Hypertonia Management for Non-ambulatory Cerebral Palsy : GMFCS level IV-V

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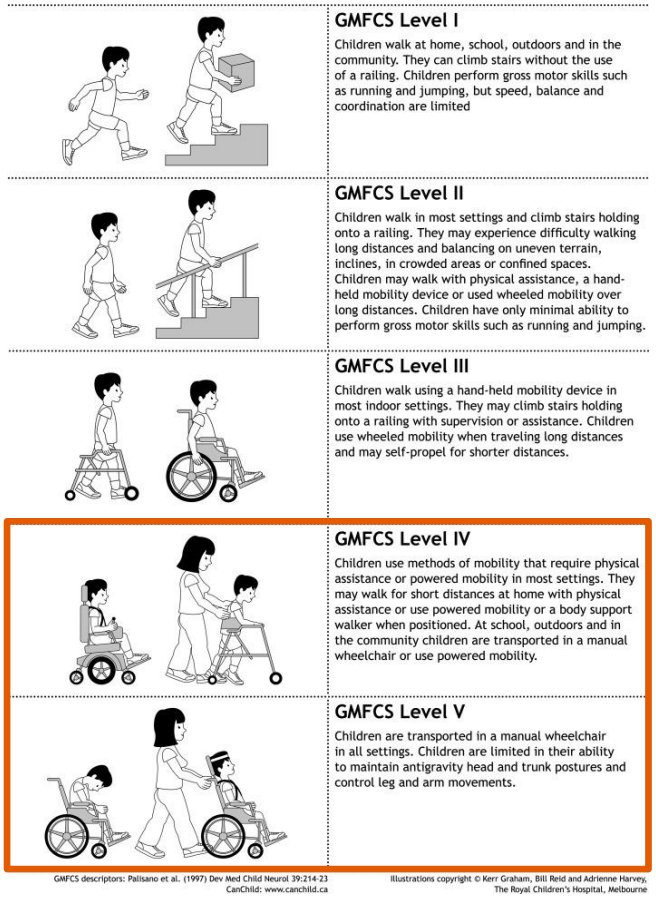
Treatment Goals for Cerebral Palsy

- Well defined and tailored to individual needs
- Adapted to specific problems

- **GMFCS I-III**
  - Focus on improving function (gait)
  - Influence pathological process

- **GMFCS IV-V**
  - Improve balance, control of sitting
  - Positioning, facilitating hygienic care
  - Easy for brace



GMFCS Level I

Children walk at home, school, outdoors and in the community. They can climb stairs without the use of a railing. Children perform gross motor skills such as running and jumping, but speed, balance and coordination are limited

GMFCS Level II

Children walk in most settings and climb stairs holding onto a railing. They may experience difficulty walking long distances and balancing on uneven terrain, inclines, in crowded areas or confined spaces. Children may walk with physical assistance, a hand-held mobility device or used wheeled mobility over long distances. Children have only minimal ability to perform gross motor skills such as running and jumping.

GMFCS Level III

Children walk using a hand-held mobility device in most indoor settings. They may climb stairs holding onto a railing with supervision or assistance. Children use wheeled mobility when traveling long distances and may self-propel for shorter distances.

GMFCS Level IV

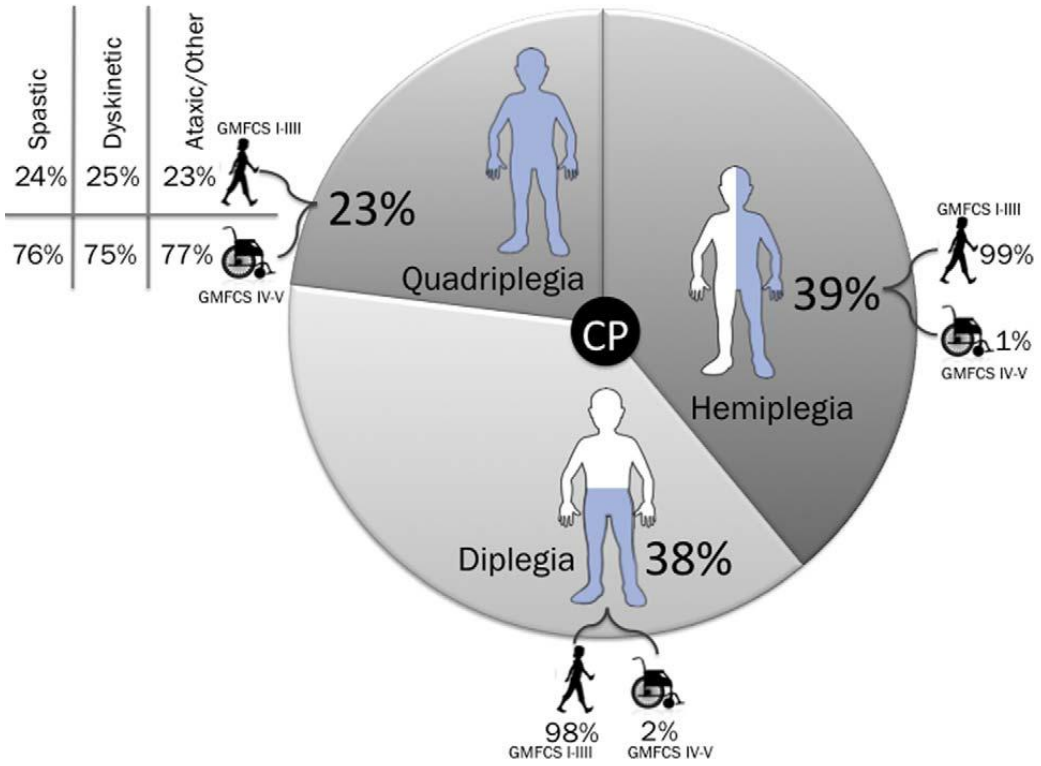
Children use methods of mobility that require physical assistance or powered mobility in most settings. They may walk for short distances at home with physical assistance or use powered mobility or a body support walker when positioned. At school, outdoors and in the community children are transported in a manual wheelchair or use powered mobility.

GMFCS Level V

Children are transported in a manual wheelchair in all settings. Children are limited in their ability to maintain antigravity head and trunk postures and control leg and arm movements.

GMFCS descriptors: Palisano et al. (1997) Dev Med Child Neurol 39:214-23
CanChild: www.canchild.ca

Proportion of Cerebral Palsy by Topography and Severity



Evidence-based diagnosis, health care, and rehabilitation for children with cerebral palsy. Novak I1. J Child Neurol. 2014 Aug;29(8):1141-56.

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Annals of Rehabilitation Medicine

Korean Database of Cerebral Palsy: A Report on Characteristics of Cerebral Palsy in South Korea

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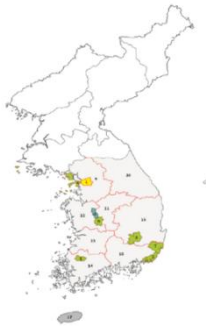
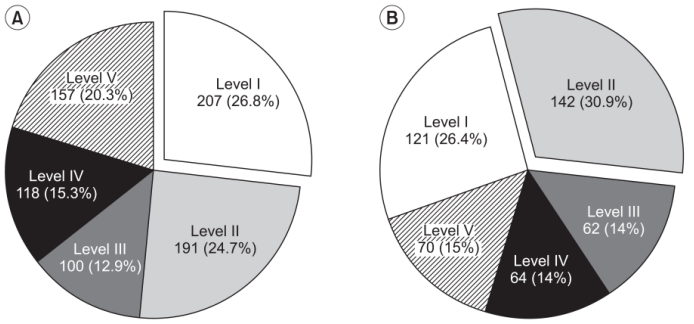


- Nationwide database of subjects with CP, a total of 773 subjects, Feb 2009 – Oct 2010
- Demography, birth history, onset and type of CP, brain magnetic resonance imaging(MRI) findings, functional ability, accompanying impairments

- Type of CP — Spastic (87.32%), Dyskinetic (5.17%), and Ataxic (1.81%)
- GMFCS level — I (26.8%), II (24.7%), III (12.9%), IV (15.3%) and level V (20.3%)
- MACS level — I (26.4%), II (30.9%), III (14%), IV (14%) and level V (15%)

Distribution of the subjects according to the type of CP

Type of cerebral palsy	Number of subjects (%)
Spastic type	675 (87.32)
Bilateral	505 (74.80)
Unilateral	164 (24.30)
Unclassified	6 (0.90)
Dyskinetic type	40 (5.17)
Dystonic	39 (97.50)
Choreoathetoid	0 (0)
Unclassified	1 (2.50)
Ataxic type	14 (1.81)
Unclassified	44 (5.70)
Total	773 (100)



Distribution of the subjects by the Gross Motor Functional Classification System (A; n+773) and the Manual Ability Classification Systems (B; n=459)

