SURGICAL MANAGEMENT OF ABNORMAL TONE

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DISCLOSURES

- No financial disclosures to report

OBJECTIVES

- Identify candidates for surgical management of increased tone
- Describe surgical options for appropriate candidates
- Identify risks, benefits, and alternatives of surgery

ABNORMAL TONE

- Spasticity
- Dystonia
- Chorea
- Athetosis
- Rigidity
- Atonia

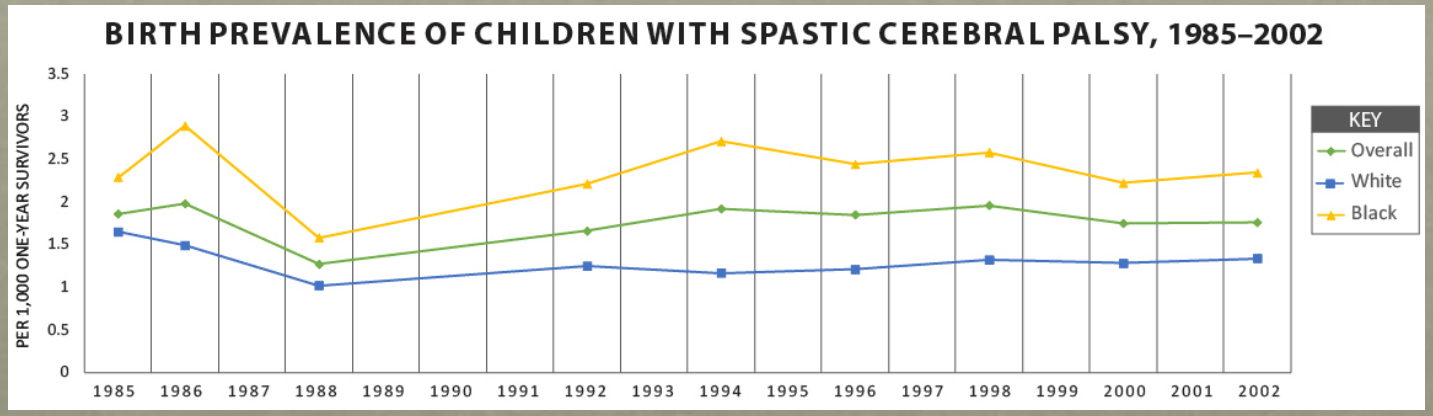
INCIDENCE

- Cerebral palsy
  - **1 in 3000 live births**
- Stroke
- Head injury
- Brain tumor
- Hereditary
- Idiopathic

TABLE. Estimated prevalence and lifetime economic costs* for mental retardation, cerebral palsy, hearing loss, and vision impairment, by cost category — United States, 2003

Developmental disability	Rate†	Direct medical costs§ (millions)	Direct nonmedical costs¶ (millions)	Indirect costs** (millions)	Total costs (millions)	Average costs per person
Mental retardation	12.0	$7,061	$5,249	$38,927	$51,237	$1,014,000
Cerebral palsy	3.0	1,175	1,054	9,241	11,470	921,000
Hearing loss	1.2	132	640	1,330	2,102	417,000
Vision impairment	1.1	159	409	1,915	2,484	566,000

* Present value estimates, in 2003 dollars, of lifetime costs for persons born in 2000, based on a 3% discount rate.
† Per 1,000 children aged 5–10 years, on the basis of Metropolitan Atlanta Developmental Disabilities Surveillance Program data for 1991–1994.
§ Includes physician visits, prescription medications, hospital inpatient stays, assistive devices, therapy and rehabilitation (for persons aged <18 years), and long-term care (for persons aged 18–76 years), adjusted for age-specific survival.
¶ Includes costs of home and vehicle modifications for persons aged ≤76 years and costs of special education for persons aged 3–17 years.
** Includes productivity losses from increased morbidity (i.e., inability to work or limitation in the amount or type of work performed) and premature mortality for persons aged ≤35 years with mental retardation, aged ≤25 years with cerebral palsy, and aged ≤17 years with hearing loss and vision impairment.



