

Management of Late Presented Perthes Disease

Kumar Amerendra Singh¹, Hitesh Shah¹

Abstract

Abstract A child with Perthes disease presents at or after an advanced stage of advanced fragmentation is considered a late presentation. The maximum femoral head deformation happens at the stage of advanced fragmentation. Preventive strategies are needed if the child presents before the advanced stage of fragmentation. The corrective strategies are deliberated if the child presents as a late presenter. The natural history of late presented Perthes is poorer than early presentation. Clinical features, investigation, various prognostic factors, management options and decision making of late presented Perthes disease have been revealed.

Keywords: Late presented Perthes disease, Hinge abduction, Severe collapse, Extrusion, Poor prognosis

Introduction

Perthes disease is self-limiting non-infective, non-inflammatory avascular necrosis of the capital femoral epiphysis. The prime aim of treatment of the Perthes disease is to maintain a spherical femoral head and to prevent secondary osteoarthritis of the hip joint. The femoral head deformation happens at the late stage of fragmentation or re-ossification stage. The femoral head deformation is related due to femoral head flattening and extrusion [1].

When the child presents before the advanced stage of fragmentation is defined as early presentation. When the child presents at or after an advanced stage of fragmentation is considered a late presentation. When the child's age of onset is less than 6 years, it is called early-onset Perthes disease [1]. When the child's age of onset is more than 8 years, it is called late-onset Perthes disease [2]. The prognosis of the Perthes disease depends on the age of onset, stage of presentation, range of motion, the extent of involvement, and the extent of extrusion [3]. The prognosis is good in early-onset, early presented cases. There is no consensus on the treatment modality for late-presented Perthes disease at or after the late stage of fragmentation.

Pathogenesis of femoral head deformation in the late stage of

fragmentation.

The ischemia of the femoral head results in synovitis of the hip joint, hypertrophy of the femoral head cartilage, and the ligamentum teres. The adductor muscle spasm and increased contents in the hip joint lead to the extrusion of the femoral head (Figure 1). When the child walks, the force from the lateral margin of the acetabulum is transmitted to the fragmented extruded femoral head which worsens the deformation (Figure 1). The newly formed bone in the late-stage lacks the trabecular pattern of bone and cannot withstand these stresses [4].

Issues with late presented cases

- Extrusion with progressive deformation of the femoral head
- Hinge abduction

Hinge abduction is defined as an impingement of the femoral head (lateral part) on the margin of the acetabulum (lateral part).

Clinical features

The typical presentation of Perthes includes restriction of hip range of movement [5]. The main difference between early and late presentations is the stiffness of the joint. In presence of hinge abduction or femoral head deformation, sudden deterioration of movements of the hip joint is the key feature. The hallmark of hinge abduction includes severe restriction of abduction, out-toeing, or in-toeing gait (due to compensatory mechanism). The axis deviation might be present in presence of femoral head deformation. The limb shortening is also marked by the severe collapse of the femoral head. Positive

¹Department of Pediatric Orthopaedics, KMC, Manipal, Manipal Academy of Higher Education, Manipal, Karnataka, India.

Address of Correspondence :

Dr. Hitesh Shah,

Professor, Department of Pediatric Orthopaedics, KMC, Manipal, Manipal Academy of Higher Education, Manipal, Karnataka, India.

E-mail: hiteshshah12@gmail.com

Submitted: 22/12/2021; Reviewed: 19/1/2022; Accepted: 23/01/2022; Published: 10/02/ 2022

© 2022 Journal of Karnataka Orthopaedic Association | Available on www.jkooonline.com | DOI:10.13107/jkoa.2022.v10i01.044

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial-Share Alike 4.0 License (<http://creativecommons.org/licenses/by-nc-sa/4.0>) which allows others to remix, tweak, and build upon the work non-commercially as long as appropriate credit is given and the new creation are licensed under the identical terms.

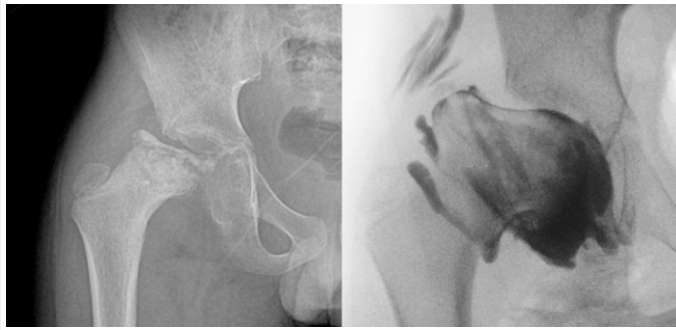


Figure 1: 35% of extrusion of the femoral head with deformation of the femoral head visualised on arthrogram

Trendelenburg sign might suggest insufficiency of abductor mechanism due to high trochanter.