Problems in Non-ambulatory CP

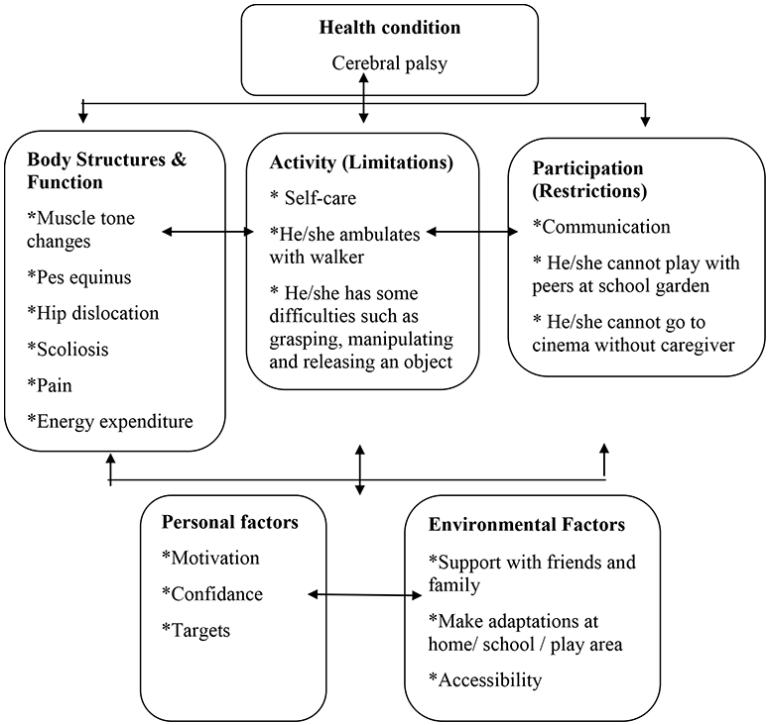
More problematic than ambulatory CP

- More severe brain damage
- Concurrent impairments

  (visual, hearing, cognition, Seizure..)
- Musculoskeletal problems

  (lever-arm dysfunction, dislocation,

  Scoliosis)
- Dysphagia, GE Reflux
- Pain



➡ Lower level on ICF-CY framework

Effect of Hypertonia in Musculoskeletal System

Altered Neurologic Function

↓

Increased Tone

↓

↓ Elasticity = ↓ Ability to Stretch to Full Range

↓

Altered Muscle Growth (↓ Length) } **Effects on Growing Bone**

↓

Contracture

Application of Abnormal Force to one

↓

A child's bone is:

'biologically' plastic

will change growth

'mechanically' plastic

will change shape

Altered Growth

- Torsion
- Angulation
- Length

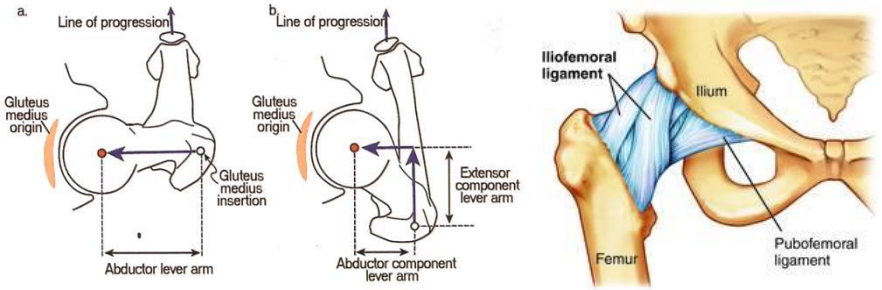
} **Lever arm dysfunction**

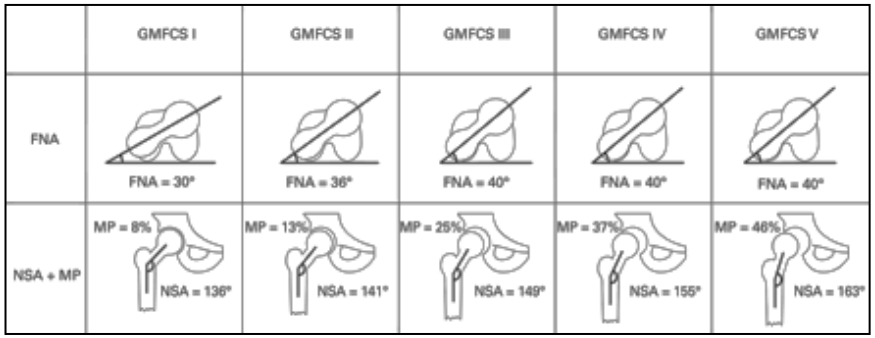
Malalignment of CP Musculoskeletal System

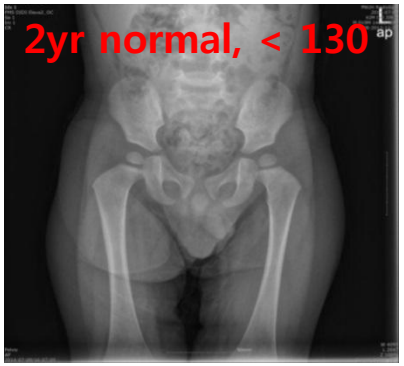
- **Femoral anteversion**
  - 30wks of gestation: 60°
  - Adulthood: 10-15°
  - Bigelow's ligament
    - reduce anteversion
    - extension of the hip joint

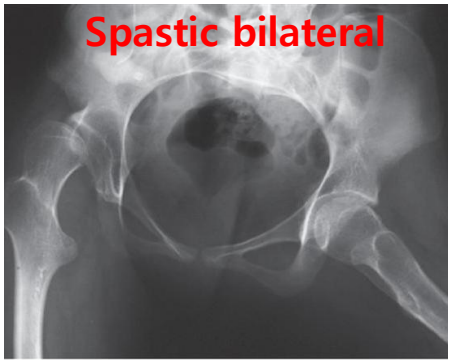
- **Coxa valga**
  - Neck-Shaft Angle
    - Birth: 135-140, Adult: 125-130

- **Muscle for coxa valga & hip D/L**
  - Iliopsoas
  - Hip adductors
  - Medial hamstrings

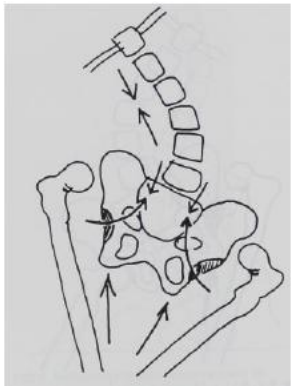








Musculoskeletal Deformity of Non-ambulatory CP

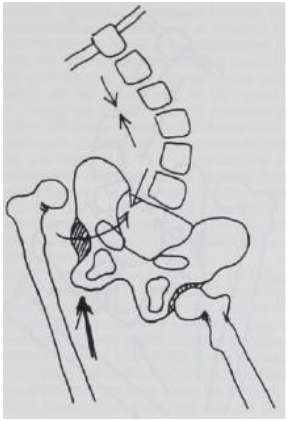


Type II: Pelvic tilt, Scoliosis, Bilateral hip dislocation

Symmetrical bilateral muscle imbalance around hip

Asymmetrical muscle tone at the trunk (persistent unilat. Galant reflex)

GMFCS V



Type III: Windswept syndrome

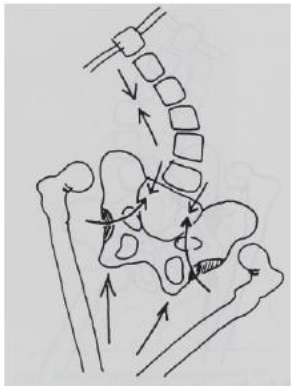
Scoliosis, pelvis tilt, and unilateral hip dislocation combination

Dislocated hip is located at the side of the concavity of the scoliosis

Bilateral CP with asymmetrical involvement of the body

Iliopsoas muscle cresses the pelvis rising at the lumbar spine

Musculoskeletal Deformity of Non-ambulatory CP



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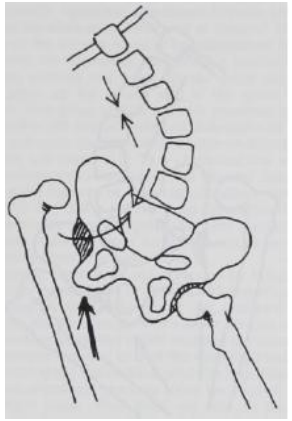


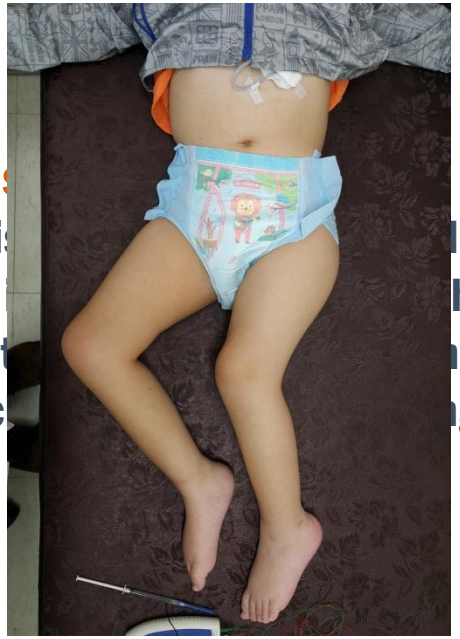
Type III: Winds

Scoliosis, pelvi…islocation combination

Dislocated hip …he concavity of the scoliosis

Bilateral CP wit…ent of the body

Iliopsoas musc…g at the lumbar spine



Incidence of Hip Displacement in Spastic CP

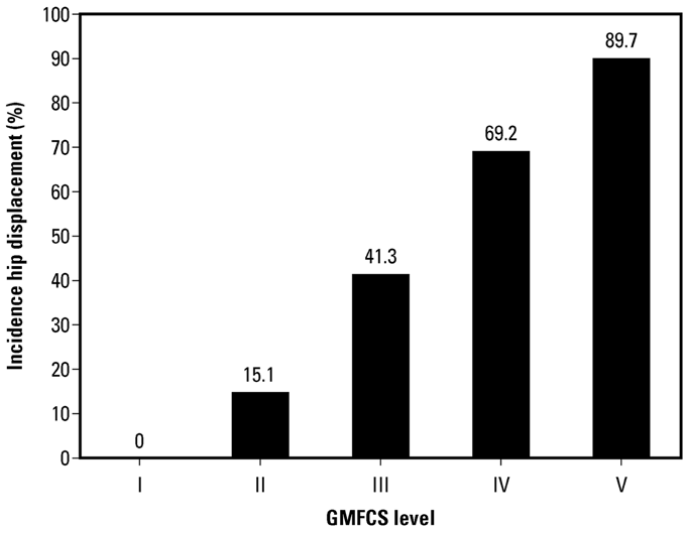
- 1% in spastic hemiplegia
- 5% in spastic diplegia
- **35% to 55% in spastic quadriplegia**

