

Perthes Disease

Evaluation and Management

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KEYWORDS

• Perthes disease • Iliac osteotomy • Femoral osteotomy • Containment • Congruent hip

KEY POINTS

- The primary goal of the treatment is to prevent irreversible femoral head deformation, incongruent hip, and femoroacetabular impingement.
- Preventable treatment strategy is more effective in the early stage (before the stage of advanced fragmentation) than in the late stage of the disease. The timing of the surgery is more important than the type of surgery.
- Epiphyseal extrusion is the most important and the only factor that modulates the preventable treatment in early Perthes disease.
- Most younger children can be managed conservatively. Surgical containment is essential for children with late-onset Perthes disease. Normal hip joint movements and absence of hinge abduction are the prerequisites for surgical containment.

INTRODUCTION

Perthes disease is one of the most common pediatric disorders. It is an aseptic, noninflammatory, self-limiting, idiopathic, avascular necrosis of capital femoral epiphysis in a child.¹ One hundred years after its first description, the exact cause of the Perthes disease is not known.² The treatment of Perthes disease may be preventive, remedial, or salvageable in nature depending on when the child is diagnosed.

The aim of treating Perthes disease is to prevent secondary degenerative arthritis of the hip in adult life, which can be achieved by preventing the femoral head from getting deformed if the child is diagnosed early, by minimizing the adverse effects of early deformation of the femoral head if it has already occurred, and by salvaging hips with established deformation of the femoral head.³

NATURAL HISTORY

Perthes is a self-limiting disorder as blood supply of the femoral head restores to normal within 2 to

4 years' duration following initial avascularization. Single or multiple recurrent episodes of interruption of blood supply of the femoral head occur. Once the blood supply to the femoral head is compromised, a series of events occur within and outside the femoral head. Avascular necrosis of part or all of the epiphysis occurs; the necrotic bone is resorbed by osteoclasts. The weakened trabeculae collapse and the epiphysis fragments. Woven bone is laid down on the periphery of the epiphysis and over a period of time this woven bone is replaced by mature lamellar bone and the epiphysis heals completely. Concomitant changes take place outside the femoral head. Hypertrophy of the synovium, ligamentum teres, and the articular cartilage occurs. These soft tissue changes along with muscle spasm initiate femoral head extrusion that tends to increase progressively. The extruded femoral head, when subjected to stresses that pass across the acetabular margin, lead to irreversible deformation of the femoral head.^{4,5} A little remodeling of the femoral head may then occur. Any residual femoral head

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Orthop Clin N Am 45 (2014) 87–97

<http://dx.doi.org/10.1016/j.ocl.2013.08.005>

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deformity and joint incongruity will then persist throughout life.^{6,7} Recent evidence has clearly shown that irreversible deformation occurs when the disease has progressed to the late stage of fragmentation or soon after.¹

Variables that make femoral head deformity worse are femoral head weakening and significant loading. Femoral head weakening correlates with the extent of head involvement.⁸ Loading depends on the patient's activity level, type of activities, and weight. Intervention should necessarily precede the onset of irreversible deformation of the femoral head.⁹

EVALUATION OF PROGNOSTIC FACTORS

It is important to know the prognostic factors that affect the final outcome of the disease.

Short-term Prognostic Factors

The factors that determine the shape of the femoral head at the time of healing of the disease include age at onset of the disease, extent of epiphyseal avascularity, extent of epiphyseal collapse, and extent of epiphyseal extrusion.^{8,10-12} Early onset of the disease, less than 50% of head involvement, less severe collapse, and the absence of epiphyseal extrusion are good indicators of the outcome. Of these factors, epiphyseal extrusion is the most important and the only factor that can be modulated by treatment.¹⁰

Long-term Prognostic Factors

The factors that predispose to the development of secondary degenerative arthritis include shape of the femoral head at the time of healing of the disease, congruency between the femur and the acetabulum, and age at onset of Perthes disease.^{8,13,14} Degenerative arthritis of the hip joint is correlated with the irregular shape of the femoral head, incongruent hip, and the late onset of the disease.

EVALUATION

Patient Evaluation

The age of onset of symptoms and duration of the disease must be determined. The exact age of onset can guide the planning of the management. Long duration of the disease may cause one to miss the timing for the preventable management strategy. History of passive smoking, which should be evaluated as maternal smoking,¹⁵ at least one smoker living in the child's household,¹⁶ and wood smoke,¹⁷ are associated with increased risk of Perthes disease.

Clinical Evaluation

The child with Perthes disease limps and complains of occasional pain in the groin, hip, or knee. These symptoms may be present for weeks or even months. The examination shows a mild limp and decreased range of motion in abduction and internal rotation. Occasionally, there may be gross limitation of all range of motion. Persistent hip stiffness is a poor prognostic sign. It is essential to gain normal range of motion of the hip joint before considering containment treatment.

Radiological Evaluation

Plain radiographs (anteroposterior [AP] and frog lateral) are useful to diagnose the stage, the extent, and the severity of involvement of the femoral head.^{8,10-12} The sequential changes of natural history can be divided into two groups: active disease and healed disease.¹ Disease is considered active when the capital femoral epiphysis looks sclerotic with or without the presence of collapse and reossification. The disease is considered healed when no remnant of avascular bone can be identified on both views. Contrast-enhanced magnetic resonance imaging can clearly define the area of the involvement in the early stage of the disease.¹⁸

ACTIVE STAGE OF DISEASE

Early Stage of the Disease

The femoral head is subjected to deformation in the late stage of fragmentation or soon after this stage, so the active stage of the disease is divided into the early and late stages. The early stage refers to the stage between early avascular necrosis and early stage of fragmentation. The late stage refers to the stage between late fragmentation and early reossification of capital femoral epiphysis.

Management

The age of the child at onset of symptoms, the extent of involvement, stage of the disease, the range of motion of the hip, and the presence of the extrusion of the femoral head must be considered for treatment in the early stage of the disease.

The age of onset can be further divided into less than 5 years, 5 to 8 years, 8 to 12 years, and more than 12 years (adolescent Perthes disease). Adolescent Perthes disease behaves differently from early onset Perthes disease.¹⁹ Nonsurgical treatment is effective and indicated for children who are young (ie, less than 5 years of age).

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