Investigations

Routine AP and frog lateral radiographs would be sufficient to diagnose late presented cases (Figure 2). The frequency of fragments is more than 2 in both views with or without new bone formation is considered a late stage. If the child presents with new bone on the lateral aspect, it is considered the re-ossification stage (stage III).

Hinge abduction can be easily picked up in the AP radiograph (Figure 2). Normally, the joint space remains similar in abduction. However, widening of medial joint space in abduction is the characteristic feature of hinge abduction (Figure 2) [6]. It can also be confirmed with a dynamic arthrogram. The medial joint space is equal in normal hip abduction, while it gradually widens in presence of hinge abduction (Figure 1). Arthrogram can also diagnose reducible or irreducible hinge abduction [7]. Another advantage of arthrogram is to choose the best congruent position and presence of anterior and lateral impingement if it is present.

Variables affecting management in late presented cases.

- Age of the child
- Presence of deformity
- The severity of restriction of range of motion
- Presence of the extrusion
- Presence of hinge abduction
- Enlargement of the femoral head



Figure 3: Anteroposterior radiograph of the pelvis with both hip joints shows 40% extrusion of the left femoral head which gets reduced in frog-lateral position radiograph.

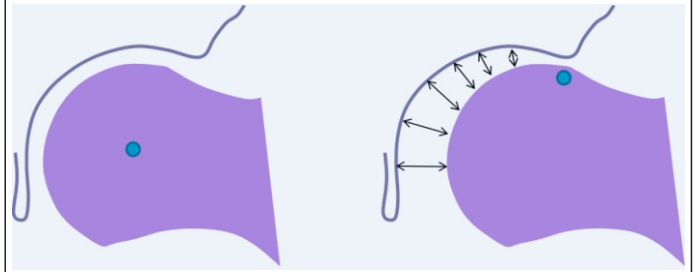


Figure 2: Schematic diagram of normal and hinge abduction show uniform medial joint space on abduction on normal, while gradual widening of medial joint space in hinge abduction

Management

The prevention of the femoral head deformity is the prime aim of treatment of early presented cases [8]. While, the aim of the treatment in late presented cases is to correct the consequences of the altered shape and size of the femoral head and prevent further deformation of the femoral head.

Treatment options in late presented cases include conservative-Non weight-bearing, symptomatic- treat after healing, valgus osteotomy, Shelf osteotomy, Chiari osteotomy, Arthrodiastasis-(articulated/non-articulated), or femoral head reduction surgery.

Conservative treatment

Non-weight bearing till the late stage of re-ossification was considered a traditional option [9]. The natural history of all surgical interventions compared to non-operative treatment is not superior in the current literature. Ideal cases for conservative treatment are late presented cases without hinge abduction and significant extrusion. The treatment is indicated in younger age of onset children with late presentation. One may treat the child symptomatically till healing and consider to deal the residual issue after complete healing. However, persistent stiffness and prolonged treatment period may be disadvantages of conservative treatment.

Containment surgery

Containment surgery is indicated at an older age of onset (more than 8 years) with late presentation. The surgeon must achieve a normal range of motion before considering containment surgery. One must rule out hinge abduction before considering



Figure 4: Left hip joint sequential radiographs showing good coverage in Perthes disease after shelf acetabuloplasty