(Miller F; 1995, Dobson F; 2002)

- 323 (86%) of 374 children with CP
- Register for the birth years 1990~1992
- follow-up durations 7 years and 8 months
- Hip displacement **90% for GMFCS level V**

(Brendan S; 2006)

HIP DISPLACEMENT IN CEREBRAL PALSY



Incidence of hip displacement (a MP of > 30%) according to the GMFCS level

Incidence of Hip Displacement in Spastic CP

- 167 spastic CP patients
- MP increased in GMFCS levels

  I-III (0.3%/yr), IV (1.9%/yr) and V (6.2%/yr)
- GMFCS level IV,

  NSA increased significantly by 3.4° /yr *(P < 0.001)*

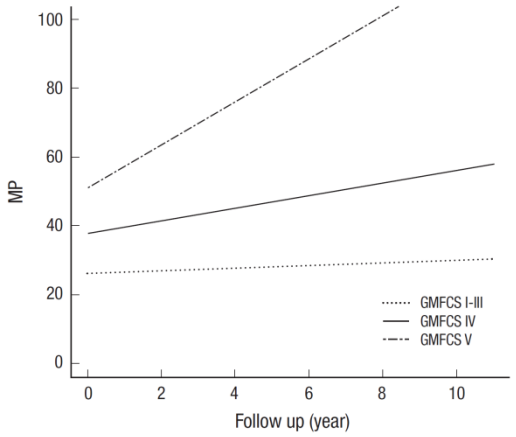


Table 3. The estimation of factors affecting MP using Linear Mixed Models

Factors	MP (GMFCS level I-III)			MP (GMFCS level IV)			MP (GMFCS level V)		
	Estimation	SE	*P* value	Estimation	SE	*P* value	Estimation	SE	*P* value
Intercept	27.3	1.5	< 0.001	45.5	4.0	< 0.001	51.7	5.0	< 0.001
Follow up	0.34	0.09	< 0.001	1.9	0.4	< 0.001	6.2	1.5	< 0.001
Sex	-3.3	1.6	0.048	-8.9	4.2	0.037	4.6	5.4	0.396

MP, migration percentage; GMFCS, gross motor function classification system; SE, standard error.

Table 4. The estimation of factors affecting NSA using Linear Mixed Models

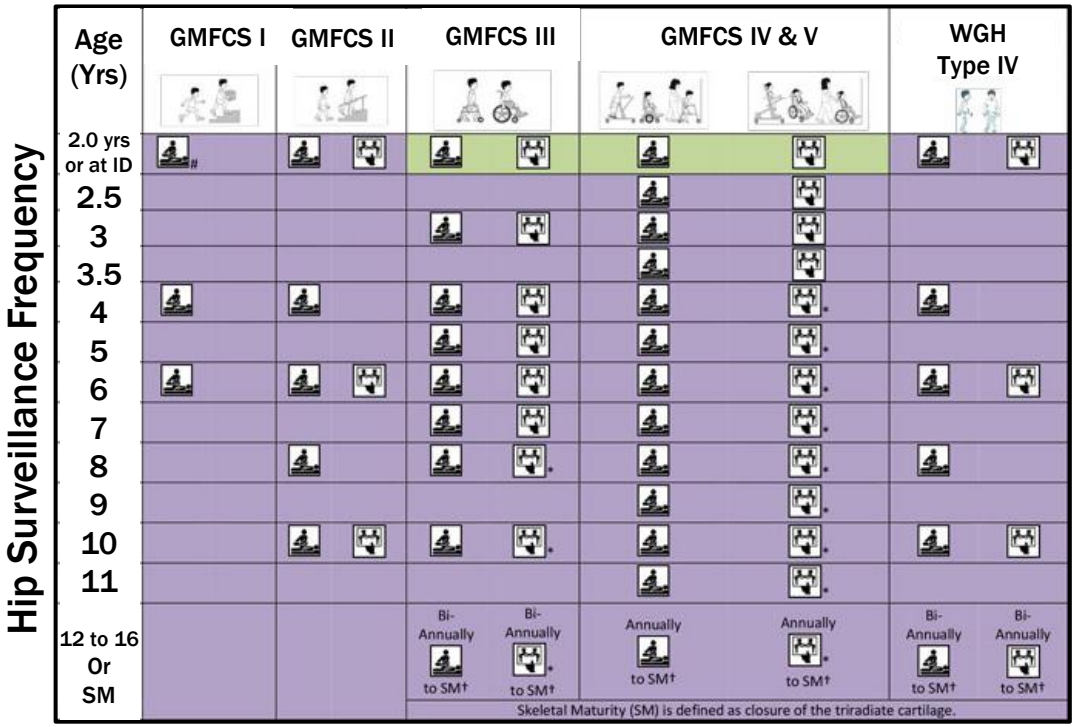
Factors	NSA (GMFCS level I-III)			NSA (GMFCS level IV)			NSA (GMFCS level V)		
	Estimation	SE	*P* value	Estimation	SE	*P* value	Estimation	SE	*P* value
Intercept	148.1	1.1	< 0.001	154.1	1.8	< 0.001	161.4	1.5	< 0.001
Follow up	0.05	0.15	0.755	3.4	0.8	< 0.001	1.1	0.7	0.1
Sex	-1.3	1.2	0.272	-0.7	1.9	0.711	-2.5	1.6	0.1

NSA, neck shaft angle; GMFCS, gross motor function classification system; SE, standard error.

(JY Park; 2016)

Hip Surveillance Guideline for Children with CP

- **In GMFCS level IV & V,**
  Annually clinical examination and AP Pelvis Radiograph until skeletal maturation (SM)



Clinical Examination AP Pelvis Radiograph

Review at 7y of age, if MP is stable, MP < 30% and gross motor function is stable,
-> GMFCS IV : discontinue until pre-puberty
GMFCS V : Continue 12-monthly

Independent of MP, when clinical and/or radiologic evidence of Scoliosis, pelvic obliquity is present
-> 6 monthly surveillance is required until SM

At skeletal maturity, if MP is abnormal, progressive scoliosis or significant pelvic obliquity is present
-> continue 12-monthly surveillance

Surveillance be continued after surgical intervention.

https://www.aacpdm.org/publications/care-pathways/hip-surveillance

Radiologic Evaluation of Hip

- **Forward or backward pelvic tilt needs to be corrected in children who have a fixed flexion deformity of the hip or a significant lumbar lordosis**

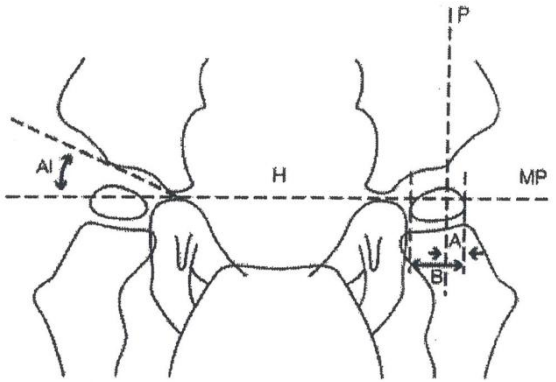


Fig. 4. Migration percentage MP = A/B X 100%.

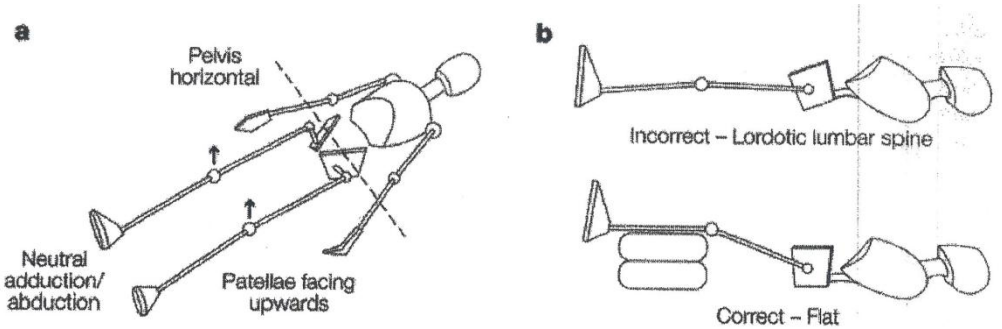


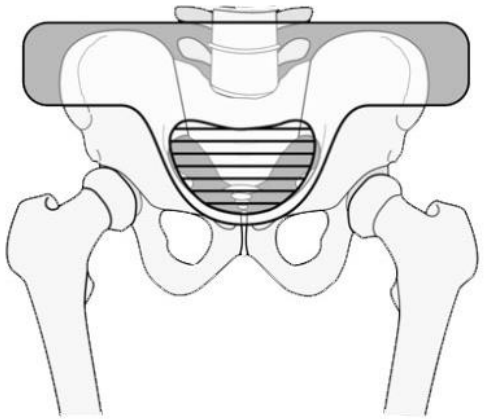
Fig. 5. Positioning for antero-posterior pelvic radiograph.

