DDH, COXA VARA & PFFD

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Developmental dysplasia of the hip (DDH)

- is a spectrum of disorders of development of the hip that present in different forms at different ages.
- The common etiology is excessive laxity of the hip capsule, with failure to maintain the femoral head within the acetabulum.
- The syndrome in the newborn consists of instability of the hip, such that the femoral head can be displaced partially (subluxated) or fully (dislocated) from the acetabulum by an examiner

- Klisic in 1989 recommended use of the term "developmental displacement of the hip" to indicate "a dynamic disorder.
- *Dislocation* is defined as complete displacement of a joint, with no contact between the original articular surfaces.
- *Subluxation* is defined as displacement of a joint with some contact remaining between the articular surfaces.
- *Dysplasia* refers to deficient development of the acetabulum

Teratologic dislocation of the hip

- is a distinct form of hip dislocation that usually occurs with other disorders.
- These hips are dislocated before birth, have limited range of motion, and are not reducible on examination.
- Teratologic dislocation of the hip is usually associated with other neuromuscular syndromes, especially those related to muscle paralysis, such as myelodysplasia and arthro-gryposis.
- The pathologic process, natural history, and management of teratologic dislocation are discussed separately.

incidence

- Dislocation (1.4/1000 births)
- Clinical finding (2.3/100 births)
- Ultrasound abnormality (8/100 births

Etiolgy

- Ligamentous laxity (often inherited)
- Breech position (especially footling)
- Postnatal positioning (hips swaddled in extension)
- Primary acetabular dysplasia (unlikely)

Conditions a/with DDH

- Torticollis (15%-20%)
- Metatarsus adductus (1.5%-10%)
- Oligohydramnios

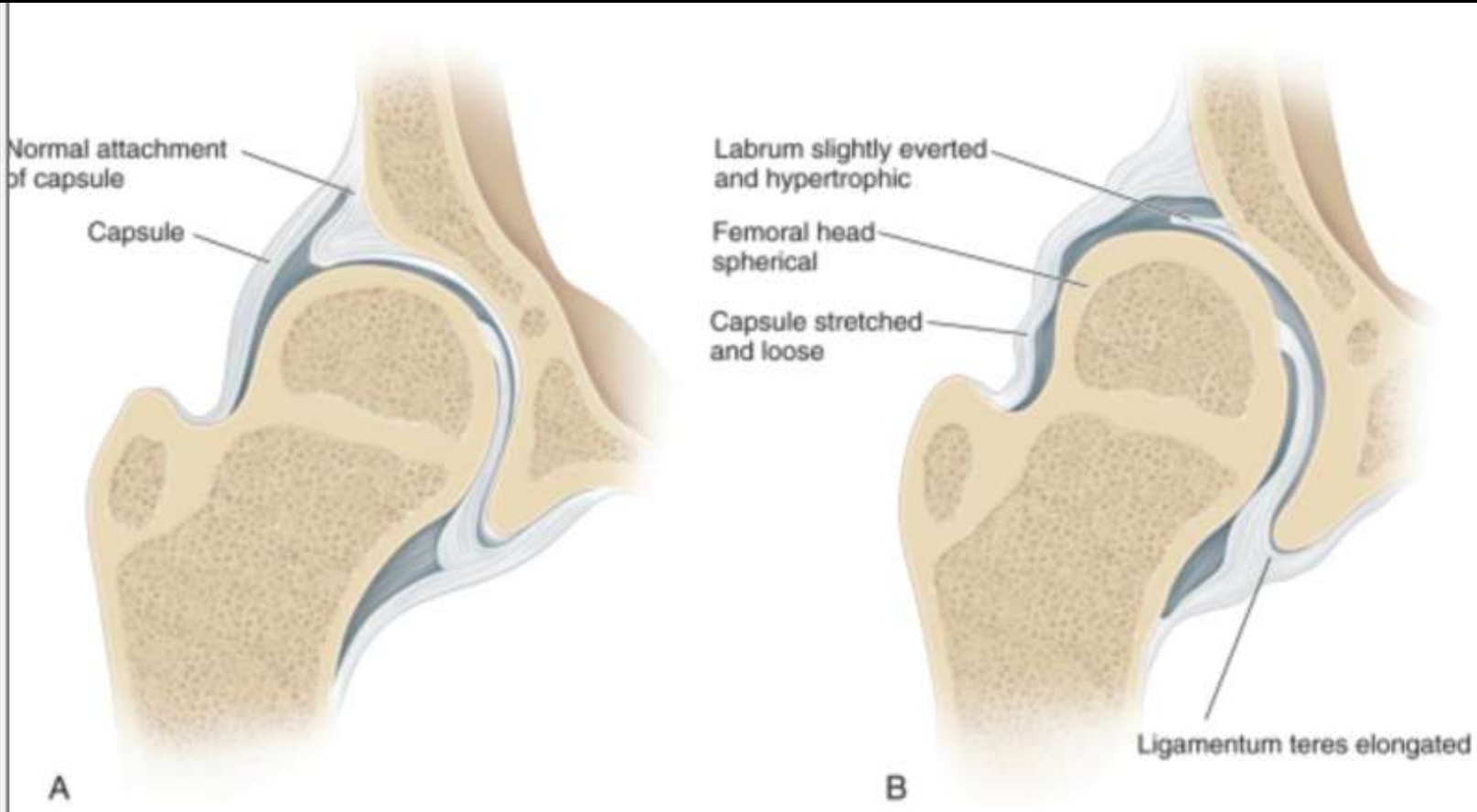


FIGURE 16-12 Pathology of the unstable hip that is subluxatable but not dislocatable. **A,** Normal hip. **B,** Subluxatable hip. Note the loose hyperelastic capsule, elongated ligamentum teres, and slight eversion of the hypertrophied acetabular rim. The femoral head is normal in shape. Excessive femoral and acetabular antetorsion may be present, causing anatomic instability of the hip joint.

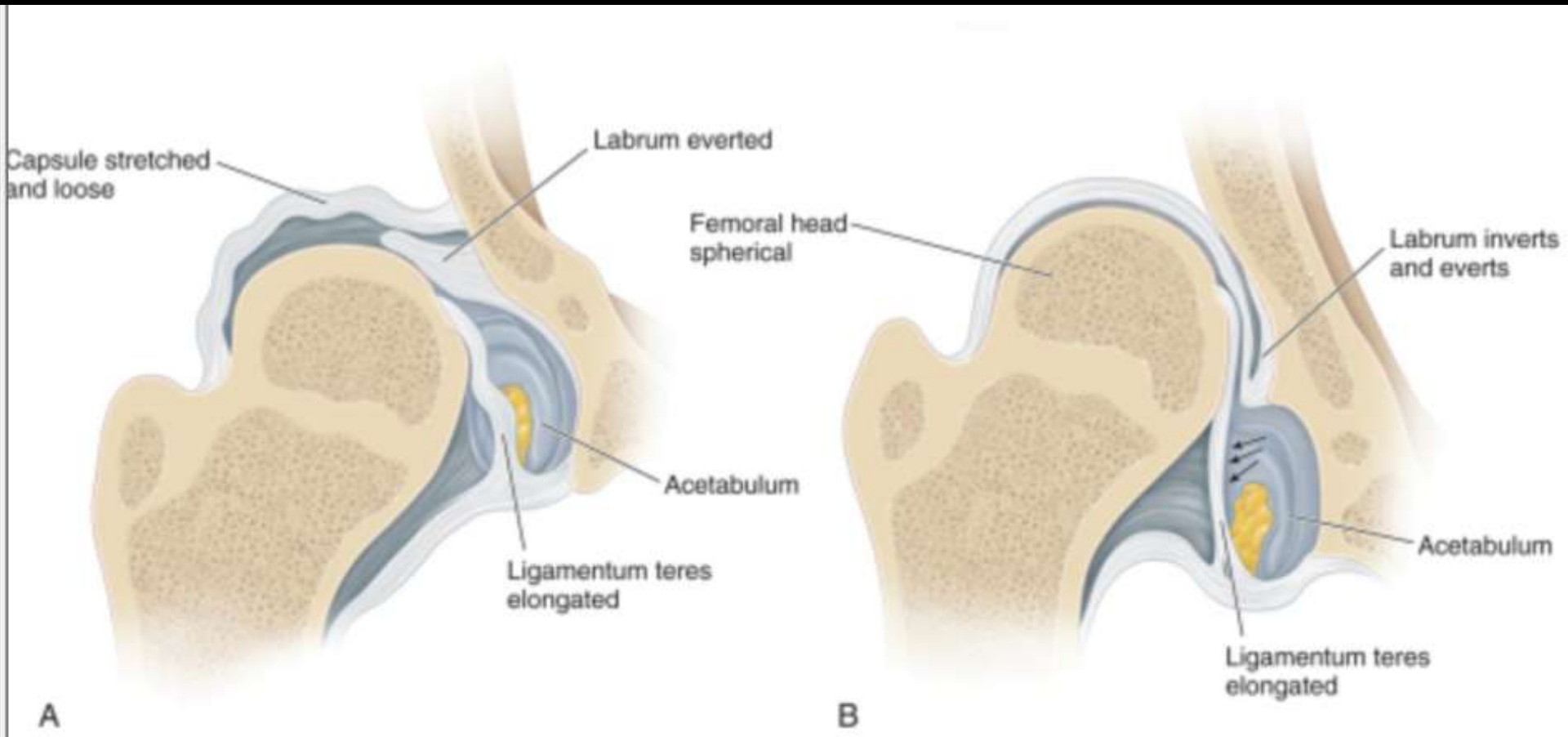
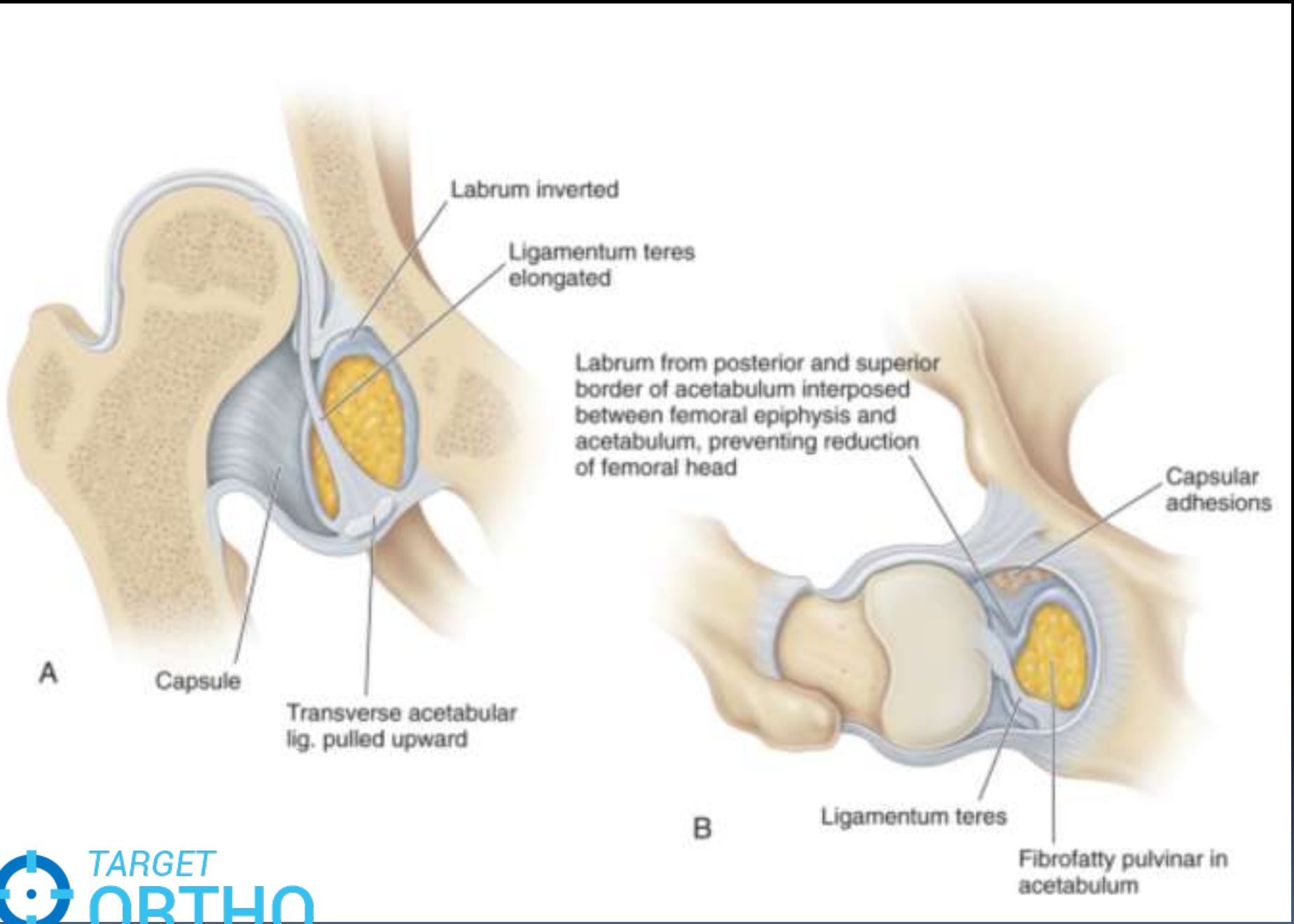
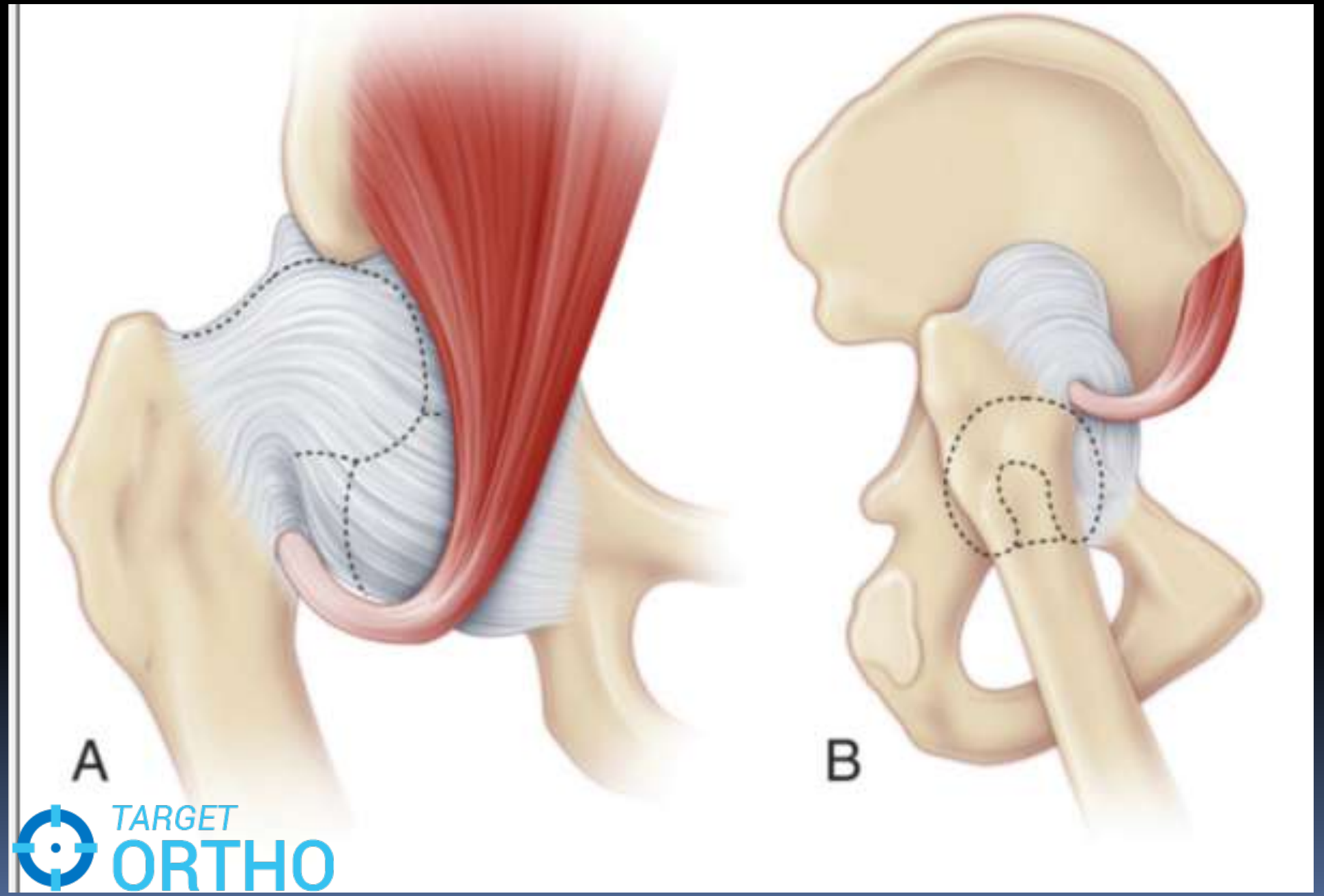
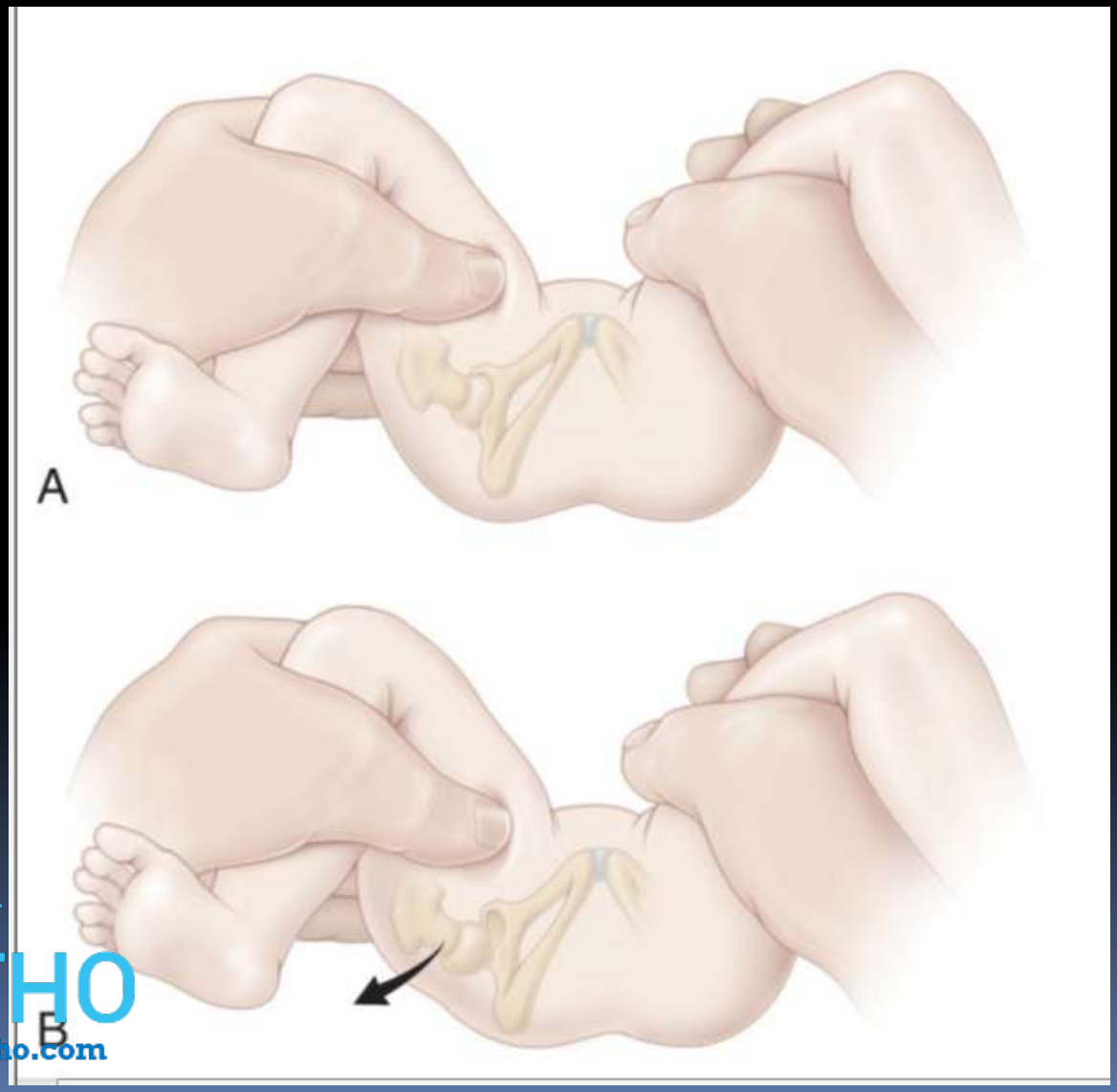


