Weill Cornell Medical College

NewYork-Presbyterian
Phyllis and David Komansky
Center for Children's Health

Pediatric
Brain and Spine Center

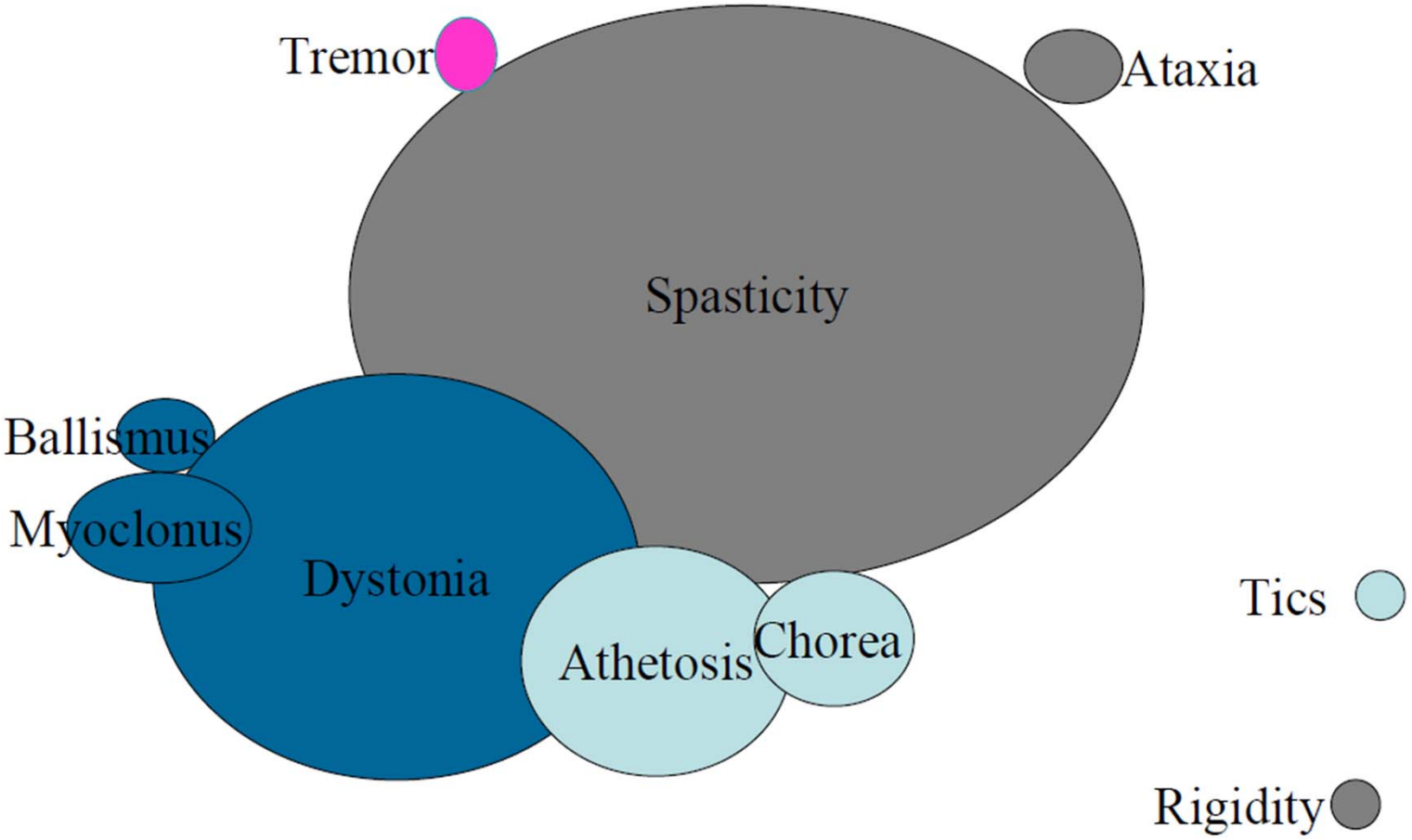
Murray Engel MD
Jeffrey P Greenfield MD PhD

