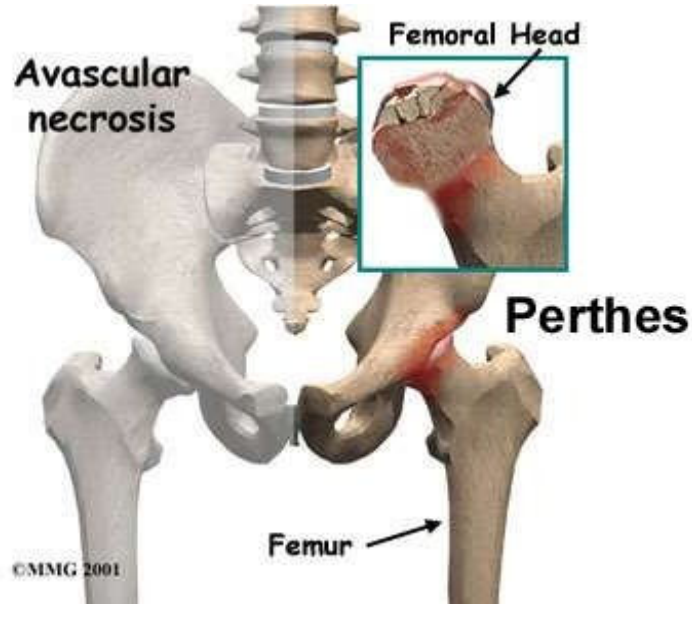
Legg–Calvé–Perthes disease

Learning objectives

Introduction to LCPD

It is a self limiting disorder of the hip produced by ischemia and varying degrees of necrosis of the femoral head.



Epidemiology

- Incidence : 1:1000
- Usual age : 4-8 years
- Boys:girls – 5:1
- Higher incidence in Caucasian, Chinese, Japanese, Inuits, Northern Europe

Aetiology-

Coagulation disorders.

Arterial status of femoral head.

Abnormal venous drainage.

Abnormal growth and development.

Trauma.

Hyperactivity or attention deficit disorder.

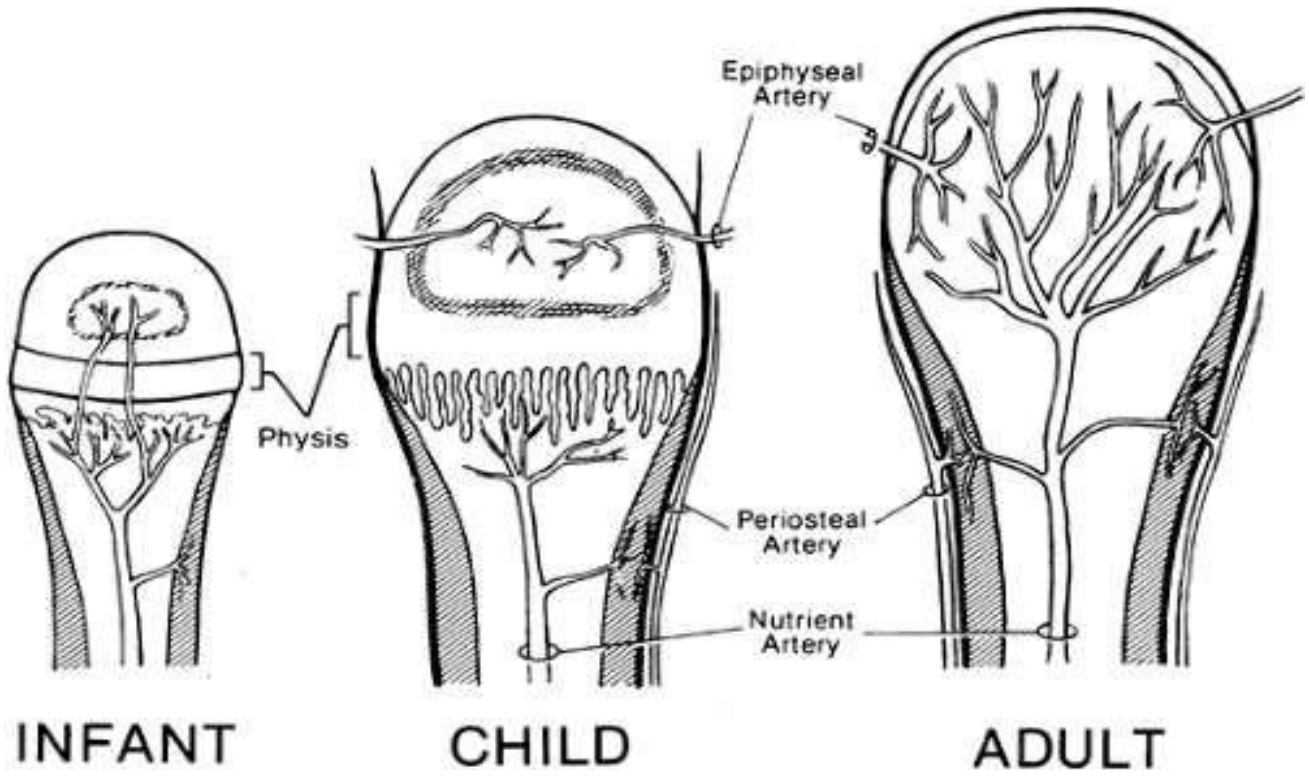
Genetic component.

