# SPASTICITY & DYSTONIA IN PATIENTS WITH CP

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# CP Definition

- A heterogeneous group of disorders of **movement and posture**, affecting primarily the **motor function**, secondary to **permanent, non-progressive pathology** of the **developing brain**

# CP Classifications

| Tone | Topographic Distribution | Function |
|---|---|---|
| Spastic | Hemiplegic | GMFCS I - V |
| Dyskinetic<br>▪ Dystonic<br>▪ Choreoathetoid | Diplegic | MACS I - V |
| Ataxic | Quadriplegic | CFCS I - V |

# Spasticity, Rigidity & Dystonia

- **Spasticity:** Velocity-dependent resistance to passive stretch, presence of spastic catch, A/W increased DTRs – caused by UMN lesion
- **Rigidity:** Velocity-independent resistance to passive stretch, equal resistance to passive stretch with extension & flexion, maintenance of limb position after passive stretch, not A/W increased DTRs – likely caused by basial ganglia lesion
- **Dystonia:** Involuntary movements/posturing of a body part (e.g., limb, torso, neck) with tactile stimulus or purposeful movement of another body part, tone is not velocity-dependent, not A/W increased DTRs – likely caused by basial ganglia lesion

# HYPERTONIA ASSESSMENT TOOL (HAT) - SCORING CHART

| HAT ITEM | SCORING GUIDELINES (0=negative or 1=positive) | SCORE 0=negative 1=positive *(circle score)* | TYPE OF HYPERTONIA |
|---|---|---|---|
| 1. Increased involuntary movements/postures of the designated limb with tactile stimulus of another body part | 0= No involuntary movements or postures observed | **0** | **DYSTONIA** |
| | 1= Involuntary movements or postures observed | **1** | |
| 2. Increased involuntary movements/postures with purposeful movements of another body part | 0= No involuntary movements or postures observed | **0** | **DYSTONIA** |
| | 1= Involuntary movements or postures observed | **1** | |
| 3. Velocity dependent resistance to stretch | 0= No increased resistance noticed during fast stretch compared to slow stretch | **0** | **SPASTICITY** |
| | 1= Increased resistance noticed during fast stretch compared to slow stretch | **1** | |
| 4. Presence of a spastic catch | 0= No spastic catch noted | **0** | **SPASTICITY** |
| | 1= Spastic catch noted | **1** | |
| 5. Equal resistance to passive stretch during bi-directional movement of a joint | 0= Equal resistance not noted with bi-directional movement | **0** | **RIGIDITY** |
| | 1= Equal resistance noted with bi-directional movement | **1** | |
| 6. Increased tone with movement of another body part | 0= No increased tone noted with purposeful movement | **0** | **DYSTONIA** |
| | 1= Greater tone noted with purposeful movement | **1** | |
| 7. Maintenance of limb position after passive movement | 0= Limb returns (partially or fully) to original position | **0** | **RIGIDITY** |
| | 1= Limb remains in final position of stretch | **1** | |

**SUMMARY SCORE – HAT DIAGNOSIS**

| | | *Check box:* |
|---|---|---|
| DYSTONIA → | Positive score (1) on at least one of the Items #1, 2, or 6 | ☐ Yes ☐ No |
| SPASTICITY → | Positive score (1) on either one or both of the Items #3 or 4 | ☐ Yes ☐ No |
| RIGIDITY → | Positive score (1) on either one or both of the Items #5 or 7 | ☐ Yes ☐ No |
| MIXED TONE → | Presence of 1 or more subgroups (e.g. dystonia, spasticity, rigidity) | ☐ Yes ☐ No |

# CP Classifications

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|---|---|---|
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| Ataxic | Quadriplegic | CFCS I - V |

# Management


Rehabilitation
Surgical Treatments
Oral Medications
Local Treatments

# Approach

- **No standardized approach** (least to most invasive approach)
- **Rehabilitation** – least invasive for all CP patients with spasticity or dystonia
- **Choice of Tx depends on:**
  1. Spasticity, dystonia or both
  2. Function (e.g., GMFCS)
  3. Muscle involvement (e.g., generalized vs. focal/segmental)
  4. Other symptoms to co-treat (e.g., clonazepam for seizures + spasticity)
  5. Age of child
  6. Realistic goals

