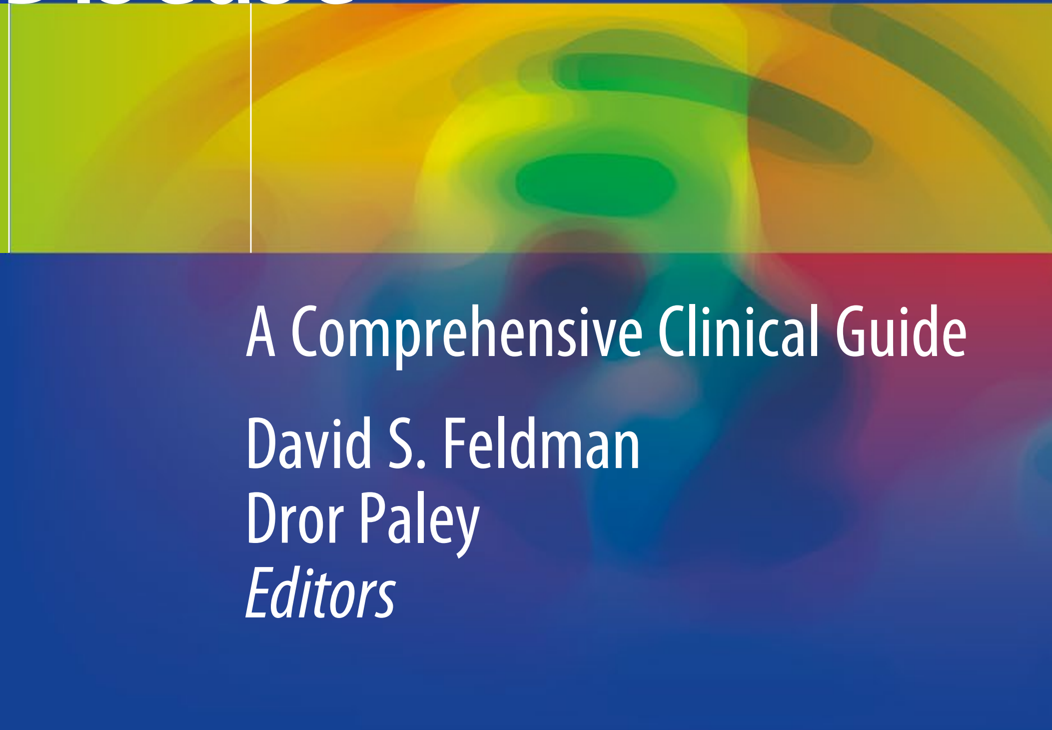
Legg-Calvé-Perthes Disease



A Comprehensive Clinical Guide

David S. Feldman
Dror Paley
Editors

Springer

Legg-Calvé-Perthes Disease

David S. Feldman • Dror Paley
Editors

Legg-Calvé-Perthes Disease

A Comprehensive Clinical Guide

Springer

Editors
David S. Feldman
Paley Orthopedic and Spine Institute
St. Mary's Medical Center
West Palm Beach, FL
USA

Dror Paley
Paley Orthopedic and Spine Institute
St. Mary's Medical Center
West Palm Beach, FL
USA

ISBN 978-1-0716-0852-4 ISBN 978-1-0716-0854-8 (eBook)
https://doi.org/10.1007/978-1-0716-0854-8

© Springer Science+Business Media, LLC, part of Springer Nature 2020
This work is subject to copyright. All rights are reserved by the Publisher, whether the whole or part of the material is concerned, specifically the rights of translation, reprinting, reuse of illustrations, recitation, broadcasting, reproduction on microfilms or in any other physical way, and transmission or information storage and retrieval, electronic adaptation, computer software, or by similar or dissimilar methodology now known or hereafter developed.
The use of general descriptive names, registered names, trademarks, service marks, etc. in this publication does not imply, even in the absence of a specific statement, that such names are exempt from the relevant protective laws and regulations and therefore free for general use.
The publisher, the authors and the editors are safe to assume that the advice and information in this book are believed to be true and accurate at the date of publication. Neither the publisher nor the authors or the editors give a warranty, express or implied, with respect to the material contained herein or for any errors or omissions that may have been made. The publisher remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

This Springer imprint is published by the registered company Springer Science+Business Media, LLC part of Springer Nature
The registered company address is: 1 New York Plaza, New York, NY 10004, U.S.A

Preface

Legg-Calvé-Perthes Disease (LCPD) has baffled treating physicians for over a century. If a patient were to be evaluated by three reputable experienced pediatric orthopedic surgeons, they would likely receive three very divergent opinions. One of my mentors, Dr. Wallace Lehman, has taught me that no matter what is done, one third of the patients will do very well, one third will do “okay,” and the other third will do poorly. Another teacher, Dr. John Wedge, taught me that LCPD is about management; no one method is utilized for treating all. Some will require simple exercise programs, while others may require extensive pelvic and femoral surgery. The treating physician must know how to manage the individual patient.

The purpose of this book as we enter the third decade of the new millennium is to outline all the current treatments of LCPD from around the world and even perhaps future developments that may change our management and outlook regarding this disease. What is obvious from the chapters that follow is that the surgeon treating this disease must have a large arsenal of interventions and procedures in his or her skill set to achieve an optimal result.

No book on LCPD could be written without controversy, as each experienced surgeon has a bias to their method and their algorithm for treatment. I hope that from these chapters one can garner and become acutely aware of all the possibilities and apply them when appropriate. It is certainly possible that in my lifetime this will be a completely medically treated disease, orchestrated with medical intervention bone repair and remodeling. However, until that time, we are left with the following book as a guide to the complex management of the uncommon disease.

West Palm Beach, FL, USA David S. Feldman, MD

About the Editors

David S. Feldman, MD is trained in Pediatric Orthopedic Surgery at the Hospital for Sick Children in Toronto and Orthopedic Surgery at NYU Hospital for Joint Diseases. Dr. Feldman practiced for over 20 years at NYU and rose to a chaired Professor of Orthopedics and Pediatrics and Chief of Pediatric Orthopedic Surgery. In 2015, he moved to the Paley Orthopedic and Spine Institute, where he can solely focus on advancing the care and treatment of several rare conditions such as Perthes, scoliosis, skeletal dysplasias, MHE, and arthrogryposis. Dr. Feldman has written numerous articles and book chapters, operates in several foreign countries, and has lectured around the world on innovation and improving outcomes in his areas of interest.

Besides being passionate about his work and his patients, Dr. Feldman loves spending quality time with his family, contemporary art, and seeing the world on his bicycle.

Dr. Feldman was born in Philadelphia and now lives in West Palm Beach, Florida, but when not there you may find him on his farm in Massachusetts.

Dror Paley, MD, FRCSC is the founder and director of the Paley Orthopedic and Spine Institute in West Palm Beach, Florida, 2009 to present and the Paley European Institute in Warsaw, Poland, 2018 to present. He was the founder and director of the Rubin Institute for Advanced Orthopedics, Baltimore, 2001–2009. He was Professor and Chief of Pediatric Orthopedics at University of Maryland, 1987–2001. Prior to that, he obtained subspecialty fellowship training in Pediatric Orthopedics, Hand Surgery, Trauma Surgery, and Limb Lengthening and External Fixation Surgery, 1985–1987. He completed orthopedic surgery residency at University of Toronto 1980–1985, and internship Johns Hopkins 1979–1980. He received his medical degree from the University of Toronto Medical School, Toronto, Canada, in 1979. He is internationally recognized for his expertise in limb lengthening and reconstruction. Dr. Paley trained under the guidance of Prof. Gavril Ilizarov, during multiple visits to Kurgan, Soviet Union, in 1986, 1987, and 1988. He introduced the Ilizarov method to the USA and Canada in 1987. He was the founder and first president of the Limb Lengthening and Reconstruction Society, 1989, and of the International Limb Lengthening and Reconstruction Society in 2015. He is the recipient of numerous awards including Gubernatorial Citation 1990, Pauwel's Medal in Clinical Biomechanics 1997, Best paper/poster award by SICOT, AAOS, POSNA, AORS; best illustrated medical textbook 2003; Health Professional of the Year 2011; Health Hero of the Year 2013; and Florida Influential Business Leader 2020. He has published over 150 peer-reviewed articles, 5 books and 60 book chapters. His notable publication is *Principles of Deformity Correction*, Springer (2002, 2005). The CORA method of deformity analysis in this text has become the gold standard for deformity planning in orthopedics. He is currently completing a book on the congenital lower limb deformities. He developed 100+ surgical procedures including SUPERhip, SUPERknee, SUPERankle, SHORDT, Paley-Weber pateloplasty, ulnarization, Paley rotationplasty, modified Judet quadricepsplasty, Paley XUNION for CPT, MHE forearm correction, and four-segment achondroplasia lengthening. He has had an interest in Perthes disease since 1989 when he first developed the method of articulated hip distraction for Perthes and continues to the present with his technique of Femoral Head Reduction Osteotomy. He developed the Multiplier method of prediction of leg length discrepancy and timing of epiphysiodesis now available using the iOS app Paley Growth. He was adjunct Professor of Orthopedic Surgery and consultant at Hospital for Sick Children, University of Toronto, 2010–2014. He currently holds an academic appointment as Professor of Orthopedics at the University of Vermont and Florida Atlantic University. He is on the Board of Governors of St. Mary's Medical Center. He has performed 25,000+ surgeries. He is fluent, writes, reads, and lectures in six languages. He runs the Paley Foundation and does mission trips around the world. He lives with his wife Jennifer and stepson Daelan and has three grown children, Benjamin, Jonathan, and Aviva and one granddaughter. His hobbies include reading history, skiing, road and mountain biking, rock climbing, and scuba diving.

Contents

Contributors

Keith Bachmann, MD Departments of Orthopaedic Surgery, and Public Health Sciences, University of Virginia, Charlottesville, VA, USA

David S. Feldman, MD Paley Orthopedic and Spine Institute, St. Mary's Medical Center, West Palm Beach, FL, USA

José Roberto Bevilacqua Guarniero, MD Ortopedia e Traumatologia, das Clínicas HCFMUSP Universidade de Sao Paulo, Sao Paulo, SP, Brazil

Roberto Guarniero, MD, PhD Ortopedia e Traumatologia, das Clínicas HCFMUSP Universidade de Sao Paulo, Sao Paulo, SP, Brazil

Yasmin D. Hailer, MD, PhD Section of Orthopedic Surgery, Department of Surgical Sciences, University Hospital Uppsala, Uppsala, Sweden

Joshua E. Hyman, MD Columbia University College of Physicians and Surgeons, Morgan Stanley Children's Hospital of New York-Presbyterian, New York, NY, USA

Nuno Craveiro Lopes, MD Orthopedic Department, Portuguese Red Cross Hospital, Lisbon, Portugal

Surya N. Mundluru, MD McGovern Medical School at the University of Texas Health Science Center, Houston, TX, USA

UT Physicians Orthopedics at Ironman Sports Medicine Institute – Texas Medical Center, Houston, TX, USA

Dror Paley, MD, FRCSC Paley Orthopedic and Spine Institute, St. Mary's Medical Center, West Palm Beach, FL, USA

Neeraj M. Patel, MD, MPH, MBS Ann & Robert H. Lurie Children's Hospital of Chicago, Chicago, IL, USA

Katherine Rosenwasser, MD New York-Presbyterian Hospital-Columbia and Cornell, New York, NY, USA

Jonathan Schoenecker, MD, PhD Vanderbilt Children's Orthopaedics, Nashville, TN, USA

Perry Schoenecker, MD Shriners Hospitals for Children, St. Louis, MO, USA

Evan Trupia, MD Morgan Stanley Children's Hospital of New York-Presbyterian, Columbia University Medical Center, New York, NY, USA

Vidyadhar Upasani, MD Department of Orthopedic Surgery, Rady Children's Hospital San Diego, San Diego, CA, USA

1

History and Epidemiology of Legg-Calvé-Perthes Disease – Who is at Risk and How do We Diagnose it?

Yasmin D. Hailer

History

The aseptic osteonecrosis of the femoral head in children, today called Legg-Calvé-Perthes disease (LCPD), was described almost simultaneously by three different authors in 1910 [1–3]. None of the authors, however, had an exact understanding regarding the underlying etiology of the disease.