Environmental influences.

As a sequel to synovitis.

Pathogenesis

- Ischemia of femoral head



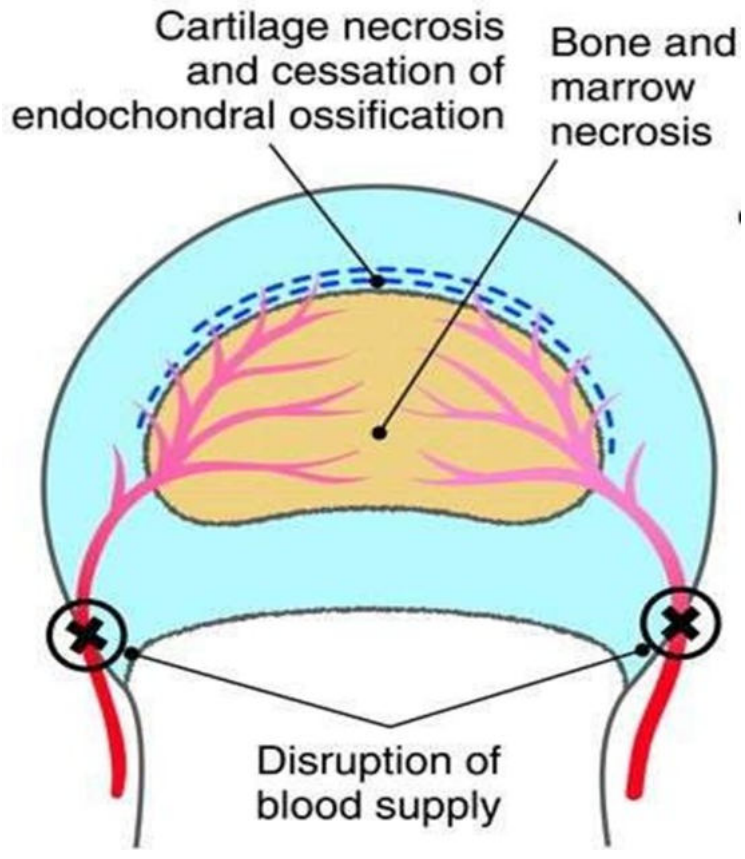
Up to 4 months	1. Metaphyseal vessels 2. Lateral epiphyseal 3. Scanty vessels in ligamentum teres
4-7 years	1. Lateral epiphyseal vessels 2. Metaphyseal supply DISSAPEAR
7 years	1. Vessels in ligamentum teres have developed

Susceptible to ischemia, as it depend entirely on lateral epiphyseal vessel.