# Approach

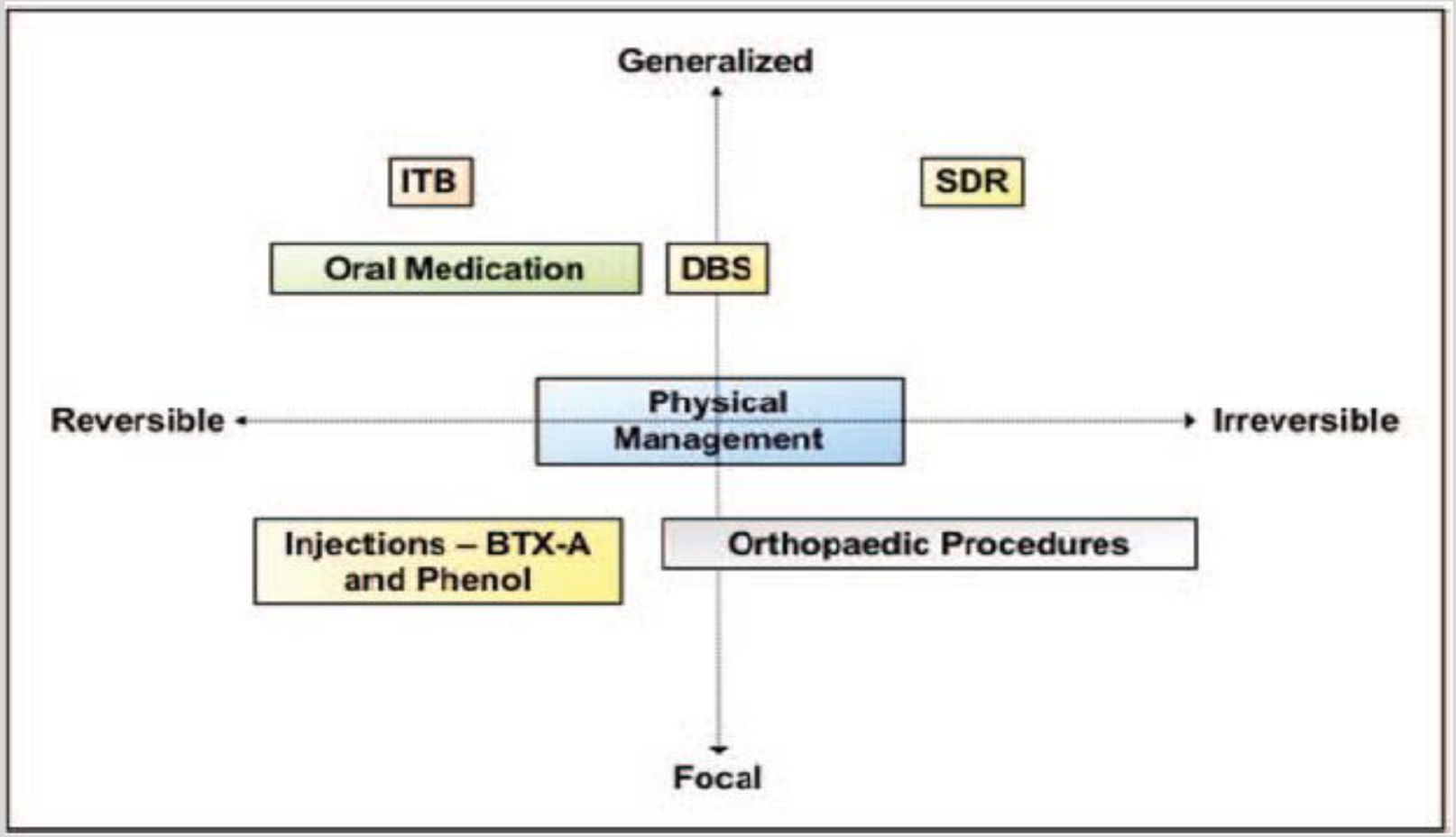
**Realistic goals (need to be discussed with the family):**

1. Improved pain (which can cause more spasticity/dystonia)
2. Improved function (e.g., GMFCS)
3. Improved care giving
4. Deferral of surgical interventions
5. Prevention of long-term MSK changes (e.g., contractures)
6. Cosmetic improvement