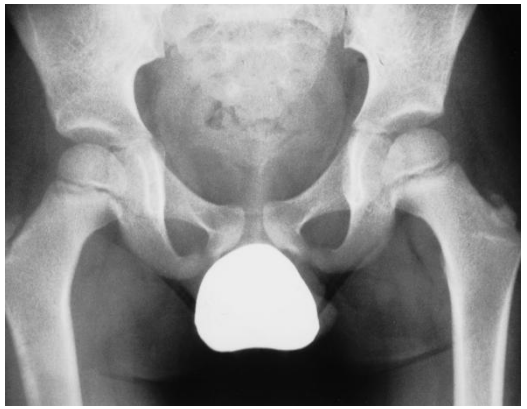
The consensus statement on Hip Surveillance for Children with Cerebral Palsy: Australian Standards of Care; 2011

Radiologic Evaluation of Hip

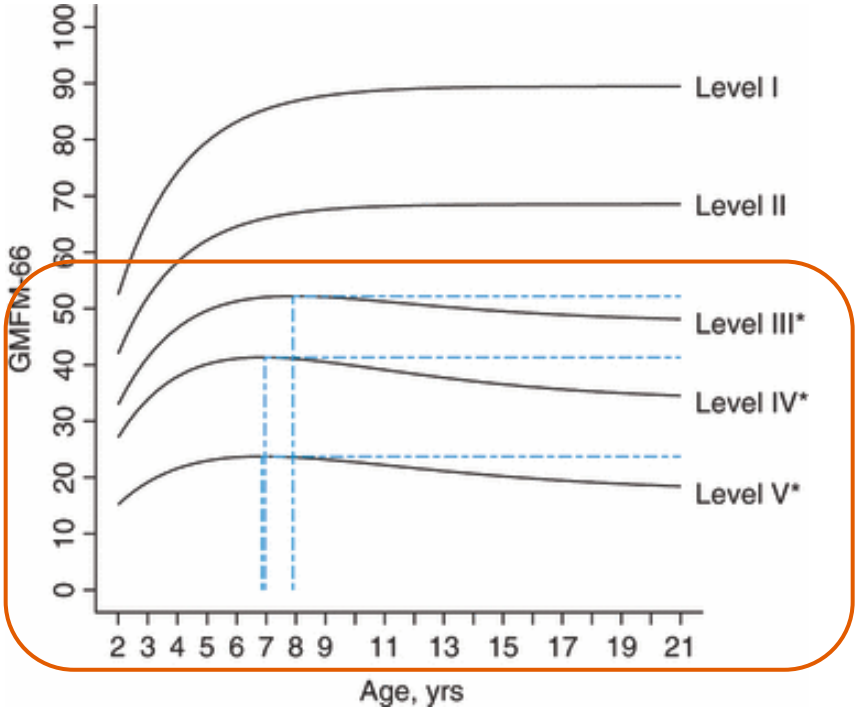
- Radiation from X-rays causes direct gonadal damage and mutation
- 0.26mGy to 2.89mGy for pelvic examination depending upon the body size
- No guideline for limitation of pelvis X-ray, dependent on clinical practice

Protect gonad from X-ray radiation !





Stable and Decline in Gross Motor Function in CP



Stability and decline in gross motor function among children and youth with cerebral palsy aged 2 to 21 years.
Hanna SE. Dev Med Child Neurol. 2009

- **GMFCS level III, IV, V**

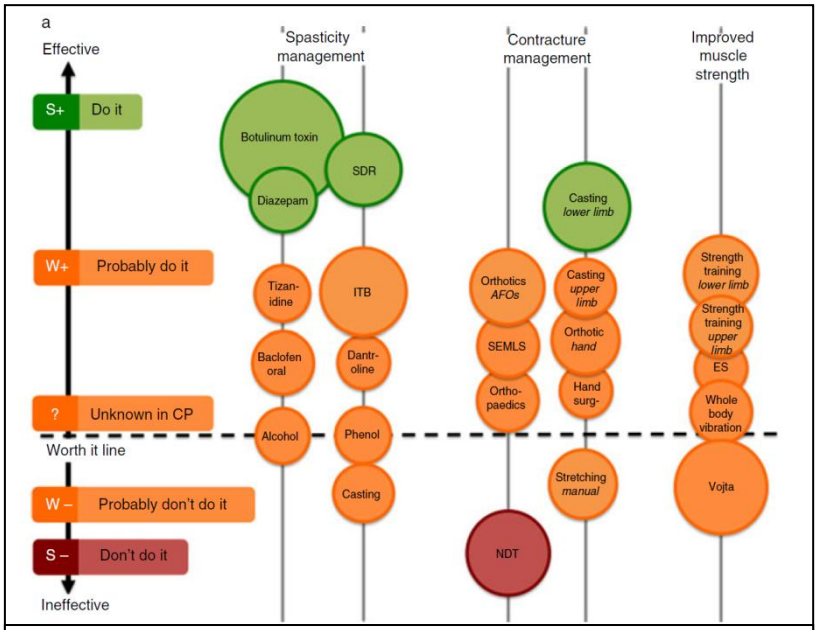
  Risk of losing motor function

- ✓ **Increased energy costs**
- ✓ **Contracture**
- ✓ **Muscle stiffness**
- ✓ **Spinal deformity**
- ✓ **Increasing imbalance**

Systematic Review of Intervention for CP

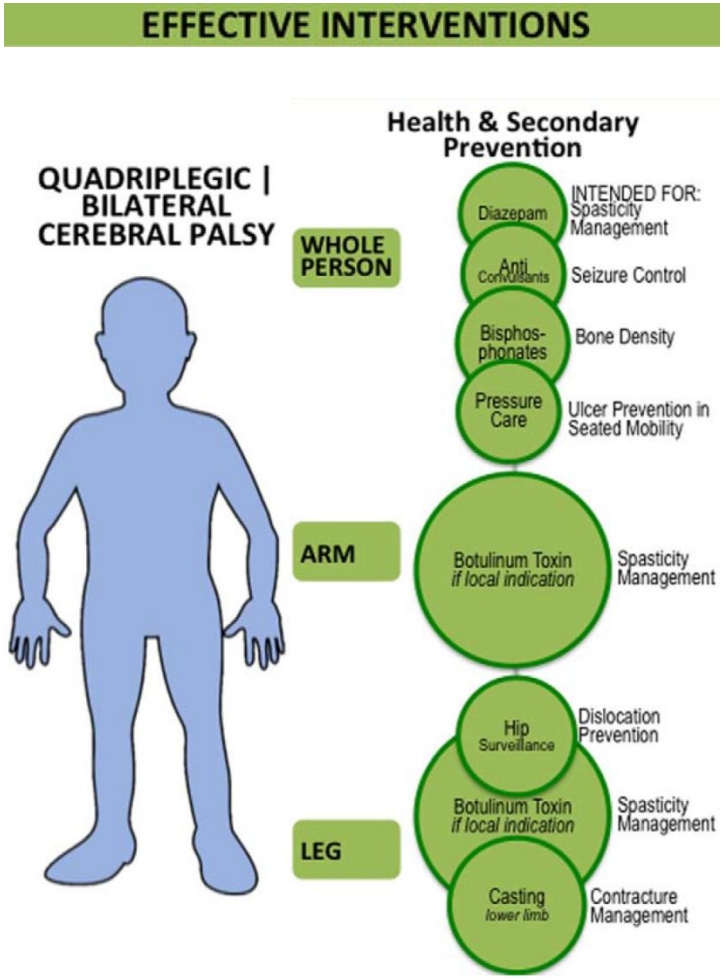
For body structures and functional level

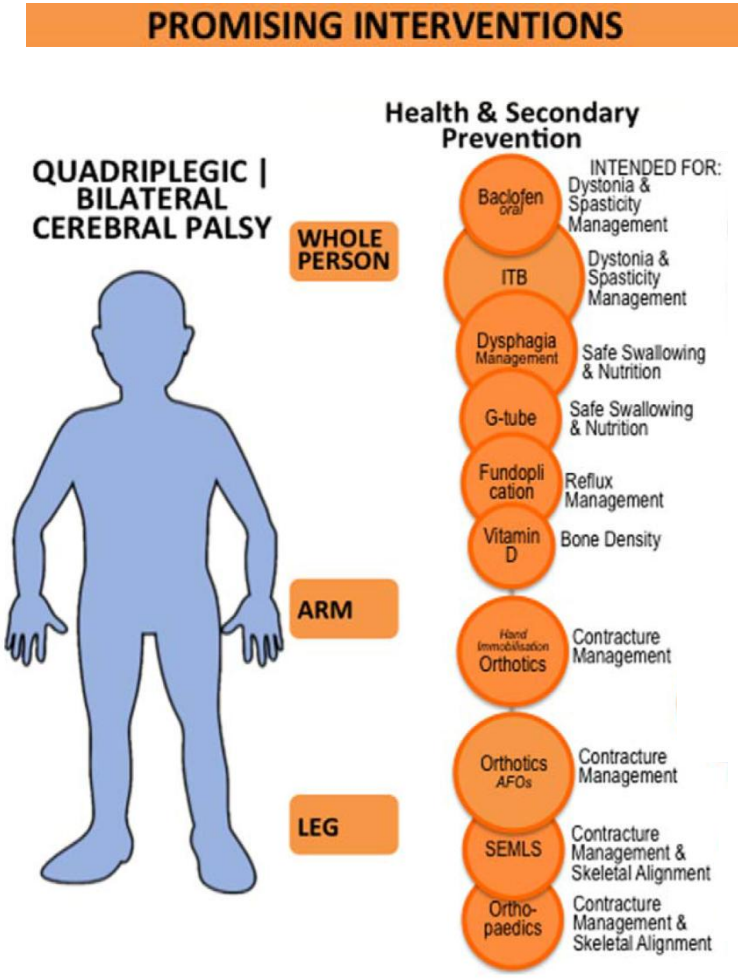
- Hip surveillance for maintaining hip joint integrity
- BoNT, diazepam, or selective dorsal rhizotomy for spasticity management
- Ankle casting for standing and/or walking
- Bisphosphonates
- Fitness training
- Pressure care



