Spasticity in the Medically Complex Child

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Spasticity

Complex Care Curriculum

Spasticity

- Learning Objectives
- Definition
- Etiology & Pathophysiology
- Impact of Spasticity & Outcome Measures
- Exam Features
- Treatment Plan
- Treatment Options
- Medications
- Intrathecal Baclofen
- Selective Rhizotomy
- Chemodenervation
- Summary
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Describe the signs, symptoms and complications of spasticity

Identify the patient with spasticity that is not optimally managed

Summarize the different pharmacologic and non-pharmacologic treatment modalities

Plan a basic approach to managing spasticity in a patient

Identify the role of the consultant (PM&R) in helping with management

Identify the patient with baclofen withdrawal and initiate management

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Spasticity =

Motor Disorder

Velocity Dependent

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Velocity dependent

Stretch dependent

UMN signs

Muscle over-activity

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Upper Motor Neuron Syndrome

Positive Symptoms

- Spasticity
- Clonus
- Flexor/extensor spasm
- Hyper-reflexia

Negative Symptoms

- Weakness
- ↓dexterity
- Paralysis
- Fatigability
- ↓movement

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Brain Injury

- traumatic brain injury, cerebral palsy, stroke, bacterial meningitis, encephalitis, tumor, MS

Spinal Cord Injury

- tumor, infection, trauma, MS

Neuromuscular

- ALS, Friederich ataxia

Genetic disorders and degenerative diseases

- Sjogren-Larsson syndrome, Tay-Sachs disease, and Rett syndrome

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Cerebral Palsy :

- Disorders of movement and posture causing activity limitations
- Non-progressive disturbances of developing brain

Underlying lesion is static, but musculoskeletal pathology is progressive

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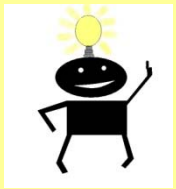
Musculoskeletal Progression in Cerebral Palsy

Static
CNS Lesion

Progressive
Musculoskeletal deformity

Upper Motor Neuron Syndrome
Spasticity and Weakness
Failure of longitudinal muscle growth
Fixed contracture
Bony torsion
Joint instability
Joint dislocation
Joint and bone degenerative changes



Must repeatedly screen children for joint dislocation as they grow

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Motor Control

Bowel/Bladder

Skeletal & Skin

Self Esteem

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- Muscle Tone
  - Ashworth Scale
  - Modified Ashworth Scale
- Muscle reaction at different velocity of stretch
  - Tardieu Scale
- Range of Motion
- Muscle Strength
- Disability/Function
- Overall Motor Function

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Ashworth Scale	
Grade	**Description**
0	No increase in tone
1	Slight increase in tone giving a 'catch' when the limb is moved in the flexion or extension
2	More marked increase in tone, but limb is easily flexed
3	Considerable increase in tone, passive movement difficult
4	Limb rigid in flexion or extension

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Modified Ashworth Scale	
Grade	**Description**
0	No increase in tone
1	Slight increase in tone - a catch and release at the end of the range of motion
1+	Slight increase in tone - catch, followed by minimal resistance in remainder of range
2	More marked increase in tone through most of range
3	Considerable increase in tone, passive movement difficult
4	Affcted parts rigid in flexion or extension

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Tardieu Scale
V1: As slow as possible (minimizing stretch reflex)*
V2: Speed of the limb segment falling under gravity**
V3: As fast as possible (faster than the rate of the natural drop of the limb segment under gravity)**
*V1 measures passive range of motion (PROM) ** V2 and V3 used to rate spasticity

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Observation of function

Identify movement disorder

Assess sensibility

Determine active and passive ROM

Evaluate spasticity, strength, and reflexes

Evaluate posture, gait, and balance

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Treatment Plan ↔ Patient Goals

Impact Patient Function?

Impact Daily Care?

