



24th Annual Meeting of European Academy of
Childhood Disability
16-19 May 2012
Istanbul, Turkey

The algorithm of Orthopaedic Management of Cerebral Palsy: When surgery, When BTx?

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Definition of Cerebral Palsy

- ✓ Motor deficit occurs due to a static nonprogressive lesion of the brain
- ✓ William John Little, 1830
“Brain injury due to oxygen deprivation to the brain at birth”
- ✓ Sir William Osler, 1889
“The Cerebral Palsies of Children”

Causes of Cerebral Palsy

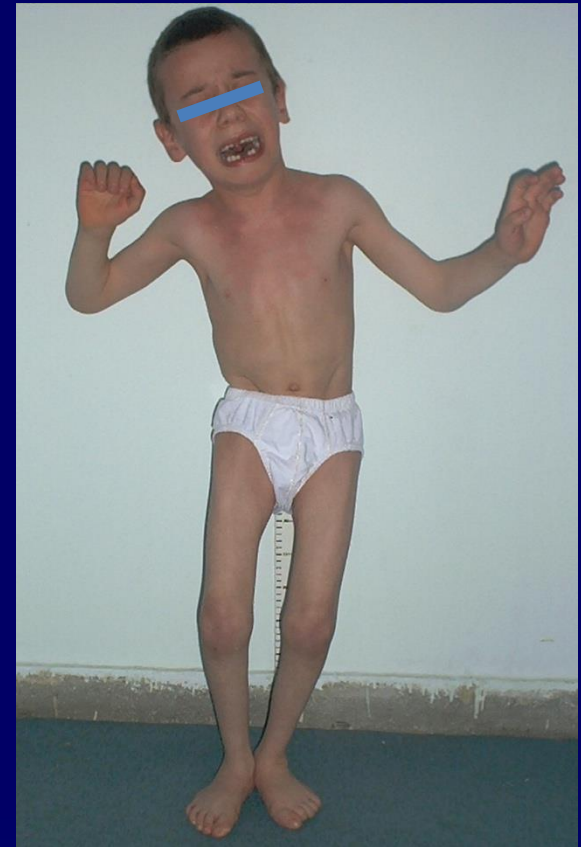
- ✓ Prenatal – 26.6%
 - ✓ Bleeding in the brain, brain infections etc.
- ✓ Perinatal – 18.5%
 - ✓ Injury during child birth with traumatic delivery
- ✓ Postnatal – 5.9%
 - ✓ Head injuries, severe jaundice etc.
- ✓ Unclassifiable – 49%

Prevalence of Cerebral Palsy

- ✓ 1 – 4 per 1000 live births
 - ✓ 2.4 per 1000 live births in developed countries
 - ✓ Invitro fertilization and improved postnatal care !
- ✓ In Turkey, 4.4 per 1000 live births

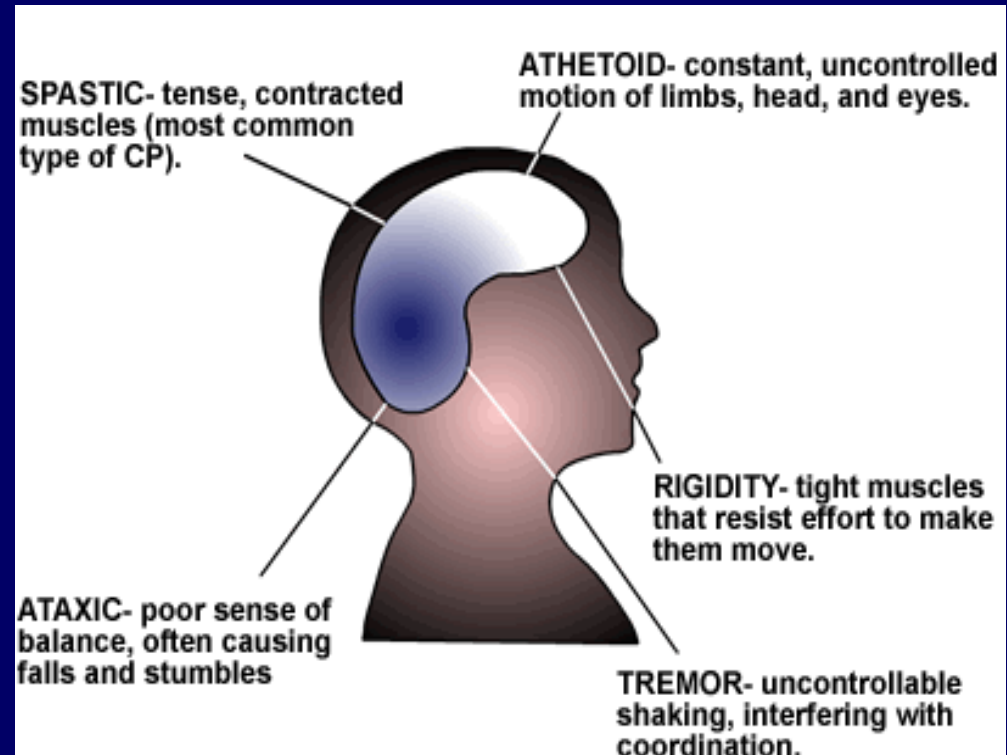
In Cerebral Palsy...

- ✓ Impossible to correct the brain damage
- ✓ Orthopaedic and biomechanical problems develop secondary to this disorder
- ✓ Some problems will continue lifelong



Types of Cerebral Palsy

- ✓ Spasticity
- ✓ Athetosis
- ✓ Ataxia
- ✓ Hypotonia
- ✓ Mixed



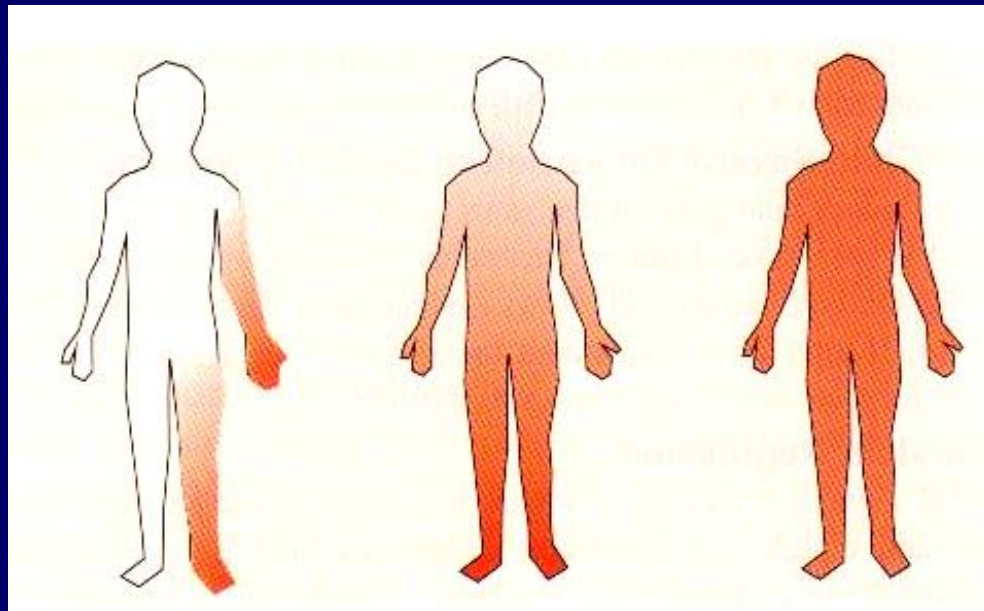
Types of Cerebral Palsy

- ✓ Spasticity
- ✓ Athetosis
- ✓ Ataxia
- ✓ Hypotonia
- ✓ Mixed



Topographical distribution

- ✓ Monoplegia
- ✓ Hemiplegia
- ✓ Diplegia
- ✓ Triplegia
- ✓ Quadriplegia
- ✓ Double hemiplegia



