Legg-Calvé-Perthes Disease — Symptoms and Classification

See online here

Unilateral or bilateral aseptic skeletal necrosis of the femoral head has a typical onset in early childhood. A common symptom is knee pain upon exertion, which may lead to a limp. How do you make the diagnosis? Which therapeutic options are there for Perthes’ disease? In the following article, we present you with all the important facts which are relevant to your exams.

Definition of Perthes Disease

Perthes disease, named after Georg Clemens Perthes (1869–1927), refers to aseptic skeletal necrosis of the pediatric femoral head. Synonymous terms are Legg-Calvé-Perthes disease and juvenile femoral head necrosis. Perthes disease was described almost simultaneously in 1910, by G.C. Perthes in Germany, J. Calvé in France, and A.T. Legg in the US.

Epidemiology of Perthes Disease

Perthes disease tied with osteochondritis dissecans is the 2nd most common aseptic skeletal necrosis disorder.
The incidence of Perthes disease is 1 in 10,000. Men are much more frequently afflicted than women, with the incidence ratio between genders being 4:1. There is a clear peak incidence between ages 5–7.

Perthes disease may occur later in life. However, this is exceptional. Most cases are unilateral. In 10% of cases, both hip joints are affected.

**Note:** The typical Perthes disease patient is a boy between ages 5–7 with unilateral manifestation.

### Etiology and Pathogenesis of Perthes Disease

Despite extensive research, the cause of the disease is not fully understood. Circulatory impairment may arise in association with retarded skeletal growth, or mechanical microtraumas, or both. In bilateral cases, there is usually a delay before the 2nd hip is affected.

Primarily, a circulatory impairment of the epiphysis of the femoral head is present. The epimetaphyseal part of the femoral head is especially affected. The circulatory impairment leads to necrotic reorganization and reconstruction of ossification centers.

Research has shown that, in many Perthes disease patients, the medial circumflex femoral artery is either absent or obliterated.

Besides vascular changes, coagulation disorders or viscosity disorders can lead to
epiphyseal skeletal necrosis. Constant cycling between construction and breakdown results from bone regeneration mechanisms.

This reconstruction phase lasts for 3–4 years. At this time, the femoral head epiphysis is less capable of handling strain. In the reconstruction phase, a high risk of deformation of the femoral head is present.

Clinical Symptoms of Perthes Disease

There is usually no pain in the early stages, but family members often notice a one-sided limp. Signs and symptoms are variable and nonspecific. The classical hip pain (seen in textbooks and exams) is a rather rare symptom. More frequently, however, affected children complain of knee pain, which only presents in association with physical activities.

**Note:** In children with knee pain, one must also examine the hips to avoid incorrect or late diagnosis.

Diagnosis

Clinical examination of Perthes disease

- Limping gait
- Limited mobility of the hip joint in internal rotation and abduction
- Distinctive FABER test signs
- Leg length difference due to adduction contracture or collapse of the epiphysis
- Positive Trendelenburg’s sign
- Transient skeletal dysplasia
- No history of trauma

Radiology in Perthes disease

Four stages of Perthes disease can be discerned from pelvic radiography images as described by Lauenstein.

<table>
<thead>
<tr>
<th></th>
<th>Initial stage</th>
<th>Condensation stage</th>
<th>Fragmentation stage</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Joint effusion, joint cavity enlargement, lateralization and shrinking of the femoral head (vascular impairment)</td>
<td>Thicken of femoral head, subchondral fracture, enlargement of joint cavity, lateralization of femoral head, fused epiphyseal centers</td>
<td>Fragmentation and flattening of the epiphysis, metaphyseal cyst, pseudocysts</td>
</tr>
<tr>
<td>2</td>
<td><strong>Image:</strong> “The natural history of Perthes’ disease.” by Terjesen T, Wiig O, Svenningsen S. License: <a href="https://creativecommons.org/licenses/by/2.5/">CC BY 2.5</a></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
4 Repair stage

Reossification of the epiphysis, enlargement, and deformation of the femoral head (mushroom-shaped), shortening, widening of the femoral neck, the elevation of the trochanter

Magnetic resonance imaging (MRI)

Conventional radiography cannot be used to detect early changes. In the absence of notable X-ray features, an MRI should be used if Perthes disease is suspected.

