**The management of Legg-Calve-Perthes disease: a scoping review with advice on initial management**

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**Abstract**

**Background:** Legg-Calve-Perthes Disease (LCPD) is a developmental disorder causing avascular necrosis of the femoral head in children, with long-term consequences that can extend into adulthood. Early diagnosis and management in primary care are crucial but challenging.

**Aim:** This review aims to provide a concise overview of the presentation, differential diagnosis, and management of LCPD, offering practical guidance for primary healthcare professionals.

**Method:** Recent literature and expert opinions were reviewed to summarise the epidemiology, diagnosis, and current management of LCPD.

**Results:** LCPD commonly presents as a painless limp in children aged between two and 14, with the diagnosis based on the clinical features and radiographic abnormalities. Management is individualised and includes non-operative care to surgery, which attempts to correct anatomical abnormalities and therefore delay the onset of osteoarthritis. The review highlights the importance of primary care in early detection, appropriate referral, and interim management.

**Introduction**

Legg-Calve-Perthes Disease (LCPD) was first described independently by three physicians in 1910 [1–3], and has since been the subject of extensive commentary, with more than 200 papers published on the disorder in the last five years alone ​[4].

It is a disorder of skeletal developmental with a global incidence of one in 1200 for children under the age of 15 [5]. The onset is associated with an indeterminate [6] vascular event that interrupts all, or part, of the blood supply of the proximal femoral epiphysis [5]. This leads to avascular necrosis​ of the femoral head, which may affect physical and mental health, in addition to social development, all of which may continue into adult life ​[7]​. ​​

LCPD usually presents with limp which may be painless [5], and primary care is often the first point of contact. It is therefore important that healthcare professionals working in this environment can identify this condition, arrange relevant investigations and appropriate referral.

Diagnosis is based on clinical and radiological findings [6], which are determined by the stage of presentation, but this may be challenging and involve exclusion of more sinister causes. Referral to a paediatric orthopaedic service is recommended, but explanation and reassurance, anti-inflammatory medication and avoidance of aggravating activities can begin in primary care [8].

This review aims to highlight the initial presentation, differential diagnosis and initial management of LCPD and describe a clinical pathway for healthcare professionals working in primary care.

**Epidemiology and potential aetiology**

LCPD can affect children at any age, but typically presents between two and 14, with peak onset between five and six ​[9]​. The condition predominantly affects males with a ratio of 5:1 [6] and affects both hips in 10-20% of cases [5].

There is no clear evidence of direct inheritance and twin studies do not demonstrate concordance ​[6]. Ethnicity, however, is a recognised risk factor [10] and LCPD is common in children of Inuit and Central European descent and unusual in East Asian and African Americans [5,10]. Geography also appears to have an effect with a 2.35 increase in incidence for each 10° increase in latitude ​[10]​. Northern European children are the most frequently affected ​[6]​ and the current incidence in the United Kingdom is estimated to be 2.48 per 100,000 [11].

Socioeconomic status is also associated with development of this condition, with the incidence in the most deprived being four times greater than the most affluent households in a single city. ​[12]​. This is also supported by a continuous decline in the UK incidence, from 12.2 per 100,000 in 1990, to 2.48 per 100,000 in 2022, which may be associated with improvement in living standards ​[13]​.  Maternal smoking during pregnancy, however, is also a recognised as a risk factor, with multiple studies reporting a dose-respondent relationship [14,15]​, and as there is a strong relationship between socioeconomic status and smoking, this may also be relevant to the change in incidence [6]. There is also an association with low birth weight, birth length below 50 cm, congenital abnormalities and Down’s syndrome [6,14,15].

Children with LCPD are 1.5 times more likely to be affected by attention deficit hyperactivity disorder (ADHD) [16] but the causal relationship is unclear [17].

The vascular aetiology of LCPD led to exploration of an association with thrombophilia and identified a proportion of patients with abnormal clotting, but without a consistent mechanism [6].

**Pathophysiology**

The cause of LCPD is unknown but is thought to involve a temporary vascular insult, which is probably multifactorial [5] and causes necrosis of the femoral epiphysis, resulting in femoral head deformity [18]. Thrombophilia [6], repeated subclinical trauma, joint overload [19], and smoking are all implicated as potentially important. Most experimental data is based on animal studies with a deterioration of material properties demonstrated in a pig model following a comparable vascular insult [18].

**Clinical presentation and differential diagnosis**

Patients present with a combination of symptoms that depend on the level of physical activity and pain threshold, the extent of an initial of joint effusion and stage of the disease [20]. Pain may not be localised to the hip and can radiate to the thigh and knee [5,21], and may affect the knee in isolation, reinforcing the necessity of examining the hip when a patient presents with knee pain, irrespective of the cause ​[22]​. The classical presentation of LCPD is limping, typically developing over a period of several weeks with no history of trauma [5,21].