Associated problems

- ✓ Seizures
- ✓ Visual impairments
- ✓ Intellectual impairment
- ✓ Learning disabilities
- ✓ Hearing problems
- ✓ Communication problems and dysarthria
- ✓ Oromotor dysfunction
- ✓ Gastrointestinal problems and nutrition
- ✓ Teeth problems
- ✓ Respiratory dysfunction
- ✓ Bladder and bowel problems
- ✓ Social and emotional disturbances

Cerebral palsy without treatment

Progression of the deformity

Hypotonia - Spasticity

Dynamic deformity

Static deformity and fix contracture

Bone and joint deformities

The goals of treatment

- ✓ Improvement of life quality
 - ✓ Motion without assistance and pain
 - ✓ Sitting and individual care
 - ✓ Functional development, independent walking and working

The goals of treatment

- ✓ To correct posture and walking
 - ✓ Balanced and stable spine and pelvis
 - ✓ Extension ability for hip and knee joints in stance phase
 - ✓ Plantigrade feet

Orthopaedic management for...



Spasticity

Contracture

Bone and joint deformities

Gross Motor Function Classification System (GMFCS)



✓ Level I



Gross Motor Function Classification System (GMFCS)



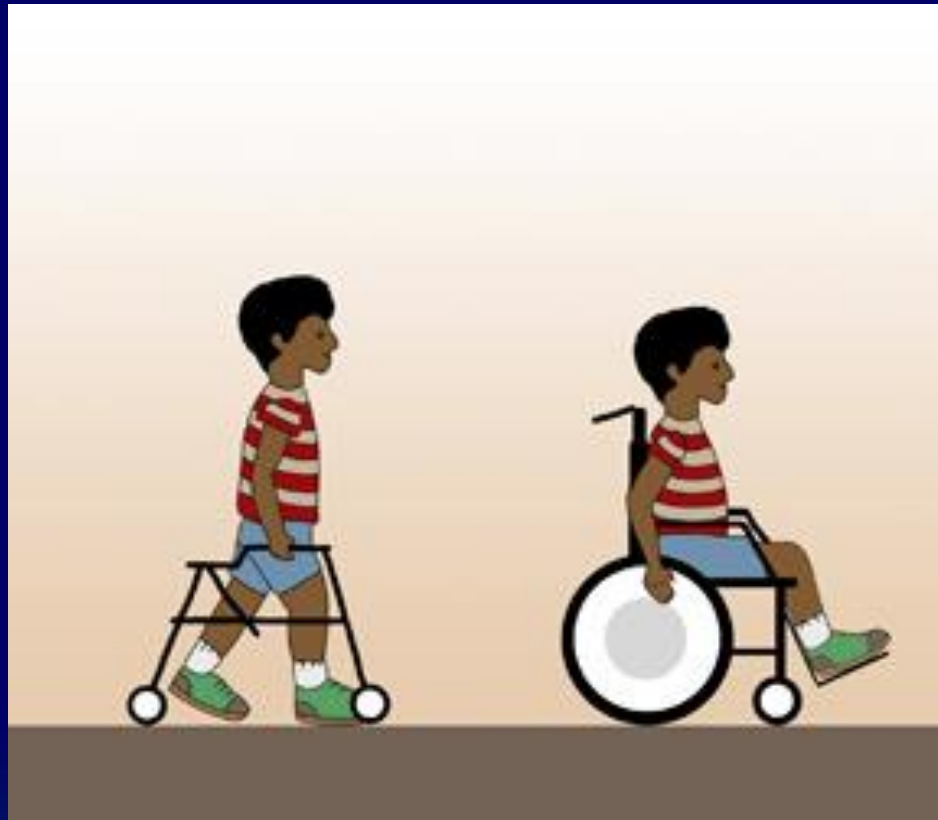
✓ Level II



Gross Motor Function Classification System (GMFCS)



✓ Level III



Gross Motor Function Classification System (GMFCS)



✓ Level IV



Gross Motor Function Classification System (GMFCS)



✓ Level V

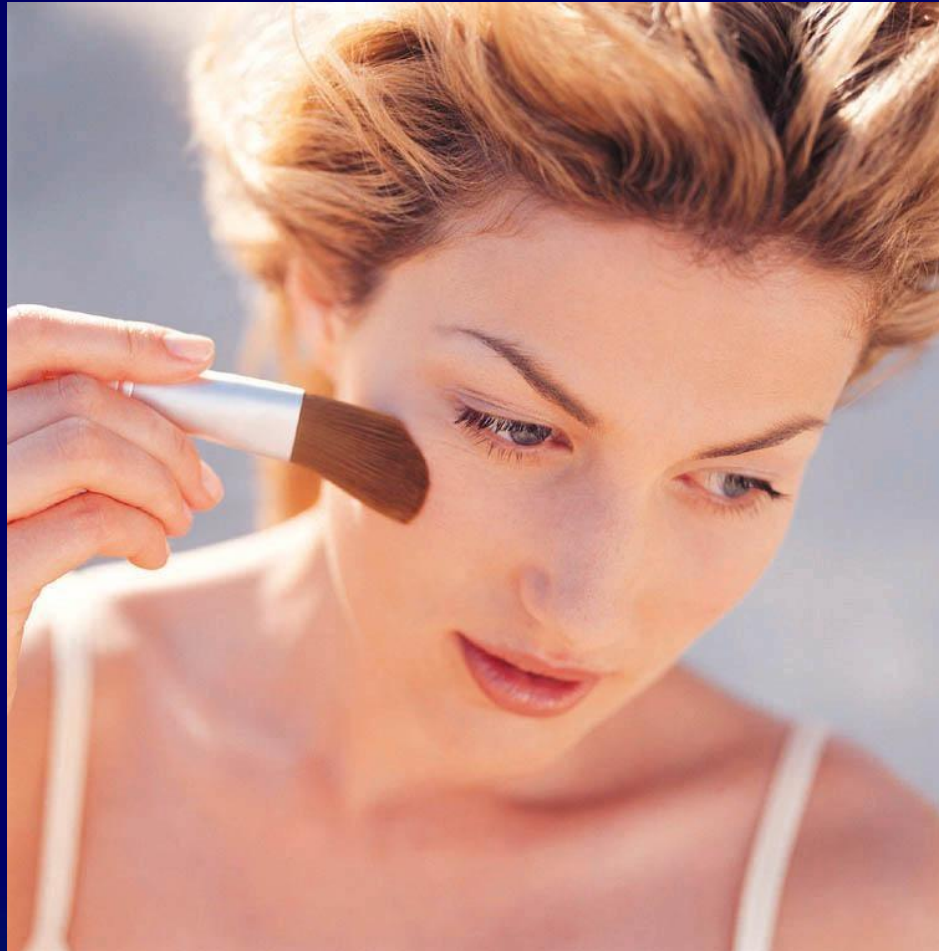


Treatment principles in Cerebral Palsy

- ✓ Primary treatment – Neurological
 - Oral medication
 - Selective dorsal rhizotomy
 - Baclofen pump
 - Botulinum toxin treatment
- ✓ Secondary treatment – Orthopaedic
 - Correction of the biomechanics
 - Correction of the anatomy

