural history of the child with CP



What is CP ?



Do no harm

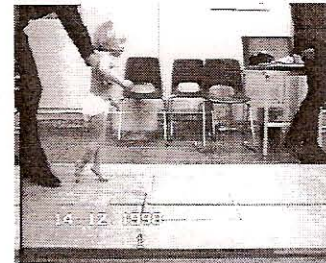
Cerebral Palsy

An insult of the developing brain that produces a disorder of movement and posture that is permanent but not unchanging

Mercer Rang

Primary = problems related to neuronal lesion

- Tone
- Balance
- Selectivity
- Strength



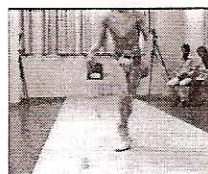
- Spasticity is velocity depending

Secondary = orthopaedic problems

- Fixed contractures



- Bony deformities (lever-arm dysfunction)



Tertiary problems



primary
To overcome problems
secondary
↓
coping responses

Children

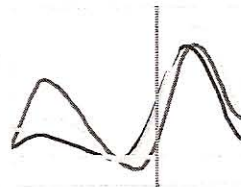
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Growth

Knee joint motion

of 3 years old child

3 trials of the same child



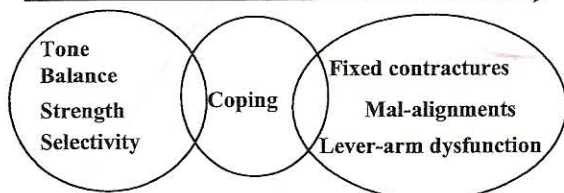
Maturity of gait



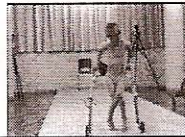
Primary problems

Maturity

Secondary problems



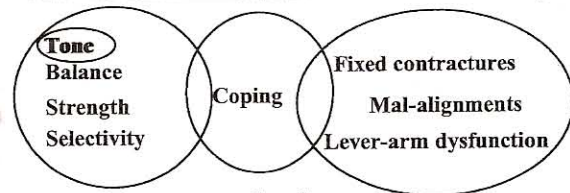
Tertiary problems



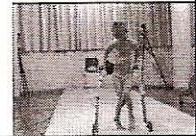
Primary problems

Maturity

Secondary problems



Tertiary problems



Spasticity

Scientific definition (Lance 1980)

Spasticity is a motor disorder
characterized

by a velocity-dependent increase in tonic stretch
reflexes (muscle tone), with exaggerated tendon jerks

resulting from

hyperexcitability of the stretch reflex,
as one component of the upper motor neuron
syndrome

Spasticity

Subjective definition (Gage 2004)

Spasticity is like love,

You know it when you feel it,

It is all embracing.

It is centrally mediated

And has important peripheral
manifestations

From Gage 2004 (personal communication with J. Oppenheim)

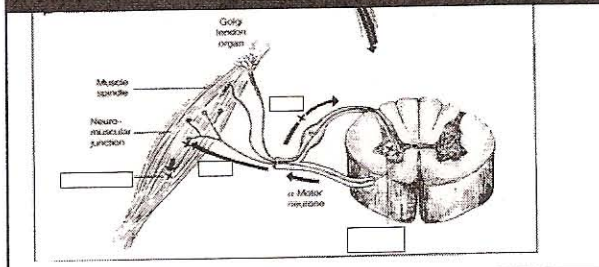
Normal muscle tone Controlled by two factors

- Excitatory signals coming from the muscles into the spinal cord, resulting in contraction
=> Reflex-arc
- Inhibitory signals coming down from the brain into the spinal cord, causing the release of a chemical, GABA, which make the muscles relax