FIGURE 16-13 Pathology of the dislocatable hip. **A,** Unstable hip. The capsule is stretched out and very loose. The ligamentum teres is markedly elongated. The labrum is definitely everted. **B,** Complete displacement of the femoral head out of the acetabulum. At the fibrocartilage-hyaline junction of the labrum with the acetabulum, there may be inversional hypertrophic changes (neolimbus; *arrows*). The femoral head is spherical. Acetabular antetorsion is usually excessive.

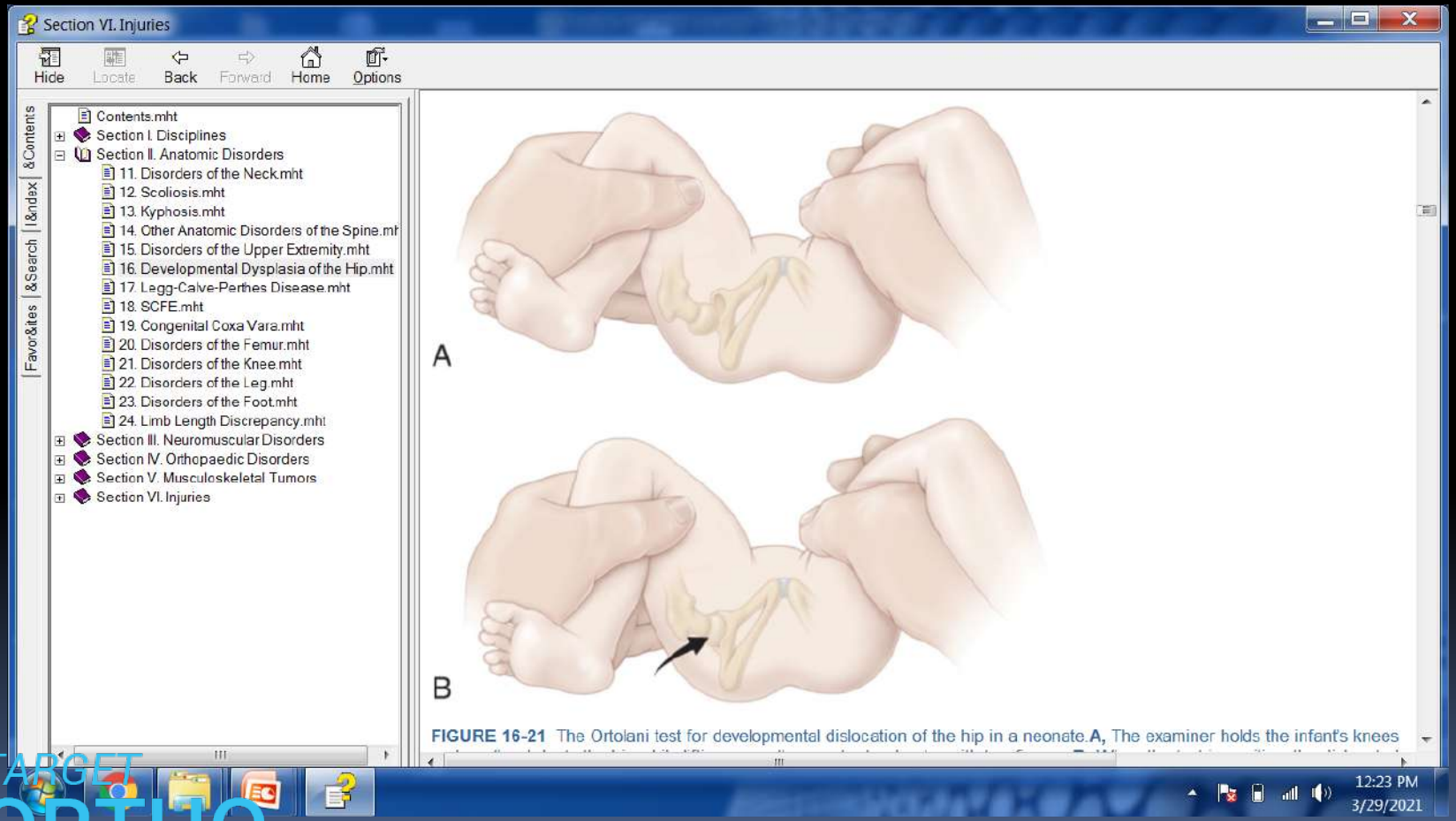




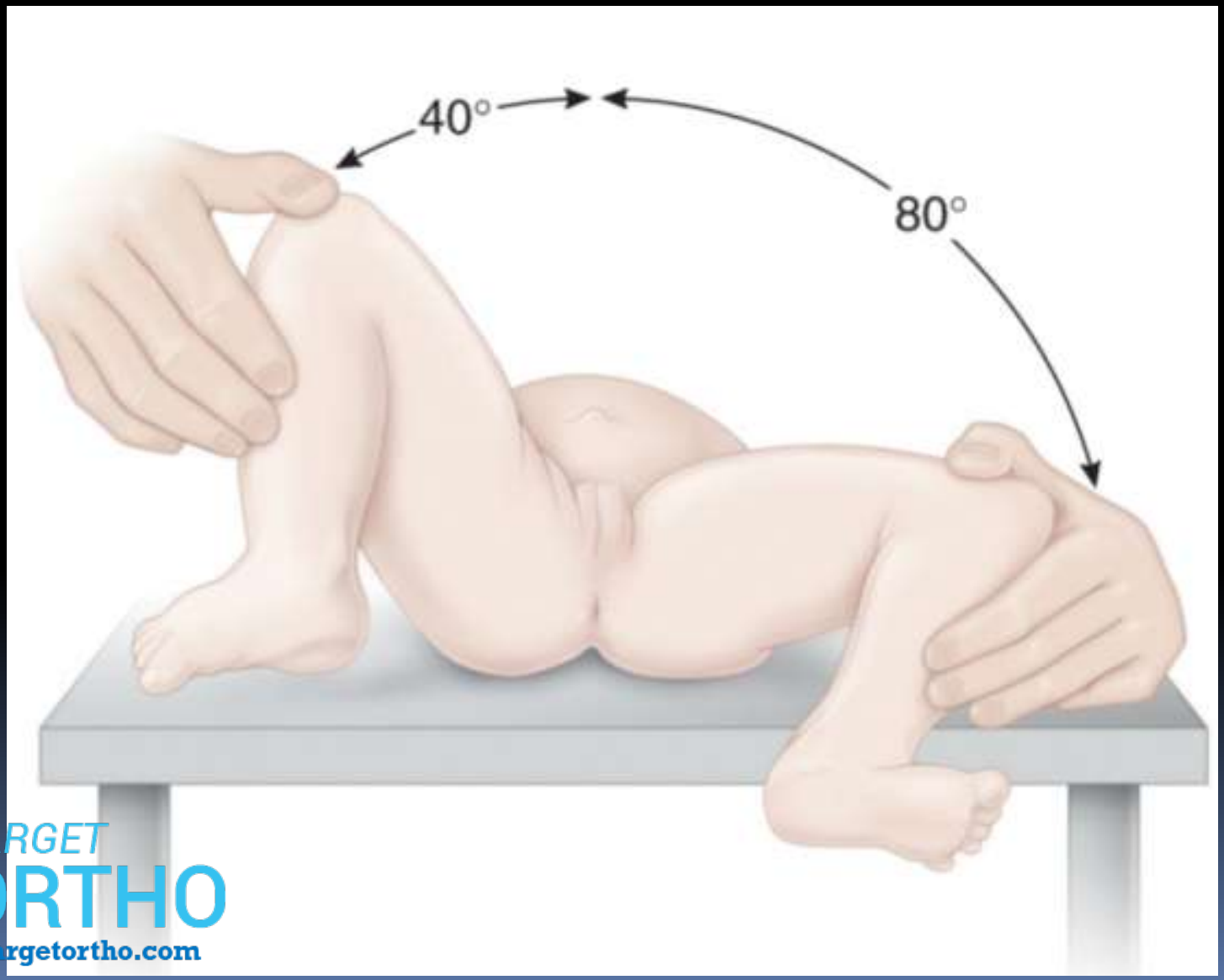
Clinical examination



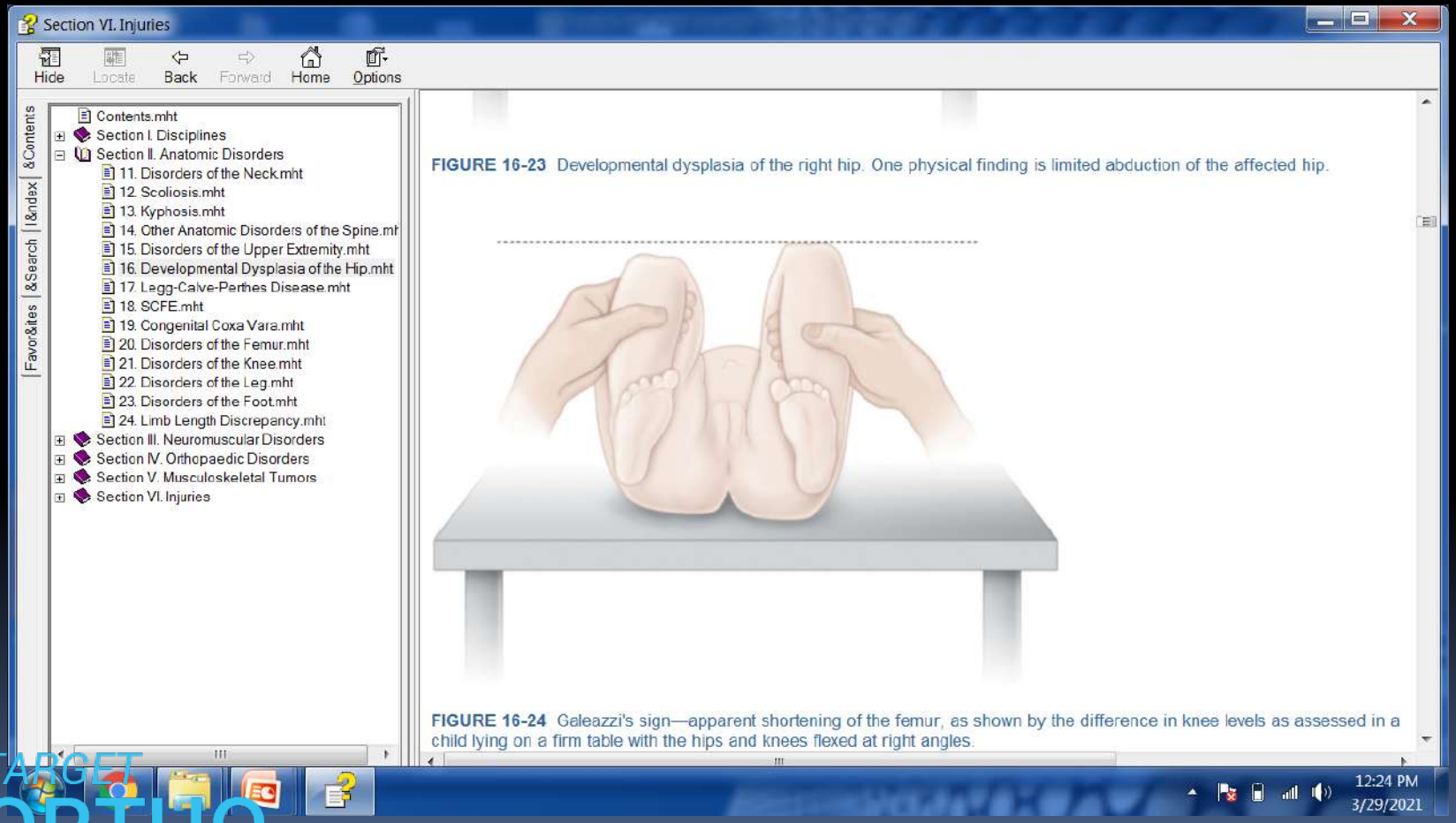
Ortolani test



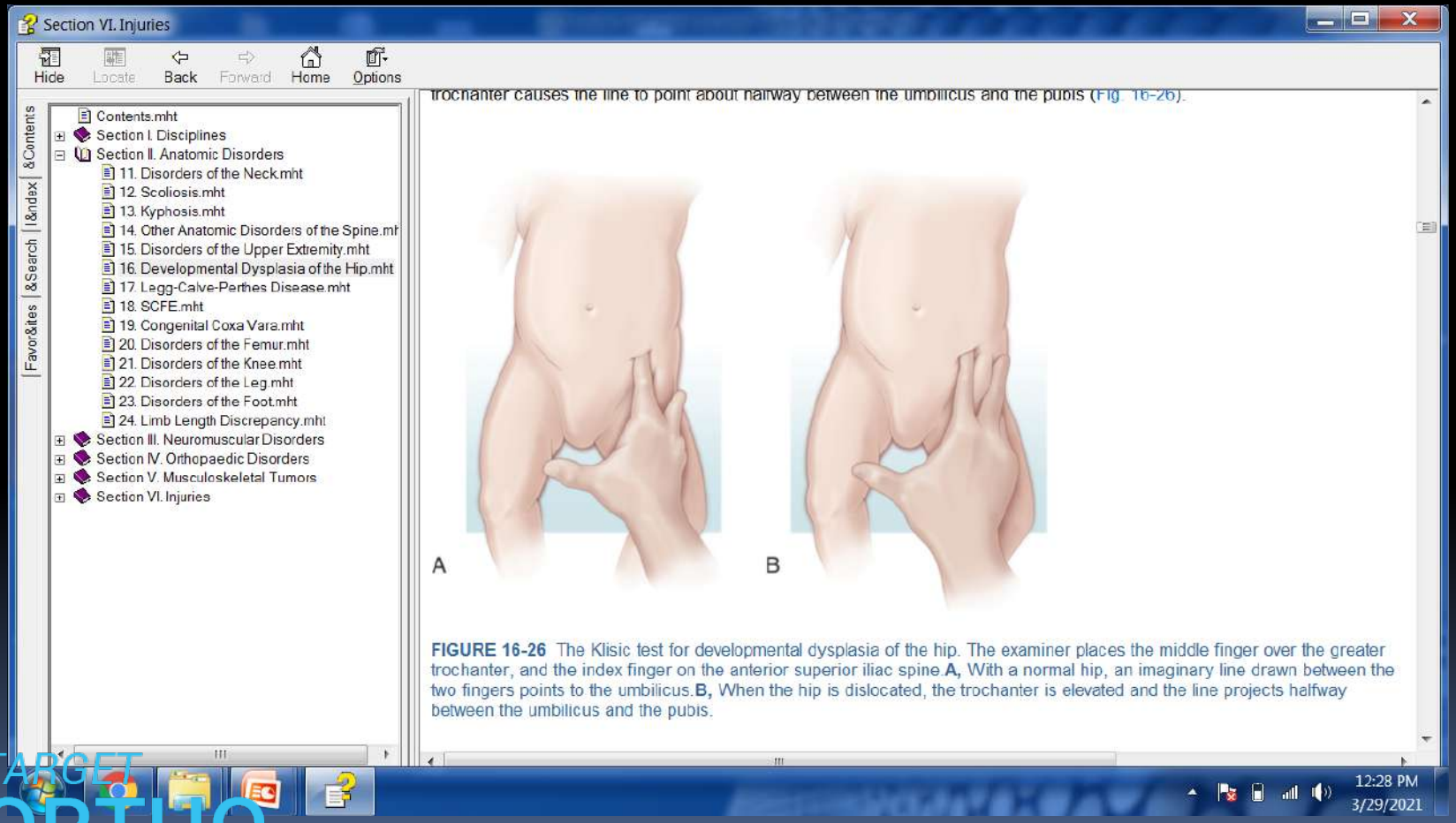
Limitation of abduction

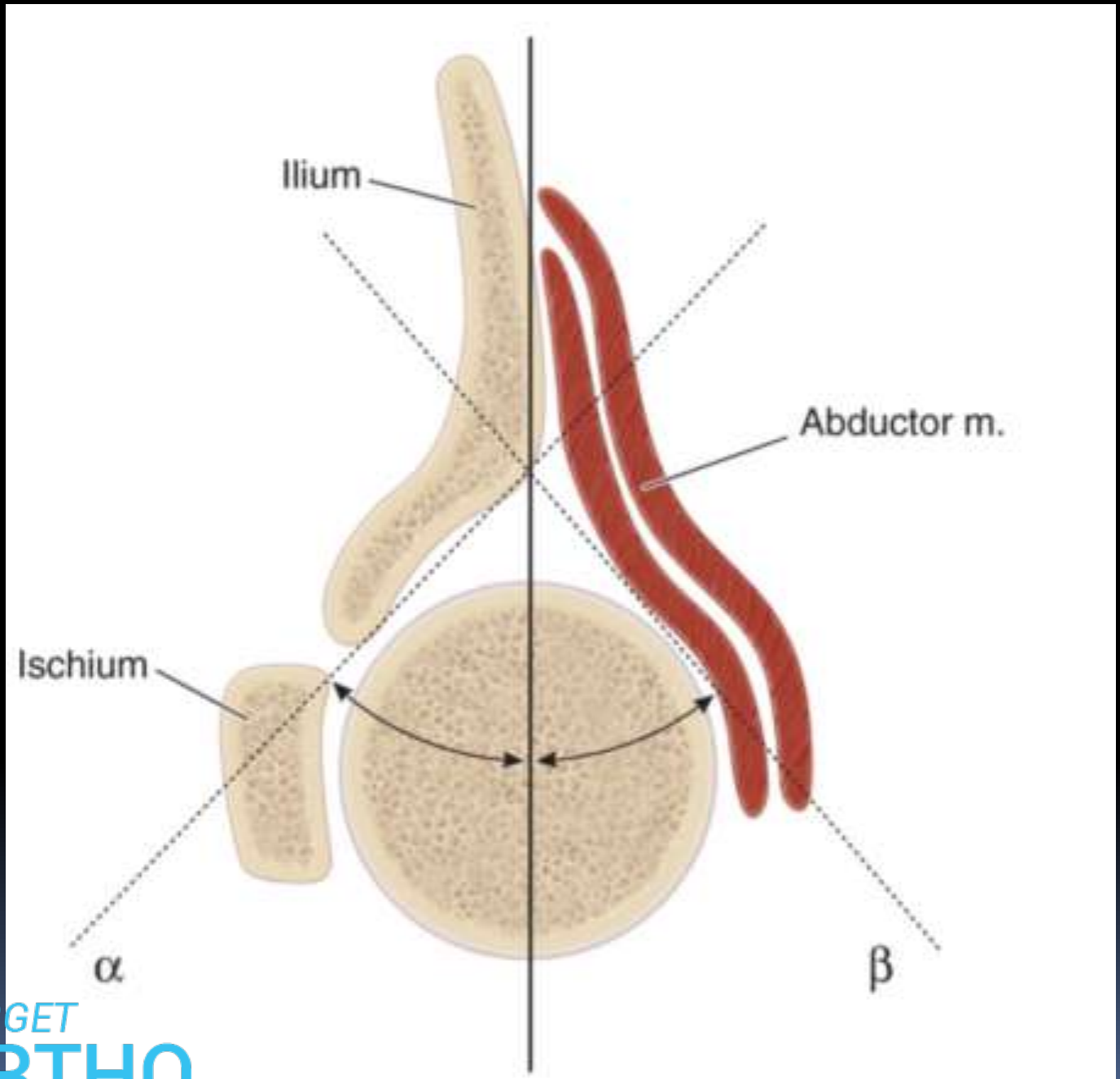


Galeazzi’s sign



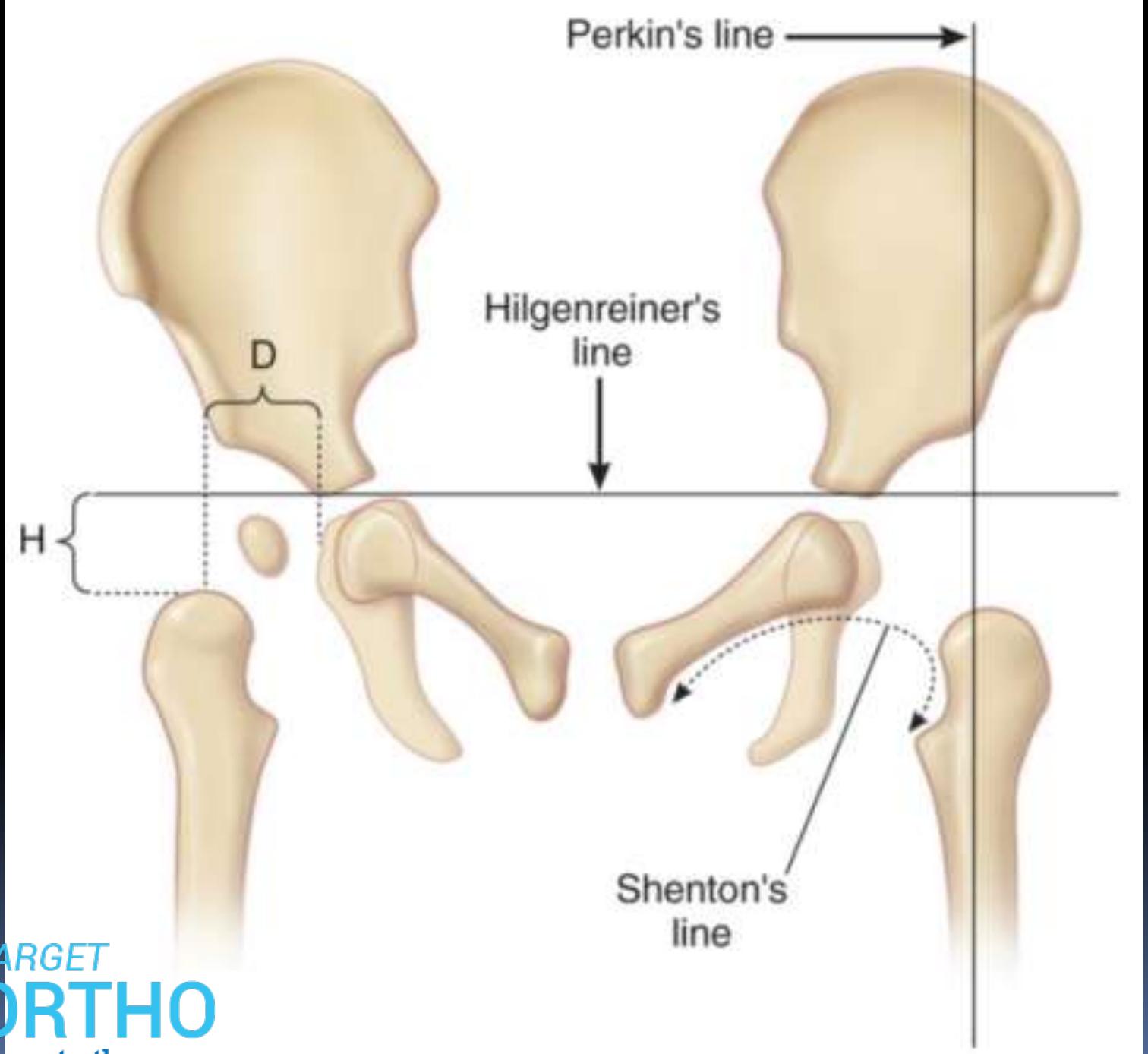
Klisic test

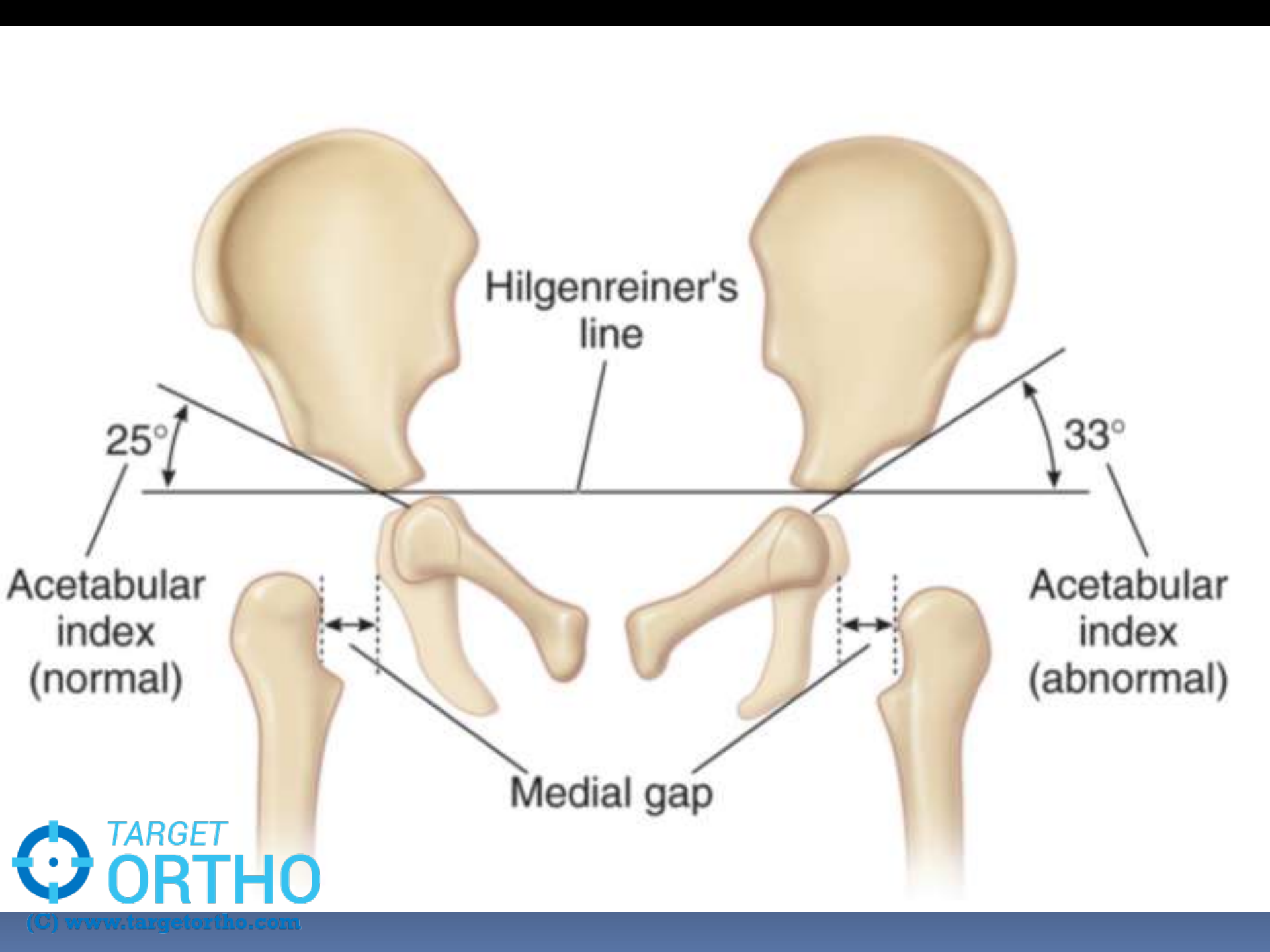




Hip

Class	Alpha Angle	Beta Angle	Description	Treatment
Standard Classification				
I	> 60°	< 55°	Normal	None
