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LEGG CALVE PERTHES DISEASE: A COMPREHENSIVE REVIEW

Nursing

Ms. Santosh Kumari* Assistant Professor, Department of Obstetrics and Gynaecological Nursing, Maharishi Markandeshwar College of Nursing, Maharishi Markandeshwar University, Kumarhatti, Solan, H.P., India.*Corresponding Author

Dr. Jasbir Kaur Dean & Principal, Department of Mental Health Nursing, Maharishi Markandeshwar College of Nursing, Maharishi Markandeshwar University, Kumarhatti, Solan, H.P., India.

ABSTRACT

Legg-Calve-Perthes disease (LCPD) is a childhood hip disorder of unknown etiology that can produce permanent deformity of the femoral head. This condition was first reported as a disease in 1910 by three independent authors: Legg, Calve and Perthes. Legg-Calve-Perthes typically occurs in children who are between 4 and 10 years old. It is five times more common in boys than in girls however, it is likely to cause more extensive damage to the bone in girls. In 10% to 15% of all cases, both hips are affected. It is a hip disorder in which the blood supply to the head of the femur interrupted (the ball in the ball-and-socket hip joint), causing it to deteriorate. Age, Child's gender, race, and genetic mutations are the risk factors for this disorder. Early diagnosis and management can help prevent the collapse of the femoral head, progressive femoral head deformity and impingement. The long-term prognosis for children with Perthes is good in most cases. After 18 to 24 months of treatment, most children return to daily activities without major limitations.

KEYWORDS

Legg-Calve-Perthes disease (LCPD), Children, Hip Disorder.

INTRODUCTION

Legg-Calve-Perthes disease (LCPD) is a childhood hip disorder of unknown etiology that can produce permanent deformity of the femoral head. This condition was first reported as a disease in 1910 by three independent authors: Legg, Calve and Perthes [1]. Since then, controversies regarding the etiology, pathogenesis, and management of LCPD have arisen, many of which remain unresolved. Recent genetic studies on a type II collagen mutation as a cause of LCPD and studies on the role of inherited thrombophilia on LCPD represent advancement, but the cause of ischemic necrosis remains unknown [2].

Legg-Calve-Perthes typically occurs in children who are between 4 and 10 years old. It is five times more common in boys than in girls however, it is likely to cause more extensive damage to the bone in girls. In 10% to 15% of all cases, both hips are affected [3]. There is no consensus for the optimum treatment. The aim of treatment is to maintain the sphericity of the femoral head and the congruency of the femur-acetabulum relationship to prevent secondary degenerative arthritis, which eventually leads to total hip arthroplasty in 5% of cases [4].

Early diagnosis and management can help prevent the collapse of the femoral head, progressive femoral head deformity and impingement [5].

OBJECTIVES

Objective of the study was to review available information regarding definition, risk factors and causes, pathogenesis, clinical manifestation, diagnostic evaluation, management and prognosis of Legg-Calve-Perthes disease (LCPD).

DEFINITION

Legg-Calve-Perthes disease: A hip disorder in children due to interruption of the blood supply to the head of the femur (the ball in the ball-and-socket hip joint), causing it to deteriorate [6].

RISK FACTORS AND CAUSES

Risk factors for Legg-Calve-Perthes disease include:

- **Age.** Although Legg-Calve-Perthes disease can affect children of nearly any age, it most commonly begins between ages 4 and 10.
- **Child's sex.** Legg-Calve-Perthes is about four times more common in boys than in girls.
- **Race.** White children are more likely to develop the disorder than are black children.
- **Genetic mutations.** For a small number of people, Legg-Calve-Perthes disease appears to be linked to mutations in certain genes, but more study is needed [7].