Intervention	ICF level: Body structures and function	Activity	Participation	Environment	Personal factors
Body structures and function interventions					
1. Anticonvulsants	G				
2. Botulinum toxin	G				
3. Bisphosphonates	G				
4. Casting (ankle)	G	Y			
5. Diazepam	G				
6. Fitness training	G	Y	Y		
7. Hip surveillance	G				
8. Pressure care	G				
9. Selective dorsal rhizotomy	G	Y	Y		

A systematic review of interventions for children with cerebral palsy: state of the evidence; Novak; 2013



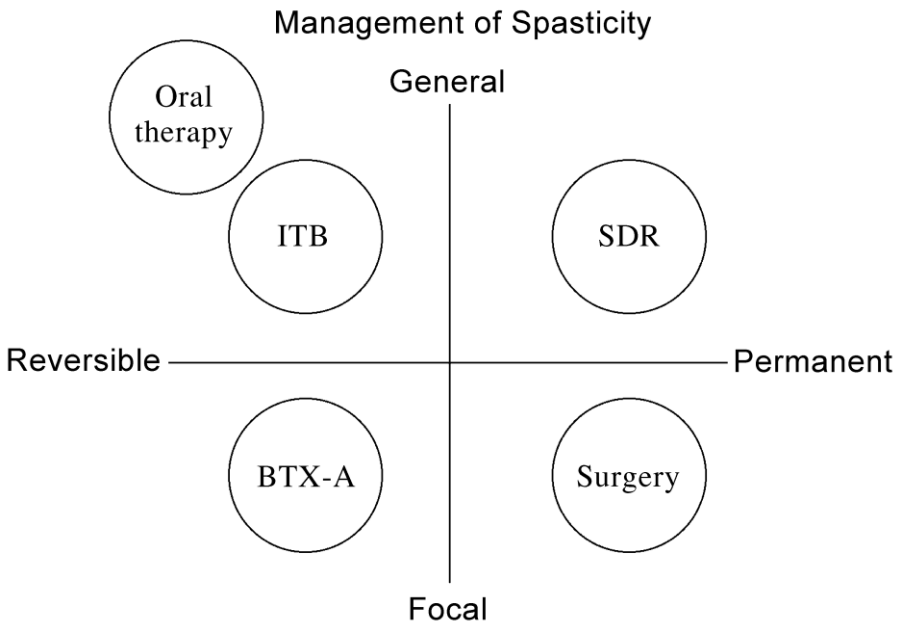


Evidence-based diagnosis, health care, and rehabilitation for children with cerebral palsy. Novak I1. J Child Neurol. 2014 Aug;29(8):1141-56.

Therapeutic Options of Spasticity

- **Elimination of aggravation conditions**

  Pain, Fatigue, Stress, Seizure, Excitement, Cold, Illness, Sleep disturbance, Immobility, Hormonal flux, GE reflux, Constipation, Voiding difficulty, Medication..



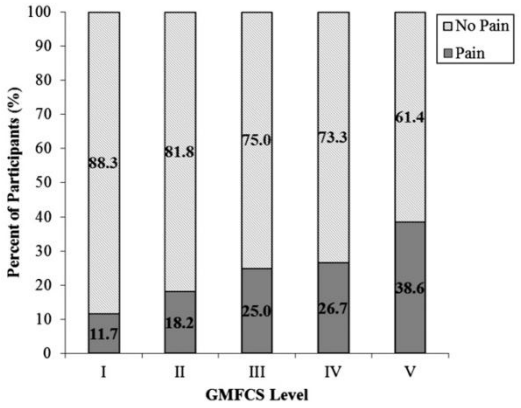
Indications for Botulinum Toxin Treatment

Localization	Unilateral CP	Bilateral ambulatory CP	Bilateral non-ambulatory CP
Upper limb	Improved function and aesthetics/appearance	N/A	Pain management Easier caring and positioning Functional and/or cosmetic Improvement of hand position
Lower limb	Improved gait	Improved gait	Pain management Easier caring and positioning Improvement of weight bearing Prevention of hip dislocation
Spine	N/A	N/A	Postural management Care Pain management

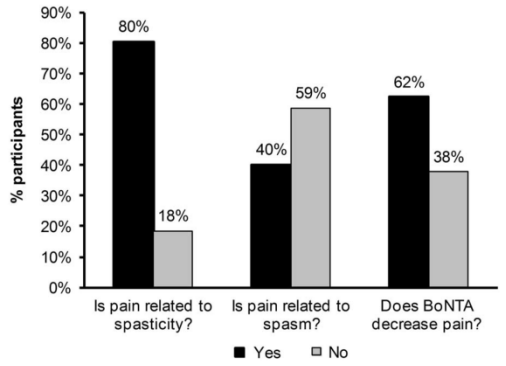
Best clinical practice in botulinum toxin treatment for children with cerebral palsy.Toxins;2015

PAIN in Non-ambulatory CP

- **Underrecognized and undertreated and negative affects QOL.**
- **Causes of pain** *(Panner et al. 2013)*
  - **In non-ambulatory CP : hip subluxation/dislocation (27%), dystonia (17%), constipation (15%), musculoskeletal(MSK) deformity, GE reflux, postop. MSK pain**
- **Hip pain in more than half of hips with migration percentage > 50%** *(Ramstad et al. 2016)*
- **BoNT treatment decreased pain (62%) in 1 month** *(Shaikh et al. 2015)*
- **26 GMFCS level V, improvement in hip pain at 3 months after BoNT** ***(p<0.001)*** *(Lundy et al. 2009)*



(Panner et al. 2013)



ure 2: Relationship between pain, spasticity, and botulinum toxin

(Shaikh et al. 2015)

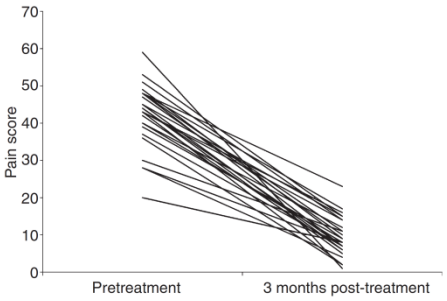


Figure 1: Individual pain profile scores before and 3 months after treatment with botulinum toxin type A.

(Lundy et al. 2009)

European Journal of Neurology 2010, **17** (Suppl. 2): 9–37 doi:10.1111/j.1468-1331.2010.03126.x

Botulinum toxin assessment, intervention and after-care for lower limb spasticity in children with cerebral palsy: international consensus statement

S. C. Love[a], I. Novak[b], M. Kentish[c], K. Desloovere[d], F. Heinen[e], G. Molenaers[f], S. O'Flaherty[g] and H. K. Graham[h]

Recommendation 3

- BoNT-A is established as effective in the management of spastic equinus to improve gait. (level A)
- BoNT-A is probably effective to improve goal attainment and function in the management of spastic equinus (level B)
- BoNT-A is similar to serial casting in the management of spastic equinus with current data being inadequate or conflicting (level U)
- BoNT-A injections to the adductor muscles is probably effective in some specific areas of goal attainment (level B)
- BoNT-A injections to the adductor muscles do not improve gross motor function (level A)
- BoNT-A injections to the adductor (and hamstring) muscles may delay hip displacement, but does not affect long-term outcome (level A)
- BoNT-A injections to multiple lower limb muscles have inadequate and conflicting data in respect of gait, goal attainment and function (level U)

Recommendation 4

- Conversion factors between different preparations of BoNT-A can lead to life threatening miscalculations and their use is strongly discouraged. Rates and sizes of reactions may be different between preparations (level A).
- Determination of dose relates to severity of spasticity, goal of treatment, size of targeted muscle, distribution of neuromuscular junctions with that muscle and previous responses to BoNT-A (if known).
- Dose should be cautiously selected in patients of GMFCS level V and any patient with dysphagia or breathing problems.
- Injection interval for serial BoNT-A should generally be no less than six months.
- Precise localization of muscle injection sites helps to improve the safety profile of BoNT-A by reducing the likelihood of unwanted toxin migration (level U)*. Use injection techniques which allow the operator to accurately isolate the target muscle (ultrasound is the preferred method).

*Expert opinion

Therapeutic Dosage of BoNT

- **In lower extremity**

Product	Dose U/kg body weight		Maximum Total Dose
	Range in literature	Recommendation	
BOTOX®	6–24 U/Kg (up to 30 U/Kg used in occasional multilevel injections)	GMFCS I–IV without risk factors: 16–20 U/Kg GMFCS V with risk factors: 12–16 U/Kg*	< 300 U [53,57] < 400–600 U [79]
Dysport®	10–30 U/Kg	20 U/Kg [52] (level B recommendation)	200–500 U [54] (level U Recommendation) < 900 U [79]

- **In upper extremity**
  - **Botox : 1-9 U/kg per treatment session**
  - **0.3-2 (forearm) up to 4 (upper arm) U/kg per muscle**

BoNT Treatment in GMFCS levels IV-V

- Moderate evidence to BoNT-A for pain reduction in undergoing hip adductor release surgery
- Weak to moderate evidence for its use in the reduction of spasticity-related pain

(Efficacy of botulinum toxin A in children with cerebral palsy in GMFCS levels IV and V: a systematic review; 2012)

- Systemic adverse effect (1~2%) in higher GMFCS levels (IV, V) and higher doses of BoNT-A
  General weakness, dysphagia, incontinence(bladder and bowel), respiratory sx.
- Respiratory sx. related with GMFCS level (IV, V), pre-existing comorbidities, bulbar function

(Systematic adverse events following botulinum toxin A therapy in children with cerebral palsy; 2010)

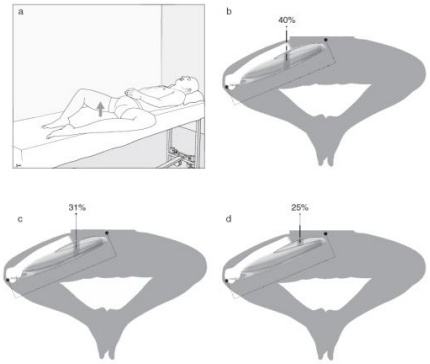
- BTA can be safely used for CP of GMFCS level V, using low doses and preferably without using sedation or anesthesia

