# CEREBRAL PALSY & MMC

Sanjay Arora

Consultant Pediatric Orthopedics
ORTHOMED HOSPITAL
Hisar

- Definition
- Classification
- Etiology
- Pathogenesis
- Evaluation and surgical management
- Medical management of spasticity

# Definition

- Permanent disorders of the development of movement and posture
- Non-progressive disturbances of the developing fetal and infant brain
- results in motor impairment

# Etiology/Pathology

- Most likely due to brain damage-

  *possible cause being anything that interferes with oxygen to the brain.*

- Damage usually occurs during fetal development, before, during, or after birth, or during infancy.

# Etiology/Pathology

- Exact cause is unknown

- Most likely due to brain damage-

  *possible cause being anything that interferes with oxygen to the brain.*

- Damage usually occurs during fetal development, before, during, or after birth, or during infancy.

# Cerebral Palsy: Etiologic

- Prenatal (70%)
  Infection, anoxia, toxic, vascular, Rh disease, genetic, congenital malformation of brain

- Natal (5-10%)
  Anoxia, traumatic delivery, metabolic

- Post natal
  Trauma, infection, toxic

# Risk Factors: Prematurity and LBW

- Approximately 50% of children with cerebral palsy have low birth weight, and 28% weigh less than 1500 g at birth.

- The prevalence of birth weight–specific cerebral palsy ranges from 1.1 per 1000 neonatal survivors weighing 2500 g or more to 78.1 per 1000 in infants weighing less than 1000 g.[417]

# Cerebral Palsy: Complications

- Spasticity
- Weakness
- Increase reflexes
- Clonus
- Seizures
- Articulation & Swallowing difficulty
- Visual compromise
- Deformation
- Hip dislocation
- Kyphoscoliosis
- Constipation
- Urinary tract infection

# Classification

- Physiological

  Spasticity:

  Hypotonia:

  Dystonia: lead pipe

  Athetosis

  Ataxic cerebral palsy

  Mixed

**Pyramidal**

- motor cortex, IC or CS tracts

**Extrapyramidal**

basal ganglia, Thalamus, Subthalamic nucleus and/or cerebellum

# Classification

- Geographical

  Hemiplegia

  Diplegia

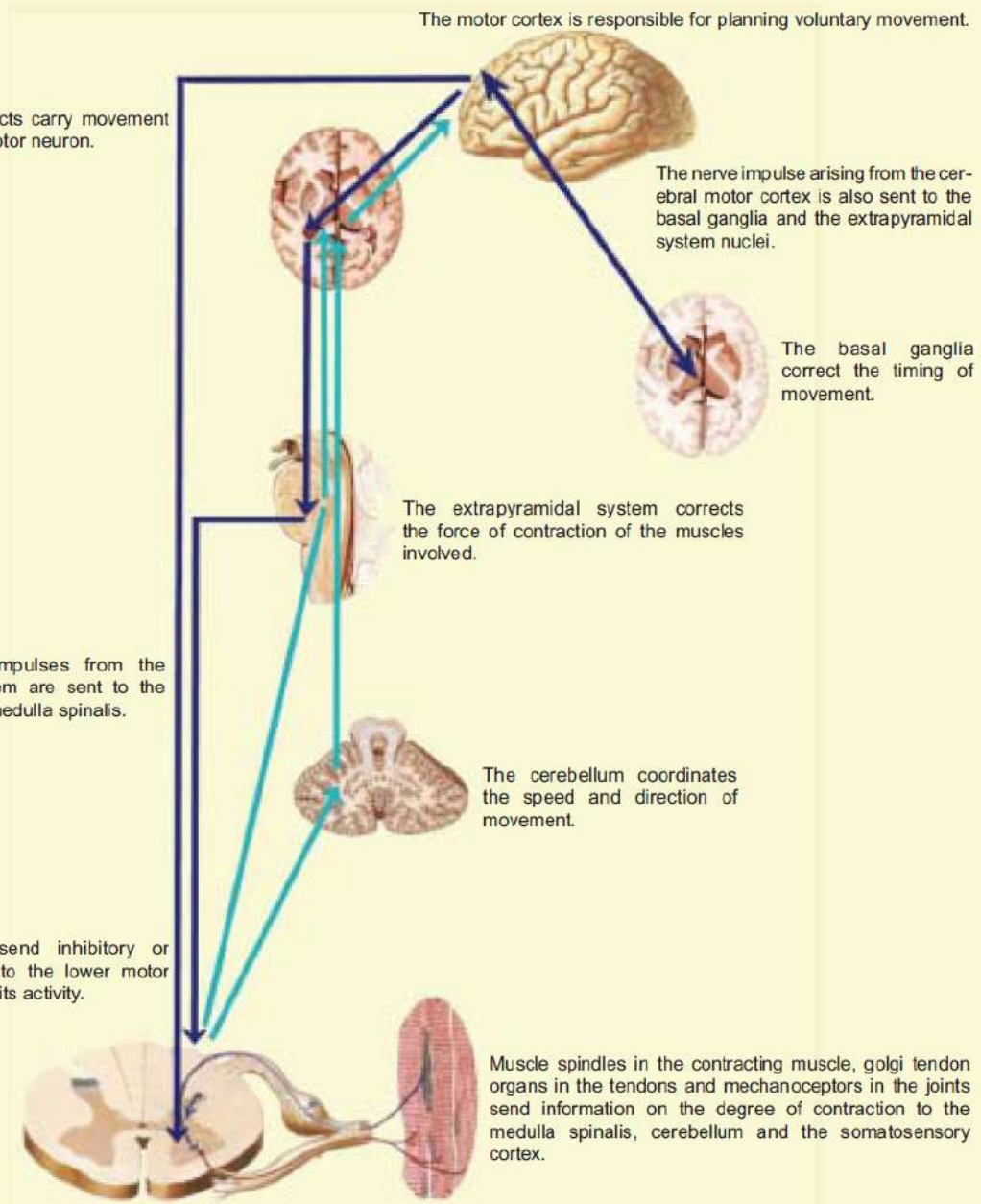
  Triplegia

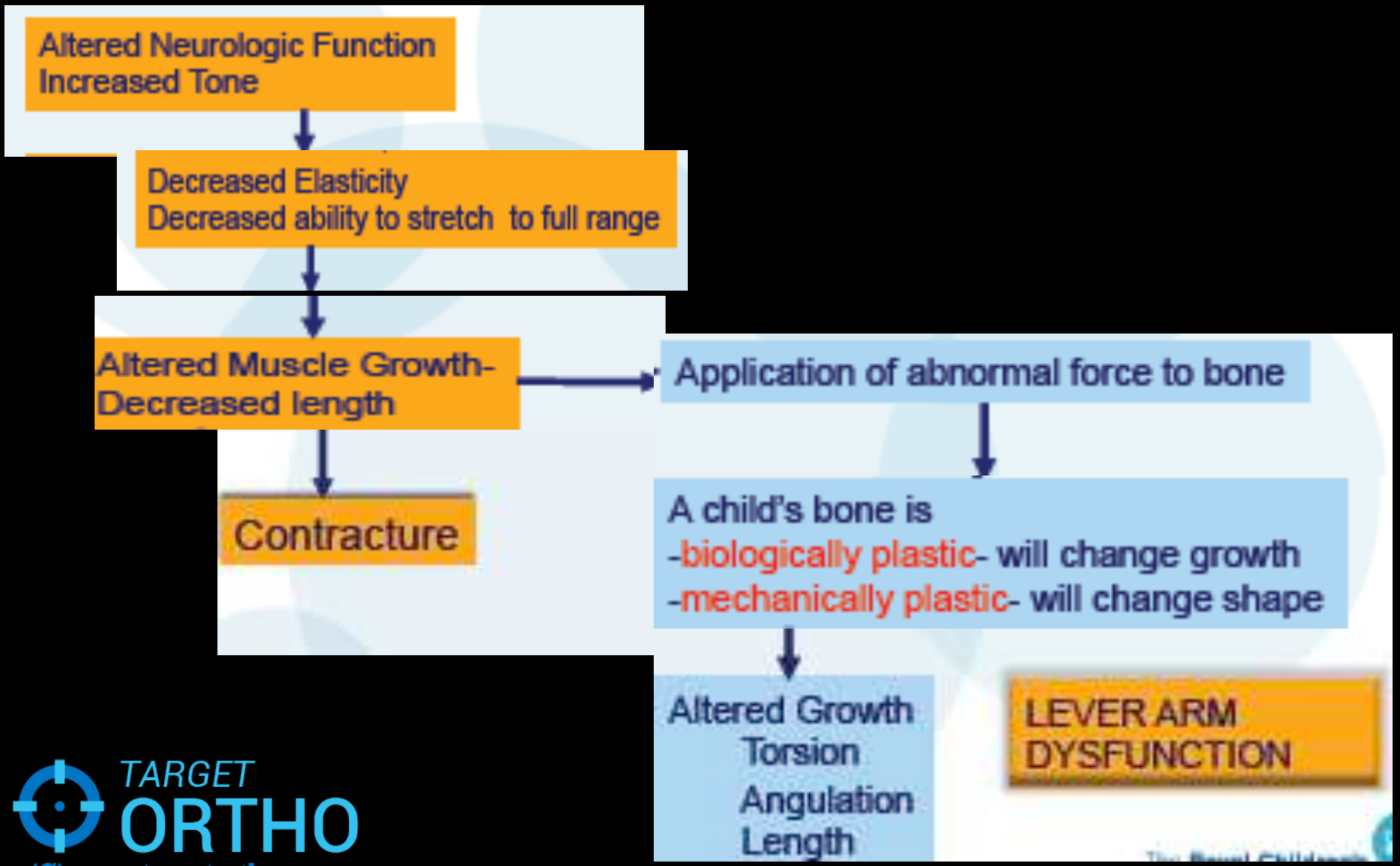
  Quadriplegia

CE

A

## Neural pathways regulating muscle contraction





# Differential diagnosis

- Spinal tumor
- Familial spastic papaparesis
- Spinocerebellar ataxia
- Congenital hypothyroidism
- Dopa responsive dystonia

# Evaluation

- Family history
- Developemental history
- Handedness
- Related medical condition

# Physical examination

- Muscle tone
- Reflexex
  - DTR
  - Infantile reflexes
- Balance, sitting and gait

# Physical examination

- Passive and active range of motion of all joints.
- Evaluation of fixed muscle contractures vs dynamic muscle contractures.
- recognition of joint contractures and/or joint subluxations and dislocations
- Spasticity grading

# Examination

- The tone is velocity dependent, which means that if a muscle is stretched rapidly, tone increases more than if the same muscle group were stretched gradually and gently.

- Dystonia is described as increased tone, which is not velocity dependent.

- tone in spasticity "clasped knife,"
- tone in dystonic cerebralpalsy "lead pipe,"

# Examination

- Athetosis is characterized by abnormal writhing movements that the patient cannot control

- Cerebellar lesions lead to ataxic cerebral palsy. The disturbed balance of these children results in a wide-based and clumsy gait. Pure ataxic cerebral palsy is rare

# Ashwoth scale

1. No increase in muscle tone.
2. Slight increase in tone giving a "catch" when affected part is moved in flexion or extension.
3. More marked increase in tone but affected part is easily flexed.
4. Considerable increase in tone; passive movement difficult.
5. Affected part is rigid in flexion or extension.

# Physical examination: Sensory

- Two-point discrimination (>15–20 mm) is the ideal to test.
- In younger children

  tactile sensitivity,

  stereognosis,

  and proprioception are easier and more accurate to test.
- The sensation is a good measure of the overall functional ability of the extremity.

# Sensory Impairment

- Evaluation of sensory capacity is difficult,
- but sensory deficits are recognized to contribute more to the overall impairment in function.
- When SSEPs were included, impairment in at least one modality of sensory function was found in 88% of children with cerebral palsy.

# Sensory Impairment

- Van Heest et al (1993)showed that 97% of the spastic limbs had a stereognosis deficit, 90% had a two-point discrimination deficit, and 46% had a proprioception deficit.

- Thus sensory deficits are the rule rather than the exception in children with spastic hemiplegia.

# Effect on growth

- Van Heest et al (1993) found a correlation between the severity of sensory impairment and the degree of growth impairment in the affected limb.

- Those children with severe stereognosis deficits had significantly smaller limbs than the children with mild or moderate stereognosis deficits.

# Prognosis for ambulation

- ability to control the head by 9 months and to sit by 24 months predicts the eventual ability to walk

- Many agree that the ability to walk plateaus by 7 years of age, thus implying that if a child is non-ambulatory at 7 years, the child will probably never become ambulatory

# PROGNOSIS for AMBULATION

## GOOD Prognosis

- Hemiplegics/ Ataxic pxs
- Achievement of all motor skills by age of 8.
- Independent sitting before 2 years
- Persistence of fewer than 3 of the primitive reflexes at age 18 months.

## POOR Prognosis

- Quadriplegics
- Did not attain independent sitting by age 4.
- Persistence of primitive reflexes beyond 18 months