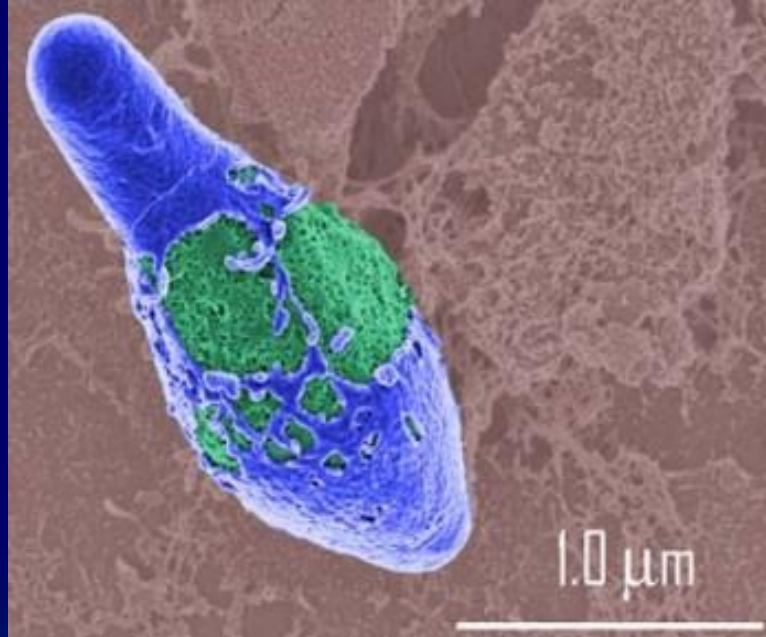
BOTULINUM TOXIN

Botulinum Toxin



Botulinum Toxin

- ✓ *Clostridium botulinum*
- ✓ Botulism (sausage intoxication)
- ✓ 7 serotype (A, B, C, D, E, F, G)



History of clinical applications

- ✓ 1973 – First applications in animals
- ✓ 1977, Scott et al. – Strabismus treatment
- ✓ 1980 – Strabismus, facial spasm, cervical dystonia
- ✓ 1987, Koman et al. – Cerebral palsy
- ✓ 1992, 1993 – Cosmetic applications

First clinical results in Cerebral Palsy



Koman LA, Mooney JF, Smith B et al. Management of cerebral palsy with botulinum A toxin: Preliminary investigation. J Pediatr Orthop 1993; 13: 489 – 495.

Indications of Botulinum Toxin

Strabismus

Cervical dystonia

Oromandibular dystonia

Blepharospasm

Hemifascial spasm

Cosmetic

Spasmodic dysphonia

(vocal cord spasm)

Specific dystonia

(writer's cramp)

Spasticity

Achalasia

Chronic anal fissures

Anal sphincter spasm

Headaches

(Migraine and cluster type)