The sensory inputs to the control system

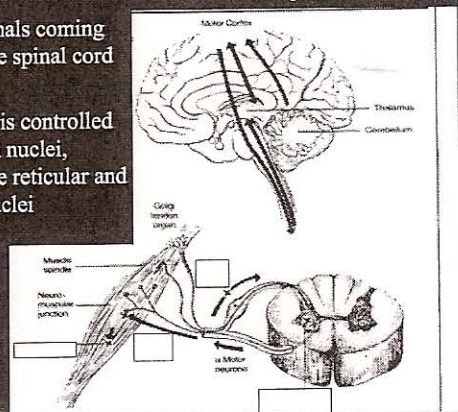
- * muscle spindles
- * Golgi tendon organs

Both sensory organs contribute to spinal cord control of muscle function and provide instantaneous information to higher control centers of function



The reflex arc is controlled by the brain

- Inhibitory signals coming down into the spinal cord
- Muscle tone is controlled by brainstem nuclei, especially the reticular and vestibular nuclei



Spasticity

Loss of descending inhibitory influences at the level of the alpha motor neuron and the spinal interneurons



Exaggerated stretch reflexes

Spasticity and fixation

Normal development:

- Proximal fixation to gain stability and to allow motion in distal joints
- Fixation (= immature pattern) for stability disappears



Child with CP: instability

- Over-use and continued use of fixation
- Prohibits selective, coordinated, dissociated movements
- Use of hypertonia in total patterns

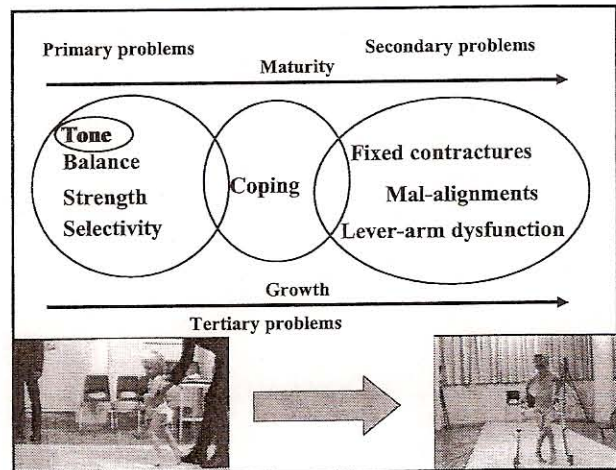


Spasticity in children is often used to overcome other problems

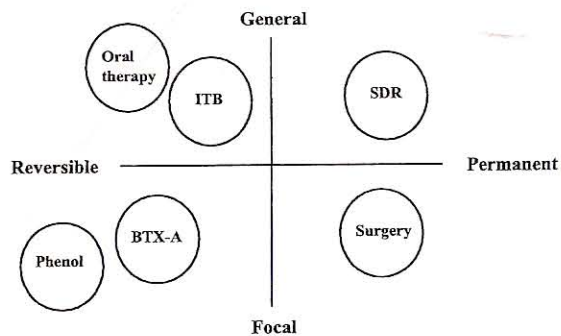
- Problem of instability
- Problem of underlying weakness
- Problem of asymmetry
- Problem of dystonia (mixed tone problem)

Spasticity is not a synonymous with upper motor neuron syndrome

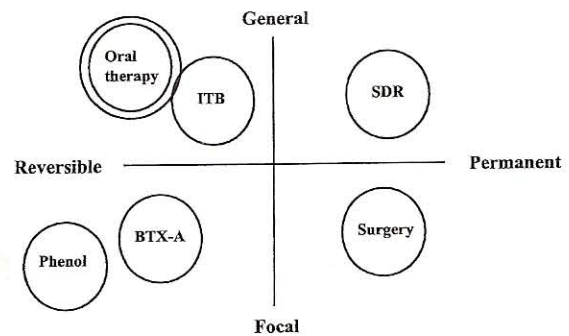
- It has been used almost synonymously with the term CP (Crothers & Paine 1988)
- Spasticity is just one feature of the Upper Motor Neuron syndrome
- Spasticity is a one part of a clinical picture
- Spasticity can arise from a multitude of lesions



