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- Idiopathic osteonecrosis of the proximal femoral epiphysis in a growing child, leading to alteration in femoral head which tends to flatten and enlarge.
- Onset: 18 months and skeletal maturity (4-12 years)
- Bilateral 10-12%
- Male : female= 4-5:1

- Limp exacerbated by activity and relieved by rest
- Pain : groin, anterior hip, laterally around GT
- H/O antecedent trauma

- Decreased ROM of hip (abduction & internal rotation)
- Restriction is transient early and becomes persistent later
- Flexion / extension less affected
- Thigh Muscle atrophy
- LLD

History

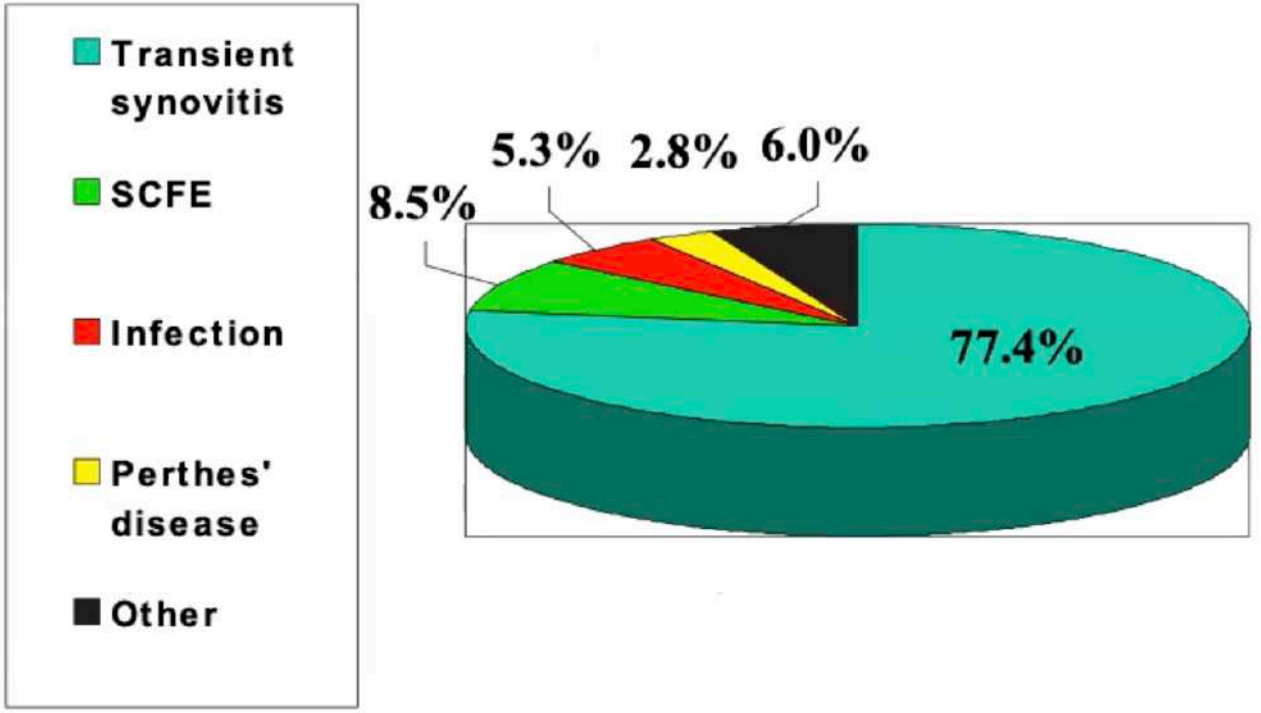
- Jacques Calve
  - 1875 -1954
- Arthur Legg -U
  - 1874 - 1939
- George Perthes -Germany
  - 1869 - 1927

- **Perthes**: called it" youthful variation of adult degenerative arthritis".
- found it self limiting, affecting capital femoral epiphysis
- **Schwarz:** first illustration of blood supply to femoral head.
- **Waldenstrom** : radiographic changes 1909

"upon" "study"

epidemiology

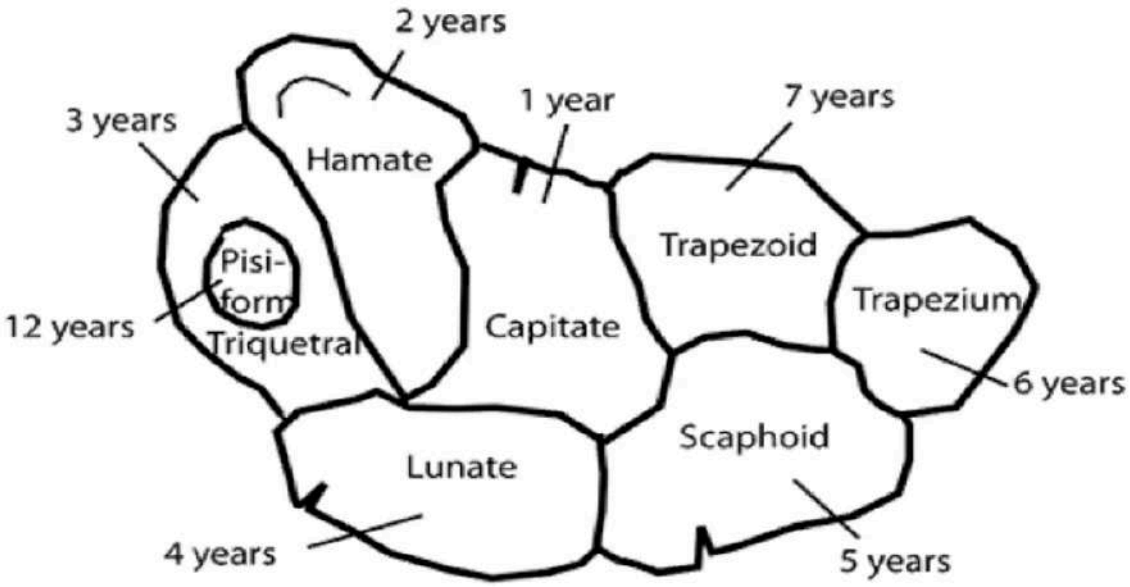
"people"



- Factors that may be etiologic
  a. trauma
  b. susceptible child
  c. hereditary factors (10%)
  d. coagulopathy
  e. hyperactivity
  f. passive smoking
- Unlikely factors
  a. endocrinopathy
  b. urban environment
  c. synovitis

Predisposed child

- **Delay In Bone age** relative to chronologic age. (carpal bone age is 2 years behind)
- Child **diagnosed before 5 years** of age- increased bone age delay
- **Radiological pause** – particular carpal affected more than others (aka **skeletal standstill**)
- **Triquetral and lunate** – MC involved
- **Bilateral cases- trapezoid**



Pathogenesis

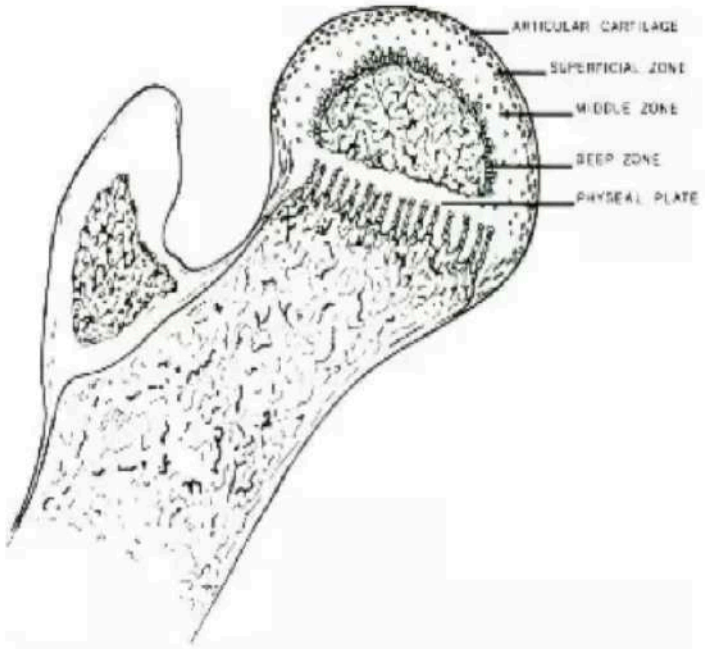


FIGURE 24-3. Proximal femur in a child.

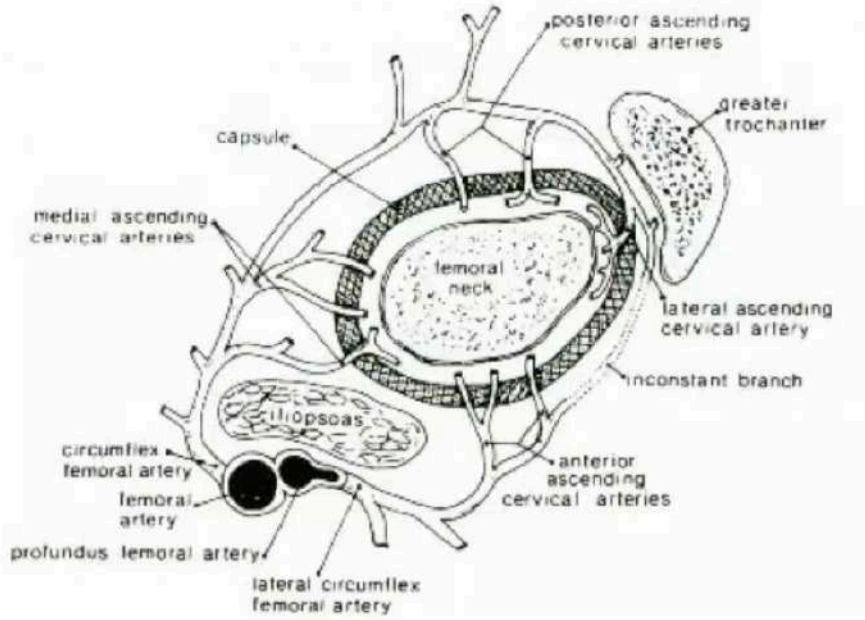
Superficial zone=like adult articular cartilage

-Epiphyseal (middle) cartilage zone= becomes thinner as skeleton matures, epiphyseal bone enlarges

-Deep thin zone= small clusters of cartilage cells that hypertrophy and degenerate

-capillaries from below

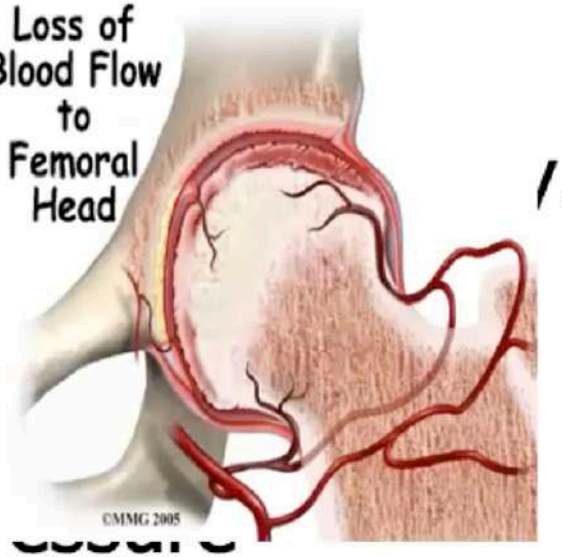
Blood supply

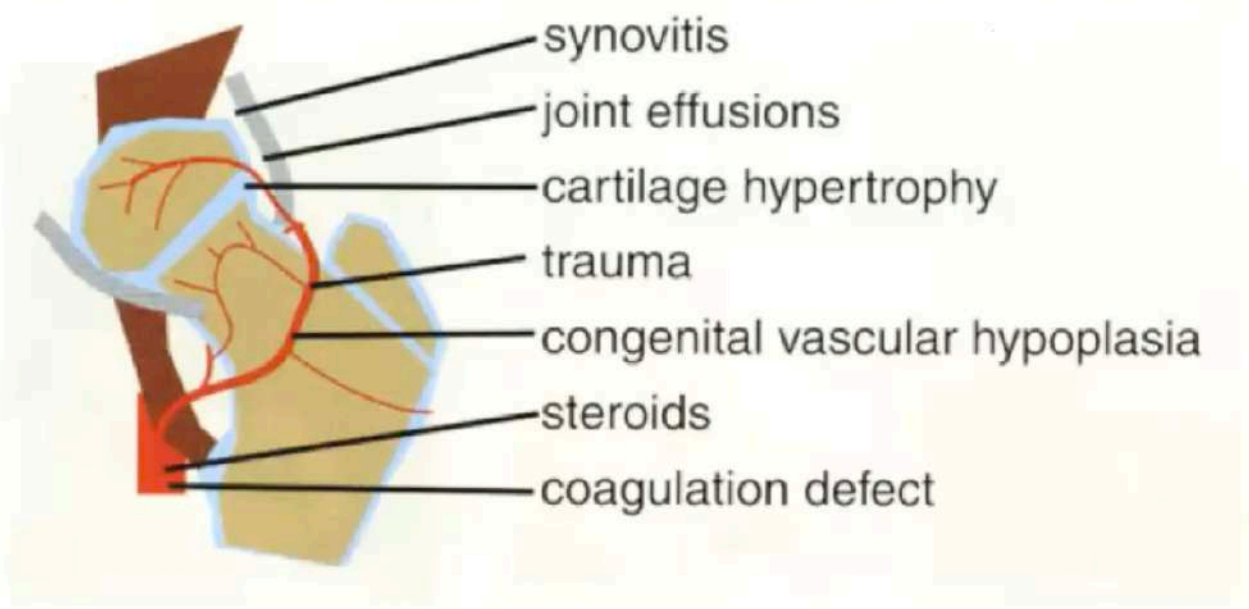


Causes

Exact cause unknown.

- **Proposed theories**.
  - Inherited protein C anc
  - Venous thrombosis
  - Arterial occlusion
  - Raised intra osseous p
  - Excessive femoral antiversion.
  - Synovitis.
  - Generalized skeletal disorder.





Pathophysiology

- The capital femoral epiphysis always is involved. In 15-20% of patients with LCPD, involvement is bilateral.
- The blood supply to the capital femoral epiphysis is interrupted.
- Bone infarction occurs, especially in the subchondral cortical bone, while articular cartilage continues to grow. (Articular cartilage grows because its nutrients come from the synovial fluid.)
- Revascularization occurs, and new bone ossification starts.

Pathophysiology

- At this point, a percentage of patients develop LCPD, while other patients have normal bone growth and development.
- LCPD is present when a subchondral fracture occurs. This is usually the result of normal physical activity, not direct trauma to the area
- Changes to the epiphyseal growth plate occur secondary to the subchondral fracture.