# Upper limb examination

- Shoulder
- Elbow
- Forearm
- Wrist
- Hand

# Spine examination

- Kyphosis
- Ex-aggregated lardosis
- scoliosis

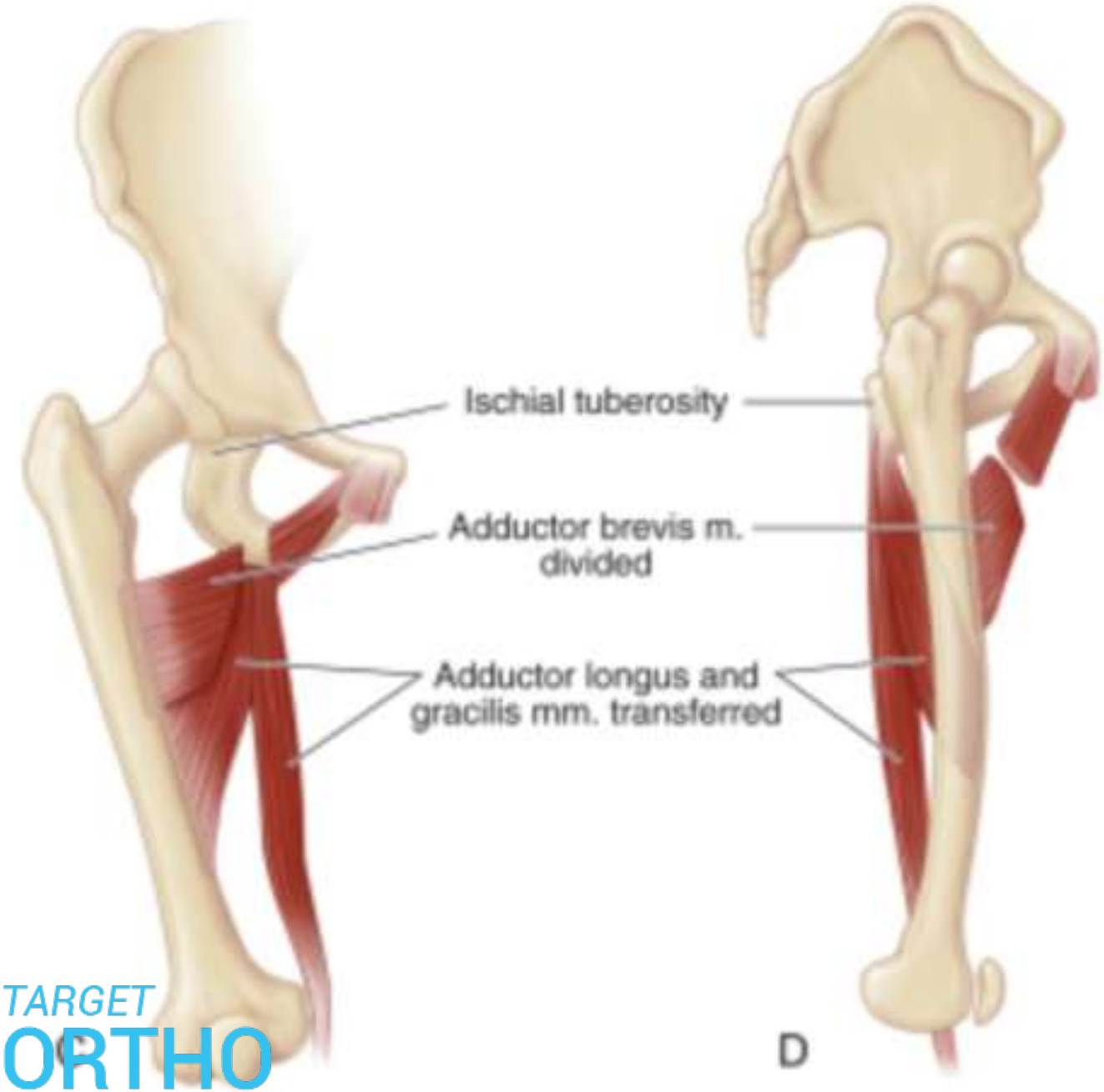
# Lower limb examination

- Adduction contracture
- **Pseudo-adduction**

- Phelp's test

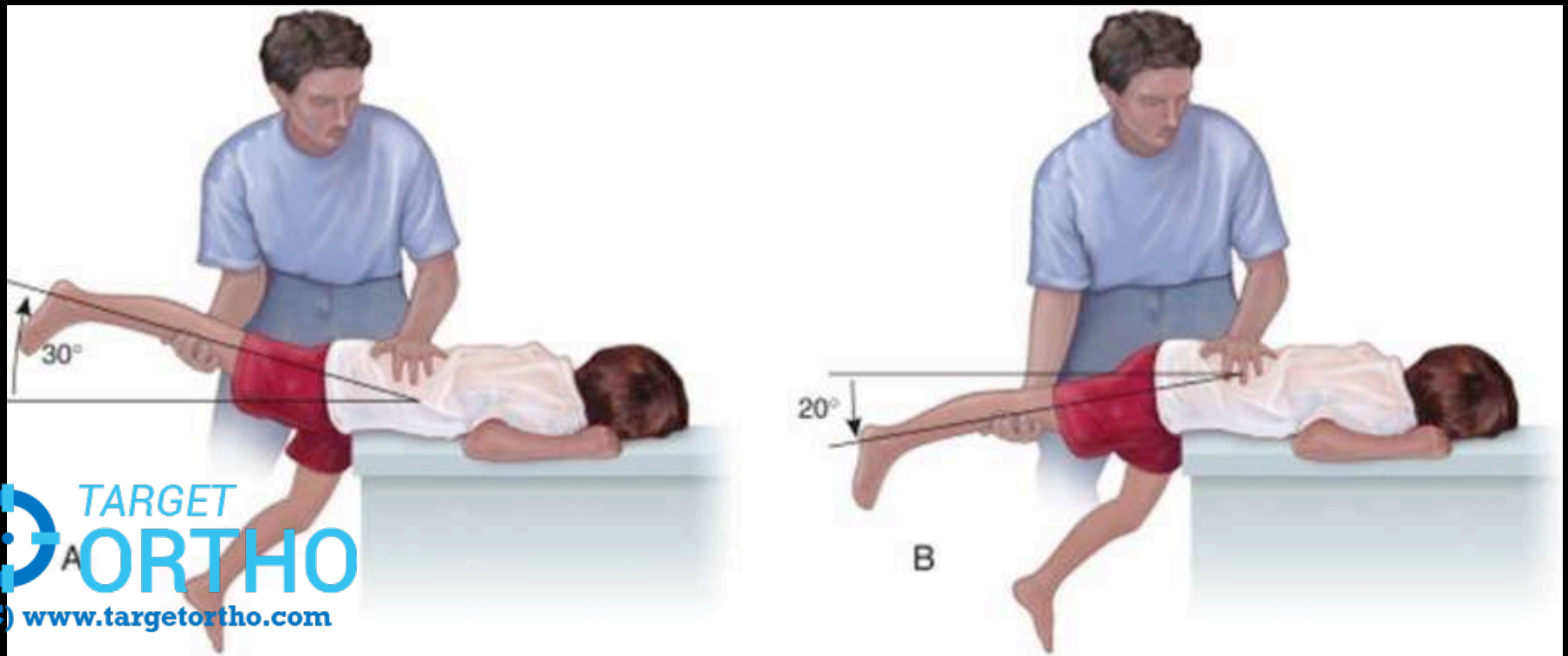
# Adduction contracture

- Adductor release
- Perry transfer
- Obturator neurectomy



# Lower limb examination

- Hip flexion contracture
  - Thomas test
  - Staheli test

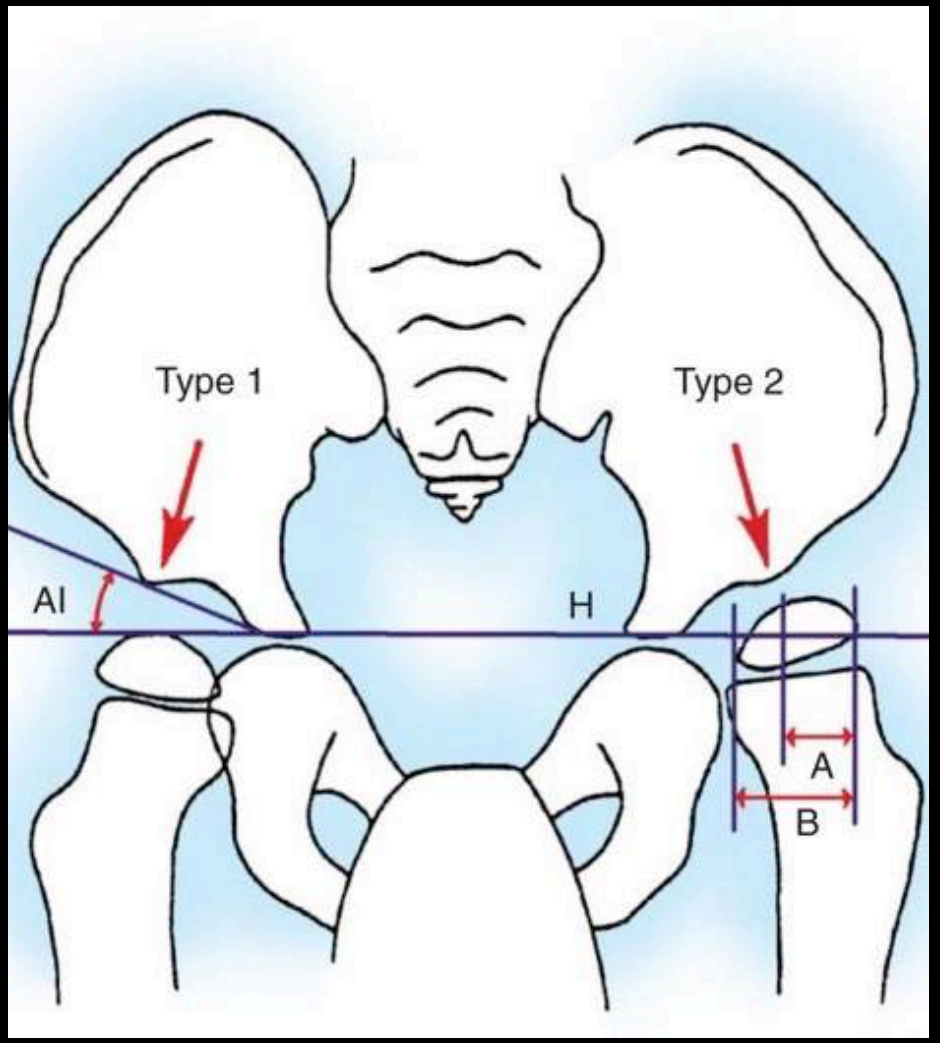


# Hip flexion contracture

- Psoas release over the brim
- Iliopsoas release at lesser trochanter