Management of Spasticity



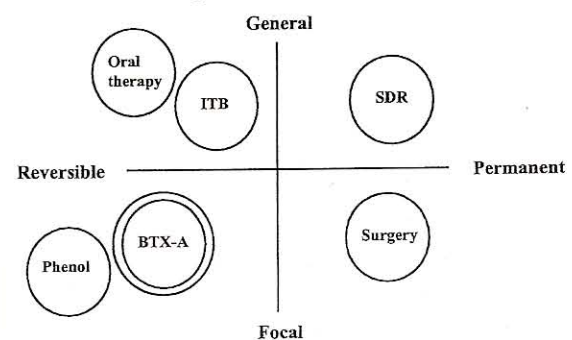
Management of Spasticity



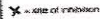
Oral medication

- Can be helpful in selected patients where *generalised tone reduction* is desired
- Effect by inhibiting excitatory neurotransmitters or augmenting inhibitory neurotransmitters at the level of the spinal cord
- Examples are oral Baclofen, Dantrolene and Diazepam
- Drugs are not selective
- Because medication administered orally must cross the blood-brain barrier => oral medication often requires doses that produce unwanted side-effects (sedation, weakness,...)

Management of Spasticity



Leaving an *in vacuo* interface as a convenient route to obtaining crystals of the chromophore, we have now extended our studies to an interface that is not only more readily accessible but also provides a more homogeneous distribution of the chromophore and hence a more uniform film. This may be the most important factor in obtaining a high quantum yield. The results of our studies are to be published in the near future.



BTX-A = neurotoxin, that affects the neuromuscular synaps by inhibiting the release of acetylcholine



Bas

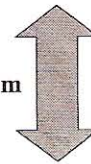
Summary: muscles that can cause the pathological pattern

- Psoas bilat.
- Rectus femoris right
- Medial hamstrings bilat.
5 R/4L U/Kg BW
- Gastrocnemius bilat.
8 R/6L U/Kg BW

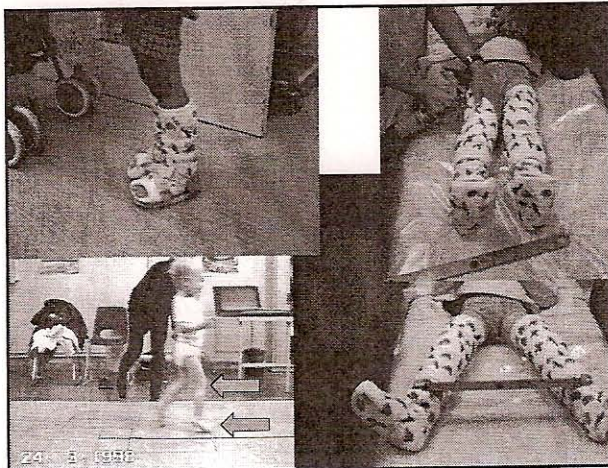


Pre- and after care

Continuum

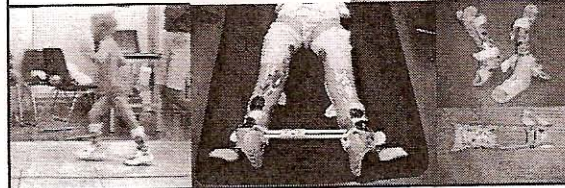
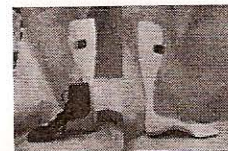


- Combined treatment with casting
- Appropriate orthotic management
- Optimal physiotherapy



BTX-A treatment Orthotic management (Bas)