IIa	50°–60°	55°–77°	Immature (<3 mo)	Observation
IIb	>50°–60°	55°–77°	>3 mo	Pavlik harness
IIc	43°–49°	>77°	Acetabular deficiency	Pavlik harness
IId	43°–49°	>77°	Everted labrum	Pavlik harness
III	<43°	>77°	Everted labrum	Pavlik harness
IV	Unmeasurable		Dislocated	Pavlik harness/closed vs. open reduction
Simplified Classification				
I	> 60°	< 55°	Normal	None
II	43°–60°	55°–77°	Delayed ossification	?
III	<43°	>77°	Lateralization	Pavlik harness
IV	Unmeasurable		Dislocated	Pavlik harness/closed vs. open reduction





Treatment

- 6 months
- 6 months to 18 months
- 18 months and above

PRINCIPLES

-The first goal is to obtain reduction

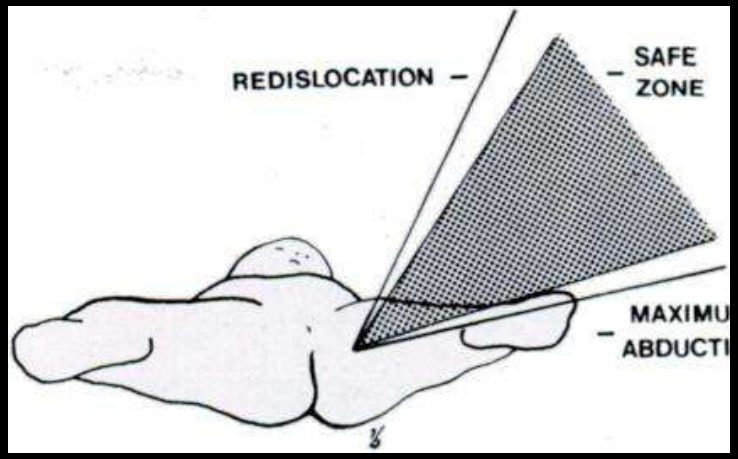
- And second is to maintain that reduction to provide an optimal environment for femoral head and acetabular development

Indications of treatment

- Most unstable hips in newborns stabilize soon after birth, some may go on to subluxation or dislocation, and some may remain located but retain anatomic dysplastic features.