Legg described trauma as a possible activator, Calvé thought it was due to an altered osteogenesis, and Perthes suggested that an inflammatory process could change the shape of the femoral head's epiphysis. It wasn't until 1963 that the name "Legg-Calvé-Perthes disease" was established through the official medical subject headings (MESH) of the national Library of Medicine [4]. Prior to this, the condition had many names, such as arthritis deformans juvenilis, coxa plana, and osteochondritis deformans juvenilis.

Although the etiology of LCPD still remains unknown, clinical and experimental evidence support the theory that the disruption of the blood supply to the femoral head is a key pathogenic event associated with the disease's process [5].

Epidemiology

The annual incidence of LCPD varies between 0.45/100,000 reported among black children in South Africa [6] and 21/100,000 for children in Liverpool, England [7]. The systematic review of Perry et al. [8] demonstrates that ethnicity plays an important role in the occurrence of the disease. The incidence of LCPD in black people is 0.5 case per 100,000 children aged <15 years and rises to 1.7 cases in mixed-raced

---

Y. D. Hailer (✉)
Section of Orthopedic Surgery, Department of Surgical Sciences,
University Hospital Uppsala, Uppsala, Sweden
e-mail: yasmin.hailer@surgsci.uu.se

D. S. Feldman, D. Paley (eds.), *Legg-Calvé-Perthes Disease*,
https://doi.org/10.1007/978-1-0716-0854-8_1

persons, and the highest incidence is found in white persons with about 10.8 cases per 100,000 children under 15 years. Furthermore, Perry et al. illustrate in the same review a correlation between the incidence of LCPD and latitude, with the disease being more common in northern countries. The possibility of detection bias playing a role in this analysis cannot be ignored, as the study compared northern Europe and the United States – both with well-established health systems – with poorer countries closer to the equator. However, the suspicion of detection bias is weakened by the study of Joseph et al. [9] showing a similar incidence of LCPD in a population of schoolchildren and a population of children diagnosed within the health-care system in the same region [8].

Many studies reveal an impact of socioeconomic status on the incidence of LCPD, with higher incidence in urban environments with greater socioeconomic deprivation [10–12]. Most studies on socioeconomic differences come from the United Kingdom and Ireland, however, a fact which might reduce their external validity. Studies focusing on passive or maternal smoking [13, 14] could not confirm an association with socioeconomic conditions.

Boys are affected four times as often as girls, and in 8–24% of patients, the disease is bilateral [15, 16]. LCPD is usually diagnosed among children younger than 15 years of age, with a peak of onset between 5 and 8 years of age [17].

Risk Factors and Prediction

A compromised blood supply to the epiphysis of the femoral head seems to be the underlying pathomechanism of LCPD, but the cause of the disturbed blood flow is still unknown. Earlier studies report thromboembolic events due to coagulation abnormalities. Factor V Leiden mutations [18, 19] and decreased levels of protein C and S [20, 21] have been reported for patients with LCPD (illustrated in Fig. 1.1), but the findings are inconsistent [22]. Perry et al. [23] found a reduced diameter and reduced blood flow of the brachial artery in patients with LCPD compared to an age- and gender-matched control group. Hailer et al. [24] conducted a register-based study and found that patients with LCPD have a higher risk for cardiovascular diseases in a life period. The fact that LCPD can be induced experimentally through hypertension in spontaneous hypertensive rats [25] makes vascular involvement plausible. To the best of our knowledge, no study investigating blood pressure in children and its association with LCPD has been published, but hypertension is a highly underdiagnosed disease in children [26] and could play an additional role in the pathogenesis of LCPD [27]. Maternal and passive smoking during pregnancy has been implicated in the pathogenesis of atherosclerosis [28, 29], hypertension among children [30], and LCPD [31]. The nicotine could provoke oxidative stress in the offspring, which could affect not only vascular formation but also growth, and could also have neurotoxic effects by disrupting neurodevelopment. Patients with LCPD were found to have had lower birth

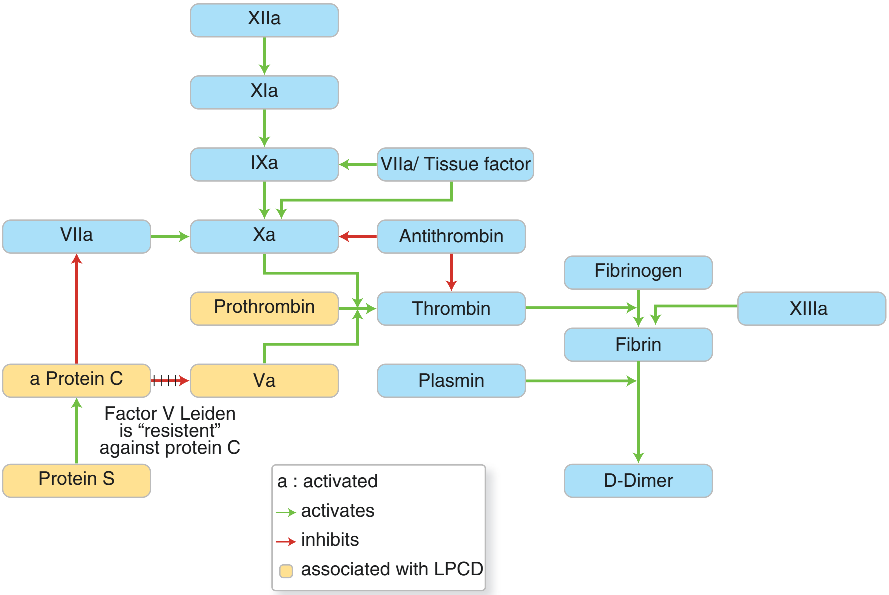


Fig. 1.1 Clotting cascade and its possible associations with LCPD

weights [32] or were shorter at birth [17] and had decreased skeletal maturation at time of diagnosis [33, 34].

Catterall developed the concept of the “susceptible child” for LCPD. He defined this as “primarily boys between ages five and nine years who are smaller than their friends and first-degree relatives and have delayed bone age. In all other aspects healthy, and very active children; many of them are hyperactive” [35]. Hyperactivity in children is hard to quantify, though in children with ADHD a hyperactive behavior pattern is well described [36]. More recent publications confirm an association between LCPD and both ADHD and high levels of physical activity [37].

An interesting issue is the pathway of insulin-like growth factor in the discussion of LCPD. Neidel et al. [38] and Matsumoto et al. [39] found lower levels of circulating IGF-1 in patients at the onset of LCPD which normalized within 2 years during the course of the disease. IGF-1 plays an important role in the growth of many tissues during childhood and could explain the delayed skeletal maturation, hyperactive behavior [5], and possibly also vascular abnormalities seen in patients with LCPD. That said, other pathways could also have a similar impact on both angiogenesis and neurodevelopment in patients with LCPD.

In summary, it seems that on the basis of a possible systemic vascular fragility, hyperactivity might trigger or catalyze LCPD in the epiphysis of the femoral head [27]. The higher risk for LCPD patients to have other osteonecroses develop, strenghthens this hyposthesis further [40].

What Makes it Worse?

To determine the final outcome, Stulberg [41] stated that the congruency of the femoral head in relation to the acetabulum at skeletal maturity is important for estimating the risk for osteoarthritis in a patient's hip before the age of 50 years. LCPD is usually diagnosed among children younger than 15 years of age with a peak of onset being between 5 and 8 years [17, 42]. The prognosis of LCPD is determined by the age at time of disease onset, with a more favorable outcome in younger patients. Wiig et al. [43] found in a register-based study that children below the age of 6 years at onset had a good outcome regardless of treatment, while Herring et al. [44] found the cutoff to be 8 years or younger. In addition to the age at onset, the extent of the necrosis seems to have a prognostic value. There are several radiological classifications of the disease [45], Catterall [46], Salter-Thompson [47], and Lateral Pillar [48] all of which are used to indicate the extent of the necrosis of the femoral head and its correlation to the prognosis. According to Nakamura et al., a decrease in range of abduction is associated with a poorer prognosis despite a young age at onset [49].

LCPD is more common in males than in females, but females have a less favorable prognosis [16, 45]. The poorer prognosis of LCPD in females could be because females reach puberty and skeletal maturity earlier than males, thus giving the femoral head less time for remodeling.

Even though it has been stated that children with bilateral disease have a worse prognosis [50], Wiig et al. found that this is only the case if the onset is sequential. Those with concurrent bilateral onset have similar outcomes as children with unilateral manifestation [51].

Symptoms and Clinical and Radiological Examination

The clinical symptoms of LCPD are variable and can be subtle, especially in its early stages. The child often presents with intermittent limping and pain in the groin, thigh, or knee. All symptoms can occur individually or appear in combination, which at times makes the diagnosis difficult to differentiate from knee pathologies or other hip pathologies such as transient synovitis (also known as coxitis simplex). Slight limping or difficulty jumping on one leg may reveal weakness of the hip muscles. This and reduced range of motion in the hip are some of the typical signs which should lead to suspicion of LCPD. The clinical examination often shows a reduced range of motion of the hip joint, especially in abduction and external rotation. The "figure of 4", or FABER's test (Fig. 1.2), is often positive in the affected hip; however, it can be subtle. In more advanced stages, a positive Trendelenburg's sign indicates a weakness of the abductor muscles.

Ultrasound examination can detect joint effusion in early stages, and blood tests such as acute-phase protein (c-reactive protein) and erythrocyte sedimentation rate (ESR) can rule out infection as a differential diagnosis. Having the diagnosis of LCPD in mind, pelvic X-rays in anterior/posterior and frog-lateral views should be

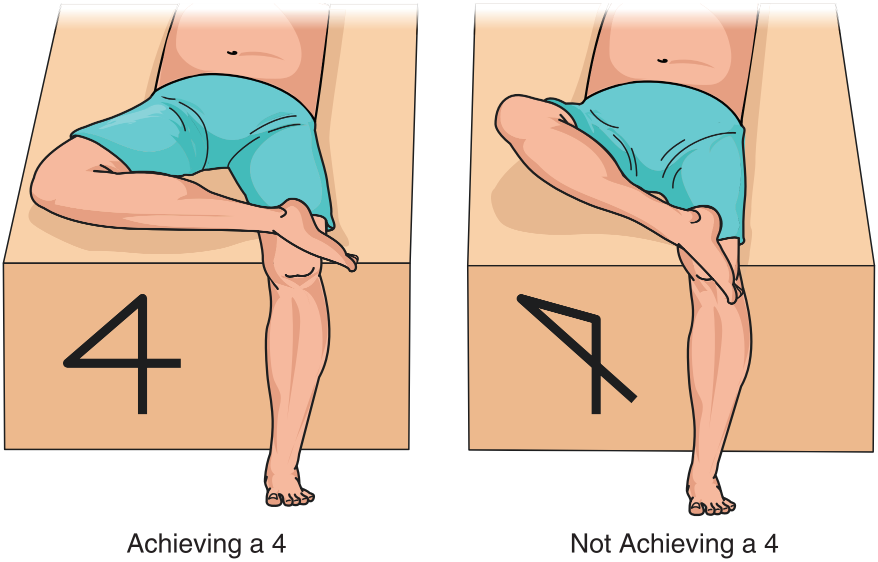
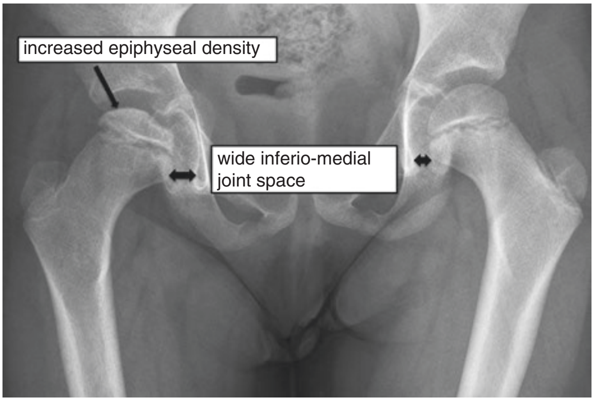


Fig. 1.2 "Figure of 4" of the right leg. (achieving a "4" on the left picture and not achieving a "4" on the right picture)

Fig. 1.3 Early radiographic findings in LCPD of the right hip (dx: right hip)



the next step. Radiographic findings are often normal in the early stage of the disease. If the suspicion of LCPD cannot be confirmed on X-ray, it is still important to inform the parents about the possibility of a LCPD diagnosis, and if the symptoms persist or reappear, the radiographic exam should be repeated.