# Approach

| | |
|---|---|
| **Rehabilitation** | All patients |
| **Oral meds** | Generalized spasticity or dystonia |
| **Botox/phenol/ ethanol injections** | Local/segmental spasticity or dystonia |
| **ITB** | Generalized spasticity or dystonia with severe motor impairment (GMFCS IV or V) + significant side effects with oral meds OR not achieving adequate response with oral meds despite maximal dose |
| **SDR** | Spastic diplegia with mild-to-moderate motor impairment (GMFCS II or III) + cognitively able to participate in post-op rehabilitation |
| **Orthopedic interventions** | Orthopedic problems in children with CP (e.g., hip pathology) |

# Approach – spasticity



# Approach – dystonia

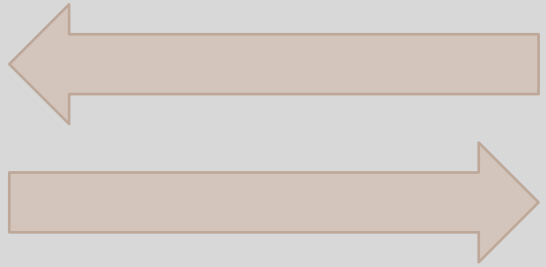
# Rehabilitation

1. **PT:**
   - Promotes range of motion, positioning/seating, coordination
   - Assessment for splinting options (e.g., AFOs)
2. **OT:** Focuses on development of fine-motor skills and improving capacity of self-care or caregiver support of tasks of daily living (e.g., feeding, toileting, dressing, personal hygiene)

# Medications

**Indications to use medications:**

- Generalized spasticity or dystonia, particularly when causing **pain** and **decreased function**



# Medications

**Spasticity:**

1. Baclofen
2. Alpha-2 agonists (e.g., tizanidine, clonidine)
3. Benzos (e.g., diazepam, clonazepam)
4. Dantrolene
5. Artane
6. Others: Gabapentin, lamotrigine, cyproheptadine, cannabinoid-like compounds (dronabinol, nabilone), CBD

**Dystonia:**

1. Baclofen
2. Levodopa
3. Artane
4. Others: Gabapentin, benzos, clonidine

# Medications

## What to watch for when starting those meds?

1. Reduced neck/trunk control
2. Increased breathing and swallowing problems (e.g., drooling)
3. Increased obstructive sleep apnea
4. Increased sedation
5. Worsening of GI symptoms (especially with baclofen)

# Medications

**Baclofen:**

- GABA agonist – mainly works at the spinal cord level
- Uses: Spasticity and dystonia
- Dose: Refer to the formulary
- Side effects: Tolerance, sedation, dizziness, confusion, weakness, fatigue, ataxia, nausea, constipation, blurry vision, renal insufficiency
- Abrupt D/C → withdrawal symptoms (e.g., increased spasticity, hallucinations, confusion, hyperthermia, **seizures**)

# Medications

**Diazepam:**

- Increases affinity to GABA – mainly works at the spinal cord and BS levels
- Uses: Spasticity and dystonia (often used for a short-term)
- Preferred in patients with seizures, anxiety or sleep problems
- Dose: Refer to the formulary
- Half life: 20-80 hr (18-28 hr with clonazepam)
- Side effects: Hypotension, sedation, memory impairment, ataxia, depression, tolerance, dependency, hypersalivation
- Abrupt D/C → withdrawal symptoms, mainly **seizures**

# Medications

**Clonazepam:**

Similar to diazepam with the following differences:

- Dose:

**< 30 Kg:** 0.1-0.2 mg/Kg/day PO div BID-TID; start at 0.01-0.03 mg/Kg/day PO div BID-TID; increase by 0.25-0.5 mg/day Q3 days; give largest dose QHS; taper gradually to D/C

**≥ 30 Kg:** 0.5-5mg PO TID; start at 0.5 mg PO TID; increase by 0.5-1 mg/day Q3 days; max 20 mg/day; taper gradually to D/C

- Half life: 18-28 hr

# Medications

**Tizanidine:**

- Centrally-acting alpha-2 agonist
- Dose: Refer to the formulary
- Preferred in patients with sleep problems and neuro-irritability
- Side effects: Hypotension, sedation, dizziness, asthenia, dry mouth, hallucinations, constipation, rarely **liver problems** (F/U with LFTs)

# Medications

**Clonidine:**

- Centrally-acting alpha-2 agonist
- Dose: Refer to the formulary
- Preferred in patients with sleep problems and neuro-irritability
- Side effects: Sinus bradycardia, hypotension, rebound hypertension, syncope