Ultrasound

- Joint effusion
- Flattening of the epiphysis
- The appearance of epiphysis contour with irregular edges
- Fragmentation may be seen

Note: The sooner the diagnosis is made, the better the chances of recovery.

Classification of Perthes Disease

For Perthes disease, 3 classification systems have been devised, based on radiological criteria: Catterall, Salter-Thompson, and Herring classification. A 4th classification system, Stulberg and Mose is used after skeletal maturity

Catterall’s classification

In Catterall’s classification (1971), the extent of femoral head involvement is in the foreground and divides axial X-ray into 4 quadrants. Unfavorable signs include:

- laterализation of hips
- lateral calcification of epiphysis
- metaphyseal involvement
- horizontal adjustment of the epiphyseal plate
- triangular rarefaction of the lateral metaphysis (Gage sign)

<table>
<thead>
<tr>
<th>Grade</th>
<th>Features</th>
<th>Affected in %</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Anterolateral quadrant is affected</td>
<td>0-25%</td>
</tr>
<tr>
<td>II</td>
<td>Up to half of the femoral head is affected</td>
<td>25-50%</td>
</tr>
<tr>
<td>III</td>
<td>Only a dorsal part remains intact</td>
<td>50-75%</td>
</tr>
<tr>
<td>IV</td>
<td>Complete involvement of femoral head</td>
<td>75-100%</td>
</tr>
</tbody>
</table>

Salter-Thompson’s classification

Salter-Thompson’s classification (1984) concentrates, above all, on the visible subchondral fracture in axial radiographs. The extension is considered a later indication of the extent of expansion. In type A, less than 50% of the femoral head dome is affected by subchondral fracture, and in type B, more than 50% is affected.

Herring’s classification

In Herring’s classification, the emphasis is placed on the identification of the lateral 3rd of the femoral head epiphysis. Three groups are distinguishable through the integrity of the lateral pillars.

| Herring Type A | Lateral pillar is unaffected |
**Herring Type B**
> 50% of lateral pillar is intact

**Herring Type C**
< 50% of lateral pillar is intact

**Stulberg and Mose classification**
This classification is used after skeletal maturity for predicting prognosis by observing the deformity of the femoral head and its congruity in relation to the acetabulum.

<table>
<thead>
<tr>
<th>Stulberg 1</th>
<th>Normal spherical head (spherical congruency → No OA)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stulberg 2</td>
<td>Spherical head with coxa magna or steep acetabulum (spherical congruency)</td>
</tr>
<tr>
<td>Stulberg 3</td>
<td>Non-spherical head (aspherical congruency → OA late adulthood)</td>
</tr>
<tr>
<td>Stulberg 4</td>
<td>Flat head and flat acetabulum (aspherical congruency → OA late adulthood)</td>
</tr>
<tr>
<td>Stulberg 5</td>
<td>Aspherical incongruence (aspherical incongruency → OA before 50 years)</td>
</tr>
</tbody>
</table>

**Therapy**

**Note:** The goal of the treatment of Perthes disease is to maintain a range of movement. The insurance of a range of motion is fundamental to a congruent hip situation. Treatment is primarily aimed at limiting progressive deformation during the repair phase to prevent a reduction in epiphyseal capacity and the femoral head slipping out from the acetabulum (loss of containment). The initiation of treatment is dependent on the age of the affected child. Physiotherapy is necessary to achieve maximal range of movement. If there is an additional contracture of adductor muscles, abduction ability can be improved through treatment with botulinum toxin in conjunction with intensive physiotherapy.

Note: For refractory disease, biologics are being experimented as an adjunct to surgery. When disease progression does occur despite interventions, joint reconstructive surgeries are indicated to relieve pain.

**Treatment of Perthes disease in children < 6 years**
As it is not uncommon for Perthes disease to be self-limiting, a wait-and-see approach is taken for children up to the age of 6. However, regular checks are mandatory.

**Treatment of Perthes disease in children > 6 years**

- Regular checks (every 3 months)
- Abduction braces
- Surgery in stages Catterall III/Salter-Thompson type B/Herring types B and C, when there is a risk that the femoral head will slip from the acetabulum or other radiological risk signs. Possible surgical techniques are:
  - Intertrochanteric varus osteotomy
  - Salter **pelvic osteotomy**: conversion of the acetabular roof
  - Tönnis **triple osteotomy** (≥ 8 years): osteotomy of the ilium, ischium, and pubis
- Postoperatively: pelvic and leg plaster, periodic radiological checks

*Picture: “Beck Survey radiograph after triple pelvic osteotomy Tönnis II” by OrthoPat. License: [CC BY-SA 3.0](https://creativecommons.org/licenses/by-sa/3.0)*
Differential Diagnosis

The following table provides an overview of the most important differential diagnosis of Perthes disease.