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Decrease Tone

Increase range of motion

Improve fit and use of orthoses

Decrease contractures

Delay surgery

Improve position for care

Improve function

Decrease caregiver burden

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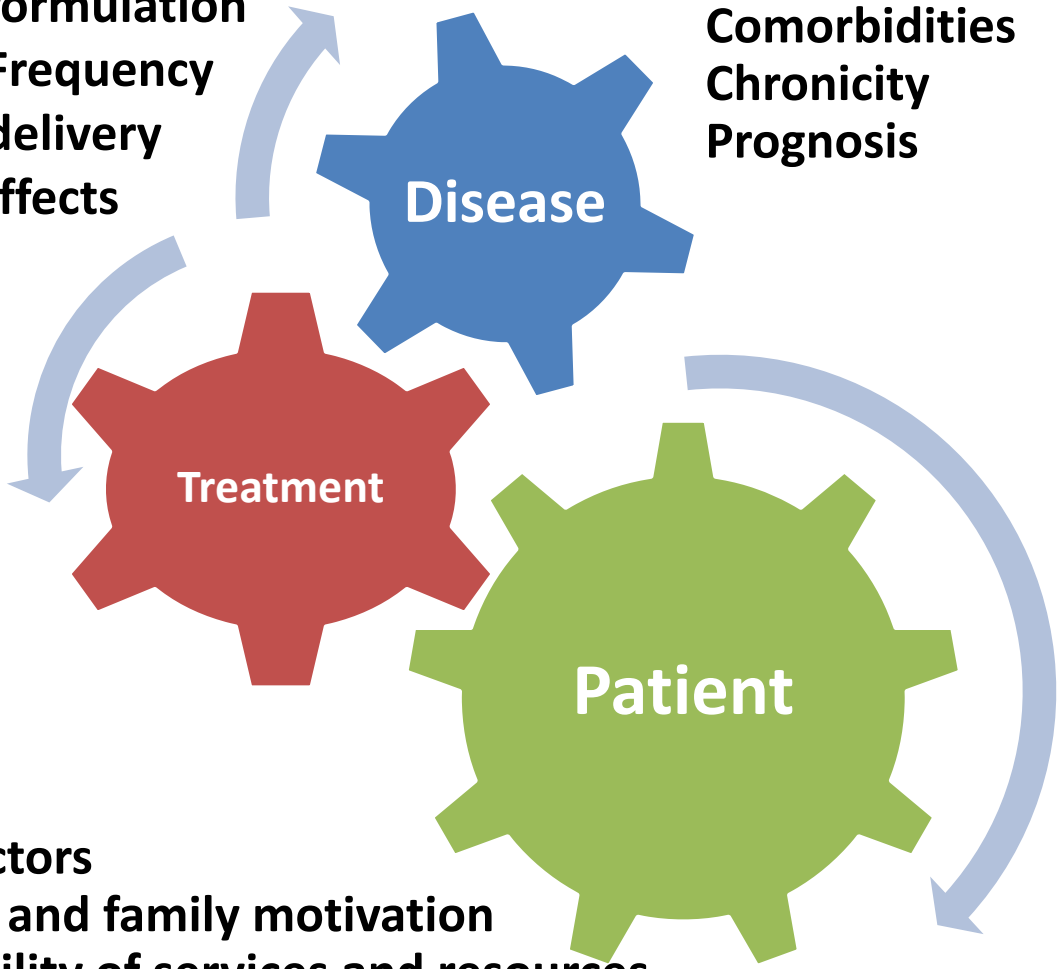
Treatment Factors
- Drug formulation
- Drug Frequency
- Drug delivery
- Side effects
- Cost

Disease Factors
- Generalized vs. Focal
- Comorbidities
- Chronicity
- Prognosis



Patient Factors
- Patient and family motivation
- Availability of services and resources
- Compliance