Pathogenesis

Avascular necrosis

↓

Temporary cessation of growth

↓

Revascularization from periphery

↓

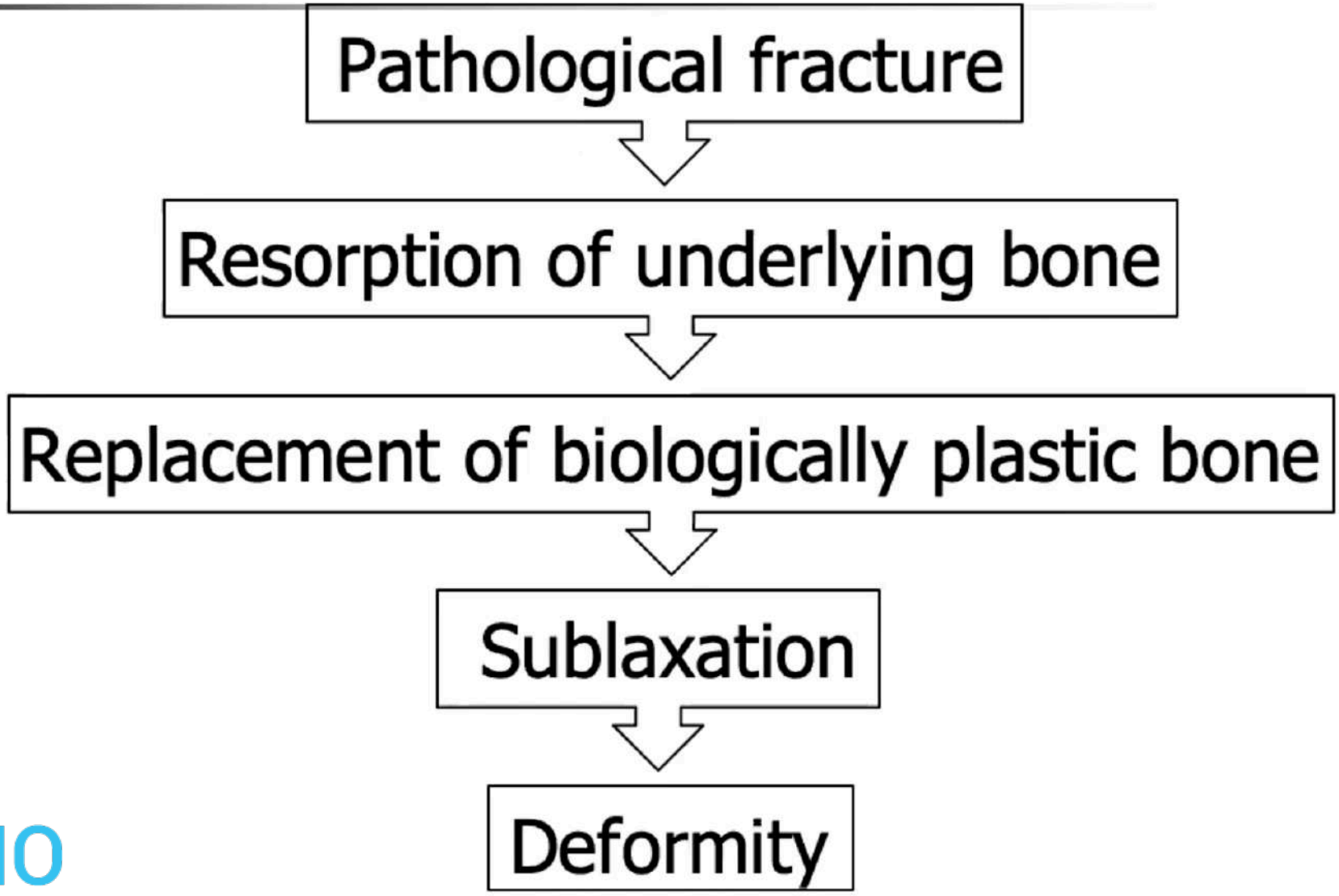
Resumption of ossification and trauma

↓

Contd.....

Pathogenesis

Contd.....



Clinical

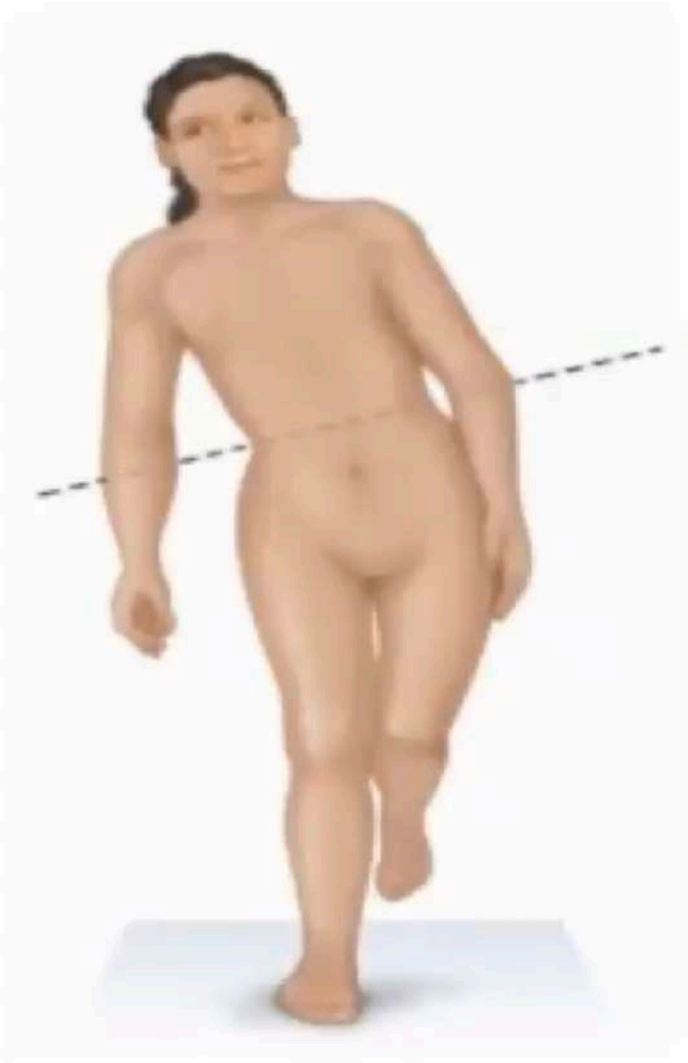
Physical:

- Painful gait
- Decreased range of motion (ROM), particularly with internal rotation and abduction
- Atrophy of thigh muscles secondary to disuse
- Muscle spasm
- Leg length inequality due to collapse

Clinical

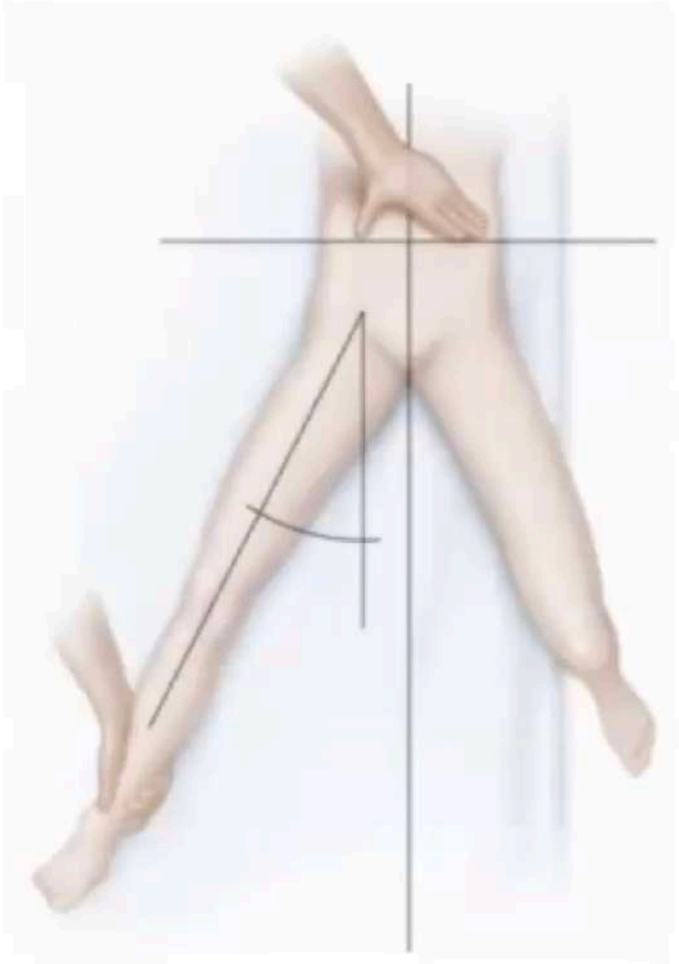
- Short stature: Children with LCPD often have delayed bone age.
- Roll test
  - With patient lying in the supine position, the examiner rolls the hip of the affected extremity into external and internal rotation.
  - This test should invoke guarding or spasm, especially with internal rotation.

Gait : trendelenberg+antalgic



Trendelenberg test +

Hip ROM



Workup

Lab Studies:

- CBC
- Erythrocyte sedimentation rate - May be elevated if infection present

Workup

Imaging Studies:

- Plain x-rays of the hip are extremely useful in establishing the diagnosis.
- Frog leg views of the affected hip are very helpful.
- Plain radiographs have a sensitivity of 97% and a specificity of 78% in the detection of LCPD
- Multiple radiographic classification systems exist, based on the extent of abnormality of the capital femoral epiphysis.
  - Waldenstrom, Catterall, Salter and Thompson, and Herring are the 4 most common classification systems.
  - No agreement has been reached as to the best classification system.

Radiographic stages

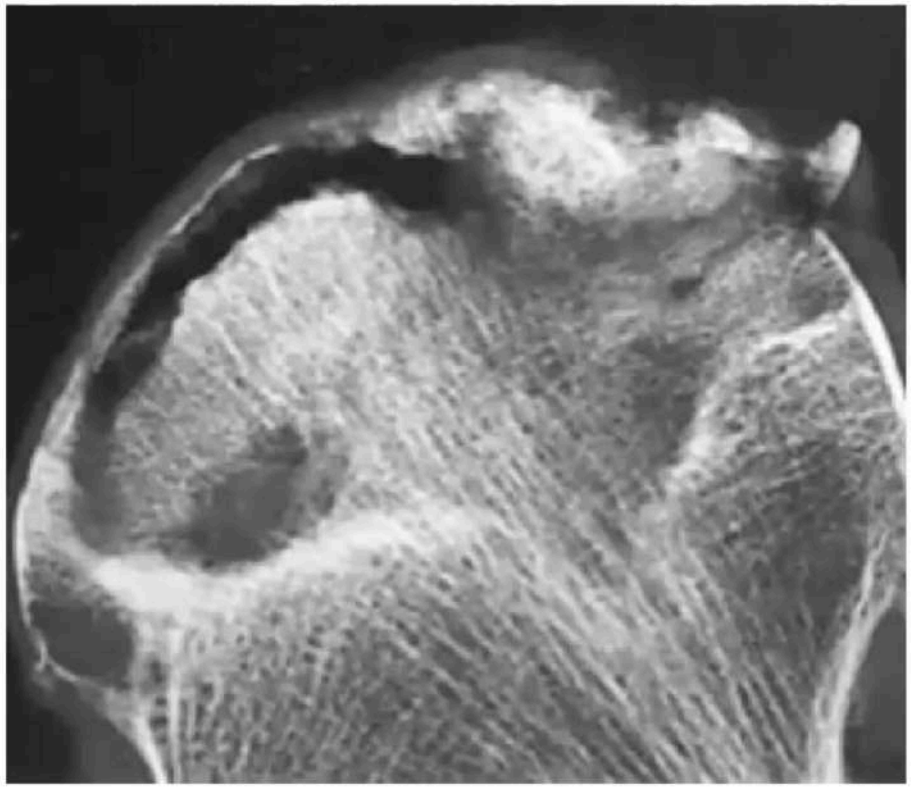
- Four Waldenstrom radiographic stages can be seen by plain x-ray. In sequence, they are as follows:
  - 1) Initial stage
  - 2) Fragmentation stage
  - 3) Reossification stage
  - 4) Healed stage
- Occurs between 2 – 4 years

1.Ischaemia

- The bony part of head dies
- Cartilage still grows
- Increase in joint space
- Waldenstrom's sign
- 6 – 12 months

2.Fragmentation

- Subchondral fracture
- New bone layed down
- Gage's sign
- 12 – 17 months



3.Reossification

- Paraphyseal ossification
- Bony bridges
- Arrests femoral neck growth

4.Remodelling

- Head may collapse
- Abnormal stress
- Coxa magna
- 6 – 24 months

Catterall classification

- **Catterall Group I**: Involvement only of the **anterior** epiphysis (therefore seen only on the frog lateral film)
- **Catterall Group II**: **Central** segment fragmentation and collapse. However the lateral rim is intact and thus protects the central involved area.
- **Catterall Group III**: The **lateral** head is also involved or fragmented and only the medial portion is spared. The loss of lateral support worsens the prognosis.
- **Catterall Group IV**: The **entire head** is involved.

Catterall's classification has a significant inter and intra observer error.

