A 4 year old with hip pain:
Legg-Calvé-Perthes Disease

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Our Patient

- A 4 year-old boy is complaining of severe L hip pain.
- The differential diagnosis of acute hip pain in children is quite broad. The categories include Tumor, Trauma, Infection, Inflammatory processes, or Infarction/mechanical derangement.
- Does this child require imaging? What would be the appropriate radiographic evaluation of this patient?
Imaging Modalities for Acute Hip Pain

- Radiographic evaluation is **necessary** in all patients with septic arthritis, skeletal injury, or tumor in the differential diagnosis.

**Diagram:**
- Plain Film
  - Ultrasound
    - MRI
    - CT
  - Bone Scan
Acute Hip Pain: Plain Films and Ultrasonography

- **Plain radiographs** - AP views of the pelvis and frog-leg views (lateral view of femur)
  - Identify bony aberrations/abnormalities, but may miss small effusions

**After Plain radiographs:**

- **Ultrasonography** -
  - If plain film is normal and you are suspicious of septic arthritis or synovitis, it can be used to identify small effusions.
  - If plain film is abnormal, it can be used to guide arthrocentesis.
Acute Hip Pain: Radionucleotide Scan

Radionucleotide scan-

- **Acute setting**: can be used to differentiate joint inflammation (septic arthritis or tissue synovitis) from osteomyelitis.
- **Chronic setting**: can identify avascular necrosis before abnormalities are visualized on plain film, as well as early tumors and myelodysplastic disease.

**Evidence Based?** 50 children with hip pain were evaluated in a prospective study that probed the usefulness of imaging protocols in the diagnosis of hip pathology. In this protocol of plain films followed, as needed, by ultrasound and three-phase radionucleotide scans, the diagnosis of 48 patients was successfully identified. *(Clinical Pediatrics 1988)*
Acute Hip Pain: MRI and CT

MRI and CT can be used if:

a) the other modalities have not identified a diagnosis
b) to provide better detail of a diagnosed abnormality

MRI - can identify signs of osteomyelitis, early Legg-Calvé-Perthes disease, early Slipped Capital Femoral Epiphysis and cartilage destruction.

- In some cases, MRI with contrast may be preferable to bone scan, though it faces complications of availability, cost, and need for sedation.

CT - may identify an intraabdominal cause of hip pain, such as appendicitis or psoas abscess.
Our Patient: Frontal Pelvis

- He received an AP film of his pelvis.
- What is the abnormality on the radiograph?

Image courtesy of Dr. Jennifer Song
Normal Hip Anatomy

Normal Blood Supply and Avascular Necrosis

Blood Supply to Head and Neck of Femur
- Head
- Neck
- Branch of the obturator artery
- Lateral femoral circumflex artery
- Medial femoral circumflex artery

Avascular necrosis

Normal Head
- Cartilage

Avascular Necrosis Head
- Necrotic bone

http://www.zimmer.co.uk/ctl?template=PC&op=global&action=1&id=7997


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There is:

- **widening** of the femoral head
- **flattening** of the femoral head (coxa plana)
- The infarction has extended across the growth plate and a **radiolucent lesion** is evident within the metaphysis
- In addition, the growth center of the femoral head has been damaged so that normal growth is arrested, and **shortening of the femoral neck** results.

AP View Pelvis

Image courtesy of Dr. Jennifer So
# Differential Diagnosis for Femoral Head Irregularity and Collapse

<table>
<thead>
<tr>
<th>Bilateral</th>
<th>Unilateral</th>
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<tbody>
<tr>
<td>Hypothyroidism</td>
<td>Legg-Calvé-Perthes-Disease*</td>
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<tr>
<td>Multiple Epiphyseal Dysplasia</td>
<td>Septic Arthritis</td>
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<tr>
<td>Spondyloepiphyseal dysplasia tarda</td>
<td>Spondyloepiphyseal dysplasia tarda</td>
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<tr>
<td>Sickle Cell</td>
<td>Sickle Cell</td>
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<td>Gaucher’s Disease</td>
<td>Gaucher’s Disease</td>
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<tr>
<td>Meyer’s Disease</td>
<td>Meyer’s Disease</td>
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<td>Eosinophilic granuloma</td>
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*Can be bilateral in 10-20% of cases

transient synovitis
Legg-Calvé-Perthes Disease (LCPD)