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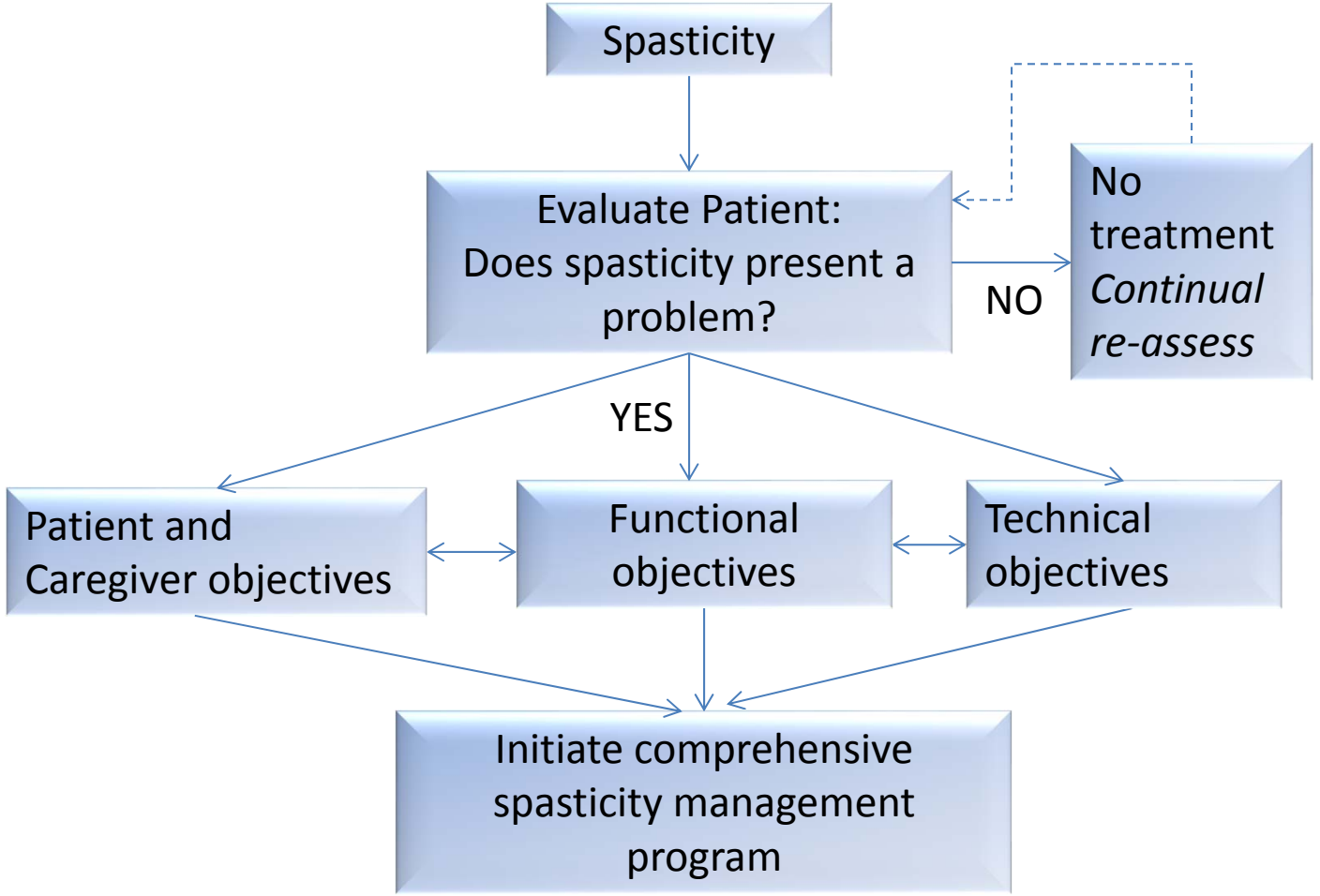

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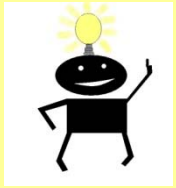
Spasticity Management Team

- Patient
- Caregiver
- Pediatrician
- Physiatrist (PMR)
- Neurologist
- Orthopedic surgeon
- Neurosurgeon
- Therapist : PT, OT, Speech
- Orthotist and Durable Medical Equipment provider
- Other: Social Work, Dietician, Education specialist



As children age, they may be more likely to need orthoses and surgery.

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What is the Role of the Physiatrist?

Function at home and in the community

Orthoses and therapeutic equipment

Altered muscle tone

Facilitating the ability of the child and family to set functional goals

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Development

Muscle Tone

Pain

Dysautonomia

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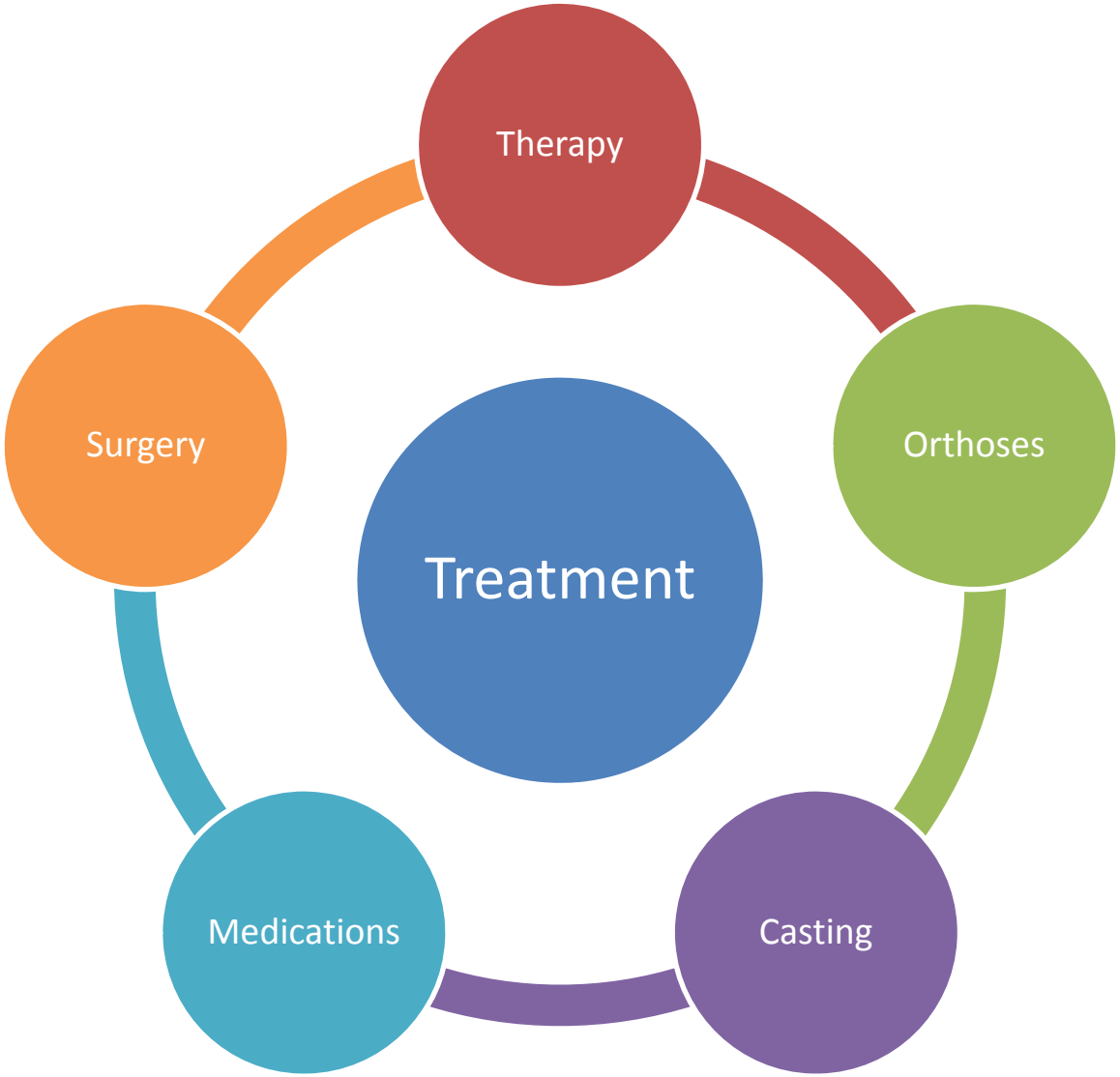