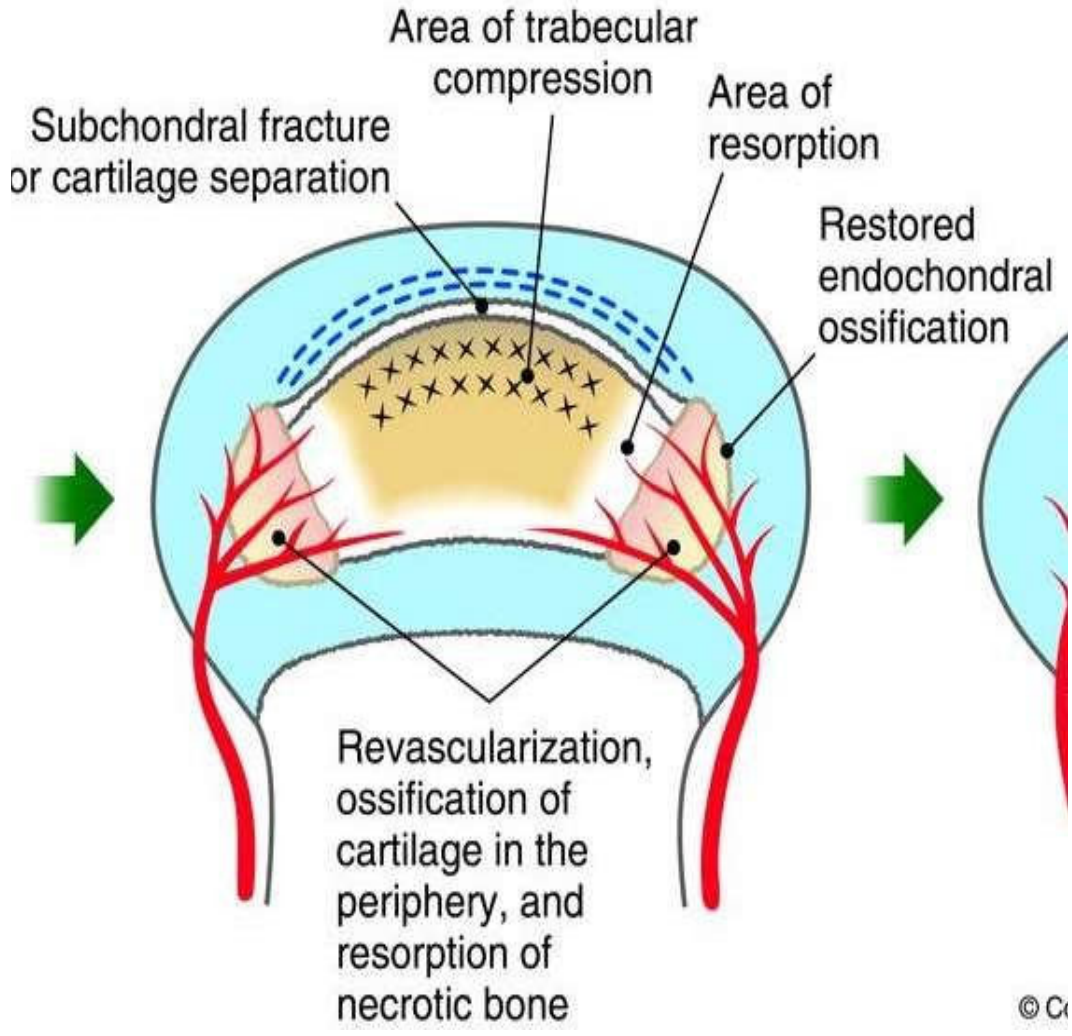
Pathology-Stage 1-Ischaemia

Pathological process → 3-4 years

- All/part of bony nucleus of femoral head is dead
- Cartilaginous part – remains viable and thicker
- Thickening and edema of synovium and capsule

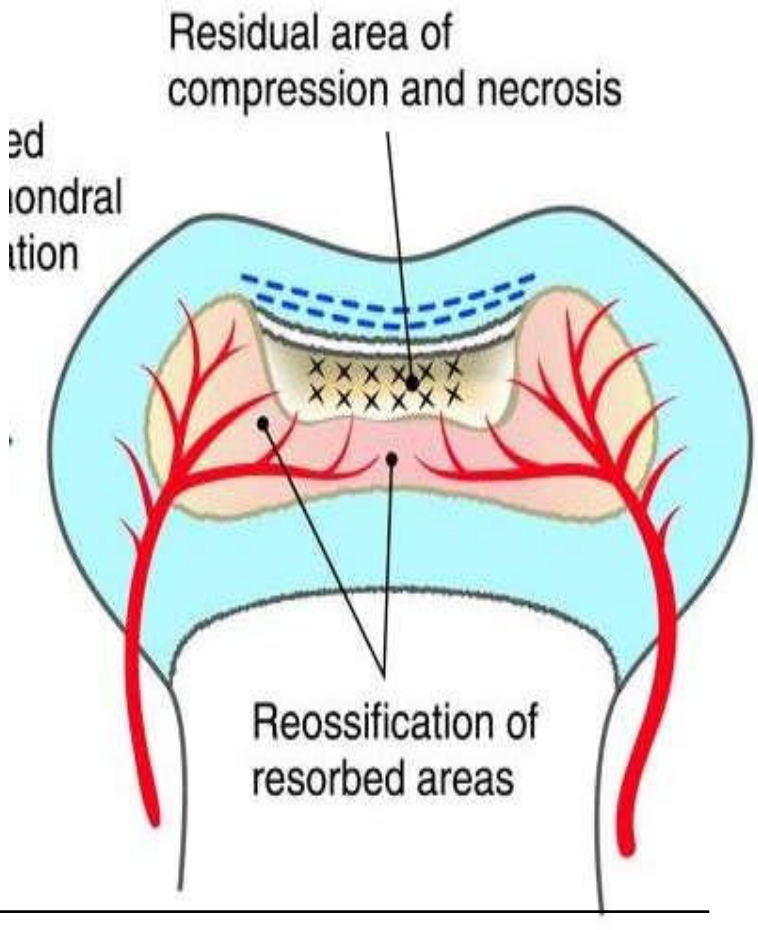


Stage 2 – revascularization and repair



Stage 3 – Distortion and remodeling

- Repair process
  - Rapid and complete : shape is restored
  - Tardy : bony collapse and growth distortion



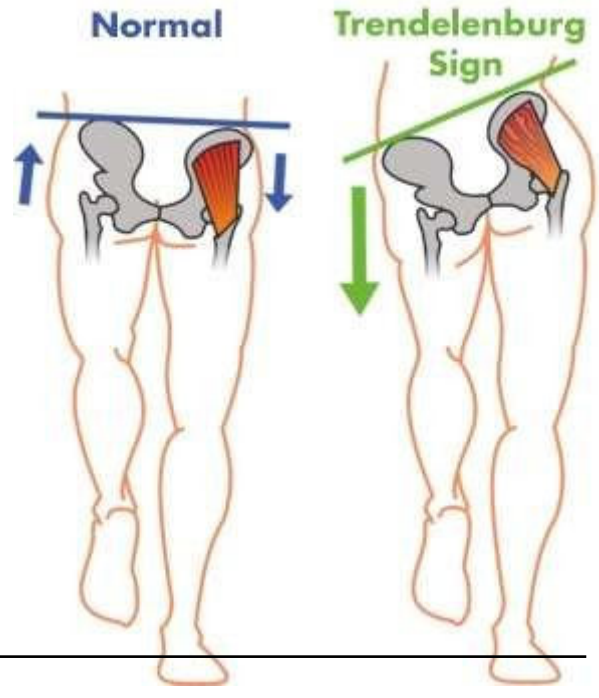
Clinical feature

Symptoms

- Typically male – 4-8 years
- Painless limping – continues for weeks or recur intermittently
- Pain in groin, thigh and knee – activity related, relieved by rest