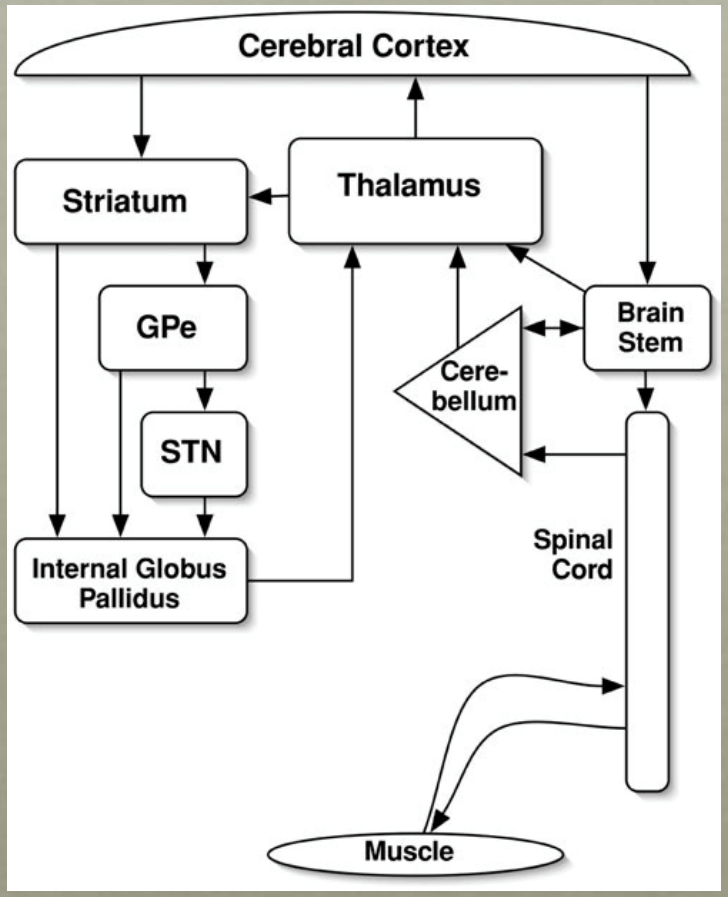
MULTIDISCIPLINARY MANAGEMENT

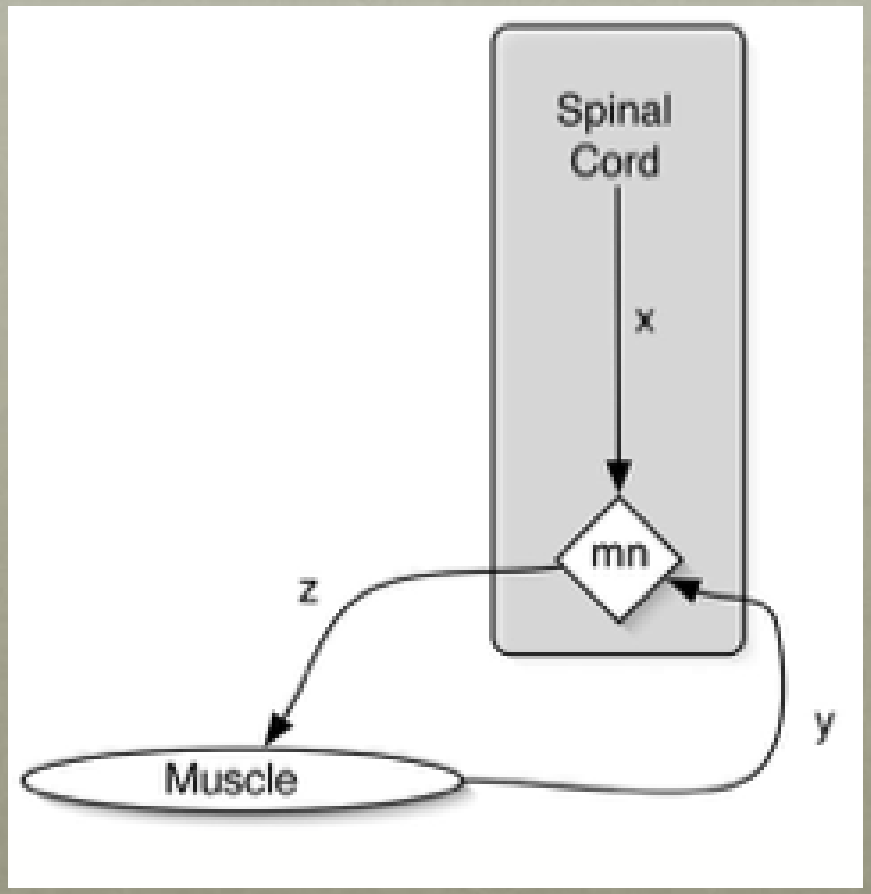
- Pediatrician
- Neurologist
- Neurosurgeon
- Physiatrist
- Physical therapist
- Occupational therapist
- Speech therapist

SPASTICITY

- "resistance to externally imposed movement that increases with increasing speed of stretch and varies with the direction of joint movement, and/or resistance to externally imposed movement that rises rapidly above a threshold speed or joint angle"
- Varies with movement, alertness, pain, anxiety
- Isokinetic, not hyperkinetic
- Most common type of abnormal tone
- Affects **60% of cerebral palsy patients**; stroke, head injury, idiopathic
- Classified by affected limbs – spastic diplegia, spastic hemiplegia, spastic quadriplegia, spastic monoplegia

SPASTICITY


Cerebral Cortex
Striatum
Thalamus
GPe
Brain Stem
Cere-bellum
STN
Spinal Cord
Internal Globus Pallidus
Muscle