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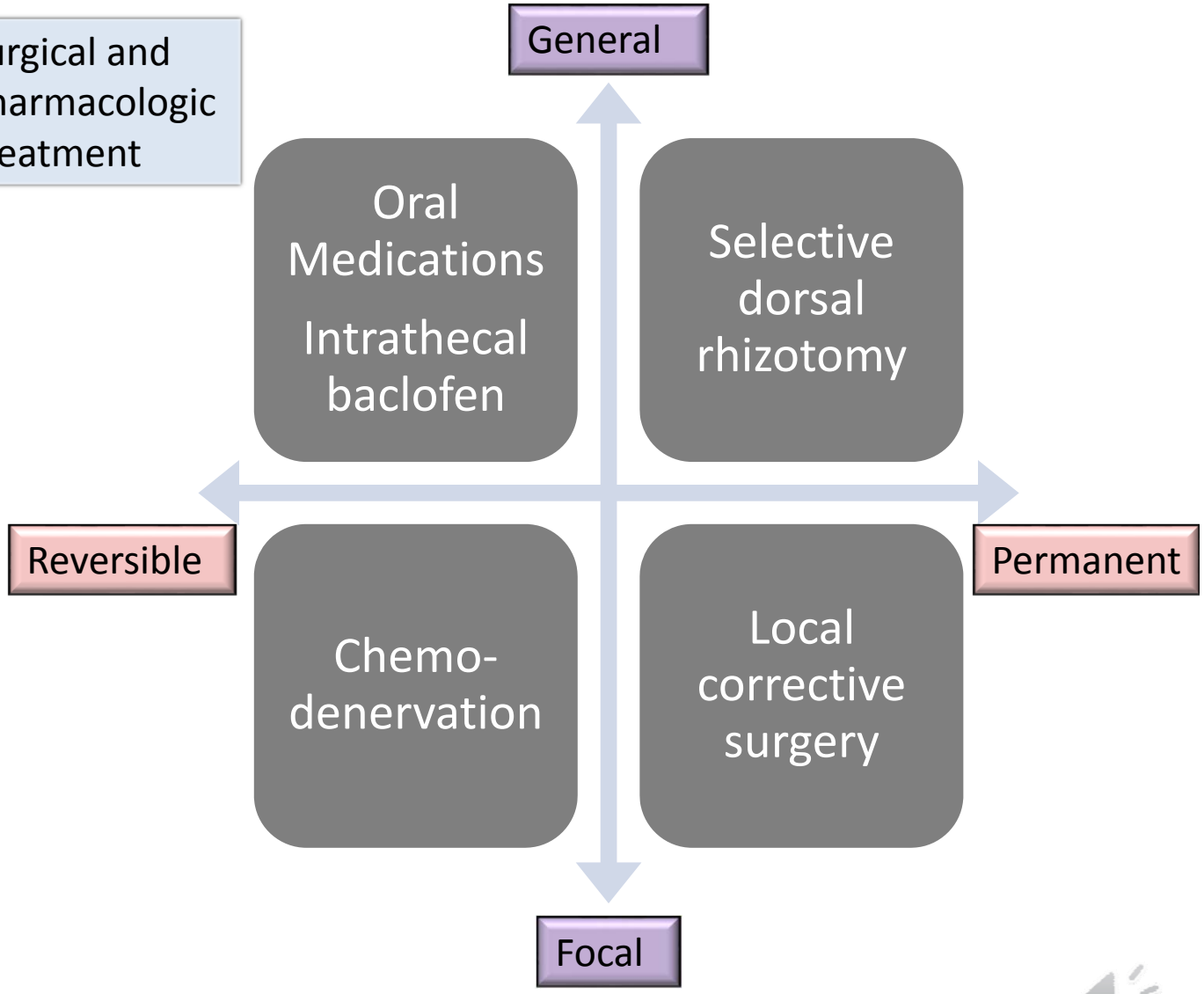
Surgical and Pharmacologic Treatment