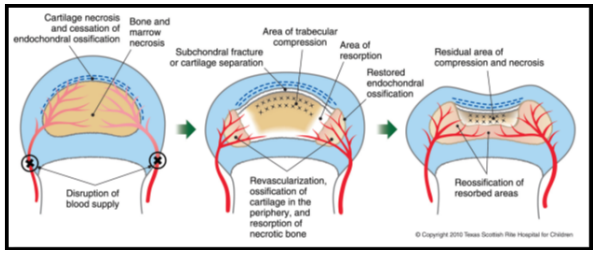

Figure 1: Histopathological changes observed in the necrotic femoral head over time. [8]

CAUSES

Legg-Calve-Perthes disease occurs when too little blood is supplied to the ball portion of the hip joint (femoral head). Without enough blood, this bone becomes weak and fractures easily (As shown in figure 1). The cause of the temporary reduction in blood flow to the femoral head remains unknown. [6,8].

PATHOGENESIS

The etiology of LCPD remains unknown. However, clinical and experimental evidence support the notion that disruption of blood supply to the femoral head is a key pathogenic even associated with the disease process. The few histopathological studies available indicate that the pathologic processes in LCPD affect the articular cartilage, bony epiphysis, physis, and metaphysis [9].

Changes in the articular cartilage are found primarily in the middle and deep layers. These changes include necrosis in the deep layer, cessation of endochondral ossification, separation of cartilage from the underlying subchondral bone, vascular invasion of the cartilage, and new accessory ossification (Figure 1). In the bony epiphysis, necrosis of the marrow space and trabecular bone, compression fracture of the trabeculae, fibrovascular granulation tissue invasion and osteoclastic resorption of the necrotic bone, and thickened trabeculae in some areas have been reported. Physeal changes are most often seen in the anterior part of the femoral head, with focal areas of growth cartilage columns extending below the endochondral ossification line. Metaphyseal changes are commonly seen during the early stages of LCPD [8].

Various tissue types have been reported, including physeal cartilage columns, fibrocartilage, fat necrosis, vascular proliferation, and focal fibrosis. The lack of availability of clinical samples for research has prompted alternative approaches, such as the use of animal models, to investigate the pathogenesis of LCPD. In particular, a piglet model has allowed more in-depth investigation of ischemic tissue damage and the

repair process. These models indicate that the induction of ischemia produces a decrease in the mechanical strength of the necrotic femoral head, resulting in a head that is softer than normal. [10]

The mechanical compromise observed in the vascular necrotic phase may be the result of necrosis of the deep layer of articular cartilage; increased mineralization of the calcified cartilage and trabecular bone, which presumably makes them more brittle; and the possible accumulation of microfractures in the necrotic bone due to the absence of viable cells to repair the microfractures produced by normal wear and tear caused by repetitive loading. Vascular invasion and subsequent resorption of necrotic bone further compromise the mechanical properties of the infarcted head in the vascular repair phase.[11]

The hip is a major load-bearing joint, and it is pertinent to consider the development of femoral head deformity in the context of hip joint loading. Data on the hip contact pressures associated with various activities of daily living in children are not available. In adults, however, a sophisticated femoral head prosthesis equipped with a strain gauge has been used to collect data following total hip replacement. [12]

Significant forces were found to act on the femoral head with various activities. Normal walking produced a hip contact pressure approximately 2.5 times body weight. Running on a treadmill (8 km/h) increased the pressure to approximately 4.5 times body weight. In a disease in which femoral head deformity is produced as the result of mechanical weakening, avoidance of activities that generate a significant increase in the hip contact pressure would seem to be reasonable. Currently, it is unknown what constitutes "significant" loading; neither is it known what effect activity restriction has on preventing deformity. [8].

CLINICAL MANIFESTATION

Signs and symptoms of Legg-Calve-Perthes disease include:

- Limping
- Pain or stiffness in the hip, groin, thigh or knee
- Limited range of motion of the hip joint
- Pain that worsens with activity and improves with rest

Legg-Calve-Perthes disease usually involves just one hip. Both hips are affected in some children, usually at different times.[8]

DIAGNOSTIC EVALUATION

During the physical exam, doctor might move child's legs into various positions to check range of motion and see if any of the positions cause pain.