Physical examination may demonstrate a limp on the affected side with limited hip movement, particularly affecting internal rotation and abduction ​[5,22]​. The opposite hip should be routinely examined, as both are affected in 10% of the cases ​[5]​. Thigh circumference may be reduced on the affected side due to disuse ​[5,22]​ and a true leg length discrepancy may be present if the epiphysis has collapsed ​[5,22]​.

The limping child is a common presentation to primary care ​[23]​ and the initial assessment is aimed to exclude alternative causes, particularly trauma, infection and malignancy [24].

Trauma is usually obvious from the history, but deliberate injury should always be considered, especially with non-ambulant children [25].

Septic arthritis usually presents with systemic symptoms and severe hip pain. There is often reluctance or inability to bear weight, and pain associated with any attempted joint movement [23,26]. The management is usually surgical drainage and children with suspected hip septic arthritis should be referred immediately to the local orthopaedic service [24].

The presentation of malignancy is usually insidious with background pain, particularly at night and not associated with activity. A palpable mass and generalised symptoms including weight loss, fevers, night sweats may not be present, particularly in the early stages [25].

Children aged older than nine with groin, thigh or knee pain or restricted hip movements, must be referred for urgent anterior-posterior and lateral radiographs to exclude slipped upper femoral epiphysis (SUFE), which is more common in older children and requires urgent surgical intervention [25].

Additional differential diagnoses are illustrated in Figure 1.

**Radiological assessment**

LCPD is usually diagnosed with anterior-posterior and lateral radiographs of both hips. ​[22,23]​ The radiographic features include asymmetrical femoral epiphyseal size, fragmentation, and bone destruction, with established deformity in longstanding cases ​[5]​. A deformed, flattened and sclerotic appearance of the femoral epiphysis on the affected side should raise suspicion for LCPD (Figure 2).

Magnetic resonance imaging (MRI) demonstrates the cartilage component of the femoral head, which is not visible on plain radiographs, [27] The addition of gadolinium contrast defines the vascular anatomy, providing information on staging and prognosis, but MRI is not usually the initial imaging modality for this condition [27].

Hip joint arthrography under general anaesthesia allows a dynamic examination of the joint including range of movement and joint stability and is often used to plan management ​[6,27]​.

**Management options and outcome**

Information and support available at the time of diagnosis helps patients and carers understand the condition, the treatment options, and the expected outcomes. This will alleviate some of the anxiety associated with an unexpected diagnosis and improve the overall care experience [28].

A recent UK Delphi study [8] emphasised the importance of a multidisciplinary approach, including physical therapy, pain management, and psychosocial support (Table 1).

Table 1 - Summary of the consensus recommendations for the supportive treatment of children with LCPD in the UK. Adapted from Galloway et al. (2024).

|  |  |  |
| --- | --- | --- |
| Early stage | Avoid | High impact exercises  Contact sport  Long-distance running  High-impact activities |
| Encourage | Regular cardiovascular exercises  Hip stretches  Swimming  Cycling |
| Late stage | Encourage | Hip-strengthening exercises  Trunk-strengthening exercises  Regular cardiovascular exercises  Stretching exercises  Swimming  Cycling  Horseback riding |
| Both stages | Encourage | Water-based exercise as self-management  Balance exercises  Pharmacological pain treatment if needed  Discussion about mental health support |
| Receive | Specialist review by an orthopaedic surgeon  Physiotherapy  Support at school  Means of contacting other patients with Perthes’ disease  Validated quality of life assessment tool and ROM assessment completed at initial assessment and regular intervals |

There are no agreed guidelines on the treatment strategy for LCPD and the options are generally subdivided into non-operative and operative management [28].

Non-operative approaches include physiotherapy to preserve hip movement. This concentrates on stretching, hip-strengthening exercises, cycling and water-based exercises [8]. A UK survey reported that 90% of clinicians refer children with Perthes’ disease to physiotherapy services [29] but a recent systematic review highlighted the limited evidence base for this treatment [30]. Additional treatment includes bracing and restriction of weight bearing [31,32]. The former involves applying braces or casts to the patient’s legs to align the femoral head within the acetabulum, thereby reducing the risk of deformity [31] and the latter aims to reduce the load on the joint during the initial stage using wheelchairs and crutches [32]. These strategies are largely of historical interest, and neither are considered part of the contemporary management [31,32].

The type of surgical approach depends on patient age, the stage of disease and surgeon’s preference [6]. Containment is the most common type of surgery and involves reorientating the femoral head and/or acetabulum. The purpose is to preserve a spherical femoral head and therefore reduce the risk of early onset osteo-arthritis [6,33] (Figure 3).

Procedures performed to relieve symptoms and postpone the definitive surgery include shelf acetabuloplasty and femoral valgus osteotomy. The former aims to support the extruded femoral head without altering the geometry of the femur or the acetabulum. The latter involves dividing the femur and reorientating the femoral head in the acetabulum [6]. It is likely that patients in this situation will require joint replacement as a young adult and

surgery must not impact on the utility of this definitive procedure.