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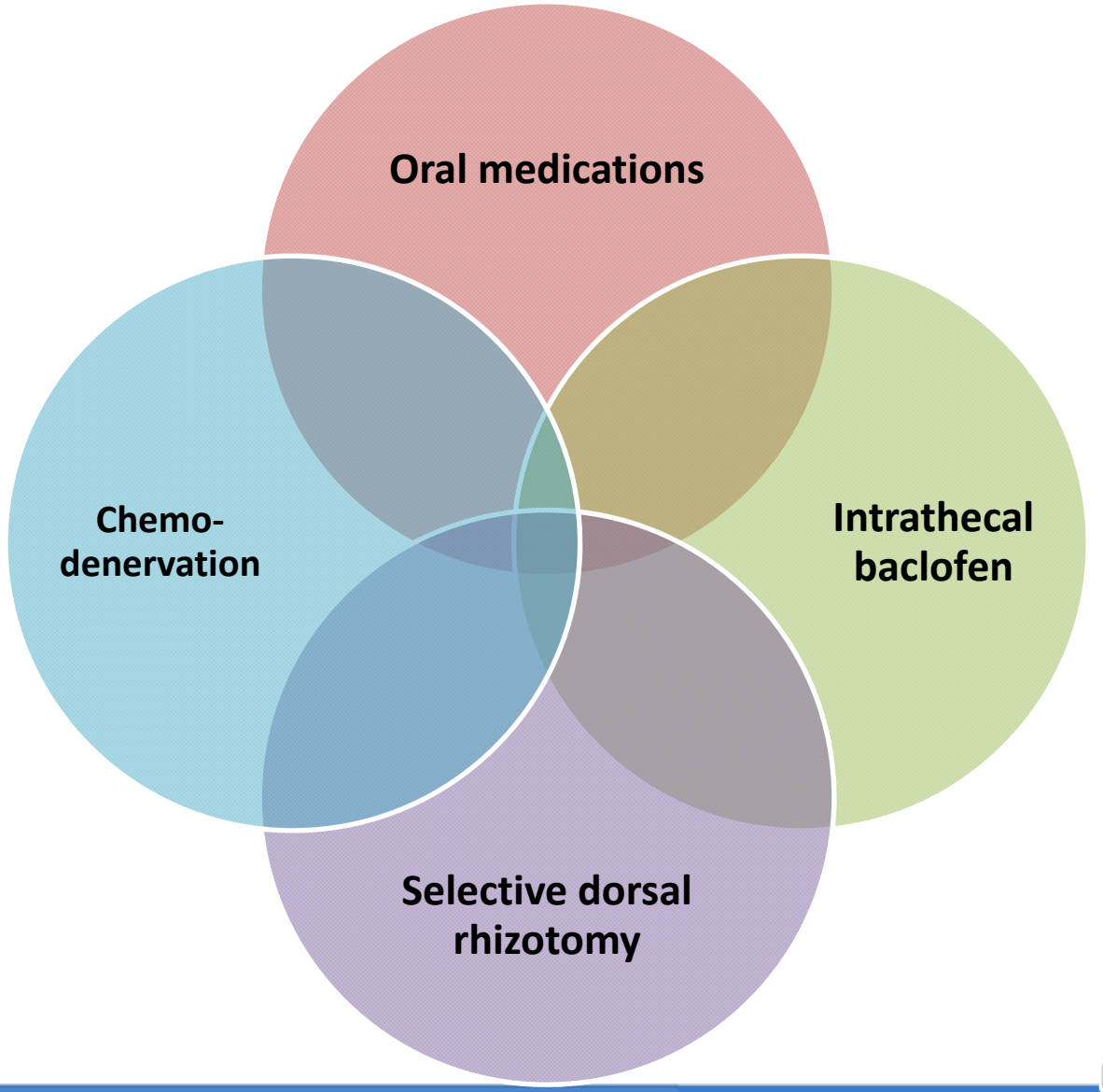