- 1. Orthopedic deformity, such as hip dislocation, contractures, or scoliosis
- 2. Impairment of activities of daily living (eg, dressing, bathing, toileting)
- 3. Impairment of mobility (eg, inability to walk, roll, sit)
- 4. Skin breakdown secondary to positioning difficulties and shearing pressure
- 5. Pain or abnormal sensory feedback
- 6. Poor weight gain secondary to high caloric expenditure
- 7. Sleep disturbance
- 8. Depression secondary to lack of functional independence

- Motor disorder characterized by a velocity-dependent increase in muscle tone

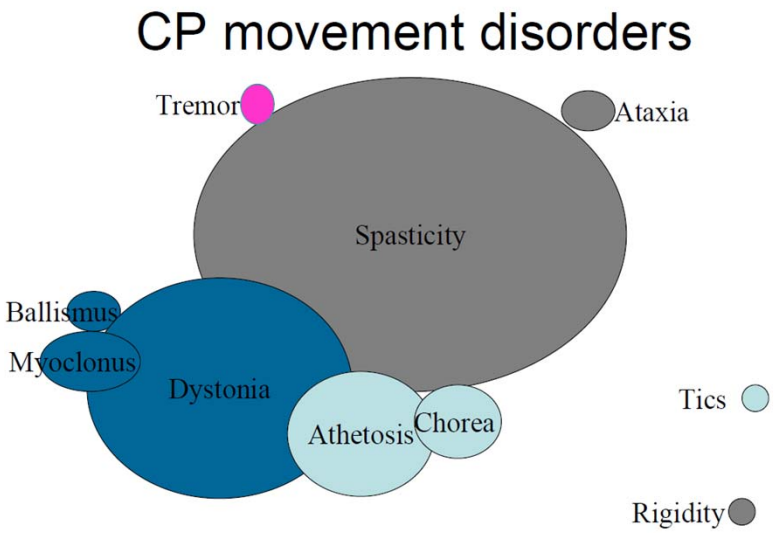
1. Rigidity
2. Spasticity
3. Dystonia
4. Tremor
5. Clonus

CP movement disorders



Spasticity may coexist with other conditions

Do not confuse with:



- 1. Rigidity – Involuntary, bidirectional, non-velocity-dependent resistance to movement
- 2. Clonus – Self-sustaining, oscillating movements secondary to hypertonicity
- 3. Dystonia – Involuntary, sustained contractions resulting in twisting, abnormal postures
- 4. Athetosis- Involuntary, irregular, confluent writhing movements
- 5. Chorea – Involuntary, abrupt, rapid, irregular, and unsustained movements
- 6. Ballism – Involuntary flinging movements of the limbs or body
- 7. Tremor – Involuntary, rhythmic, repetitive oscillations that are not self-sustaining

Goals of spasticity management

1. To improve function related to the activities of daily living, mobility, the ease of care by caregivers, sleep, cosmesis, and overall functional independence

2. To prevent orthopedic deformity, the development of pressure areas, and the need for corrective surgery

3. To reduce pain

4. To allow the stretching of shortened muscles, the strengthening of antagonistic muscles, and the appropriate orthotic fit

- Lance: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. (from Young, Spasticity:A review, Neurology, 44(Supp 9),Nov 1994, S12-20)

- Positive symptoms- exaggerated deep tendon reflexes, dystonia and contractures
- Negative symptoms-paresis and fatiguability
- Variations occur depending on which part of the CNS is damaged.
- Lesions just of corticospinal tract cause Babinski and paresis, without dystonia (internal capsule)
- Lesions of parasagittal perirolandic area(often traumatic) severely dystonic early on