Signs

- Hip pain with passive range of movement
- Reduced range of movement (abduction & internal rotation)
- Hip flexion contracture
- Leg length discrepancy
- Mild muscle wasting – thigh, calf,
- Tredenlenburg test ; positive



Investigation

- X-ray of both hips (AP & Frog lateral view)
- Bone scan
- CT scan – follow up
- Arthrography : to see congruity, head deformity and determine method of treatment
- Blood inflammatory marker
  - FBC
  - ESR
  - CRP

X-ray

- Widening of joint space
- Sclerosis
- Necrotic phase : increase density of ossific nucleus
- Fragmentation : alternating patches of density and lucency
- Lateral uncovering of femoral head
- Acetabular remodelling

Waldenström classification based on radiographic changes

<u>Stage 1 (increased density)</u>

- ossific nucleus smaller and denser
- subchondral fracture
- radiolucencies in the metaphysis

Caffey's sign

→Subchondral fracture in the anterolateral aspect of the femoral capital epiphysis

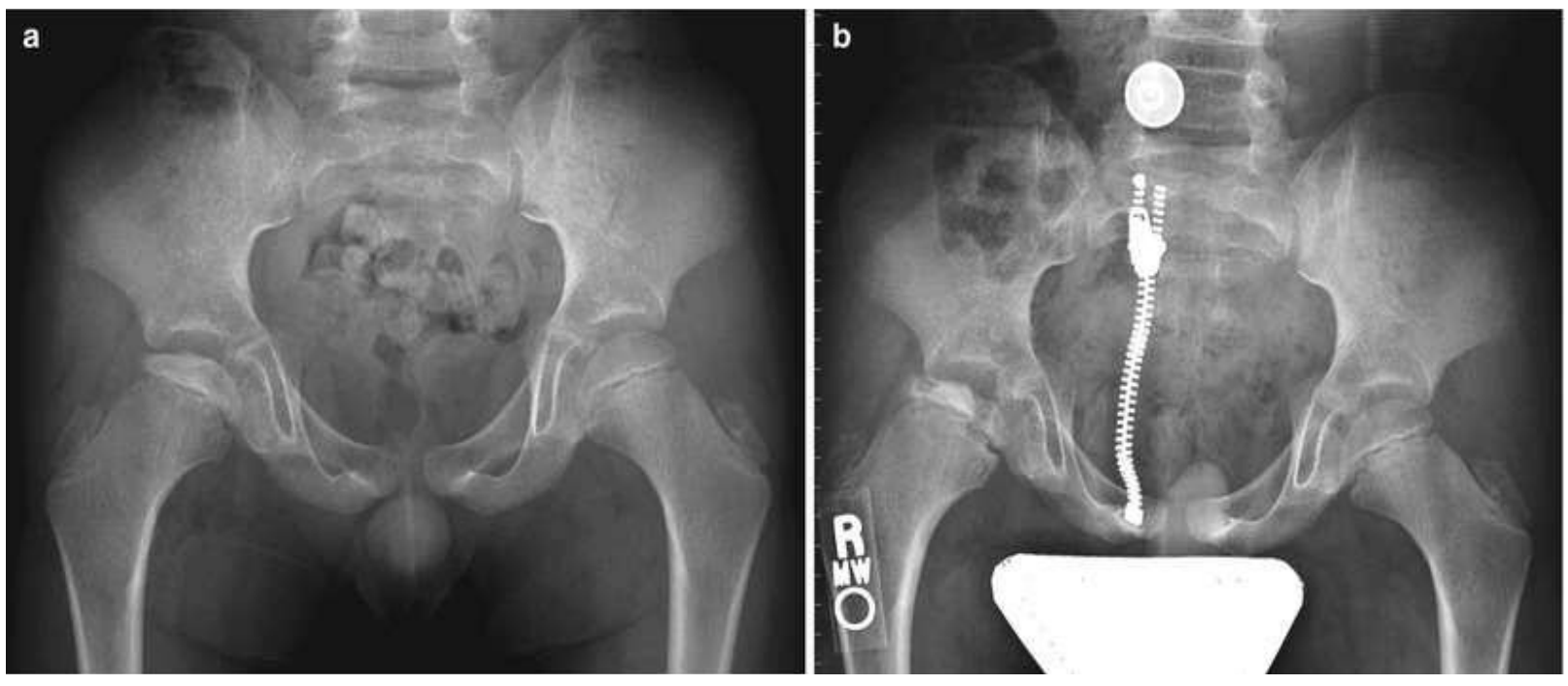
→Produces crescentic radiolucency

Waldenström classification based on radiographic changes

<u>Stage 2 (fragmentation and revascularization)</u>

- lucency in epiphysis
- pillars are demarcated
- metaphyseal changes resolve

-acetabular contour change



Stage 3 (healing or reossification stage)