- Idiopathic osteonecrosis of the femoral head described independently in 1910 by Legg, Calvé, and Perthes.
- Rare, affecting 1 in 1200 children
- Affects mostly males, only about 1:4-5 are girls.
- About 10-20% of all diagnosed develop the disease in both hips.
  - Most of these children are very active and often very athletic.
- The age of diagnosis is usually between 2 and 12 years old, with the average age of 6.
- Legg-Calvé-Perthes children tend to be of shorter stature.
Pathophysiology of LCPD

- Approximately 10 percent of cases are familial
- Symptoms include painless limp, pain, and restriction in movement at hip.

**Proposed causes: (controversial)**
- An unusually high frequency of factor V Leiden and inherited coagulopathies has been noted in some reports among patients with LCP, suggesting thrombophilia as a contributor to avascular necrosis.
- Structural abnormalities of epiphyseal cartilage
- Association with prenatal secondhand smoke exposure and birth weight less than 2.5 kg in boys.
Evolution of LCPD

- Early on, you can have normal frontal pelvis radiographs.

- **Three Stages of LCPD:**
  1. Ischemia disrupts growth and femoral head becomes more dense with possible fracture of supporting bone
  2. Fragmentation and reabsorption of bone
  3. Reossification when new bone has regrown, often with residual deformity when new bone reshapes.

- Antero-lateral head most affected

Frontal Pelvis views of Femoral Head

http://nonf.org/perthesbrochure/perthes-brochure.htm
Companion Patient Frontal Pelvis

Initial Presentation: Painless Limping

○ What abnormality do you see?

At Presentation

Ten Months later

AP View Pelvis

Image courtesy of Dr. Carolynn DeBenede
Identifying Abnormalities

1. Radiolucent lesion at femoral head
2. Fragmentation and reabsorption of bone
3. Flattening, Widening and Reossification with residual deformity

At Presentation          Ten Months later

AP View Pelvis

Image courtesy of Dr. Carolynn DeBenede
Radiographic Findings in LCPD

1. Small femoral ossification nucleus
2. Lateral displacement of the femoral ossification nucleus
3. Fissuring and fracture of the femoral ossification nucleus
4. Flattening and sclerosis of the femoral ossification nucleus
5. Metaphyseal changes: widening and shortening of femoral neck
Example #1: Lateral displacement of Femoral ossification nucleus

A very early finding!

Displacement may range from 1-4mm laterally.

Seen in majority of cases

Image courtesy of Dr. Carolynn DeBenedectis
Example #2:
Fissuring and fracture of the femoral ossification nucleus

- Radiolucent areas are seen, beginning in the anterior margin of the epiphysis.
- The fracture fragment is clearly seen.

Image courtesy of Dr. Carolynn DeBenedectis.
Companion Patient:

Evolution of LCPD on Radionucleotide Scan

Normal Hip RN Scan

Legg-Calvé-Perthes RN Scan

• At this phase, radionucleotide scan shows decreased perfusion to the femoral head.
2 years later, hip radiograph showed:
- continued flattening and fragmentation of the L femoral head
- broadening of the femoral neck
- Patient had hip stiffness and limited range of motion.
Catterall Classification

- Used to predict prognosis by graded involvement of femoral head and proximal structures

- Groups I and II have a better prognosis

- Groups III and IV have a relatively poor prognosis
The principle of treatment is protection of the joint. If the joint is deeply seated within the acetabulum and normal joint motion is maintained, a reasonably good hip can result.

This can range from conservative management (NSAIDS) to surgical intervention.
He required osteotomy (to maintain femoral head within the acetabulum).
While femoral head is still somewhat flattened, it is mostly covered by the acetabulum.
References

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