Imaging tests

These types of tests, which are vital to the diagnosis of Legg-Calve-Perthes disease, might include:

- **X-rays.** Initial X-rays might look normal because it can take one to two months after symptoms begin for the changes associated with Legg-Calve-Perthes disease to become evident on X-rays. Doctor will likely to recommend several X-rays over time, to track the progression of the disease.
- **MRI.** This uses radio waves and a strong magnetic field to produce very detailed images of bone and soft tissue inside the body. MRIs often can visualize bone damage caused by Legg-Calve-Perthes disease more clearly than X-rays can, but are not always necessary. [13].

MANAGEMENT

In Legg-Calve-Perthes disease, the complete process of bone death, fracture and renewal can take several years. The types of treatment recommended will depend on the:

- Age when symptoms began
- Stage of the disease
- Amount of hip damage

As Legg-Calve-Perthes disease progresses, the ball part of the joint (femoral head) weakens and fragments. During healing, the socket part of the joint can serve as a mold to help the fragmented femoral head retain its round shape.

For this molding to work, the femoral head must sit snugly within the socket. Sometimes this can be accomplished with a special type of leg cast that keeps the legs spread widely apart for four to six weeks.

Some children require surgery to help keep the ball of the joint snug within the socket. This procedure might involve making wedge-shaped cuts in the thigh bone or pelvis to realign the joint.

Surgery usually isn't needed for children younger than 6. In this age group, the hip socket is naturally more moldable, so the ball and socket typically continue to fit together well without surgery. [13]

Legg-Calve-Perthes surgical treatment

If non-surgical treatments prove inadequate, child may need surgery to hold the head of the thigh bone in the hip socket (containment). Surgery involves reorienting the affected bones (osteotomy) and stabilizing the realignment with screws and plates.

The two most important factors that determine the outcome are the child's age (usually, the younger the better) and how much of the head of the thigh bone is affected by the condition. [14]

Other treatments

Some children, particularly very young ones, might need only conservative treatments or observation. Conservative treatments can include:

- **Activity restrictions.** No running, jumping or other high-impact activities that might accelerate hip damage.
- **Crutches.** In some cases, child may need to avoid bearing weight on the affected hip. Using crutches can help protect the joint.
- **Physical therapy.** As the hip stiffens, the muscles and ligaments around it may shorten. Stretching exercises can help keep the hip more flexible.
- **Anti-inflammatory medications.** Doctor might recommend ibuprofen (Advil, Motrin, IV others) or naproxen sodium (Aleve) to help relieve child's pain.[13]

PROGNOSIS

The long-term prognosis for children with Perthes is good in most cases. After 18 to 24 months of treatment, most children return to daily activities without major limitations. [15]

CONCLUSION

Legg-Calve-Perthes, it is a hip disorder in children due to interruption of the blood supply to the head of the femur (the ball in the ball-and-socket hip joint), causing it to deteriorate, typically occurs in children who are between 4 and 10 years old. So early diagnosis and management can help prevent the collapse of the femoral head, progressive femoral head deformity and impingement. The long-term prognosis for children with Perthes is good in most cases. After 18 to 24 months of treatment, most children return to daily activities without major limitations.