- Because it is not possible to predict the outcome of unstable hips in newborns, all newborns with clinical hip instability, as manifested by a positive Ortolani or Barlow sign, should be treated.

Concept of SAFE CONE



- Initially called as SAFE ZONE by Mubaraq
- But SAFE CONE is better term as it is 3 D movement
- the arc between the angle of adduction that would allow redislocation, and the angle of abduction that can be comfortably attained
- and same time sagital and rotations should be considered

OPTIONS

- Harness
- Splints
- spica

- Triple dipers
- Abduction dipers

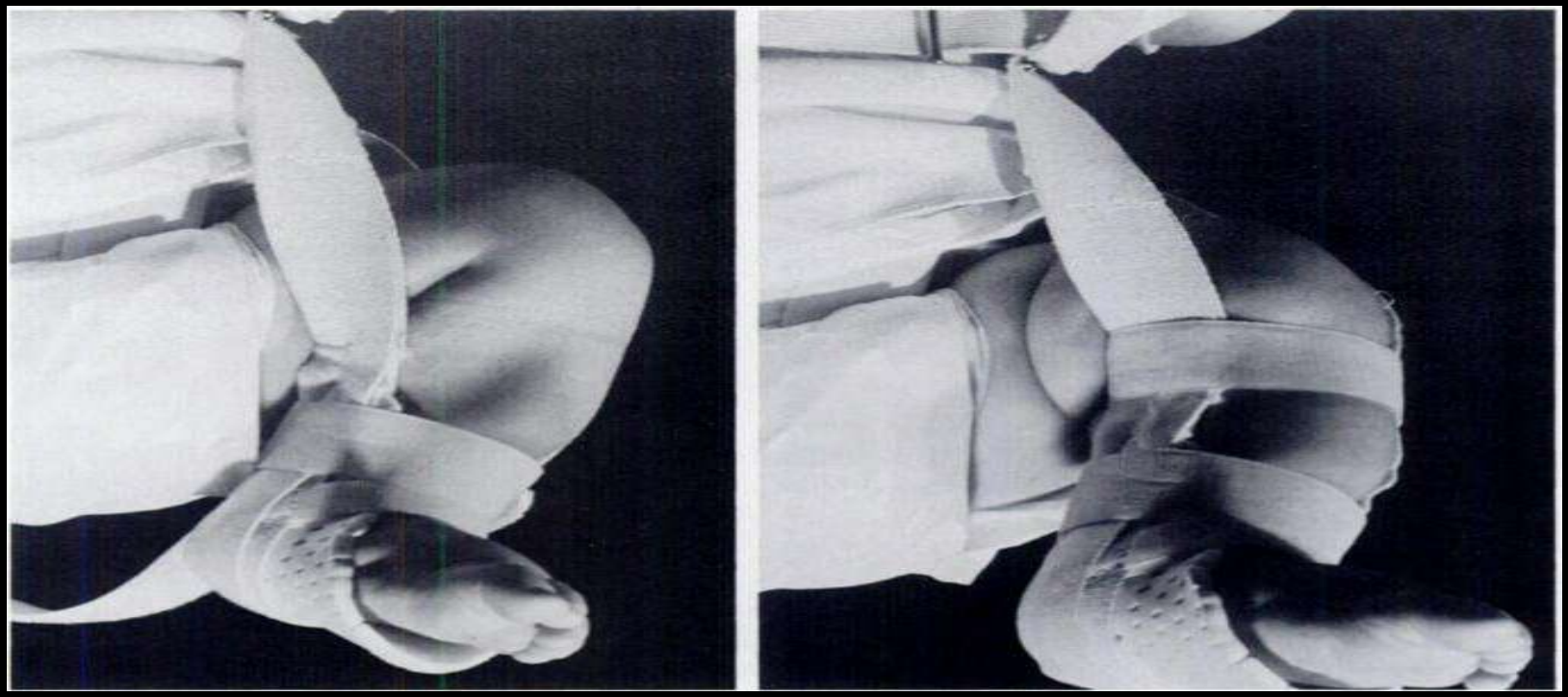
  NO ROLE

Pavlic Harness

- first used by Arnold Pavlik in 1944 for the treatment of DDH
- Dynamic flexion abduction type of orthosis
- Generally made from cotton cloths with velcro straps

How to apply on the baby

- The harness is applied with the child supine and in a comfortable undershirt.
- The chest strap is fastened first, allowing enough room for three fingers to be placed between the chest and the harness.
- The shoulder straps are buckled to maintain the chest strap at the nipple line.
- The feet are placed in the stirrups one at a time.



- The hip is placed in flexion (90 to 110 degrees), and the anterior flexion strap is tightened to maintain this position.
- The line of pull of the flexion straps must be lateral enough (e.g., along the anterior axillary line)
- Finally, the lateral strap is loosely fastened to limit adduction, not to force abduction.
- Excessive abduction to ensure stability is unacceptable.
- The knees should be 3 to 5 cm apart at full adduction in the harness

Imaging

- When imaging is used to aid in assessing hip position during treatment with a Pavlik harness, ultrasonography appears to be superior to anteroposterior radiography for assessing hip position.

- Clinical examination agreed with hip ultrasonography for hip positionin 100% of hips.

- Interpretation of radiographs agreed with ultrasonography in only 49% of cases in which the hip was judged to be dislocated and in 82% of cases in which the hip was judged to be reduced

Song KM, Lapinsky A. J Pediatr Orthop. 2000;20(3):317-9.

Imaging with pavlik harness

- Weekly visit
- USG on application
- USG 3wks if not satisfactory, discard harness & change modality of treatment
- Repeat USG every 2- 3 wks till sonographic maturation

- Radiographs are useful at the following times: immediately after the initiation of treatment, after any major adjustment in the harness, 1 month after weaning begins, at 6 months old, and at 1 year old for residual dysplasia or AVN

Four basic patterns of persistent dislocation

Superior –further flexion of the hip is indicated

Inferior - decrease in flexion is indicated

lateral- observe and USG follow up

posterior -persistent posterior dislocation is difficult to treat, and Pavlik harness treatment frequently is unsuccessful

Duration and weaning

- full-time harness wear is equal to the age at which stability is attained plus 2 months.
- Weaning is started by removing the harness for 2 hours each day.
- This time is doubled every 2 to 4 weeks until the device is worn only at night.
- Night bracing can be continued until the hip is normal with imaging
- ultrasound documentation can be used throughout the treatment period to verify the position of the hip.

Advantages

- The major advantages of the Pavlik harness are that it allows
- ultrasound observation of the reduction
- allows diapers to be changed without its removal,
- spontaneous reduction without rigid fixation, permits
- inexpensive and
- easy to use, radiolucent,lightweight

