

PERTHES DISEASE

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DEFINITION

- It is a condition in which the blood supply of the capital femoral epiphysis is disrupted, resulting in epiphyseal osteonecrosis and chondronecrosis with cessation of growth of the ossific nucleus.

EPIDEMIOLOGY

Onset:

Between 2- 12 years,
with a peak in 6 to 8
years of age.

Male prevalence:
4 or 5 times more
likely in boys than
in girls

Due to incomplete
anastomosis in
boys

Involvement:

Bilateral
in 10% to 12% of
pts

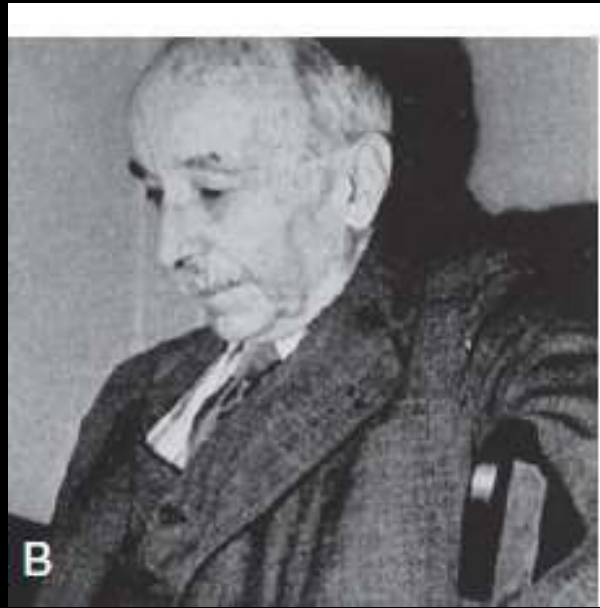
that too affecting the hips
sequentially and not
simultaneously

HISTORY

- LCPD was independently recognized as a distinct entity in 1910 by



Arthur Legg of
United States



Jacques Calvé of
France



Georg Clemens
Perthes of Germany



Henning Waldenström
of Sweden

AETIOLOGY

Cause for the vascular insult is unknown

Genetic & several environmental factors have been implicated.

Currently, some suggests that Perthes disease pts have a disorder of clotting mechanism, with deficiency of factors - protein C or S & factor V Leiden mutation.

Box 14.2 Etiology of Legg-Calvé-Perthes Disease

FACTORS THAT MAY BE ETIOLOGIC

Susceptibility in a child

Trauma

Hyperactivity

Socioeconomic deprivation

Passive smoking

Coagulopathy

Type II collagenopathy

FACTORS UNLIKELY TO BE ETIOLOGIC

Hereditary factors

Endocrinopathy

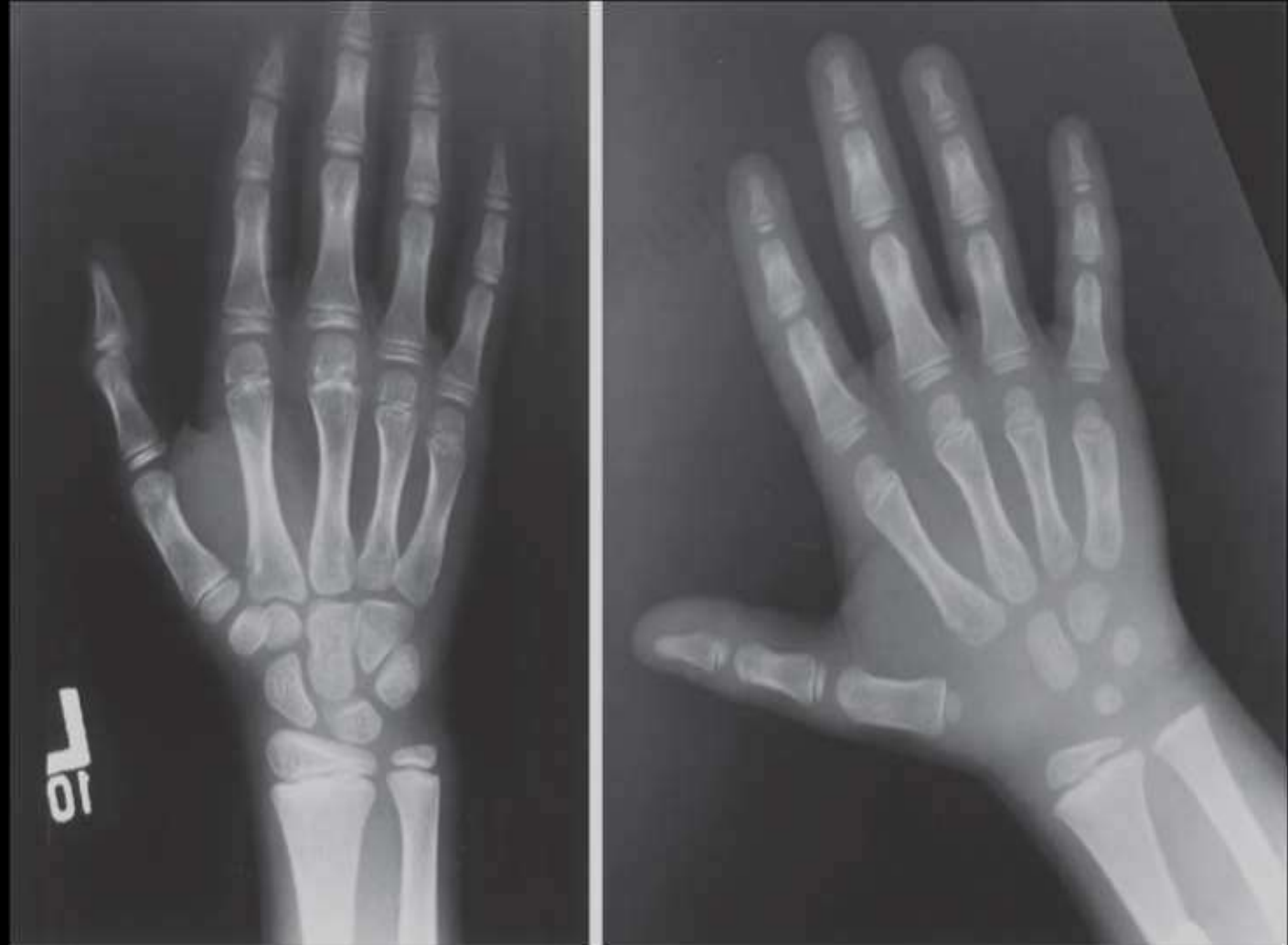
Urban environment

Synovitis

“THE PREDISPOSED CHILD”

Delay in bone age relative to the patient’s chronological age is the most commonly observed abnormality & is seen in early years of the disorder.

The carpal bone age is frequently 2 years or more behind the chronological age.

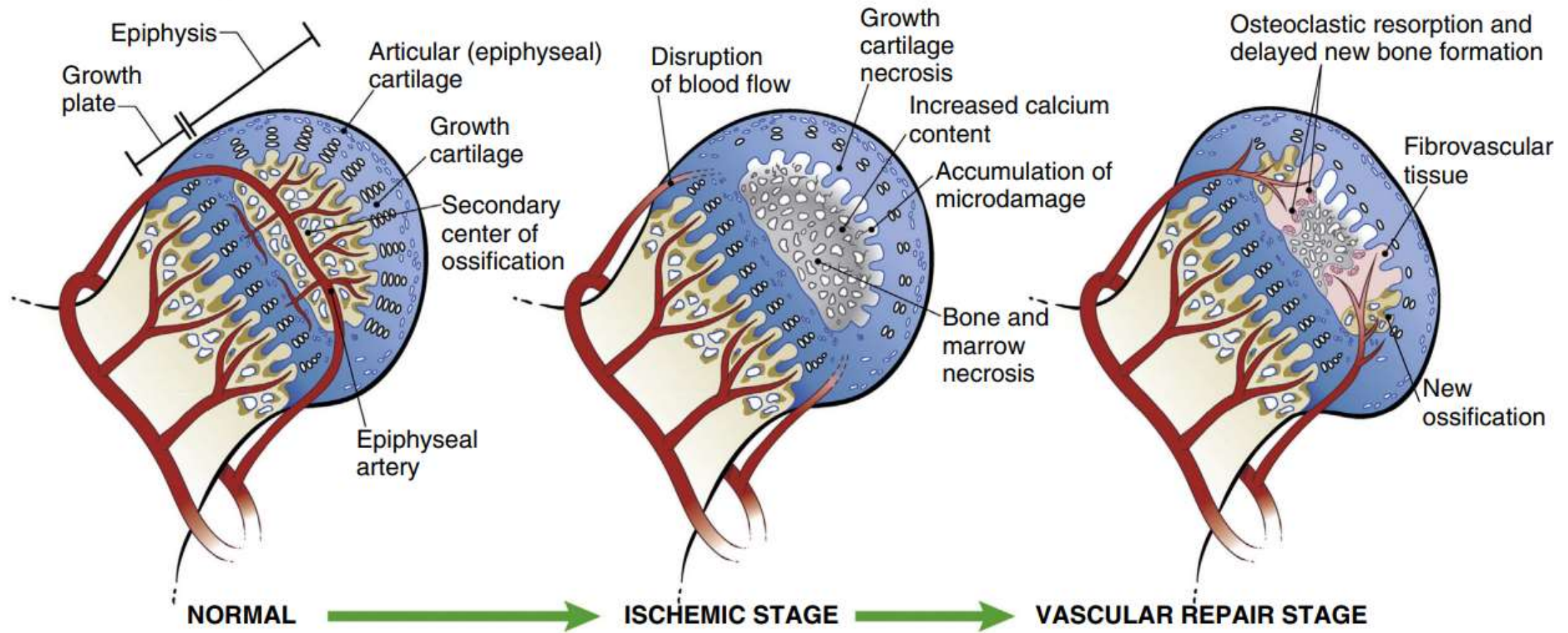


- Children < 5 years age showed an increase in bone age delay over the following 4 to 5 years, whereas those diagnosed after 8 yrs of age had less delay over the ensuing years.
- A “**radiologic pause**” in bone age has also been observed, with particular carpals
 - affected more than others.
- This phenomenon had previously been referred to as “**Skeletal Standstill.**”
- Although growth of the triquetrum & lunate is significantly delayed, the capitate & hamate are not affected.