The long-standing assumption that most patients affected by LCPD are expected to have good or excellent long term functional outcome ​[34]​ has recently been challenged. Studies have consistently identified inferior long-term radiological outcomes in females and patients who are older at the onset of disease ​[7]​. Additional factors that predispose to osteoarthritis include flattening (coxa plana) and enlargement (coxa magna) of the femoral head and premature growth plate closure, all causing hip joint incongruency [5].

Recent studies report a high proportion of patients with osteoarthritis and poor functional outcomes at 20-year follow up, with up to 50% of patients having radiological signs of osteoarthritis at the age of 50 ​[7]**​**.These are predominately retrospective, observational studiesand methodologically robust, long-term cohort studies utilising standardised outcome measures are required ​[35]​.

**Novel therapies and research directions**

LCPD is amongst the top five research priorities of the British Society for Children’s Orthopaedic Surgery ​[36], with an emphasis on prevention and an improved understanding of aetiology [6]. The International Perthes Study Group (IPSG) has begun prospective international data collection [37] to quantify risk factors including geography and ethnicity [6]. Animal studies using bisphosphonates and bone morphogenetic proteins [38] have demonstrated a positive effect in preserving the shape of the femoral head [39,40] and a human randomised control trial using bisphosphates is in progress [41]. Other potential treatments, including biologics and autologous stem cell transplants, are also being investigated *in vitro* [39] and the role of surgical containment of the femoral head is also an area of current research [6,11,42].

**Referral pathway for the General Practitioner**

Healthcare professionals working in primary care are often the first point of contact [23], and a step-by-step guide is outlined in Figure 4 to assist with recognition and referral.  Once LCPD is suspected, patients and families should be referred to specialist orthopaedic services. In the interim, primary care support is vital, and this can involve referral to physiotherapy, prescribing analgesia, signposting mental health support and arranging school support. Providing patient centred literature may also alleviate anxiety and improve the overall care experience [28]. This support promotes greater understanding of the condition, treatment options and expected outcome. [26]. STEPS, a registered charity working with people affected by childhood lower limb conditions, has produced a comprehensive patient-centred LCPD factsheet and handbook (<https://www.stepsworldwide.org/wp-content/uploads/2021/03/JN113xxx-STEPS-PERTHES-DISEASE-BOOKLET-V10.pdf>) [43].

Children affected by LCPD should remain active, although there is no consensus on the appropriate level of activity [8]. Regular cardiovascular exercise and water-based activities are recommended but forms of high impact and contact sport may be inappropriate. This depends on the disease stage and usually forms part of the discussion with an orthopaedic surgeon [8].

Pain management is of paramount importance in the management of LCPD, as more than half of the patients present with pain [21]ffective analgesia reduces the disease burden and can limit absence from school [28]. A combination of simple analgesia such as paracetamol and ibuprofen should be used [8], but opiates are not recommended [8].

School attendance and performance may be affected by pain, reduced mobility and hospital visits and access to a named support worker is an advantage during the course of the disease[8].

Mental health support is also important [8] as a visible limp and exclusion from recreational activities may cause anxiety, impact sleep [44], with recent evidence suggesting that this continues into adulthood[7]. Whilst access to paediatric mental health services may be limited [45,46] alternative support should be considered. This includes self-help strategies and organisations that promote family and peer support [43]. STEPS and the Perthes Kids Foundation coordinate activities for children affected by LCPD [47] and the IPSG manages a list of Facebook groups that facilitates contact between patients and parents [48].

**Summary**

LCPD is an uncommon disorder, which may cause significant long-term disability, impacting a child's physical, mental, and social development. It often presents as a painless limp, and healthcare professionals working in primary care are often the first point of contact.

Diagnosis is generally made using plain radiographs that demonstrate an alteration of the shape of the femoral head. MRI may provide additional information about the unossified cartilage component and vascularity and assist with management planning, which includes non-surgical and surgical options.

Non-surgical methods include physiotherapy with a focus on maintaining hip mobility, and surgical treatment aims to correct the hip alignment. Despite treatment, patients are susceptible to osteoarthritis in later life.

Many patients present to primary care in the early stages, and it is essential for health care workers to recognise symptoms and refer to orthopaedic services for specialist management. Interim primary care support is vital, and this includes referral for physiotherapy, analgesia, and signposting mental health support, school support, and patient centred literature.

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**Figure legend:**Figure 1 – Differential diagnoses divided by age group for children presenting with a new-onset limp

Figure 2- A case of LCPD affecting the right hip in a x year-old patient demonstrating flattening and sclerosis of the femoral epiphysis

Figure 3 – Surgical containment for LCPD affecting the right hip.

Figure 4 - Pathway for suspected LCPD

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