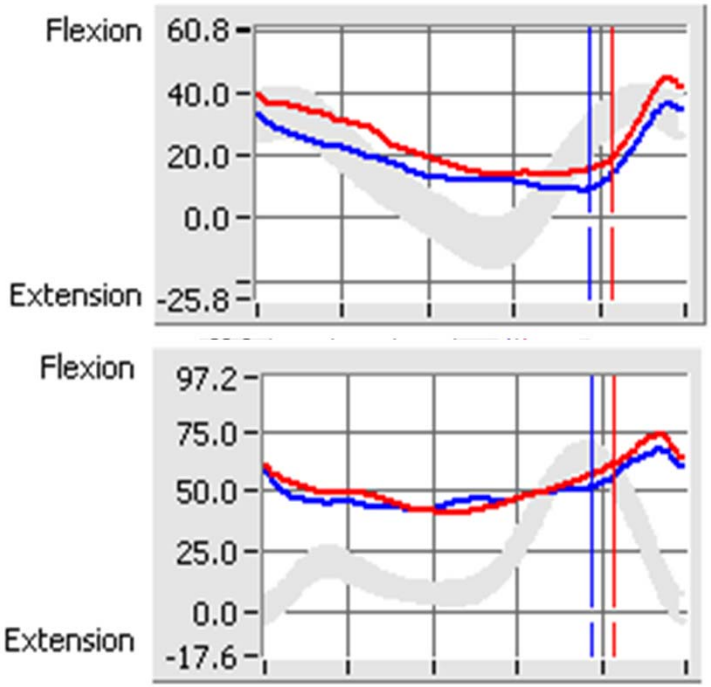
- "Group of disorders of movement and posture, causing activity limitation, that are attributed to nonprogressive disturbances that occurred in the developing fetal or infant brain. The motor disorders are often accompanied by disturbances of sensation ,cognition,communication,perception &/or behavior, &/or a seizure disorder." Bax et al DMCM, 47(8):571-575,2005

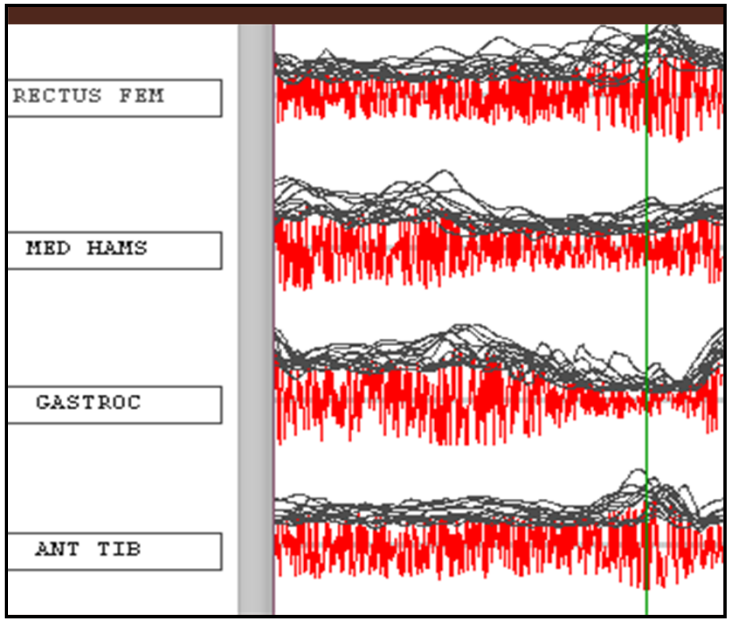
- Prematurity: Periventricular Leukomalacia
- HIE(Hypothermia treatment)
- Intrauterine infections, chorioamnionitis
- Maternal coagulopathy- Pre or perinatal stroke and hemmorhage
- Development Brain Malformations
- Incidene 1.5 to 2.5/1000 births(stable incidence due to greater neonatal survival)

- Hemiplegia- One arm and leg(stroke)
- Quadriplegic-Severe involvement all limbs (Severe HIE or brain malformations)
- Diplegic-Both legs primarily- often some arm involvement(PVL-premature)
- Dyskinetic-Chorea,athetosis,dystonia- Often with spasticity-Kernicterus,metabolic
- Ataxic(hypotonic)-Heterogeneous causes