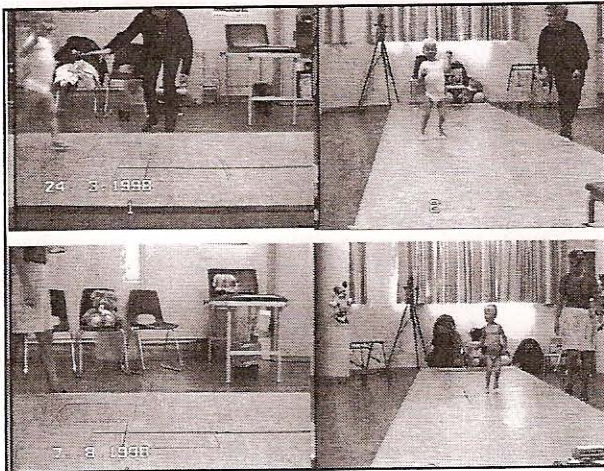
- Day: leafsprings bilat.
- Night: KAFO + abduction-exorotation rod



Physiotherapy

- Pre BTX-A injections
- Post BTX-A injections
- Long term adapted physical therapy

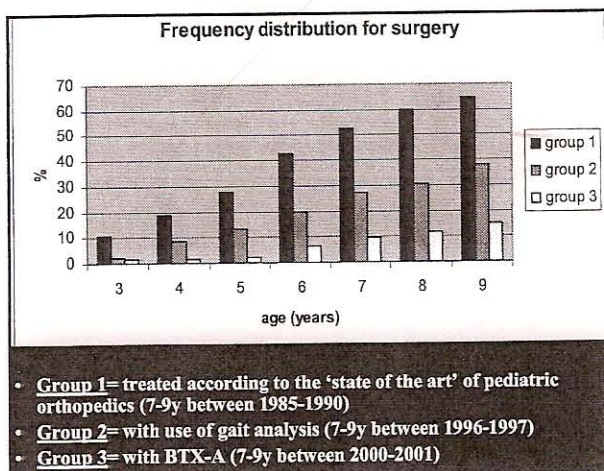
Outcome of BTX-A treatment



What is the impact of the use of BTX-A in the patient population of children with CP ?

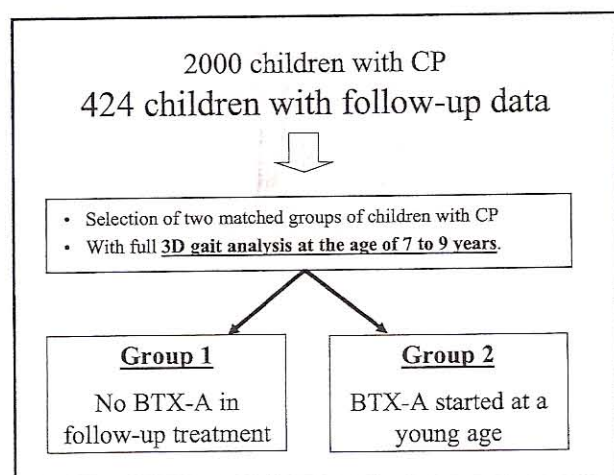
Retrospective study

- 2000 patients with CP at the University Hospital of Pellenberg, born between 1975-2001
- Selection of 424 children: Longitudinal follow-up, treated according to the current 'state of the art' (orthoses, PT, casts, tone reduction, surgery)



BTX-A can delay and reduce surgery

What about the overall condition of these children ?



SUMMARY

Group 1 (no BTX-A)

➤ More pronounced pathological pattern, with secondary (fixed) problems

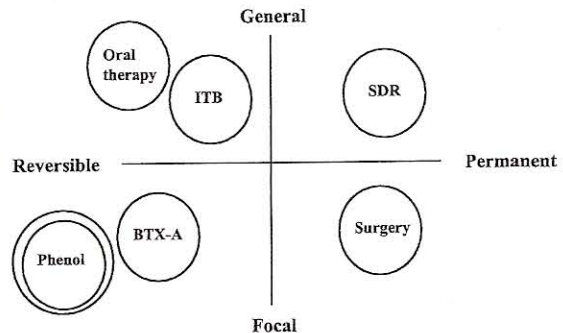
Group 2 (with BTX-A)

➤ Gait less defined by secondary problems (less fixed problems)

Conclusion:

We have objective evidence that we can influence, in a positive way, the natural history of CP by applying BTX-A treatment according to the integrated approach