It is not until 2–3 months after the onset of symptoms that the subtle findings typical for LCPD appear on radiographs: the epiphysis is sclerotic with increased density. Further, the inferomedial joint space can be widened (Fig. 1.3), and in about 25% of cases, a subchondral fracture line is visible (Fig. 1.4). Radiographically, LCPD progresses through four stages: condensation,

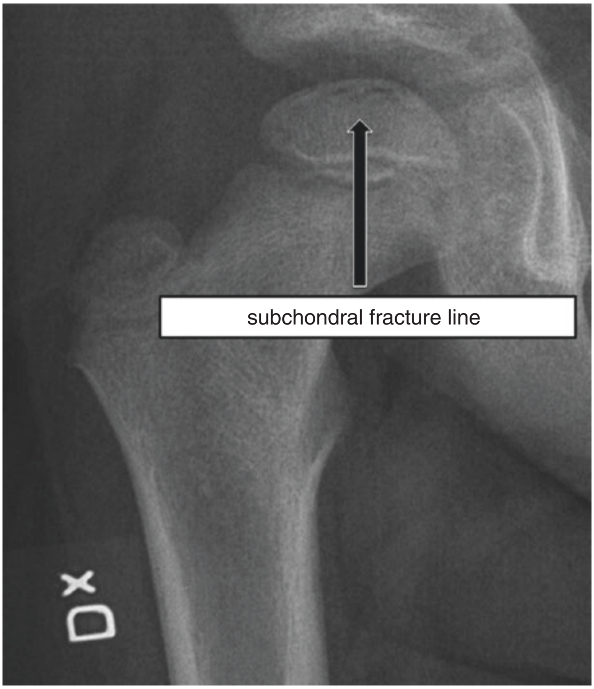


Fig. 1.4 Subchondral fracture line in LCPD (dx: right hip)

fragmentation, repairing, and healing [52]. Joseph et al. modified this categorization in a more detailed classification [53] in order to identify the optimal timing for possible containment surgery. To determine the prognosis of LCPD, Herring et al. suggest focusing on the lateral aspect of the epiphysis in the fragmentation stage of the disease [48].

Magnetic resonance imaging (MRI) is an option in cases with strong suspicion of LCPD. MRI makes the diagnosis obvious, even in early stages when X-rays fail to show the typical changes of the femoral head [54], and is considered to be superior to bone scintigraphy [55]. Kim et al. recommend gadolinium-enhanced MRI with a subtraction technique in order to detect early ischemia and provide information about the blood flow in the epiphysis of the femoral head [56]. The advantage of this technique compared to non-contrast MRI is that it allows one to determine the full extent of the osteonecrosis of the epiphysis and by this the involvement of the lateral pillar before the collapse. This method may also be utilized to predict the amount of collapse based upon the pattern of ischemia [57].

Arthrography of the hip has been recommended as a decision-making tool for containment surgery. While it can contribute useful information, the need for arthrography in LCPD is controversial, and decisions on its use come down to the surgeon preference [58–60].

In patients with confirmed LCPD, a complete blood count will reveal possible other causes of a necrosis of the femoral head such as leukemia or sickle cell anemia. In patients with a family history of LCPD or clotting disorders, further blood exams are recommended in order to detect a possible factor V Leiden mutation, prothrombin G20210A mutation, or elevated levels of factor VIII [61].

References

1. Legg AT. An obscure infection of the hip joint. Boston Med Surg J. 1910;162:202–4.
2. Calvé J. Sur une forme particuliere de coxalgie greffe sur des deformations caracteristiques de l'extremite superieure de femur. Revue de chirurgie. 1910;42:54–84.
3. Perthes G. Über arthritis deformans juvenilis. Deutsche Zeitschrift für Chirurgie. 1910;11:111–59.
4. Loder RT, Skopelja EN. The epidemiology and demographics of Legg-Calve-Perthes' disease. ISRN Orthop. 2011;2011:504393.
5. Kim HK. Legg-Calve-Perthes disease: etiology, pathogenesis, and biology. J Pediatr Orthop. 2011;31(2 Suppl):S141–6.
6. Purry NA. The incidence of Perthes' disease in three population groups in the Eastern Cape region of South Africa. J Bone Joint Surg Br. 1982;64(3):286–8.
7. Hall AJ, Barker DJ, Dangerfield PH, Taylor JF. Perthes' disease of the hip in Liverpool. Br Med J (Clin Res Ed). 1983;287(6407):1757–9.
8. Perry DC, Machin DM, Pope D, Bruce CE, Dangerfield P, Platt MJ, et al. Racial and geographic factors in the incidence of Legg-Calve-Perthes' disease: a systematic review. Am J Epidemiol. 2012;175(3):159–66.
9. Joseph B, Chacko V, Rao BS, Hall AJ. The epidemiology of Perthes' disease in South India. Int J Epidemiol. 1988;17(3):603–7.
10. Hall AJ, Barker DJ, Lawton D. The social origins of Perthes' disease of the hip. Paediatr Perinat Epidemiol. 1990;4(1):64–70.
11. Kealey WD, Moore AJ, Cook S, Cosgrove AP. Deprivation, urbanisation and Perthes' disease in Northern Ireland. J Bone Joint Surg Br. 2000;82(2):167–71.
12. Perry DC, Bruce CE, Pope D, Dangerfield P, Platt MJ, Hall AJ. Perthes' disease of the hip: socioeconomic inequalities and the urban environment. Arch Dis Child. 2012;97(12):1053–7.
13. Gordon JE, Schoenecker PL, Osland JD, Dobbs MB, Szymanski DA, Luhmann SJ. Smoking and socio-economic status in the etiology and severity of Legg-Calve-Perthes' disease. J Pediatr Orthop B. 2004;13(6):367–70.
14. Bahmanyar S, Montgomery SM, Weiss RJ, Ekbom A. Maternal smoking during pregnancy, other prenatal and perinatal factors, and the risk of Legg-Calve-Perthes disease. Pediatrics. 2008;122(2):e459–64.
15. Salter RB. The present status of surgical treatment for Legg-Perthes disease. J Bone Joint Surg Am. 1984;66(6):961–6.
16. Guille JT, Lipton GE, Szoke G, Bowen JR, Harcke HT, Glutting JJ. Legg-Calve-Perthes disease in girls. A comparison of the results with those seen in boys. J Bone Joint Surg Am. 1998;80(9):1256–63.
17. Wiig O, Terjesen T, Svenningsen S, Lie SA. The epidemiology and aetiology of Perthes' disease in Norway. A nationwide study of 425 patients. J Bone Joint Surg Br. 2006;88(9):1217–23.
18. Eldridge J, Dilley A, Austin H, El-Jamil M, Wolstein L, Doris J, et al. The role of protein C, protein S, and resistance to activated protein C in Legg-Perthes disease. Pediatrics. 2001;107(6):1329–34.
19. Balasa VV, Gruppo RA, Glueck CJ, Wang P, Roy DR, Wall EJ, et al. Legg-Calve-Perthes disease and thrombophilia. J Bone Joint Surg Am. 2004;86-A(12):2642–7.
20. Glueck CJ, Glueck HI, Greenfield D, Freiberg R, Kahn A, Hamer T, et al. Protein C and S deficiency, thrombophilia, and hypofibrinolysis: pathophysiologic causes of Legg-Perthes disease. Pediatr Res. 1994;35(4 Pt 1):383–8.

21. Levin C, Zalman L, Shalev S, Mader R, Koren A. Legg-Calve-Perthes disease, protein C deficiency, and beta-thalassemia major: report of two cases. J Pediatr Orthop. 2000;20(1):129–31.
22. Lopez-Franco M, Gonzalez-Moran G, De Lucas JC Jr, Llamas P, de Velasco JF, Vivancos JC, et al. Legg-Perthes disease and heritable thrombophilia. J Pediatr Orthop. 2005;25(4):456–9.
23. Perry DC, Green DJ, Bruce CE, Pope D, Dangerfield P, Platt MJ, et al. Abnormalities of vascular structure and function in children with Perthes disease. Pediatrics. 2012;130(1):e126–31.
24. Hailer YD, Montgomery SM, Ekbom A, Nilsson OS, Bahmanyar S. Legg-Calve-Perthes disease and risks for cardiovascular diseases and blood diseases. Pediatrics. 2010;125(6):e1308–15.
25. Hirano T, Iwasaki K, Yamane Y. Osteonecrosis of the femoral head of growing, spontaneously hypertensive rats. Acta Orthop Scand. 1988;59(5):530–5.
26. Hansen ML, Gunn PW, Kaelber DC. Underdiagnosis of hypertension in children and adolescents. JAMA. 2007;298(8):874–9.
27. Hailer YD. Legg-Calvé-Perthes Disease – Is it just the hip? Epidemiological, Clinical and Psychosocial studies with special focus on Etiology. Ph.D. thesis, Uppsala: Uppsala University; 2014.
28. Kosecik M, Erel O, Sevinc E, Selek S. Increased oxidative stress in children exposed to passive smoking. Int J Cardiol. 2005;100(1):61–4.
29. Aycicek A, Ipek A. Maternal active or passive smoking causes oxidative stress in cord blood. Eur J Pediatr. 2008;167(1):81–5.
30. Lawlor DA, Najman JM, Sterne J, Williams GM, Ebrahim S, Davey SG. Associations of parental, birth, and early life characteristics with systolic blood pressure at 5 years of age: findings from the Mater-University study of pregnancy and its outcomes. Circulation. 2004;110(16):2417–23.
31. Garcia Mata S, Ardanaz Aicua E, Hidalgo Ovejero A, Martinez GM. Legg-Calve-Perthes disease and passive smoking. J Pediatr Orthop. 2000;20(3):326–30.
32. Lappin K, Kealey D, Cosgrove A, Graham K. Does low birthweight predispose to Perthes' disease? Perthes' disease in twins. J Pediatr Orthop B. 2003;12(5):307–10.
33. Weiner DS, O'Dell HW. Legg-Calve-Perthes disease. Observations on skeletal maturation. Clin Orthop Relat Res. 1970;68:44–9.
34. Burwell RG, Dangerfield PH, Hall DJ, Vernon CL, Harrison MH. Perthes' disease. An anthropometric study revealing impaired and disproportionate growth. J Bone Joint Surg Br. 1978;60-B(4):461–77.
35. Catterall A. Thoughts on the Etiology of Perthes' diseases. Iowa Orthop J. 1984;4:34–6.
36. Biederman J, Faraone SV. Attention-deficit hyperactivity disorder. Lancet. 2005;366(9481):237–48.
37. Hailer YD, Nilsson O. Legg-Calve-Perthes disease and the risk of ADHD, depression, and mortality. Acta Orthop. 2014;85(5):501–5.
38. Neidel J, Schonau E, Zander D, Rutt J, Hackenbroch MH. Normal plasma levels of IGF binding protein in Perthes' disease. Follow-up of previous report. Acta Orthop Scand. 1993;64(5):540–2.
39. Matsumoto T, Enomoto H, Takahashi K, Motokawa S. Decreased levels of IGF binding protein-3 in serum from children with Perthes' disease. Acta Orthop Scand. 1998;69(2):125–8.
40. Hailer YD, Hailer NP. Is Legg-Calvé-Perthes Disease a Local Manifestation of a Systemic Condition? Clin Orthop Relat Res. 2018;476:1055–64.
41. Stulberg SD, Cooperman DR, Wallensten R. The natural history of Legg-Calve-Perthes disease. J Bone Joint Surg Am. 1981;63(7):1095–108.
42. Perry DC, Skellorn PJ, Bruce CE. The lognormal age of onset distribution in Perthes' disease: an analysis from a large well-defined cohort. Bone Joint J. 2016;98-b(5):710–4.
43. Wiig O, Terjesen T, Svenningsen S. Prognostic factors and outcome of treatment in Perthes' disease: a prospective study of 368 patients with five-year follow-up. J Bone Joint Surg Br. 2008;90(10):1364–71.
44. Herring JA, Kim HT, Browne R. Legg-Calve-Perthes disease. Part II: prospective multicenter study of the effect of treatment on outcome. J Bone Joint Surg Am. 2004;86-A(10):2121–34.