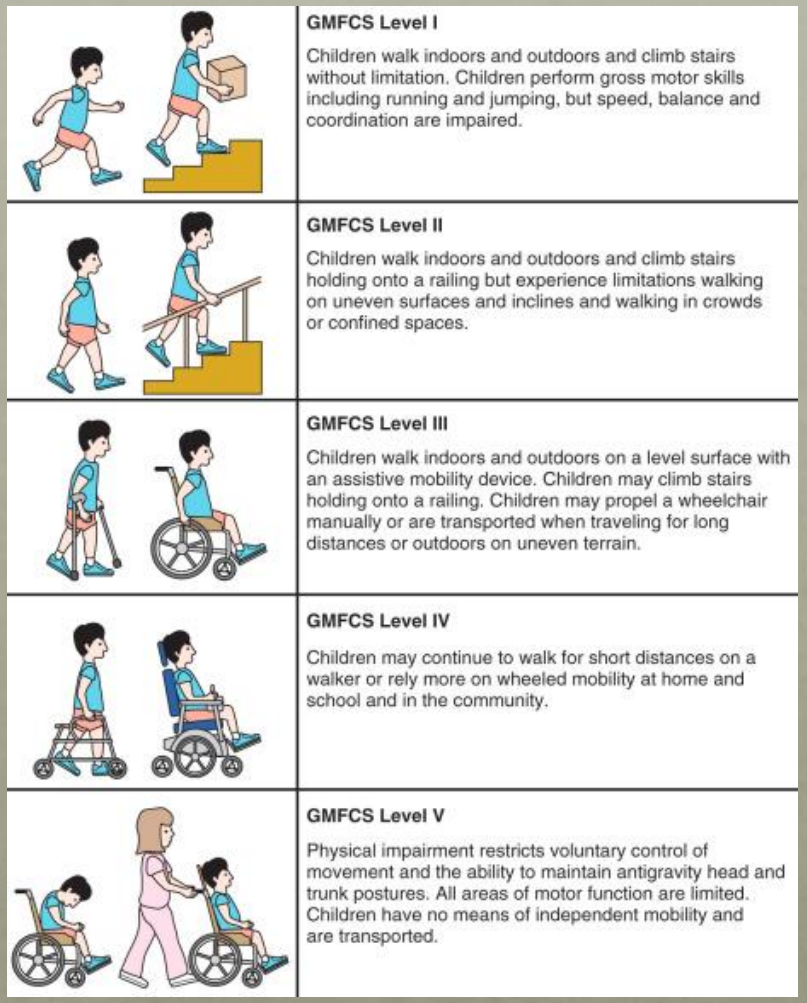

Spinal Cord
x
mn
z
y
Muscle

MANAGEMENT

- Goals of treatment
  - Improve function
  - Facilitate care
  - Reduce contractures
  - Reduce pain
- Oral medications
  - Baclofen (GABA agonist), diazepam, tizanidine
- Botox injections
- Therapy
- Surgery

EVALUATION

Grade	Description
0	no increase in muscle tone
1	slight increase in muscle tone, manifested by a catch and release or by minimal resistance at the end of the range of motion when the affected part(s) is moved in flexion or extension
1+	slight increase in muscle tone, manifested by a catch, followed by minimal resistance throughout the remainder (less than half) of the ROM
2	more marked increase in muscle tone through most of the ROM, but affected part(s) easily moved
3	considerable increase in muscle tone, passive movement difficult
4	affected part(s) rigid in flexion or extension



GMFCS Level I

Children walk indoors and outdoors and climb stairs without limitation. Children perform gross motor skills including running and jumping, but speed, balance and coordination are impaired.

GMFCS Level II

Children walk indoors and outdoors and climb stairs holding onto a railing but experience limitations walking on uneven surfaces and inclines and walking in crowds or confined spaces.

GMFCS Level III

Children walk indoors and outdoors on a level surface with an assistive mobility device. Children may climb stairs holding onto a railing. Children may propel a wheelchair manually or are transported when traveling for long distances or outdoors on uneven terrain.