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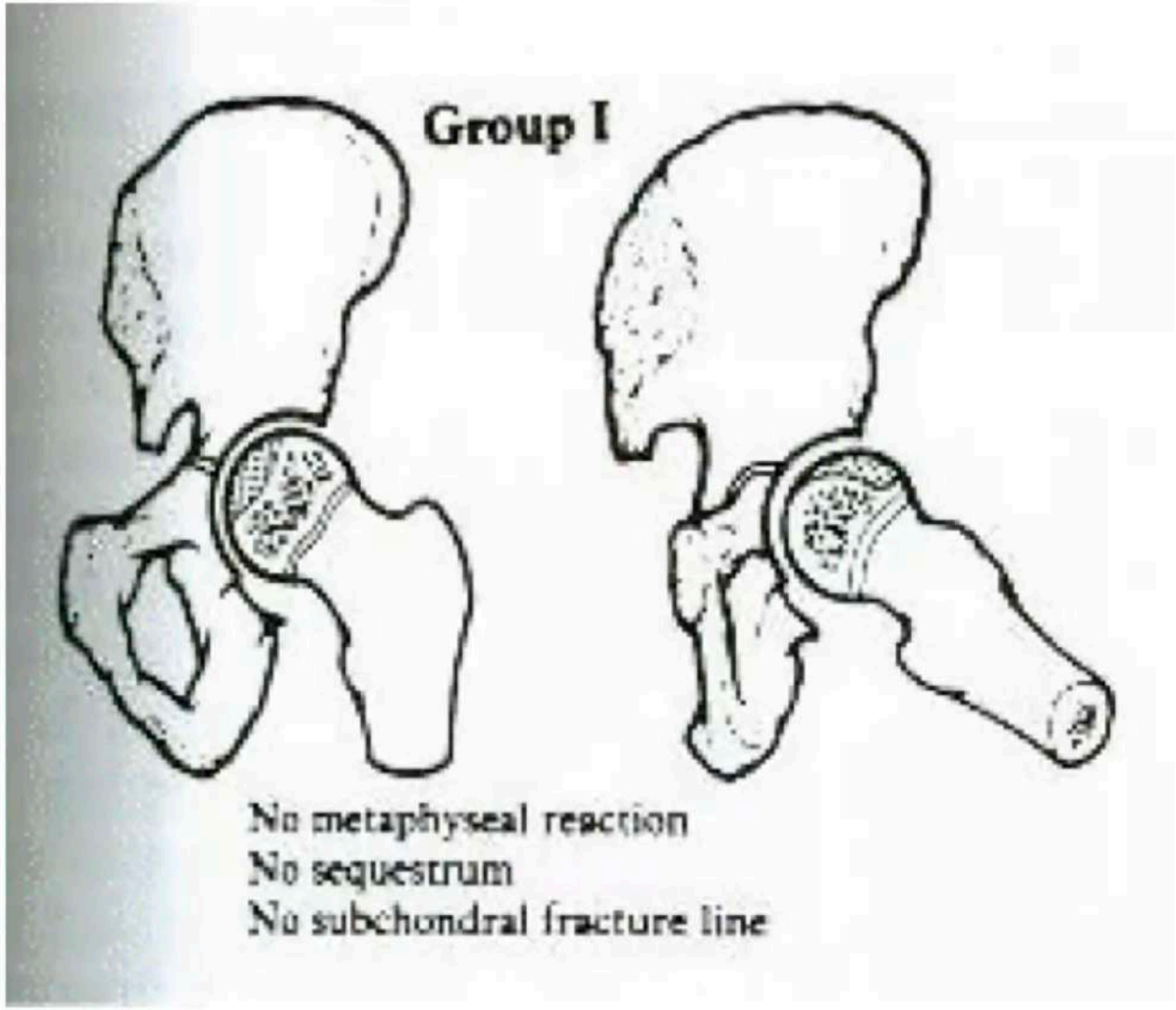
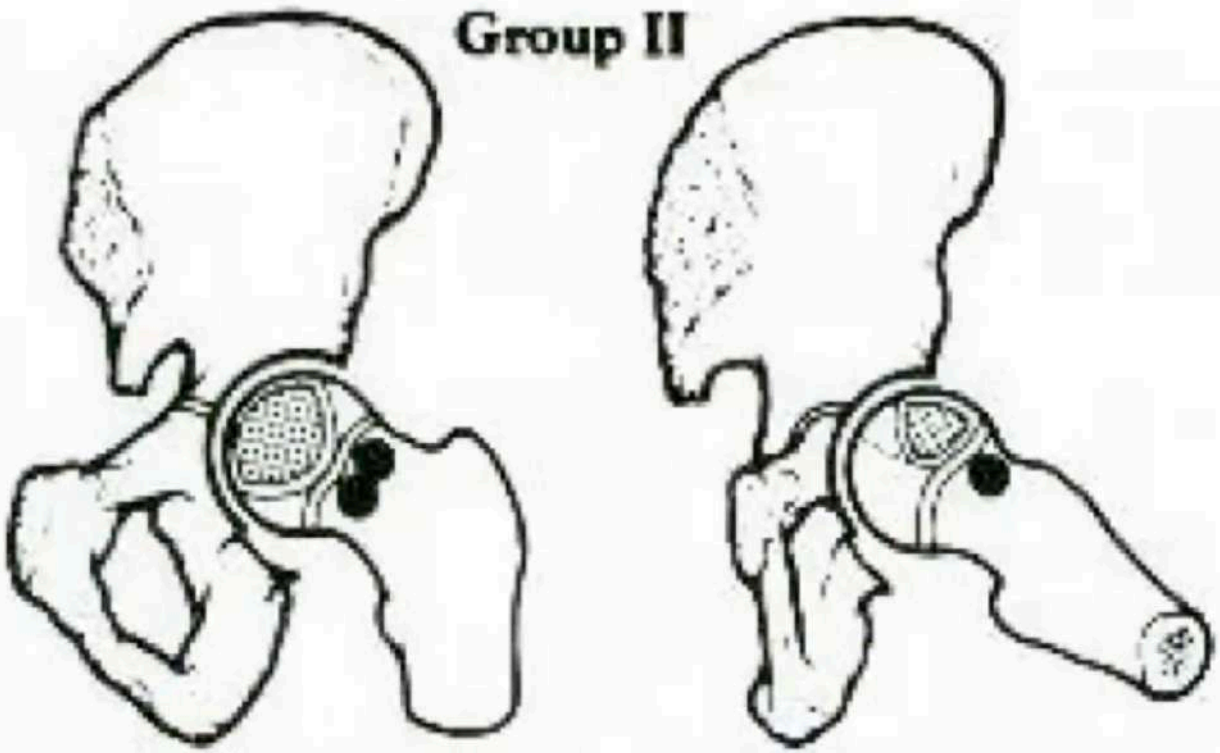
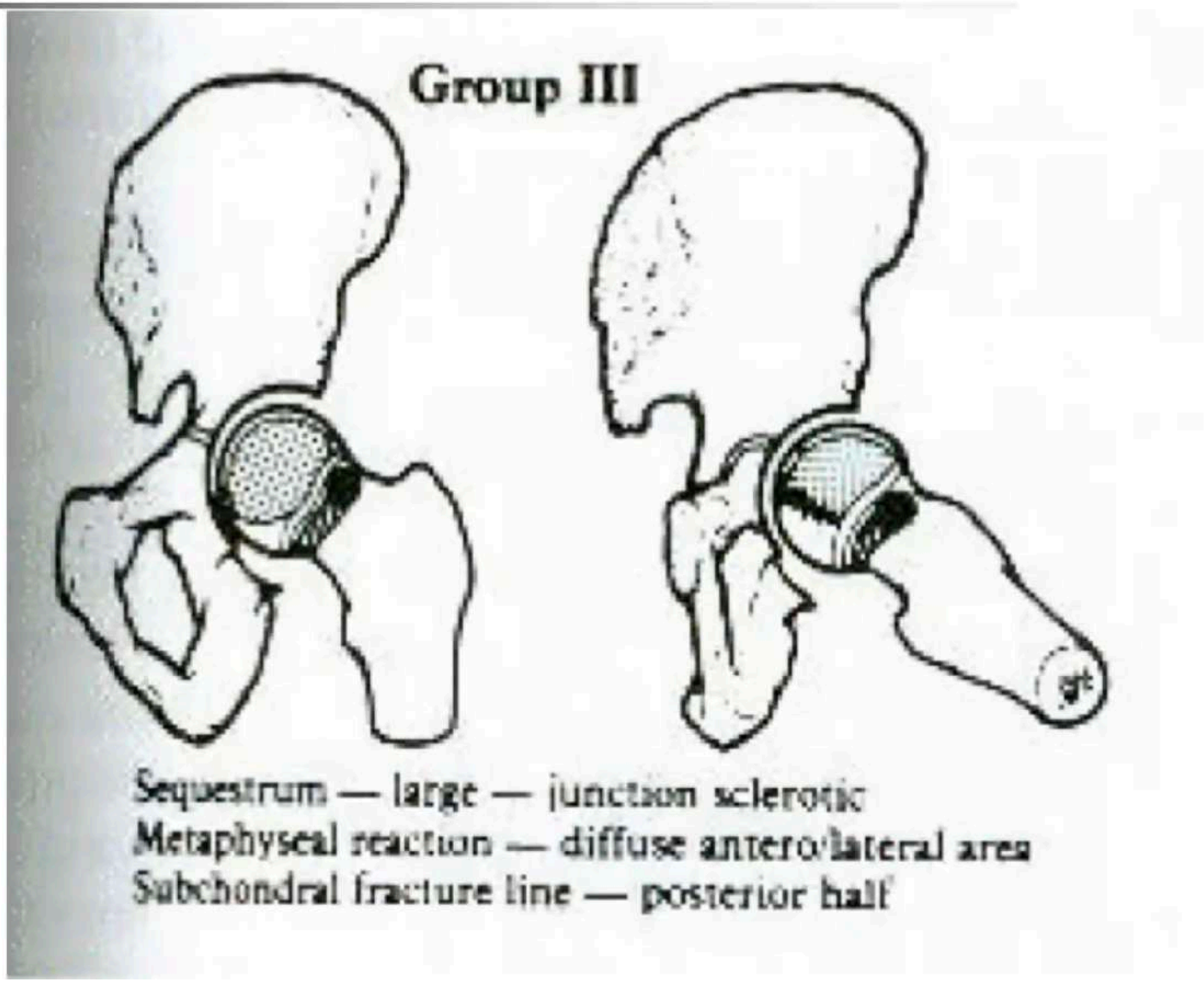


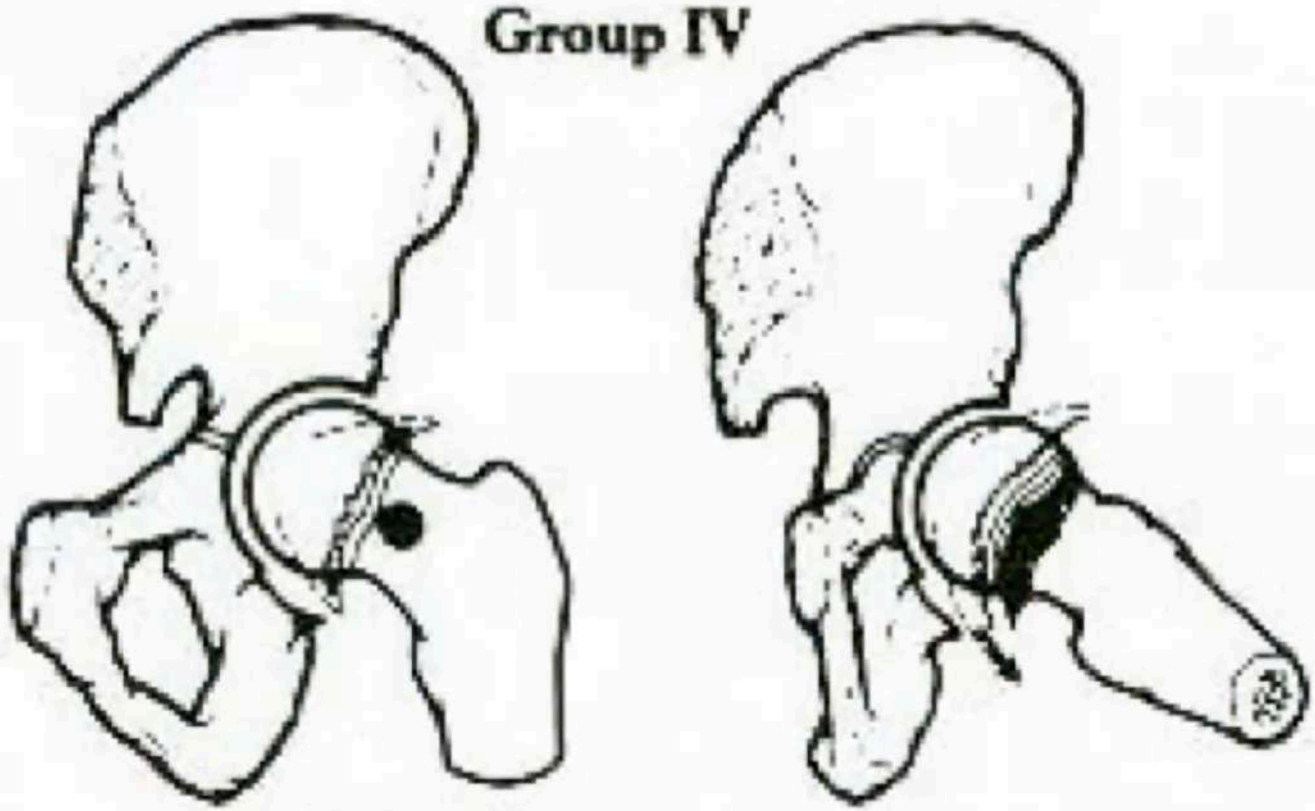
Catterall classification



Catterall classification



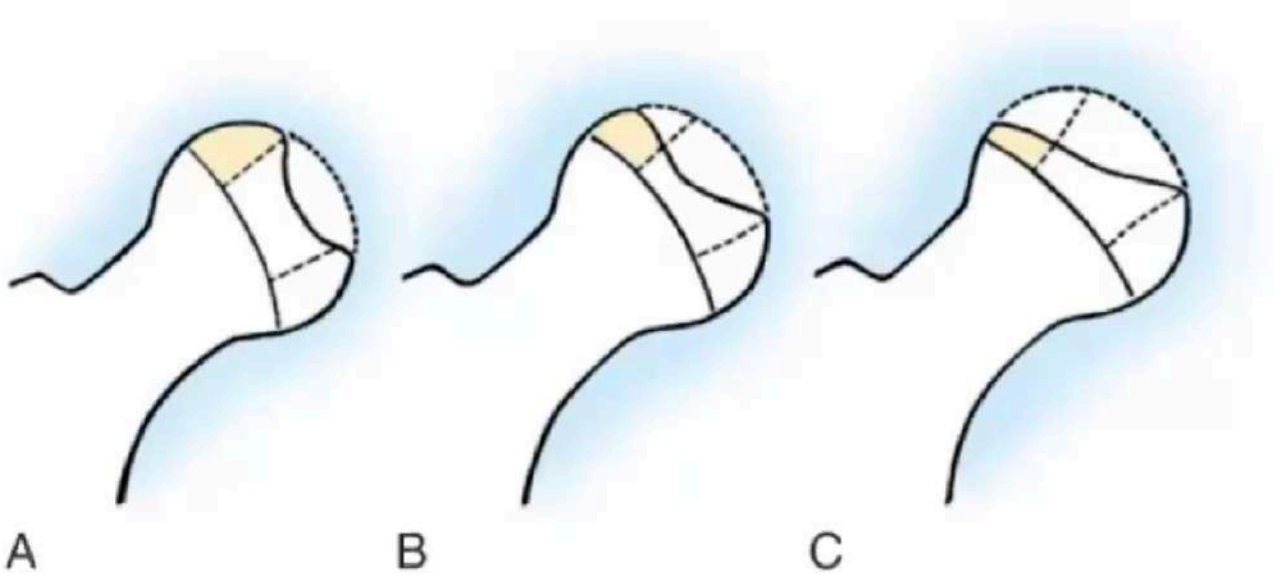
Catterall classification



Whole head involvement
Metaphyseal reaction — central or diffuse
Posterior remodelling