# Medications

**Artane:**

- Anticholinergic
- Formulations: 2 mg and 5 mg tabs; Elixir 2 mg/5 mL
- Dose:
  - Start at 1 mg BID x7 days, then increase every 3 days by 1-2 mg/day, if daily dose > 10 mg → div TID-QID (not BID), continue this way up to target dose (6-40 mg/day), max daily dose 100 mg/day; taper gradually to D/C
- Preferred in patients with sialorrhea
- Side effects: Dryness of mouth, blurred vision, anhidrosis, dizziness, mild nausea, nervousness

# Medications

**Dantrolene:**

- Works at the muscle; inhibits calcium release from the sarcoplasmic reticulum and thus causing muscle weakness
- Dose: 0.5 mg/Kg/dose BID, increase to 3 mg/Kg/dose QID, max 400 mg/day
- Side effects: Sedation, paresthesia, GI symptoms (e.g., nausea, vomiting, diarrhea, rarely hepatotoxicity)
  - LFTs should be performed before treatment and periodically thereafter.

# Medications

**Levodopa:**

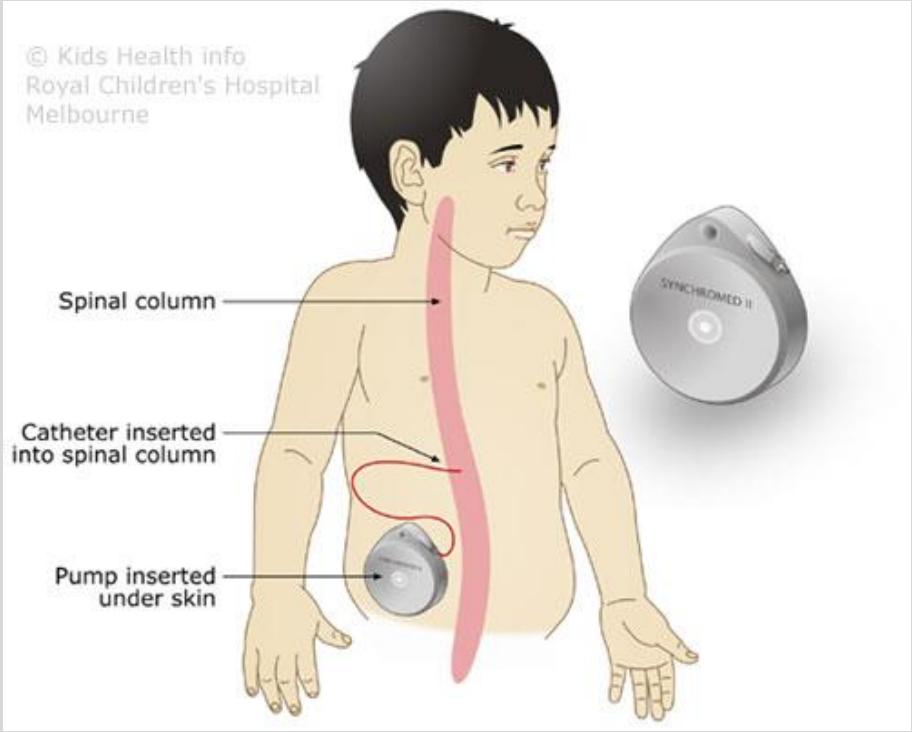
- Dopaminergic
- Comes in various combinations of Carbidopa:Levodopa
- Common example: Sinemet 25/100 (Carbidopa 25 mg/Levodopa 100 mg)
- Dose:
  - Calculate dose according to the Levodopa component
  - Start at 1 mg/Kg/day div BID, gradual increase to 4-5 mg/Kg/day div BID (some authors recommend 10 mg/Kg/day), max 600 mg/day

# ITB

**Indication to use ITB:** Generalized spasticity or dystonia with severe motor impairment **(GMFCS IV or V)** + significant side effects with oral meds OR not achieving adequate response with oral meds despite maximal dose