45. Catterall A. The natural history of Perthes' disease. J Bone Joint Surg Br. 1971;53(1):37–53.
46. Catterall A. Natural history, classification, and x-ray signs in Legg-Calve-Perthes' disease. Acta Orthop Belg. 1980;46(4):346–51.
47. Salter RB, Thompson GH. Legg-Calve-Perthes disease. The prognostic significance of the subchondral fracture and a two-group classification of the femoral head involvement. J Bone Joint Surg Am. 1984;66(4):479–89.
48. Herring JA, Neustadt JB, Williams JJ, Early JS, Browne RH. The lateral pillar classification of Legg-Calve-Perthes disease. J Pediatr Orthop. 1992;12(2):143–50.
49. Nakamura J, Kamegaya M, Saisu T, Kakizaki J, Hagiwara S, Ohtori S, et al. Outcome of patients with Legg-Calve-Perthes onset before 6 years of age. J Pediatr Orthop. 2015;35(2):144–50.
50. Van den Bogaert G, de Rosa E, Moens P, Fabry G, Dimeglio A. Bilateral Legg-Calve-Perthes disease: different from unilateral disease? J Pediatr Orthop B. 1999;8(3):165–8.
51. Wiig O, Huhnstock S, Terjesen T, Pripp AH, Svenningsen S. The outcome and prognostic factors in children with bilateral Perthes' disease: a prospective study of 40 children with follow-up over five years. Bone Joint J. 2016;98-b(4):569–75.
52. Waldenström H. The first stages of coxa plana. J Bone Joint Surg Am. 1938;20(3):559–66.
53. Joseph B, Varghese G, Mulpuri K, Narasimha Rao K, Nair NS. Natural evolution of Perthes disease: a study of 610 children under 12 years of age at disease onset. J Pediatr Orthop. 2003;23(5):590–600.
54. Kaniklides C, Lonnerholm T, Moberg A, Sahlstedt B. Legg-Calve-Perthes disease. Comparison of conventional radiography, MR imaging, bone scintigraphy and arthrography. Acta Radiol. 1995;36(4):434–9.
55. Uno A, Hattori T, Noritake K, Suda H. Legg-Calve-Perthes disease in the evolutionary period: comparison of magnetic resonance imaging with bone scintigraphy. J Pediatr Orthop. 1995;15(3):362–7.
56. Kim HK, Kaste S, Dempsey M, Wilkes D. A comparison of non-contrast and contrast-enhanced MRI in the initial stage of Legg-Calve-Perthes disease. Pediatr Radiol. 2013;43(9):1166–73.
57. Kim HK, Wiesman KD, Kulkarni V, Burgess J, Chen E, Brabham C, et al. Perfusion MRI in early stage of Legg-Calve-Perthes disease to predict lateral pillar involvement: a preliminary study. J Bone Joint Surg Am. 2014;96(14):1152–60.
58. Klisic P, Blazevic U, Seferovic O. Approach to treatment of Legg-Calve-Perthes disease. Clin Orthop Relat Res. 1980;150:54–9.
59. Devalia KL, Wright D, Sathyamurthy P, Prasad P, Bruce C. Role of preoperative arthrography in early Perthes disease as a decision-making tool. Is it really necessary? J Pediatr Orthop B. 2007;16(3):196–200.
60. Kamegaya M, Saisu T, Takazawa M, Nakamura J. Arthrographic indicators for decision making about femoral varus osteotomy in Legg-Calve-Perthes disease. J Child Orthop. 2008;2(4):261–7.
61. Vosmaer A, Pereira RR, Koenderman JS, Rosendaal FR, Cannegieter SC. Coagulation abnormalities in Legg-Calve-Perthes disease. J Bone Joint Surg Am. 2010;92(1):121–8.

Staging and Classification of Severity of Legg-Calvé-Perthes Disease and Its Relevance

2

Joshua E. Hyman, Katherine Rosenwasser, and Evan Trupia

Legg-Calvé-Perthes disease (LCPD) remains controversial regarding treatment and outcome. In order to bring uniformity to the discussion of management of LCPD, researchers and clinicians must be able to reliably stratify LCPD patients by disease severity and stage at presentation. The intervention offered and ultimately the treatment outcome may be impacted by the disease stage and severity. It is essential to control for these confounding variables by grouping patients accurately and precisely, regardless of observer.

Classification systems are used throughout the field of orthopedic surgery for a multitude of reasons. The goals of a useful system include accurate and thoughtful characterization of a problem, a suggestion of prognosis and/or natural history of a problem, and, ideally, information regarding potential treatment options. Aside from these overarching goals, successful classification systems also must be reproducible, reliable, and validated [1]. To date, classification systems of LCPD serve to describe the stage of the disease (Waldenstrom and Elizabethtown) and prognosticate (lateral pillar and Salter-Thompson) or define the outcome (Stulberg). Assisting these classifications and likely altering them in the future will be a scintigraphic classification [2] and more recently an MRI classification [3]. Neither has been used routinely as a stand-alone classification.

J. E. Hyman (✉)
Columbia University College of Physicians and Surgeons, Morgan Stanley Children's Hospital of New York-Presbyterian, New York, NY, USA
e-mail: jh736@cumc.columbia.edu

K. Rosenwasser
New York-Presbyterian Hospital-Columbia and Cornell, New York, NY, USA

E. Trupia
Morgan Stanley Children's Hospital of New York-Presbyterian, Columbia University Medical Center, New York, NY, USA

D. S. Feldman, D. Paley (eds.), *Legg-Calvé-Perthes Disease*,
https://doi.org/10.1007/978-1-0716-0854-8_2

In the case of LCPD, previously validated classification systems have focused primarily on disease severity [4]. These have included the Catterall, Herring, and Salter-Thompson systems, which have the ability to prognosticate outcomes and relate their importance to treatment.

Catterall Classification

Catterall proposed a four-group system based on the degree of epiphyseal involvement. It was the first of its kind to highlight the relationship between the extent of head involvement and outcomes [5]. Catterall group I represents involvement of the anterior head (no more than 25%) only, with neither sequestrum nor epiphyseal collapse. Group II also involves the anterior head (up to 50%) but also includes a sequestrum. In group III, up to 75% of the head is involved, mainly sparing sections of the epiphysis lying medial and lateral to the central segment [6], and in group IV, 100% of the head is affected. The system is applied during the fragmentation stage when there is clear demarcation of the necrotic segment of bone from the remaining viable femoral head [7]. Groups I and II in his series were deemed to be benign, needing little treatment, while groups III and IV may benefit from containment treatment (Fig. 2.1).

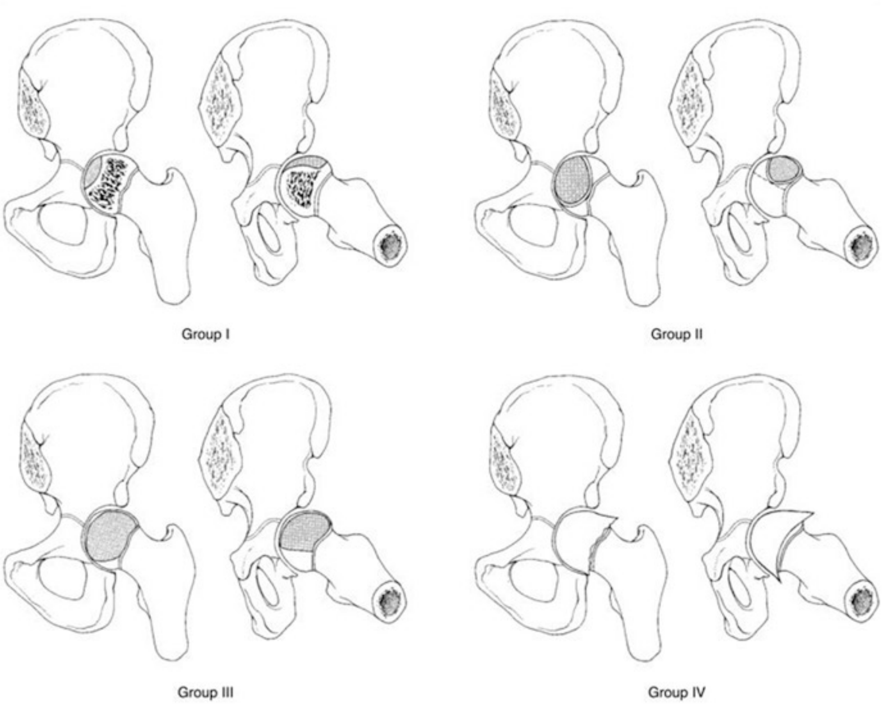


Fig. 2.1 The Catterall classification. (From Skaggs and Tolo [1]. Reprinted with permission)

Additionally, the Catterall classification describes radiographically identifiable "head-at-risk" signs that may be associated with poor outcomes. These are the radiolucent "v" in the lateral portion of the epiphysis, calcification lateral to the epiphysis, lateral subluxation of the femoral head, and a horizontal physis [6, 8].

Initial studies by Hardcastle et al. found the Catterall system to have poor reliability, but reliability improved once groups II and III were combined [6]. Though multiple studies have noted moderate to substantial interobserver and intraobserver reliability [4, 9–16], criticism remains as to whether or not this system can be widely applied successfully. A more recent analysis attempted to determine intra- and interobserver reliability of Catterall as compared to Herring, Salter-Thompson, and Stulberg and showed that this system may have applications in evaluating radiographs taken *prior* to treatment but loses utility when implemented postoperatively [17].

Herring (Lateral Pillar) Classification

The lateral pillar or Herring classification system groups patients according to the height of the capital femoral epiphysis at the early stages of fragmentation and has been shown to provide prognostic information regarding final femoral head sphericity [18]. The classification was originally described as a three-category system with group A demonstrating no involvement of the lateral pillar, group B with greater than 50% of lateral pillar height maintained, and group C with less than 50% of height maintained [18]. It was later modified to include a fourth group, the "B/C border" which includes femoral heads with a poorly ossified lateral pillar or those with exactly 50% height loss [6] (Fig. 2.2).

A study by Herring et al. found near-perfect agreement among 6 pediatric orthopedists' classification of 20 sets of radiographs [6], which represents an improvement over the reliability of the Catterall system. However, as this classification relies upon the fragmentation stage of LCPD in the same manner as the Catterall, the lateral pillar system has limitations for the early-presenting patient. In addition, surgeons must also be aware that up to 1/3 of patients will change categories over time. Because of this, the patient must undergo a "wait and classify" approach until radiographs stabilize prior to treatment decisions being made. This is of particular importance to the older patient, for whom there is higher risk for poor outcomes [19]. Another limitation of this classification is that it may not be applicable to children over age 12 as the remodeling stage will not occur, thus leading to asphericity of the femoral head and poor outcomes [19]. A recent study by Rampal et al. showed, however, that from a radiological perspective Herring's classification has high prognostic value and good reproducibility, despite these limitations, and may be considered useful in determining a head "at-risk," which correlates well with outcome [20].

Thus, applications of the Herring lateral pillar classification are best kept to patients under age 12 during the fragmentation stage of disease and may serve to be a valid tool in identifying surgical candidates.