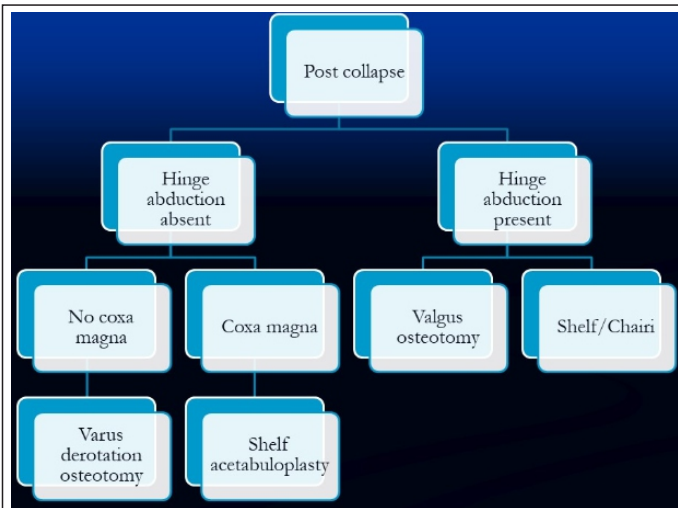


Figure 5: Flowchart for the treatment of hinge abduction in Perthes disease

containment surgery. If hinge abduction is suspected, one must check under anesthesia regarding the reducibility of hinge abduction. If the femoral head is well centered without pressing the lateral margin of the acetabulum, it is reducible hinge abduction (Figure 3). However, in presence of reducible hinge abduction, the outcome would be poorer than no hinge abduction.

The containment can be achieved by either varus osteotomy or shelf osteotomy (Figure 4) [10]. The size of the femoral head is similar to the size of the acetabulum; one can consider femoral varus osteotomy. If the size of the femoral head is bigger than the acetabulum, one can consider shelf acetabuloplasty. Lateral shelf acetabuloplasty can prevent the subluxation of the femoral head and stimulate lateral acetabular growth. Shelf acetabuloplasty is indicated in older children and children with a bigger femoral head (suspected coxa magna).

Non-containment surgery

Containment surgery does not give benefit children with irreducible hinge abduction. When the femoral head does not center the acetabulum, the lateral part of the femoral head enforces pressure on the lateral margin of the acetabulum, it is defined as irreducible hinge abduction. Femur valgus osteotomy, articulated hip distraction, shelf and Chiari acetabuloplasty, and femoral head reshaping are options to treat such a scenario (Figure 5) [2].

Valgus osteotomy

When the hip joint is congruent in adduction, and non-congruent in neutral or abduction, femur valgus osteotomy is commonly indicated. It reinstates joint congruity and decreases femoroacetabular impingement [11].

Valgus osteotomy relocates the abnormal hinge segment away from the acetabulum margin; it escalates the weight-bearing

surface under the acetabulum; it improves the neck-shaft angle; it maintains the abductor muscle length.

The degree of valgus should be judged by the maximum joint congruent position in adduction. Valgus osteotomy may also be combined with other plane deformities. Valgus extension osteotomy is effective when hinge abduction is combined with flexion deformity of the hip with a soft anterior hinge that can be contained within the acetabular socket. When the anterior hump is relatively large and non-containable, extension combination is not indicated because the hump may cause anterior impingement. If the position of the hip is better in internal rotation, external rotation osteotomy can have been added with a valgus osteotomy, similarly, the congruency of the hip is good in external rotation with internal rotation osteotomy can be done.

The disadvantage of valgus osteotomy includes higher chances of subluxation and increased instability of the hip. Various surgeries (shelf, or Chiari osteotomy) can be added to attain acceptable coverage [12, 13].

When all range of movements is restricted, the valgus osteotomy is contraindicated. In presence of a stiff hip with fixed soft tissue contracture, femur osteotomy and acetabulum osteotomy are not possible. Arthrodiastasis is a useful option in such cases [2] it is typically indicated in the child with stiff hips and non-reducible hinge abduction. Arthrodiastasis (articulated hip distractions) can be combined with soft tissue release.

Outcome evaluation

The outcome of Perthes disease is evaluated with Stulberg classification [14]. Stulberg I or II might be infrequent with late presented cases. The outcome following any intervention in late presented cases is uncertain compared with preventable surgery performed in the earlier stage of the disease. The outcome is poorer with older onset child, severe collapsed, presence of femoral head deformation, very stiff hip with the presence of severe deformity of the hip. The outcome is better in absence of poor prognostic factors.

Conclusion

The treating physicians should not wait for late cases. Containment must be achieved very early in the course of the disease. Many variables (age of onset, range of motion, presence of femoral head deformity, hinge abduction, extrusion of the femoral head, and enlargement of the femoral head) are to be considered in late-presented cases. It is better to prevent the non-containable hip. It is very difficult to get the spherical head in late-presented cases. The natural history of the non-containable hip is poor. Preventing the non-containable hip is better than trying to cure it.