**Evidence:** Probability effective

**Limitation:** Size of the device that needs to be implanted, and therefore most younger children are not applicable



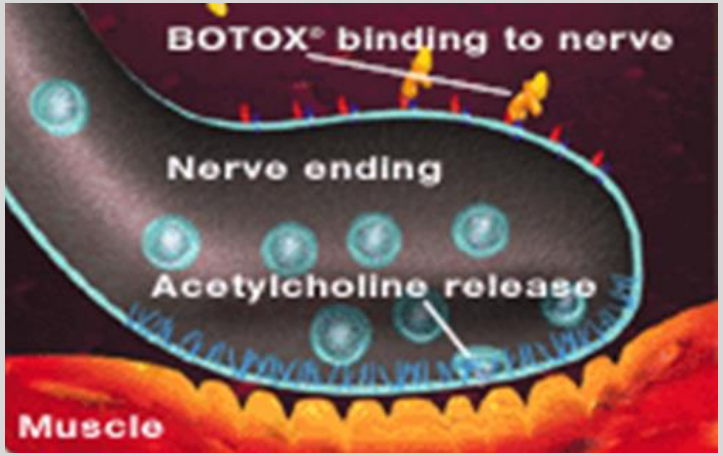
# ITB

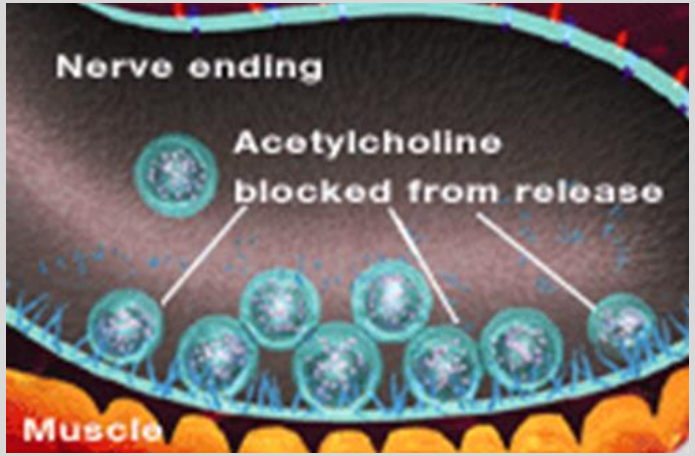
**Side effects:**

- 25-30% experience significant complications requiring surgical management in 3 years
  - System (pump malfunction, catheter blockage/breakage/leakage, CSF leak, seroma, back pain at the site, pump pocket/suture site infection/inflammation)
  - General (seizures, withdrawal symptoms due to pump malfunction, progressive scoliosis)

# Botulinum Toxin

- **Botulinum toxin A (Botox) & B**
- **MoA:** Inhibits acetylcholine release at the neuromuscular junction



# Botulinum Toxin

- **Evidence:**
  - **Botulinum toxin A:**
    - **Effective in reducing spasticity & dystonia in children with CP**
    - Improves function & ROM – mainly of the arms (no evidence of its use to maintain ROM)
    - Possibly improves pain
    - No evidence of hip dislocation prevention
    - No evidence of long-term positive changes
    - Has cosmetic considerations
  - **Botulinum toxin B:** Insufficient evidence to support its use as a treatment for spasticity/dystonia in children with CP

# Botulinum Toxin

**Indication:** Local/segmental spasticity or dystonia

- Gan be given in generalized spasticity or dystonia, but to be localized to specific muscle groups

# Botulinum Toxin

**Which muscles will Botox be most affective at?**

Muscles that have a **dynamic range** and are **not contracted** as measured in the modified Tardieu scale (R1:R2)

**R1 = Resistance 1 = first resistance on quick movement**

**R2 = Resistance 2 = end range**

The more contracted the muscle, the smaller the difference between R2 and R1 → the less effective the treatment

# Botulinum Toxin

**Timeline:**

- Starts working after **a few days**
- Peaks at **~2 weeks**
- Lasts **~3 months**