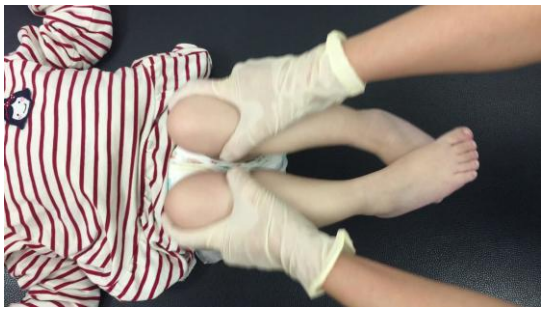
(Focal treatment of spasticity using botulinum toxin A in cerebral palsy cases of GMFCS level V: evaluation of adverse effects; 2014)

Motor Endplate-targeted BoNT Injection

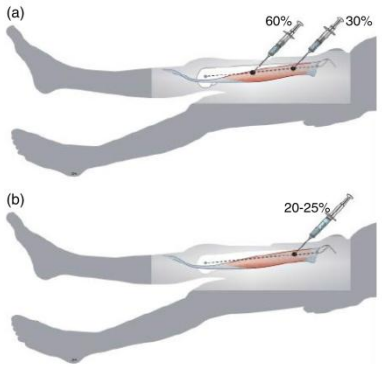
A line from the symphysis pubis to the medial knee joint line

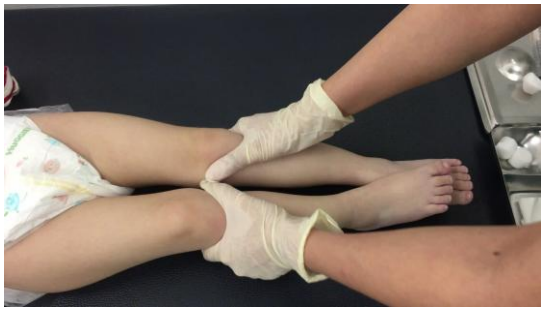
- **Hip Adductors**
  - Adductor longus 31%
  - Adductor brevis 22.4%
  - Adductor magnus 38.1%





- **Gracilis**
  - 30%, 60% 2 points





Localization of the motor endplate zone in human skeletal muscles of the lower limb, Developmental Medicine & Child Neurology (2010)

Motor Endplate-targeted BoNT Injection

- **Iliopsoas**

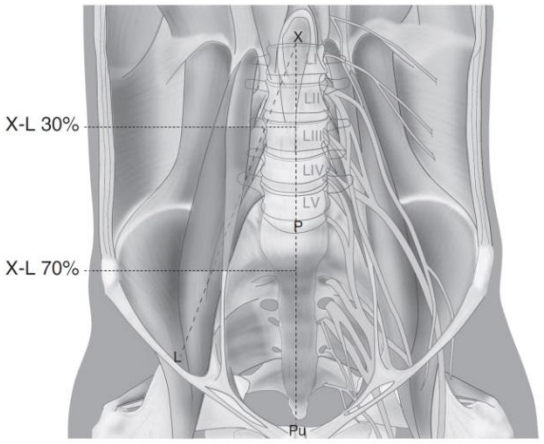


Figure 14: Optimal injection area for the psoas muscle, according to Van Campenhout et al.[29] X, Th12; P, sacral promontory; Pu, pubis; X–L, location where psoas muscle passes under inguinal ligament.

- **Rectus femoris**

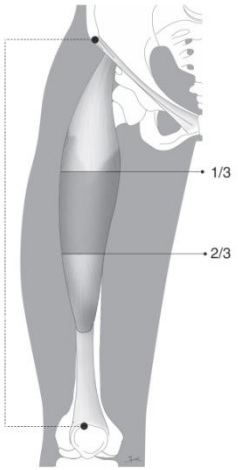


Figure 12: Optimal injection area for the rectus femoris muscle.

- **Hamstrings**

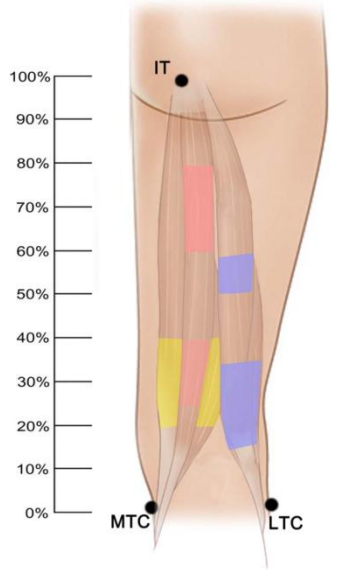


Fig. 5. Targeted botulinum toxin injection point for the hamstring muscles. The arborized portions of the biceps femoris long head and short head were at distances of 50–60% and 15–30%, respectively (blue shaded), the semitendinosus proximal and distal zones were at distances of 25–40% and 60–80% (red shaded), and the semimembranosus was at 20–40% (yellow shaded). [Color figure can be viewed in the online issue, which is available at wileyonlinelibrary.com.]

Localization of the motor endplate zone in human skeletal muscles of the lower limb, Developmental Medicine & Child Neurology (2010)

Intramuscular Nerve Distribution of the Hamstring Muscles:Application to Treating Spasticity, Clinical Anatomy (2016)

Muscles selection for BoNT injection

Position	Target muscle
Flexed hip	Iliopsoas Rectus femoris
Flexed knee	Hamstring Gastrocnemius
Adducted thigh	Adductor Gracilis Hamstring
Shoulder Adducted, Int. rotated	Pectoralis Latissimus dorsi Teres major Subscapularis
Shoulder Ext. rotated, Retracted	Infraspinatus Post. deltoid
Opisthotonus	Upper trapezius Paraspinal muscle

Sialorrhea (Drooling)

- Ant. drooling : unintentional loss of saliva from the mouth
- Post. drooling : Into the pharynx possibly creating a risk of aspiration
- Normal phenomenon until 18~24 months of age, pathological drooling after age 5 yrs

- Possible causes

  Inadequate head and trunk control

  Inadequate frequency of swallowing

  Constant open mouth position (poor m. function and coordination): BG injury

  Reduce the sensory cues needed to trigger a swallow

  Gastroesophageal reflux

Management of Sialorrhea

- **Pharmacologic treatment (20%)**

  Benztropin: not recommended under 3 yrs, positive effect on drooling *(Camp-Bruno 1989)*

  Scopolamine patch: contraindication under 7 years

  Trihexiphenidyl 1mg bid → increase

  Glycopyrrolate (Robinul) 0.01~0.04mg/kg, 4~19yrs: improvement *(Mier et al. 2000)*

  Oxybutinin (Ditropan, Obutin) S/E: dry mouth, constipation, vomiting, voiding difficulty..

- **Botulinum toxin**

  Previously undertaken a 3-month trial of pharmacological therapy with no effect

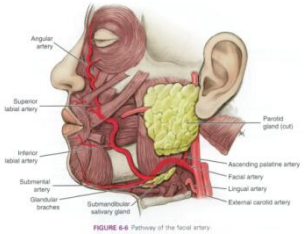
  5 units/kg is a safe effective dose

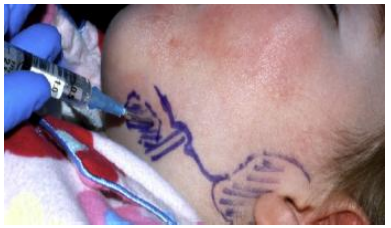
  Flow without stimulation (resting) : submandibular 65% (sero-mucous), parotid 23%, sublingual 4%

  Flow with stimulation (eating, chewing) :

  parotid 69% (serous)

  submandibular 26%, sublingual 5%





Oral Medication

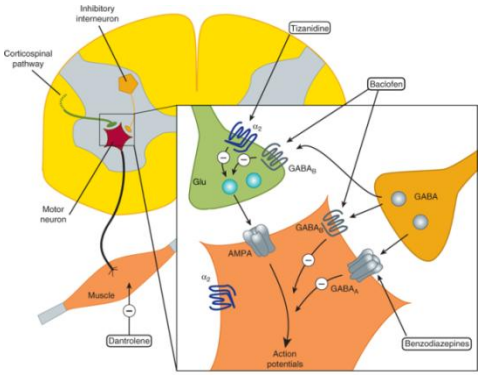
- Painful spasm, decreased sleep, seizure, dystonia
- Mild tone reduction, Widespread spasticity
- Musculoskeletal development

- Disadvantages of systemic effect and side effect
- Often based on personal experience and the impact of benefit versus potential side effects

Oral Medication for Spasticity Management

Medication	Site of Action	Mechanism	Dosage	Cautions
Diazepam	Supraspinal	Facilitate transmission at GABA-A receptors	0.12 - 0.8 mg/kg/d	-Risk of dependence -Risk of withdrawl
Baclofen	Spinal cord	Analogue of GABA-B	2.5 -10 mg/d Max 40~60 mg/d	Withdrawl GERD Seizure
Dantrolene	Skeletal muscle	Inhibit release of Ca from SR	0.5(1~3) mg/kg/d 6 – 8(12) mg/kg/d	Hepatotoxicity
Tizanidine	Brain / spinal cord	A2 agonist Hyperpolarization of motor neuron	2 mg at bedtime (>10yrs)	Hepatotoxicity (rare)

Diazepam



- Facilitate the activity of gamma-aminobutyric acid ($GABA_A$)
- Well absorbed, distributed into the brain according to lipid solubility
- Decrease sleep latency, stages 1&2 ⬆, stage 3 & slow-wave sleep & REM ⬇
- Should be considered for short-term treatment (Level B)

- **Single dose at bedtime** reduces hypertonia and muscle spasm and improves posture and mobility in young children with CP.
- Repeated short courses of 6-12 weeks with a 3-4 week drug holiday in between precludes the possibility of tolerance with long-term use.