Evaluation of Tone: Clinical Rating Scales

- Modified Ashworth scale
  - 0 to 4 scale of tone intensity
- Oswestry scale
  - rates stage and distribution of tone
  - "useful" vs. "non-useful" movement
- Degree of adductor muscle tone (Snow)
- Tardieu scale
  - measures tone vs. velocity


Flexion
60.8
40.0
20.0
0.0
Extension
-25.8
Flexion
97.2
75.0
50.0
25.0
0.0
Extension
-17.6


RECTUS FEM
MED HAMS
GASTROC
ANT TIB

No meaningful plan can be formulated without first determining the goals of treatment including:

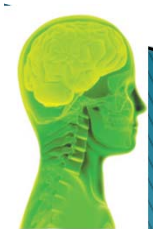
- patient/caregiver goals
- functional goals
- technical goals

- Increased ROM
- Decrease energy expenditure
- Decreased spasm frequency
- Decreased pain

- Improved mobility
- Improved gait
- Improved orthotic fit
- Improved positioning
- Increased ease of hygiene
- Improved cosmesis

- Neurologist
- Physical Therapist and Occupational Therapist
- Physiatrist
- Orthopedic surgeon
- Family and other caregivers
- Coordinator/administrator
- Wheelchair clinic, gait lab, orthotics clinic, counseling, social work

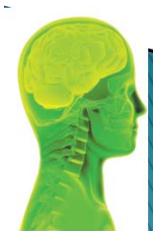
- Neurosurgeon



Weill Cornell Medical College

NewYork-Presbyterian
Phyllis and David Komansky Center for Children's Health
Pediatric
Brain and Spine Center

- Rehabilitation
- Oral medication
- Intrathecal baclofen
- Chemodenervation
- Neurosurgery
- Orthopedic surgery



Weill Cornell Medical College

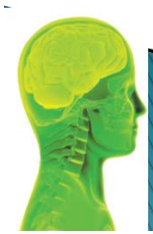
NewYork-Presbyterian
Phyllis and David Komansky Center for Children's Health
Pediatric Brain and Spine Center

- Segmental Activity
- Alpha motor neuron
- 1a EPSPs
- 1a inhib interneuron
- Renshaw inhibition
- Presynaptic inhibition
- Polysynaptic paths

- Neurotransmitter
- Acetylcholine(Nicotin)
- Glutamate
- Glycine
- Glycine
- GABA
- Serotonin,TRH

NEUROTRANMITTER AGONISTS

Neurotransmitter	Agonist
Acetylcholine-N	Nicotine
Glutamate(EAA)	NMDA,AMPA
Glycine	Threonine
GABA A receptor	Benzos, Barbs
GABA B receptor	Baclofen
Serotonin	Buspirone, Imitrex



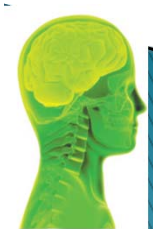
Weill Cornell Medical College

NewYork-Presbyterian
Phyllis and David Komansky Center for Children's Health
Pediatric
Brain and Spine Center

NEUROTRANSMITTER ANTAGONISTS IN SPINAL CORD

- Neurotransmitter
- Acetylcholine
  Glycine
- GABA A
- GABA B
- Serotonin
- Opioids
- ?Agmatine

- Antagonist
- Trimethaphan
- Strychnind
- Picrotoxin
- Baclofen
- Tricyclics
- Naloxone
- Alpha adrenergic agonist (Clonidine, tizanidine)



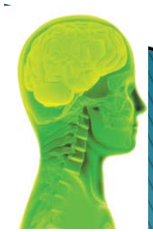
Weill Cornell Medical College

NewYork-Presbyterian
Phyllis and David Komansky Center for Children's Health
Pediatric
Brain and Spine Center

- Different medications act at different locations in CNS; ?polypharmacy
- Side effect profile as important as benefits, especially in children eg sedation
- Will benefit also result in functional loss (Decreased spasticity but increased weakness)

- Structural analogue of GABA that inhibits mono and polysynaptic spinal reflexes
- Binds to GABA B receptor–Coupled to Ca & K channels, pre and postsynaptically
- Presynaptic membrane hyperpolarized & influx Ca restricted with decrease NT release in excitatory spinal path(decrease alpha motor neuron activity). Other effects (gamma inhibition,decrease muscle spindle activity).