# Botulinum Toxin

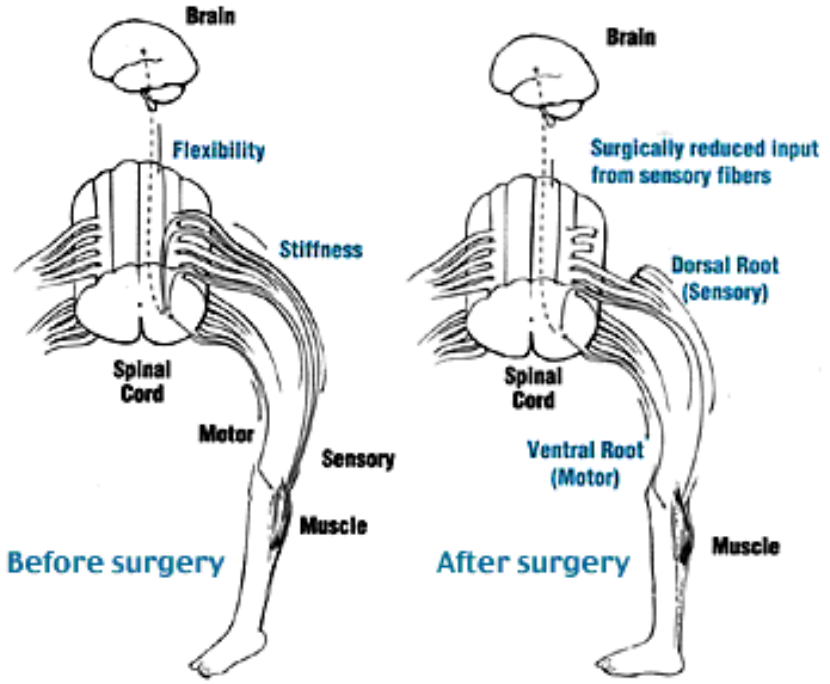
- **Side effects:**
  - Antibody formation: Repeated, high-dose injections are more likely to result in antibody formation than less frequently repeated, low-dose injections
  - Generalized weakness, particularly when given for generalized spasticity/dystonia

# Phenol/ethanol injections

- Effective treatment to reduce spasticity/dystonia
- Can be combined with Botox
- Advantage:
  1. **Longer acting than Botox (6-8 months)**
  2. **Immediate onset**
- Side effects:
  - Can cause paresthesia when used on sensory nerves
  - Technically, slightly more difficult, requires EMG guidance

# SDR

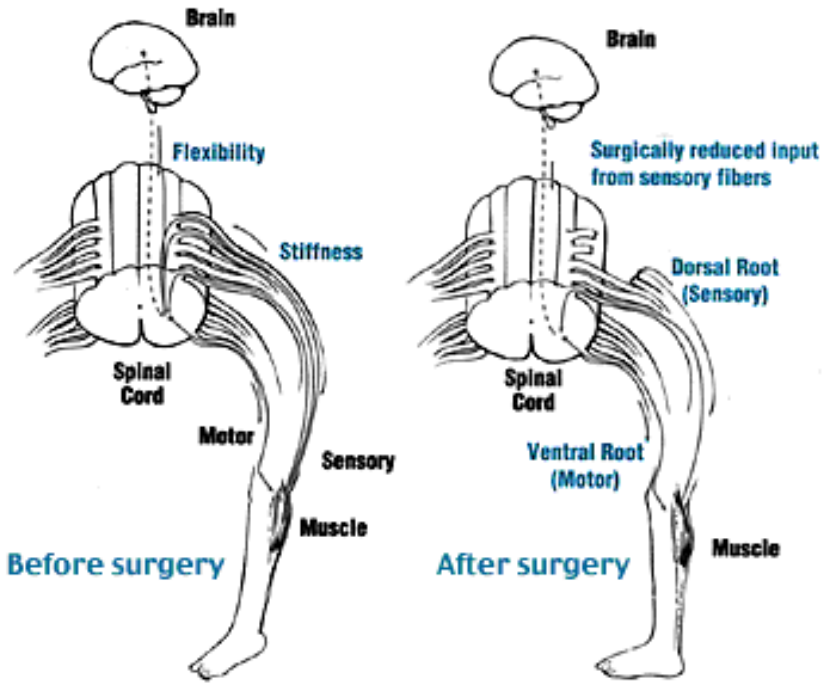
- Neurosurgical, irreversible intervention
- Involves cutting nerve roots L2-S2
- **Indication:** Spastic diplegia with mild-to-moderate motor impairment **(GMFCS II or III)** + cognitively able to participate in post-op rehabilitation
- **Age:** 4-8 years
- No role in dystonia management



# SDR

## Complications:

- Transient urinary incontinence (1.2 – 24%) – permanent incontinence rare
- Transient dysesthesias (2.5-40%)
- Permanent hypoesthesia (0-6%)
- Back pain – long term (4-7%)
- Progressive hip subluxation (not clear of causality)
- Progressive scoliosis – more an issue in non-ambulatory spastic quadriplegic patients