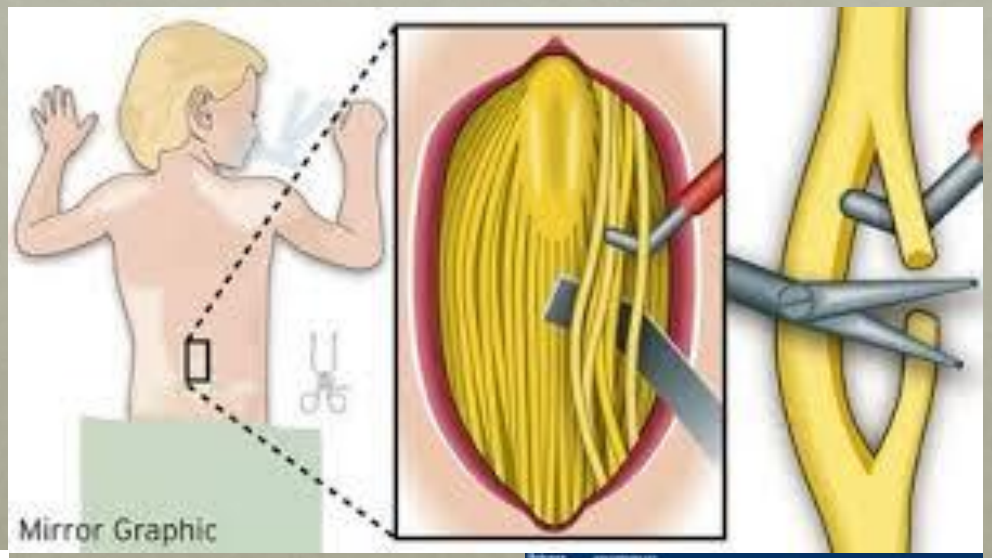
GMFCS Level IV

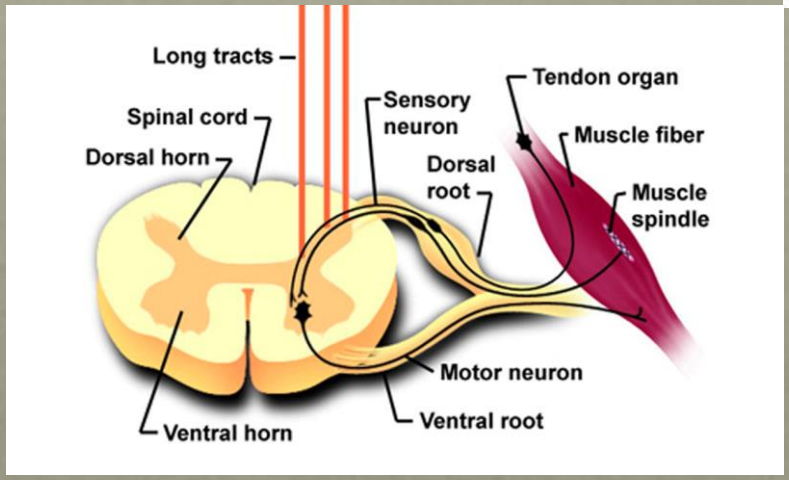
Children may continue to walk for short distances on a walker or rely more on wheeled mobility at home and school and in the community.

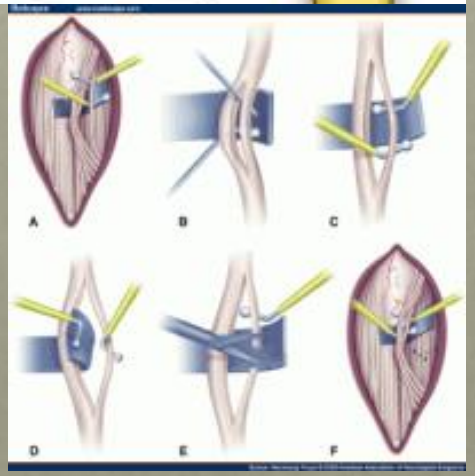
GMFCS Level V

Physical impairment restricts voluntary control of movement and the ability to maintain antigravity head and trunk postures. All areas of motor function are limited. Children have no means of independent mobility and are transported.

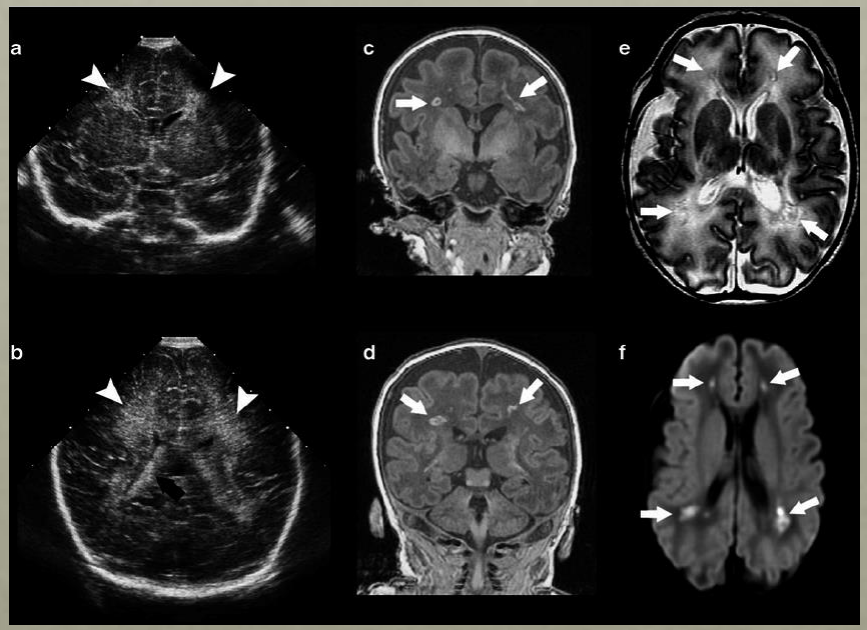
SELECTIVE DORSAL RHIZOTOMY

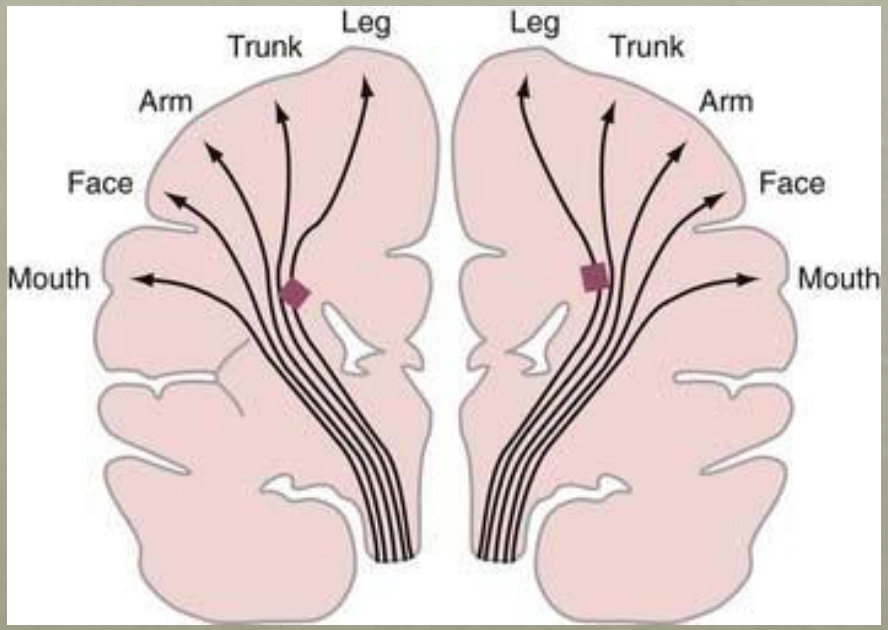






SELECTIVE DORSAL RHIZOTOMY





INDICATIONS/ CONTRAINDICATIONS

TABLE 227-1 Indications for Selective Dorsal Rhizotomy for Spastic Cerebral Palsy