Hyperhidrosis

Low back pain

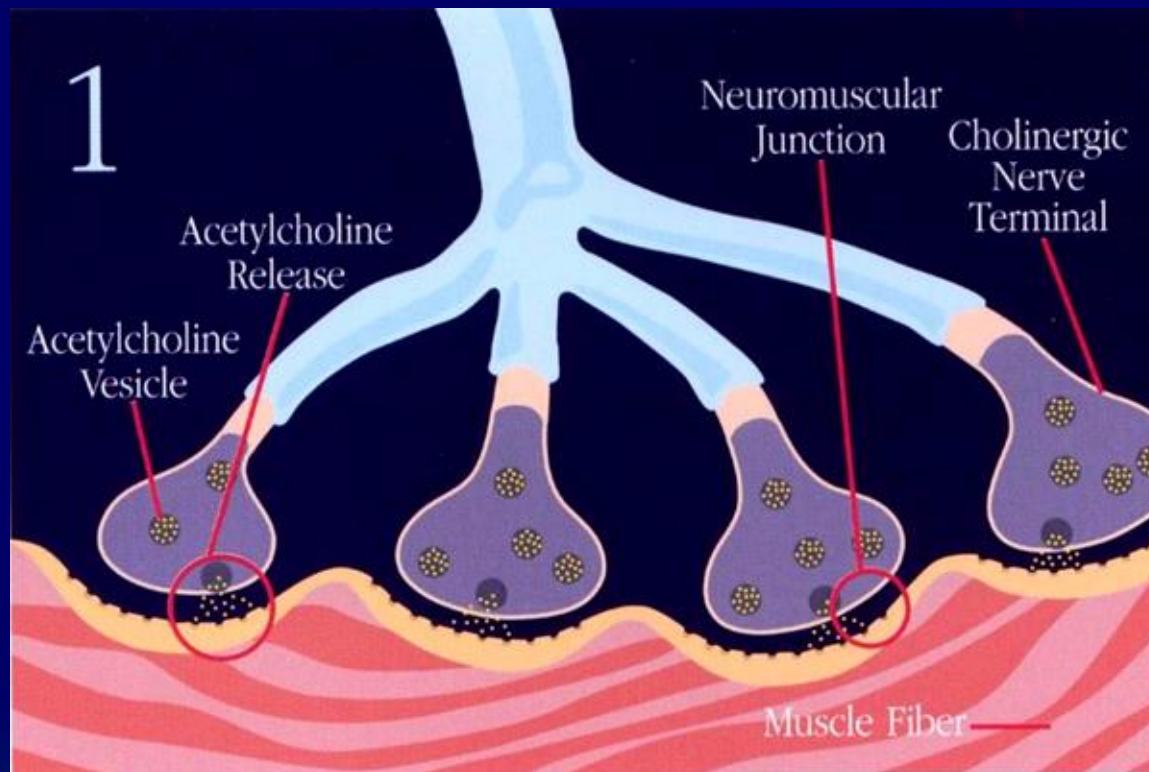
Myofascial pain syndrome

Tic treatment

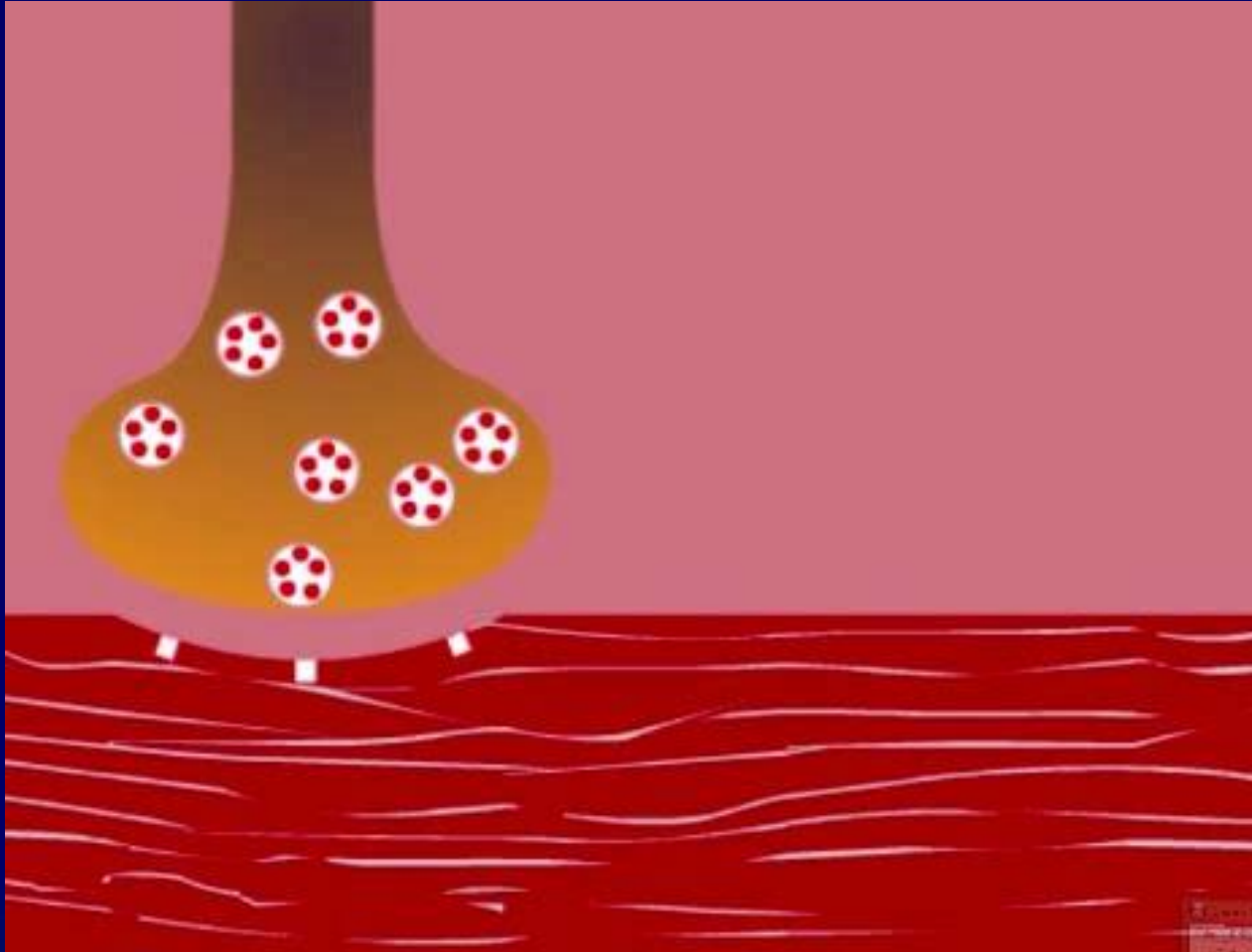
Spastic bladder and sphincters

The mechanism of effect

- ✓ Inhibits acetylcholine release at the neuromuscular junction



The mechanism of effect



The goals of Botulinum Toxin treatment



Weaken the group of spastic muscles

Control of spasticity

Prevent the development of static deformity
(contracture)

The goals of Botulinum Toxin treatment

Weaken the spastic (agonistic) muscle
Strengthening antagonistic muscle



BALANCE OF MUSCLE FORCES

The goals of Botulinum Toxin treatment (oriented to the joints)

- ✓ Prevent development of deformities
- ✓ Increase active and passive joint range of motions
- ✓ Facilitate physical therapy
- ✓ Reduction of the number and complexity of future surgery; delay surgery until the optimal timing is achieved
- ✓ Increase the compliance to orthotics
- ✓ Reduction of pain due to spasticity
- ✓ Low energy consumption during gait

Except spasticity ...

- ✓ Provide perianal, axillar and palmar hygiene
- ✓ Facilitate wearing and mobility
- ✓ Provide correct sitting posture
- ✓ Reduction of using systemic medications
- ✓ Hyperlacrimation (sialore) treatment
- ✓ Torticollis, shoulder dislocation, scoliosis etc.

Evaluation of patient

- ✓ Severity of disease
- ✓ Spasticity
- ✓ Functional capacity
- ✓ Life quality of patient and family
- ✓ Socioeconomic status of patient, family and social security system
- ✓ Intended surgeries

Appropriate patient

- ✓ Dynamic deformity
- ✓ Pain due to spasticity
- ✓ Pain after surgery or cast applications
- ✓ Deformity secondary to muscle imbalance

Inappropriate patient

- ✓ Presence of contracture or static deformity
- ✓ Allergy and hypersensitivity
- ✓ Severe muscle weakness
- ✓ Resistant patients to the previous injections
- ✓ Patients used aminoglycoside, curare, calcium channel blockers

Inappropriate patient

- ✓ Incompetible patient and family ???
- ✓ Age ???
 - ✓ < 2 years and > 6 years for lower extremity
 - ✓ < 4 years for upper extremity

Terminology in application

Body weight (kg)

Concentration (U/cc)

Total dose (U/kg)

Dose per muscle (U/muscle)

LD50 – Lethal dose

Amount of injection sites per muscle

Agonist/antagonist muscle

