LEGG-CALVE’PERTHES DISEASE

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Perthes’ disease is a disorder of childhood, describes idiopathic avascular necrosis of the proximal femoral epiphysis.

Incidence is only 1 in 1000 in children, it should always be considered in the differential diagnosis of hip pain in young children.

Patients are usually 4–8 years old and often show delayed skeletal maturity;

boys are affected five times as often as girls.

Bilaterally affected hips are seen in approximately 10%, more common in girls (In bilateral disease hips do not follow the same time course and are often in different stages of the disease process).
- Risk factors:
  low birth weight
  exposure to second hand cigarette smoke,
  short body length at birth,
  family history,
  low socioeconomic status

Pathophysiology:
The pathophysiology of LCPD is not fully understood but it may include a vascular insult and uncoupling of the bone metabolic process.
Femoral Head Blood Supply – In Pediatric Age Group

- Up to the age of 4 months, the femoral head is supplied by:
  - (1) **metaphyseal vessels** which penetrate the growth disc
  - (2) **lateral epiphyseal vessels** running in the retinacula
  - (3) scanty vessels in the ligamentum teres.
The metaphyseal supply gradually declines until, by the age of 4 years, it has virtually disappeared; by the age of 7, however, the vessels in the ligamentum teres have developed.

Between 4 and 7 years of age the femoral head may depend for its blood supply almost entirely on the lateral epiphyseal vessels whose situation in the retinacula makes them susceptible to stretching and to pressure from an effusion.

The precipitating cause is probably an effusion into the hip joint following either trauma, of which there is a history in over one-half of the cases, or a non-specific synovitis.
Pathology: -

- The **pathological process takes 2–4 years to complete**, passing through three stages.

- **Stage 1**: bone death: following one or more episodes of ischaemia, part of the bony femoral head dies; it still looks normal on plain x-ray but it stops enlarging.

- X-ray → subtle increase in space between acetabular socket and head of femur, even though it may appear normal!

- **Subtle Widening of joint space**

  - **Stage 2**: revascularization and repair: new blood vessels enter the necrotic area and new bone is laid down on the dead trabeculae, producing the appearance of increased density on the x-ray. Some of the necrotic parts are resorbed and replaced by fibrous tissue (producing the x-ray appearance of epiphyseal ‘fragmentation’)

- **Stage 3** (distortion & remodeling)
# Symptoms and Signs:

- The patient – usually a boy of 4–8 years – complains of:
  - Hip, knee or groin pain exacerbated by movement.
  - Reduced range of motion, particularly in abduction & internal rotation.
  - Limping Child./ antalgic gait
  - Atrophy of thigh muscles may occur from disuse and an inequality of leg length.
Diagnosis:-

➤ (x-ray)

➤ • early changes:
  - widening of the joint ‘space’.
  - Joint effusion
  - increased radiographic density in the bony epiphysis (classic feature ).

➤ • later change:
  - Flattening, false ‘fragmentation’
  - lateral displacement of the epiphysis with rarefaction and broadening of the metaphysis (Reduction in density of metaphyseal bony tissue).
  - Varus deformity of the femoral neck( angle between the neck and the shaft of the femur is reduced to less than 120 degrees(normal femoral neck–shaft angle is 160 degrees at birth, decreasing to 125 degrees in adults)…..occurs when the bone tissue in the neck of the femur is softer than normal, causing it to bend under the weight of the body
prognosis

- Various prognostic grading systems are employed, based mainly on x-ray appearances.

- **Herring classification** is based on the severity of structural disintegration of the lateral pillar of the femoral epiphysis.
**TREATMENT** (The primary aim of treatment is to prevent deformity to the femoral head before the remodeling phase)

- As long as the hip is painful the child should be in bed, with severe symptoms sometimes requiring a period of **skin traction** applied to the affected leg protect the shape of the femoral head, and restore normal hip movement. Once Pain has subsided (3 weeks) movement is encouraged

  - /containment

  - Orthotic devices (bracing)

  - Surgery (Osteotomy),

  - Note: The main long-term problem with this condition is that it can produce a permanent deformity of the femoral head, which increases the risk of developing osteoarthritis in adults
DIFFERENTIAL DIAGNOSIS

• The commonest cause of hip pain in children is a non-specific transient synovitis – the so-called *irritable hip*.

• Diagnosis: Ultrasound may show a joint effusion, but the x-rays are always normal.

• Symptoms: last for 1–2 weeks and clear up completely.

• The child should be kept in bed until pain disappears and the effusion resolves.
Thank you