BLOOD SUPPLY OF FEMORAL HEAD-

- Lateral ascending cervical artery which is a branch of medial circumflex femoral artery while advancing between the trochanter and capsule ,it courses through a narrow passage ,an area particularly constricted in children younger than 8 years age
- The sub-capsular ring of vessels is found to be incomplete more often in boys than in girls

Proposed Pathogenesis of Femoral Head Deformity Following Ischemic Necrosis



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FIG. 14.13 Proposed pathogenesis of femoral head deformity. Disruption of blood flow to the growing femoral head produces cell death of osteoblasts, osteocytes, osteoclasts, bone marrow cells, and chondrocytes in the deep layer of the articular cartilage. The ischemic damage produces cessation of growth of the epiphysis and increased calcium content of the necrotic bone, thus making it more brittle to accumulate microdamage with mechanical loading. With initiation of revascularization, a predominance of osteoclastic resorption is observed with delayed bone formation. The net bone loss further weakens the epiphysis and makes it more prone to deformation with loading. The fragmentation stage of the disease is marked by an imbalance of bone resorption and bone formation. (© 2011, Texas Scottish Rite Hospital for Children, Dallas.)

CLASSIFICATION OF PATHOGENESIS

Waldenström classification determines the four stages all cases follow:

1) Initial Stage—sclerotic epiphysis with joint widening (X-rays may not show changes for 4-6 months)



Linear fractures in subchondral area of femoral head is known as **Salter's sign**, **Caffey's sign** or **Waldenstrom's sign**.



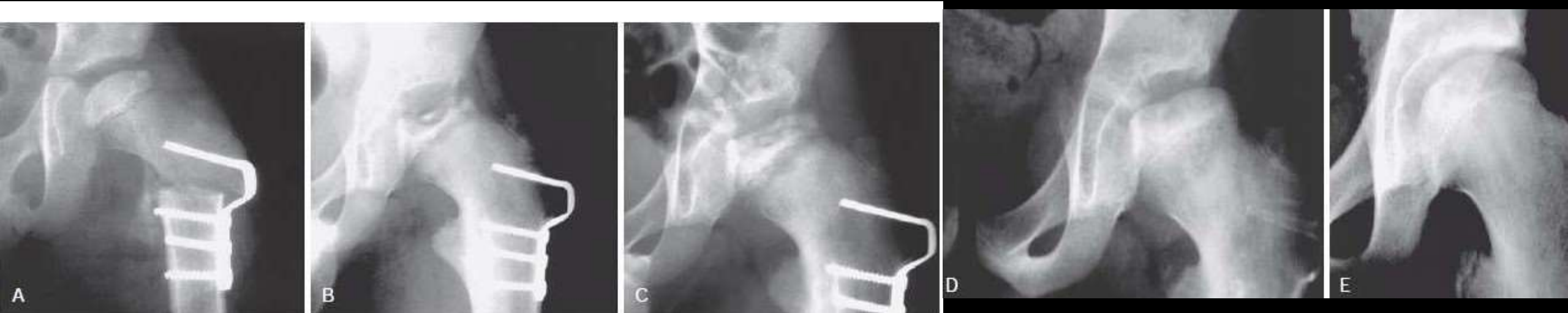
2) Fragmentation Stage—
due to bone resorption and
collapse (lateral pillar
classification based on this stage)



3) Healing/
Reossification Stage-
new bone appears
(may last up to 18
months)



4) Remodelling—
continued remodelling until maturity



SYMPTOMS

1) Limp- Most common presentation

Combination of an antalgic gait and a Trendelenburg gait

Trendelenburg test is positive on the affected side

Exacerbated by activity & alleviated with rest

2) Pain- May be located in the groin, proximal thigh, knee, or laterally around the Greater Trochanter.

3) Possible history of antecedent trauma

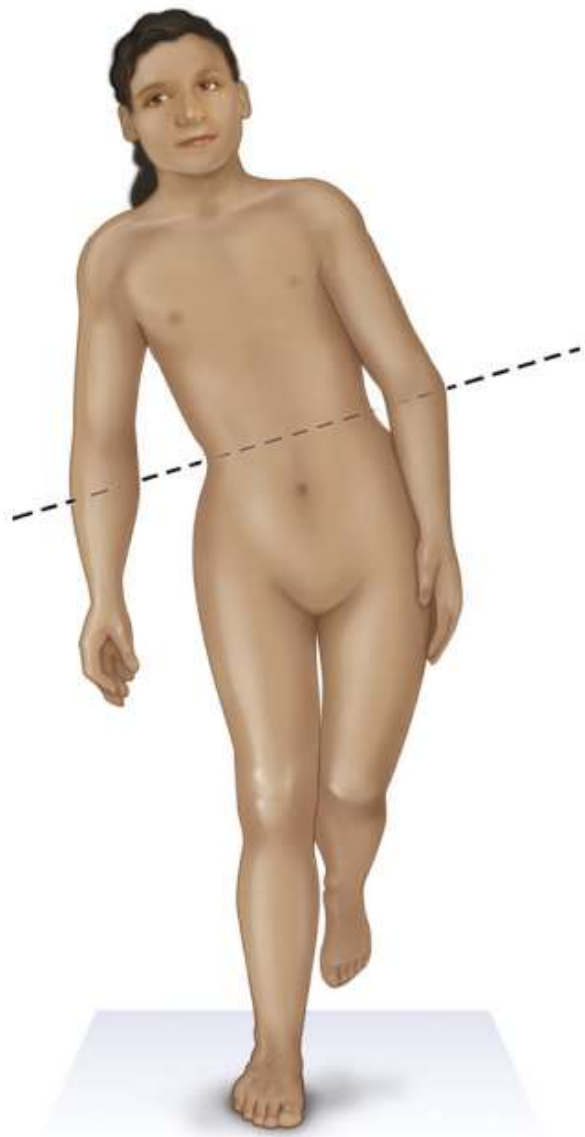


FIG. 14.16 Illustration of a typical Legg-Calvé-Perthes limp, which is a combination of an antalgic gait and a Trendelenburg gait. In the stance phase of gait, the patient leans the body over the involved hip to decrease the force of the abductor muscles and the pressure within the hip joint.

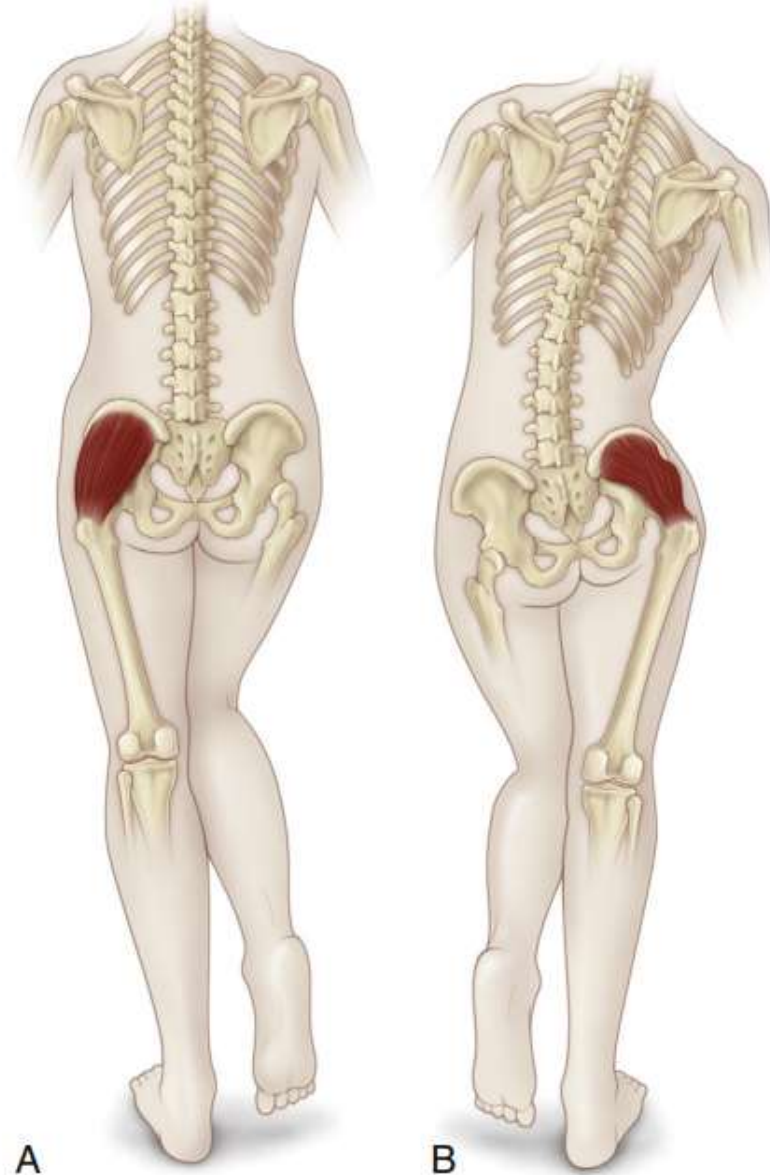


FIG. 14.17 Positive Trendelenburg test result. (A) As the patient stands with the weight on the normal hip, the pelvis is maintained in the horizontal position by contraction and tension of the normal hip abductor muscles. (B) As the patient stands with the weight on the affected hip, the pelvis on the opposite normal side drops as a result of weakness of the hip abductor muscles.

SIGNS

- Abductor limp
- Decreased ROM of hip, especially on abduction & internal rotation (decreased ROM is transient early in the disease, persistent later on)
- Flexion and extension less affected
- Positive Logroll test.