Total dose can differ depending on...

- ✓ The size of the injected muscle
- ✓ Body weight of the patient
- ✓ Number of neuromuscular junctions in the muscle
- ✓ Formulation of the toxin and injected volume
- ✓ Acquired tolerance

Attention for lethal dose !!!

- ✓ The most potent toxin !!!
- ✓ Lethal dose for human ???
- ✓ In primats LD50 39-56 U/kg



Commercial forms

✓ Botulinum toxin A

- ✓ Botox
- ✓ Dysport
- ✓ Xeomin



✓ Botulinum toxin B

- ✓ Myobloc/Neurobloc



Commercial forms



	BOTOX	DYSPORT
Serotype	A	A
Company	Allergan (US)	Ipsen (UK)
Efficacy (in 1 unit)	3 - 5	1
Equivalent dose	1	3 – 5
Vial containing	100 U toxin	500 U toxin
Preparation	Dilution with 0.9 % NaCl	Dilution with 0.9 % NaCl
Maximum usability	24 months	12 months
Storage	2 - 8°C	2 - 8°C
Exposure time after dilution	4 hours	8 hours

Dose ranges – Upper extremity

Clinical problem	Muscles injected	Dose range (U/kg)	Amount of injection sites per muscle
Shoulder adduction and internal rotation	Pectoralis Latissimus dorsi Teres major Subscapularis	1-2	1-3
Elbow flexion	Brachioradialis Biceps Brachialis	1-2	1-4
Forearm pronation	Pronator quadratus Pronator teres	0.5-2	1
Wrist flexion	Flexor carpi radialis Flexor carpi ulnaris	1-2	1
Thumb in palm	Flexor pollicis longus Adductor pollicis Flexor pollicis brevis/opponens	0.5-1	1
Fist position	Flexor digitorum profundus Flexor digitorum superficialis	1-2	1-2
Intrensec plus position	Lumbrical/interosseous	0.5-1	1

Dose ranges – Lower extremity

Clinical problem	Muscles injected	Dose range (U/kg)	Amount of injection sites per muscle
Hip flexion	iliacus Psoas Rectus femoris	2-6	1-3
Hip adduction	Adductors	2-4	1-3
Knee flexion	Medial hamstrings Lateral hamstring	2-6	2-3
Knee extension	Quadriceps	3-6	1-3
Equinovarus deformity	Gastrocnemius	3-6	2-5
	Soleus	1-3	1-2
	Tibialis posterior		
	Tibialis anterior		
	Flexor digitorum longus/brevis Flexor hallucis longus		
Parallel hallux	Extensor hallucis longus	1-2	1

Guide for dose in children

- ✓ Above 2 years old (between 10-30 kg)
 - ✓ Total maximum dose – 26 U/kg
 - ✓ Large muscle groups – 3-6 U/kg (maximum dose per muscle 100 U)
 - ✓ Small muscle groups – 1-2 U/kg (maximum dose per muscle 50 U)
 - ✓ Maximum dose/injection site – 50 U
 - ✓ Repeat of injection $\geq$ 3 months

Graham HK et al., Gait Posture 2000; 11:67-79.