# Hip subluxation or dislocation

- In response to muscle imbalance
- Add & Flex >> Abd & ext
- Remodelling of anteversion angle
- Neck shaft angle
- Loss of range of motion
- Reimer's migration index
- Shanton's line
- Acetabular index

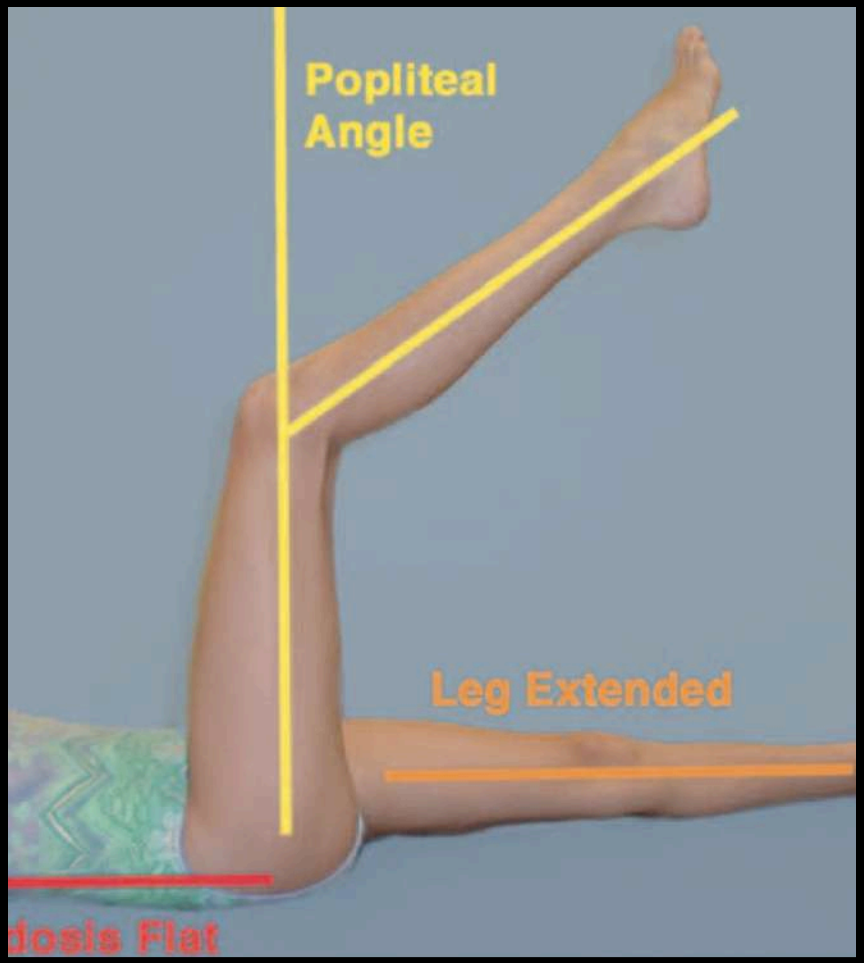


# Hip subluxation or dislocattion

- Soft tissue release
- Reduction and reconstruction
- Salvage surgery
- Hip arthrodesis
- Total hip arthroplasty

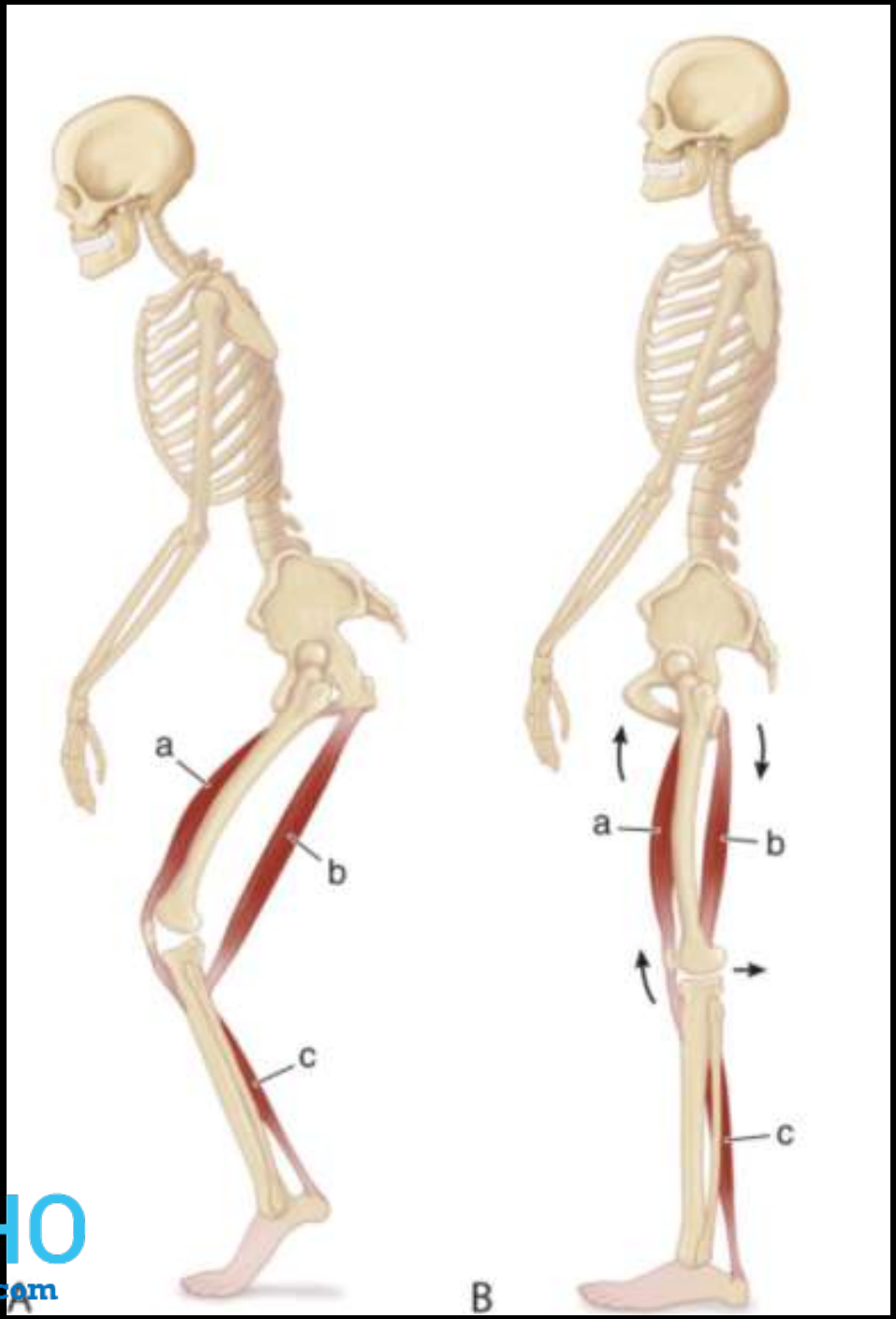
# Knee

- Flexion deformity
- Hamstring spasticity
- Knee contracture
- Quadriceps power
- Prone rectus test
- Patella alta