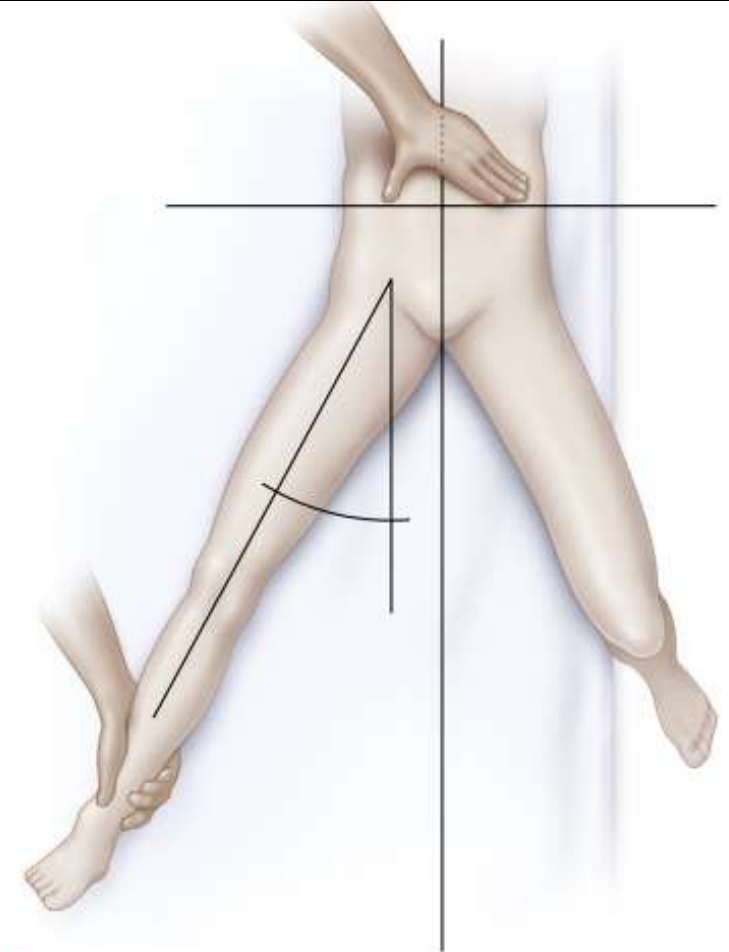


FIG. 14.18 Hip abduction is best examined in extension with the patient supine. Dropping the uninvolved leg over the side of the examining table helps stabilize the pelvis. (Reproduced with permission from Herring JA. *Legg-Calvé Perthes Disease*. Rosemont, IL: American Academy of Orthopaedic Surgeons; 1996.)

Table 14.2 Association Between Clinical Findings and Radiographic Stages of Disease.

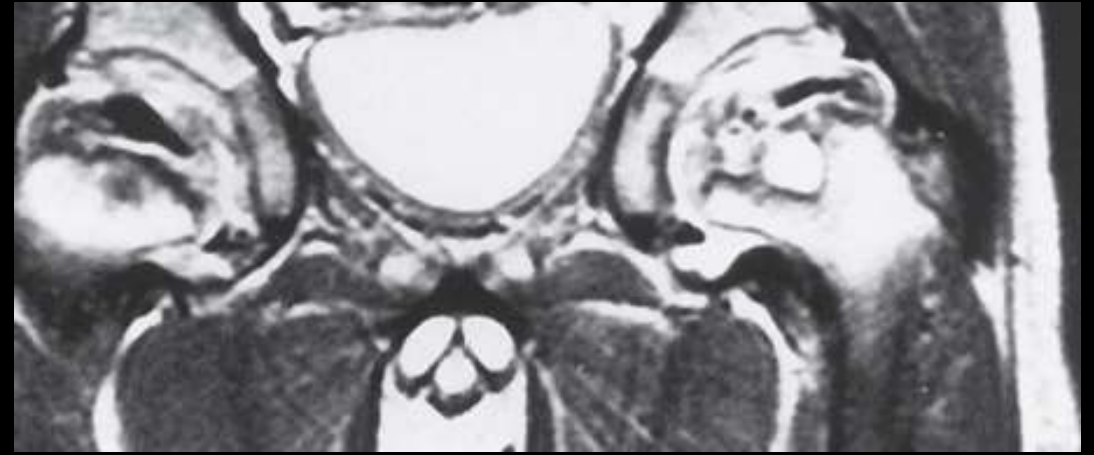
Stage	Clinical Findings	Radiographic Changes
Increased density stage	Limp and pain variable, often mild and intermittent	Increased density of femoral head, with/without subchondral fracture
Fragmentation stage	Pain and limp may worsen; may lose range of motion	Head shows fragmentation and resorption, 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; some develop impingement symptoms	May develop osteochondrotic lesion or femoroacetabular impingement

INVESTIGATIONS

- X-rays
- MRI
- CT Scan
- USG
- Technetium Bone Scan

X-RAYS:

1) Changes in the Metaphysis: Metaphyseal cyst



2) SAGGING ROPE SIGN

-Radiodense line in proximal femoral metaphysis

-Metaphyseal response to physeal damage



FIG. 14.23 The sagging rope sign, a ropelike, radiodense line across the intertrochanteric area (*arrow*), represents the edge of the enlarged femoral head, or coxa magna.

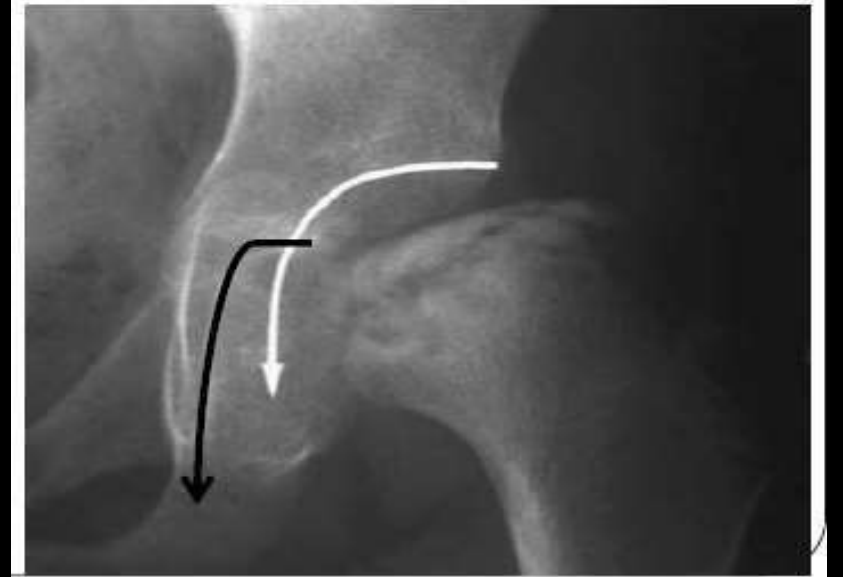
3) Changes in the Physis:
Premature physeal closure



4) Changes in the Acetabulum

-Osteoporosis of acetabular roof

-Premature fusion of triradiate cartilage (bicompartimentalisation)



5) Head-At-Risk Signs

- Lateral subluxation of the femoral head
- A radiolucent V in the lateral aspect of the epiphysis (the Gage sign)
- Calcification lateral to the epiphysis
- Horizontal physeal line.
- The presence of these signs increased the chances of a poor outcome



Saturn Phenomenon

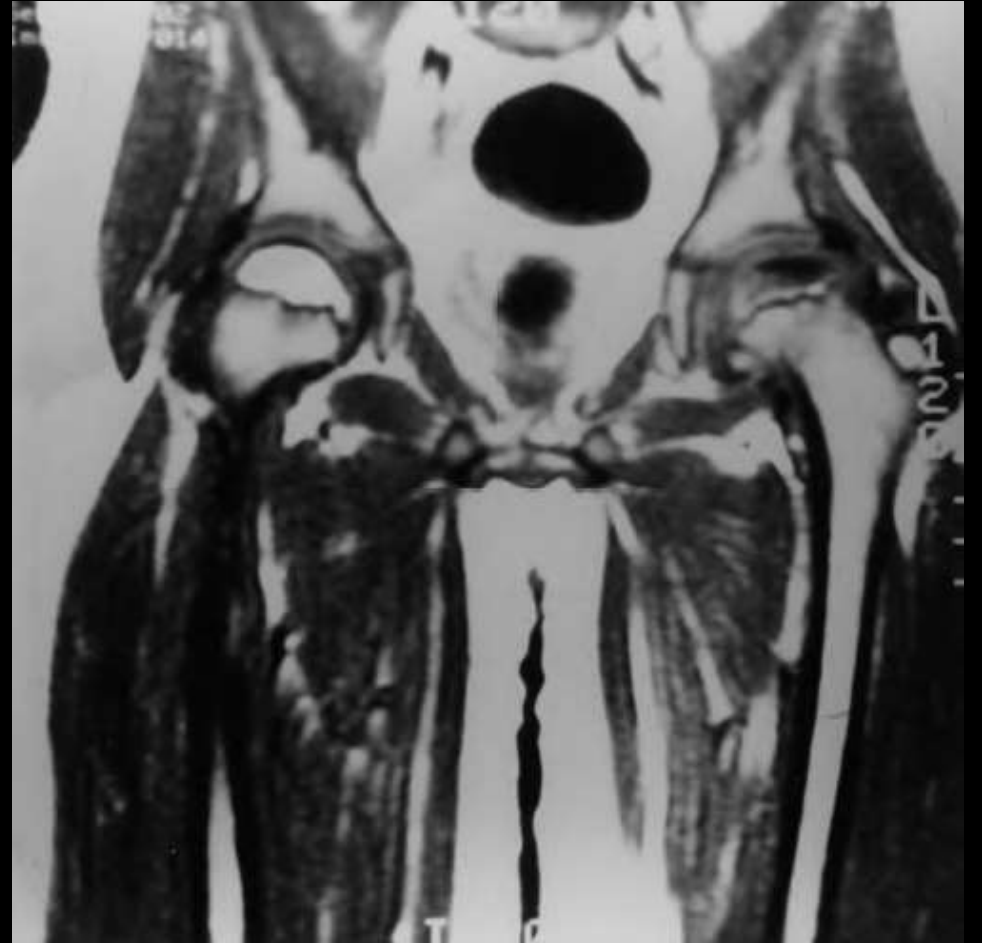


FIG. 14.41 The Saturn phenomenon, a radiolucent ring occasionally seen in young children with Legg-Calvé-Perthes disease. When this pattern is present, the outcome is often poor. Note flattening of the epiphysis medially and laterally, with the central fragment having the shape of a flying saucer.