- Studies most in spinal cord(MS)-spasms
- Benefit with decreased spasticity, spasms
- Side effects; Increased weakness, fatigue, attention and memory issues, seizures (also with sudden withdraw),elevated LFTs
- Half life 3.5 hours(may need 4/day)
- Up to 30- 60 mg in children(Gracies)



Weill Cornell Medical College

NewYork-Presbyterian
Phyllis and David Komansky
Center for Children's Health

Pediatric
Brain and Spine Center

- Sustained or repetitive involuntary muscle contractions that produce abnormal but patterned postures and movements of different parts of the body(Fahn)
- Syndrome of sustained muscle contraction ,frequently causing twisting and repetitive movements or abnormal posture(Dystonia Foundation)

- DYT1(DMD)-AD- Deletion 9q34 gene, Protein torsin-Deep brain stimulation of globus pallidus
- DYT5-Dopa-Responsive Dystonia-Segawa diurnal fluctuations, foot dystonia presents in childhood(mimic CP)-AD with marked phenotypic variability- 14q22.1-2, Protein GTP cyclohyrolase-Treated with Carbidopa 25/100mg

- Most common etiology associated with CP Also multiple genetic causes(eg Wilson's, glutaric aciduria), Stroke, Medication
- Carbidopa- At times partial response
- Anticholinergics
- Baclofen
- Botulinum

- Injectable therapy
- Results in local muscle weakening
- Temporary and titratable
- Agents include:
  - Botulinum toxin
  - Phenol
  - Ethyl alcohol

- Produced by the bacterium, *Clostridium botulinum*
- Seven serotypes (A–G); only "A" and "B" approved for clinical use
- Trade names of BTX-A:
  - BOTOX® (Allergan)
  - DYSPORT® (Ipsen, Ltd.)
- Conversion ratio: 1 Unit BOTOX~3-5 Units Dysport
- Trade name of BTX-B, from Elan:
  - Myobloc™ in USA
  - NeuroBloc® in Europe

- Injected directly into overactive muscle
- Focal, temporary chemodenervation
- Onset usually within 24–72 hours, maximum effect at approximately 2 weeks
- Clinical benefit usually >12 weeks; may be extended with adjunctive therapy
- Preliminary BTX-B results in spasticity indicate clinical efficacy; autonomic side effects in some patients

Delivered via an implantable, programmable pump

For management of severe spasticity due to:

- Cerebral palsy
- Stroke
- Brain injury
- Spinal cord injury
- Demyelinating disease

$GABA_b$ agonist

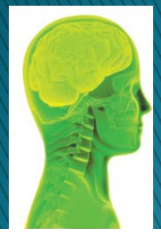
- Decreases interneuronal firing at the spinal cord level