Diagnosis—spastic diplegia or quadriplegia
Age—2 to 40 years
History of premature birth or neonatal asphyxia
Emerging locomotive functions
Potential for significant postoperative functional gain

TABLE 227-2 Contraindications to Selective Dorsal Rhizotomy for Spastic Cerebral Palsy

CP associated with intrauterine encephalitis
Mixed CP with predominant dystonia or ataxia
CP caused by a widespread neuronal migration disorder
Severe head injury and hypoxic encephalopathy, such as after drowning
Familial spastic paraplegia and other progressive neurological disorders
Severe basal ganglia damage in children younger than 5 years
Severe thoracolumbar scoliosis
Severe lumbar lordosis
Multiple previous muscle and tendon releases
Profound motor impairment with no head control
Psychiatric disorders in adults
Lack of commitment to carry out postoperative therapy

RISKS

- Bleeding
- Infection
- CSF leak
- Spinal deformity

- https://youtu.be/IcFcDgNEwUE

- https://youtu.be/wA7TKfI2WeM

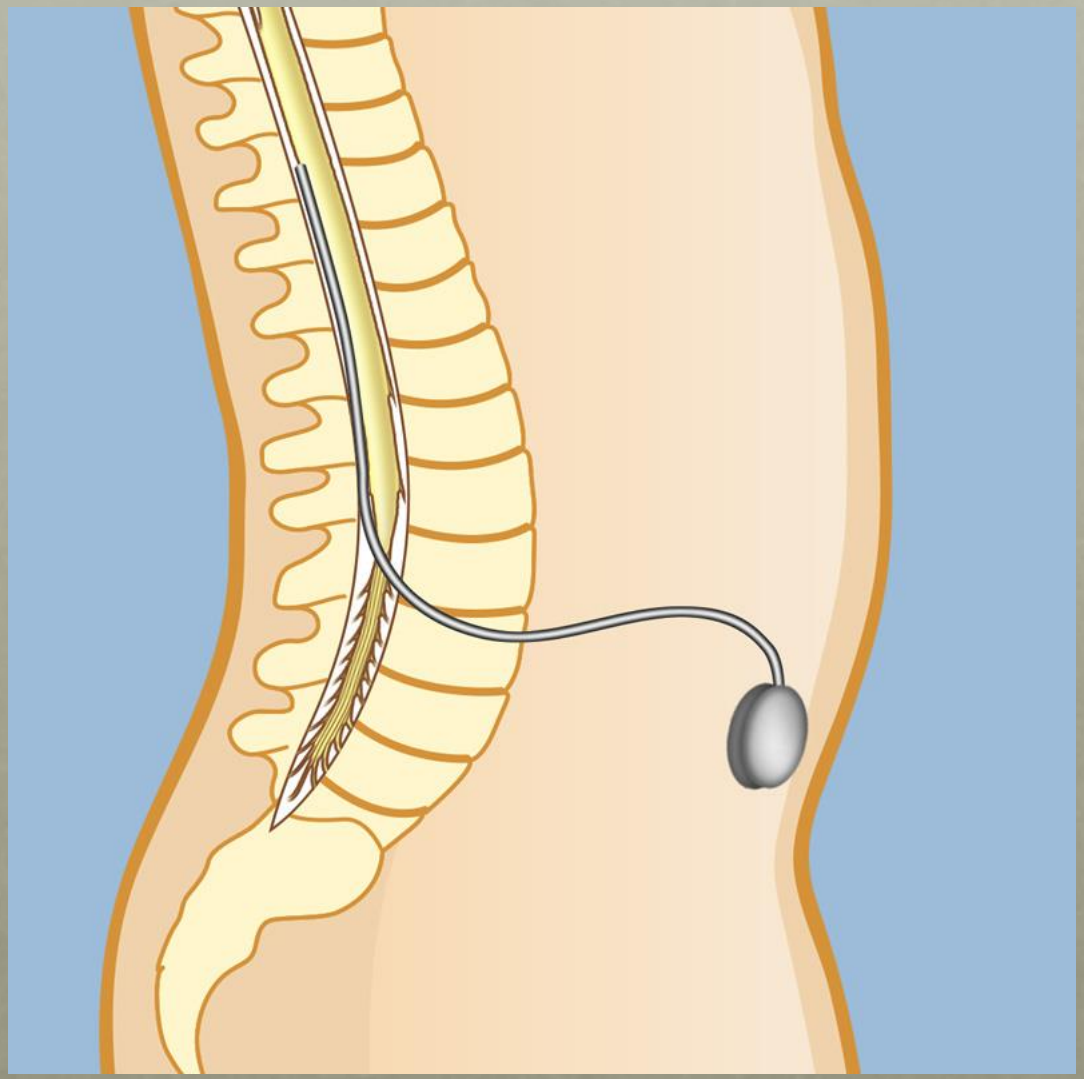
INTRATHECAL BACLOFEN PUMP

- Intrathecal catheter with subcutaneous pump/reservoir
- 20cc or 40cc reservoir
- Suitable for pts >10kg