Widening of the femoral head before fragmentation, the (a sclerotic epiphysis surrounded by a ring of lucency

MRI

- MRI is an accurate investigation for diagnosis of early Perthe's disease.
- The varying modalities of MRI, such as perfusion and diffusion MRI and delayed gadolinium-enhanced MRI (dGEMRIC), may offer new insights into the pathophysiology and prognostication of this disorder



USG

-Sometimes used in early stages of LCPD to demonstrate joint effusion and in later stages to assess femoral head shape.

-Can provide a good profile of the cartilaginous femoral head (comparable with that of arthrography) and allows subsequent observation of deformation of the head without the need for radiographs

Technetium scan with pinhole collimation.

Scintigraphy used to classify revascularization as either recanalization of existing vessels (the A track or pathway) or neovascularization, meaning new vessel formation (the B track or pathway)

A track has a favorable prognosis, whereas patients on pathway B generally had a poorer prognosis and often required surgical intervention

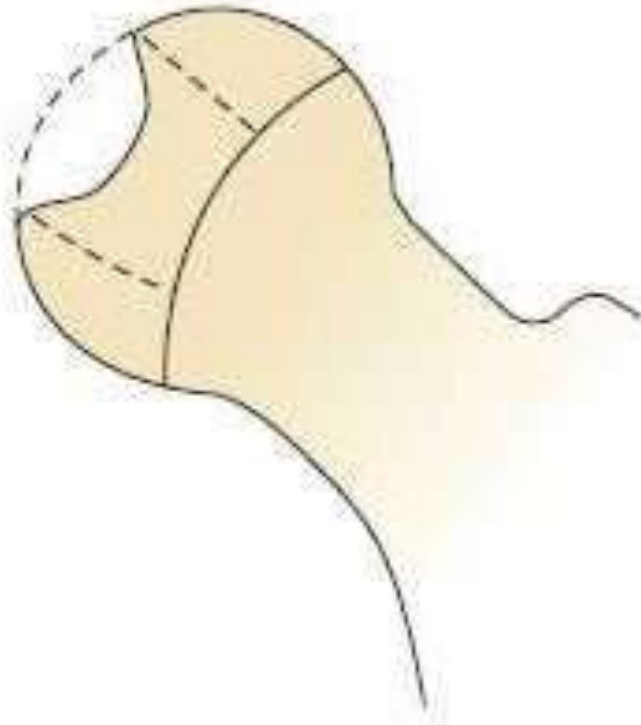


CLASSIFICATIONS

Most common prognostic classification used is Herring classification, also known as Lateral Pillar Classification.

Based on involvement of lateral pillar of capital femoral epiphysis during the fragmentation stage

Lateral pillar classification of Legg-Calvé-Perthes disease



Normal pillars

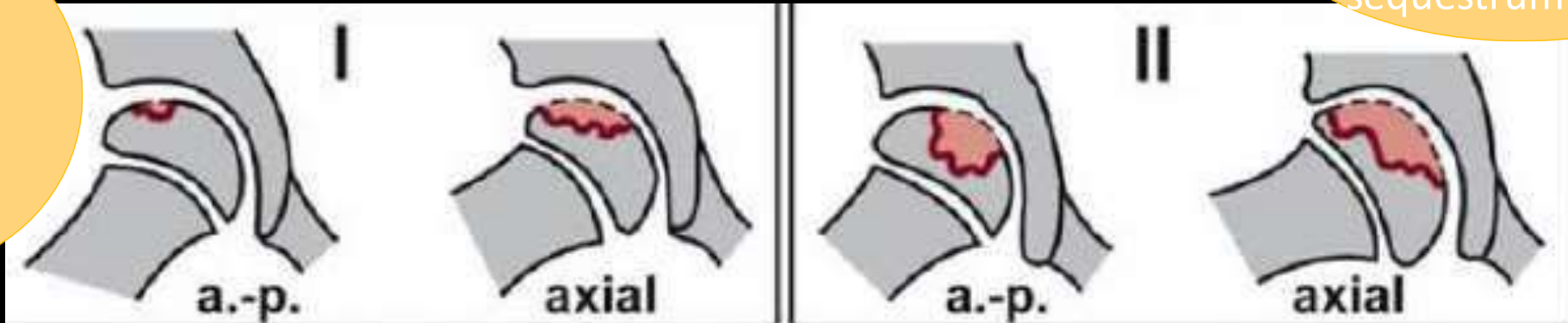
Normal pillars were derived by noting the lines of demarcation between the central sequestrum and the remainder of the epiphysis on the anteroposterior radiograph



FIG. 14.32 Lateral pillar classification: *group A*, no loss of height in the lateral pillar; *group B*, partial collapse (<50%) of the lateral pillar; *group B/C border*, intermediate between B and C, approximately 50% collapse, thin pillar, irregular; *group C*, more than 50% collapse of the lateral pillar.

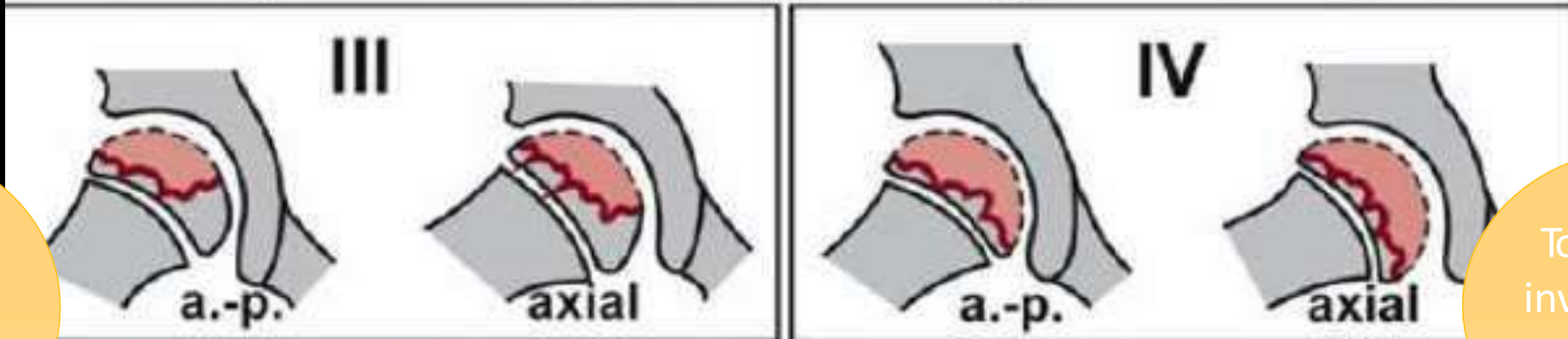
Catterall Classification

Anterior epiphysis only involved



Involvement of anterior epiphysis with central sequestrum

Most of the epiphysis is "sequestrated"