# Orthopedic Interventions

Options:

- Tendon lengthening/contracture release
- Tendon transfer
- Osteotomy

# How to decide on a treatment?

1. Full assessment
   - Age, CP classification based on tone, topographic distribution & function (e.g., GMFCS), ROS (other symptoms to co-treat)
2. Discussion about realistic goals for the patient (6 goals)
3. Use the ICF framework including the F-Word approach to implement the goals and goal oriented therapy

# How to decide on a treatment?

**WHO ICF Framework (with F-words)**

Child: care, health and development
Review Article
doi:10.1111/j.1365-2214.2011.01338.x

## The 'F-words' in childhood disability: I swear this is how we should think!

P. Rosenbaum* and J. W. Gorter*†

**CanChild* Centre for Childhood Disability Research, McMaster University, Hamilton, ON, Canada, and
†NetChild Network for Childhood Disability Research, Utrecht, the Netherlands

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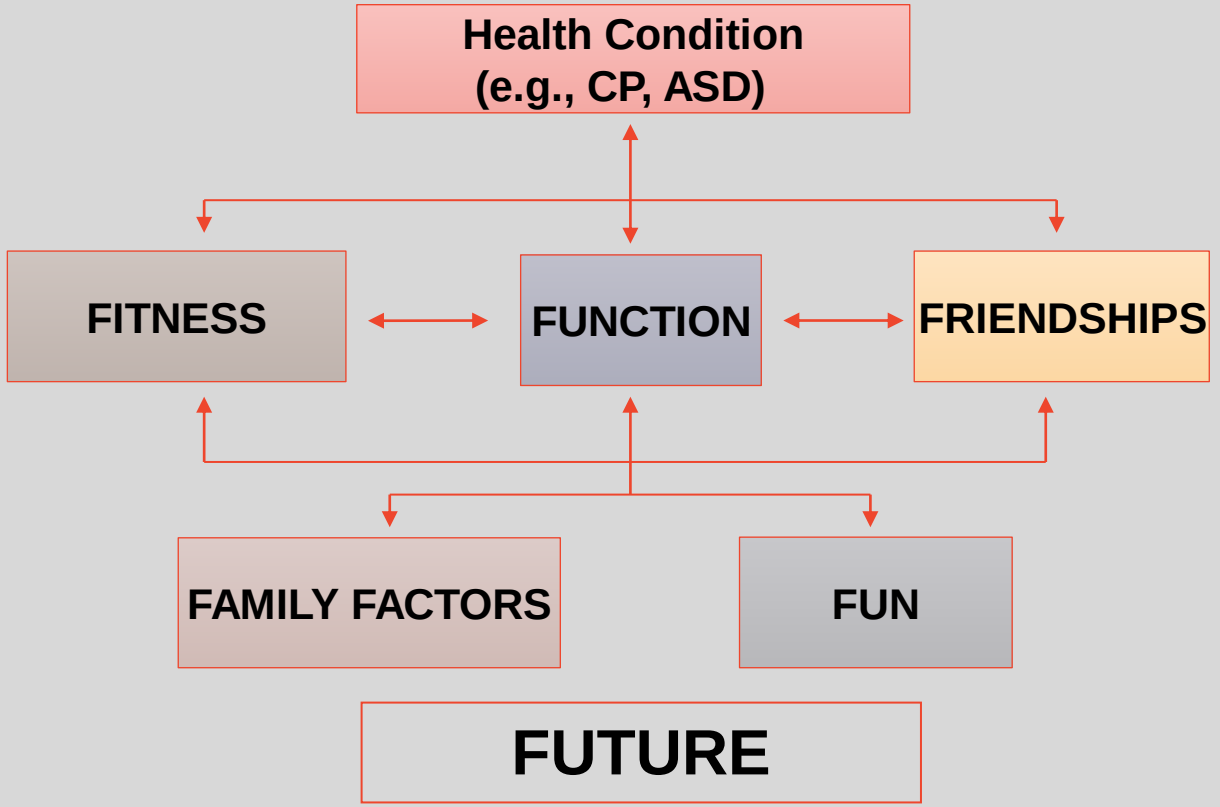
# How to decide on a treatment?

**WHO ICF Framework (with F-words)**



# Thanks!

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