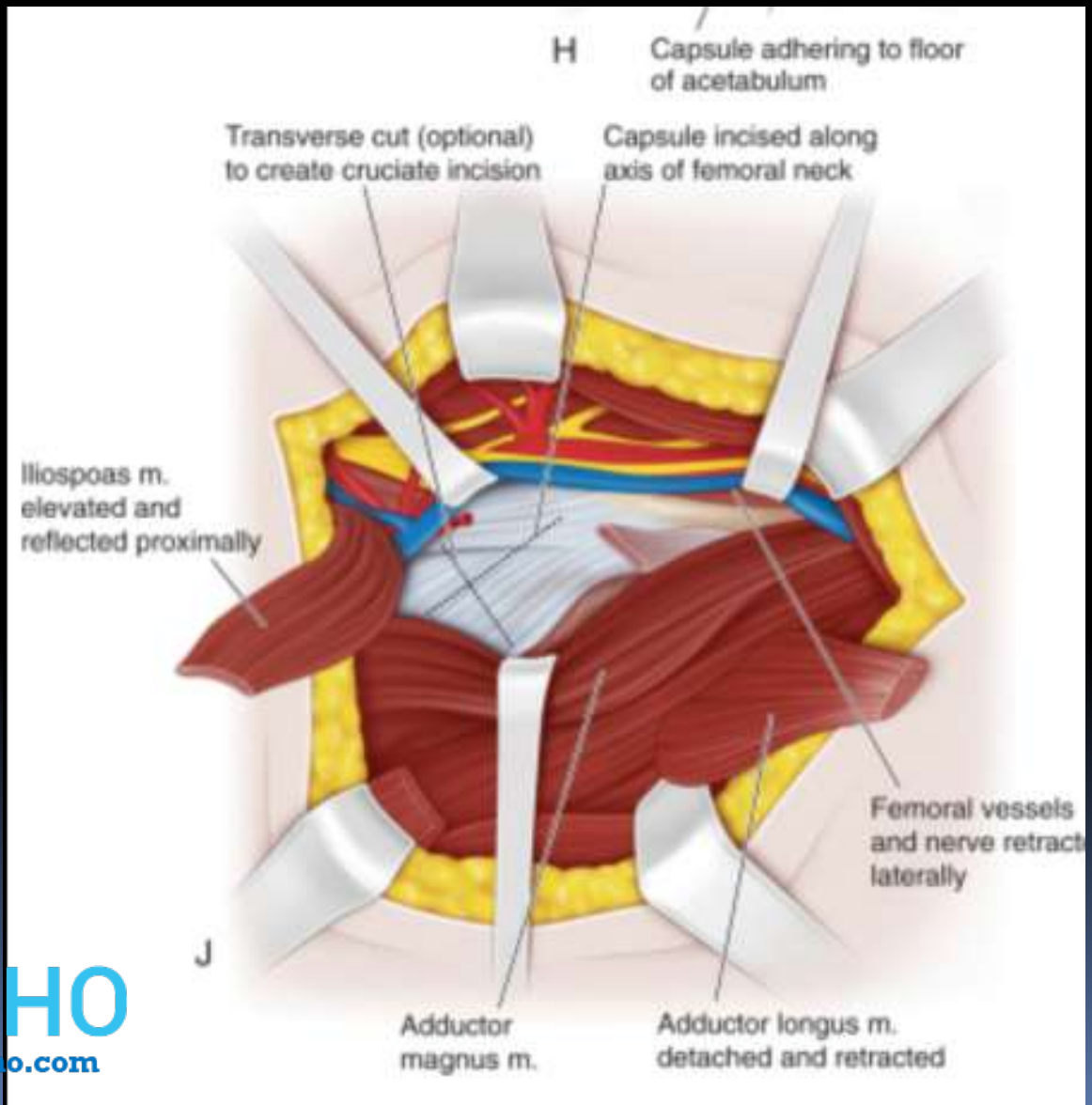
Age 6 months to 2 years of age

- Close reduction
- Arthrogram

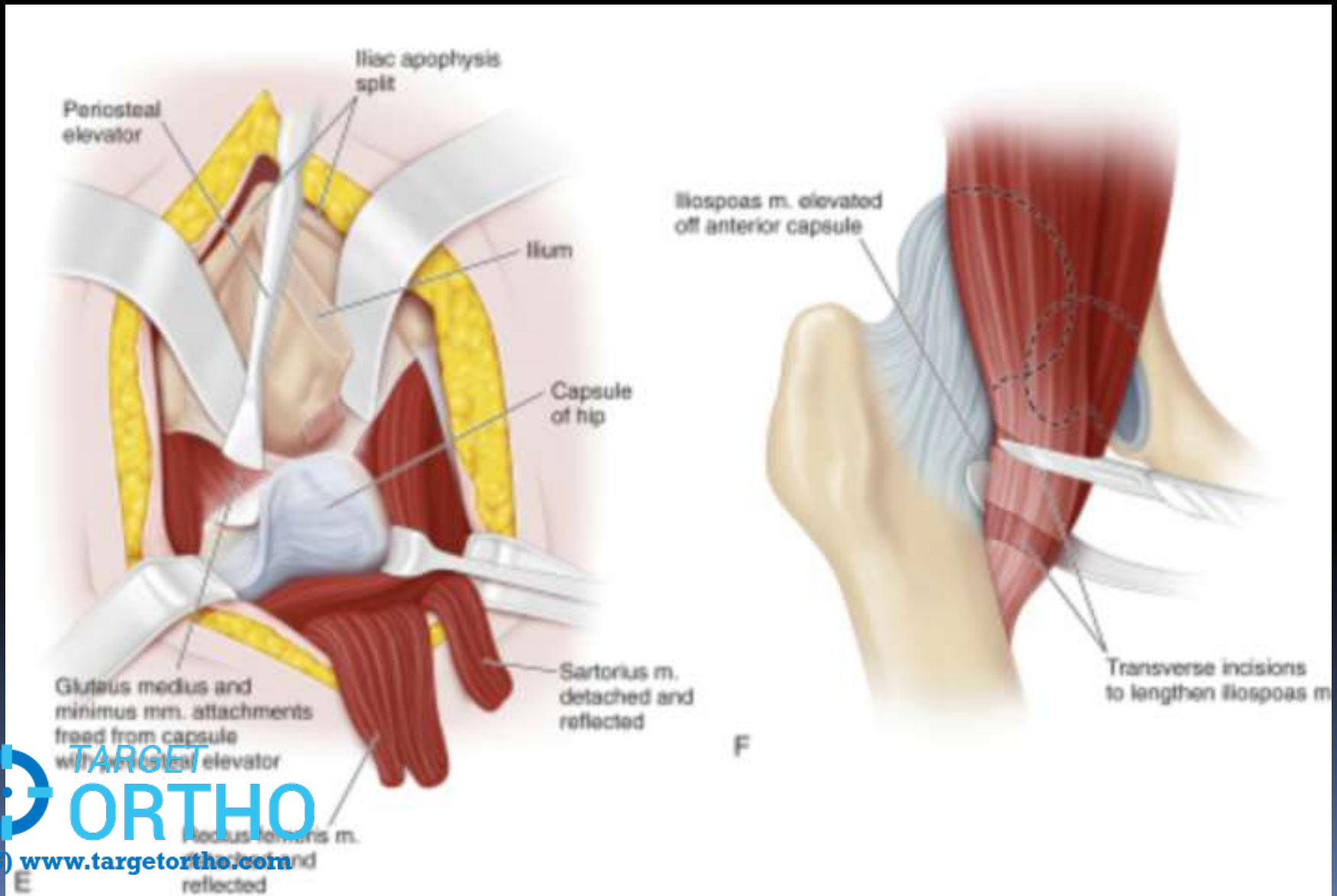
- Stable and unstable reduction

- Pop hip spica
- CT or MRI

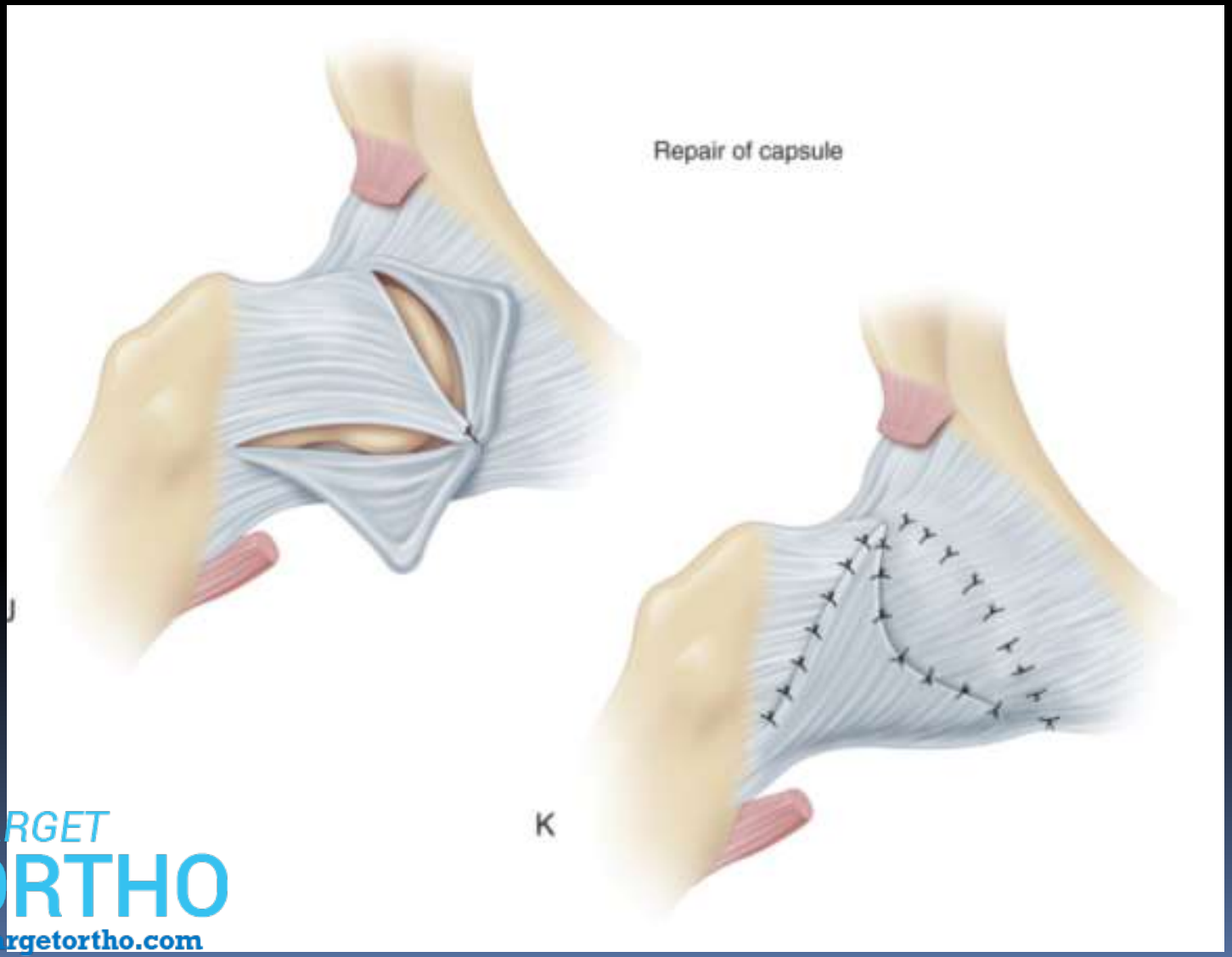
Open reduction- medial



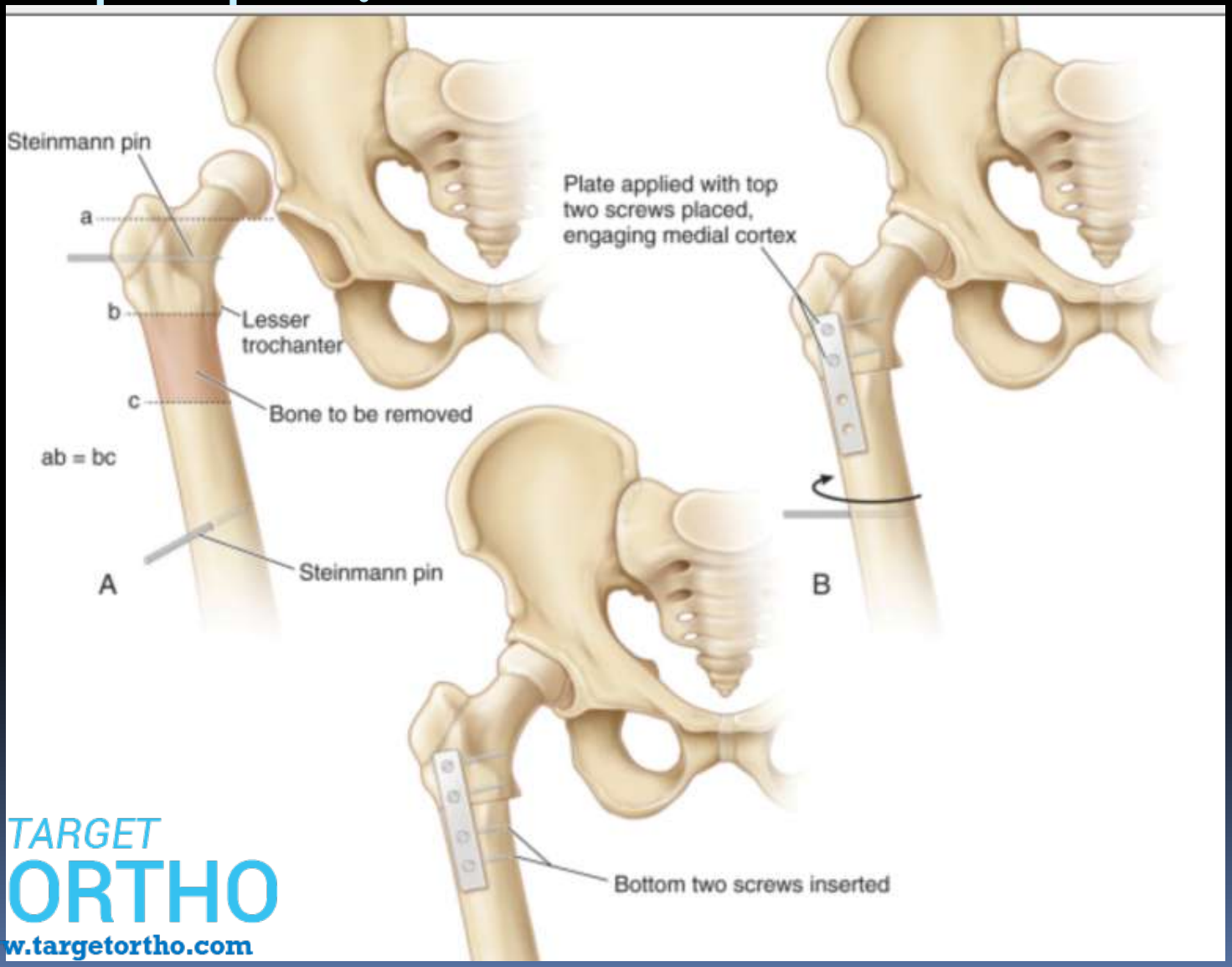
Anterior approach



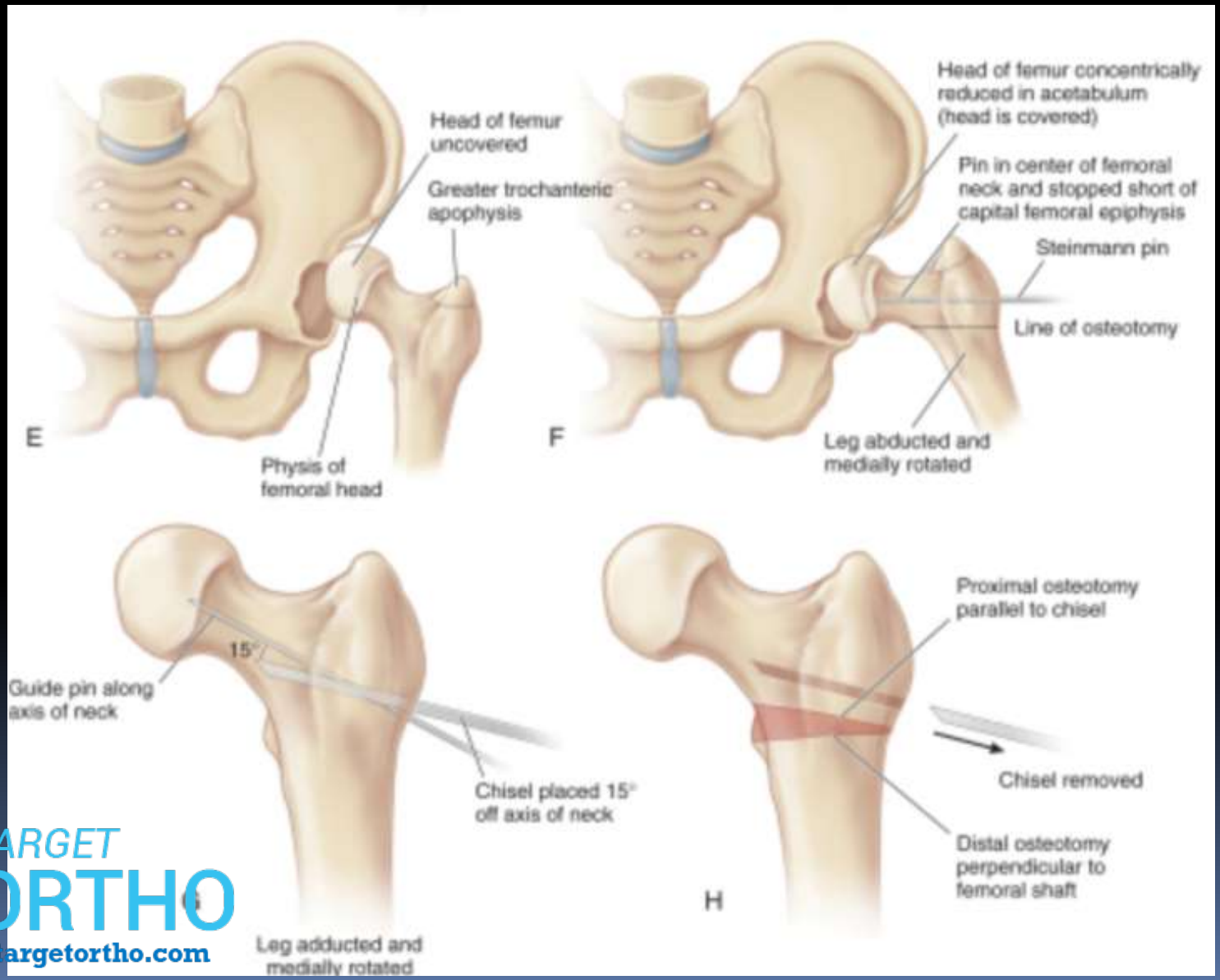
Capsulorraphy



Open reduction and femur



VDRO



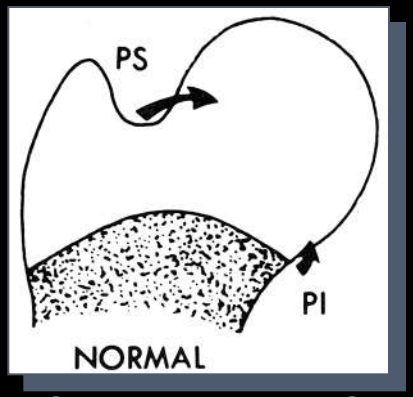
Prevalence of AVN Occurrence 2° to Tx of DDH

- Involving fem capital epiphysis & prox fem growth plate - 4-10%

- More likely to be severe following closed reduction in infancy/very young child (<6-9 mos)

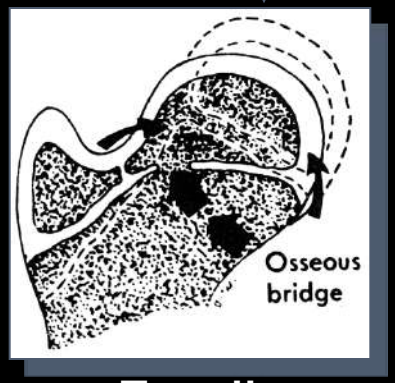
- Extreme position occludes circulation 2° to direct pressure on relatively soft head &/or kinking medial circumflex across the psoas tendon