Lateral Pillar Classification (Herring Classification)

- 3 groups:
  - A) no lateral pillar involvment
  - B) >50% lat height intact
  - C) <50% lat height intact

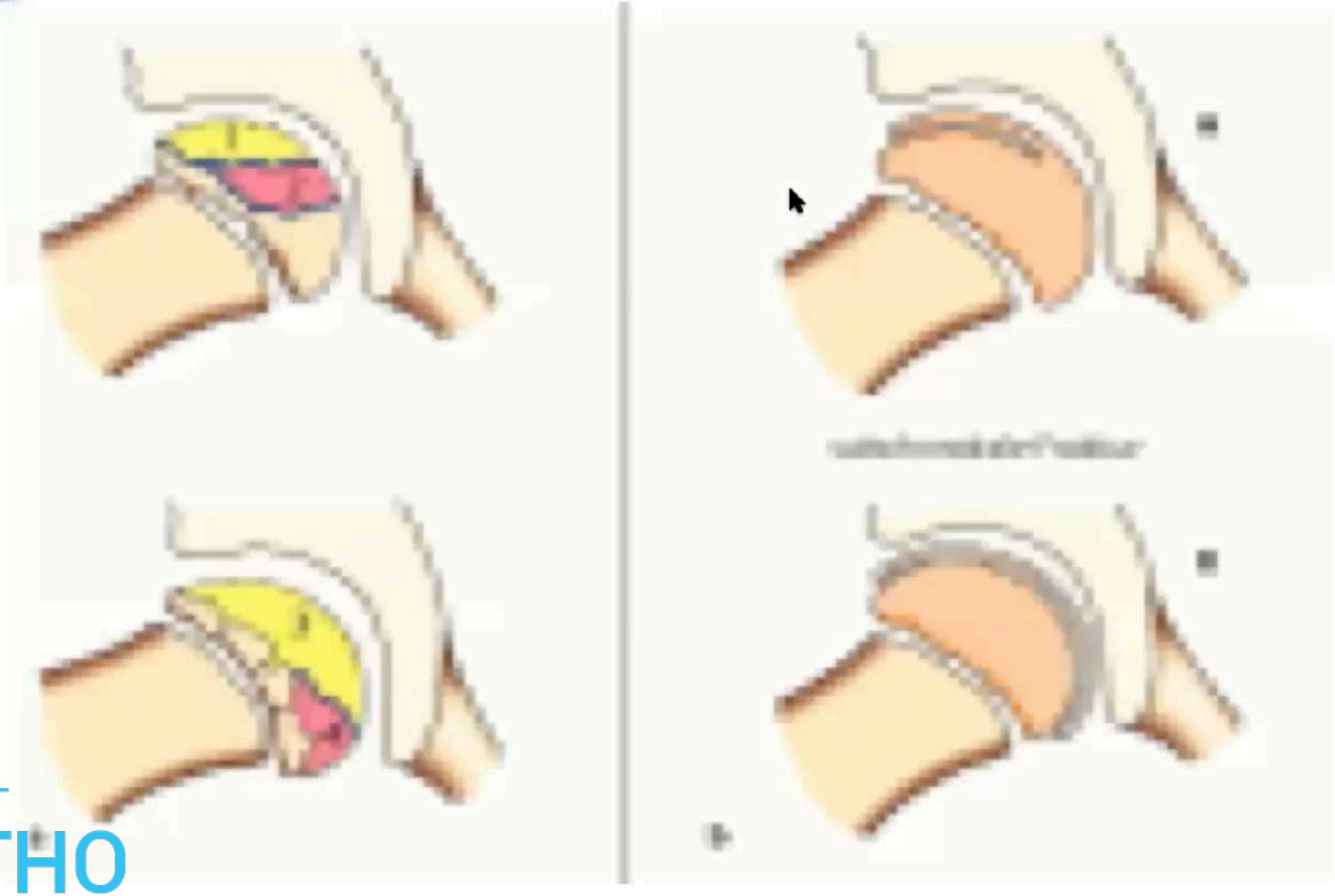


Salter and Thompson Classification

- Salter and Thompson recognized that Catterall's first two groups and second two groups were distinct and therefore proposed a two part classification.
- **Salter & Thompson Group A**: Less than 1/2 head involved.
- **Salter & Thompson Group B**: More than 1/2 head involved.
- Again the main difference between these two groups is the integrity of the lateral pillar.

Subchondral fracture

Salter and Thompson Classification



Stulberg Classification

- **Class 1**
  - completely normal
- **Class 2**
  - Spherical (< 2mm). Coxa Magna. Short Neck. Increased Sharp angle
- **Class 3**
  - Elliptical > 2mm, but not flat
- **Class 4**
  - Flat femoral head, flat acetabulum
- **Class 5**
  - Flat femoral head, round acetabulum

Catterall's Head at risk signs

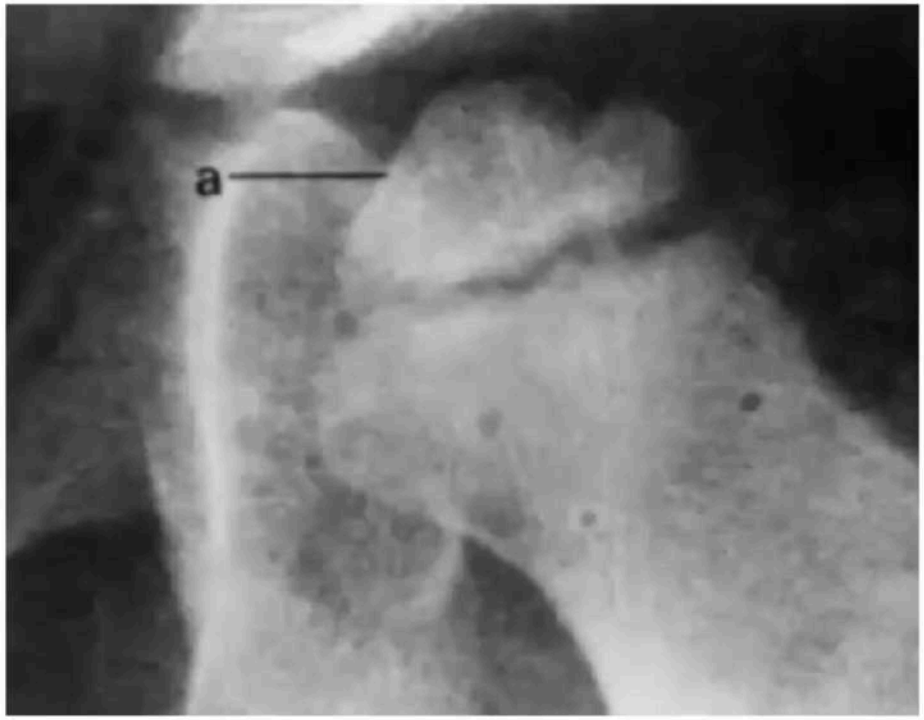
- Latral subluxation of the head
- Whole of the head involved
- Calcification lateral to epiphysis
- Metaphyseal cysts
- Gage's sign- 'V' shaped lucency in the lateral epiphysis.
- Horizontal physis

corelation

Stage	Clinical Findings	Radiographic Changes
Increased density stage	Limp and pain variable, often mild and intermittent	Increased density of femoral head, with/without subchondral fracture
Fragmentation stage	Pain and limp may worsen; may lose range of motion	Head shows fragmentation, may lateralize and flatten
Reossification stage	Limp and pain gradually resolve, range of motion improves	Femoral head gradually reossifies; flattening of head may improve
Healed stage	Occasional limp; occasional locking, popping	May develop osteochondrotic lesion

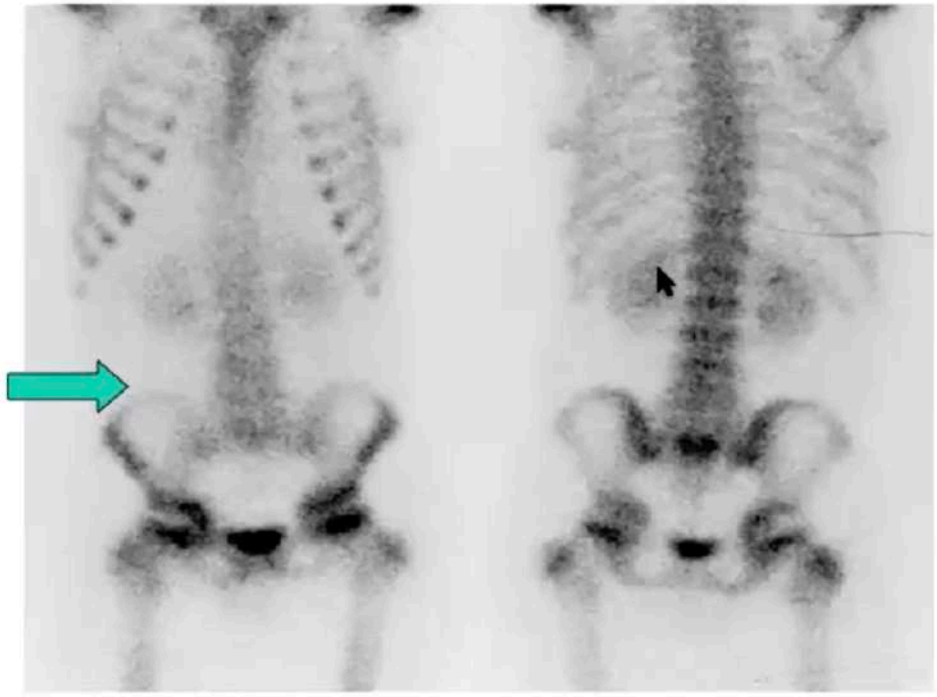
Sagging rope sign

Dimple sign



Workup

- Technetium 99 bone scan - Helpful in delineating the extent of avascular changes before they are evident on plain radiographs.
  - The sensitivity of radionuclide scanning in the diagnosis of LPD is 98%, and the specificity is 95%.
- Dynamic arthrography - Assesses sphericity of the head of the femur.



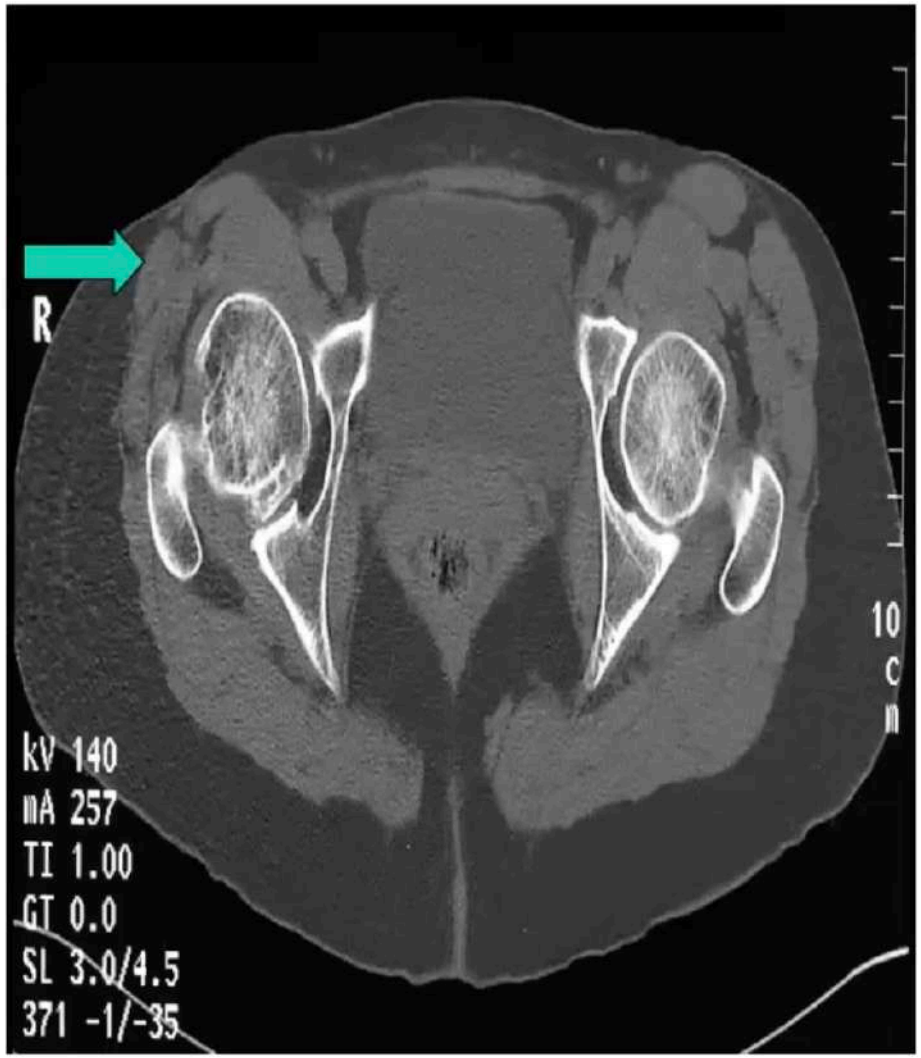
Ultrasonography in transient synovitis and early Perthes' disease

- Ultrasonography may provide significant diagnostic clues to differentiate early Perthes' from transient synovitis.

T Futami, Y Kasahara, S Suzuki, S Ushikubo, and T Tsuchiya
Journal of Bone and Joint Surgery - British Volume, Vol 73-B, Issue 4, 635-639

CT Scan

- Staging determined by using plain radiographic findings is upgraded in 30% of patients.
- Not as sensitive as nuclear medicine or MRI.
- CT may be used for follow-up imaging in patients with LPD.



MRI

- It allows more precise localization of involvement than conventional radiography.
- MRI is preferred for evaluating the position, form, and size of the femoral head and surrounding soft tissues.
- MRI is as sensitive as isotopic bone scanning.

Differential diagnosis

- Other causes of AVN
  .sickle cell disease
  .hemoglobinopathies
  .thalassemia
  .steroid
  .traumatic hip dislocation
  . DDH treatment
- Epiphyseal dysplasia
  .MED
  .SED
  .mucopolysacch.
  .hypothyroidism

- Osteochondromatosis
- Metachromatosis
- Schwartz-jampel syndrome
- Trichorhinophalangeal syndrome
- Marteaux-lamy syndrome

Outcome variables

- Age
- Extent of involvement
- Duration
- Remodeling potential
- Premature physeal closure
- Type of treatment
- Stage of disease at treatment.

