SERIAL IMAGING OF A CHILD WITH LEGG–CALVE-PERTHES DISEASE AT BETHESDA HOSPITAL, YOGYAKARTA: A CASE REPORT

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ABSTRACT

**Background:** Legg-Calve-Perthes disease (LCPD) is a rare condition of idiopathic avascular necrosis of the femoral head, mostly affects children under fifteen old. Most cases only affect one femoral head; bilateral involvement only occurs in about 15% of cases. Early diagnosis and prompt intervention play an important role in the prognosis to prevent degenerative damage on the bones and joints.

**Objective:** To describe a case of Legg-Calve-Perthes disease in a 9-year-old boy in Bethesda Hospital, Yogyakarta.

**Case Description:** A 9-year-old boy came with one month history of worsening pelvic pain to Bethesda Hospital, Yogyakarta. He also showed limping and limited mobility. Initial imaging studies did not find any abnormalities. Further follow-up with MSCT scan related to the complaint of persistent pain showed defects of left hip joint indicating Legg-Calve-Perthes disease. The patient undergone skin traction treatment which shows modest improvement on serial radiographic examinations during hospital admission.

**Conclusion:** The rarity of LCPD warrant thorough imaging studies on children with unexplainable severe hip joint pain. Treatment should be initiated as early as possible after diagnosis to prevent further bone damage and worsening prognosis. Conservative management with skin traction would benefit the patient in restoring normal anatomy and optimal joint congruence to prevent degenerative damage to the joints.

**Keywords:** Legg-Calve-Perthes Disease, Multidetector Computed Tomography, case report, Indonesia
INTRODUCTION
Legg-Calve-Perthes Disease (LCPD) is a rare condition of idiopathic avascular necrosis of the femoral head, with an annual incidence of 0.2-19.1 per 100,000 among children under fifteen years of age. Most cases only affect one femoral head; bilateral involvement only occurs in about 15% of cases. Early diagnosis and prompt intervention play an important role in the prognosis to prevent degenerative damage on the bones and joints. LCPD affects boys three to four times more often than girls and is more common in children with low birth weight, exposure to maternal smoking during pregnancy, low economic status, and white ethnicity. This report describes a case of LCPD in a 9-year-old boy with pelvic pain and limp gait.

Clinical Findings And Diagnostic Assessment
On the physical examination, there was tenderness and reduced ROM (range of motion) of the left hip joint. Initial anteroposterior x-ray of the pelvic showed no abnormalities and appropriate bone age (Figure 1). Furthermore, the follow-up multi slice CT-scan of the same area showed defects of the posterior part of fasciae on the left hip joint (Figure 2). The defect might indicate fissures or deformities. The patient was further treated with working diagnosis of Legg-Calve-Perthes disease (LCPD) and differential diagnosis of osteochondritis dissecans.

CASE DESCRIPTION
Patient Information
On the 6 July 2019, A 9-year-old male was brought to Bethesda Hospital, Yogyakarta with severe pelvic pain in the past month. The pain was throbbing with VAS (visual analogue scale) 8/10. The pain was initially felt in the inguinal region which was radiated to the left hip; it was aggravated by small movement causing a limp gait. He was discharged with painkillers prescription. On the second visit on 20 July 2019, the patient still had persistent pain unresponsive to pain medications.

Figure 1. Anteroposterior x-ray of the pelvic region on first visit.
Therapeutic Interventions
The patient was admitted on the second visit and was treated on the second day of admission (22 July 2019) with skin traction along with appropriate analgesics.
Follow-up And Outcomes
On the fifth day after skin traction, first follow-up MSCT showed an expanding joint space with a hyperdense, thin and irregular left femoral head and multiple small cysts under the hypophyseal line of the left femoral head (Figure 3). This follow-up MSCT confirmed the diagnosis of LCPD.

Two weeks after skin traction, the pain was still present and further MSCT showed clearer signs of avascular necrosis (irregularity of the joint head and acetabulum) of left femoral head with a modest improvement compared to previous CT scan (Figure 4). The patient was discharged after 1 month of treatment.
Further follow-up MSCT one months after the initial skin traction (29 August 2019) showed more irregularity and subperiosteal sclerosis of left femoral head compared to previous visit (Figure 5). The patient still experienced moderate pain and stiffness on the left hip joint and continue the treatment at Bethesda Hospital until this report was submitted.