AVN in Assoc. w/ Tx of DDH

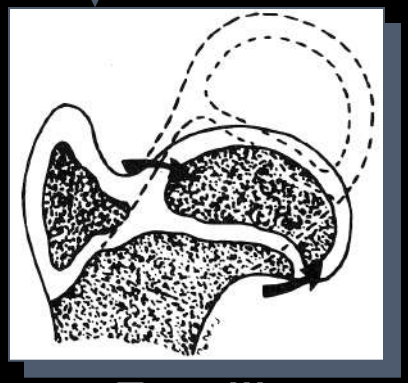


Bucholz & Ogden

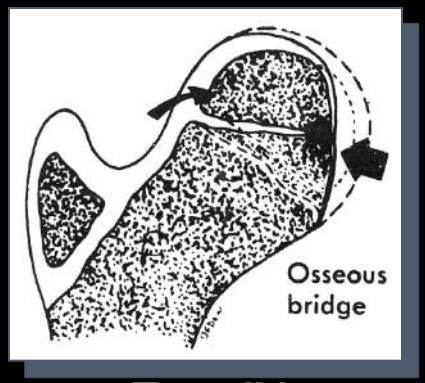
Type I



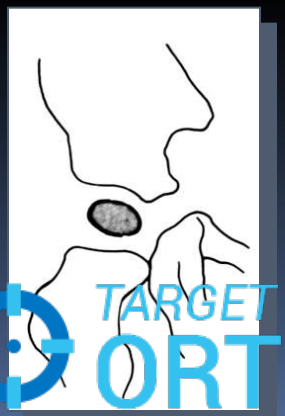
Type II



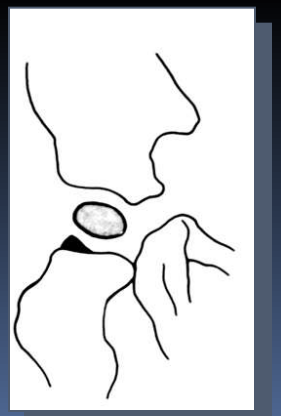
Type III



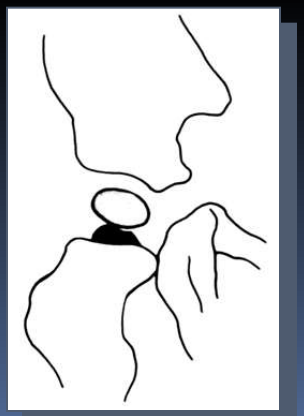
Type IV



Group I

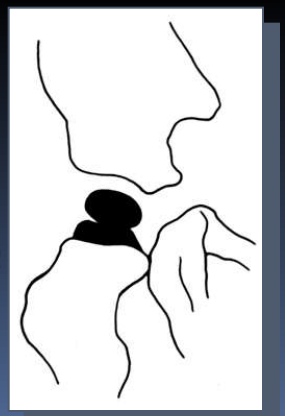


Group II

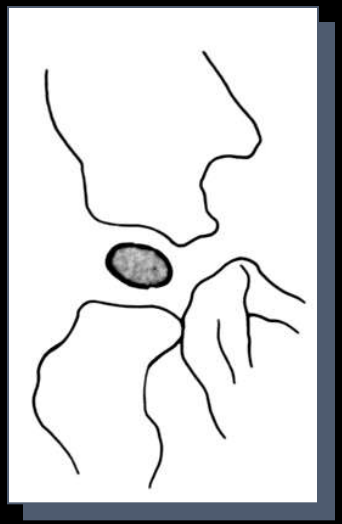


Group III

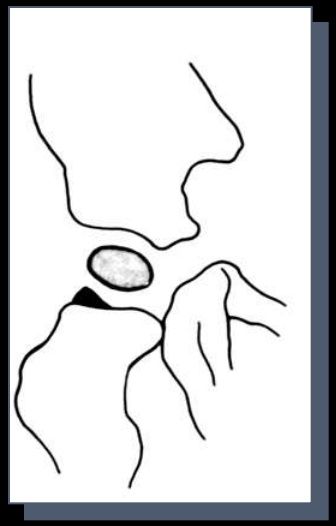
Kalamchi & MacEwen



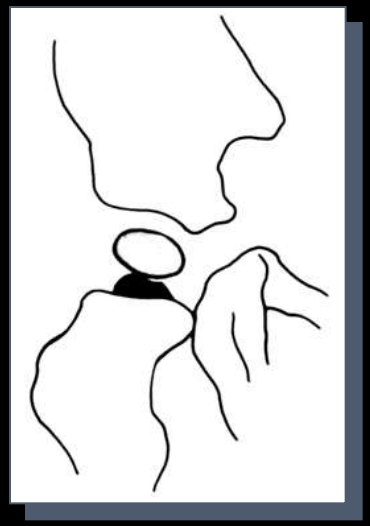
Group IV



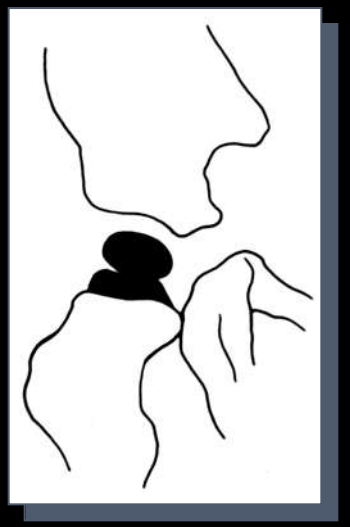
Group I – 40%



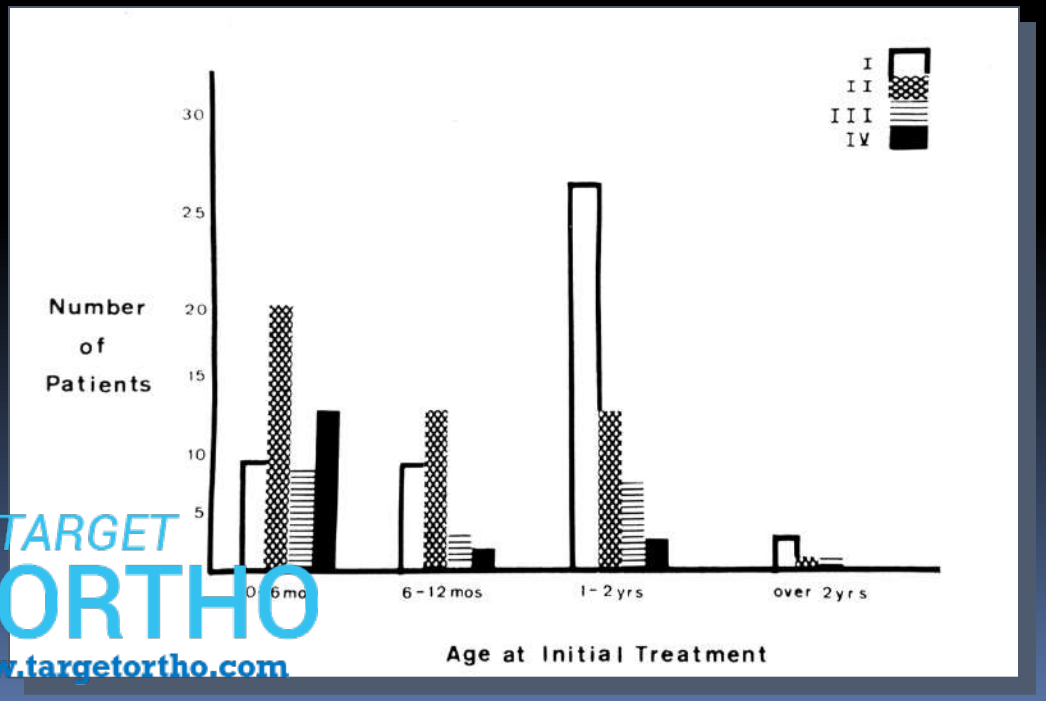
Group II - 40%



Group III – 10%

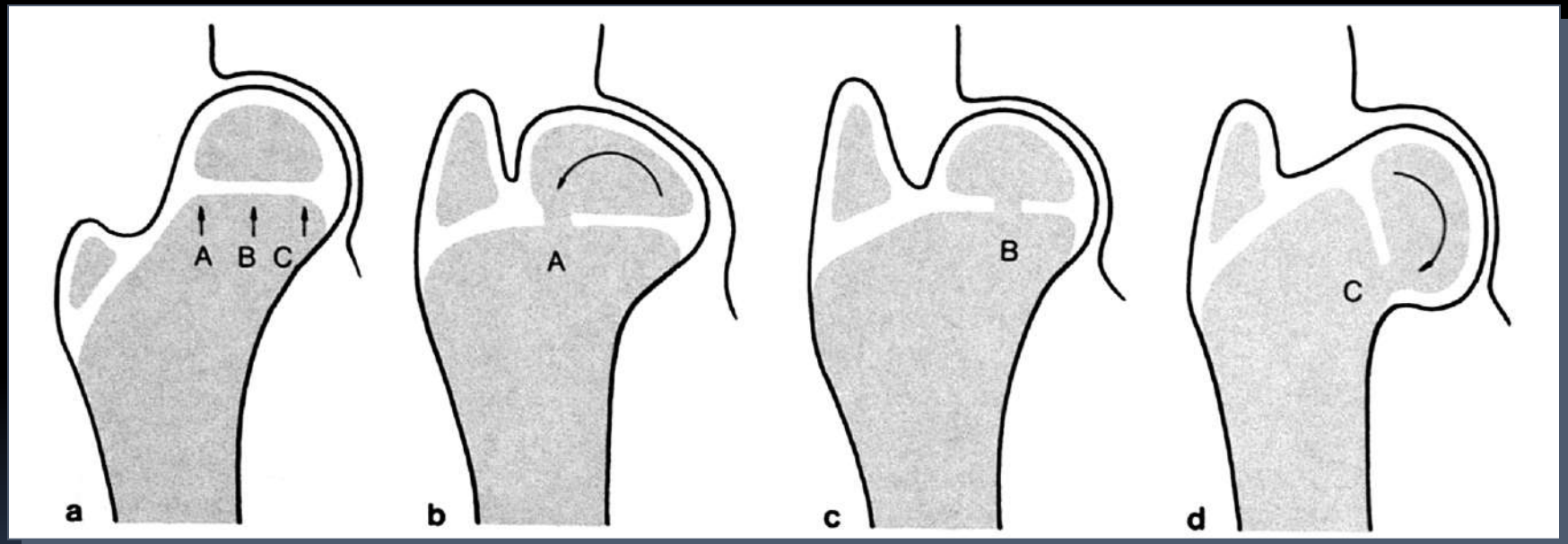


Group IV – 10%



Kalamchi & MacEwen

How Does Femoral Head Grow Following Partial Necrosis of Various Segments of the Physis?



Short & valgus

Short

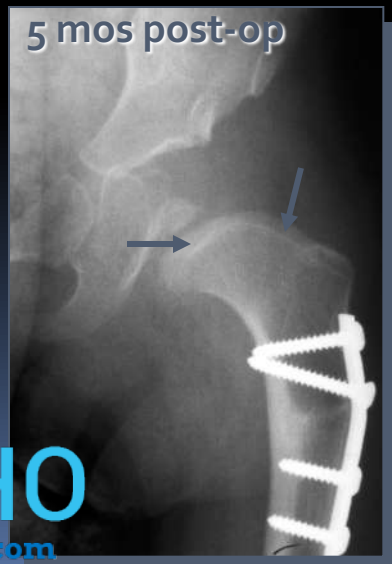
Short & varus

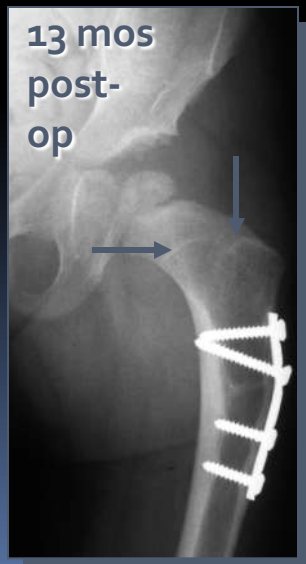
Siffert in Tönnis: Congenital Dysplasia & Dislocation of the Hip 1984:268.

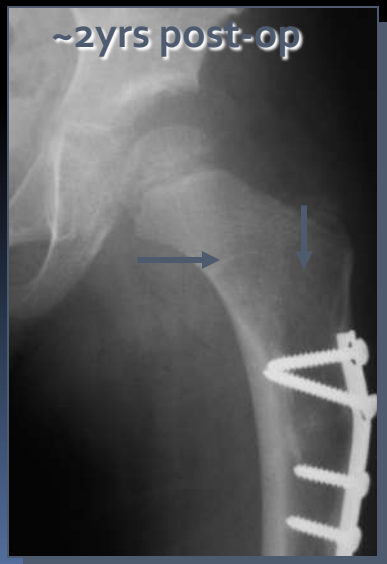
AVN in Assoc. w/ Tx of DDH

How does prox fem normally grow after successful CR?

→ look for "O'Brien's lines"



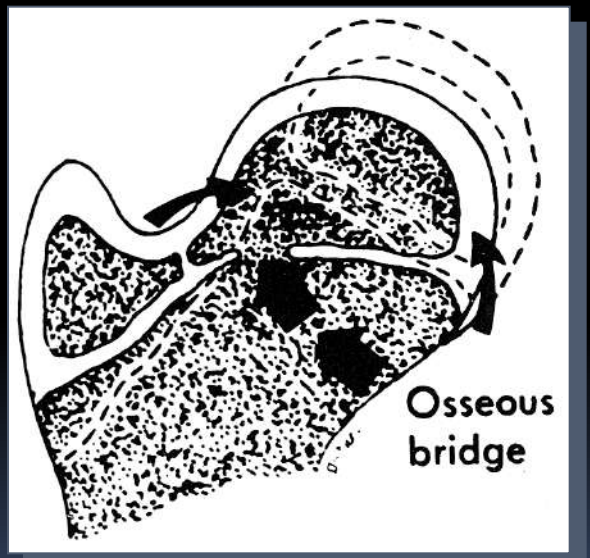




Type/Group II Growth Disturbance

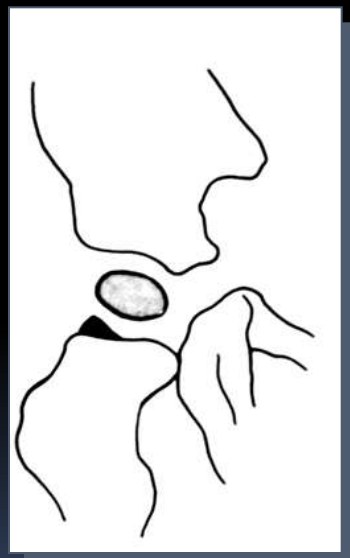
(Becomes apparent late, after 8-9 yrs)

Bucholz & Ogden



Type II

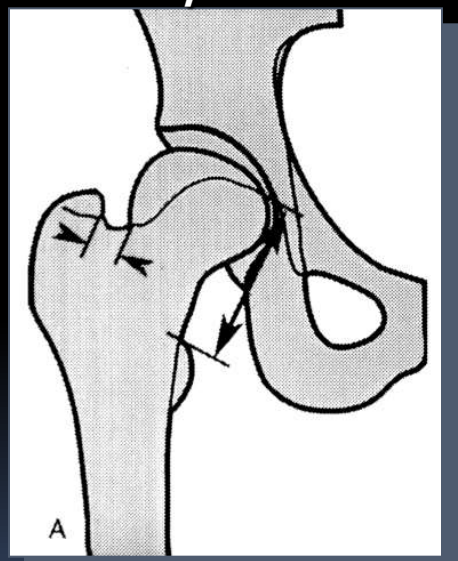
Kalamchi & MacEwen



Group II

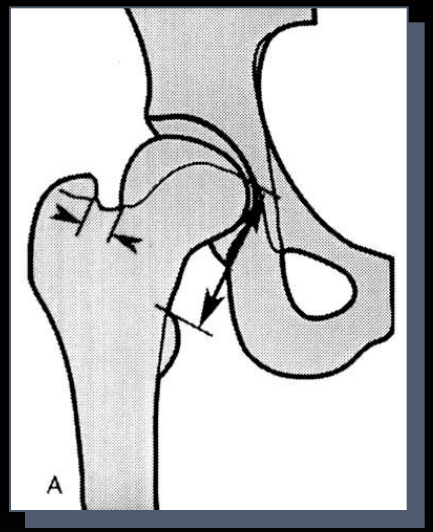
Sieffert

Oh, et. al.



Treatment for Group II Growth Disturbance Deformity

- For coxa valga
  - Proximal medial physeal arrest
    Kalenderer A & Tümer B; EPOS, April 6th, 2006
  - Proximal femoral varus osteotomy
- For acetabular dysplasia
  - Pelvic osteotomy as needed



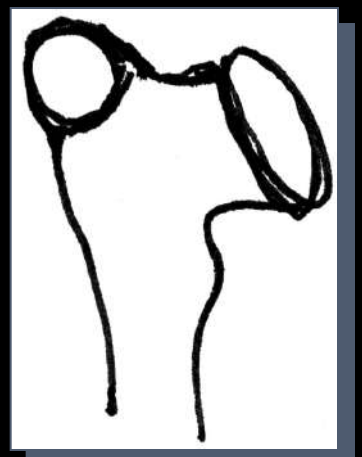
Oh C-W, Guille JT, Kumar SJ, Lipton GE, MacEwen GD. CORR 434:86, 2005

- Better outcome have been if done later & correct both the valgus (PFO) & acetabular dysplasia (pelvic ost)

Treatment for Group III or IV Growth Disturbance Deformity

Normal

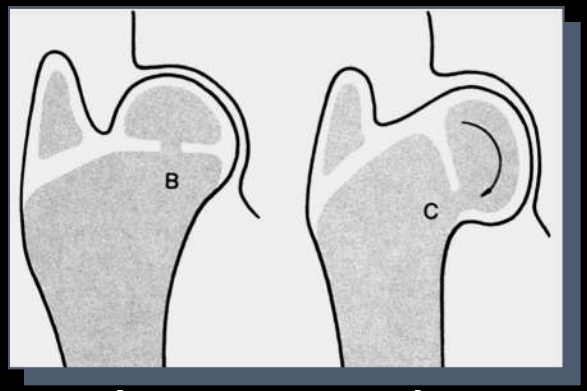
Short

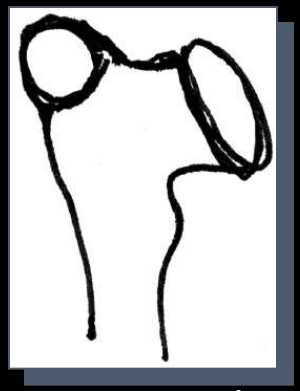


Short & varus

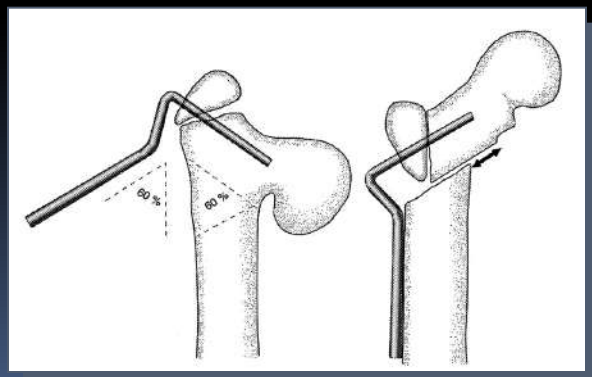
- **For short neck, varus & troch "overgrowth"**
  - Troch arrest
  - Troch transfer
  - Tx limb length discrepancy
  - Valgus osteotomy
- **For acetabular dysplasia**
  - Pelvic osteotomy
  - Shelf