- Catheter tip can be cervical or thoracic
- Can titrate dose to effect

IT baclofen – 396ug/day = CSF level 400ng/ml

PO baclofen - 60mg/day = CSF level 12ng/ml



RISKS

- Mechanical failure
- Catheter fracture/migration
- Infection
- **CSF leak**
- Need for lifelong follow up
- Risk of withdrawal or overdose

- https://youtu.be/JdwCbH77MYg

SUMMARY

Table 76.3 Treatments for Spasticity

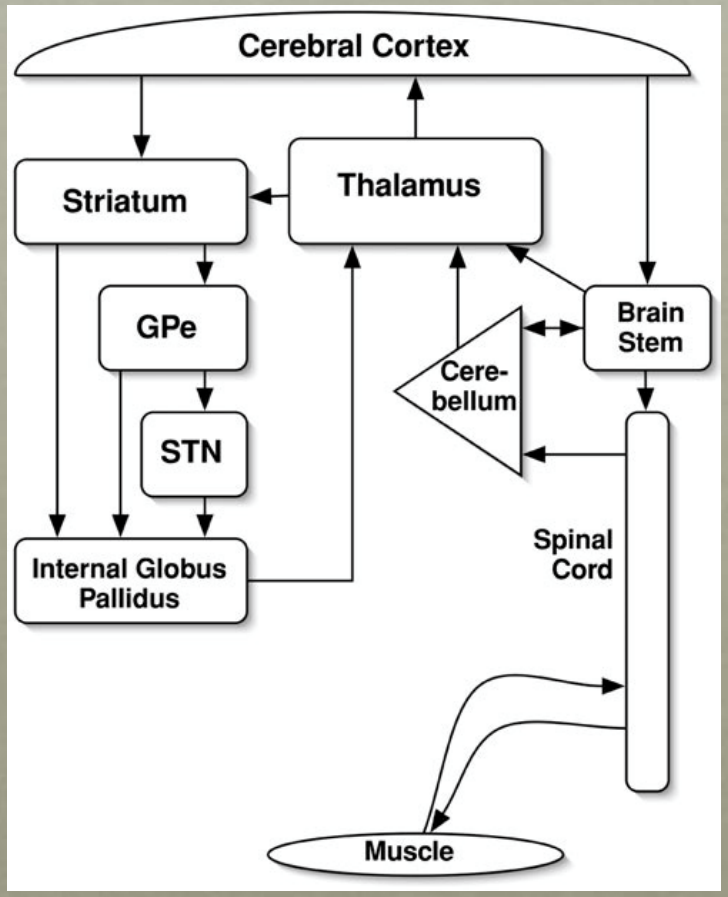
Method	Age (y)	Diagnosis	Characteristics	Expected results	Follow-up care	Outcome	Side effects, risk
Oral medications	Any age, most often 2 to 5	Spastic quadriplegia, traumatic brain injury	Diffuse spasticity	Mild decrease in spasticity in arms and legs	PT, OT as needed	SPRs or ITB often needed later	Drowsiness
Botox injections	Any age, less often older than 10	Spastic diplegia, spastic quadriplegia	Isolated spasticity; too young for ITB, SPRs	Decrease in spasticity in injected muscle(s) for 2 to 4 months	PT, OT to increase range of motion and to increase strength	Improved gait, sometimes improved arm function	None with usual doses
Rhizotomy (SPRs)	4 to 8 (most common), rare after 16	Spastic diplegia or quadriplegia, *capable* of ADLs	Good leg strength, no severe contractures, motivation for PT	Marked, permanent, nonadjustable decrease in spasticity	Extensive PT, OT	Improved walking, improved ADLs, decrease in orthopedic operations	Infection, 2%; wound, 1%; CSF leak, 3%
	Older than age 3, before multiple contractions	Spastic diplegia or quadriplegia, *not capable* of ADLs	Severe leg spasticity interfering with care		Minimal	Easier care	Infection, 2%; wound, 1%; CSF leak, 3%
Baclofen (ITB)	Older than age 3, big enough to insert pump	Spastic quadriplegia, spasticity in legs greater than or equal to spasticity in arms, *capable* of ADLs	Severe spasticity, positive response to test dose, spasticity limiting function	Adjustable decrease in spasticity	Frequency of PT, OT depends on goals	Improved walking, improved ADLs, improved speech, decrease in orthopedic operations, easier care	Infection, 5 to 10%; wound, 5 to 10%; CSF leak, 5 to 10%
		Spastic quadriplegia, *not capable* of ADLs	Spasticity interfering with care		Minimal PT	Easier care	
		Posttraumatic brain injury	Severe spasticity in arms or legs, usually more than 1 year after injury		PT, OT for range of motion		

Abbreviations: ADLs, activities of daily living; CSF, cerebral spinal fluid; ITB, intrathecal baclofen; OT, occupational therapy; PT, physical therapy; SPRs, selective posterior rhizotomies.