(Mathew et. al.; Pediatric Rehabilitation; 2005)

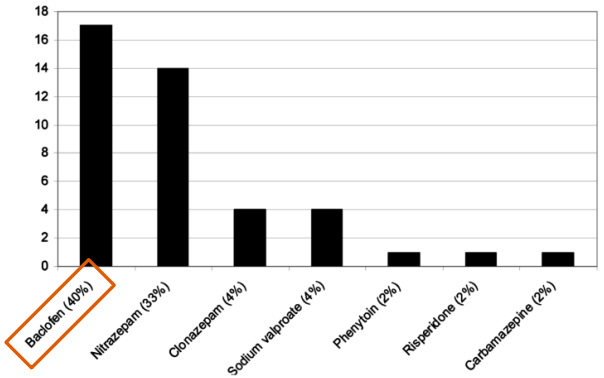
Sleep Disturbance in CP

- Obstructive sleep apnea
  - Gastroesophageal reflux
  - Obesity
  - Excessive secretion
- Dysregulation of circadian rhythm (e.g., melatonin)
  - Intellectual disability
  - Visual impairment
  - Abnormalities of motor and tone
  - Epilepsy and pain

Baclofen

- Gamma-aminobutyric acid B ($GABA_B$) agonist
- Both intrathecal and oral delivery routes
- CNS depressant properties – sedation /c tolerance, somnolence, ataxia
- Proconvulsive effect - EEG changes by the baclofen increase *(Rinoaldis, 2010)*

❖ Side effect of excessive drooling ?



Number of children taking drugs which are known to exacerbate drooling

(J. Montgomery, 2016)

❖ Effect in neonatal hypertonia ?

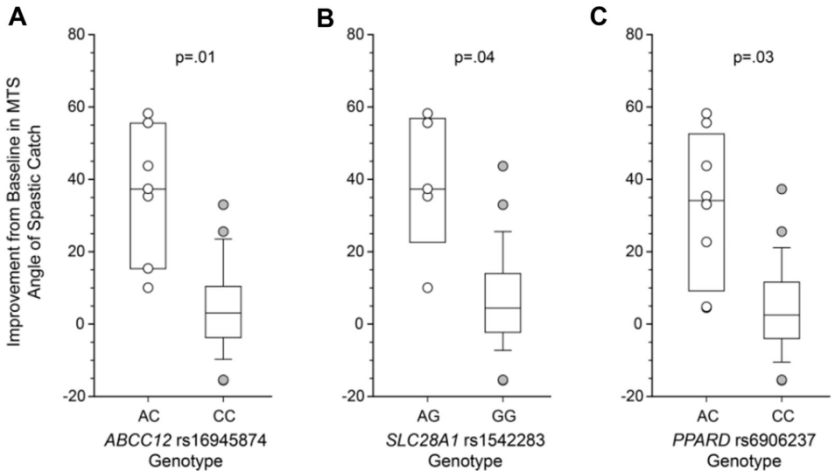
29 infants (mean GA 25.7 ± 1.9 wks)
Started age of 86.4 ± 3.6 days
Baclofen 1 mg/kg/d, 2-4 wks
Modified Ashworth Scale

No overall decrease in tone during oral baclofen in hypertonic preterm neonates.
Limitations of dosage, assessment..

(E. Schulz, 2012)

Pharmacogenomic Variability of Oral Baclofen Clearance and Clinical Response in Children with CP

- 49 spastic CP
- SNP of ABCC9 (rs11046232, heterozygous AT versus the reference TT genotype), a 2-fold increase in oral baclofen clearance (mean 0.51 standard deviation 0.05 L/h/kg for the AT genotype versus 0.25 0.07 L/h/kg for the TT genotype, adjusted P < .001)
- Clinical responses were associated with decreased spasticity by Modified Tardieu Scale in allelic variants with SNPs ABCC12, SLC28A1, and PPARD.



Physical Medicine & Rehabilitation; 2017

Oral Medication for Dystonia Management

Medication	Mechanism	Dosage	Cautions
Trihexyphenidyl	Antagonized acetylcholine receptor	2.5 mg/d Max. 15 mg/d	Dry mouth, blurry vision, dizziness, nausea, anxiety, glaucoma, anhidrosis, neuroleptic malignant syndrome, tardive dyskinesia
Clonazepam	Facilitate transmission at GABA-A receptors	2-4mg/d	Sedation, confusion, depression, ataxia, dependence
Cardidopa /Levodopa	Inhibit peripheral dopamine decarboxylation	10/100 mg bid 25/100 mg tid	Dyskinesia, bradykinesia, hypotension, memory impairment, confusion, hallucination

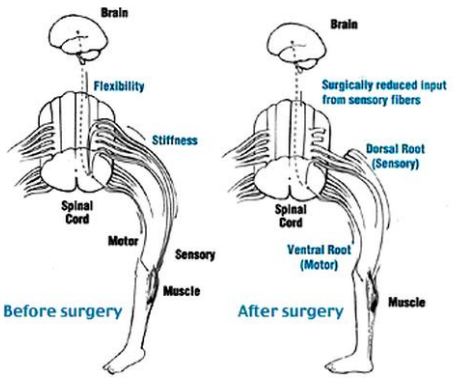
Intrathecal Baclofen Pump (ITB pump)

2014 척수강내 약물주입펌프이식술 (PUMP) 보험급여 혜택

Ref) 건강보험요양급여비용 **(2014년 7월판)** 사단법인대한병원협회 / 보건복지부 고시 제**2014-84**호

항 목	자-484 척수강내 약물주입펌프이식술 (Implantation of Intrathecal Drug Infusion Pump)
	자-484 척수강내 약물주입펌프이식술의 **세부사항 인정기준**
세부인정사항	척수강내 약물주입펌프이식술은 다음과 같은 경우에 요양급여를 인정함. [다 음] 가. **6개월 이상의 적절한 통증치료**(약물치료와 신경차단술 등)에도 효과가 없고, 심한 통증(VAS 통증점수 7 이상)이 지속되는 불인성 통증이 있는 경우 나. 고용량의 모르핀(1일 200mg) 경구투여나 또는 동등 역가의 타 마약성 진통제 투여를 하였음에도 통증이 제어되지 않는 암성통증(VAS 통증점수 7 이상)으로 **여명이 1년 이상으로 예상되는 경우** 다. 모르핀 또는 타 마약성 진통제의 부작용 등 약물투여를 할 수 없는 암성통증(VAS 통증점수 7 이상)으로 여명이 1년 이상으로 예상되는 경우 라. 적절한 경직치료(약물치료 등)에도 불구하고 경직척도(MAS)가 하지 3등급 이상 또는 상지 2등급 이상인 중추신경계 손상에 의한 경직(spasticity)으로 시험적 약물주입술에서 1등급 이상 호전된 경우

Selective Dorsal Rhizotomy (SDR)



- **Indication**

  Group I : ambulation

  Group II : patient care, contracture, subluxation, dislocation, deformity, skin ulcer, voiding care

Pre-operative scissoring

M/8 yrs
Spastic bilateral CP
GMFCS level IV
Difficulty in perineal care

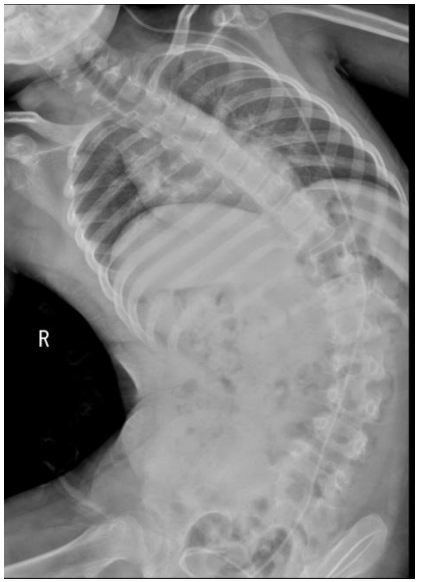
SDR

Post-operative tone reduction

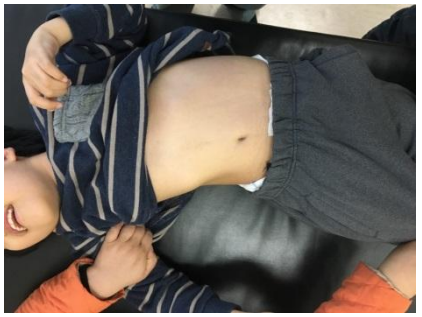
Clinical Case

Case 1.

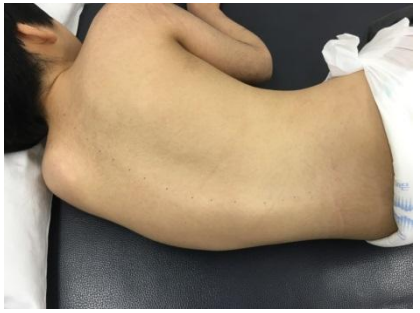
- F/12y, BW 24kg
- Spastic bilateral CP, GMFCS IV
- Lt. scoliosis, Cobb's angle 70°



- Ambulation with assist until 8 yr
- Aggravation of scoliosis since W/C bounded



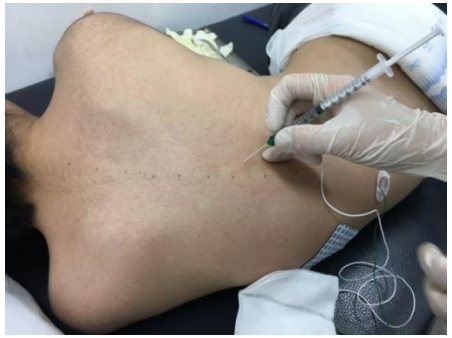
- '척추측만증이 심해서 앉은 자세유지가 어렵다.'