Treatment for Group III & IV Growth Disturbance Deformity





- **For short neck, varus & troch "overgrowth":** troch arrest, troch transfer &/or valgus ost

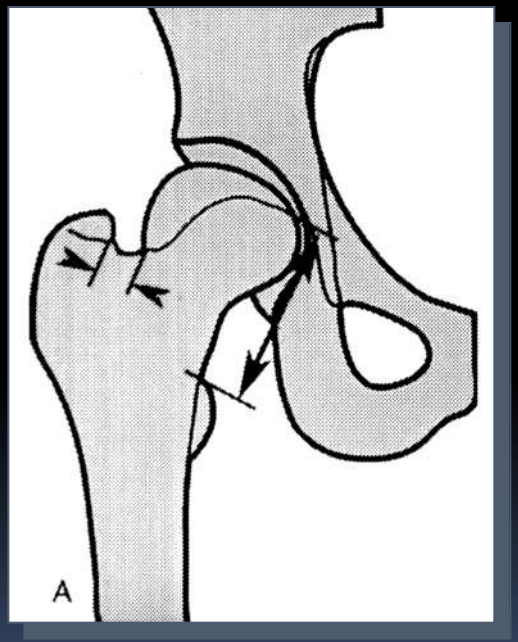


- **For acetabular dysplasia**: pelvic osteotomy, shelf

Treatment for Group II Growth Disturbance Deformity

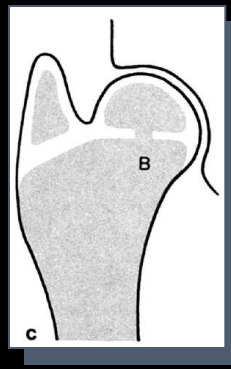
- For coxa valga
  - Proximal medial physeal arrest

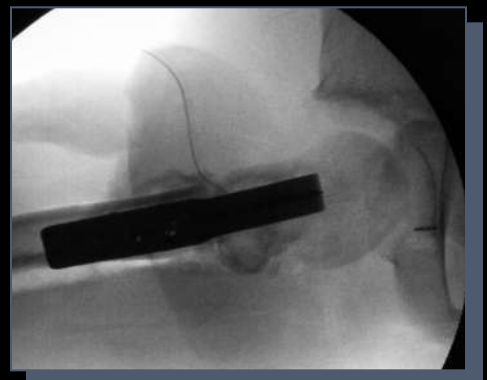
    Kalenderer A & Tümer B; EPOS, April 6th, 2006
  - Proximal femoral varus osteotomy
- For acetabular dysplasia
  - Pelvic osteotomy as needed
- Delay until 8 or 9 years of age



Oh C-W, Guille JT, Kumar SJ, Lipton GE, MacEwen GD. CORR 434:86, 2005

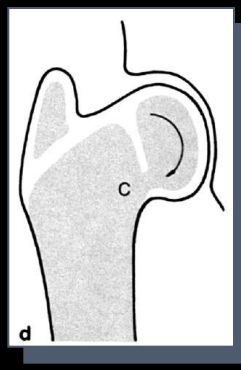
- Short neck, varus, troch overgrowth & LLD

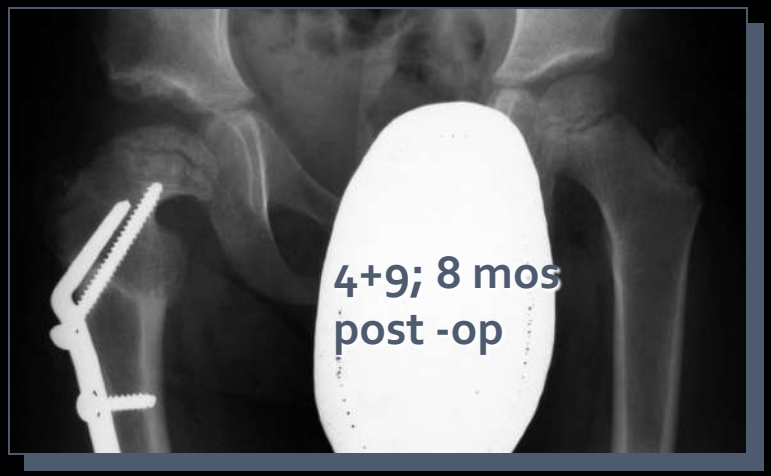


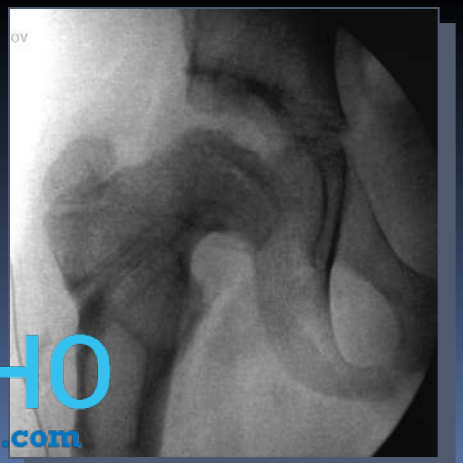


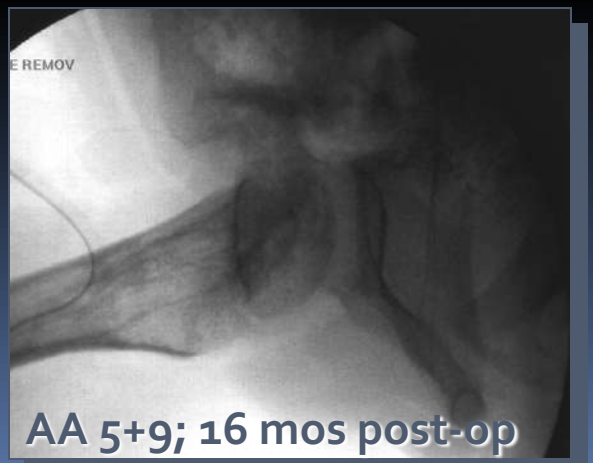
AVN in Assoc. w/ Tx of DDH

- Short neck, varus, troch overgrowth & LLD









Coxa vara

- Congenital coxa vara is a developmental abnormality characterized by a primary cartilaginous defect in the femoral neck with an abnormal decrease in the femoral neck–shaft angle, shortening of the femoral neck, relative overgrowth of the greater trochanter, and shortening of the affected lower limb

Etiology

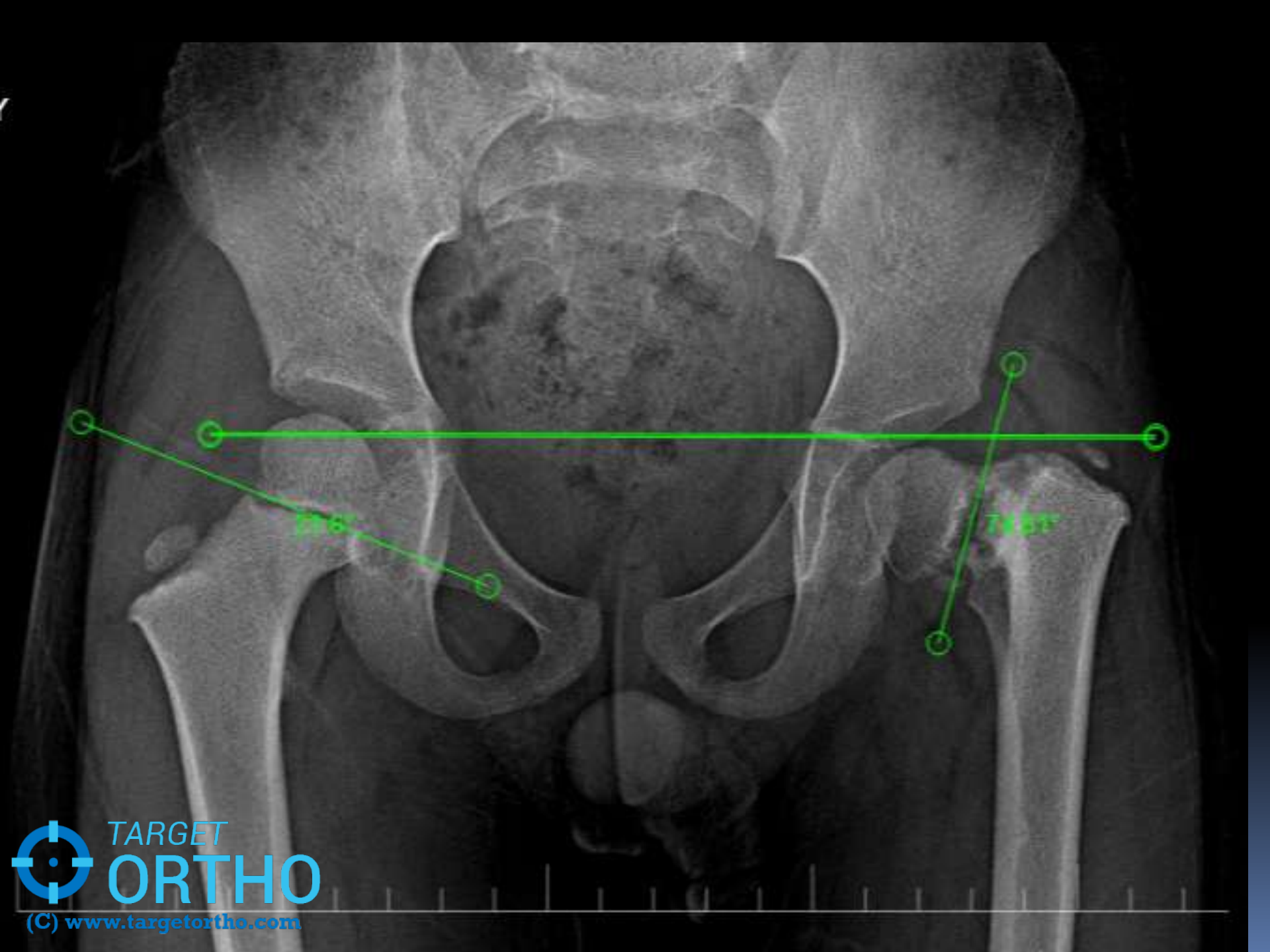
- The disorder is frequently present in skeletal dysplasias with known genetic causes,
- such as cleidocranial dysostosis;
- metaphyseal dysostosis,
- Jansen type;
- and spondylometaphyseal dysplasia, especially Kozlowski type, all of which are autosomal dominant disorders

Clinical features

- Painless limp
- Shortening
- Difficulty in cross leg sitting
- Decreased internal rotation and abduction

- Evidence of a generalized skeletal dysplasia should be sought

Radiographic findings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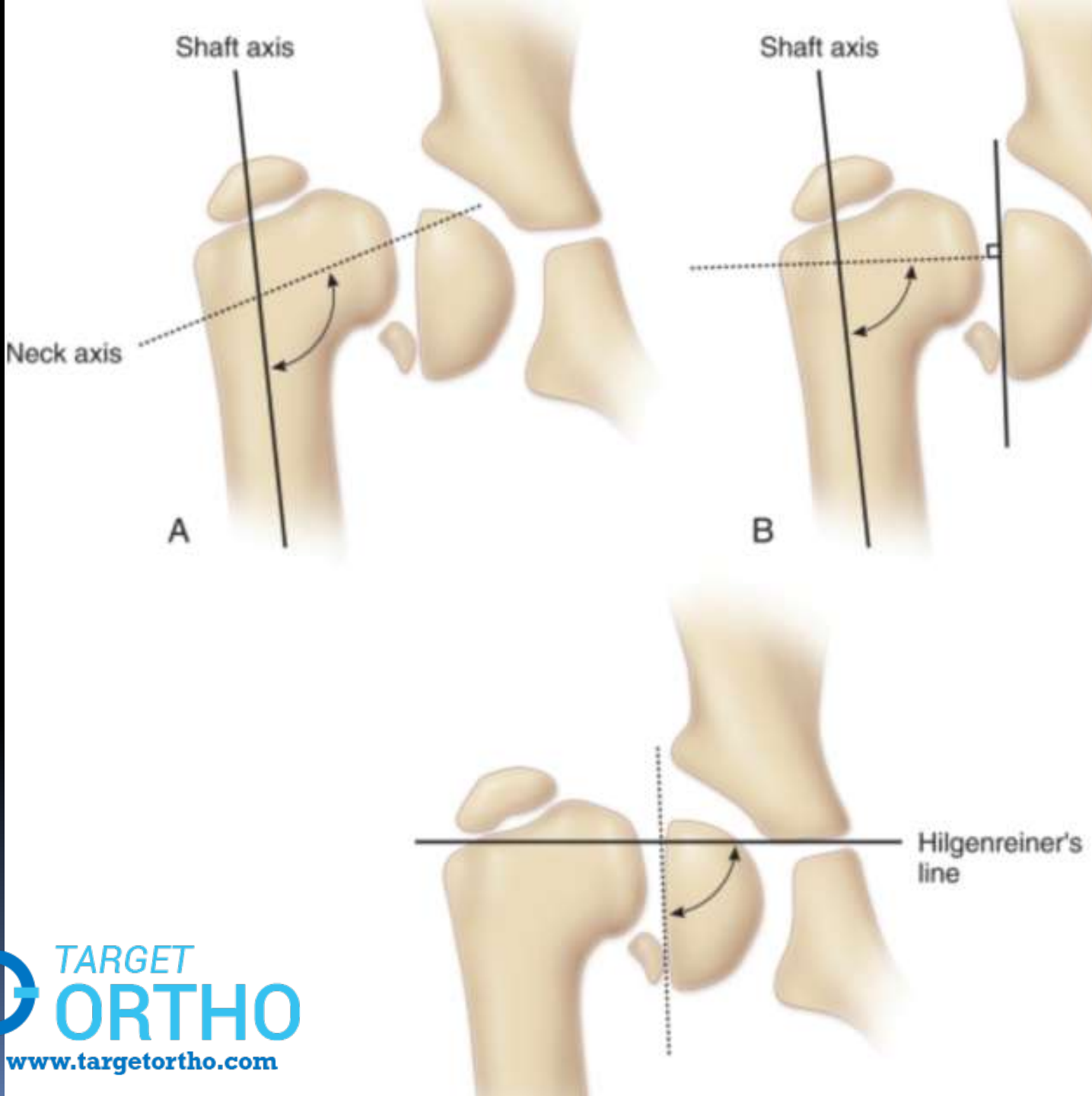
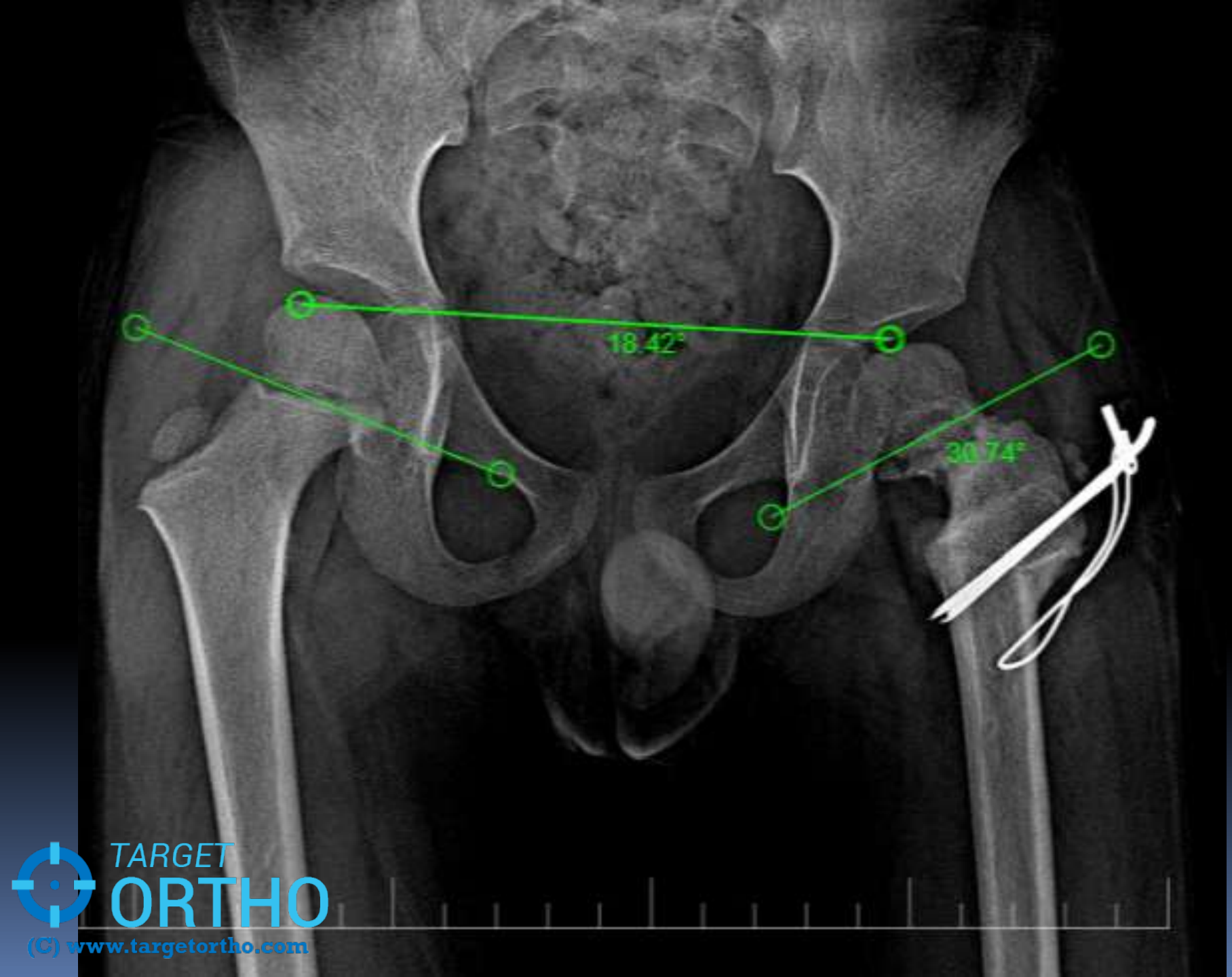