Dose modifiers

	Dose can be decreased	Dose can be increased
Body weight	Low	High
Possible duration of treatment	Chronic	Acute
Muscle bulk	Very small	Very large
Number of injected muscles simultaneously	Many	One or a few
Ashworth score	Low	High
Concern about muscle weakness	High	Low
Results of previous therapy	Too much weakness	Inadequate response

Application of Botulinum Toxin

- ✓ Intramuscular injection to the spastic muscle
 - ✓ Electromyography
 - ✓ CT/MRI
 - ✓ USG
 - ✓ Palpation



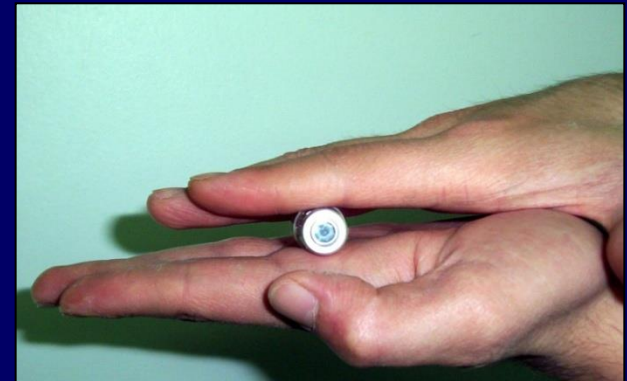
Application of Botulinum Toxin

- ✓ **Palpation is enough...**
 - Graham HK, Gait and Posture, 2000
 - Molenaers G, J Bone Joint Surg Am. 2006
- ✓ **USG is required...**
 - Berweck S, Neuropediatrics, 2002
 - Willenborg MJ, J Pediatr Orthop, 2002
- ✓ **Electromyography is required...**
 - Chin TY, J Pediatr Orthop. 2005
- ✓ **If applied appropriate, all methods are acceptable...**
 - Schroeder AS, Neurotox Res. 2006

Application of Botulinum Toxin

- ✓ Cold chain !!!
- ✓ Dilution with 1, 2, 4 or 5 cc. saline (%0.9 NaCl)
- ✓ Be carefull for denaturation

Don't shake the vial !!!



Application of Botulinum Toxin

✓ Insulin syringe (1cc/1ml)



✓ 22 gauge needle



Application of Botulinum Toxin

- ✓ Repeat physical examination under general anesthesia



Application of Botulinum Toxin

✓ Sterility is important !



Application of Botulinum Toxin



Application of Botulinum Toxin

- ✓ Lower extremity



Application of Botulinum Toxin

- ✓ Lower extremity



Application of Botulinum Toxin

- ✓ Lower extremity



Application of Botulinum Toxin

- ✓ Upper extremity



Must keep in mind...

- ✓ Antispastic effect appears within 12 to 72 hours
- ✓ Becomes maximum at 10 to 14 days
- ✓ Effect lasts for 3 to 6 months
- ✓ Can be repeated after 3 months
- ✓ No more than 6 times injection at the same site

Postinjection follow-up

- ✓ Physical therapy
- ✓ Casting
- ✓ Orthotics



Response to toxin can differ depending on...

- ✓ Presence of contacture
- ✓ Severity of spasticity
- ✓ Amount of blocked neuromuscular junction
- ✓ Status of antagonistic muscles
- ✓ Physical rehabilitation protocol

Side effects of Botulinum Toxin

- ✓ Temporary muscle weakness
- ✓ Pain, swelling or rash on injection area
- ✓ Infection
- ✓ Influenza-like symptoms
- ✓ Autonomic dysfunction like incontinence or constipation

ORTHOPAEDIC SURGERY

Orthopaedic surgical procedures

- ✓ Lengthen contracted myotendinous units
- ✓ Balance joint forces
- ✓ Transfer motor power
- ✓ Fuse unstable joints by arthrodesis
- ✓ Correct bony deformity to improve biomechanical alignment
- ✓ Reduce joint subluxation and dislocation to improve joint congruency
- ✓ Diminish painful spasticity
- ✓ Maintain, restore or stabilise spinal deformity

Secondary treatment - Orthopaedics

- ✓ Correction of the biomechanics
 - ✓ Muscle-tendon lengthening and transfers
 - Single even multilevel surgery (SEMLS) in children after 6 years of age
 - Minimize the hospitalization time and duration of rehabilitation

Secondary treatment - Orthopaedics

- ✓ Correction of the biomechanics

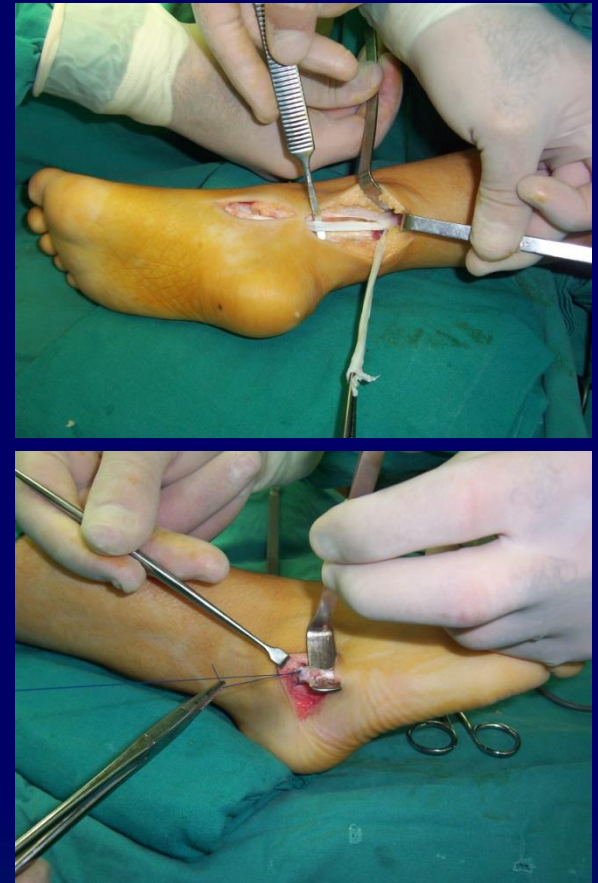
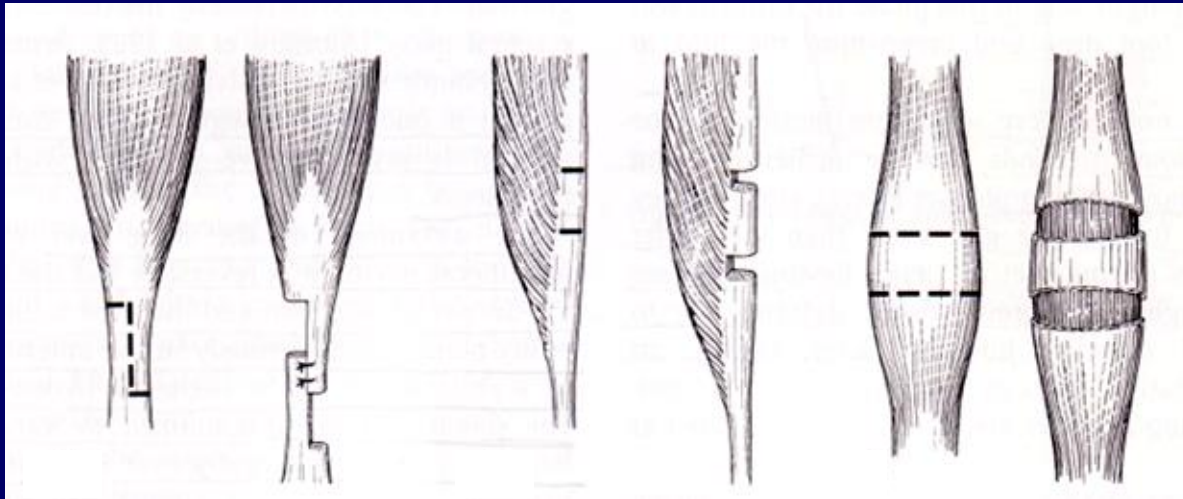
- ✓ Lever arm dysfunction