Management of Spasticity



Phenol neurolysis

- Neurolytic block
- Focal, reversible tone reduction
3 – 12 months
- Side-effects:
– dysesthesias

Management of Spasticity

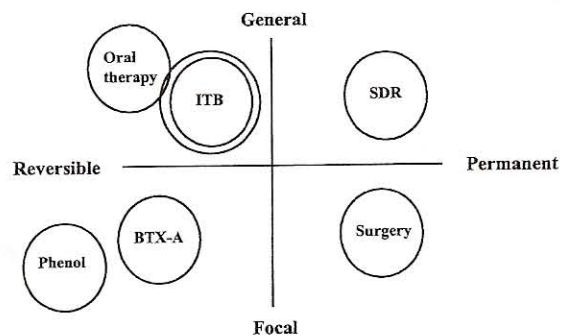
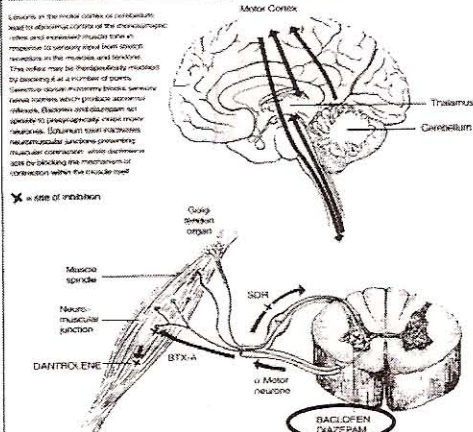
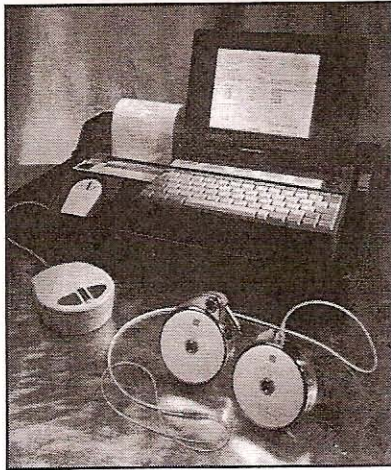


Figure 1: Pathways involved in spasticity

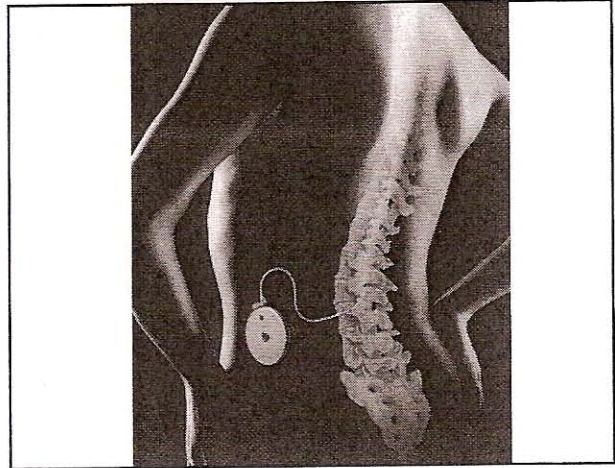


Intrathecal baclofen (ITB) Working mechanism

- GABA-analogue
(GABA = inhibiting neurotransmitter with key role in tone regulation)
- Tone reduction by inhibition of the spinal stretch reflex (binding of GABA-B receptors)
- Oral baclofen: only 3% in CSF (cerebro spinal fluid), often lack of sufficient tone-reduction due to side-effects
- Intrathecal baclofen: 95 % in CSF, less side-effects
- Plasma concentration 100 X less than for oral medication



- Diameter of 6.5 cm
- 2 types:
 - 18 ml, 10 ml: 17% smaller
 - NEW 20ml, 40ml
- Battery for 3.5 to 7 years (recent models until 10 years)
- Pump is programmed by external computer to regulate dosage
- Filling of pump by injection