REFERENCES

1. Kim HKW, Legg-Calve-Perthes Diseases, Review, Journal of the American Aademy of Orthopedic Surgeon [Internet] 2010 Nov ;[18][11]: p676–86.
2. Pavone V, Chisari E, Vescio A, Lizzio C, Sessa G, Testa G. Aetiology of Legg-Calvé-Perthes disease: A systematic review [Internet]. Vol. 10, World Journal of Orthopaedics. Baishideng Publishing Group Co; 2019 [cited 2020 Dec 26]. p. 145–65. Available from: /pmc/articles/PMC6429000/?report=abstract.
3. American Acadamy of Surgeons, Perthes Disease - Legg-Calve-Perthes - OrthoInfo - AAOS [Internet]. 2019 Oct [cited 2020 Oct 29]. Available from: https://orthoinfo.aaos.org/en/diseases--conditions/perthes-disease.
4. Larson AN, Sucato DJ, Herring JA, Adolfsen SE, Kelly DM, Martus JE, et al. A prospective multicenter study of Legg-Calvé-Perthes disease: Functional and radiographic outcomes of nonoperative treatment at a mean follow-up of twenty years. J Bone Jt Surg - Ser A [Internet]. 2012 Apr 4 [cited 2020 Dec 26];94(7):584–92. Available from: https://pubmed.ncbi.nlm.nih.gov/22488614/
5. Shah H. Perthes disease. Evaluation and management [Internet]. Vol. 45, Orthopedic Clinics of North America. Orthop Clin North Am; 2014 [cited 2020 Dec 26]. p. 87–97. Available from: https://pubmed.ncbi.nlm.nih.gov/24267210/.
6. Definition of Legg-Calve-Perthes disease [Internet]. [cited 2020 Dec 26]. Available from: https://www.medicinenet.com/legg-calve-perthes_disease/definition.htm.
7. Legg-Calve-Perthes disease - Symptoms and causes - Mayo Clinic [Internet]. [cited 2020 Dec 26]. Available from: https://www.mayoclinic.org/diseases-conditions/legg-calve-perthes-disease/symptoms-causes/syc-20374343.
8. Legg-Calvé-Perthes Disease : JAAOS - Journal of the American Academy of Orthopaedic Surgeons [Internet]. [cited 2020 Dec 26]. Available from: https://journals.lww.com/jaaos/pages/articleviewer .aspx?year=2010&issue=11000&article=00005&type=Fulltext.
9. Catterall A, Pringle J, Byers PD, Fulford GE, Kemp HB, Dolman CL, et al. A review of the morphology of Perthes' disease. J Bone Jt Surg - Ser B [Internet]. 1982 [cited 2020 Dec 26];64(3):269–75. Available from: https://pubmed.ncbi.nlm.nih.gov/6807991/.
10. Kim HKW, Su PH. Development of flattening and apparent fragmentation following ischemic necrosis of the capital femoral epiphysis in a piglet model. J Bone Jt Surg - Ser A [Internet]. 2002 Aug 1 [cited 2020 Dec 26];84(8):1329–34. Available from: https://pubmed.ncbi.nlm.nih.gov/12177261/
11. Pringle D, Koob TJ, Kim HKW. Indentation properties of growing femoral head following ischemic necrosis. J Orthop Res [Internet]. 2004 [cited 2020 Dec 26];22(1):122–30. Available from: https://pubmed.ncbi.nlm.nih.gov/14656670/
12. Kim HKW. Legg-Calve-Perthes Disease. J Pediatr Orthop [Internet]. 2011 Sep [cited 2020 Dec 26];31(SUPPL. 2):S141–6. Available from:

http://journals.lww.com/01241398-201109001-00006
13. Legg-Calve-Perthes disease - Diagnosis and treatment - Mayo Clinic [Internet]. [cited 2020 Dec 28]. Available from: https://www.mayoclinic.org/diseases-conditions/legg-calve-perthes-disease/diagnosis-treatment/drc-20374348
14. Legg-Calve-Perthes Disease | Treatments | Boston Children's Hospital [Internet]. [cited 2020 Dec 28]. Available from: https://www.childrenshospital.or g/conditions-and-treatments/conditions/l/legg-calve-perthes-disease/treatments
15. Perthes Disease - Legg-Calve-Perthes - OrthoInfo - AAOS [Internet]. [cited 2020 Dec 28]. Available from: https://orthoinfo.aaos.org/en/diseases--conditions/perthes-disease