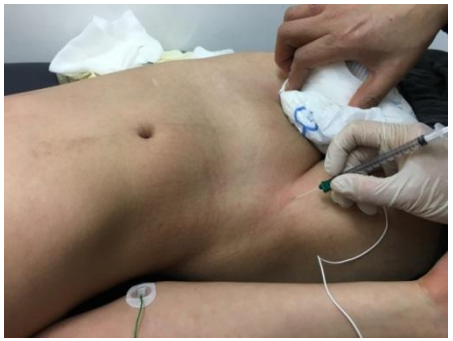
Case

- Symmetric hypertonia around hip, bilateral (scissoring)
- Asymmetric trunk tone increased in Rt. side

- Botulinum Toxin injection
  - Rt. paraspinal muscle 80 unit
    (20 unit x 4 points, T10~S1)
  - Both iliacus 30 unit, each point
  - Both med. hamstring 80 unit, each point
  - Total 300 unit

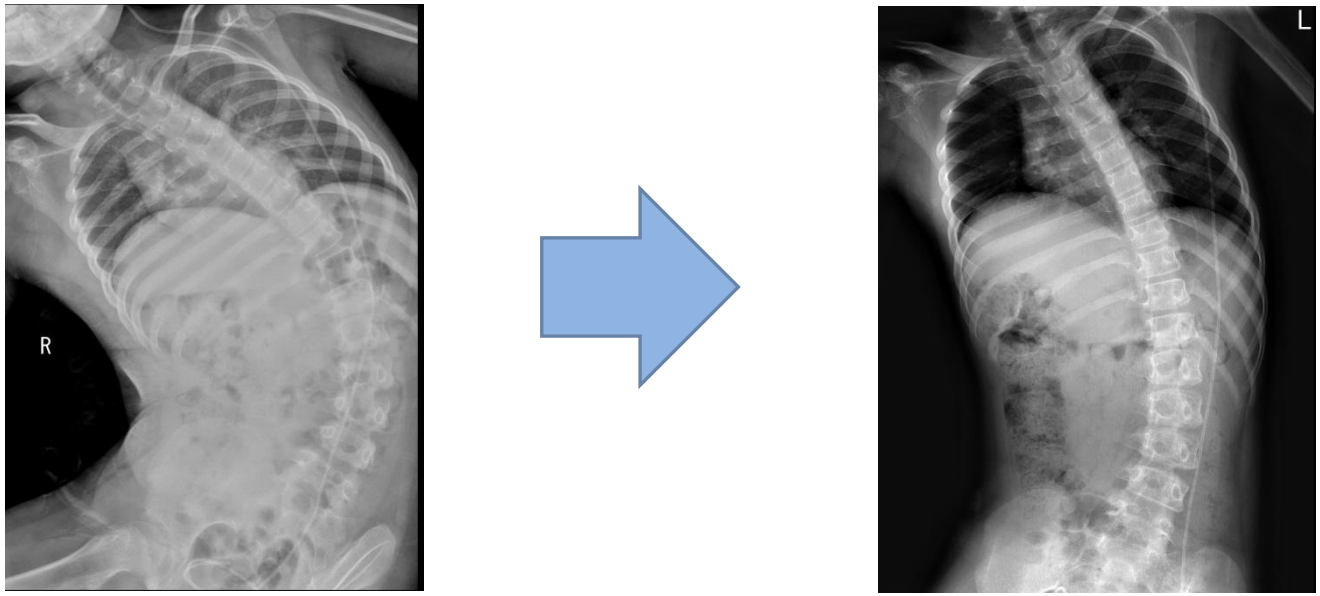


- Chemoneurolysis
  - Both obturator nerve
  - 50% alcohol , 3cc, each point



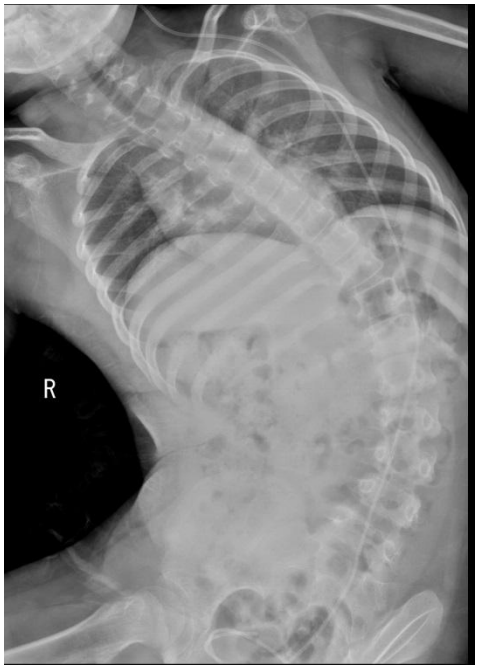
Case

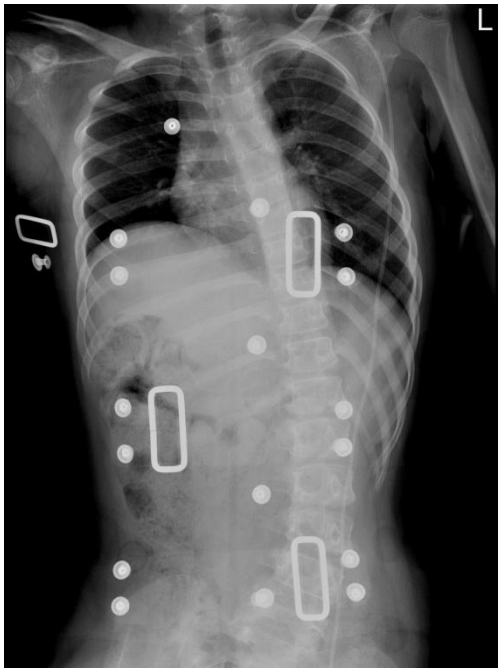
- Post-injection assessment
- Cobb's angle 70^{o} -> 35^{o}



Case

- TLSO and seating system application





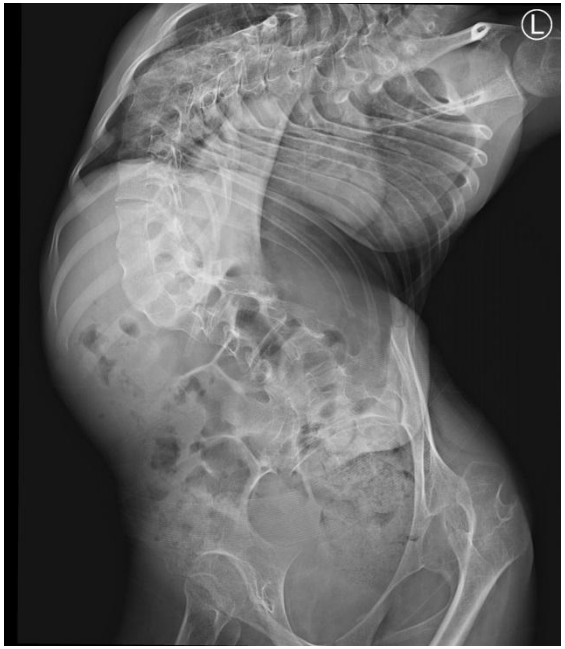
Case 2.

- **M/18y, BW 22kg**
- **Spastic bilateral CP, GMFCS level V**
- **Severe scoliosis, Windswept hip**
- **Seizure**
- **Intellectual disability**
- **Dysphagia –** 누워서 유동식만 먹는다.

- ‘경직이 심해진다.’ ‘많이 아파한다.’

Case 2.

- Previous medication
  - Anti-epileptic drug – 30mg phenobarbital
  - Anti-spastic drug – long-term baclofen 5mg tid
  - Analgesic drug - Acetaminophen 320mg bid
- Intervention
  - Oral medication changed to diazepam, dantrolene
  - Botulinum toxin injection on hip and paraspinal m.



Case 2.

- ‘약이나 음식을 먹고 나면 헛구역질을 하고 경직이 심해진다.’
  - **GE reflux** 의심 하에 **PPI trial**

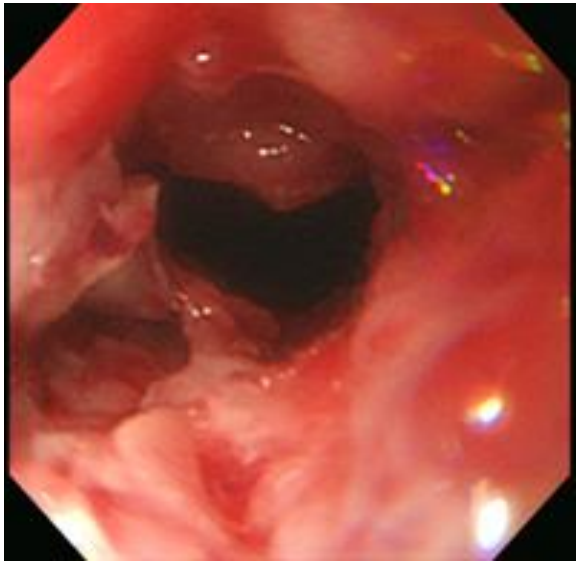
Case 2.

- ‘약이나 음식을 먹고 나면 헛구역질을 하고 경직이 심해진다.’
  - **GE reflux** 의심 하에 **PPI trial**

- **Upper gastrointestinal endoscopy**
  - **Mid-esophageal adenocarcinoma**

    d/t chronic GE reflux

    rare case in young age



Thank you for listening.