PROGNOSIS

- 60% of kids do well without Rx
- **AGE** is key prognostic factor:
  - <6y= good outcome regardless of Rx
  - 6-8y= not always good results with just containment
  - >9y= containment option is questionable, poorer prognosis, significant residual defect

Poor Prognosis

- Sex
- Bone age
- Uncovering of Femur head
- Percentage involvement
- Adduction contracture
- Weight

Treatment

- **Goals of treatment**
  - Achieve and maintain ROM
  - Relieve weight bearing
  - Containment of the femoral epiphysis within the confines of the acetabulum
  - Traction

Treatment

- Non Operative
  - Symptomatic
  - Conservative
- Operative

Non-operative Treatment

- Improve ROM 1st
- Bracing:
  - Removable abduction orthosis
  - Pietrie casts
  - Hips abducted and internally rotated
- Wean from brace when improved x-ray healing signs

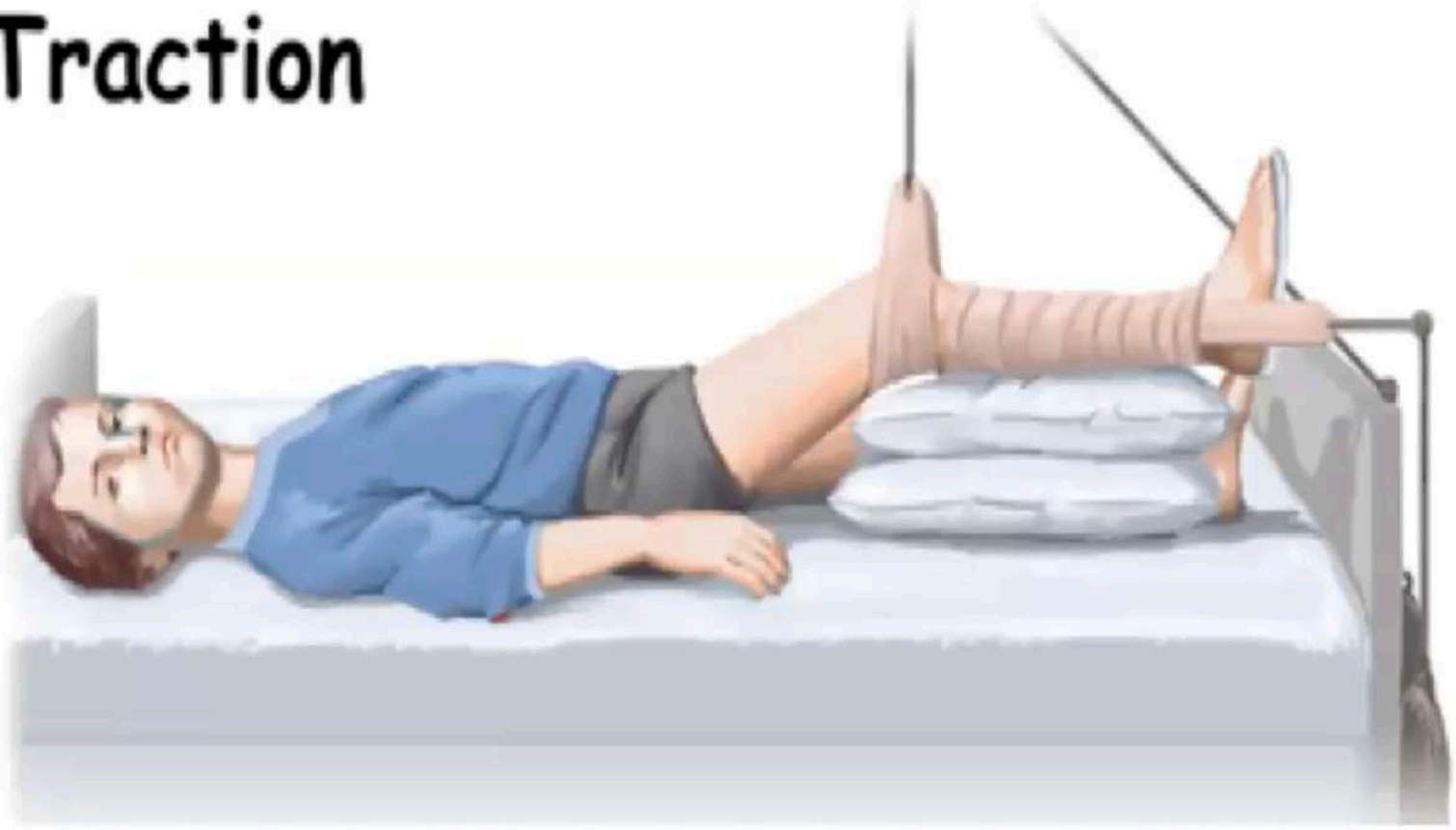
Medications

- Medical treatment does not stop or reverse the bony changes.
- Appropriate analgesic medication should be given.
- *Nonsteroidal anti-inflammatory drugs*
  - **Ibuprofen**
  - Adult dose: 200-400 mg PO q4-6h; not to exceed 3.2 g/d.
  - Pediatric dose:

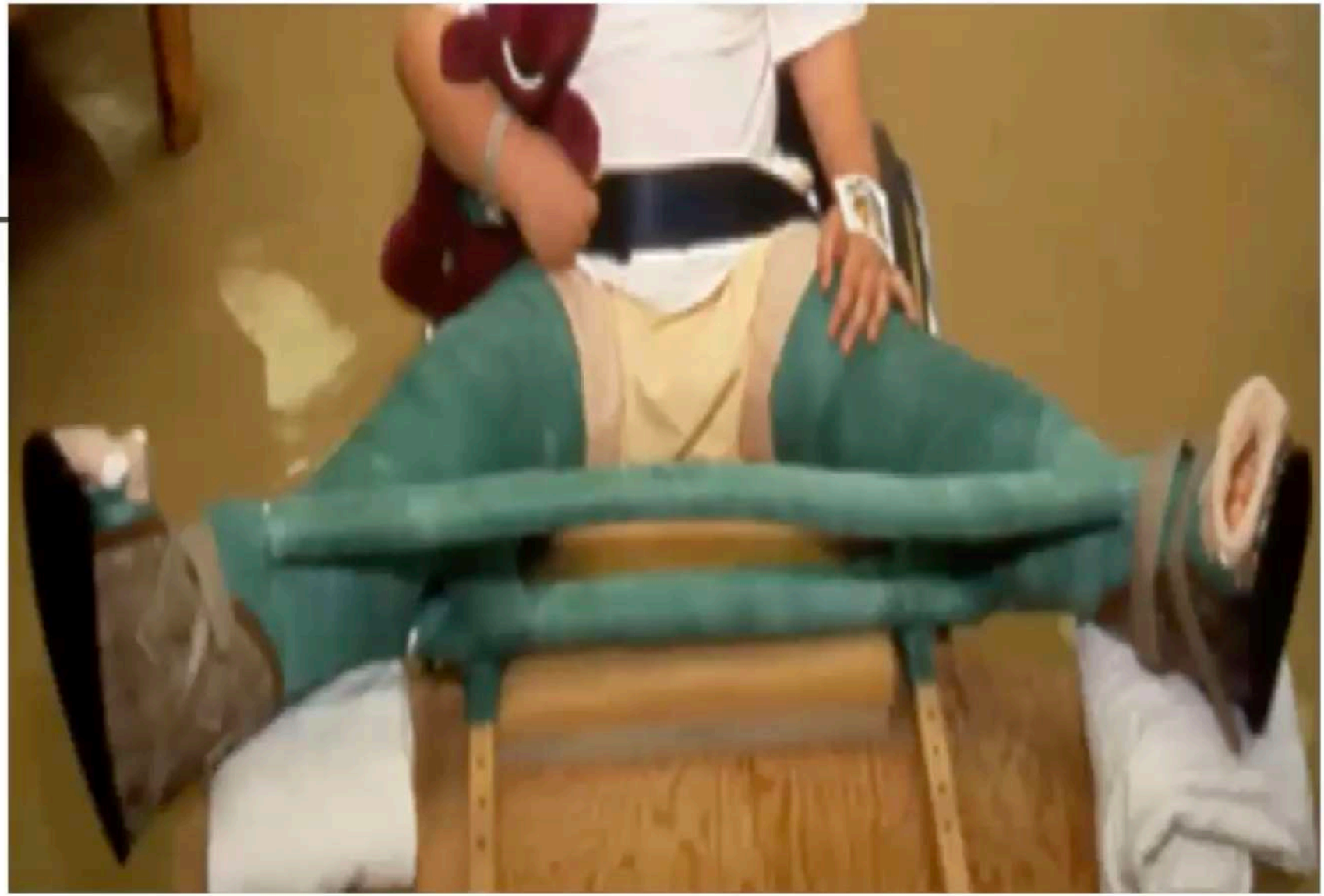
    6 months to 12 years: 20-40 mg/kg/d PO divided tid or qid; start at lower end of dosing range and titrate upward; not to exceed 2.4 g/d
    >12 years: Administer as in adults.

Non surgical containment

Traction



Scotish Rite abduction brace



Japenes modification of petrie abduction cast

Non-operative Treatment

- Check serial radiographs
  - 3-4 months with ROM testing
- Continue bracing until:
  - Lateral column ossifies
  - Sclerotic areas in epiphysis gone
- Cast/brace uninvolved side

Operative Treatment

- If non-op Rx cannot maintain containment
- Surgically ideal pt:
  - 6-9yo
  - Catterral II-III
  - Good ROM
  - <12months
  - In collapsing phase

Surgical Treatment

- Surgical options:
  - Tenotomy
  - Varus derotation femoral osteotomy
  - Acetabular (innominate) osteotomy to cover head

    -salter, pemberton, chiari, staheli(shelf)
  - Excise lat extruding head portion to stop hinging abduction
  - Recostructive surgeries.
  - Arthrodiastasis

