Legg-Calve-Perthes Disease

Osteonecrosis of the proximal femoral epiphysis in a growing child with subsequent remodelling of the regenerated bone.

- In 1910 independently described by
  - Legg in USA
  - Perthes in Germany
  - Calve in France
ETIOLOGY

- UNKNOWN
- Arterial- obstruction of intra and extra osseous vessels
- Venous – abnormal venous drainage, increased metaphyseal pressure
- Disturbance of clotting mechanism, Hypofibrinolysis
- Transient Synovitis
  - 4% patients develop perthes

- OTHER THEORIES
  - inflammation
  - infection
  - trauma
Incidence

- Affects 1 in 9000 children
- More common in boys 4:1
- 80% age 4 – 8 years (2 – 12)
- Familial in 10%
- Both hips in 10 – 12%
- More common in lower socioeconomic
- Rare in black africans
Pathophysiology

- Blood supply to Fetal Femoral Head
  metaphyseal, epiphyseal vessels, Ligamentum teres
- Metaphyseal supply gradually decreases at age 4
- Vessels in Ligamentum teres develop at age 7
- Between age 4 – 7 blood supply depend solely on lateral epiphyseal vessels,
  susceptible to external pressure, hip effusion
  increased intracapsular pressure, vessel occlusion

Repeated episodes result in AVN
Clinical Features

- Typically a boy 4-8 year c/o groin, inner thigh or knee pain and start limping.
- Painless intermittent limp.
- Limitation hip movements IR / Abd.
- Catteral`s sign – passive hip flexion cause ER.
- Trendelenburg Gait
- Short Limb
- Wasting of thigh and buttock.
Investigations

- Radiographs  AP  Frog Lat View
- Isotope Bone Scan.  T99 , Demonstrate avascularity of femoral head before any radiological feature is evident. Useful for diagnosis and monitoring.
- MRI Scan . Sensitive to detect infarction and extent of necrosis.
- Arthrography. Help to determine position of containment and osteotomy.
4 Stages of perthes (2 – 4 years)

1- Ischaemia/Necrosis
   Part of femoral head dies, Head stops enlarging, cartilage still nourished, widening of medial joint space, dense femoral head.

2- Fragmentation/ Resorption
   Trabeculae in dead bone fracture    CRESCENT SIGN
   Lysis of bone suprolateral corner   GAGE`S SIGN
   return of blood supply           Femur head separate into pieces

3- Reossification/ Healing
   New bone formation and Femoral Head take a new shape
   Lucent areas are replaced by New Woven Bone.

4- Remodelling/ Deformity
   Femoral head is fully reossified    Repair process completed
   X-Ray shows residual deformity, Flattening and lateral displacement of epiphysis
   Coxa magna, Gradual remodelling untill maturity, Acetabulum remodels
   Coxa breva
Crescent sign

Subchondral radiolucent line in lat view

The length of subchondral line indicates extent of femoral head involvement
Gage’s sign

Lytic area on lat aspect of epiphysis and adjacent metaphysis

Indicates poor prognosis
Sagging rope sign

Sclerotic line curving across
Old Perthes
Femoral neck due to distortion
And remodelling of fem head
Head at Risk: RADIOLOGICAL

- Gage`s Sign
- Calcification lateral to epiphyses
- Metaphyseal Cyst Formation
- Lateral Subluxation
- Horizontal Physis
- Uncovering of the Femur Head
  > 3mm in excess of opposite side
Head at Risk: CLINICAL

- Progressive loss of movement
- Females have worse prognosis than males
- Bone age > 6 years at presentation
- Adduction contracture
- Flexion with abduction
- Heavy child
Herring’s Lateral pillar classification

Group A
Normal ht lat pillar

Group B
Lat pillar maintained 50 % ht

Group C
Loss of lat pillar ht > 50 %
Catterall Classification

Group I
- No metaphyseal reaction
- No sequestrum
- No subchondral fracture line

Group II
- Sequestrum present—junction clear
- Metaphyseal reaction—anterolateral
- Subchondral fracture line—anterior half

Group III
- Sequestrum—large—junction sclerotic
- Metaphyseal reaction—diffuse anterolateral area
- Subchondral fracture line—posterior half

Group IV
- Whole head involvement
- Metaphyseal reaction—central or diffuse
- Posterior remodelling
Deformities At Maturity

- Coxa Magna
- Hanging Rope Sign
- Coxa Breva
- Coxa Irregularis
- Osteochondritis dissecans
Stulberg Classification of the End Result *(JBJS 1981. 63-A:1095-1108)*

<table>
<thead>
<tr>
<th>Class</th>
<th>Description</th>
<th>Arthritis Outcome</th>
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</thead>
<tbody>
<tr>
<td>Class 1</td>
<td>completely normal</td>
<td>Arthritis does not develop</td>
</tr>
<tr>
<td>Class 2</td>
<td>Spherical (&lt; 2mm). Coxa Magna. Short Neck. Increased Sharp angle.</td>
<td>Arthritis does not develop</td>
</tr>
<tr>
<td>Class 3</td>
<td>Elliptical &gt; 2mm, but not flat</td>
<td>Mild to moderate OA</td>
</tr>
<tr>
<td>Class 4</td>
<td>Flat femoral head, flat acetabulum</td>
<td>Mild to moderate OA</td>
</tr>
<tr>
<td>Class 5</td>
<td>Flat femoral head, round acetabulum.</td>
<td>Severe OA &lt; 50yrs age</td>
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Mose Sphericity Measurement

- Concentric ring Template
- 2 mm spacing
- Exact fit 1 circle – Good
- < 2mm – Fair
- > 2mm – Poor
## Treatment

- **Aim** – Resolve symptoms and get spherical, well covered femoral Head (containment), maintain range of motion.
- Bed rest, restricted activity
- PWB NWB with crutches
- Skin Traction
- NSAID
- Physical Therapy
- Hip Containment (Abd. Brace, Muscle Release, Osteotomy)
- Abduction Devices  
  - Cast/Braces/Splint
  - Petrie Abduction Cast/Atlanta Scottish Cast/Broomstick Cast
Petrie Cast
Operative Containment

- Pelvic Osteotomy
  - salter
  - chiari
  - shelf
- Proximal Femoral Osteotomy, Varus Osteotomy
- Late Surgery for Deformity
  - cheilectomy (over growth greater trochanter)
- Late Surgery for Osteoarthriticis
Treatment recommended by Herring (JBJS 76A 448-458, 1994) & Alder Hey:

<table>
<thead>
<tr>
<th>Age Range</th>
<th>Treatment</th>
<th>Description</th>
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<tbody>
<tr>
<td>&lt;6yrs</td>
<td>Observe</td>
<td></td>
</tr>
<tr>
<td>6-8yrs</td>
<td>Observe</td>
<td>Herring A</td>
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<tr>
<td></td>
<td></td>
<td>Herring B pts with bone age &lt; 7yrs</td>
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<tr>
<td></td>
<td>Containment</td>
<td>Herring B with bone age &gt; 7yrs</td>
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<tr>
<td></td>
<td></td>
<td>Herring C</td>
</tr>
<tr>
<td>&gt;8yrs</td>
<td>Surgery</td>
<td>Most are Herring B/C</td>
</tr>
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