Pathology

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

Aseptic Skeletal Necrosis of the Pediatric Femoral Head

Perthes’ disease (named after Georg Clemens Perthes (1869 – 1927)) refers to aseptic skeletal necrosis of the paediatric 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

Second most Common Aseptic Skeletal Necrosis Disorder

Perthes’ disease ties with osteochondritis dissecans as the second most common aseptic skeletal necrosis disorder.
The incidence of Perthes’ disease is 1 : 10,000. Males are much more frequently afflicted than females, the incidence ratio between genders is 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**

**Identifying the Causes of Perthes’ Disease**

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

There is primarily a circulatory impairment of the epiphysis of the femoral head. The epimetaphyseal part of the femoral head is especially affected. The circulatory impairment leads to necrotic reorganisation/reconstruction of ossification centers.

Research has been able to show that, in many Perthes’ disease sufferers, the medial circumflex femoral artery is either absent or obliterated.
Besides vascular changes, coagulation disorders or viscosity disorders can lead to epiphyseal skeletal necrosis. A 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, there is a high risk of deformation of the femoral head.

Clinical Symptoms of Perthes’ Disease

Signs and Symptoms of Perthes’ Disease

There is often 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, you must also examine the hips, in order to avoid incorrect or late diagnoses.

Diagnostics

Clinical Examination of Perthes’ Disease

- Limping gait
- Limited mobility of hip joint in internal rotation and abduction
- Distinctive FABER test signs
- Leg length difference resulting through adduction contracture/collapse of the epiphysis
- Positive Trendelenburg’s sign

Transient skeletal dysplasia
4 stages of Perthes’ disease can be discerned from pelvic radiography appearances as described by Lauenstein.

<table>
<thead>
<tr>
<th></th>
<th>Initial stage</th>
<th>Joint effusion, joint cavity enlargement, lateralization and shrinking of the femoral head (vascular impairment)</th>
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<tbody>
<tr>
<td>1</td>
<td>Condensation stage</td>
<td>Thickening of femoral head, subchondral fracture, enlargement of joint cavity, lateralization of femoral head, fused epiphyseal centers</td>
</tr>
<tr>
<td>2</td>
<td>Fragmentation stage</td>
<td>Fragmentation and flattening of the epiphysis, metaphyseal cyst, pseudocysts</td>
</tr>
<tr>
<td>3</td>
<td>Repair stage</td>
<td>Resossification of the epiphysis, enlargement and deformation of the femoral head (mushroom-shaped), shortening, widening of femoral neck, elevation of trochanter</td>
</tr>
</tbody>
</table>

**Magnetic Resonance Imaging**

Conventional radiography can’t be used to detect early changes. In the absence of notable X-ray features, an MRI should be used, if you suspect Perthes’ disease, and there are complex differential diagnoses.

**Ultrasound**

- Joint effusion
- Flattening of the epiphysis
- 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, three classification scheme has been devised, based on radiological criteria: **Catteral**, **Salter** and **Thompson**, and **Herring** classification. A fourth classification: **Stulberg** and **Mose** classification is used after skeletal maturity.

**Catteral’s Classification of Perthes’ Disease**

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

- lateralization of hips
- lateral calcification of epiphysis
- metaphyseal involvement
- horizontal adjustment of epiphyseal plate
- triangular rarefication of lateral metaphysis (Gage sign)
Salter und Thompson’s Classification of Perthes’ Disease

Salter and Thompson’s classification (1984) concentrates, above all, on the visible subchondral fracture in axial radiographs. The extension is considered 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 of Perthes’ Disease

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

<table>
<thead>
<tr>
<th>Herring Type A</th>
<th>Lateral pillar is unaffected</th>
</tr>
</thead>
<tbody>
<tr>
<td>Herring Type B</td>
<td>&gt; 50 % of lateral pillar is intact</td>
</tr>
<tr>
<td>Herring Type C</td>
<td>&lt; 50 % of lateral pillar is intact</td>
</tr>
</tbody>
</table>

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 with acetabulum.

<table>
<thead>
<tr>
<th>Stulberg 1</th>
<th>Normal Spherical Head (spherical congruency → No OA)</th>
</tr>
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<tbody>
<tr>
<td>Stulberg 2</td>
<td>Spherical head with coxa magna/breva 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>

Therapeutics

Note: The goal of treatment of Perthes’ disease is to maintain range of movement. The insurance of range of movement is the fundament of a congruent hip situation. Treatment is primarily aimed at limiting progressive deformation during the repair phase. This is 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. In order to achieve maximal range of movement, physiotherapy is necessary. 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 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’s 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
- Operation in stages Catterall III/Salter-Thompson B/Herring BC, 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

Differential Diagnoses

Similar Presentations as Perthes’ Disease

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

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Description</th>
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<tbody>
<tr>
<td>Coxitis fugax</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>bacterial coxitis</td>
<td>pronounced pain, joint drain/paracentesis is indicated</td>
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<tr>
<td>juvenile Osteonecrosis</td>
<td>vascular necrosis with a known underlying disease (e.g. sickle cell anemia, thalassemia, coagulopathy)</td>
</tr>
<tr>
<td>septic arthritis</td>
<td>acute malaise, flexed hips, abducted and externally rotated, signs of inflammation, greater joint effusion, synovitis, bone marrow edema</td>
</tr>
<tr>
<td>juvenile rheumatoid arthritis</td>
<td>Fever, positive ANA, rash, atrophy of the thigh muscles, synovitis, ischemic signs in the femoral head (in long-term cases)</td>
</tr>
<tr>
<td>Slipped capital femoral epiphysis</td>
<td>Dislocation of the femoral head in caudal and posteromedial direction, metadiaphyseal bone marrow edema, enlargement and distortion of the growth plate</td>
</tr>
<tr>
<td>Hypothyreose</td>
<td>Determine thyroid values!</td>
</tr>
<tr>
<td>Pituitary dysfunction</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 coxarthroses resulted in 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 at 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 female patient 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 K., a 6-year-old boy accompanied by his mother, is at your pediatric practice consultation. She reports that her son often complains of pain in the left knee while playing. Additionally, the kindergarten teacher, and herself at home, have 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. This 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

Genzwürker et al. (2014): AllEX – Alles fürs Examen. Thieme Verlag.


**Correct answers:** 1B, 2C, 3E

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