- new bone formation
- homogenous epiphysis

Stage 4 (remodelling)

- femoral head is reossified and remodels
- acetabular remodelling

Classification

According to radiologic stage of disease –

Waldenström classification

According to prognostic outcome –

– Herring lateral pillar

According to defining outcome –

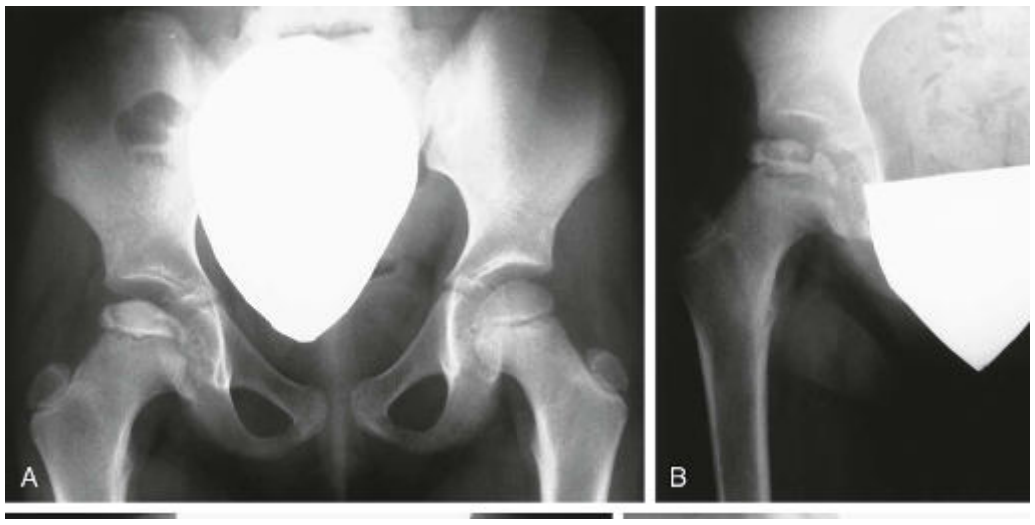
Stulberg classification

The Herring lateral pillar classification–

lateral pillar not affected	**>50% of height of lateral pillar preserved**	**<50% of height of lateral pillar preserved**