Treatment

- Valgus osteotomy of some form is recommended in hips with an H-E angle of 60 degrees or greater,
- is not usually required in patients with an angle less than 45 degrees,
- and may or may not be required in patients with angles between 45 and 59 degrees
- Pauwel's intertrochanteric osteotomy

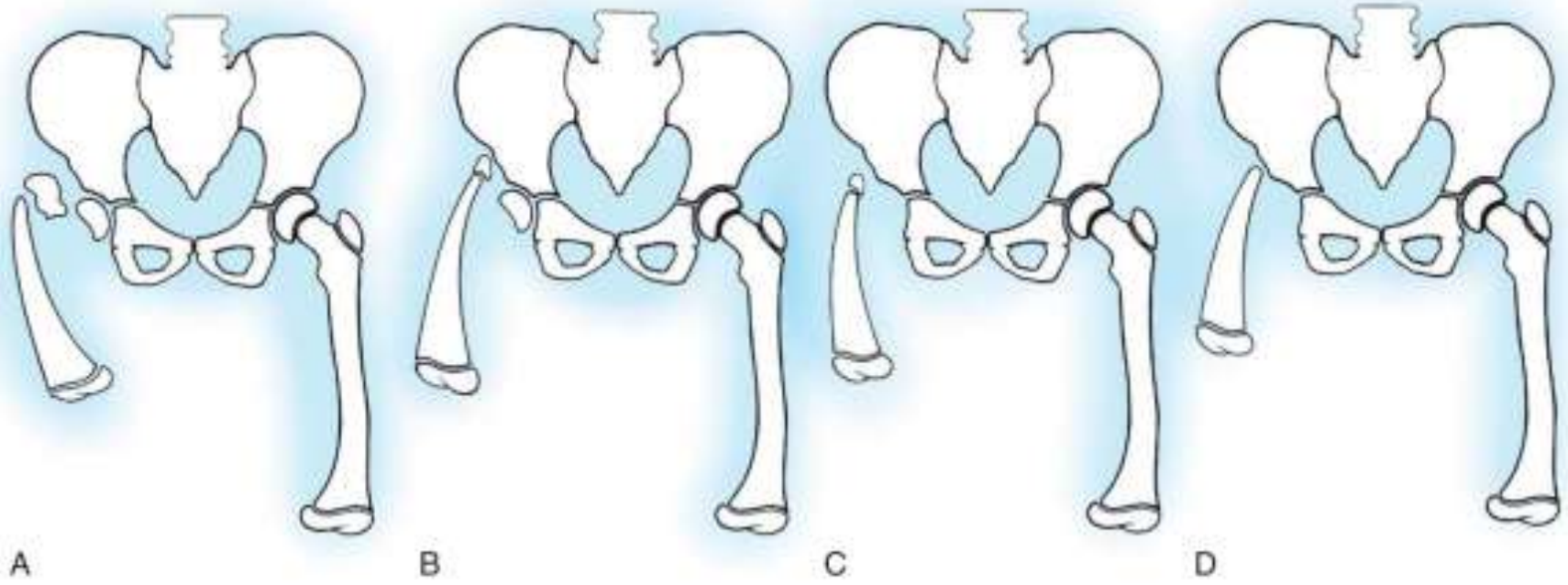


PFFD

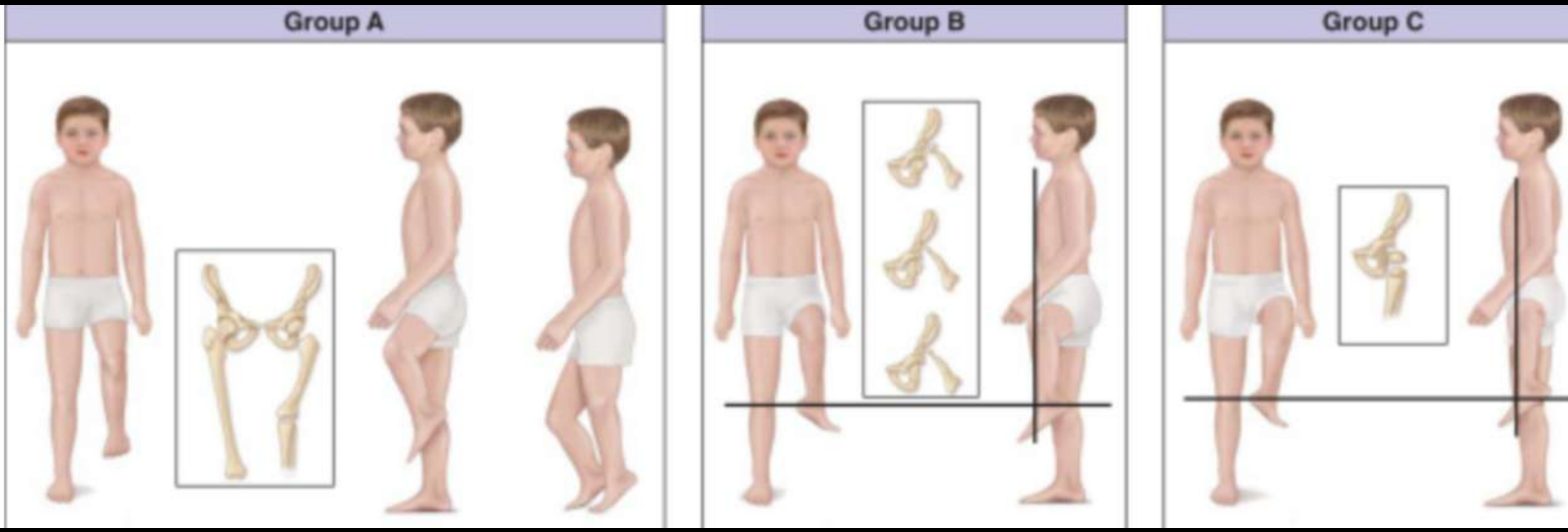
- one of several terms used to describe a deformity in which the femur is shorter than normal and there is apparent discontinuity between the femoral neck and shaft.

OTHER CONGENITAL ANAMOLIES

- clubfoot,
- congenital heart anomalies,
- spinal dysplasia,
- and facial dysplasias



A		Present	Normal	Short	Bony connection between components of femur Femoral head in acetabulum Subtrochanteric varus angulation, often with pseudarthrosis
B		Present	Adequate or moderately dysplastic	Short, usually proximal bony tuft	No osseous connection between head and shaft Femoral head in acetabulum
C		Absent or represented by ossicle	Severely dysplastic	Short, usually proximally tapered	May be osseous connection between shaft and proximal ossicle No articular relation between femur and acetabulum
D		Absent	Absent Obturator foramen enlarged	Short, deformed	None



GROUP A: Congenital short femur

Group B: Aitken A,B,C

Group C: Aitken D

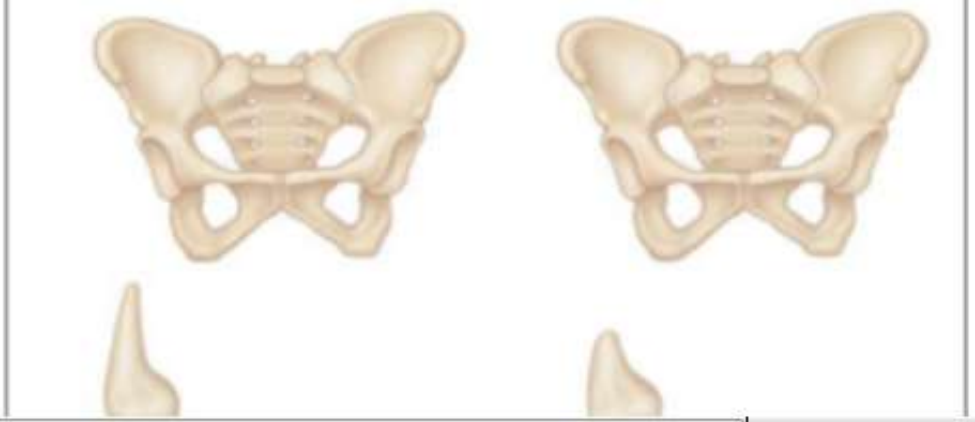
Type 1: Intact Femur, Mobile Hip and Knee

Normal ossification

Delayed ossification

Type 2: Mobile Pseudarthrosis

Type 3: Diaphaseal Deficiency of Femur



Clinical features

- The affected thigh is extremely short, the hip is flexed and abducted, the limb is externally rotated, there is often flexion contracture of the knee, and the foot is usually at the level of the contra-lateral knee.
- Dimple over greater trochanter
- Although the hip abductors and extensors are present, they are foreshortened and unable to function properly because of the abnormal anatomy of the proximal femur.

Clinical features

- In approximately 45% of cases, the patient also has ipsilateral fibular hemimelia of the affected limb, with a short tibia and an equinovalgus deformity of the foot.

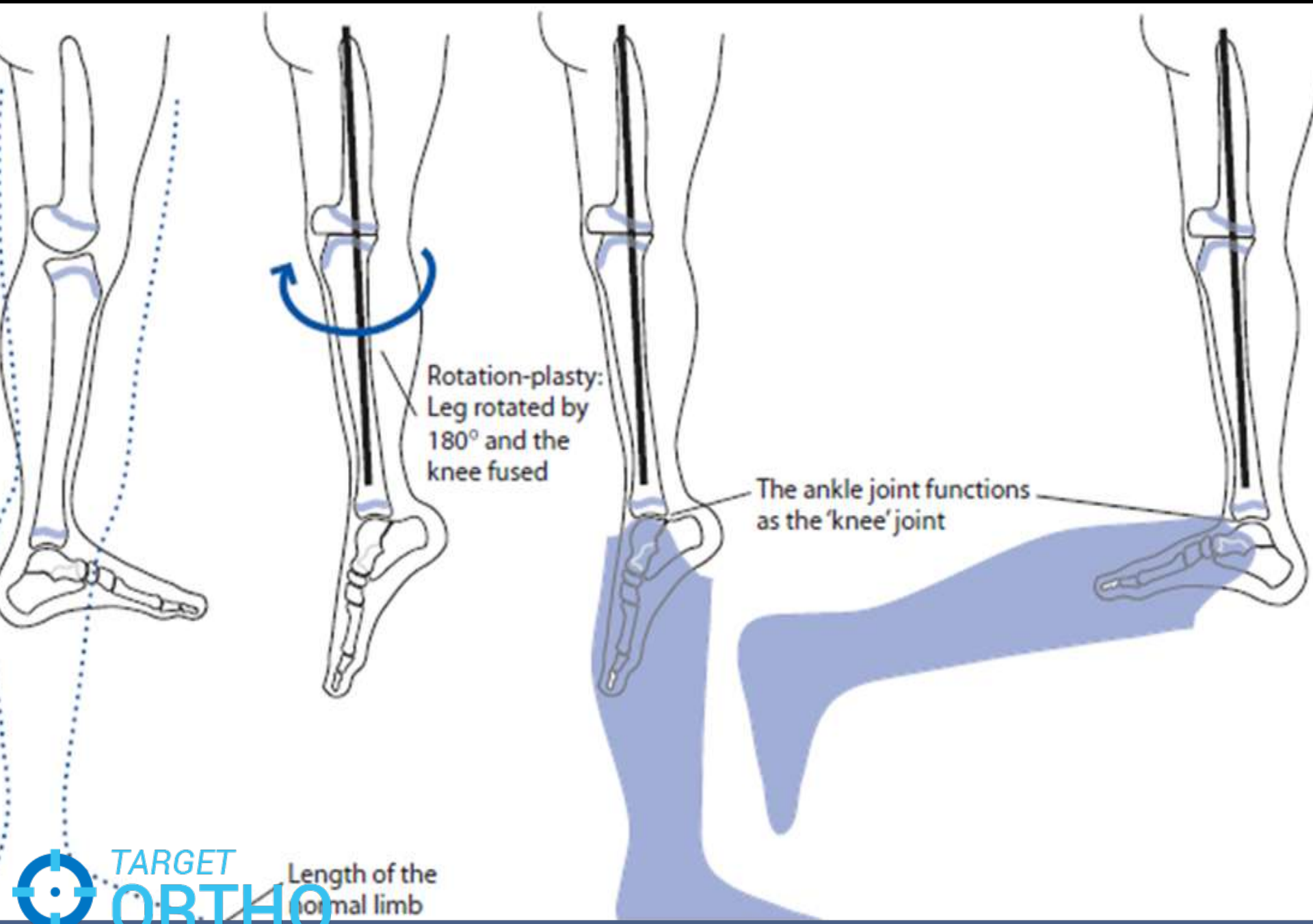
- Lateral rays of the foot may be missing. The disorder may be accurately diagnosed prenatally with sonography.

Clinical features

- Dysplasia of the hip and pelvis
- Genu valgum
- Knee instability
- Shortening
- Associated deficiencies of the leg

Aims of treatment

- To provide reassurance and education for the family
- To enable the child walk and optimize the gait pattern
- Eliminating deformities of the hip and knee
- Making the best use of the available components of the limb
- Achieving limb length equality by adulthood



INDICATIONS				
Well-formed foot + Foot being used for prehensile function (e.g. in children with major upper limb deficiency with no useful hand function) ⇩	Well-formed foot + Foot not performing prehensile function + Parents willing for rotationplasty ⇩	Well-formed foot + Foot not performing prehensile function + Parents not willing for rotationplasty + Parents not willing for amputation ⇩	Well-formed foot + Foot not performing prehensile function + Parents not willing for rotationplasty + Parents willing for foot amputation ⇩	Deformed and non-functional foot ⇩
Retain foot + No knee fusion + Correction of knee deformities interfering with prosthetic fitting + Extension prosthesis accommodating the foot	Retain foot + Van Nes rotationplasty + Below-'knee' prosthesis	Retain foot + Knee fusion + Extension prosthesis accommodating the foot	Syme's amputation + Knee fusion + Above-knee prosthesis	Syme's amputation + Knee fusion + Above-knee prosthesis

INDICATIONS		
Acetabulum and femoral head and neck not formed ⇩	Acetabulum and femoral head and neck formed + No ossification defect in neck of femur + Coxa vara ⇩	Acetabulum and femoral head and neck formed + Ossification defect in neck of femur + Coxa vara ⇩
No intervention	Subtrochanteric valgus osteotomy	Subtrochanteric valgus osteotomy + Screw across neck
TREATMENT		