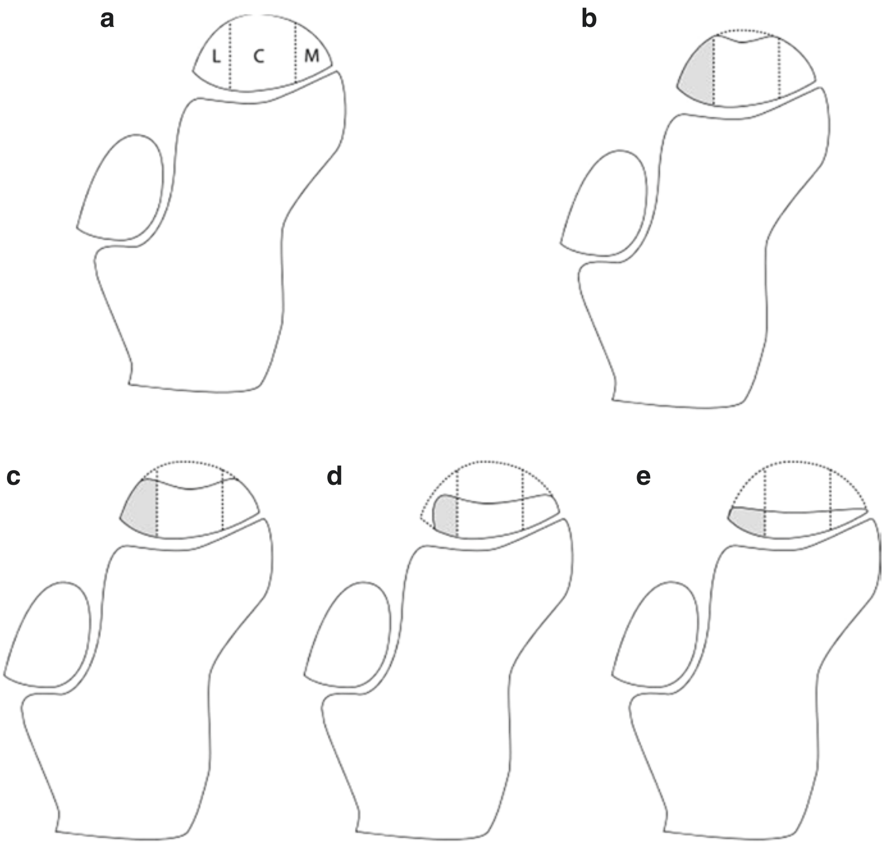


Fig. 2.2 (**a**–**e**) The drawings show a pediatric hip, identifying the three Herring pillars and the height of each Herring class hip. (**a**) The three pillars in the Herring lateral pillar classification are: L = lateral pillar; C = central pillar; and M = medial pillar. (**b**) In Herring Group A, 100% of lateral pillar height is maintained. (**c**) In Herring Group B, greater than 50% of the lateral pillar height is maintained. (**d**) In this example of Herring Group B/C border, the tall and narrow type is shown with slightly more than 50% of the lateral pillar height maintained on a narrow base. (**e**) In Herring Group C, less than 50% of the lateral pillar height is maintained. (From Kollitz and Gee [7]. Reprinted with permission)

Conway in 1993 described a scintigraphic classification attempting to determine whether one can predict by a bone scan early in the disease if there will be lateral pillar collapse [2]. While this showed promise in the published paper, it has not been widely used as prognostic tool for treatment. Kim in 2014 described using perfusion MRI early in the disease to predict the lateral pillar classification [3]. This too will need validation before being widely accepted as a new classification.

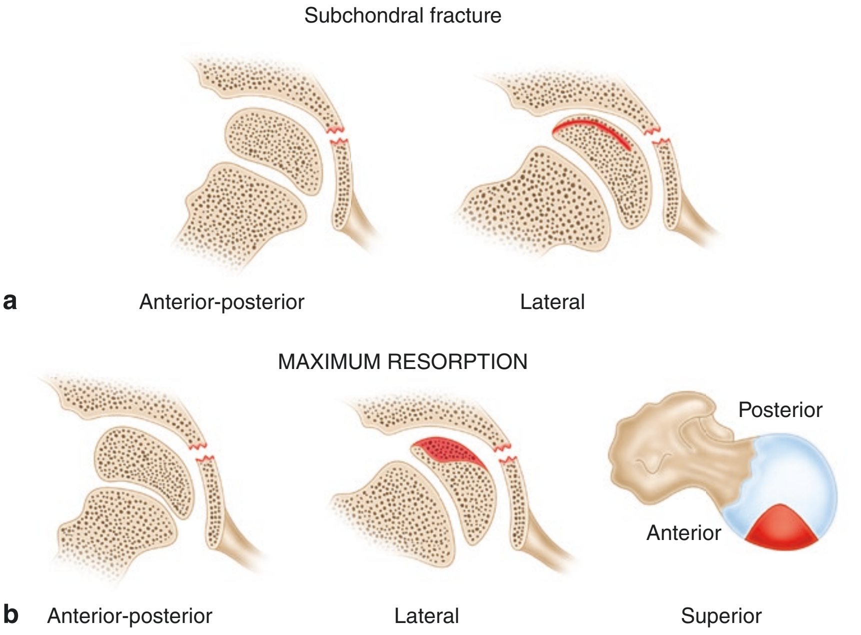


Fig. 2.3 The Salter-Thompson classification. (From Mahadeva et al. [22]. Reprinted with permission)

Salter-Thompson Classification

Based on an experimental model of LCPD in young pigs, the Salter-Thompson classification is a two-group system based on the extent of a radiographic crescent sign, or subchondral fracture [21]. It posits that the fracture initiates a resorptive stage and may indicate the timing of clinical onset of disease. In group A, less than half of the femoral head is involved, and in group B, more than half is involved. In their series of 1057 children (1264 hips), 376 hips were identified that included this subchondral fracture, all of which correlated with subsequent extent of femoral head resorption [21]. While this sign is usually present in the early stage of fragmentation, the utility of this classification system is restricted in that many patients do not possess a fracture at the time of presentation and the time course of radiographic changes in patients that do show this sign is relatively brief [6] (Fig. 2.3).

The utilization of this system lies in early detection of clinically symptomatic disease. Despite this, there are significant limitations to both its practical use and its performance in follow-up radiographic studies of this patient population. As the subchondral fracture line exists only in early presentation, this classification is unable to identify or guide treatment for latecomers. Additionally, studies that attempt to validate this system have difficulty when comparing series of radiographs from the same patient, as the fracture line is no longer identifiable during follow-up [22]. Studies of both the Herring and Salter-Thompson classifications have found interobserver and intraobserver reliability ranging from moderate to substantial [4, 9, 10, 12–14, 23, 24]. Reliability was notably better in initial radiographic comparisons, but in a second set of images, it was noted that genuine comparisons could not be made [22].

Stulberg Classification

The Stulberg classification system, first described in 1981, sought to categorize the final hip configuration in patients with LCPD in a descriptive manner. Class I and II hips retain a spherical femoral head shape, class III began to see a shift towards an ovoid shape, and class IV and V hips demonstrate a flat shape. In Stulberg's original paper, he was able to correlate these shapes at skeletal maturity with characteristic clinical course during the active phase of the disease. Herring et al. further elucidated these class descriptions in defining a class II head as round and fitting within 2 mm of a circle on AP and frog-leg lateral x-rays, class III as out of round by more than 2 mm on either view, and class IV as showing at least 1 cm of flattening over the weight-bearing surface [6]. Class I and II hips show spherical congruency and are generally not at great risk of arthritis development, class III and IV hips show aspherical congruency and may develop mild to moderate arthritis in adulthood, and class V hips show aspherical incongruency, with the highest risk of early-onset arthritis [25].

The utility of this system lies in its potential ability to predict the onset of degenerative joint disease. The potential insight into the long-term prognosis of these patients is something that was lacking in other available classification systems for LCPD [26]. Early reports by Herring were able to show high inter- and intraobserver reliability with this classification system [6], though Neyt et al. called the quality of these analyses into question. In their 1999 study, they found that Stulberg's system lacked reliability and continued to show marked interobserver variability [26].

In its ability to predict outcomes and rate residual deformity, the Stulberg system remains a gold standard in the classification of LCPD. However useful it may be, a main limitation of this system is that it has no role in determining the stage of disease and thus dictating treatment decision-making algorithms. As it is widely known that staging and timing of intervention with respect to disease progression has an impact on ultimate functional outcome, it remains an imperfect system.

Waldenstrom and Modified Waldenstrom (Elizabethtown) Classification

The Waldenstrom classification is a staging classification for LCPD [27]. In his original paper, Waldenstrom noted that the femoral epiphysis produces characteristic radiographic changes as the disease progresses and classified these changes into four temporal stages: osteonecrosis, fragmentation, reossification, and healed [28]. To more clearly describe the various changes in the femur and acetabulum and increase clinical utility, Joseph et al. further modified the first three stages of the Waldenstrom classification (modified Waldenstrom or Elizabethtown) into early (A) and late (B) [29]. A preliminary study among three surgeons at their center found substantial interobserver and intraobserver reliability [29].

After their review of the natural history of LCPD showed that the amount of femoral head deformity and epiphyseal extrusion is demonstrably greater during later versus early stages of fragmentation, Joseph et al. published several retrospective studies showing that children who were treated surgically during early fragmentation had a greater chance of retaining femoral head sphericity versus children operated on during later stages [30, 31]. With early evidence confirming that differentiating between early and late stages of fragmentation is clinically important, Joseph et al. advocated the use of the modified Waldenstrom system to guide treatment decision-making.

The modified Waldenstrom system consists of seven stages. Stage IA demonstrates a mostly or entire sclerotic epiphysis without loss of height. Stage IB demonstrates similar sclerosis but with the addition of epiphyseal height loss without fragmentation. Stage IIA shows early fragmentation with one or two fissures visible on the AP or frog-leg lateral x-ray. Stage IIB demonstrates advanced fragmentation with no bone present lateral to the epiphysis. Stage IIIA begins to show new bone formation lateral to the site of fragmentation. Stage IIIB represents increased bone formation covering one third of the width of the epiphysis, and stage IV demonstrates complete healing without radiographic evidence of avascular bone [32] (Fig. 2.4).

To confirm the modified Waldenstrom system's broader applicability and further establish the classification as an important clinical and research tool, Hyman et al. were able to demonstrate substantial to near-perfect agreement between 20 surgeons from 4 countries across North America and Europe, with a wide range of experience and training, assessing 80 pairs of radiographs encompassing the full range of disease [32]. In this study, weighted kappa, which takes into account the relative distance between two observations, was used to reflect the importance of surgeons' ability to differentiate between very early and very late stages of disease. Nonetheless, it also remains critical for surgeons to demonstrate an ability to distinguish between stages IIA and IIB (early and late fragmentation, respectively), as there are relevant clinical implications. To that end, interobserver and intraobserver agreement remained substantial after reorganizing surgeon responses into two groups: early (stage IIA or lower) and late (stage IIB or higher) LCPD. This is a significant conclusion, as previous research by Joseph et al. has shown that final femoral head sphericity is improved if surgery occurs prior to the late stage of fragmentation (IIB) [31]. To provide higher level of evidence supporting this hypothesis, the International Perthes Study Group (IPSG) is currently enrolling patients in prospective studies using the modified Waldenstrom classification as a reliable clinical and research instrument. It is also expected that these studies will provide information regarding the utility of the full, seven-stage classification.

Stage IA:
All or part of the epiphysis is sclerotic. There is no loss of height of the epiphysis.

Stage IB:
There is sclerosis of the epiphysis. There is loss of epiphyseal height. There is no fragmentation of the epiphysis.

Stage IIA:
The epiphysis has just begun to fragment. one or two vertical fissures are present either in the AP view or the Lauenstein frog-lateral view.

Stage IIB:
Fragmentation is advanced. There is no new bone lateral to the fragmented epiphysis.

Stage IIIA:
Early new bone is visible lateral to the fragmented epiphysis (arrows). The texture of the new bone is not normal; it is more “porotic” and covers less than a third of the width of the epiphysis.

Stage IIIB:
New bone is normal texture covers more than one third of the width of the epiphysis.

Stage IV:
Healing is complete. There is no radiographic evidence of avascular bone.

Fig. 2.4 The modified Waldenstrom (Elizabethtown) classification. (From Hyman et al. [8]. Reprinted with permission)

Works Cited

1. Garbuz DS, Masri BA, Esdaile J, Duncan CP. Classification systems in orthopaedics. J Am Acad Orthop Surg. 2002;10(4):290–7. http://www.ncbi.nlm.nih.gov/pubmed/15089078.
2. Conway JJ. A scintigraphic classification of Legg-Calvé-Perthes disease. Semin Nucl Med. 1993;XXIII:274–95.
3. Kim HK, et al. Perfusion MRI in early stage of Legg-Calvé-Perthes disease to predict lateral pillar involvement: a preliminary study. J Bone Joint Surg Am. 2014;96:1152–60.