DYSTONIA

- "a movement disorder in which involuntary sustained or intermittent muscle contractions cause twisting and repetitive movements, abnormal postures, or both"
- Hyperkinetic
- Focal, segmental, hemidystonia, generalized
- Primary (hereditary – DYT genes) vs secondary (CP, stroke, head injury, tumor)
- Affects 15-25% of cerebral palsy patients

DYSTONIA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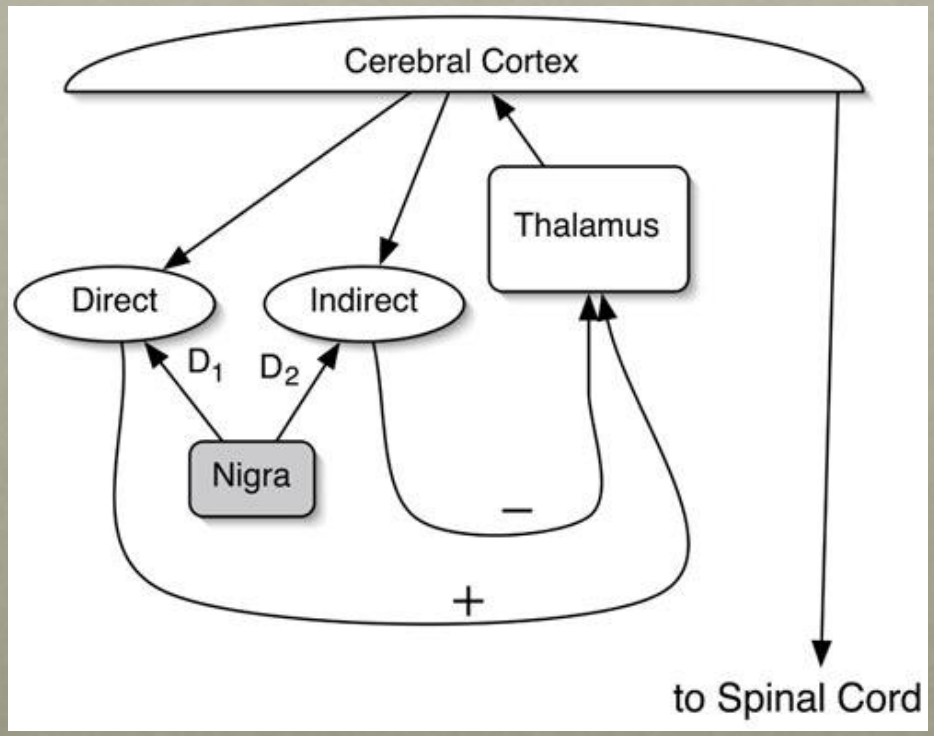
Cerebral Cortex
Thalamus
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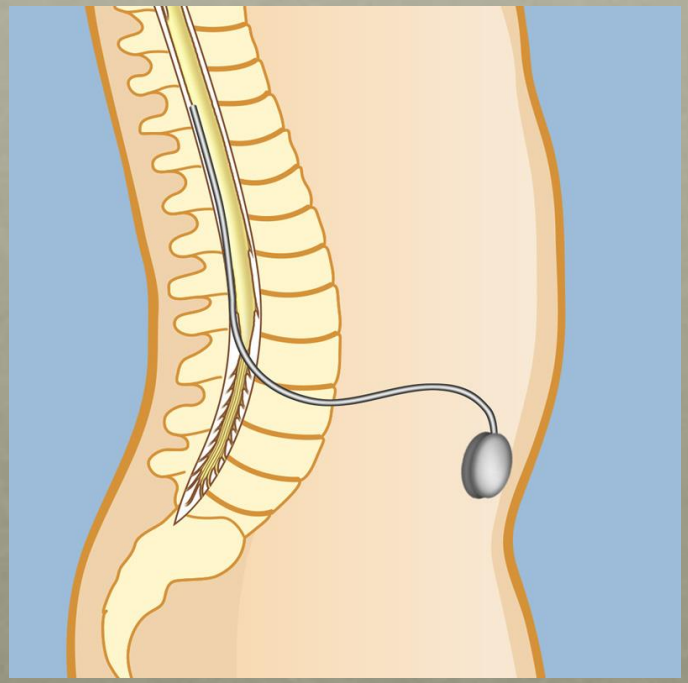
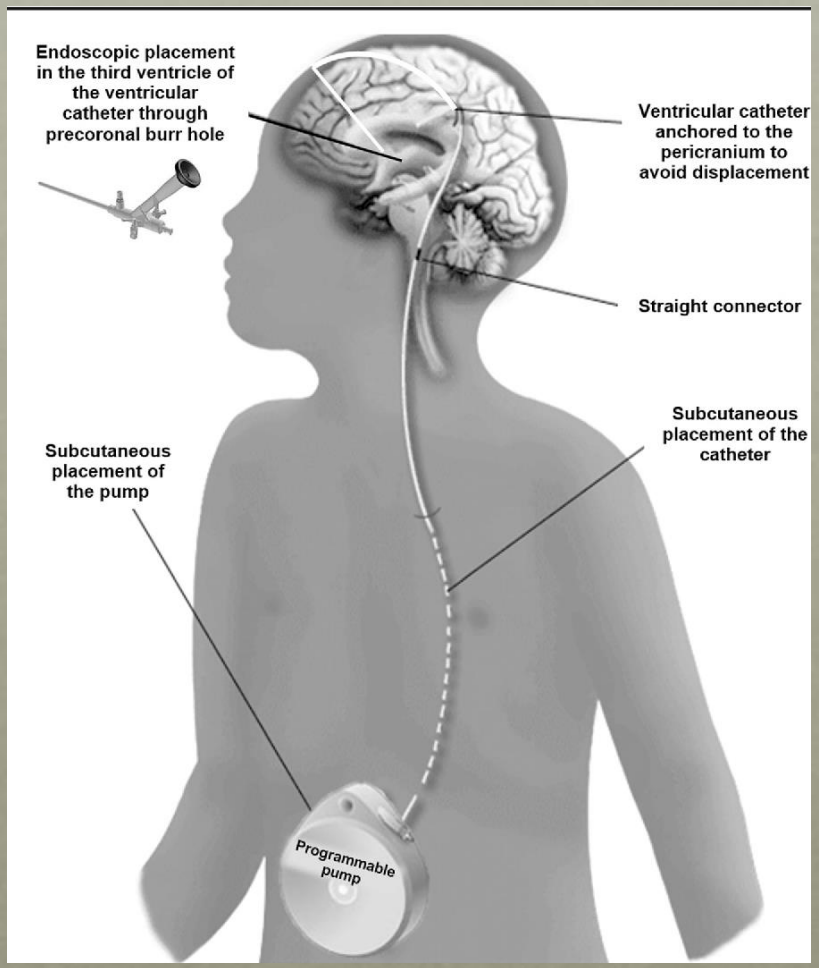
MANAGEMENT