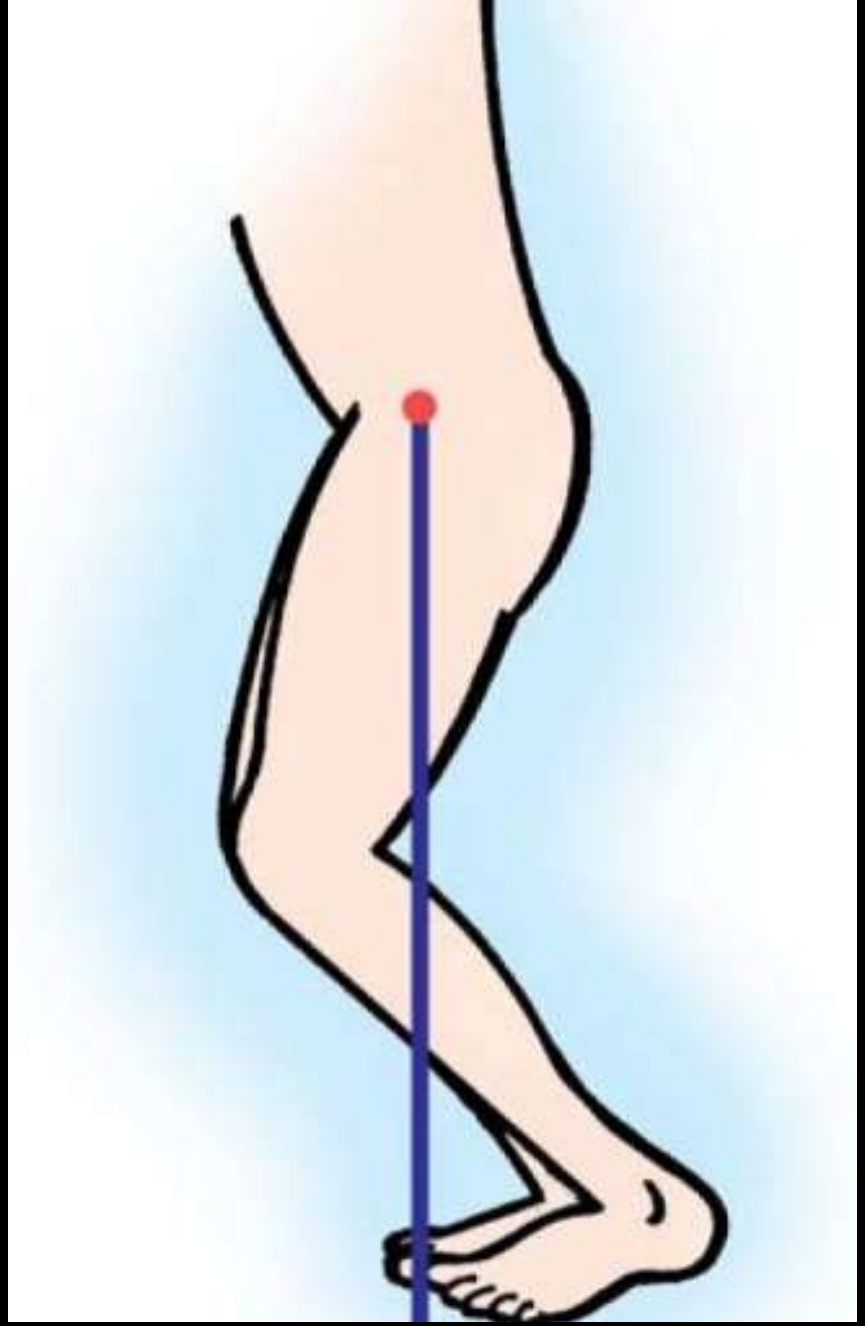


# Knee

- Hamstring lengthening
- Posterior capsule release
- Patellar tendon reefing
- Distal transfer of rectus femoris
- Femur shortening
- Supracondylar extension osteotomy
- ST transfer ao adductor tubercle
- Egger's transfer

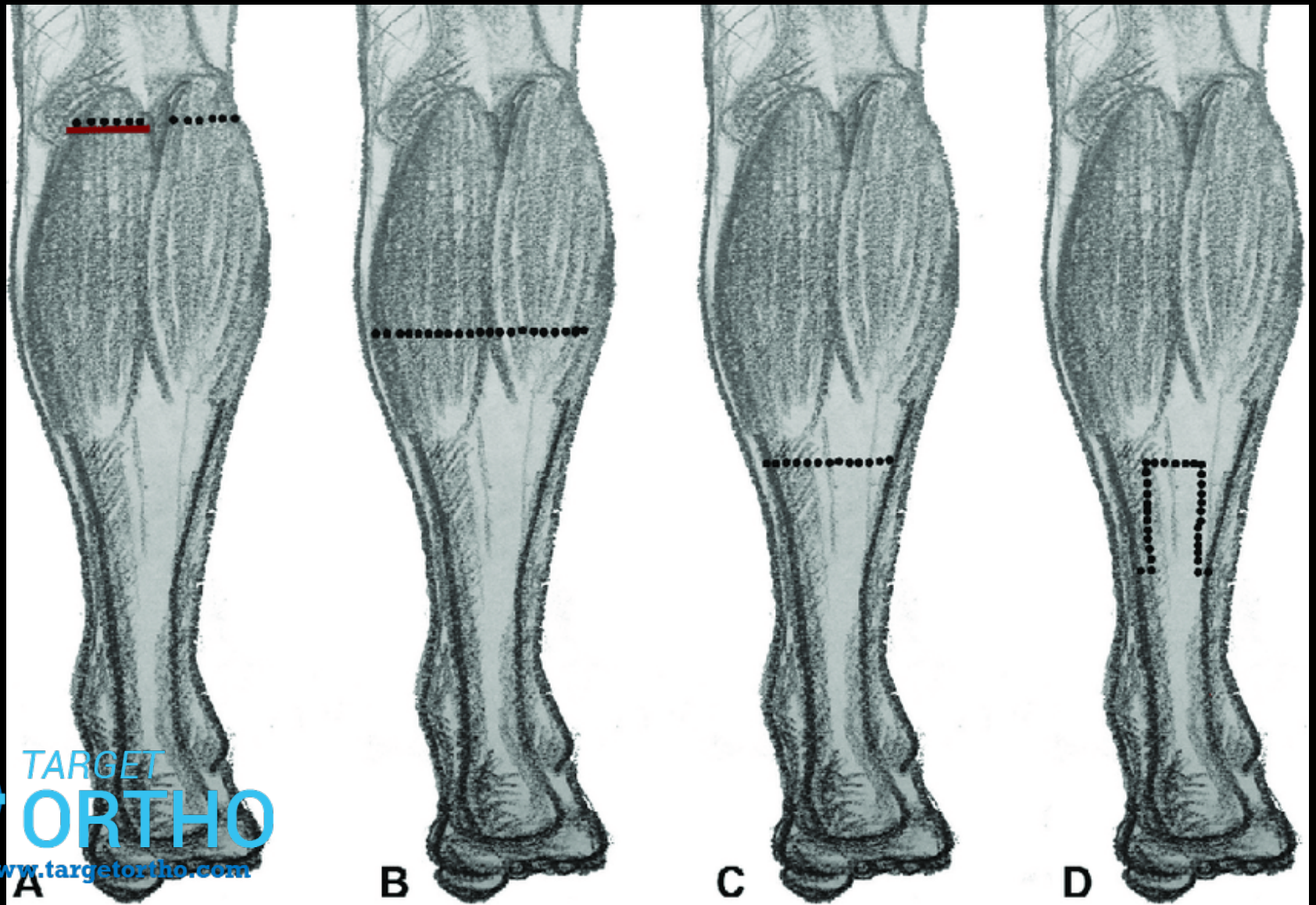


- Hip and knee relations



# Foot

- Equinus



# foot

- Equinovarus
- Lengthening  or recessionTib post
- Split tendon transfer
- Osteotomy of calcaneus