4. Park MS, Chung CY, Lee KM, Kim TW, Sung KH. Reliability and stability of three common classifications for Legg-Calvé-Perthes disease hip. Clin Orthop Relat Res. 2012;470:2376–82. https://doi.org/10.1007/s11999-012-2314-7.
5. Catterall A. The natural history of Perthes' disease. J Bone Joint Surg Br. 1971;53-B(1):37–53. http://web.jbjs.org.uk/cgi/content/abstract/53-B/1/37%0Ahttp://web.jbjs.org.uk/cgi/reprint/53-B/1/37.pdf.
6. Herring JA, Kim HT, Browne R. Legg-Calve-Perthes disease. Part I: classification of radiographs with use of the modified lateral pillar and Stulberg classifications. J Bone Joint Surg Am. 2004;86-A(10):2103–20. https://doi.org/10.1097/BPO.0000000000000157.
7. Kim HKW, Herring JA. Pathophysiology, classifications, and natural history of Perthes disease. Orthop Clin North Am. 2011;42(3):285–95. https://doi.org/10.1016/j.ocl.2011.04.007.
8. Lloyd-Roberts GC, Catterall A, Salamon PB. A controlled study of the indications for and the results of femoral osteotomy in Perthes' disease. J Bone Joint Surg Br. 1976;58(1):31–6. http://www.ncbi.nlm.nih.gov/pubmed/1270493.
9. Nathan Sambandam S, Gul A, Shankar R, Goni V. Reliability of radiological classifications used in Legg-Calve-Perthes disease. J Pediatr Orthop B. 2006;15(4):267–70. https://doi.org/10.1097/01202412-200607000-00006.
10. Pietrzak S, Napiontek M, Tomaszewski M. Catterall and Herring classifications in assessing Perthes disease – inter- and intra-observer study. Ortop Traumatol Rehabil. 2004;6(5):561–6. http://www.ncbi.nlm.nih.gov/pubmed/17618203.
11. de Billy B, Viel JF, Monnet E, Garnier E, Aubert D. Interobserver reliability in the interpretation of radiologic signs in Legg-Calvé-Perthes disease. J Pediatr Orthop B. 2002;11(1):10–4. https://doi.org/10.1097/01202412-200201000-00003.
12. Wiig O, Terjesen T, Svenningsen S. Inter-observer reliability of radiographic classifications and measurements in the assessment of Perthes' disease. Acta Orthop Scand. 2002;73(5):523–30. https://doi.org/10.1080/000164702321022794.
13. Ritterbusch JF, Shantharam SS, Gelinas C. Comparison of lateral pillar classification and Catterall classification of Legg-Calve-Perthes' disease. J Pediatr Orthop. 1993;13(2):200–2.
14. Simmons ED, Graham HK, Szalai JP. Interobserver variability in grading Perthes' disease. J Bone Joint Surg Br. 1990;72-B(2):202–4.
15. Christensen F, Søballe K, Ejsted R, Luxhøj T. The Catterall classification of Perthes' disease: an assessment of reliability. J Bone Joint Surg Br. 1986;68(4):614–5. http://www.ncbi.nlm.nih.gov/pubmed/3733840.
16. Hardcastle PH, Ross R, Hamalainen M, Mata A. Catterall grouping of Perthes' disease. An assessment of observer error and prognosis using the Catterall classification. J Bone Joint Surg Br. 1980;62-B(4):428–31. http://www.ncbi.nlm.nih.gov/pubmed/7430217.
17. Agus H, Kalenderer O, Eryanilmaz G, Ozcalabi IT. Intraobserver and interobserver reliability of Catterall, Herring, Salter-Thompson and Stulberg classification systems in Perthes disease. J Pediatr Orthop B. 2004;13(3):166–9. https://doi.org/10.1097/00009957-200405000-00005.
18. Herring JA, Neustadt JB, Williams JJ, Early JS, Browne RH. The lateral pillar classification of Legg-Calvé-Perthes disease. J Pediatr Orthop. 1992;12(2):143–50. https://doi.org/10.1097/01.bpb.0000279028.74397.02.
19. Kollitz KM, Gee AO. Classifications in brief: the herring lateral pillar classification for Legg-Calvé-Perthes disease. Clin Orthop Relat Res. 2013;471(7):2068–72. https://doi.org/10.1007/s11999-013-2992-9.
20. Rampal V, Clément JL, Solla F. Legg-Calvé-Perthes disease: classifications and prognostic factors. Clin Cases Miner Bone Metab. 2017;14(1):74–82. https://doi.org/10.11138/ccmbm/2017.14.1.074.
21. Salter RB, Thompson GH. Legg-Calvé-Perthes disease. The prognostic significance of the subchondral fracture and a two-group classification of the femoral head involvement. J Bone Joint Surg Am. 1984;66(4):479–89. http://www.ncbi.nlm.nih.gov/pubmed/6707027.
22. Mahadeva D, Chong M, Langton DJ, Turner AM. Reliability and reproducibility of classification systems for Legg-Calve-Perthes disease: a systematic review of the literature. Acta Orthop Belg. 2010;76(1):48–57.

23. Akgun R, Yazici M, Aksoy MC, Cil A, Alpaslan AM, Tumer Y. The accuracy and reliability of estimation of lateral pillar height in determining the herring grade in Legg-Calve-Perthes disease. J Pediatr Orthop. 2004;24(6):651–3.
24. Podeszwa DA, Stanitski CL, Stanitski DF, Woo R, Mendelow MJ. The effect of pediatric orthopaedic experience on interobserver and intraobserver reliability of the herring lateral pillar classification of Perthes disease. J Pediatr Orthop. 2000;20(5):562–5. https://doi.org/10.1097/01241398-200009000-00003.
25. Stulberg SD, Cooperman DR, Wallensten R. The natural history of Legg-Calvé-Perthes disease. J Bone Joint Surg Am. 1981;63(7):1095–108. http://www.ncbi.nlm.nih.gov/pubmed/7276045.
26. Neyt JG, Weinstein SL, Spratt KF, et al. Stulberg classification system for evaluation of Legg-Calvé-Perthes disease: intra-rater and inter-rater reliability. J Bone Joint Surg Am. 1999;81-A(9):1209–16.
27. Waldenstrom H. The first stages of the coxa plana. Acta Orthop Scand. 1934;5(1–4):1–34. https://doi.org/10.3109/17453673408991314.
28. Waldenström H. The definite form of the coxa plana. Acta Radiol. 1922;1:384–94.
29. Joseph B, Varghese G, Mulpuri K, Narasimha Rao KL, Nair NS. Natural evolution of Perthes disease: a study of 610 children under 12 years of age at disease onset. J Pediatr Orthop. 2003;23(5):590–600.
30. Joseph B, Nair NS, Narasimha Rao KL, Mulpuri K, Varghese G. Optimal timing for containment surgery for Perthes disease. J Pediatr Orthop. 2003;23(5):601–6. https://doi.org/10.1097/01241398-200309000-00006.
31. 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;14(1):10–5. https://doi.org/10.1097/01202412-200501000-00002.
32. Hyman JE, Trupia EP, Wright ML, et al. Interobserver and intraobserver reliability of the modified Waldenström classification system for staging of Legg-Calvé-Perthes disease. J Bone Joint Surg Am. 2015;97(8):643–50. https://doi.org/10.2106/JBJS.N.00887.

3

Treatment of Legg-Calvé-Perthes Disease by Maintaining Abduction to Allow Dynamic Hip Modeling Through Range-of-Motion Therapy

Perry Schoenecker and Jonathan Schoenecker

Rationale

Legg-Calvé-Perthes disease (LCPD) is an ailment of skeletal immaturity, which manifests as idiopathic necrosis of the proximal femoral epiphysis, physis, and, in severe cases, the metaphysis. The segregation of the epiphyseal vasculature from the metaphyseal vasculature of the proximal femur during development and the relatively marginal epiphyseal vasculature make the epiphysis uniquely susceptible to osteonecrosis [1]. The disease follows a protracted course, which is readily observed by plain radiography stages I–IV, and is characterized by sclerosis, fragmentation, reossification, and remodeling, respectively (Fig. 3.1a–d) [2, 3].

The relatively benign course of the disease in children younger than 6 years suggests that the age of the patient is one of the most important factors in disease severity. In older children, disease severity is initially correlated with the extent of proximal femoral involvement, but outcome is ultimately associated with the resultant geometry and condition of the articular cartilage of the hip [4, 5]. Although healing occurs in many patients with little or no permanent change in the morphology of the femoral head and acetabulum, long-term studies have shown that a substantial change develops in approximately 50% of patients, which leads to premature degenerative joint disease and disability [6]. Loss of containment and the development of a physeal bar contribute to poor hip geometry. Femoroacetabular impingement/subluxation and the extent of revascularization of the epiphysis are among the factors that influence the health of the articular cartilage. Late LCPD, especially in the setting of a physeal bar, characteristically includes coxa magna, a short femoral

P. Schoenecker (✉)
Shriners Hospitals for Children, St. Louis, MO, USA
e-mail: pschoenecker@shrinenet.org

J. Schoenecker
Vanderbilt Children's Orthopaedics, Nashville, TN, USA

D. S. Feldman, D. Paley (eds.), *Legg-Calvé-Perthes Disease*,
https://doi.org/10.1007/978-1-0716-0854-8_3

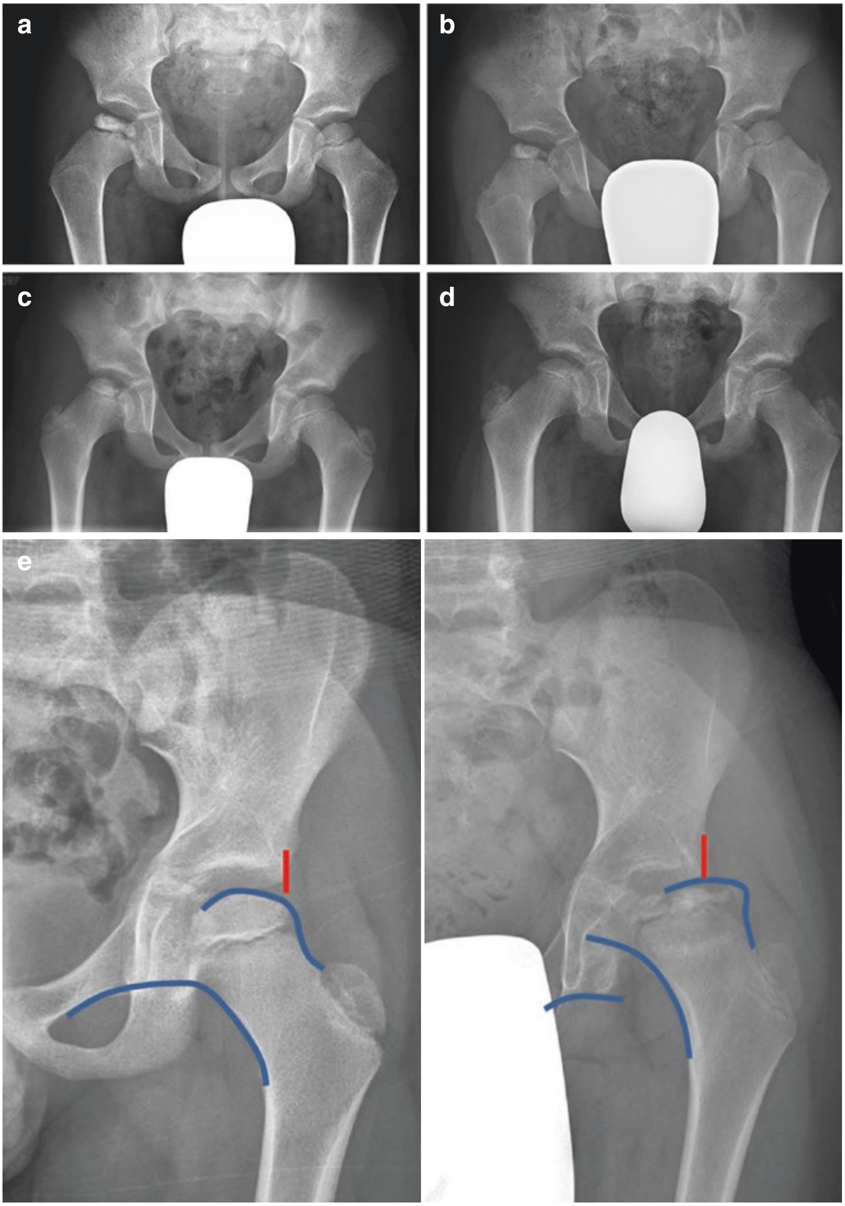


Fig. 3.1 (**a**–**d**) Anterior-posterior radiographs of a patient treated for acute LCPD of the left hip showing sclerotic stage I (**a**), fragmentation stage II (**b**), reossification stage III (**c**), and remodeling stage IV (**d**). (**e**) Comparative anterior-posterior views of the hip; note progression from stage I (**a**) (contained located with Shenton's line intact) to stage II (**b**) (uncontained, subluxated with Shenton's line broken). (**f**, **g**) (Fluoro spots from intraoperative arthrogram): The femoral head surface is relatively aspherical (**f** abducted anterior-posterior). There is no focal surface deformity. The femoral head moves from a laterally subluxated position to a more medially reduced (**g** neutral anterior-posterior) and contained location