- Goals of treatment
  - Improve function
  - Facilitate care
  - Reduce contractures
  - Reduce pain
- Oral medications
  - Baclofen, trihexyphenidyl (Artane), dopa
- Botox injections
- Surgery

INTRATHECAL BACLOFEN PUMP

- Intrathecal catheter with subcutaneous pump/reservoir
- 20cc or 40cc reservoir
- Suitable for pts >10kg





DEEP BRAIN STIMULATION

- Most experience is with adult movement disorders (Parkinson's disease, essential tremor)
- Has been used in pediatric dystonia
- Appears more effective for **primary** dystonia
- Typically target is globus pallidus interna
- Modulates the cortical-basal ganglia-cortical loop

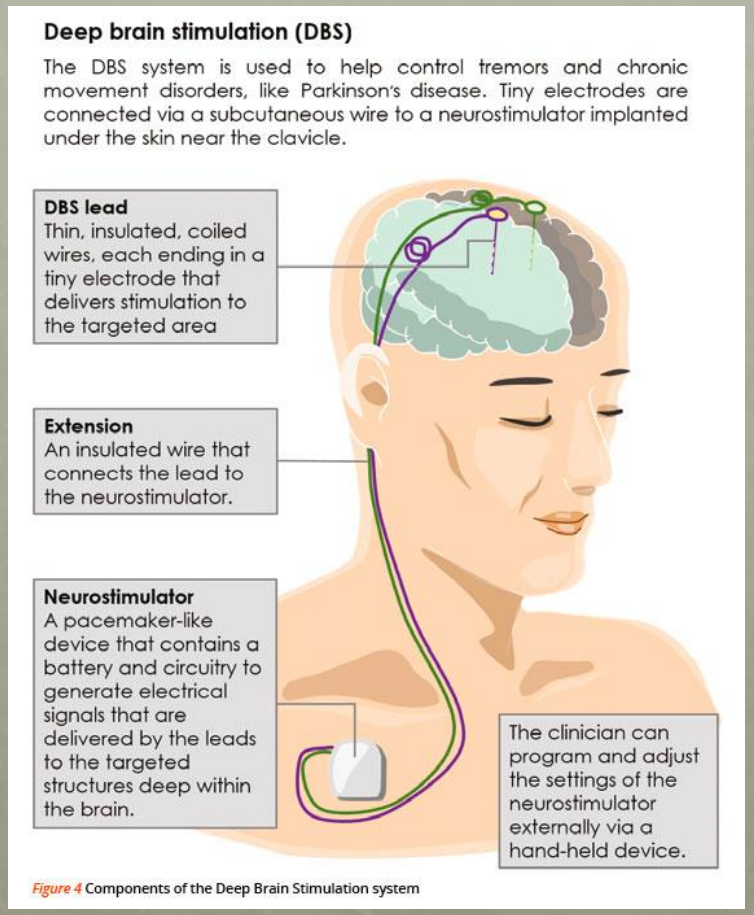


Figure 4 Components of the Deep Brain Stimulation system

- https://youtu.be/jtyCTk3OBpQ

ATHETOSIS/CHOREA

- Athetosis = “slow, distal, wormlike, writing movements of the extremities, with fanning or hyperextension of the digits”
- Chorea = “a state of excessive, spontaneous movements, irregularly timed, nonrepetitive, abrupt in character, involuntary, rapid, brief, unsustained, dancelike movements”
- Do not respond well to surgical management – important to distinguish from dystonia

- https://youtu.be/XCzdUiSo8Y8

UNM TONE CLINIC

- First Friday of every month
- Staff
  - James Botros, MD and Barbara Bell, NP, Neurosurgery
  - John Phillips, MD, Neurology
  - Denise Taylor, MD, PM&R
  - Angela Kouri, PT
  - Catherine Burke, PT
- Send referrals!

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