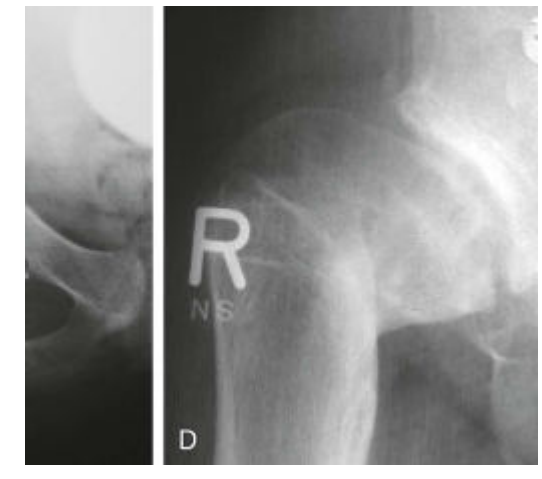
Herring stage A

Good prognosis



Herring stage B

Herring stage C



Prognostic features

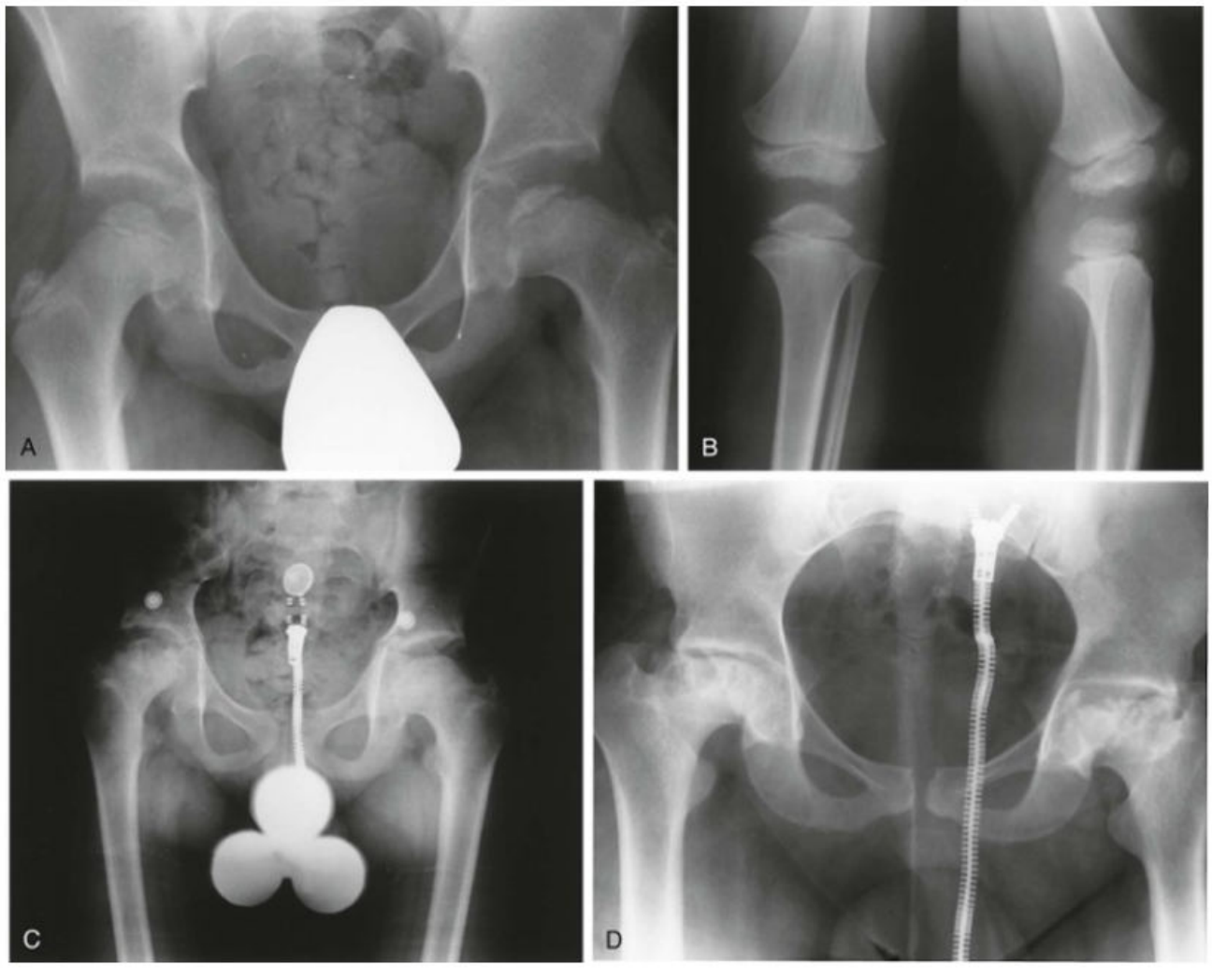
- Child under 6 years – excellent
- Age on higher side
- Progressive loss of hip motion more so abduction
- Obese child
- Progressive uncovering of the epiphysis

Differential diagnosis

- Multiple epiphyseal dysplasia
- Spondyloepiphyseal dysplasia
- Mucopolysaccharidoses
- Hypothyroidism
- Other Causes of Avascular Necrosis
  - Sickle cell disease
  - Steroid medication
  - Sequela of traumatic hip dislocation
  - Treatment of developmental dysplasia of the hip
- Septic arthritis

D/D-Hypothyroidism

D/D-Multiple Epiphyseal dysplasia



Management-Principles

1. Prevent deformity to femoral head before remodelling phase
2. Restore and maintain ROM
3. Concept of containment
4. Relief of symptoms

Guidelines to treatment

- Decision are based on :
  - Stage of disease
  - Prognostic x-ray classification
  - Age and clinical feature particularly range of abduction and extension

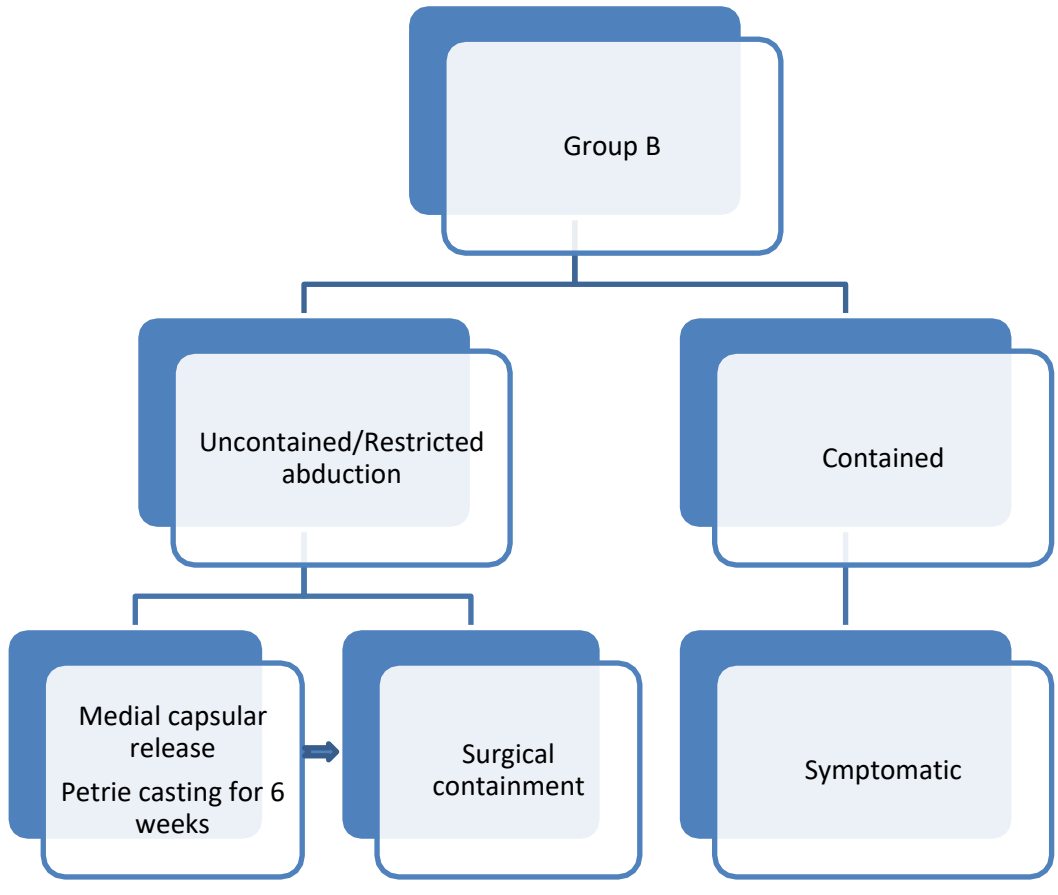
Guidelines by Herring (1994)