Oral dose of 60 mg ⇒ lumbar concentration of 24µg/L

Intrathecal infusion of 600 µg/day ⇒ lumbar concentration of 1240µg/L

- cervical concentration approximately 1/4th lumbar

Most patients maintained on 300-800µg/day



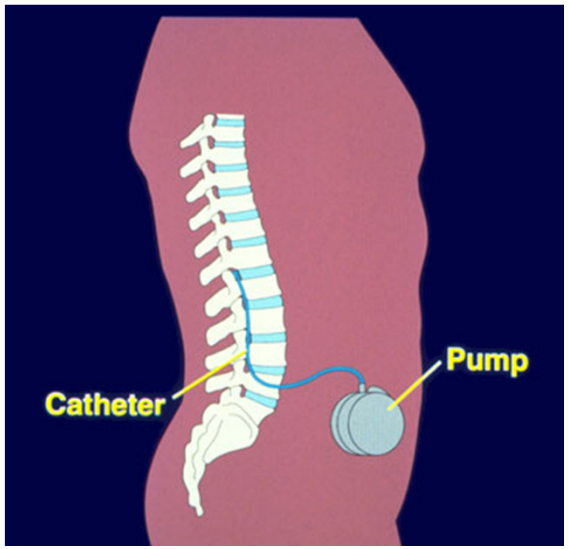
Weill Cornell Medical College

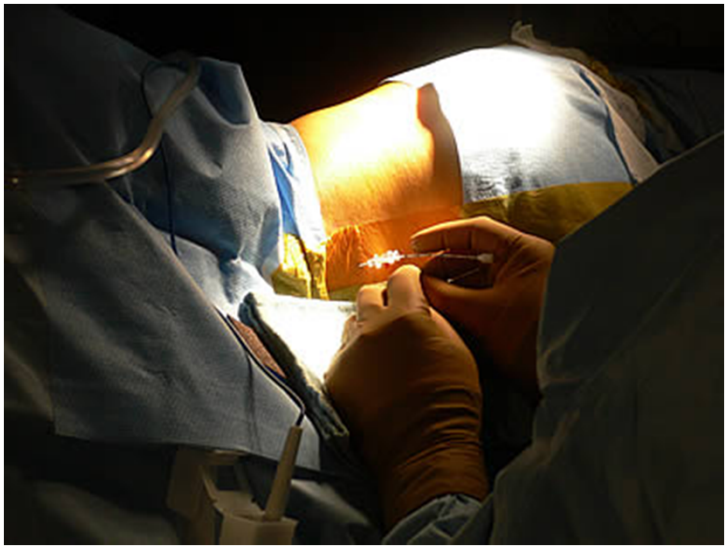
NewYork-Presbyterian
Phyllis and David Komansky Center for Children's Health
Pediatric Brain and Spine Center

- Pump controlled via radio-telemetry link from an external programmer
- Allows control of rate, mode, and pattern
  of infusion
- Dosage titration, schedule revision

- Indicated for patients with severe spasticity in whom oral medications have proven ineffective
- Age of 3-4 or older and at least 30-35 pounds
- Responsive to the trial dose
- Patient and family have commitment to program

- Pump implanted in a subcutaneous or subfascial pouch in the abdomen just under the rib cage
- Catheter is placed in the lumbar spine (usually L3 or L4) with a 15 or 16 gauge Touhy needle
- Tip placement at the T10–T11 level is confirmed using fluoroscopy





- Dose adjustments performed in office using portable computerized programmer via a telemetry wand
  - Adjustable parameters include: drug concentration, reservoir status, alarms, infusion rate, and pattern,
    e.g., more at night and less during school
- Dose may be increased by up to 15% on each occasion on a weekly basis
- Temporal adjustments for functional needs
- Adjustments in adjunctive therapies

Side effects are similar to oral medication and may be minimized by adjusting dose:

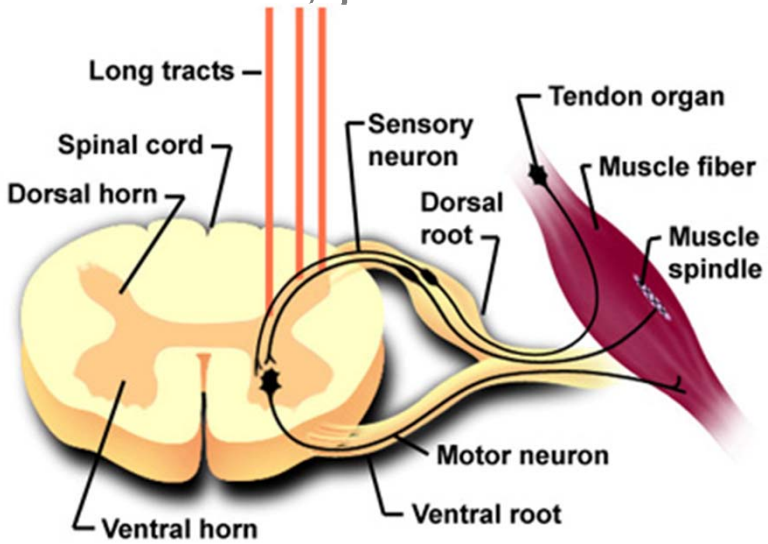
- Hypotonia
- Somnolence
- Nausea/vomiting
- Headaches
- Dizziness