Total head involvement



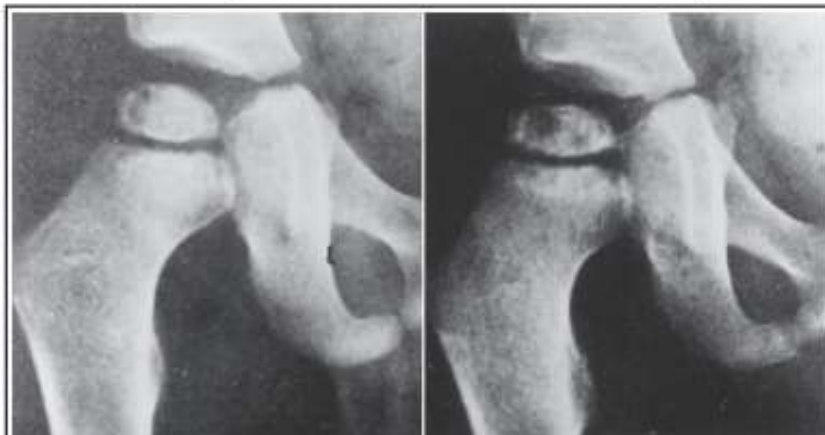
A Group I



AP



Frog-leg lateral
Group III



At onset

After 4 months

B Group II



D Group IV

Salter-Thompson Classification

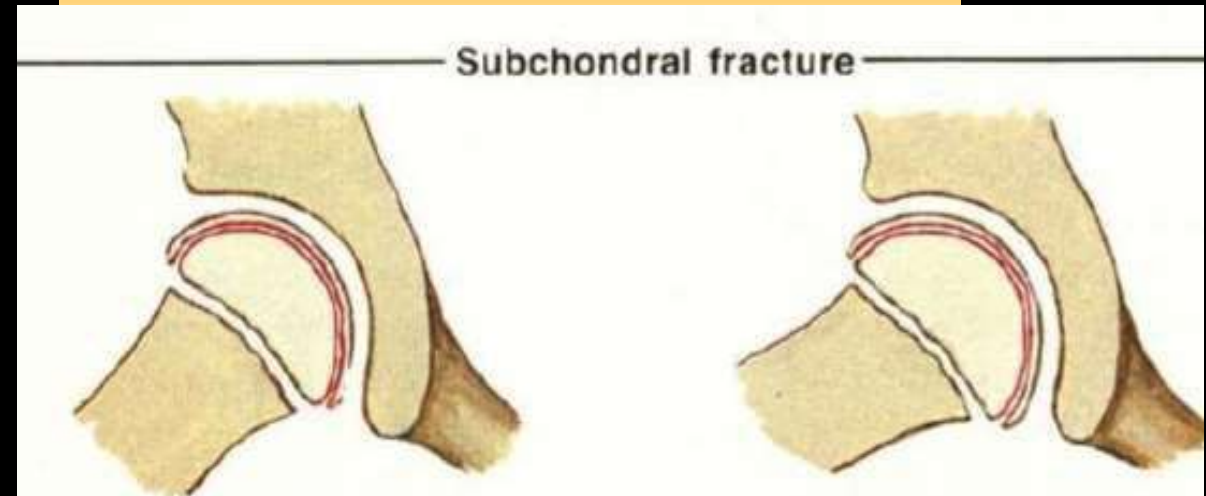
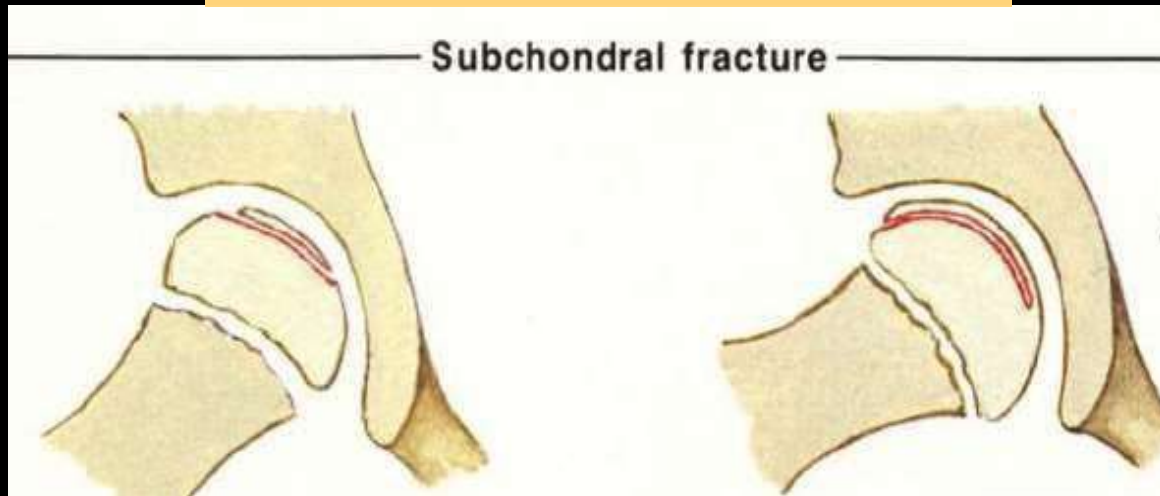
Based on extent of subchondral fracture

Grp A:

<1/2 head involvement

Grp B:

>1/2 head involvement



Classification of End Results

1) Mose classification

Based on the fitting contour of healed femoral head to a template of concentric circles

Deviation in Shape	Outcome
<1 mm	Good
1-2 mm	Fair
>2 mm	Poor

2) Stulberg Classification

Table 14.4 Modified Stulberg Classification System.¹⁴⁵

Group	Radiographic Findings ^a
I	Femoral head normal
II	Femoral head round, within 2 mm of circle, same circle both views
III ^b	Femoral head ovoid, acetabulum matches head
IV ^b	Femoral head flattened more than 1 cm on weight-bearing areas, acetabulum also flattened
V ^c	Femoral head collapsed, acetabulum not flattened

^aAnteroposterior and frog-leg lateral radiographs.

^bGroups III and IV termed congruous incongruity.

^cGroup V termed incongruous incongruity.

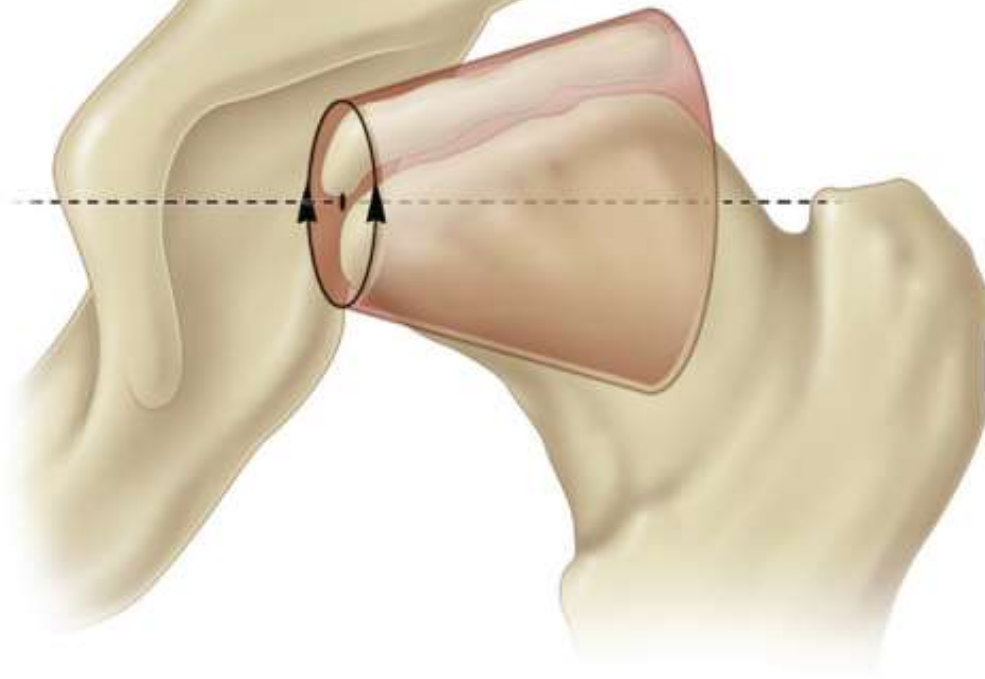


FIG. 14.39 Illustration of the concept of congruous incongruity in Stulberg group IV hips, in which the femoral head assumes a cylindrical shape. There is an almost normal range of flexion and extension but no available rotation or abduction. Often the hip externally rotates with flexion and returns to a neutral position with extension. This motion has been likened to the movement of a cow's hip, which has a similar shape. (Redrawn from Schultz KP, Dustmann HO. Morbus Perthes: Ätiopathogenese, differential diagnose, therapie und prognose. Berlin: Springer; 1991.)



FIG. 14.38 A hip with a Stulberg III result. Anteroposterior radiograph obtained 11 years after onset of the disease shows the femoral head fully reossified and ovoid. (From Herring JA. *Legg-Calvé Perthes Disease*. Rosemont, IL: American Academy of Orthopaedic Surgeons; 1996.)



FIG. 14.40 An example of a Stulberg V result. The femoral head is irregular, with hills and valleys, whereas the acetabulum has a smooth contour.

Poor Prognostic Risk Factors

- Extent of uncovering of the femoral head,
- Catterall group III or IV, lateral calcification,
- Lateral head displacement (using the head-to-teardrop Distances)
- Widening of the femoral neck in the early stages of the disorder.

Box 14.6 Differential Diagnosis of Legg-Calvé-Perthes Disease

KNOWN CAUSES OF AVASCULAR NECROSIS

Sickle cell disease

Other hemoglobinopathies (e.g., thalassemia)

Chronic myelogenous leukemia¹⁵⁹

Steroid medication

Sequela of traumatic hip dislocation

Treatment of developmental dysplasia of the hip

Septic arthritis

SKELETAL DYSPLASIAS MIMICKING PERTHES

Multiple epiphyseal dysplasia

Spondyloepiphyseal dysplasia

Mucopolysaccharidoses

Hypothyroidism

OTHER SYNDROMES

Osteochondromatosis

Metachondromatosis

Schwartz-Jampel syndrome

Trichorhinophalangeal syndrome

Maroteaux-Lamy syndrome

Martolf syndrome⁸¹

Stickler syndrome³⁴¹

Multiple epiphyseal dysplasia

Always bilateral

Symmetric involvement

Acetabular changes
are usually seen

Metaphyseal cysts absent

Changes in other joints
like shoulders and knees

Perthes disease

Bilateral in a small
proportion

If bilateral, usually
metachronous
findings with both
hips at different
stages of the disease

Acetabular changes
uncommon

Metaphyseal cysts
are common

Other joints are
normal

TREATMENT

CURRENT TREATMENT APPROACH

- Latest treatment approaches are based on the age of the patient.
- We categorise the patient into 4 age groups-
 - 1) Below 6 years
 - 2) 6 to 8 years
 - 3) 8 to 11 years
 - 4) >11 years

Below 6 years of age:

- Management focuses upon:
 - -Pain relief
 - -Short-term use of anti-inflammatory medications
 - -Bed rest
- Operative treatment like osteotomies have no added benefit.

6 to 8 years of age:

- Best management is controversial.
- Lateral pillar A- Symptomatic treatment
- Lateral pillar B- Petrie cast for 6 weeks, followed by Femoral varus osteotomy or wide abduction brace.

8 to 11 years of age:

- Consideration of surgical treatment if child presents after 8 years of age, in the initial stage with more than 50% head involvement.
- -Femoral varus osteotomy (62% success rate)
- -Salter inominate osteotomy (41% success rate)
- -Combined Femoral and Salter osteotomy
- -Triple pelvic osteotomy
- Non-operative treatment for Lateral column type C

>11 years of age:

- Success rate is not predictable in this age group, regardless of treatment modality due to poor healing and remodelling potential.

Symptomatic Therapy:

- The primary means of treating symptoms related to LCPD are bed rest (with or without traction) or local rest by non–weight bearing on the affected hip.
- A wheelchair, crutches, or a walker is prescribed for the non– weightbearing treatment.
- Short-term use of NSAIDs drugs for pain and discomfort may also be
- of benefit.
- Prolonged use of these drugs is best avoided given that they may negatively affect new bone formation

Concept of Containment Treatment

- The objective of containment is hold the femoral head in the acetabulum during the period of “**biological plasticity**” while necrotic bone is resorbed and living bone is restored through “**creeping substitution**” so that during repair and remodelling the head becomes almost spherical.