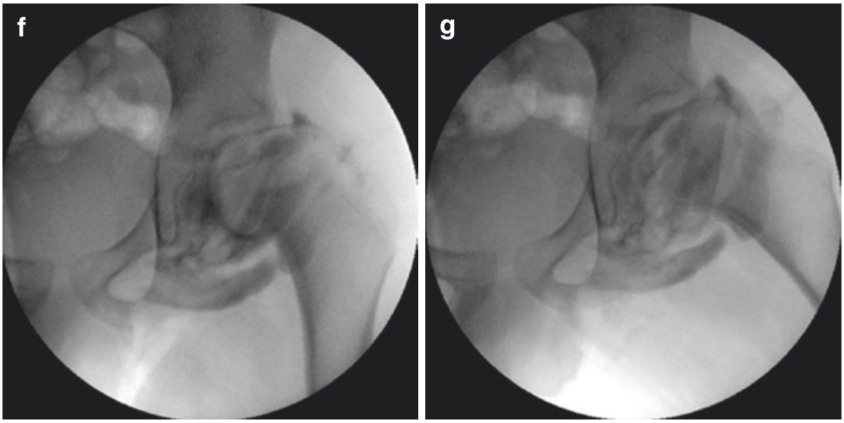


Fig. 3.1 (continued)

neck, and a high-riding greater trochanter with variable acetabular dysplasia [3, 7]. These deformities may cause impingement between the anterior head-neck junction and the anterior aspect of the acetabular rim and external impingement of the posterosuperior greater trochanter and ischium. In severe LCPD, secondary acetabular dysplasia may also include paradoxic joint instability transposed on femoroacetabular impingement. These geometric deformities instigate degradation of the chondrolabral junction and articular cartilage. Hip preservation surgeries are capable of addressing some, but not all, of these late deformities to achieve impingement-free and stable, normal hip range of motion in an effort to delay the development of osteoarthritis. Therefore, the optimal treatment is to minimize geometric deformity of the proximal femur and acetabulum and promote complete revascularization while avoiding the development of a physeal bar during acute LCPD (stages I and II, Fig. 3.1a, b) [3, 7].

Because no current therapies exist that alter the extent of revascularization of the epiphysis or the development of a physeal bar, containment is the only factor associated with outcome that is treatable. Initially, containment is attained by any combination of range-of-motion therapy, bracing, activity modification, and anti-inflammatory drugs. Surgical management is considered if containment cannot be maintained by nonsurgical methods. Proposed surgical containment methods range from adductor tenotomy and abduction casting to femoral and pelvic osteotomies. When considering methods of containment, the unique biology of LCPD healing must be considered. Importantly, and unique to the occurrence of osteonecrosis of the proximal femur in patients younger than 10, during fragmentation stage II, there are both bone resorption and metaplastic development of a neocartilage anlage, in both the proximal femur and often also the acetabulum (Fig. 3.1b) [8–10]. Due to its plasticity, this cartilage anlage ultimately determines the fate of the hip geometry. During revascularization/ossification, stage III (Fig. 3.1c), the cartilaginous anlage also serves to promote subsequent revascularization and ossification [3]. Considering the unique biology of LCPD healing, especially the plasticity of the hip during stage II, the principal theory of LCPD treatment is to utilize hip motion to maintain and promote femoroacetabular congruency.

The goal of initial intervention, ideally during stage I, is to aggressively achieve and maintain containment to permit maximal range of motion (ROM) of the hip. Given that these patients most commonly develop an adduction contracture (inability to fully abduct the hip), therapy is directed toward resolving this contracture. Subsequently, initiating in stage II and carrying on through stage III, the patient undergoes dynamic hip modeling through full ROM therapy. Thus, optimal treatment of LCPD requires two phases of treatment: containment and ROM hip modeling. The authors maintain that, regardless of the method of containment, continued dynamic modeling of hip cartilage through ROM therapy during stage II is the single most important variable in maximizing the geometric relationship of the femoral head and acetabulum.

Containment is readily achieved through stretching, bracing/casting, and adductor tenotomy. This comprehensive non-osteotomy approach involves serial evaluation and intervention depending on severity of contractures [11]. The objective of maintaining hip abduction motion is to prevent femoral head extrusion lateral to the rim of the acetabulum and the potential for the associated occurrence of problematic hinge abduction (Figs. 3.1g, 3.9c, d, and 3.10c, d) [13]. Although, like all therapies proposed in LCPD, outcomes of this approach are optimal if performed prior to cartilage deformity occurs during stage II. An additional advantage of this protocol is that it offers clinical improvement even if applied during stages II and III. This treatment strategy provides dynamic protection from potential anterior/lateral femoral head deformation, and enables sustained satisfactory hip joint motion. Continued femoral head and acetabular growth potentiates congruent remodeling of both the capital femoral epiphysis and acetabulum.

Indication

The natural history of LCPD is the inherent protracted course, and need for treatment of the involved hip to minimize deformity is discussed in detail with the family. Above all else, the importance of obtaining and maintaining satisfactory hip abduction motion is emphasized. Decision-making regarding the initiation of, altering, and ending treatment is based on the patient's passive adduction contracture (loss of hip abduction) of the involved hip. Hip abduction is assessed with one hand fixed on both anterior superior iliac spines and second passively abducting the involved lower extremity. The end point of functional hip passive abduction is determined when the examiner feels the pelvis being tilted into abduction. Comparison of the involved to the inherent abduction ROM of the uninvolved hip will help in appreciating the restriction of passive abduction of the involved hip (typically 10–15 ° abduction on the involved compared to 35–40 ° on the uninvolved hip). Considering the notable changes that occur in abduction during the protracted course of LCPD, parents and physical therapists play an important role in serial examination of the patient and therefore must also learn how to assess passive hip abduction.

For patients presenting with passive hip abduction >30 ° (adduction contracture of <10 °), home/physical therapy treatment regimen is initiated. The parents and physical therapist are instructed in the technique to maintain/improve passive hip abduction. If in the 4–6-week follow-up exam, >30 ° passive hip abduction is maintained (adduction contracture <10 °), similar care is continued with serial evaluation, monitoring both the clinical course and radiological progression of LCPD to stage II every 2–3 months. For patients whose passive hip abduction is maintained >30 °, continued follow-up is necessary until stage III (reossification).

More typically, patients with acute LCPD initially present with passive hip abduction of <30 (an adduction contracture of >10 °). Treatment to restore abduction is initiated. This approach includes exam under anesthesia, adductor tenotomy, and application of a Petrie cast (see below). Prior to performing the above procedure, the patient is measured for a hip abduction clamshell orthosis to be applied following the completion of abductor casting (see below).

OR Technique

Following the administration of anesthesia, there typically will be a marked improvement in passive hip abduction of the involved hip. In contrast, if there is extensive lateral head deformity with subluxation noted preoperatively on x-ray and/or MRI, there will be relatively less improvement in the passive abduction examination in the operating room [11]. Regardless, the degree of abduction to achieve with the Petrie cast is determined both with a clinical exam and c-arm with or without an arthrogram. First, abduction is assessed, confirming femoral head containment occurs within the acetabulum with a static and dynamic fluoroscopic examination (Fig. 3.2a–c). An arthrogram is beneficial to assure that the abduction position allows for complete acetabular coverage of the lateral cartilaginous anlage of the femoral head. This is performed through a medial, or anterior, hip joint injection, utilizing a 20-gauge, 2.5-inch spinal needle. Despite an often-noted lateral head deformity, with passive abduction, the femoral head will typically completely reduce into the acetabulum as visualized with the arthrogram (Figs. 3.1g, h, 3.9c, d, and 3.10c, d) [13].

If the patient's hip abduction is insufficient to easily obtain complete coverage of the femoral head by the acetabulum, and there is a contracture of the adductor longus, a tenotomy is performed. The adductor crease is sterilized and isolated with "square toweling." The tenotomy is performed through either a percutaneous or open technique, depending on surgeon preference. The authors prefer a percutaneous technique utilizing a less than 1 cm incision over the origin of the adductor longus. The point of a #15 blade is used to palpate and transect the adductor longus tendon. Specifically, with the blade aligned in the direction of the tendon, it is moved slightly laterally until the tip of the blade falls off the edge of the tendon. The blade is then rotated medially 90 ° directed toward the adductor longus, and the

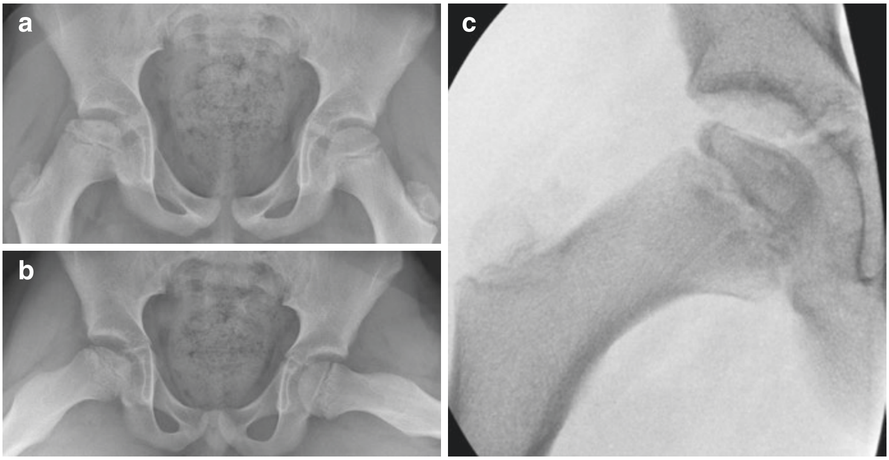


Fig. 3.2 (**a**–**c**) (Anterior-posterior **a** and frog-lateral **b** radiographs): presenting stage I acute LCPD at time of initial presentation with adduction contracture of >10°; (fluorospot **c** at time of initial EUA): satisfactory confirmation the femoral head is reduced in the acetabulum as is often possible with stage I Perthes without the need for arthrography

tendon is transected with the hip in an abducted position. If passive abduction motion is notably restricted and the arthrographic study does not show the femoral head reduction into the acetabulum, (hinge abduction) containment with an adductor tenotomy and Petrie casting is insufficient [11]. In these rare and difficult cases, the authors have used alternative surgical approaches to restore femoroacetabular congruency. These details are beyond the scope of this chapter.