# Foot

- Equinovalgus

- Calcaneus osteotomy
- Subtalar arthrodesis
- Triple arthrodesis

# GROSS MOTOR FUNCTIONAL CLASSIFICATION

| | |
|---|---|
| LEVEL 1 | Walks without restriction, Limitations in high-level skills |
| LEVEL 2 | Walks without devices, Limitations in walking outdoors |
| LEVEL 3 | Walks with devices, Limitations walking outdoors |
| LEVEL 4 | Limited mobility, Poor mobility outdoors |
| LEVEL 5 | Very limited self-mobility, even with assistive technology |

| | |
|---|---|
| Primary deficit due to brain lesion | • Muscle tone- spasticity, dystonia<br>• Balance<br>• Strength<br>• Selectivity<br>• Sensation |
| Secondary impairment | Contractures (equinus, adduction)<br>Deformities (Scoliosis |
| Tertiary impairment | • Adaptive or compensatory mechanisms<br>• Knee hyperextension in stance |

Motor Type

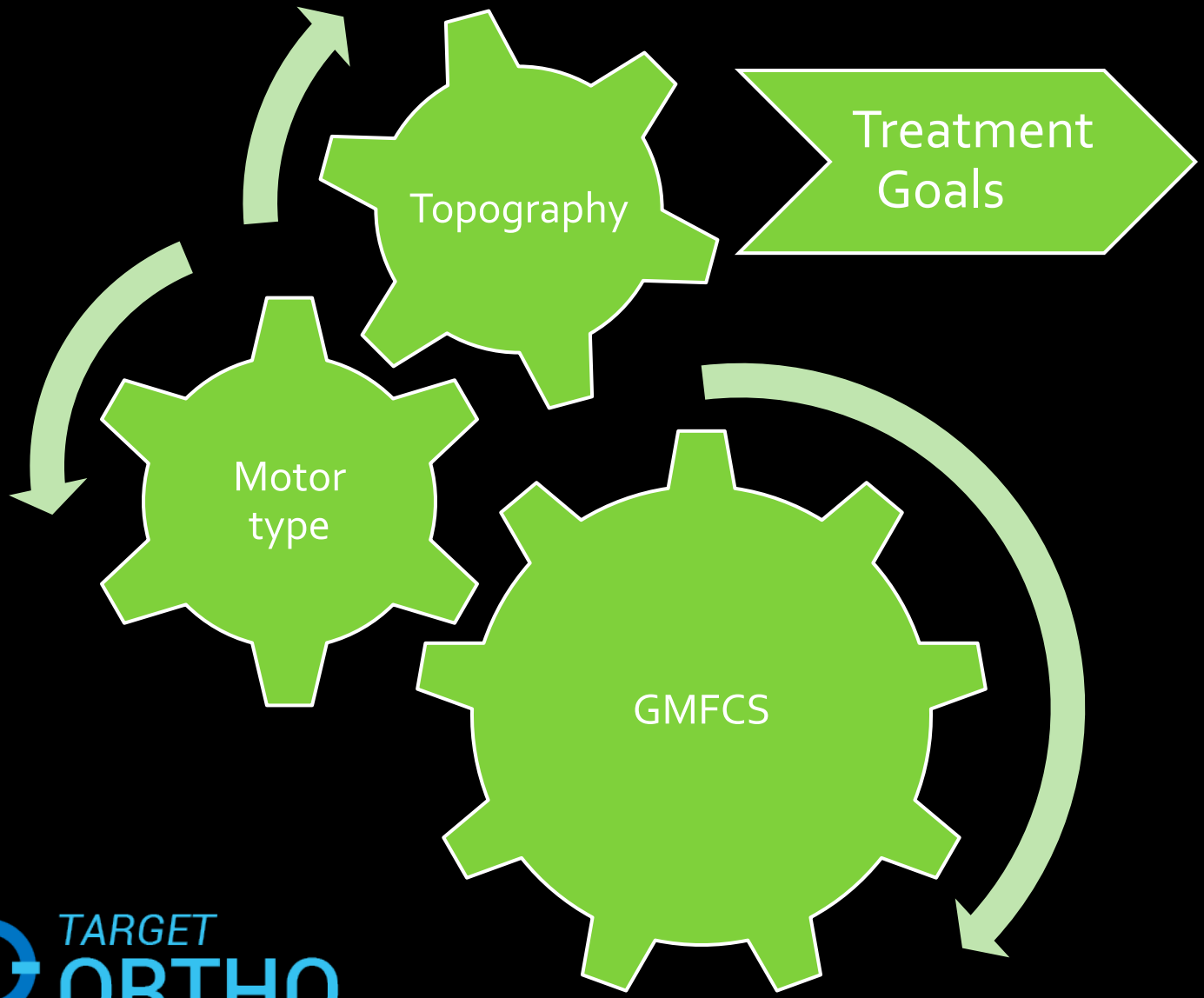
- Spastic
- Dyskinetic
- Mixed

Body part involved

- Diplegia
- Mono, Hemiplegia
- Total Body

Function

- GMFCS
- FMS



# Gait Correction Surgery: Aims

- GMFCS level I-III
  - Correct deformity
  - Improve the gait pattern
  - Maintain (? Improve) function

# Aim of orthopaedic surgery

- GMFCS level IV
  - Flexible, enlocated hips, good sitting
  - Pain-free braceable feet for transfers and minimal ambulation

# Aim of orthopaedic surgery

- GMFCS level V
  - Enlocated pain-free hips
  - Spinal alignment
  - Optimal sitting posture