Broomstick plasters/ Petrie casts

Maintains the hips in approximately 45 degrees of abduction and 5 to 10 degrees of internal rotation with the knees slightly flexed.

The patient walks by using crutches in front and back

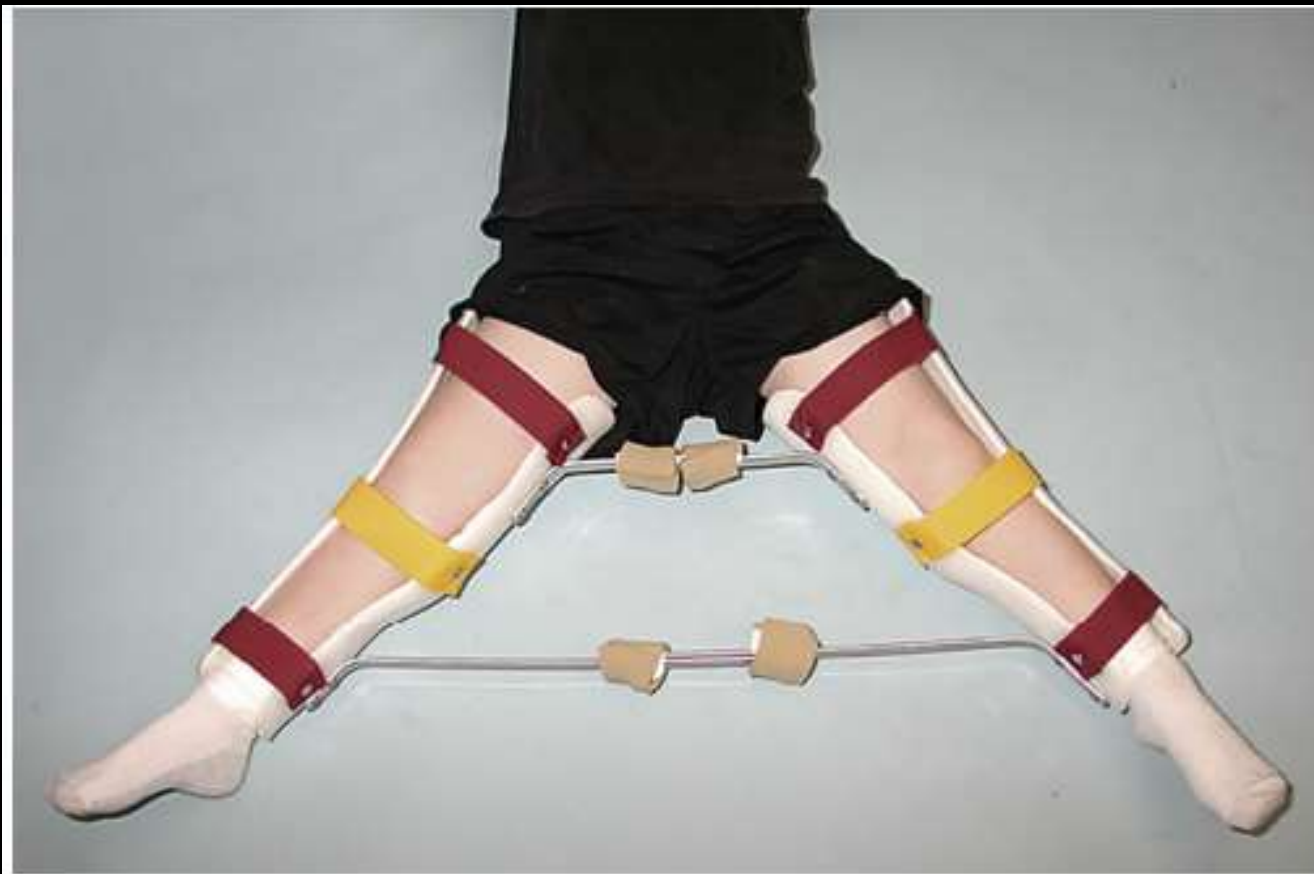
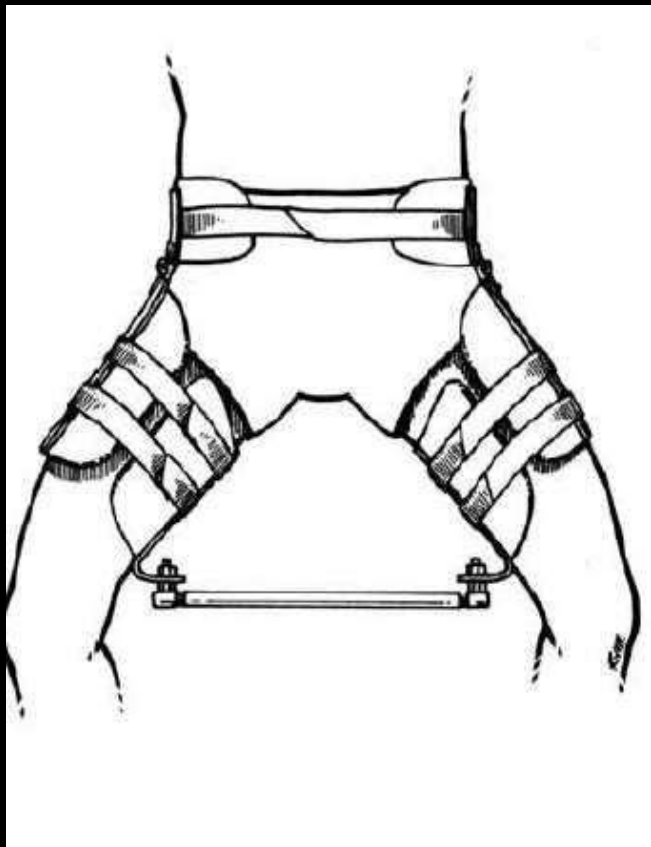


FIG. 14.45 Photograph of a patient wearing a wide abduction brace called an A-frame brace. This brace is used at home after school and at night time after the removal of Petrie casts to maintain good hip abduction. An in-brace radiograph is obtained at the initiation of bracing to confirm that the affected femoral head is contained. This brace is to be used while the patient is recumbent. It is recommended that the brace is worn for 12 hours per day initially. If a good hip abduction is maintained, use of the brace can be decreased to 8 hours per day and then discontinued.

Atlanta Scottish Rite Brace





Toronto brace, with its universal joints, was designed to keep the hip abducted while allowing hip and knee flexion. Thus the patient can both sit and stand

Evaluation of the Concept of Containment Treatment

- To assess the dynamic effects of various containment devices, Rab conducted gait analyses in children with LCPD.
- Use of Petrie casts resulted in an increase in anterior and lateral coverage of the femoral head and a reduction in posterior and medial coverage.
- By establishing hip flexion and external rotation of the limb, the Atlanta brace provided greater posterior coverage than lateral coverage.

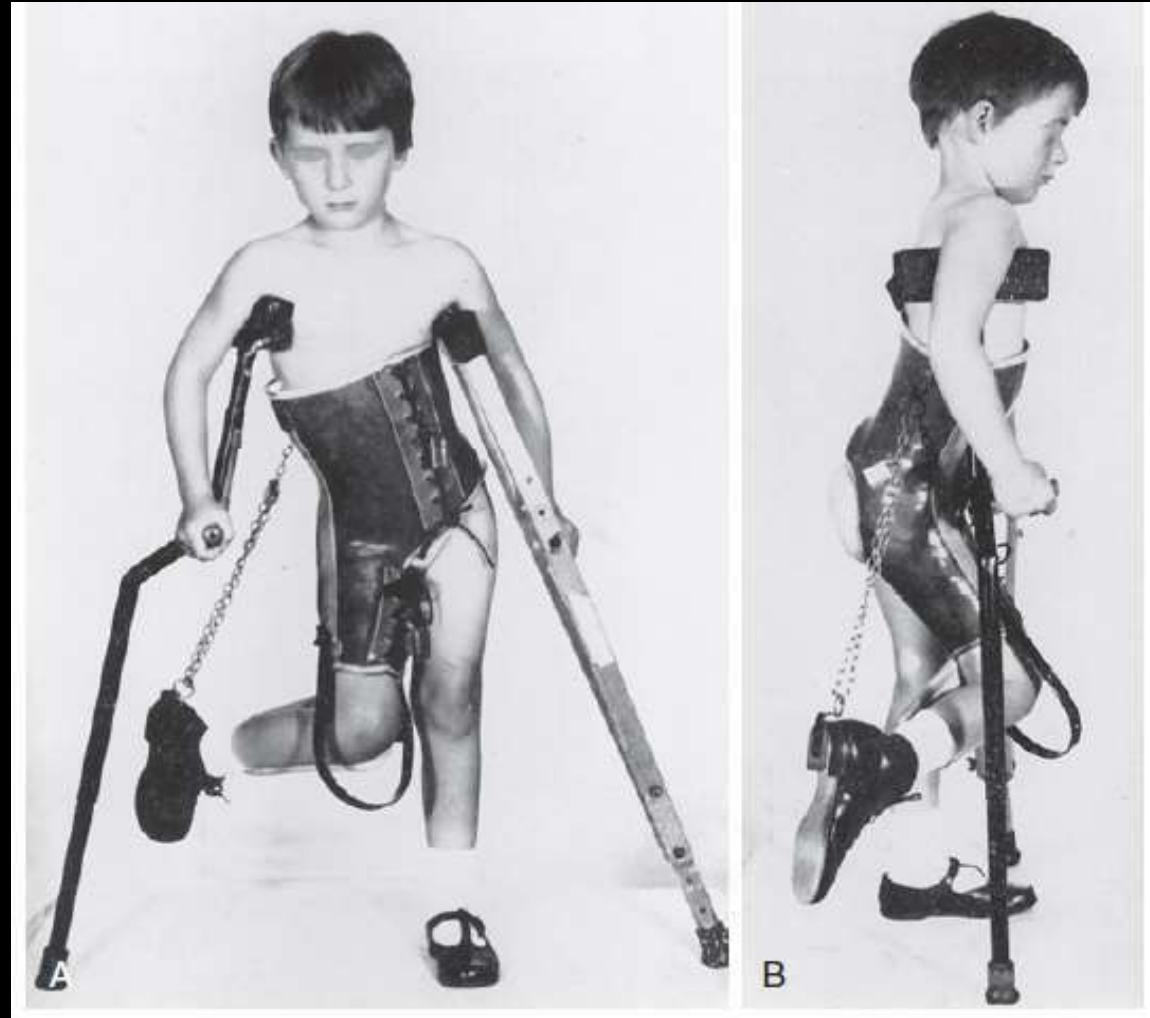
Variable	Indications for containment	Contraindications for containment
Age	<p>In children under 7 years at onset if there is demonstrable extrusion</p> <p>In children over 7 years contain early (even before any extrusion is evident) as extrusion almost invariably occurs in these older children</p>	In children under 7 years without any extrusion
Extent of involvement	If half or more than half of the epiphysis is avascular	If less than half of the epiphysis is avascular
Stage of the disease	Stage of avascular necrosis or the stage of fragmentation	Stage of regeneration or healed stage
Extrusion	<p>If extrusion is present (in child under 12 years at onset of disease)</p> <p>OR</p> <p>Even if extrusion is absent in children aged between 7 and 12 years</p>	If extrusion is absent (in children under 7 years at onset of disease)
Range of hip motion	Normal	Restricted