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Medication	Mechanism of Action	Most Common Side Effects
Baclofen	- Spinal cord - GABA B agonist	- Lowers seizure threshold
Benzodiazepines: Diazepam,clonazepam	- GABA A agonist - Spinal cord, brain	- Drooling
Tizanidine (Zanaflex)	- Alpha-2 agonist - Spinal cord, brain	- Hypotension - Hepatotoxicity
Dantrolene	- Calcium channel blocker - Muscle	- Hepatotoxicity
Trihexyphenidyl (Artane)	- Anticholinergic - Central muscarinic receptors	- Dry mouth

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Medication	Initial Dose	Maximum per day	Doses/ Day
Baclofen	2.5-5 mg x 2-3/day 0.6 mg/kg/d tid for <12 mo	40-90 mg (age dependant)	3-4
Diazepam	0.12-0.8mg/kg		3-4
Clonazepam	0.01-0.03 mg/kg/day	0.1-0.2 mg/kg/d	2-3
Tizanidine	2 mg/day	36 mg/day	2-3
Dantrolene	25 mg/day (0.5mg/kg daily)	12 mg/kg/day or 400 mg/day	2-4
Artane	1 mg/day	15 mg/day	2

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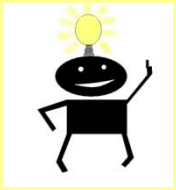
Sedation is one of the common side effects with all oral medications

To minimize potential sedation, always start low and titrate up slow



Abrupt withdrawal can result in seizures

If NPO, can use IV Diazepam

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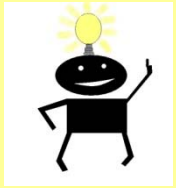
First Line: Oral Baclofen

AVOID MEDICATION ERROR

✓Concentration is variable

✓1 ml ≠ 1 mg



When prescribing oral baclofen, the dose MUST be verified to avoid fatal medication errors!

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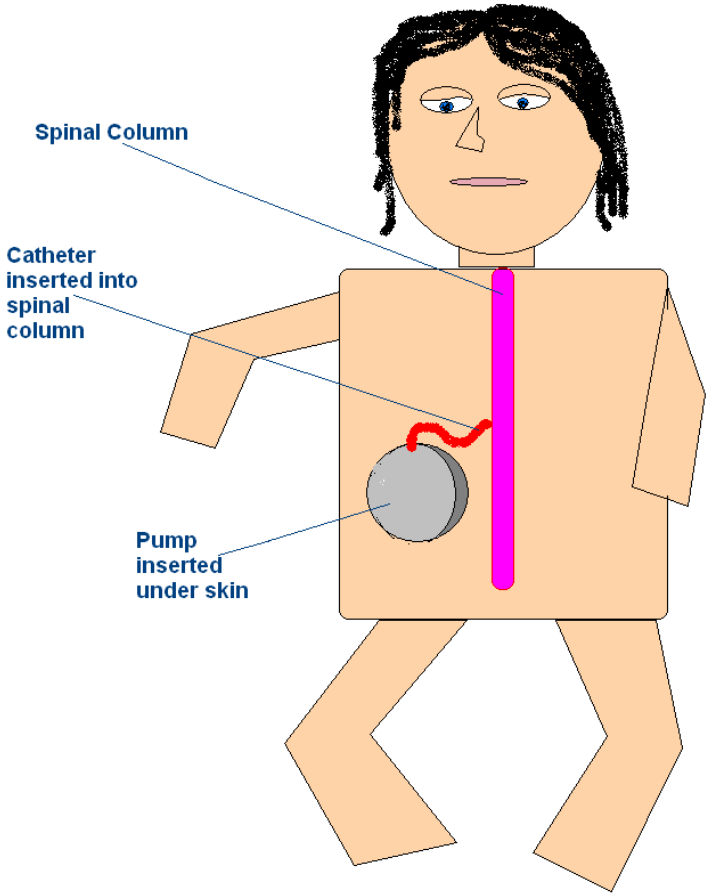
Intrathecal baclofen:

50x response of oral baclofen



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Who is a candidate?