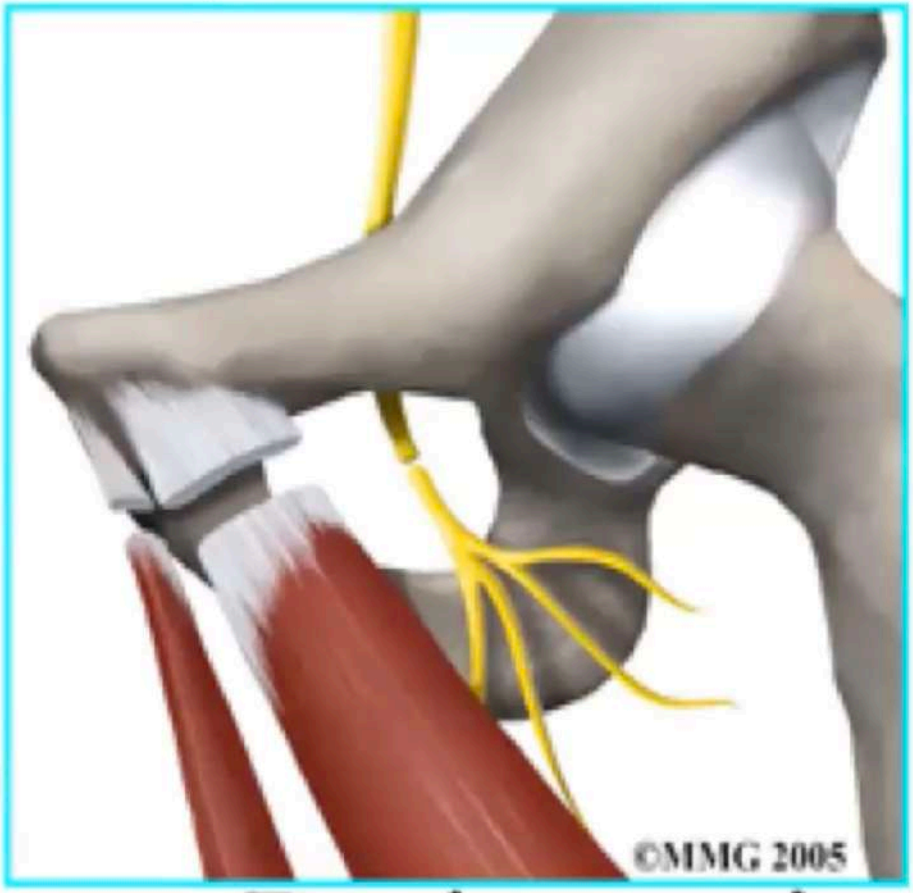
Tenotomy

Incision

Obturator nerve

Gracilis muscle

Adductor longus muscle

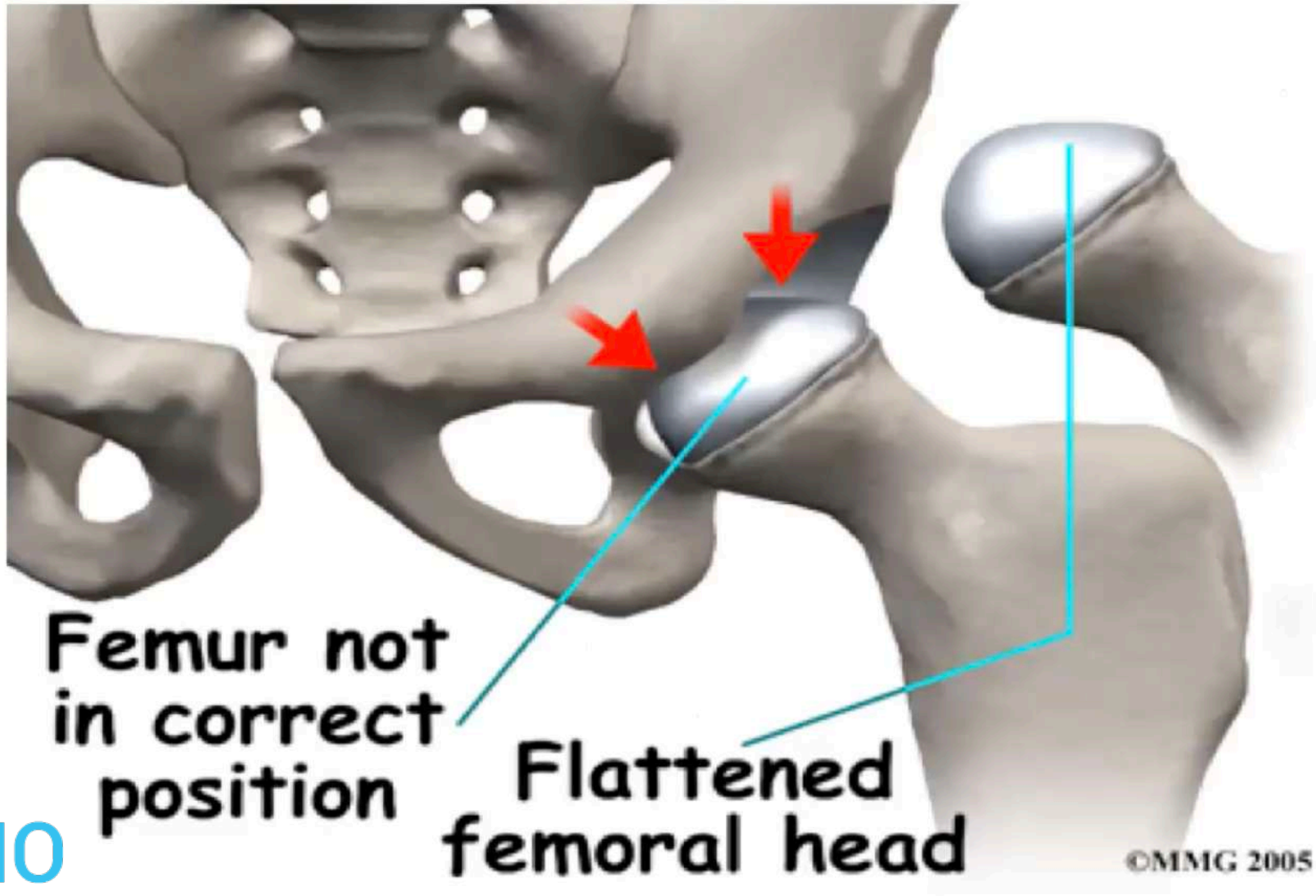


Tendons and nerve divided

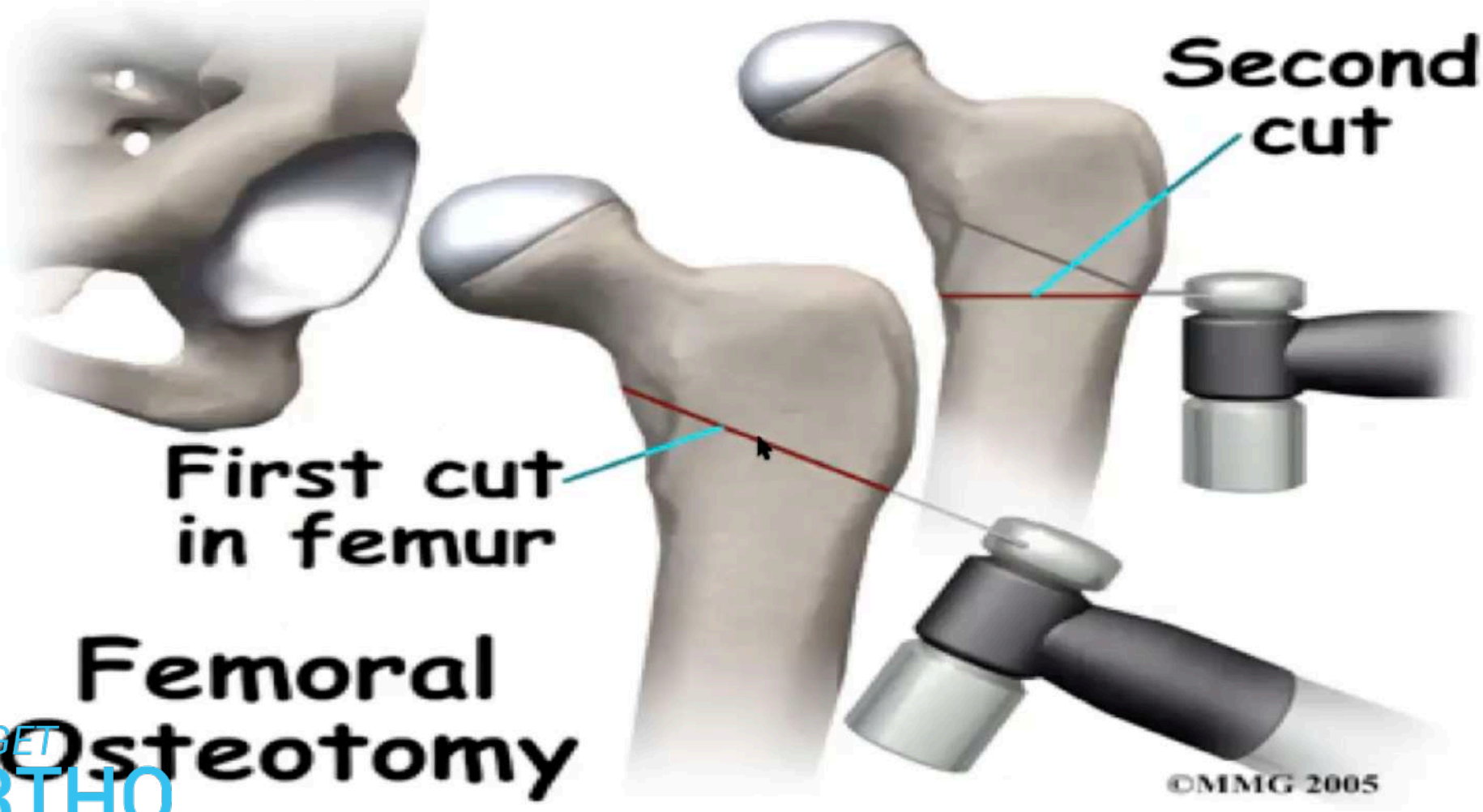
Rational behind "containment"

- Salter has demonstrated the" ***biologic plasticity"*** of the femoral head in pigs following a vascular insult.
- Containment of the head within the acetabulum is reported to encourage spherical remodelling during the reossification and subsequent phases.
- However if there is total head involvement and the lateral pillar collapses then the effect of containment is probably less.
- Therefore it seems that the extent of involvement of the head is the critical factor and containment simply optimizes the situation.

Surgical containment



Medial closing wedge osteotomy



Varus Osteotomy

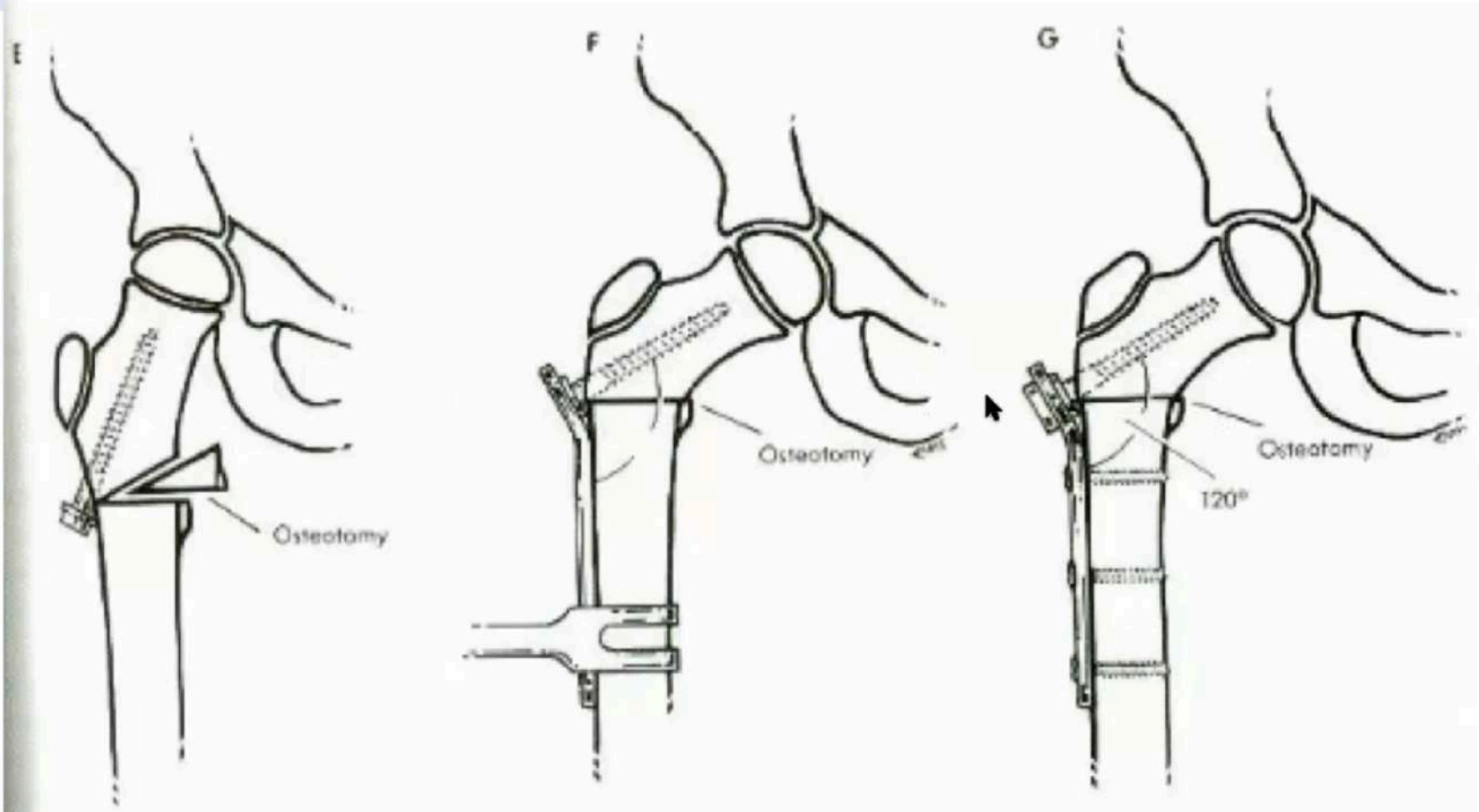
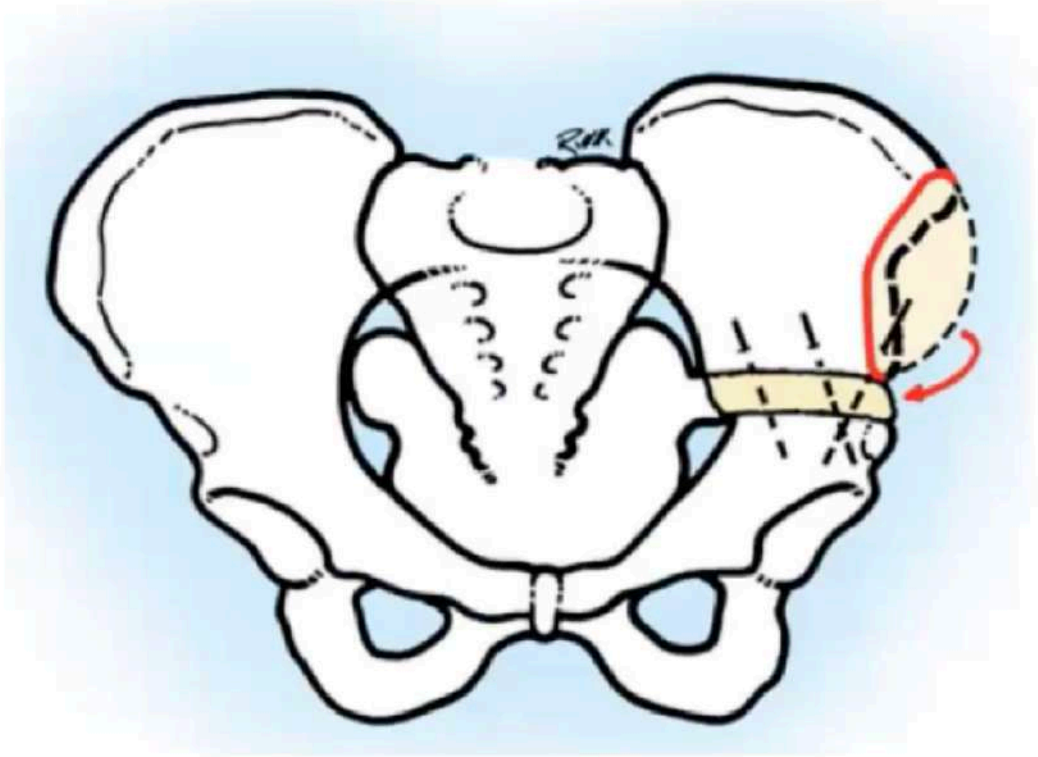


Fig. 24-24, cont'd E, Removal of wedge. F, Attachment of side plate. G, Final attachment of side plate.

Varus Osteotomy

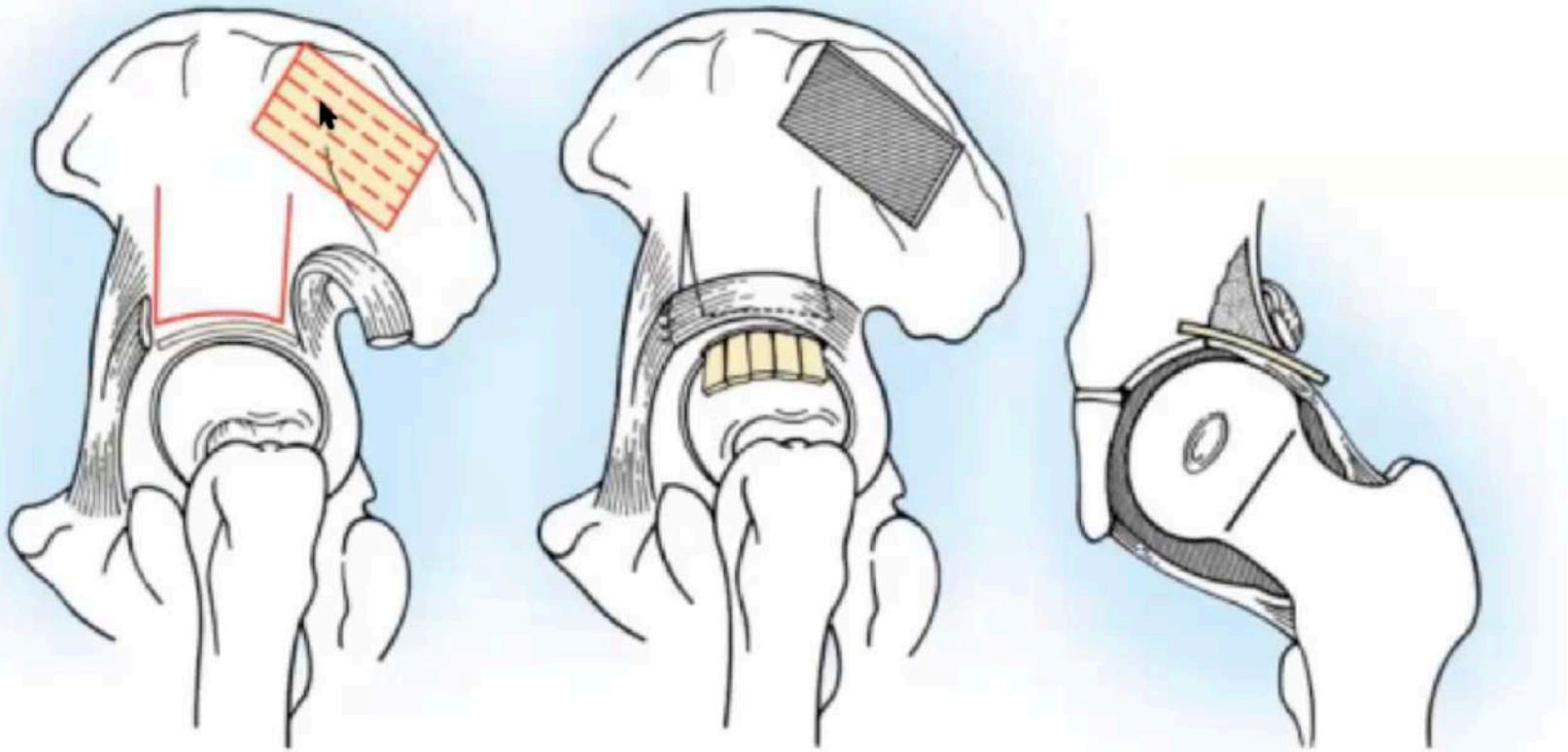
Varus osteotomy can also be done by lateral opening wedge osteotomy