Corrective osteotomies for foot, leg and pelvis

Additional to soft tissue procedures and orthosis for lower extremity

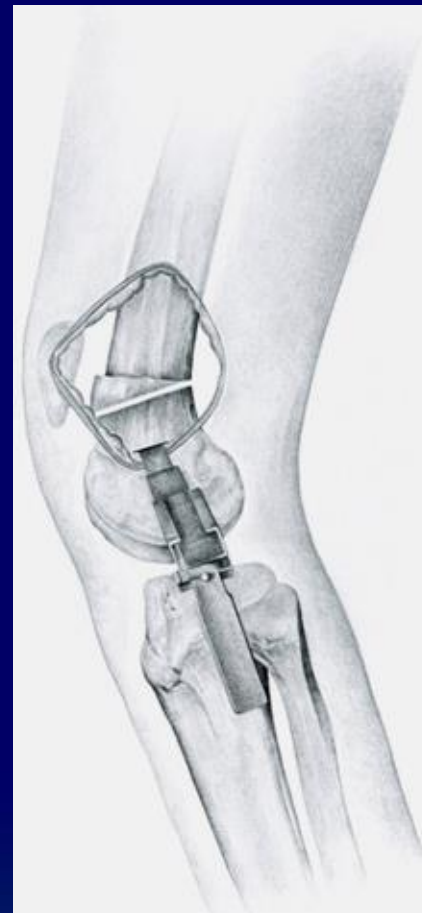
Secondary treatment - Orthopaedics

✓ Correction of the biomechanics



Secondary treatment - Orthopaedics

✓ Correction of the biomechanics



Secondary treatment - Orthopaedics

- ✓ Correction of the biomechanics



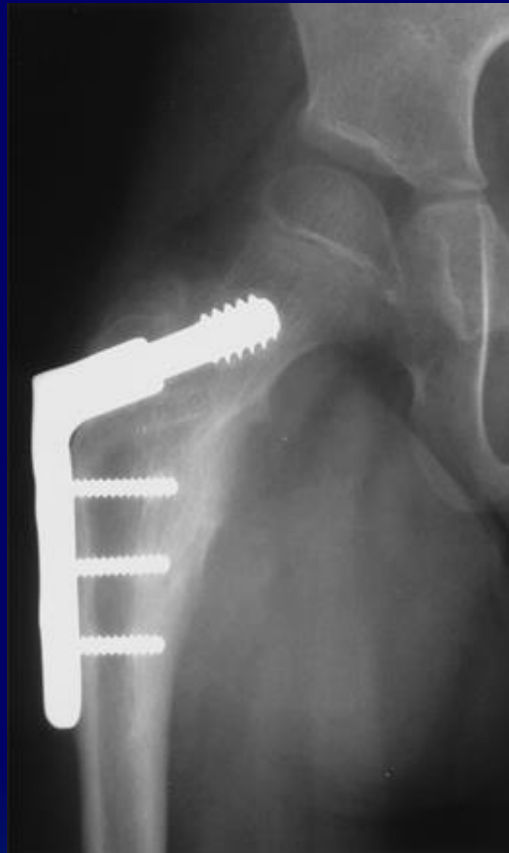
Secondary treatment - Orthopaedics

- ✓ Correction of the biomechanics



Secondary treatment - Orthopaedics

- ✓ Correction of the biomechanics



Secondary treatment - Orthopaedics

- ✓ Correction of the biomechanics



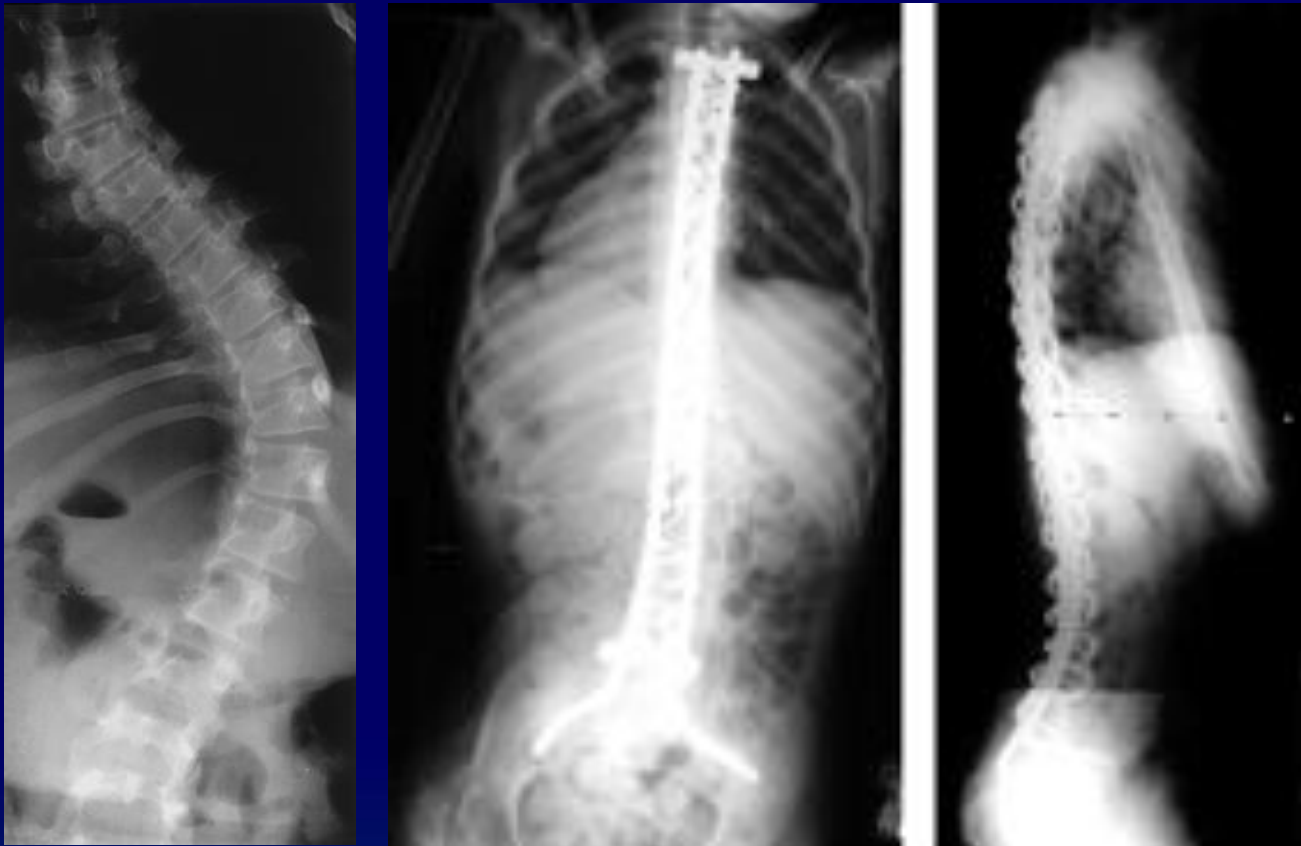
Secondary treatment - Orthopaedics

- ✓ Correction of the anatomy
 - ✓ Mostly for quadriplegic, sometimes for diplegic patients
 - ✓ Scoliosis
 - ✓ Hip dislocation
 - ✓ Joint contractures
 - ✓ Foot problems



Secondary treatment - Orthopaedics

- ✓ Correction of the anatomy

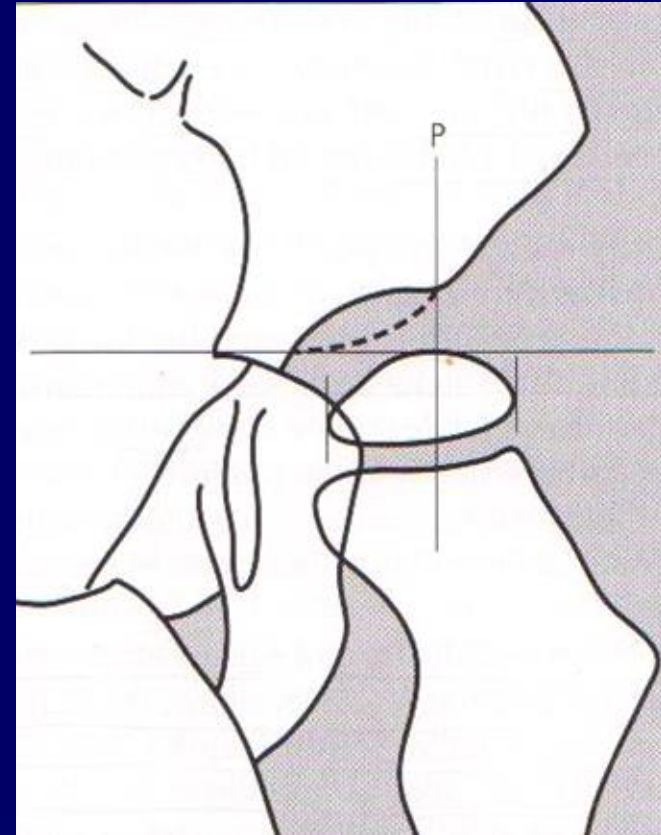


Secondary treatment - Orthopaedics

- ✓ Correction of the anatomy
 - ✓ Hip at risk
 - ✓ Severe spasticity
 - ✓ Scissoring gait
 - ✓ Inability to stand and walk independently
 - ✓ $< 30^\circ$ hip abduction, when hips extended
 - ✓ $> 20^\circ$ hip flexion contracture

Secondary treatment - Orthopaedics

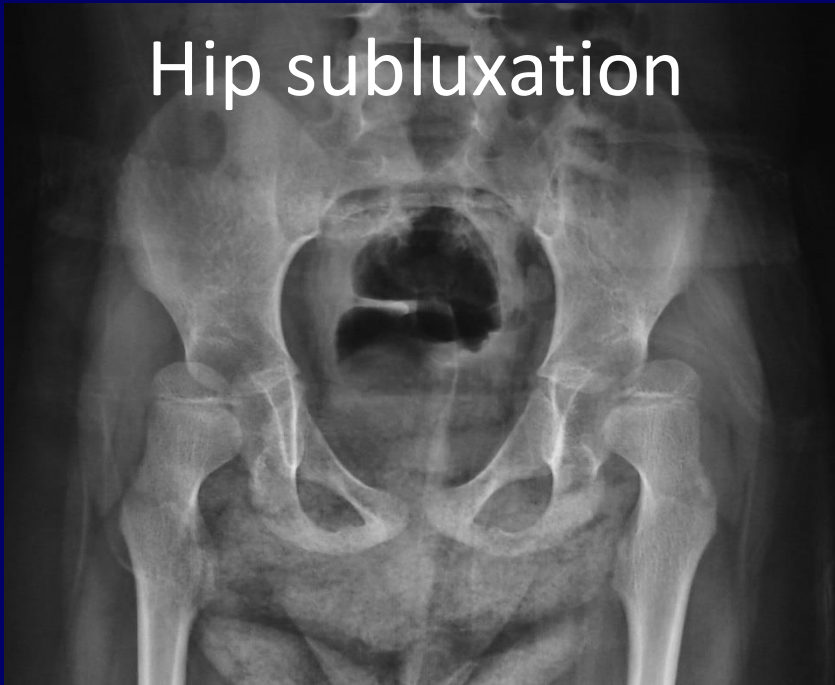
- ✓ Correction of the anatomy
 - ✓ Follow-up of hip at risk
 - ✓ Pelvis AP radiograph at 6 months interval
 - ✓ Reimer instability (migration) index