- Severe, generalized tone
- Not successfully managed with oral medications
- Improvement with test dose of intrathecal baclofen given via lumbar puncture

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PROS:

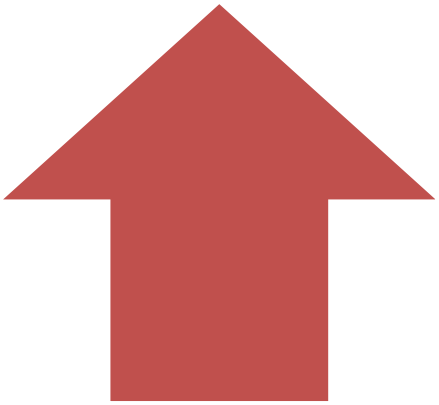
GABA inhibition without side effects in the brain

Programmable to set best dose

CONS:

Complications of device

High maintenance of pump



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Mechanical

- CSF leak
- Catheter malfunction
- Infection

Medical

- Overdose
- Withdrawal

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Baclofen withdrawal

Early signs:
- Pruritis
- Dysphoria
- Irritability
- Spasticity
- Tachycardia
- Fever
- Hypertension
- Respiratory Distress

Late signs:
- Hyperthermia
- Seizures
- Rhabdomyolysis
- DIC
- Altered Mental Status
- Psychomotor agitation
- Respiratory Distress
- Multisystem Organ Failure

Death

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Troubleshooting Pump Failure

1. Medications
2. Radiographs
3. Spiral CT

Call Neurosurgeon!

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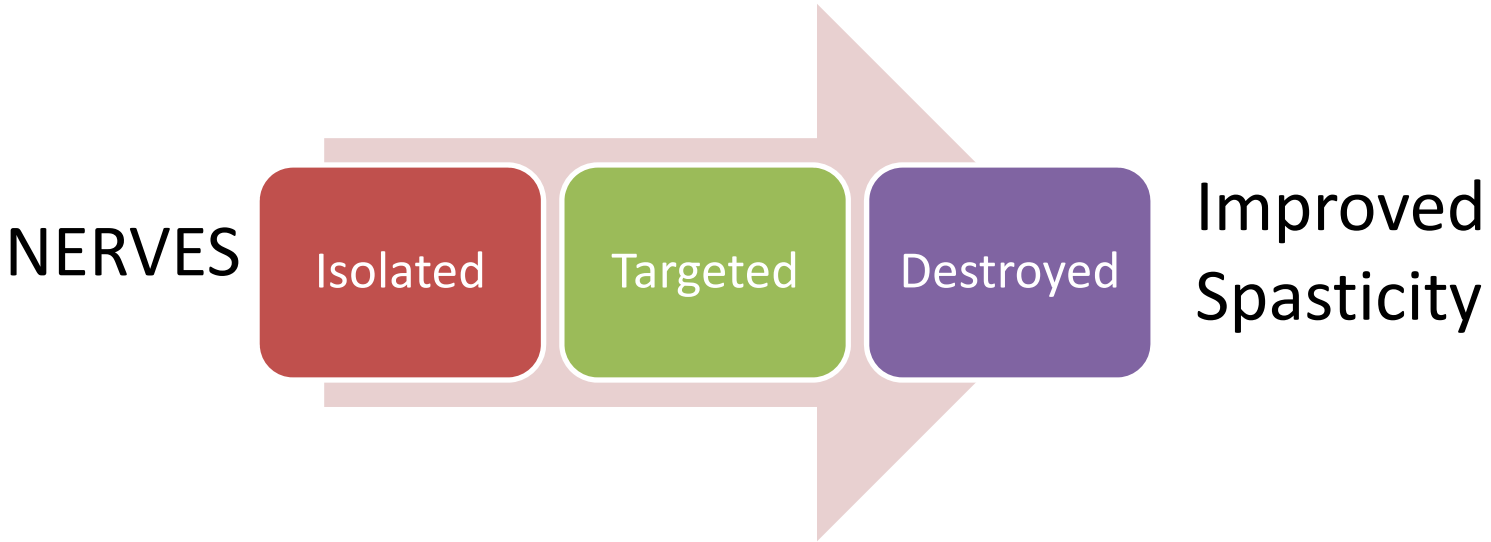
What is Selective Rhizotomy?



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Who is a Good Candidate?

- Ages between 4 and 7 years
- "Pure" spasticity
- Ambulatory
- Absence of fixed contractures
- Cooperation with intensive therapy