Nonsurgical Containment Using Orthotic Devices



Snyder sling

Birmingham brace



Newington brace



SURGICAL CONTAINMENT

1) Femoral Varus Osteotomy

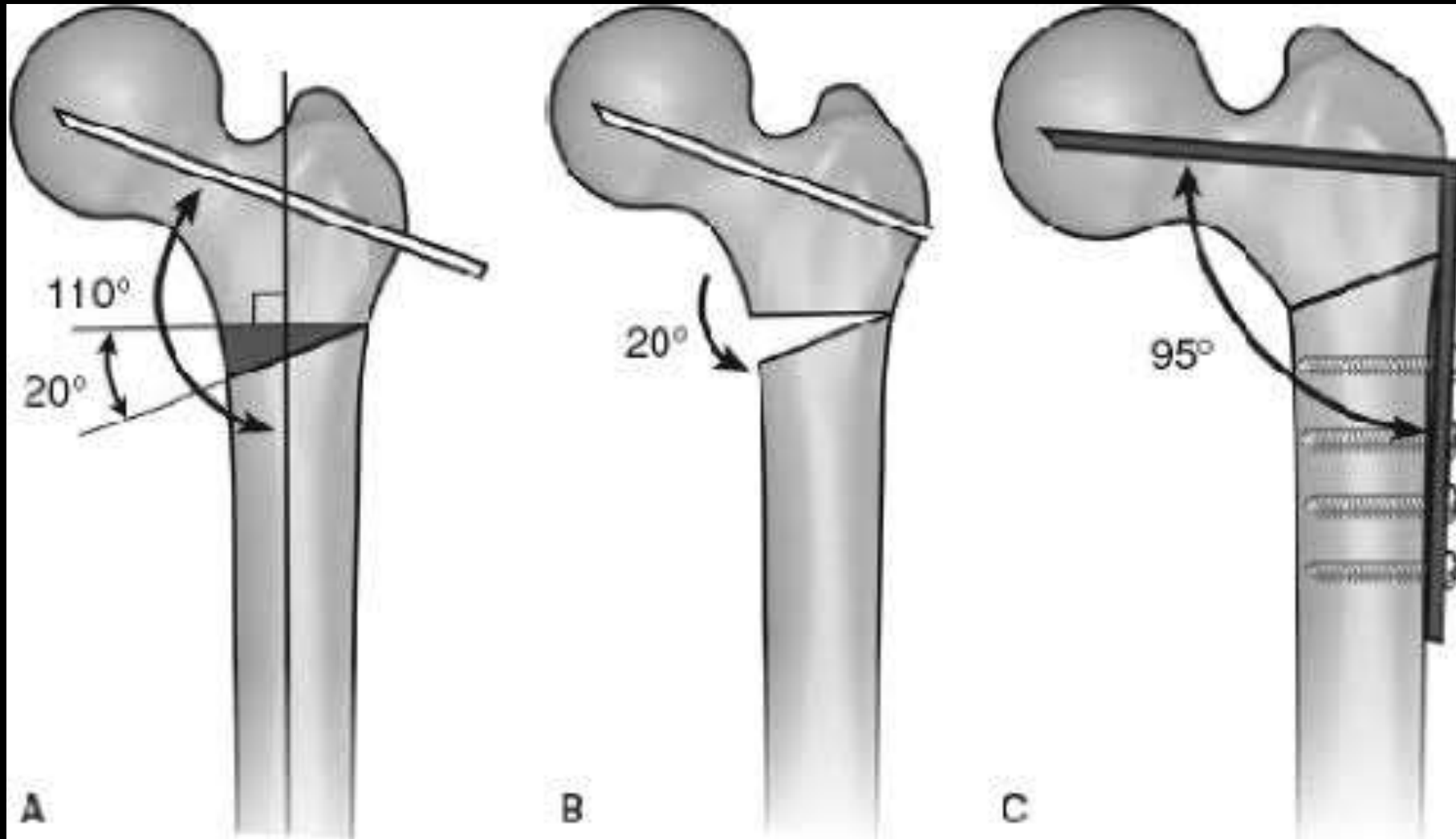
- Indications:
- >8 years of age at onset
- Lateral pillar type B or B/C in fragmentation stage.

- Before femoral osteotomy, patient should have a reasonable range of motion.

- 10 to 15 degree correction in early stage disease and greater degree done in later stage of the disease.

Complications:

- Excessive post-operative varus
- Failure of varus to remodel
- Persistent external rotation of limb even after rotational osteotomy
- Shortening of extremity
- Trochanteric overgrowth
- Fracture while removing plate
- Delayed union
- Non-union



FEMORAL VARUS OSTEOTOMY



2) Innominate Osteotomy(Salter's)

Indications:

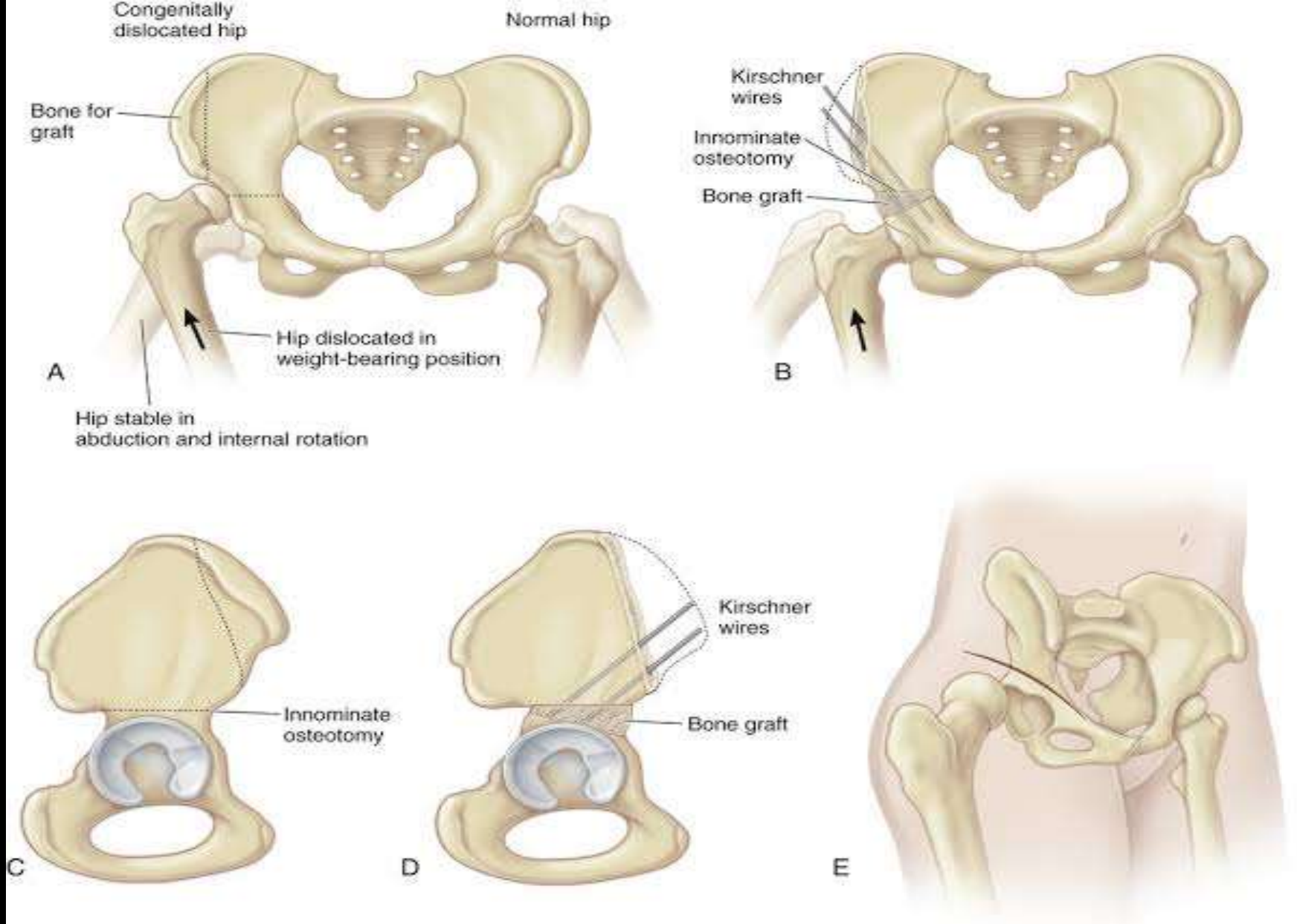
- Onset after 6 years of age
- Moderate to severe affected head
- Failure of containment.