# CONCEPTS

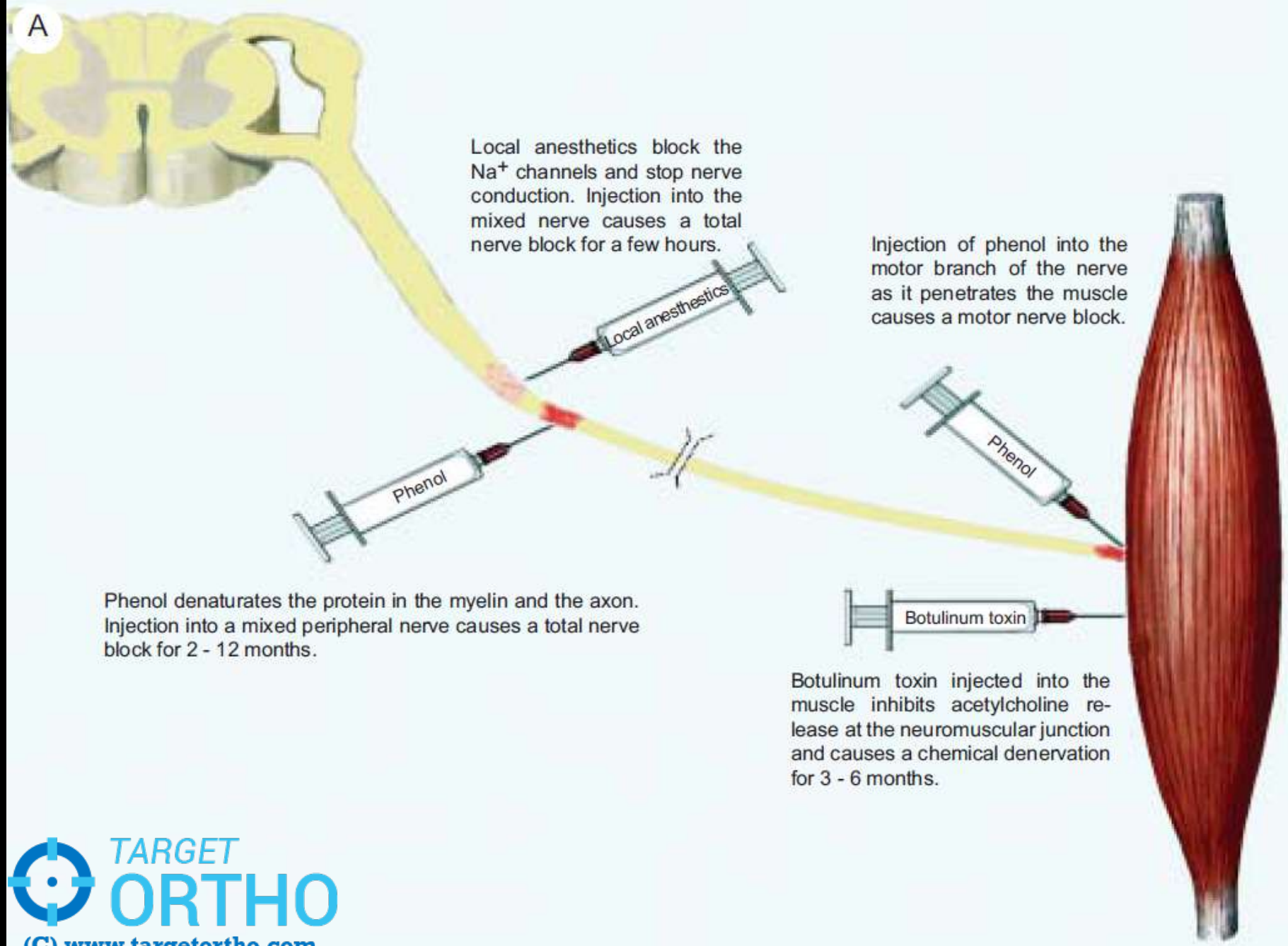
- Monoarticular muscles-stabilizers of joints iliacus,adductors,gluteals.
- Polyarticular muscles –movers of joints affect gait and movement psoas, gastrocnemius, hamstrings,
- Musculotendinous junction –growth plate of muscles --needs stretch for growth

# Management of Hypertonia

- Oral Medications
  a. Baclofen
  b. Dantrolene
  c. Diazepam
- Specific Medications for Dystonia:
  a. Trihexyphenidyl HCl
  b. Levodopa-carbidopa (Sinemet)

- Botox Injections
- Phenol Injection

❖ Reappraisal of spasticity or hypertonia in a growing child with CP every 6 months is necessary.



# Botulinium Toxin Type A (BOTOX)

- Effective in improving ROM and reducing tone.
- Also effective in improving motor control.
- Best results: Botox + Serial casting

- Indications for Use of Botox:
  a. Calf injection for dynamic equinus persistent throughout the gait cycle
  b. Hamstring injection for dynamic knee flexion
  c. Adductor injection for scissoring
  d. Diagnostic measures before surgery
  e. Management of focal limb dystonia
  f. Analgesia for pain and spasm in the perioperative period
  g. In the UE, persistent thumb in palm, wrist posture preventing hand use, and elbow flexion.

# HOW DO WE MANAGE SHORTENING??

- Stretching
- Splints
- Serial casts
- Surgery

# NEUROSURGICAL INTERVENTIONS

I. Selective Dorsal Rhizotomy (SDR)
II. Intrathecal Baclofen (ITB) Pump
III. Stereotactic ablation of selected thalamic nuclei
IV. Chronic ES of the Cerebellum or Posterior Columns
    - Has shown promise in adults with dystonia.

# Selective Dorsal Rhizotomy

- Since 1980's
- Reduces spasticity by interrupting the sensory input in the dorsal horn.
- Ideal Candidates for SDR:
  1. Premature child with spastic diplegia.
  2. Good balance
  3. Good selective motor skills
  4. Aged 4 or 5 years
  5. With minimal contactures
  6. Able to walk unassisted
- Athetosis – contraindication for SDR
- Dystonia – can become more problematic post SDR

- Two groups of children who benefit from selective dorsal rhizotomy:

1. Spastic Diplegics (Borderline ambulators)
   - The goals of surgery : better gait and leg function
2. Severe Spastic Quadriparetics
   - increase their independence by allowing them to sit for longer periods of time, use a potty seat, or power a wheelchair on their own.

# Intrathecal Baclofen

- In addition to spasticity reduction, it also has impact on dystonia.

- Complications with ITB are as high as 50%.
  - Improvements in technology with catheters and pumps might reduce the mechanical risk.
  - Experience in management.
  - Abrupt withdrawal in children must be treated aggressively.