- Child <6 years(age at onset)
- Symptomatic treatment-Rest/Analgesics
- Operative intervention-no added benefit

Age at onset 6-8 years

Group A

Symptomatic



Age at onset 8-11 years

- Surgical treatment gives better outcome
- In very early stage when fragmentation yet to appear,do perfusion MRI
- If severe ischaemic changes-Surgical containment
- Group B,B/C-Surgical containment

Symptomatic

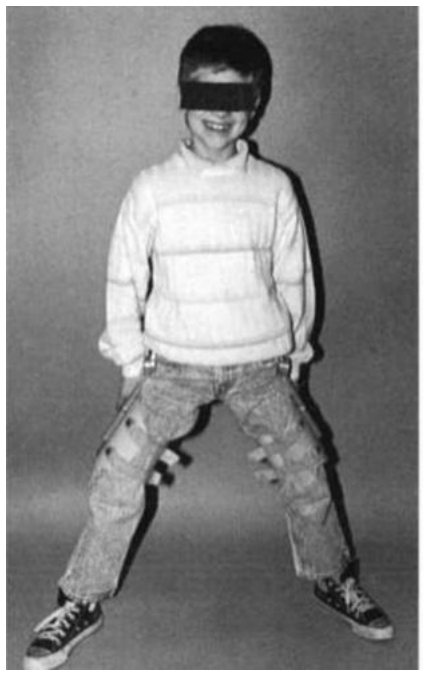
- Pain control
- Hospitalization for bed rest and short period traction
- Gentle exercise to maintain movement

Containment

Harrison and Menon stated ;

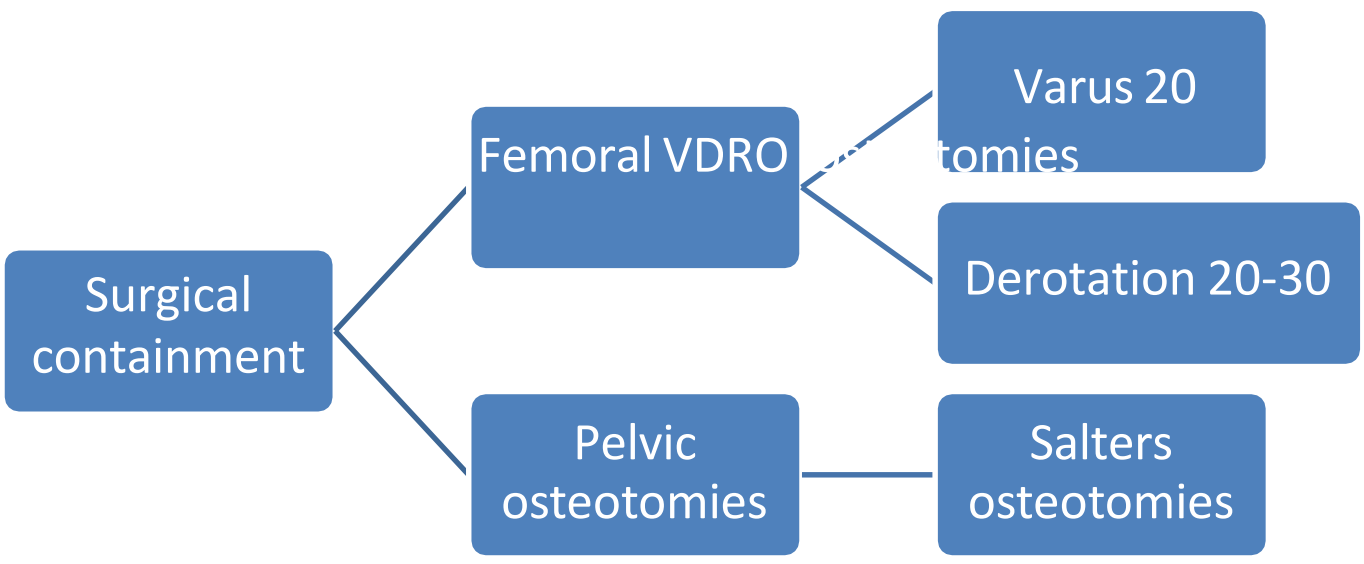
‘if the head is contained within the acetabular cup, then like jelly poured into a mold the head should be the same as the cup when it is allowed to come out after reconsitution ‘