Pre-requisites:

- Minimum deformity
- Non-irritable hip
- Hip has to be able to abduct to 45 degrees
- Good range of motion.

Complications:

- Loss of fixation with displacement of distal fragment
- Lengthening of the leg
- Decreased hip flexion
- Joint stiffness
- Hinge abduction



INOMINATE/ SALTER'S OSTEOTOMY



3) Triple Pelvic Osteotomy

Combines standard Salter's inominate osteotomy with high ischial and high pubic osteotomy near the acetabulum

Indications:

- Loss of containment
- Requirement of abduction >25 degree

3) Combined Femoral and Inominate Osteotomy

Indications:

- Lateral subluxation
- Lateral calcification and metaphyseal changes

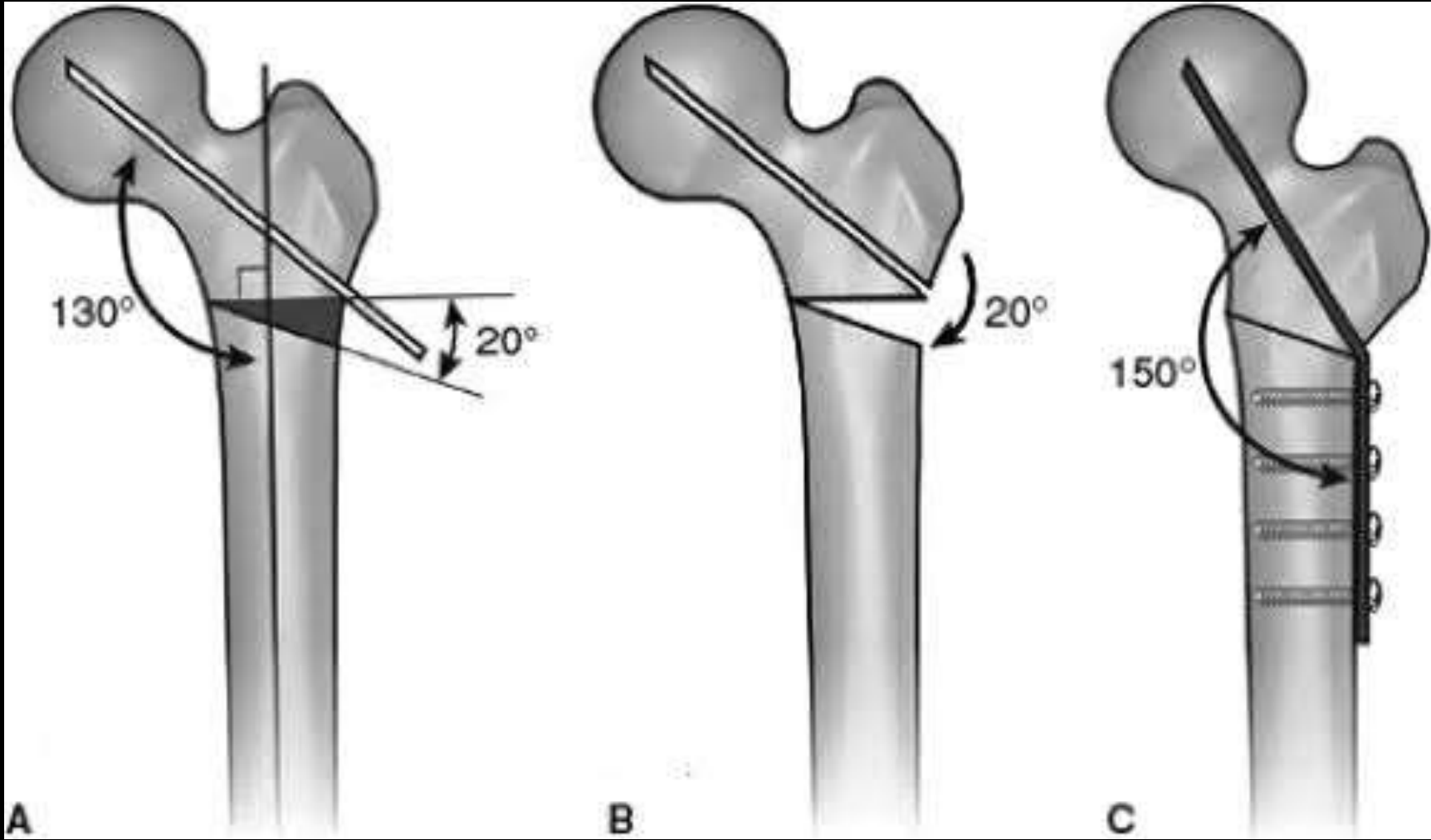
4) Valgus Osteotomy

Indications:

Treatment of hinge abduction due to deformed femoral head

Advantage:

Improves the roundness of femoral head



VALGUS OSTEOTOMY



5) Shelf Arthroplasty

Indications:

- Lateral subluxation of femoral head
- Insufficient coverage of femoral head
- Hinge abduction of hip

Shelf procedure- Gill technique
- Staheli technique

SHELF ARTHROPLASTY



Complications:

- Loss of hip flexion secondary to an excessively wide augmentation graft
- Inadequate hip coverage due to thin graft
- Dysaesthesia of lateral femoral cutaneous nerve

6) Chiari Osteotomy

Indications:

- Usually reserved for treating the healing femoral head that remains lateralised.
- Older children with painful hip, significant head deformity and incongruity between head and acetabulum.

CHIARI PELVIC OSTEOTOMY



Conservative

- Physiotherapy exercises, mobilization under anesthesia, botox injections
- Abduction splint, braces and casts

Operative

- Improving range of motion by soft tissue procedures (tenotomies, etc.)
- Femoral side osteotomies (valgus or varus trochanteric)
- Pelvis (pelvic side (salter, dega or pemberton, triple, Chiari, etc.)
- Combination of above

Fig. 3: Methods of containment of hip in Perthes disease with subluxation

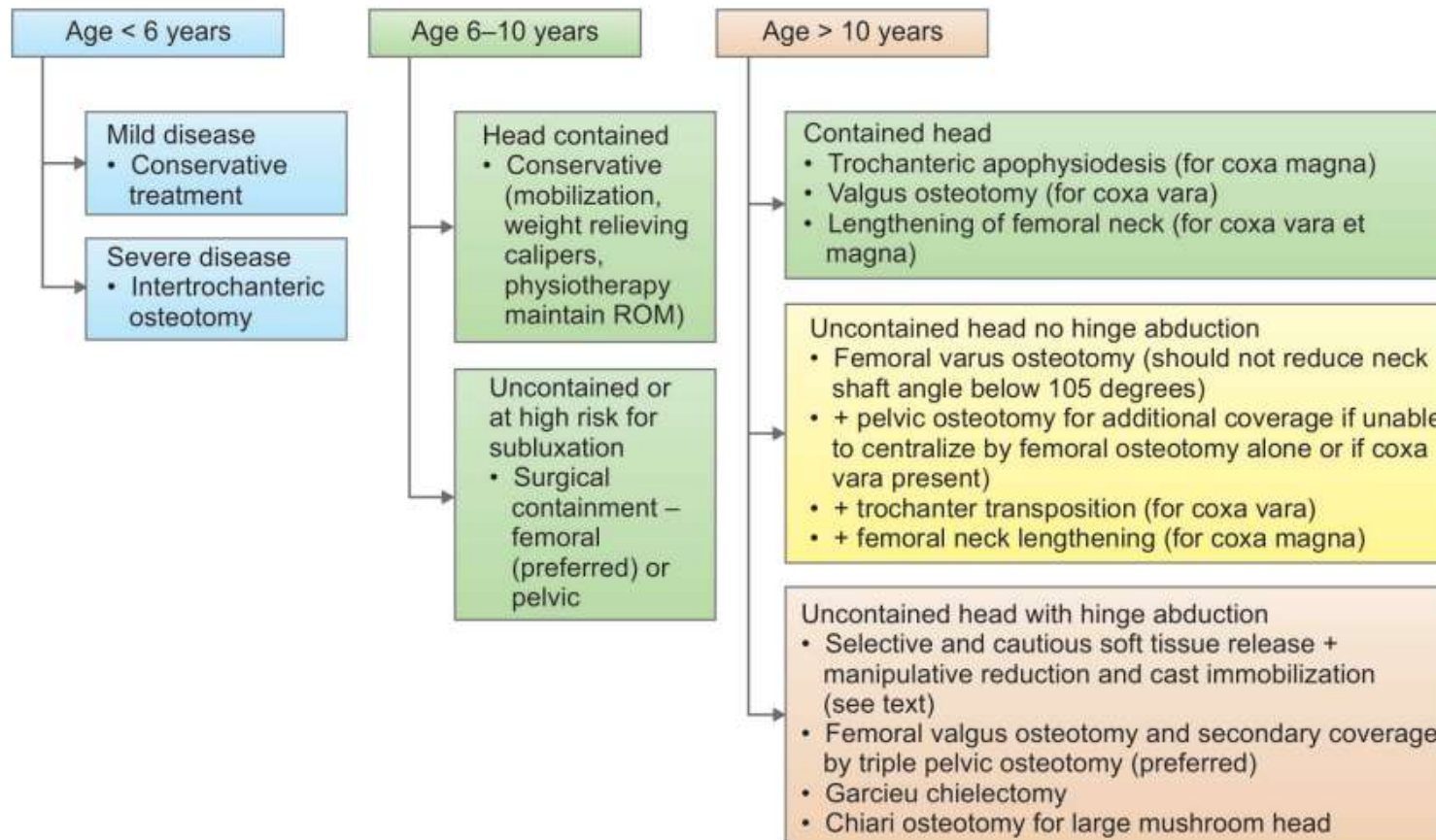


Fig. 4: General treatment algorithm for management of Perthes disease in different age groups

SEQUELAE OF PERTHES DISEASE

- Osteochondrotic lesion
- Femoroacetabular impingement
- Osteoarthritis

THANK YOU