<table>
<thead>
<tr>
<th>Condition</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Coxitis fugax</strong></td>
<td>Acute, self-limiting (3–10 days), often in boys &gt; 4 years, synovitis and joint effusion, no bony changes</td>
</tr>
<tr>
<td><strong>Bacterial coxitis</strong></td>
<td>Pronounced pain, joint drain/paracentesis is indicated</td>
</tr>
<tr>
<td><strong>Juvenile osteonecrosis</strong></td>
<td>Vascular necrosis with a known underlying disease (e.g., sickle cell anemia, thalassemia, coagulopathy)</td>
</tr>
<tr>
<td><strong>Septic arthritis</strong></td>
<td>Acute malaise, flexed hips, abducted and externally rotated, signs of inflammation, greater joint effusion, synovitis, bone marrow edema</td>
</tr>
<tr>
<td><strong>Juvenile rheumatoid arthritis</strong></td>
<td>Fever, positive antinuclear antibodies (ANA), rash, atrophy of the thigh muscles, synovitis, ischemic signs in the femoral head (in long-term cases)</td>
</tr>
<tr>
<td><strong>Slipped capital femoral epiphysis</strong></td>
<td>Dislocation of the femoral head in the caudal and posteromedial direction, metadiaphyseal bone marrow edema, enlargement and distortion of the growth plate</td>
</tr>
<tr>
<td><strong>Hypothyrosis</strong></td>
<td>Determine thyroid values!</td>
</tr>
<tr>
<td><strong>Pituitary dysfunction</strong></td>
<td>Proportioned growth retardation, retardation of growth zones</td>
</tr>
</tbody>
</table>

Prognosis and Course of Perthes Disease

The child’s age at the point of diagnosis is the main determinant for the course of the disease (the younger, the better). Long term studies of conservative therapy showed that serious coxarthrosis occurred in 15–20% of cases. Clinical risk factors for a prognostically unfavorable course are:

- being overweight
- severe disabilities
- female gender

Retrospective studies of later results showed that over 80% of hips showed good results up until the 4th decade of life.

Review Questions

The correct answers are located below the sources.

1. A 7-year-old boy is accompanied by his mother to your practice. Based on clinical symptoms, you suspect Perthes disease in the young patient. Which of the symptoms would most likely not be typical of juvenile femoral head necrosis?

   A. limping gait
B. restricted mobility of the hip joints for external rotation and adduction
C. four distinct characters
D. leg length difference by adduction/collapse of the epiphysis
E. positive Trendelenburg’s sign

2. A 33-year-old woman developed an autoimmune disease 9 years ago with a severe course. She has since been treated with systemic corticosteroids. For about half a year, the patient has noticed increasingly painful movement restrictions. On clinical examination, you notice that the rotation is absent at the hip, and a flexion contracture of 10° is present. Imaging measurements show a rounded femoral head and an absent joint space. What is the most likely cause of these changes in the hip joint?

A. hip dysplasia
B. coxitis fugax
C. femoral head necrosis
D. septic arthritis
E. Perthes disease

3. Alexander, a 6-year-old boy, is accompanied by his mother to a pediatric practice consultation. She reports that her son often complains of pain in the left knee while playing. Additionally, the kindergarten teacher and the mother at home has noticed that Alexander has developed a slightly limping gait. On clinical examination, you find no pathological symptoms on his left knee. However, when you test the range of motion of the hip joints, Alexander reports feeling slight pain, which leads to a radiograph of the left hip with the following diagnosis:

A. juvenile osteonecrosis
B. coxa valga
C. left-sided Perthes disease
D. left-sided hip dysplasia
E. left-sided bacterial coxitis

References

Leitlinien zum Morbus Perthes
Morbus Perthes: Diagnostische und therapeutische Prinzipien. Ärzteblatt 2009

Correct answers: 1B, 2C, 3E

Legal Note: Unless otherwise stated, all rights reserved by Lecturio GmbH. For further legal regulations see our legal information page.