ITB Indications

Movement disorders

Spasticity
Dystonia
Mixed tone

more generalised spasticity

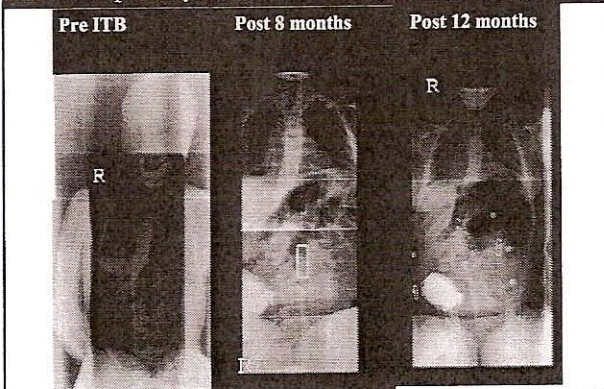
ITB contra-indications

- Too small to support pump
 - Nutritional status (?)
 - New pump somewhat smaller
- disorders without a positive response to a trial infusion of ITB (double blind)
- Poor trunk control / scoliosis (?)

Rapid progression of scoliosis

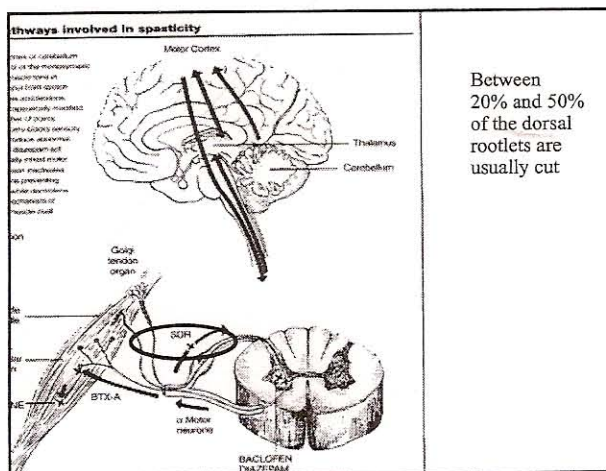
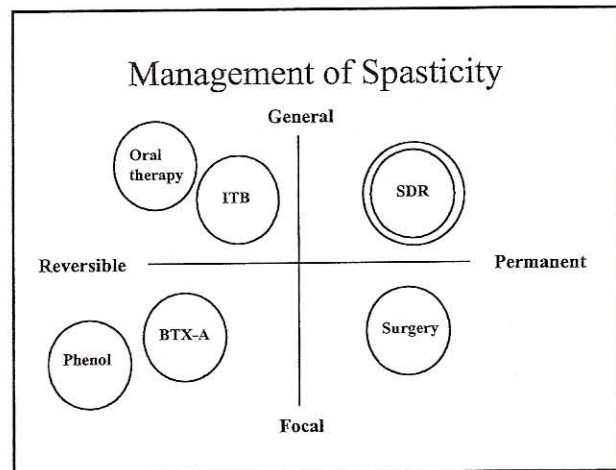
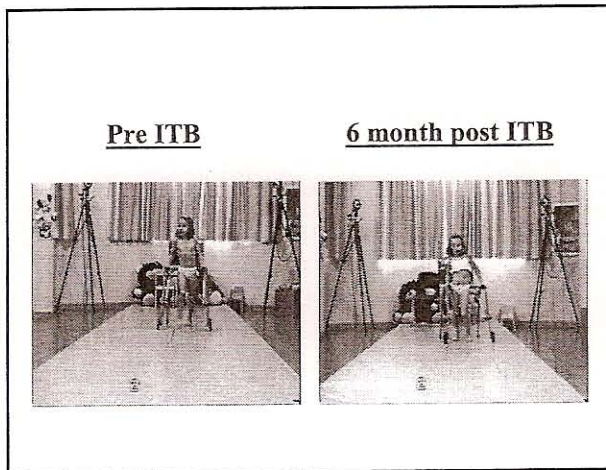
Example: 13 years old child with spastic quadriplegia