**DISCUSSION**

Legg-Calvé-Perthes disease (LCPD) is a disorder of the femoral head characterised by repetitive cycles of bone necrosis, repair and remodelling which usually started with osteonecrosis due to severed vascular supply. The necrosis would cause localised synovitis, cartilage hypertrophy and teres ligament hypertrophy. The changes lead to muscle spasm which pull the femoral head lateral of the acetabulum. The deformity is worsened with gravitational pull of the body weight. Each cycle could take up to two years. In the time of its discovery, LCPD was classified as idiopathic, although recently some genetic studies found the potential connection of mutation in COL2A1 gene (which regulates the production of type II collagen) among children with LCPD.1

Most of the patients are children, shorter than their peers and experienced pelvic pain, limp gait and limited mobility.1,4,5 Patients often present with limping that is worsened by strenuous physical activity and relieved by rest. The second most frequent complaint is pain located in the groin or pelvic area. The pain is often referred to the knee or anterior thigh which can obscure the diagnosis. The pain is usually worse at night. Patients may be smaller in stature than their peers.5

Physical examination findings vary depending on the stage of the disease and become more pronounced with worse deformities of the femoral head. The limping in children is usually a combination of the antalgic gait and the Trendelenburg style. The child often leans his body over the affected hip to reduce abductor motion and pressure in the hip joint.5

During the initial stages, there is a variable reduction in pelvic movement due to muscle spasms or pelvic synovitis. Early in the disease process, distress occurs in the internal rotational movement. With
time, clinical manifestations may vary widely. Patients with mild disease may experience only a slight loss of ROM and can quickly regain normal leg mobility. Severe disease may cause greater loss of motion, especially in internal rotation and hip abduction.5

Diagnosis of LCPD requires radiologic examinations such as X-rays or CT scans. Early radiographs may be normal or show smooth flattening of the femoral head. Sclerosis and subchondral fractures may occur.6,7 As the disease progresses, the femoral head will fragment and collapse. The metaphysis becomes prominent. In the healing phase, LCPD can cause the femoral neck to become short and thick, which is called the coxa magna deformity (Figure 3).6,7

The main goals of management in LCPD are to restore the pelvic ROM, maintain the position of the femoral head inside the acetabulum, prevent further deformity, control pain and prevent osteoarthritis. After diagnosis, prompt treatment and referral to a paediatric orthopaedic surgeon is needed.8 Initial management to relieve symptoms include activity restriction and non-steroidal anti-inflammatory drugs (NSAIDs). Guidelines advise conservative care for children less than 8 years, reserving surgical procedures for older patients. To reduce pain and increase ROM, horizontal skin traction is used on both legs. Traction is done until the pain disappears, and ROM is restored, usually needing 7 to 9 days.9 Other nonoperative treatments include crutches and casting. Additional recommended nonsurgical modalities are physical therapy and orthotics. Braces, such as Scottish Rite braces, help position the femur in the abducted position while allowing free knee movement.3,8

The main surgical procedures used in LCPD are proximal femoral osteotomy and pelvic osteotomy. The goal of surgery is to restore normal anatomy and optimal joint congruence to prevent degenerative damage to the joint. Additional procedures that may be required are isolated proximal femoral osteotomy-osteochondroplasties, labrum procedure, and acetabuloplasties. 3

Generally, the classification of LCPD is not for treatment purposes, but rather prognostications with some available published prognostic scoring, for example a widely used Stulberg classification which predict the occurrence of arthritis in long term (Table 1).

<table>
<thead>
<tr>
<th>Class</th>
<th>Spherical congruency</th>
<th>Description</th>
<th>Radiologic Aspect</th>
<th>Prognosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Spherical congruency</td>
<td>Normal</td>
<td>Spherical head with one or more of the following findings: coxa magna, short femoral neck, upper located great trochanter, obliquus acetabulum</td>
<td>Good</td>
</tr>
<tr>
<td>II</td>
<td>Spherical congruency; Loss of head shape &lt;2mm</td>
<td>Non-spherical but not flat</td>
<td>Flat head and acetabulum</td>
<td>Good</td>
</tr>
<tr>
<td>III</td>
<td>Aspherical congruency; Loss of head shape &gt;2mm</td>
<td>Flat head, normal neck and acetabulum</td>
<td>Poor: moderate arthritis</td>
<td></td>
</tr>
<tr>
<td>IV</td>
<td>Aspherical congruency</td>
<td>Flat head and acetabulum</td>
<td>Poor: moderate arthritis</td>
<td></td>
</tr>
<tr>
<td>V</td>
<td>Aspherical incongruency</td>
<td>Flat head, normal neck and acetabulum</td>
<td>Bad: severe early arthritis</td>
<td></td>
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</tbody>
</table>
The most important predictors of treatment outcome for LCPD are the shape of the femoral head and how well it fits into the acetabulum. Based on the Stulberg staging, the case described belongs to class 1 to 2. These classes have a good prognosis because the head of the femur is still intact and congruent with the acetabulum is good. Patients rarely experience arthritis in the long term.10

CONCLUSION

LCPD is a rare disease, with an incidence of 0.5-19.1 per 100,000 people, requiring early diagnosis and immediate management. Radiological examinations with pelvic x-ray and hip joint MSCT can help make a diagnosis, assess the severity of disease and identify resulting structural abnormalities. Treatment is recommended as early as possible to prevent further bone damage and worsening prognosis. The management of LCPD is by osteotomy and conservative therapy (skin traction) with the aim of restoring normal anatomy and optimal joint congruence to prevent degenerative damage to the joints.

CONFLICT OF INTERESTS AND FUNDING RESOURCES

The authors declare no conflict of interests and external resource of funding for this case report.

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