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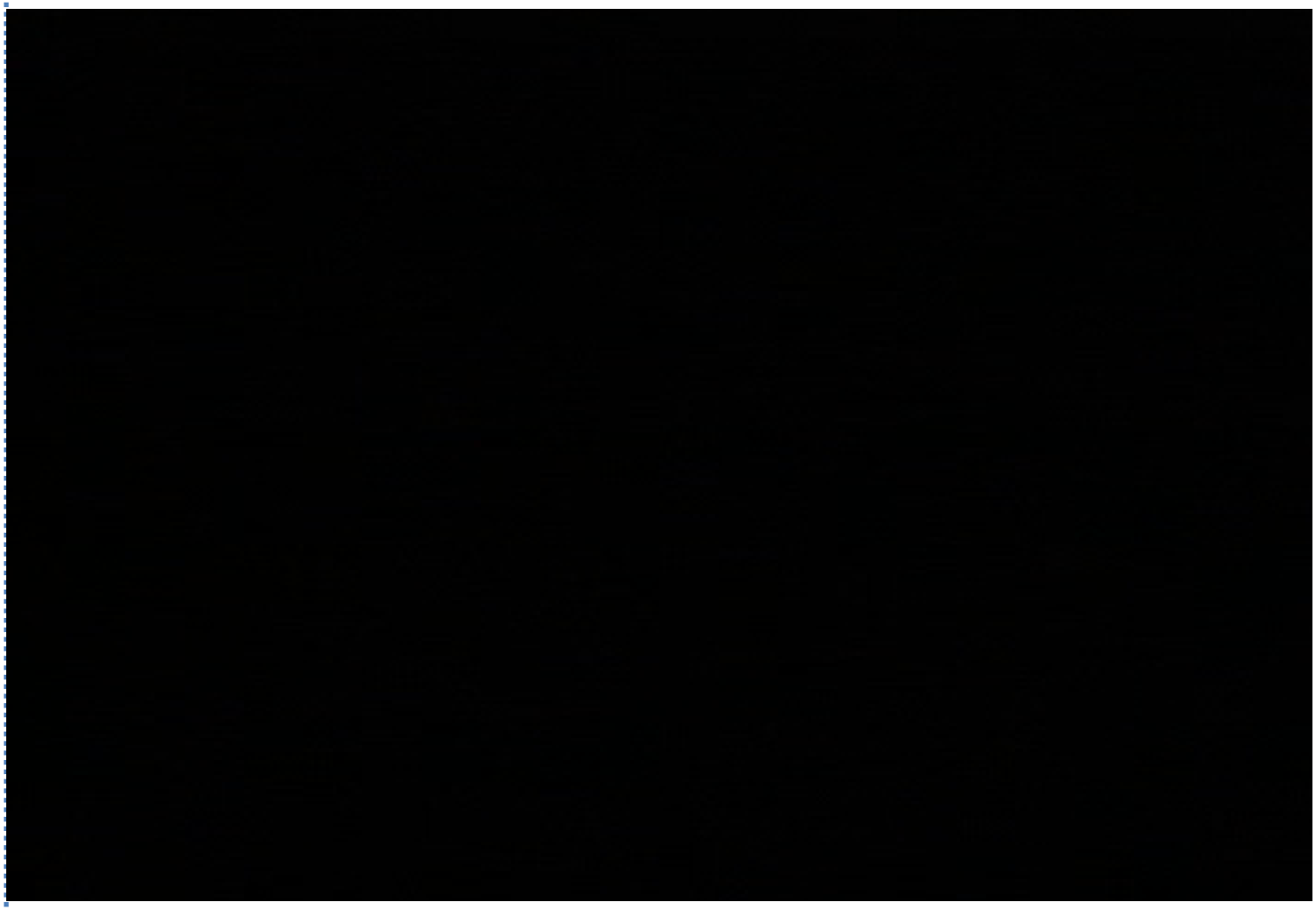

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	Botulinum Toxin Type A	Phenol
Mechanism of Action	Selective motor denervation at the NM junction (presynaptic block of ACh release)	Non-selective chemical neurolysis in injected nerve
Onset	24-72 hours	< 1 hour
Duration	6-12 weeks (3-6 months functional outcome)	2-36 months
Side Effects	Weakness	Dysesthesia, skin necrosis
Pros	Easy to inject Limited dose of toxin	Low cost
Cons	Cost	Difficult to inject

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Advantages

- Temporary*
- Minimally invasive
- Increase ROM
- Learn normal movement patterns
- Safely repeated
- Mimic surgical outcome
- Allows age-selective timing of surgery

Disadvantages

- Temporary*
- May decrease functional tone
- Total body dose of toxin limited (BTX-A)
- Cost (BTX-A)

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Who is a Good Candidate?

Focal increase in muscle tone

Absence of fixed contractures

Absence of bony/joint problems

No underlying bleeding disorder

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Timing of Botulinum Treatment

Early Years

- May allow postponement, simplification, avoidance of surgery

Later Years

- May provide pain relief, improved ease of care, functional goals

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Spasticity = 1 component of the UMN syndrome

Spasticity = velocity dependent increased resistance to passive stretch

Underlying lesion is static, but musculoskeletal pathology is progressive

Treatment plan is not “one-size fits all”, but should be customized to patient’s goals

Most spasticity medications CANNOT be stopped abruptly. Should be weaned to avoid seizures, or switched to IV if patient is NPO.

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