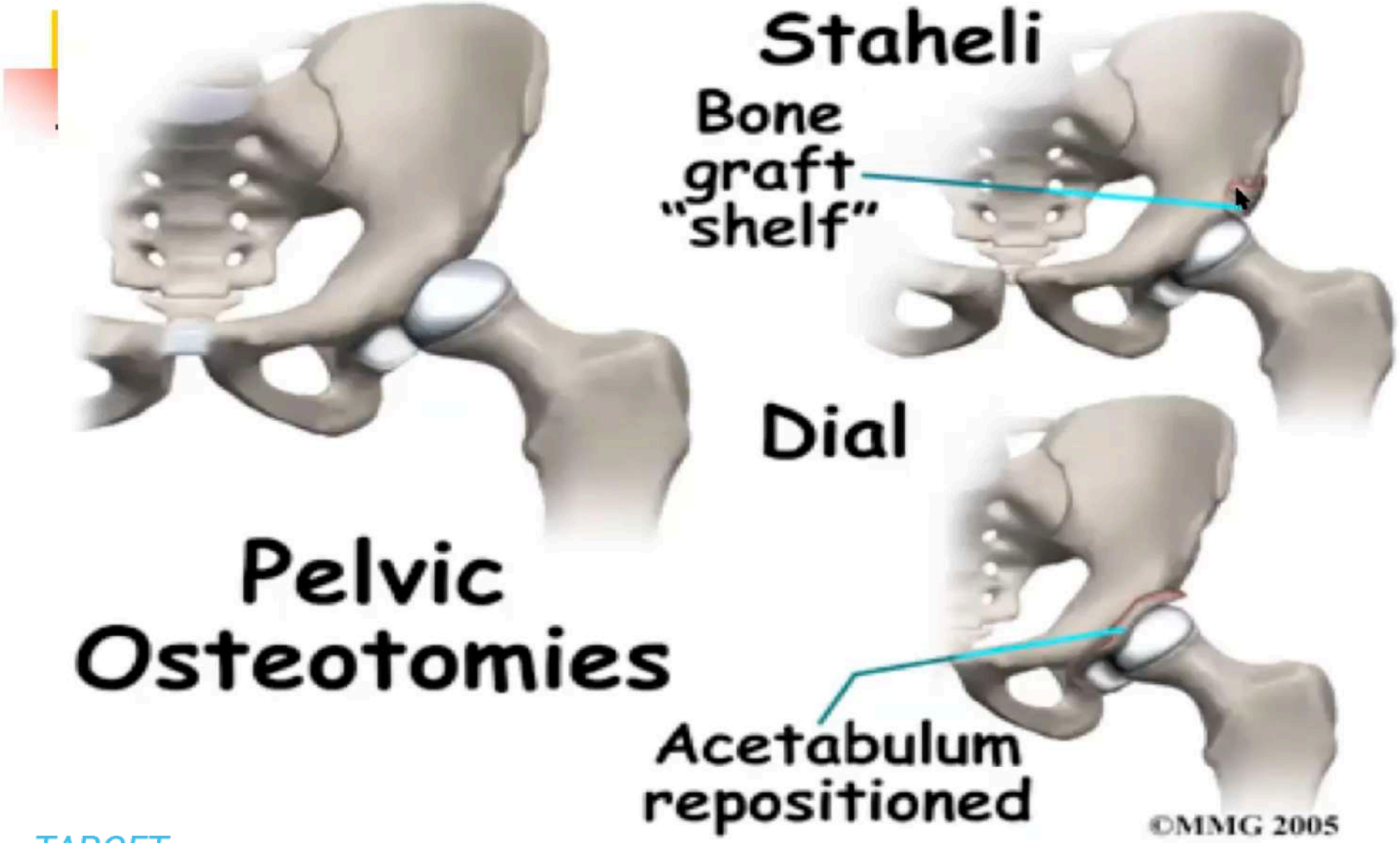
Innominate osteotomy

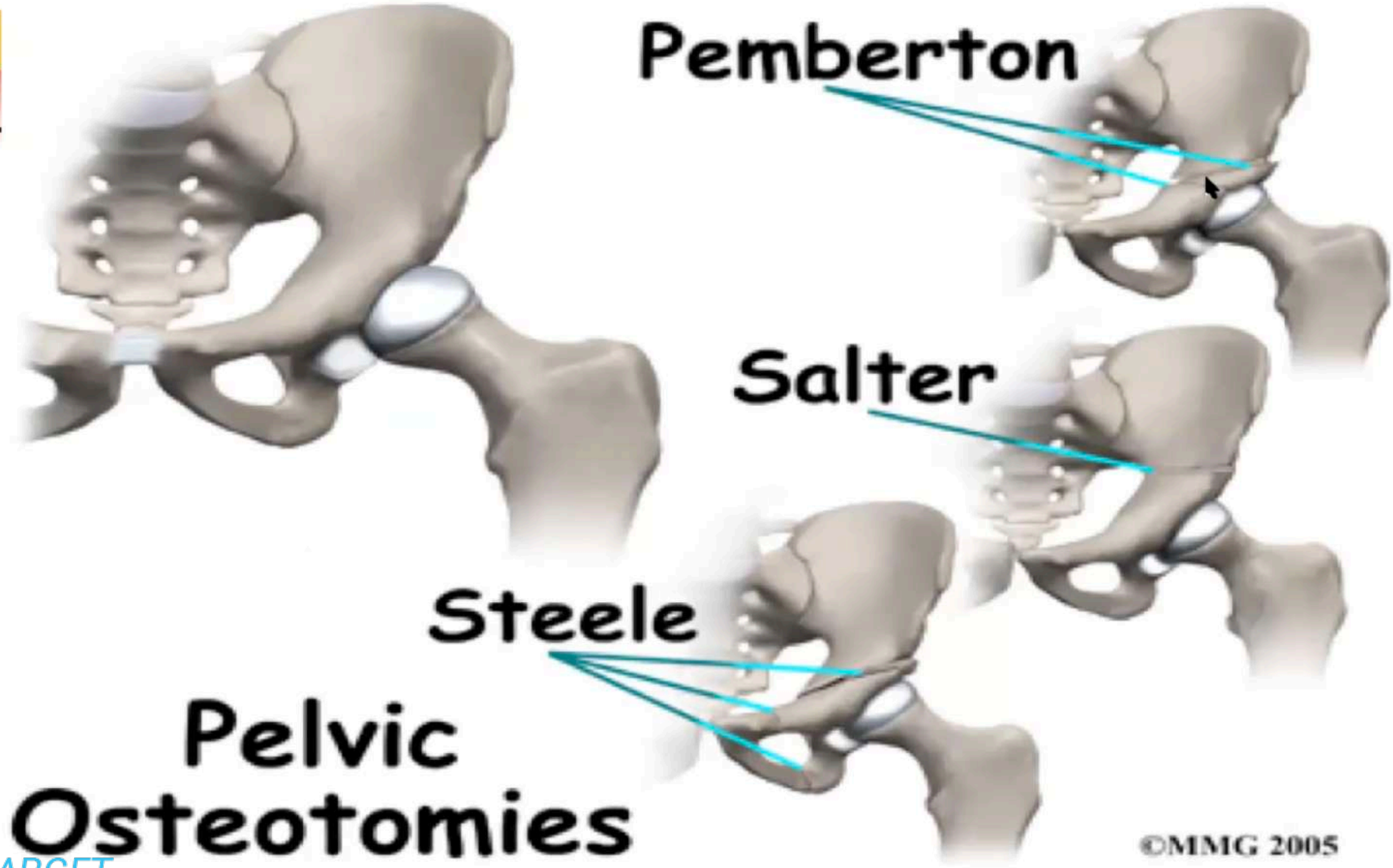


Innominate osteotomy using quadrangular graft

Lateral shelf acetabuloplasty

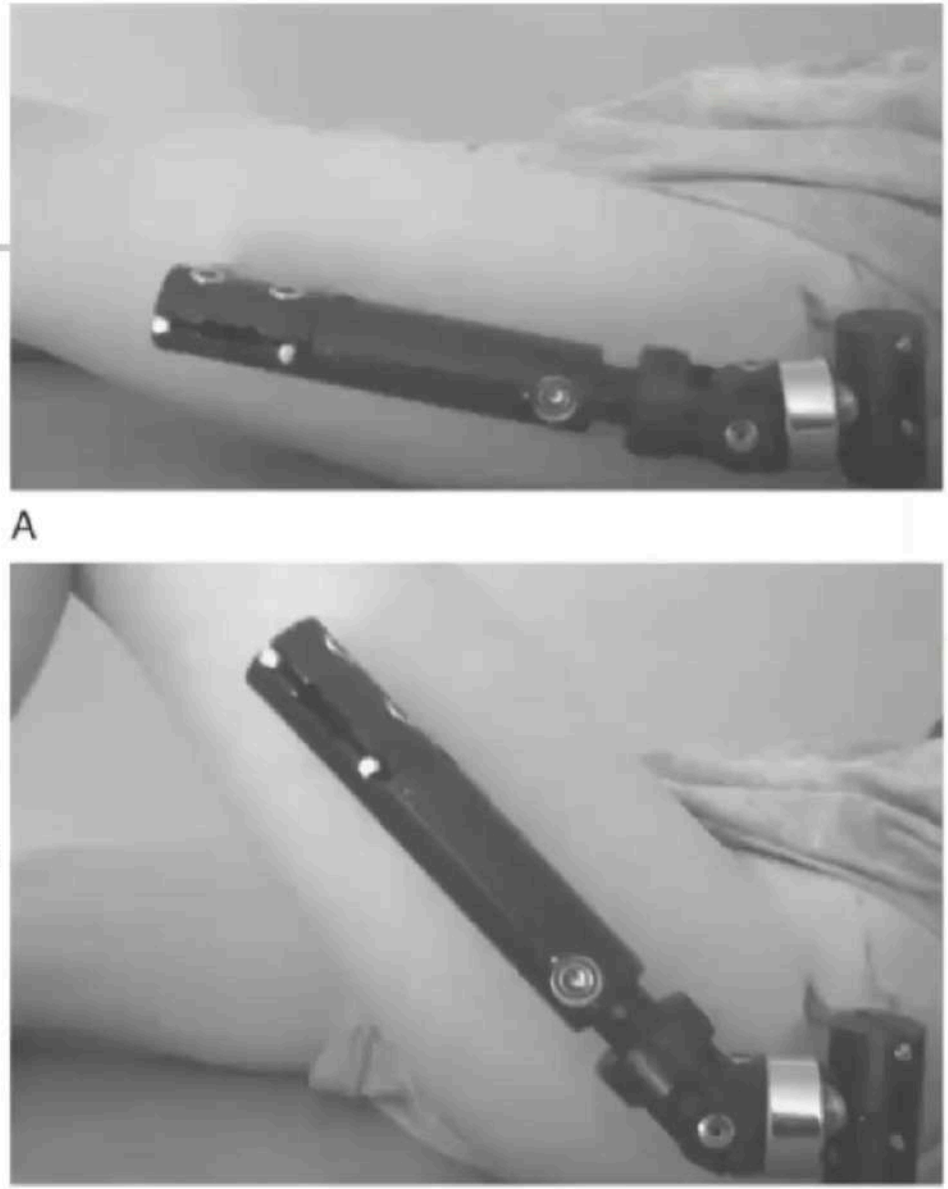






Arthrodiastasis

- The rationale behind arthrodiastasis is that distraction of the joint not only widens, but also unloads the joint space, reduces the pressure on the femoral head, allows fibrous repair of articular cartilage defects, and preserves congruency of the femoral head.



A and B, Hinged external fixator (Orthofix) for arthrodiastasis.