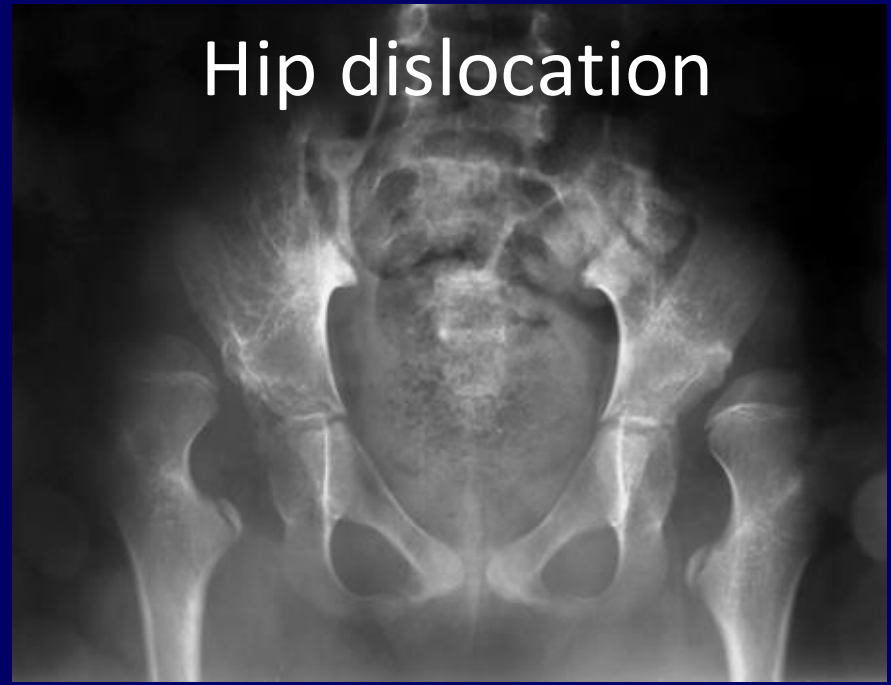
Secondary treatment - Orthopaedics

- ✓ Correction of the anatomy

Hip subluxation



Hip dislocation



Secondary treatment - Orthopaedics

- ✓ Correction of the anatomy



Secondary treatment - Orthopaedics

- ✓ Correction of the anatomy



Upper extremity surgery

- ✓ Ideal surgical candidate
 - ✓ Voluntary hand use
 - ✓ Good sensation
 - ✓ Intelligent
 - ✓ No athetosis
- ✓ Surgery between 6 – 12 years of age
- ✓ Mostly hemiplegic, some times quadriplegic patients

Upper extremity surgery

- ✓ Shoulder
 - ✓ Adduction – internal rotation
 - ✓ Lengthening
 - ✓ Derotational osteotomy
- ✓ Elbow
 - ✓ Flexion contracture greater than 60°
 - ✓ Biceps lengthening

Upper extremity surgery

✓ Forearm

✓ Pronator spasticity or contracture

- ✓ Transfer of pronator teres to the supinator
- ✓ Pronator release
- ✓ Release of biceps aponeurosis

✓ Wrist

✓ Flexion and ulnar deviation

- ✓ Flexor lengthening
- ✓ Tendon transfer
- ✓ Proximal row carpectomy
- ✓ Wrist fusion

Upper extremity surgery

- ✓ Finger
 - ✓ Flexor spasticity or contracture; sometimes swan-neck deformity
 - ✓ Flexor-pronator origin release or lengthening
- ✓ Thumb
 - ✓ Thumb-in-palm deformity
 - ✓ Release adductor and flexor tendons

Botulinum Toxin vs Surgery

✓ Similarities

- ✓ Improvement in gait pattern
- ✓ Multilevel treatment
- ✓ Adjunctive management
- ✓ Multidisciplinary team involvement

Molenars G et al. Single event multilevel botulinum toxin type A treatment and surgery: similarities and differences. Eur J Neurol 2001; 8(Suppl 5): 88 – 97.

Botulinum Toxin vs Surgery

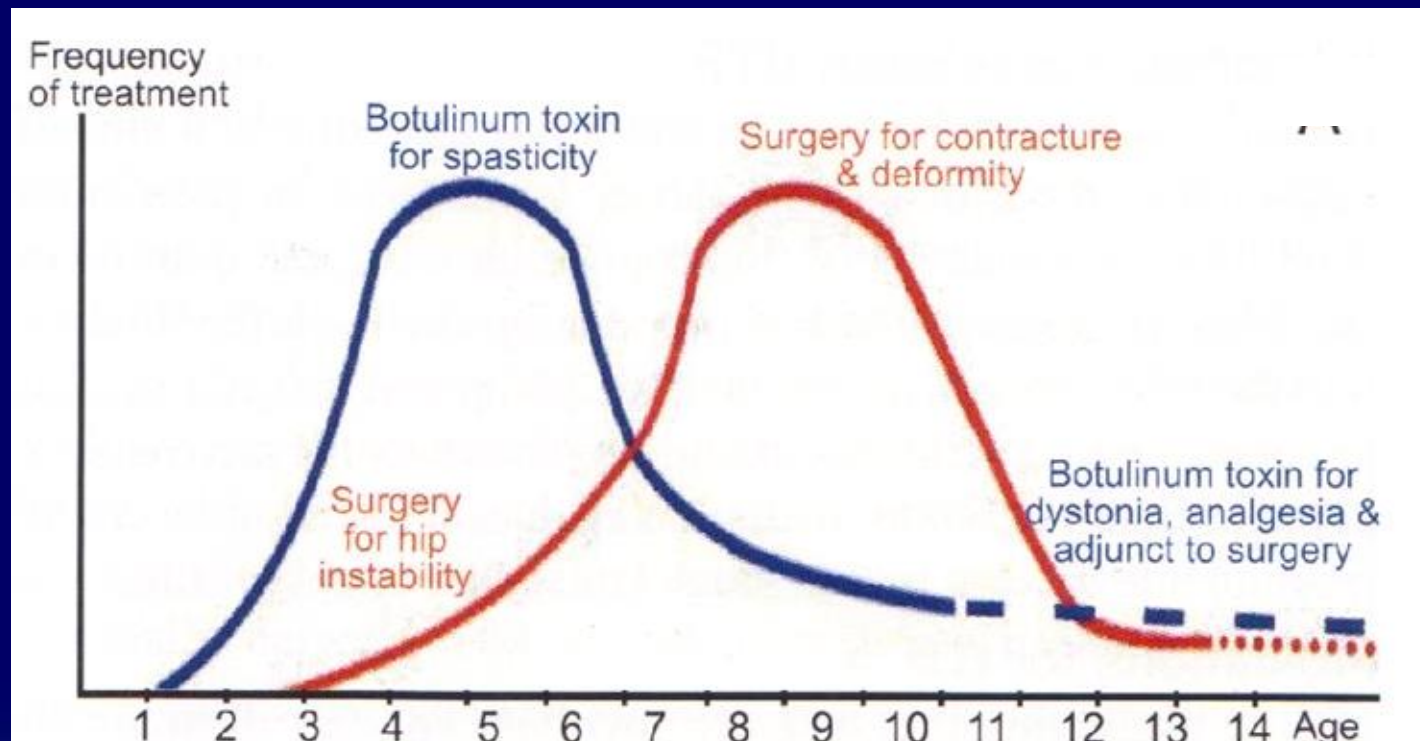
✓ Differences

- ✓ Age at time of intervention
- ✓ Pretreatment status
- ✓ Repeated applications
- ✓ Reversibility

Molenars G et al. Single event multilevel botulinum toxin type A treatment and surgery: similarities and differences. Eur J Neurol 2001; 8(Suppl 5): 88 – 97.

Summary

- ✓ Multilevel treatment at an early age; especially with Botulinum toxin for spasticity



Yalçın S, Berker N. The help guide to cerebral palsy. Istanbul, 2005.

Summary

- ✓ Prevention is better than cure
- ✓ Combined with physiotherapy, casting and orthotics
- ✓ Prevent, delay, fine-tune and/or minimize the surgical intervention

Don't forget...

- ✓ Multidisciplinary team evaluation and working
 - ✓ Physical rehabilitation
 - ✓ Pediatric neurology
 - ✓ Neurosurgical consultations
 - ✓ Ophtalmology
 - ✓ Psychiatry
 - ✓ Hand rehabilitation
 - ✓ Occupational therapies

Thank you...