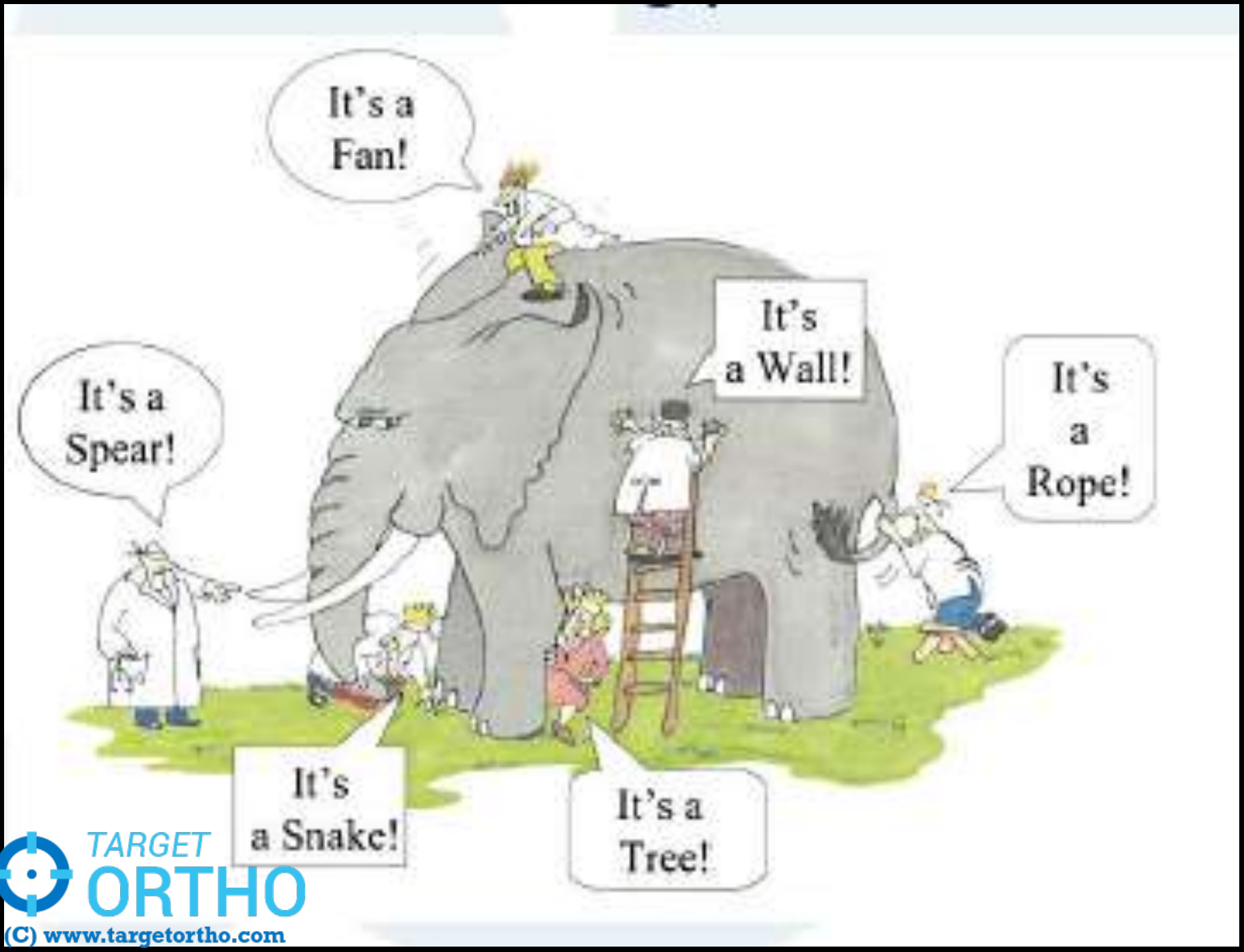
# PROGNOSIS for AMBULATION

## GOOD Prognosis

- Hemiplegics/ Ataxic pxs
- Achievement of all motor skills by age of 8.
- Independent sitting before 2 years
- Persistence of fewer than 3 of the primitive reflexes at age 18 months.

## POOR Prognosis

- Quadriplegics
- Did not attain independent sitting by age 4.
- Persistence of primitive reflexes beyond 18 months



# Spinal Dysraphism

- complex group of developmental abnormalities of the spine and neural axis in which there is a nerve tissue anomaly,
- usually combined with bony anomalies of the vertebral column

- Myelomeningocele is one of the more severe forms of *spinal dysraphism*,
- meningocele,
- lipomeningocele,
- and caudal regression syndrome

# Myelomeningocoele occulta

- overlying sinus,
- fatty deposit,
- or hemangioma

- Tethered cord syndrome

# Spinal cord

- Dysplasia of the spinal cord is invariably present.

- The cord may be

  (1) cystic or cavitated,

  (2) solid but degenerated and disorganized,

  (3) grossly proliferated

# Classification

- Thoracic
- Upper lumbar
- Lower lumbar
- sacral

# Thoracic

- Flail limbs
- congenital scoliosis,
- developmental scoliosis,
- and progressive congenital deficiency kyphosis.

# Uppar lumbar

- hip flexor power and some adductor power, but no motor control of the knees or feet

- No difference in ambulation potential as compared to thoracic level

# Lower thoracic

- Patients with lower lumbar lesions have greater hip adductor strength and, more important, quadriceps power to provide active knee extension.
- Those with L5 functioning have a functioning tibialis anterior, and they may have medial hamstring function as well.
- Hip strength is usually adequate to allow these patients to walk with the hips unbraced, that is, with knee-ankle-foot orthoses (KAFOs).

# Sacral level

- near-normal knee function and more stable hip, foot, and ankle function

- Their partial paralysis and insensate skin can lead to a number of foot problems, however, including cavovarus deformity, clawtoes, and neurogenic ulcers

# complications

- Latex allergy
- Pressure sores
- Infection
- Fracture

# Orthopaedic management

- Foot
- Knee
- Hip

- Foot calcaneus deformity
- Valgus deformity of ankle
- Rotational deormities

# Hip dislocation

1. simple release of the iliopsoas tendon with adductor release,
2. posterior transfer of the adductor muscle mass on the ischium to convert it into more of a hip extensor,
3. transfer of the iliopsoas tendon posterolaterally to convert it to a hip abductor (Sharrard procedure), and
4. transfer of the external oblique to the trochanter to recruit a hip abductor from the anterior abdominal wall.

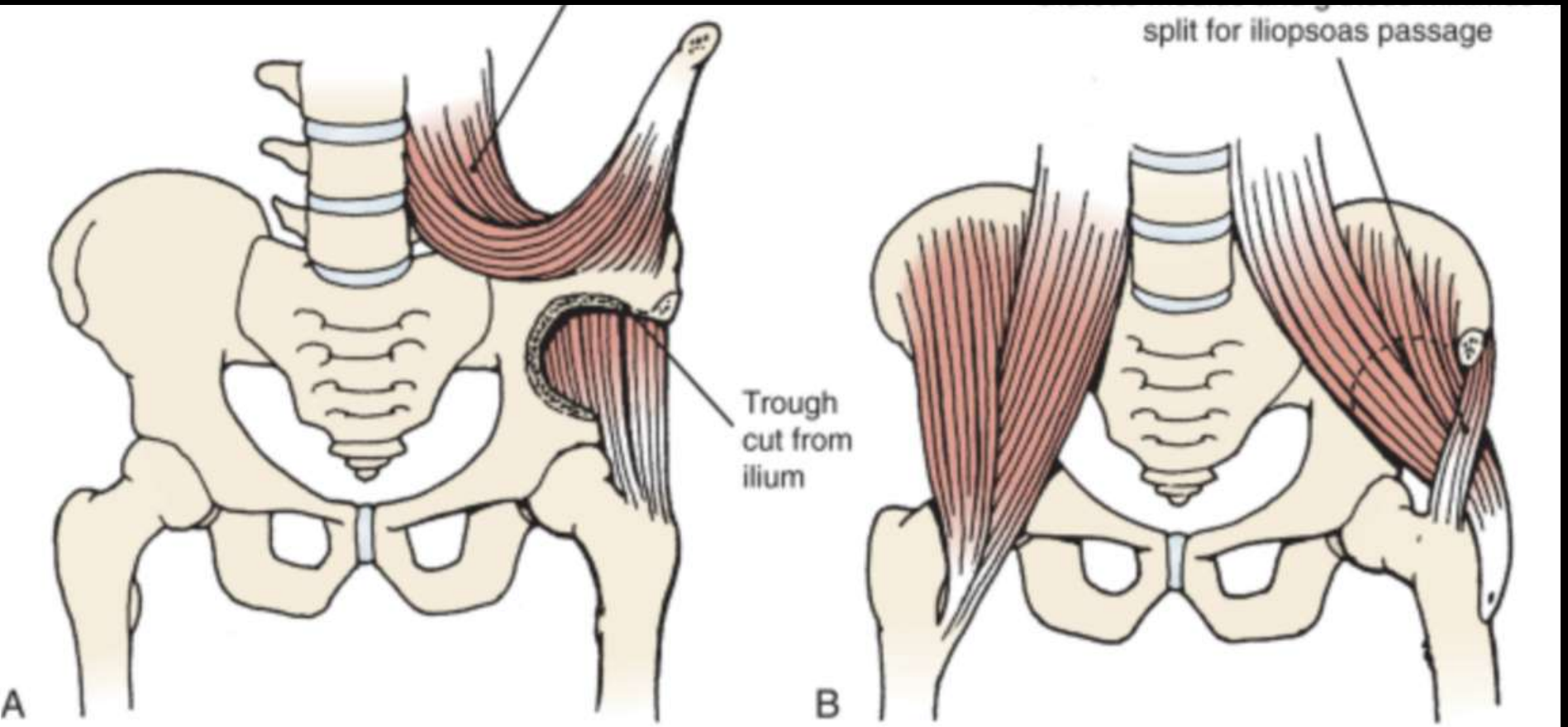



**FIGURE 26-12** Mustard transfer of the iliopsoas. **A,** The iliacus muscle is mobilized with a portion of the lesser trochanter, and a trough is made in the ilium. **B,**