- Catheter and procedural complications
- Infection
- Overdose potential
  - Respiratory depression
  - Loss of consciousness
- May be associated with a worsening of scoliosis in some patients

- Interruption of reflex arc
- Sectioning of afferent nerve rootlets L-2 to S-2
- EMG guidance, selected roots only

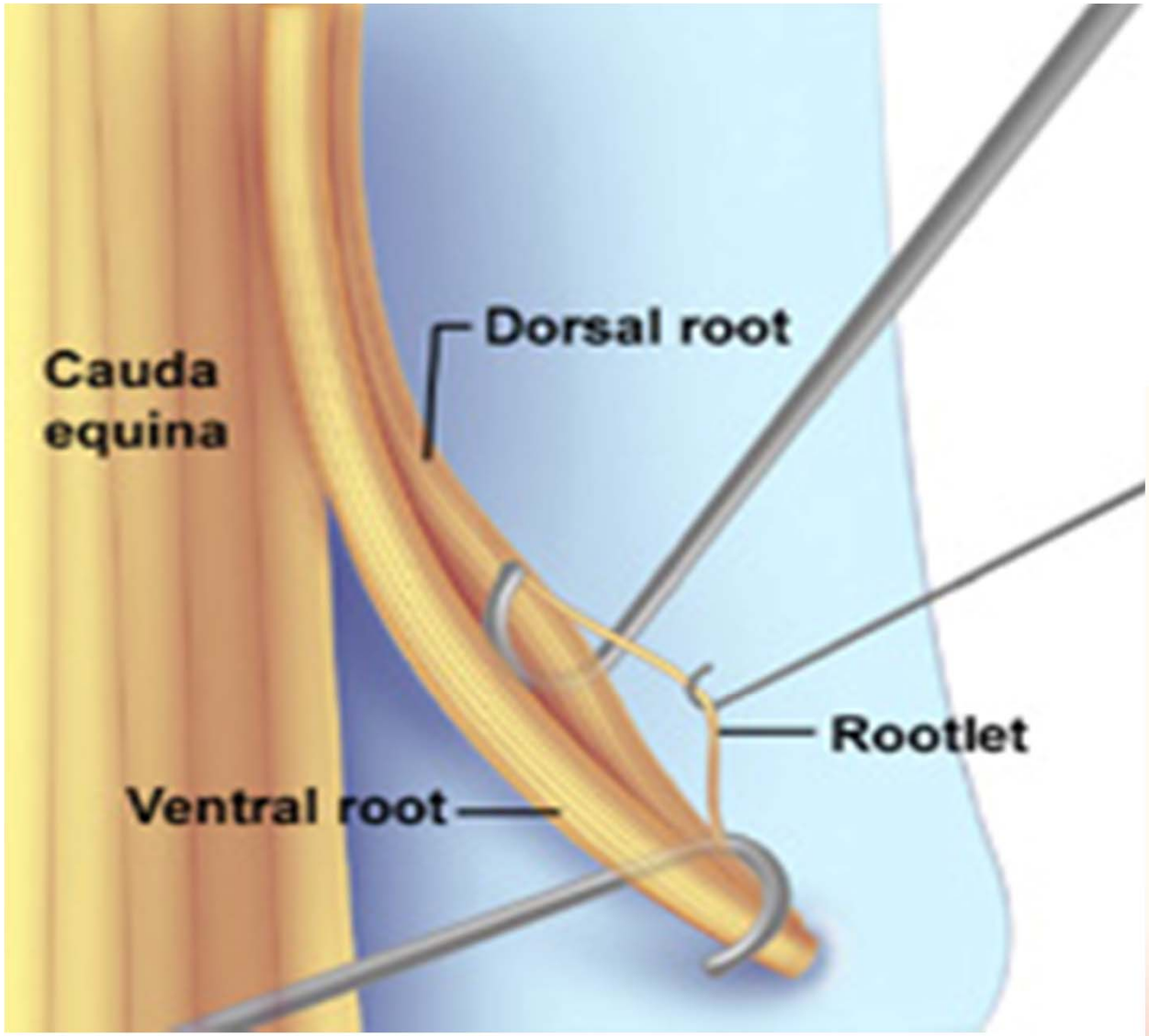
Treatment goals:

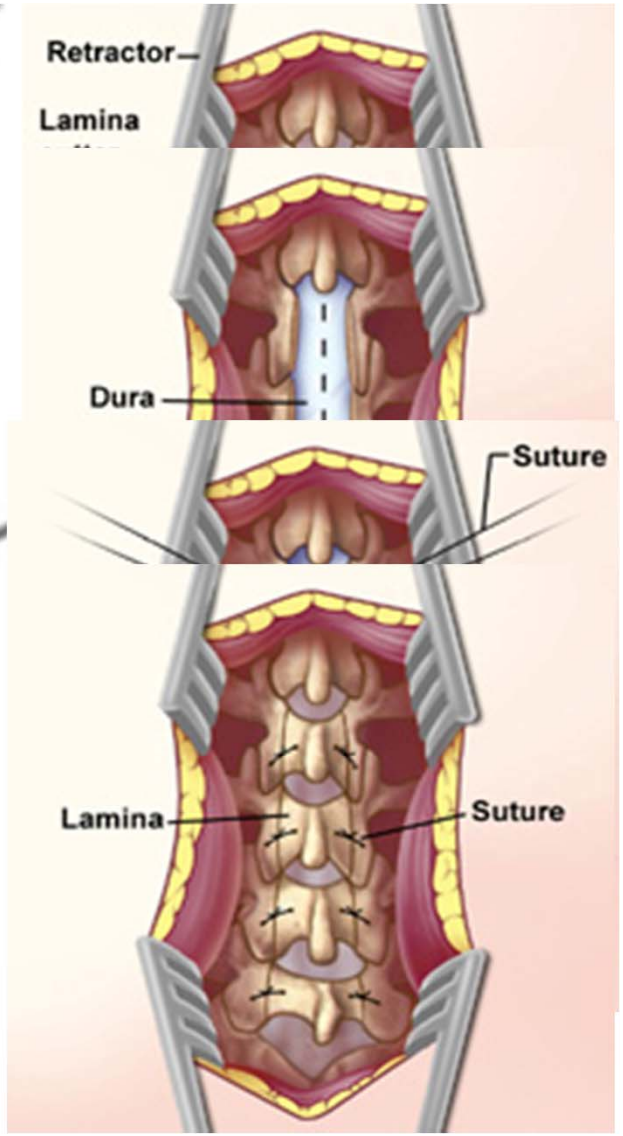
Improve gait and mobility; facilitate care; prevent contractures or bony deformities



- Pure spasticity
- No evidence of dystonia (fluctuating tone) or rigidity (constant resistance to force)
- No evidence of truncal hypotonia
- Good head control
- Best age is between 4 and 6

- Spasticity of spinal cord origin
- Athetosis
- Rigidity
- Poor trunk control
- Severe weakness


Cauda equina
Dorsal root
Rootlet
Ventral root


Retractor
Lamina
Dura
Suture
Lamina
Suture

- Pain management very important
- Physical therapy
  - Intensive PT within one week
  - 3–5 times per week depending on the center
- Ambulatory patients begin walking as soon as possible
- Strengthening is a key component after rhizotomy

HOSPITAL FOR SPECIAL SURGERY

- 6 year old
- Diagnosed with Cerebral Palsy
- Gait Analysis Video from Hospital for Special Surgery

Weill Cornell Medical College

NewYork-Presbyterian
Phyllis and David Komansky Center for Children's Health
Pediatric Brain and Spine Center

What does Sofia have?

Spastic hemiplegia
Spastic diplegia
Spastic quadriplegia
Dystonia
Choreoathetosis

Time to laugh