Greater trochanteric overgrowth

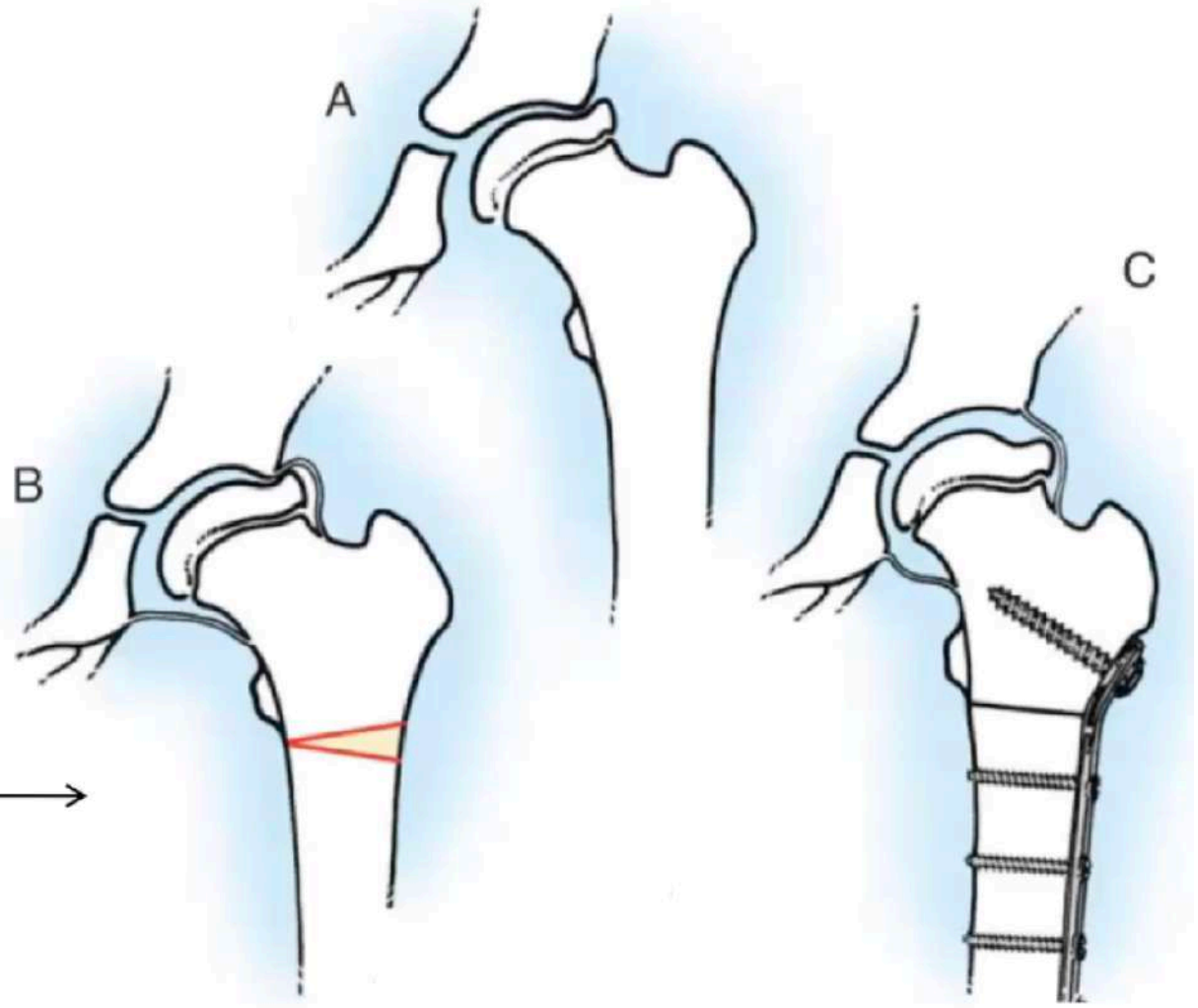
- The trochanteric overgrowth can be dramatic on radiographs but several studies have shown that a Trendelenberg gait does not always occur.
- If it does occur, and is significant, then trochanteric advancement may improve the gait.
- An alternative is to perform a trochanteric arrest at an earlier date but this assumes that the first statement will not apply to the particular child.

RECONSTRUCTIVE SURGERY

- **Hinge abduction.** - valgus subtrochanteric osteotomy.
- **Malformed femoral head** in late group III or residual group IV. - Garceau's cheilectomy.
- **Coxa magna.** - shelf augmentation
- A **large malformed femoral head with lateral sublaxation.** - Chiari's pelvic osteotomy.
- **Capital femoral physeal arrest.** - trochanteric advancement or arrest.

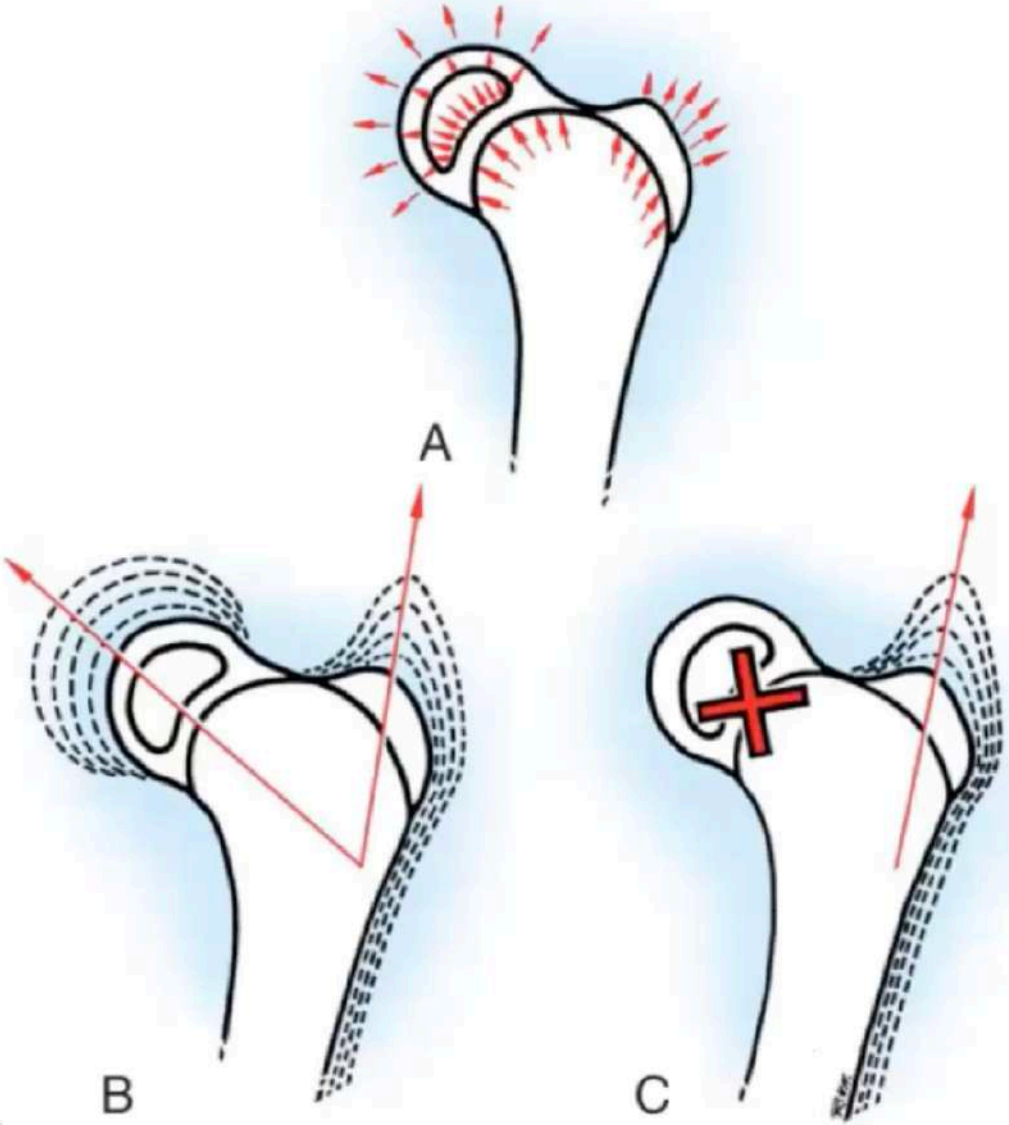
Valgus Extension Osteotomy

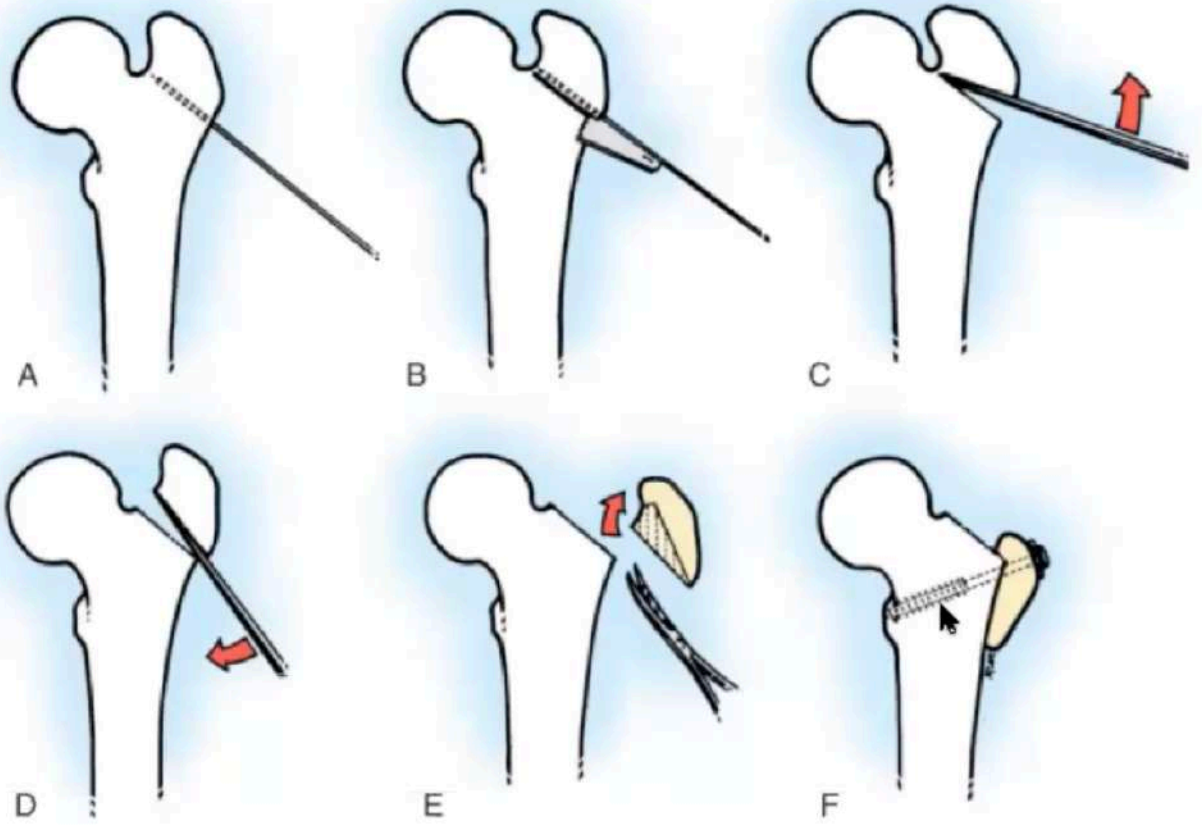
Hinged abduction of the hip is an abnormal movement that occurs when the deformed femoral head fails to slide within the acetabulum.



Valgus osteotomy to reduce hinge abduction

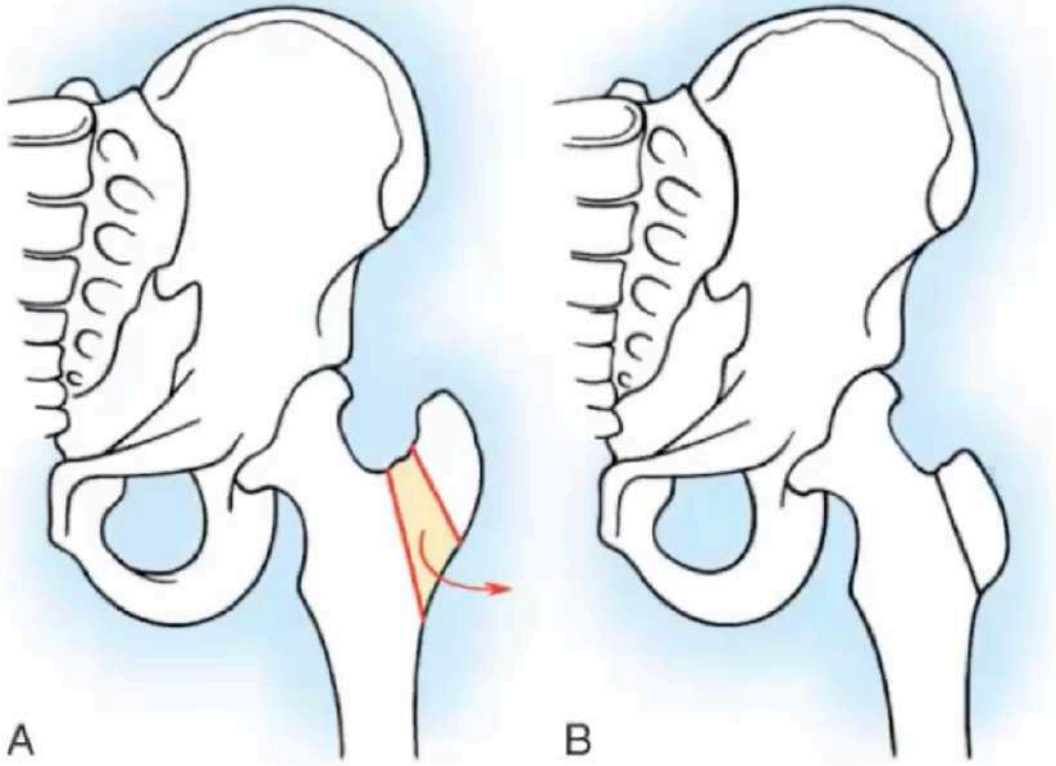
Trochanteric Overgrowth





A-F, Trochanteric advancement for trochanteric overgrowth

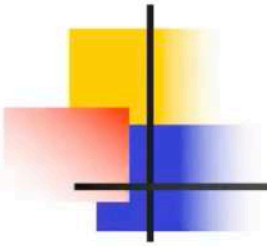
After initial osteotomy of greater trochanter, trapezoidal wedge of bone is removed.



Trochanteric osteotomy for trochanteric overgrowth

Summarised approach

- Onset on or before 8 years- symptomatic treatment
- After 8 years, lateral pillar groups A & C- symptomatic
- After 8 years, lateral pillar B- surgery



- Surgical choices- femoral varus osteotomy, acetabular osteotomy, combined(onset after 9 years)
- Late measures for established head & acetabular flattening, adducted hip, short leg gait – femoral valgus osteotomy

- Mechanical symptoms: hip arthroscopy, removal of osteochondrotic fragment, surgical dislocation for incongruity

SUMMARY

- For patients less that 6 years old the prognosis is good for the majority.
- If they are stiff or painful they respond to bed rest, traction and pain relieving anti-inflammatory medication.
- There is no evidence that abduction splints or surgical intervention is warranted in the majority of these younger patients.

SUMMARY

- If they are between 6 and 8 and are in lateral pillar group C then the result of intervention are equivocal.
- Children presenting with Perthes disease at age 9 or older often have lateral pillar B or C and a poor prognosis.
- The trend is towards early containment of these hips although stiffness can be a problem following early pelvic (Salter's) osteotomy.

Follow-up

- Initially, close follow-up is required to determine the extent of necrosis.
- Once the healing phase has been entered, follow-up can be every 6 months.
- Long-term follow-up is necessary to determine the final outcome.

Complications

Femoral

- Shortening
- stiffness
- Malrotation
- Limp
- Positive trendelenburg

Pelvic

- Lenghtening
- Stiffness
- Chondrolysis
- Failure of containment

Prognosis

- The younger the age of onset of LCPD, the better the prognosis.
- Children older than 10 years have a very high risk of developing osteoarthritis.
- Most patients have a favorable outcome.
- Prognosis is proportional to the degree of radiologic involvement.