References

- Shah H. Perthes disease: evaluation and management. *Orthop Clin North Am.* 2014 Jan;45(1):87-97. doi: 10.1016/j.jocl.2013.08.005. Epub 2013 Sep 26. PMID: 24267210.
- Kim SS, Lee CW, Kim HJ, Kim HH, Wang L. Treatment of Late-Onset Legg-Calve-Perthes Disease by Arthrodiastasis. *Clin Orthop Surg.* 2016 Dec;8(4):452-457. doi: 10.4055/cios.2016.8.4.452. Epub 2016 Nov 4. PMID: 27904729; PMCID: PMC5114259.
- Grzegorzewski A, Szymczak W, Synder M, Drobniewski M. Czynniki prognostyczne w chorobie Perthesa [The prognostic factors in perthes disease]. *Chir Narzadow Ruchu Ortop Pol.* 2006;71(3):177-82. Polish. PMID: 17131722.
- Joseph B. Management of Perthes' disease. *Indian J Orthop.* 2015 Jan-Feb;49(1):10-6. doi: 10.4103/0019-5413.143906. PMID: 25593353; PMCID: PMC4292319.
- Shah, H. (2014). Perthes Disease. *Orthopedic Clinics of North America*, 45(1), 87–97. doi:10.1016/j.jocl.2013.08.005.
- Farsetti P, Benedetti-Valentini M, Potenza V, Ippolito E. Valgus extension femoral osteotomy to treat "hinge abduction" in Perthes' disease. *J Child Orthop.* 2012 Dec;6(6):463-9. doi: 10.1007/s11832-012-0453-8. Epub 2012 Nov 16. PMID: 24294308; PMCID: PMC3511687.
- Nakamura J, Kamegaya M, Saisu T, Kenmoku T, Takahashi K, Harada Y. Hip arthrography under general anesthesia to refine the definition of hinge abduction in Legg-Calvé-Perthes disease. *J Pediatr Orthop.* 2008 Sep;28(6):614-8. doi: 10.1097/BPO.0b013e3181842013. Erratum in: *J Pediatr Orthop.* 2013 Oct-Nov;33(7):774. PMID: 18724196.
- Joseph B, Price CT. Principles of containment treatment aimed at preventing femoral head deformation in Perthes disease. *Orthop Clin North Am.* 2011 Jul;42(3):317-27, vi. doi: 10.1016/j.jocl.2011.04.001. Epub 2011 May 6. PMID: 21742143.
- Iwamoto M, Nakashima Y, Nakamura T, Kohno Y, Yamaguchi R, Takamura K. Clinical outcomes of conservative treatment with a non-weight-bearing abduction brace for Legg-Calvé-Perthes disease. *J Orthop Sci.* 2018 Jan;23(1):156-160. doi: 10.1016/j.jos.2017.09.012. Epub 2017 Oct 2. PMID: 28982606.
- Joseph B, Rao N, Mulpuri K, Varghese G, Nair S. How does a femoral varus osteotomy alter the natural evolution of Perthes' disease? *J Pediatr Orthop B.* 2005 Jan;14(1):10-5. doi: 10.1097/01202412-200501000-00002. PMID: 15577301.
- Freeman RT, Wainwright AM, Theologis TN, Benson MK. The outcome of patients with hinge abduction in severe Perthes disease treated by shelf acetabuloplasty. *J Pediatr Orthop.* 2008 Sep;28(6):619-25. doi: 10.1097/BPO.0b013e3181804be0. PMID: 18724197.
- Wright DM, Perry DC, Bruce CE. Shelf acetabuloplasty for Perthes disease in patients older than eight years of age: an observational cohort study. *J Pediatr Orthop B.* 2013 Mar;22(2):96-100. doi: 10.1097/BPB.0b013e32835b5726. PMID: 23164833.
- Dammerer D, Braitto M, Ferlic P, Kaufmann G, Kosiol J, Biedermann R. Long-term clinical and radiological outcome in patients with severe Legg-Calvé-Perthes disease after Chiari pelvic osteotomy: a mean of 14 years follow-up. *Hip Int.* 2021 Feb 10;1120700020988150. doi: 10.1177/1120700020988150. Epub ahead of print. PMID: 33566697.
- Herring JA, Kim HT, Browne R. Legg-Calve-Perthes disease. Part I: Classification of radiographs with the use of the modified lateral pillar and Stulberg classifications. *J Bone Joint Surg Am.* 2004 Oct;86(10):2103-20. PMID: 15466719.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the Journal. The patient understands that his name and initials will not be published, and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

How to Cite this Article

Singh KA, Shah H | Management of Late Presented Perthes Disease | Journal of Karnataka Orthopaedic Association | January-February 2022; 10(1):05-08.