Pre ITB Post 8 months Post 12 months



ITB-procedure => Integrated approach

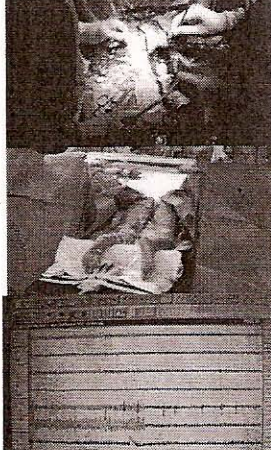
- Multi-disciplinary team
- Longitudinal follow-up pre ITB
- Multilevel BTX-A before ITB =>
 - Evaluation of effect of tone reduction
 - Focal versus general tone reduction
- Optimal after-care
 - Intensive rehab program => focus on strengthening
 - Optimal orthotic management
 - More involved children => optimal positioning
 - Functional children => prevention of crouch



SDR

University Hospital Pellenberg

- Laminoplasty L1-L5
- Stimulation of sensory rootlets (L1-S2)
- EMG monitoring of major lower limb muscles and physical evaluation
- Rootlet is cut when there is a pathological response
 - Crescendo response
 - Long lasting response
 - Activation of multiple or contralateral muscles
 - Abnormal EMG pattern
- Average SDR of 29.9% (25.9-35.9)
- Intensive rehabilitation post SDR



SDR: indications

University Hospital Pellenberg

- Pure bilateral spasticity (diplegia)
- Ex-prematurity, periventriculaire leucomalacie
- Good strength and selectivity and good trunk control
- No previous surgical interventions with muscle lengthening
- Good motivation and cooperation (intensive rehabilitation!)
- Age preferably >5 years (cooperation) and <10 years (secondary problems)

SDR Literature

- Large variation in patient selection (diplegia, hemiplegia and quadriplegia)
- Large variation in percentage of rhizotomy
- Large variation in surgical technique (laminectomy en laminoplasty)
- Large variation in procedure of selectivity (monitoring of muscles, use of clinical information)
- Variation in level of rhizotomy
 - S1 / S2 ??? (soleus !!!, strength of gluteus maximus)
 - L5 ??? (Gluteus and hamstrings on same level)
- Variation in aftercare

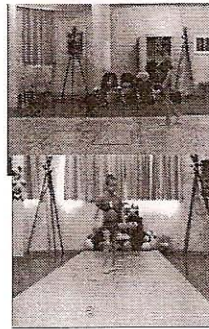
Longterm risks of SDR

- Irreversible treatment !!!!!
- Weakness
- Crouch pattern
- Correction of lever-arm dysfunction
 - later
 - before

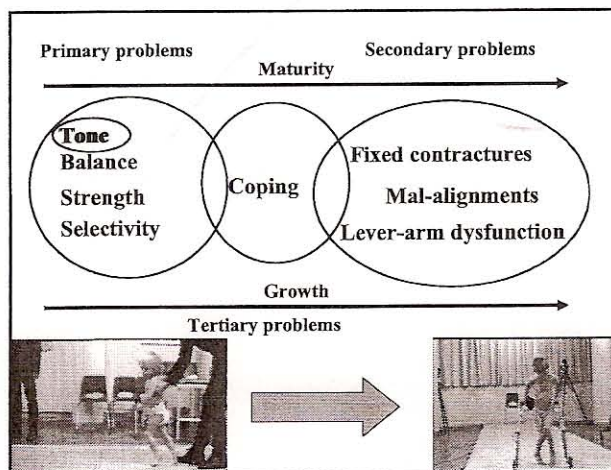
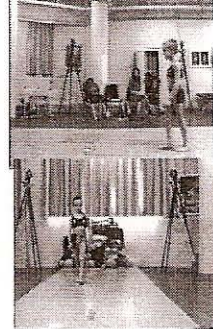
Example SDR:

8 y old child with spastic diplegia

PRE SDR



1 year POST SDR



Thank you