Containment – non operative

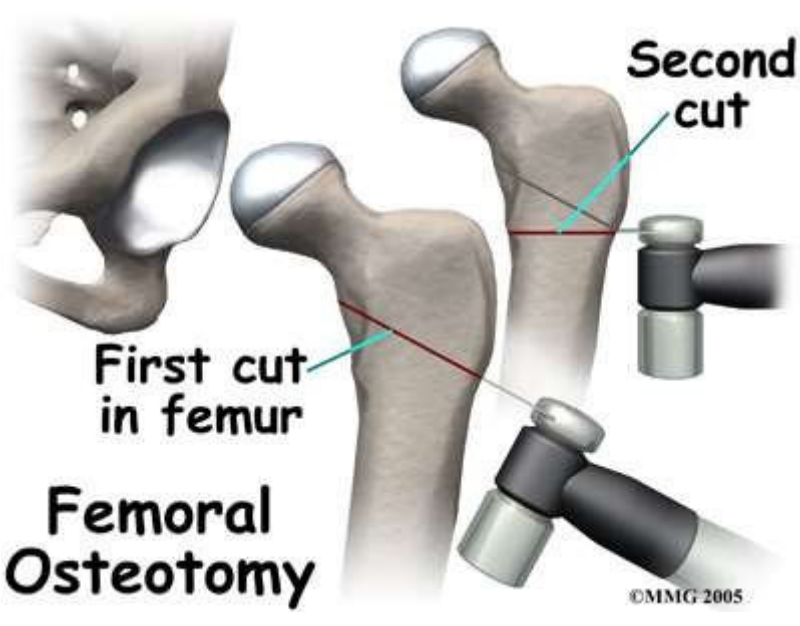


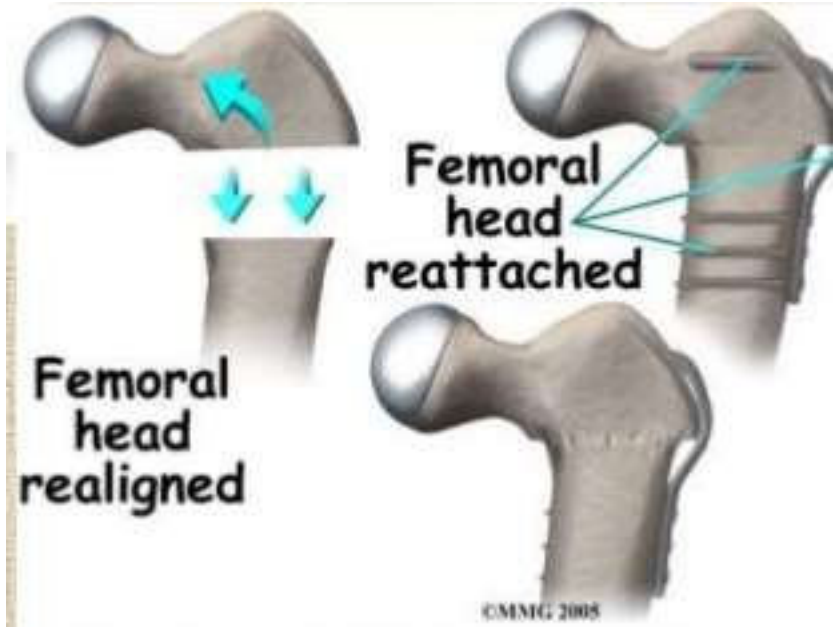
Containment – surgical

- Done before irreversible deformation of femoral head occurs (early in fragmentation stage)



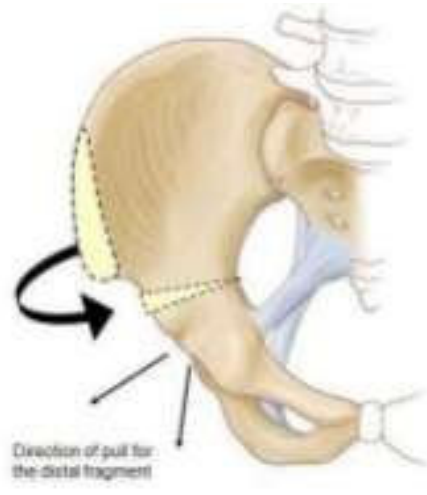
Femoral varus derotational osteotomy





Pelvic osteotomy

Innominate osteotomy using quadrangular Graft (Canale et al)



Salter innominate osteotomy

Summary