Petrie type hip abduction cast is applied (Fig. 3.3a–d). This cast consists of bilateral cylinder casts fixed distally with two wooden dowels in enough bilateral hip abduction so as to contain the involved femoral head within the acetabulum [11]. The cylinder casts extend from 2–3 cm above the medial malleolus to the upper thigh. Stockinette and felt cuffs are applied to assure edge of cast comfort. The cast is constructed of fiberglass, applied with both knees flexed about 10 ° and supracondylar molding to minimize the tendency for the casts to slide distally (Fig. 3.3a). After the casts have set up, the extremities are abducted to the degree determined which contains the involved hip (Fig. 3.3b). The casts are temporarily secured in this position by taping a wooden dowel to the casts close to the ankles. The c-arm is used to confirm containment has been achieved of the involved hip (Fig. 3.3c). Having documented satisfactory hip positioning, two wooden dowels are secured, one posteriorly and one anteriorly with fiberglass to the cylinder casts just above the malleoli (Fig. 3.3d). Patients are instructed how to stand for transfer (with crutches) and typically discharged on the day of cast application. Most patients return to school in a few days after cast application using a wheelchair to facilitate moving about in school.

Fig. 3.3 (**a**–**d**) (Modified Petrie cast fabrication)

Abduction/Range-of-Motion Care

The cast is removed at 5 weeks, and the previously measured custom hip abduction "A-frame" orthosis is applied (Fig. 3.4a, b). That containment of the involved hip is achieved is assessed by an anterior-posterior pelvic x-ray (Fig. 3.4c). The A-frame is constructed so that if necessary abduction and adduction can be adjusted. Most importantly, the parents are re-instructed by the treating surgeon on adductor stretching exercise protocol (Fig. 3.5a). It will be relatively easier to obtain and maintain satisfactory hip abduction with less severe radiographic deformities than

Fig. 3.4 (**a**–**c**) The cast is removed at 5 weeks and a custom pre-fabricated A-frame orthosis (**a**) is applied. The orthosis consists of two polymer clamshell type cylinders attached firmly to an adduction/abduction adjustable stainless steel (1/8″ by 5/8″) A-type frame. Following the initial A-frame fitting (**b**), an anterior-posterior pelvic radiograph (**c**) is taken to assure there is adequate containment of the involved femoral head. If not, the brace frame is adjusted

hips with more severe lateral head deformities. Instructions are given on standing for transfers, weight bearing as desired with crutches when in the A-frame. Again, most patients prefer using a wheelchair at school. The family/patient is instructed on the initial brace protocol, 18 hours in and 6 hours out of orthosis. Patients that are >6 years of age are mobilized with crutches when out of the brace. When initially using crutches, patients are instructed on the importance of being non-weight-bearing on the involved hip. Patients <6 years of age are allowed out of the brace also for 6 of 24 hours, but because of the predictable lack of compliance with protected bearing on the involved hip are not given crutches. Patients return for follow-up examination in 4–5 weeks. The purpose of this exam is to assure that passive hip abduction is maintained (Fig. 3.5b, c).

Fig. 3.5 (**a**–**c**) Parents and care providers, respectively, perform and monitor hip abduction motion regimen/effectiveness. (**d**, **e**) A unique (custom-built) goniometer utilized by parents of an 8-year-old boy with acute Perthes disease of the left hip. The movable pegs allowed for effective observation of resolution of the adductor contracture of the left hip

For patients who maintain passive hip abduction >30 ° (adduction contracture of <10 °), home/physical therapy treatment regimen is re-emphasized. In addition, brace time may be decreased as long as passive hip abduction remains >30. When out of brace, patients are to remain non-weight-bearing on the involved extremity. These patients are instructed to return to clinic every 2–3 months to reassess the passive hip abduction and AP and frog radiographs of the involved hip to determine the Perthes radiological stage. Patients who do not maintain passive hip abduction >30 ° (adduction contracture of <10 °) are seen more frequently. The parents and physical therapist are reinstructed on the technique of adductor stretching so as to improve hip abduction. Brace fit and wearing compliance are reviewed. Again, the patient and family are instructed on non-weight-bearing on the involved side when walking with crutches. These patients are followed every 4–6 weeks to assure they regain passive hip abduction. As soon as they achieve optimal passive hip abduction, the treatment plan for patients with hip abduction >30 ° (adduction contracture of <10 °) is followed.

The Problematic Hip

Generally, maintenance of passive hip abduction is more difficult in hips with more extensively involved radiographic stages I and early II and in older and/or heavier patients. For these patients who on close follow-up observation do not regain the passive hip abduction >30 ° (adduction contracture of <10 °) from the first casting, a repeat EUA and reapplication of the Petrie cast should be performed. The cast application with anesthesia again assures obtaining satisfactory abduction. If an adductor tenotomy was not performed when applying the first cast, it typically would be done at the second abduction cast application. Occasionally hip abduction >30 ° cannot be achieved with only an adductor tenotomy. In these cases we have performed a more extensive medial adductor muscle and/or capsular release to achieve medial head reduction, eliminating problematic hinged abduction. The cast is removed again at 4–5 weeks and the patient is reassessed. Depending on the passive hip abduction at that visit, the appropriate protocols described above are followed. For the patient who has had more chronic synovitis and is slower to regain motion, the end goal of sustained improvement in abduction is still typically achieved, but perhaps 3–4 months later than patients with less problematic synovitis [*see case examples* Figs. 3.6, 3.7, 3.8, 3.9, and 3.10].

Duration of Treatment

Once the patient enters stage II, therapy changes to not only maintain hip abduction but perform complete ROM therapy in all planes of motion of the hip to maximally model both the femoral head and acetabulum. The family and/or physical therapy

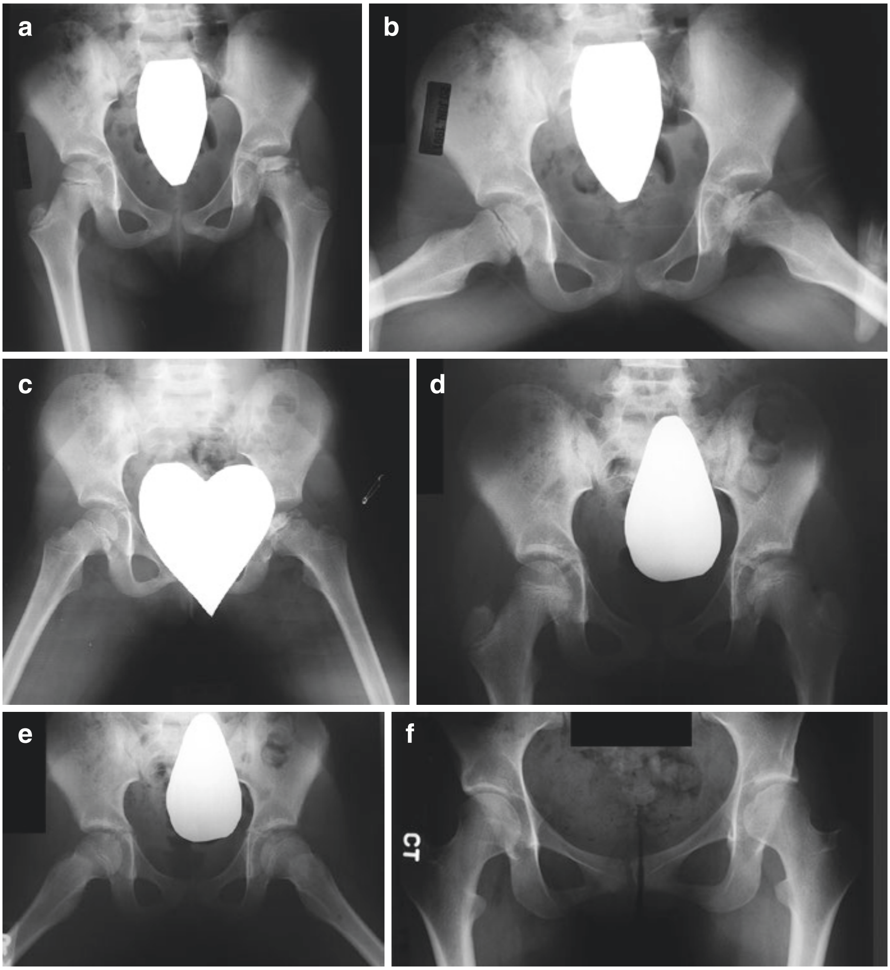


Fig. 3.6 (**a**–**c**) (Anterior-posterior **a** and frog-lateral **b** radiographs of a 7 + 6-year-old female with a 4-month history of limping and progressive left hip pain): There is acute LCPD involving the left femoral capital epiphysis, which is sclerotic (stage Ib) with noted collapse laterally (**a**) and anteriorly (**b**). The femoral head is subluxated, Shenton's line broken. The right hip is normal in appearance. There is a 25 ° adductor contracture of the left hip. Following adductor tenotomy and Petrie casting, anterior-posterior left hip radiograph (**c**) confirming containment by A-frame orthosis. Patient was treated with A-frame alternating with crutches and protected weight-bearing on involved hip and ROM protocol. (**d**, **e**) (Anterior-posterior **d** and frog-lateral **e** radiographs taken 2 years later): The femoral epiphysis has reossified and is relatively spherical. The femoral head and acetabulum appear congruent. (**f**) (Anterior-posterior standing pelvic radiograph at age 17): Both femoral heads appear spherically normal; there is no residual deformity of the left femoral neck. This is a spherically congruent Stulberg type I hip

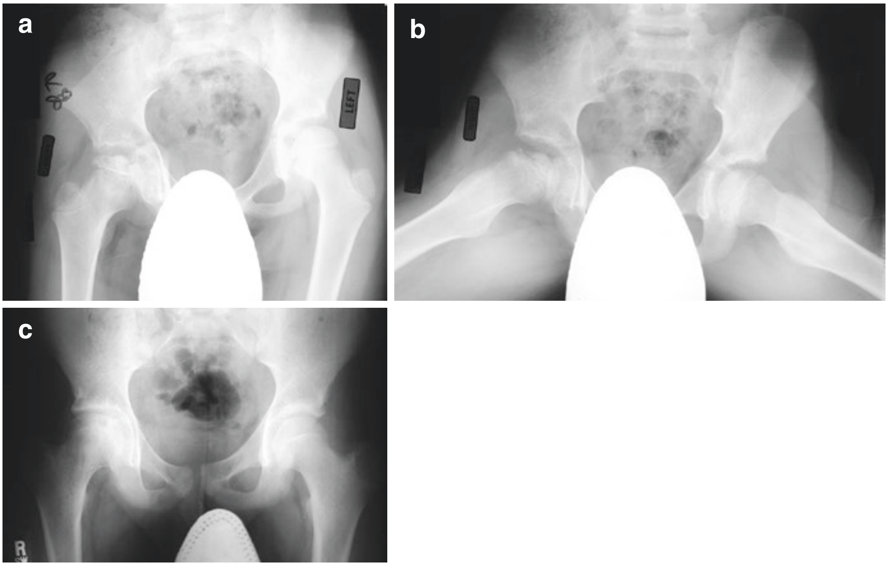


Fig. 3.7 (**a**, **b**) (Anterior-posterior **a** and frog-lateral **b** radiographs of a 9-year-old male with noted limping and right hip stiffness): There is acute Perthes disease involving the right capital femoral epiphysis in early fragmentation stage. There is a 30 ° right hip adductor contracture. The patient was treated with adductor tenotomy, abduction casting, and then A-frame orthosis alternating with crutches and protected weight-bearing on the involved hip and ROM protocol. (**c**) (Anterior-posterior standing pelvic radiograph at age 16): The right hip Perthes disease has fully reossified (healed stage), the femoral head is 0–1 mm out of round, and the neck slightly widened. This is a spherically congruent Stulberg type II hip

should perform this daily. If possible, recreational swimming is advocated. As long as these patients do not lose passive hip abduction, time out of brace may be gradually increased, again ambulating with crutches and protected weight-bearing when out of brace. Patients are followed every 2–3 months until they have radiographic signs of entering stage III (revascularization/ossification). When early stage III (healing) is present on radiographs, patients resume weight-bearing, and the brace may be discontinued, again as long as hip abduction motion is maintained. Patients are allowed to return to physical education at school as progression of stage III healing is noted on follow-up radiographs. In general, for patients who have not had difficulties in achieving passive abduction, a relatively fast course of treatment might be completed in 12–15 months [11]. Typically this would be an example of a 7-year-old patient with a moderate hip deformity (Fig. 3.6a–f). In contrast an 8–9-year-old patient presenting with a more severe deformity may need treatment for 15–18 months (Fig. 3.10a–i). Patients with greater deformity to begin with usually require longer treatment.

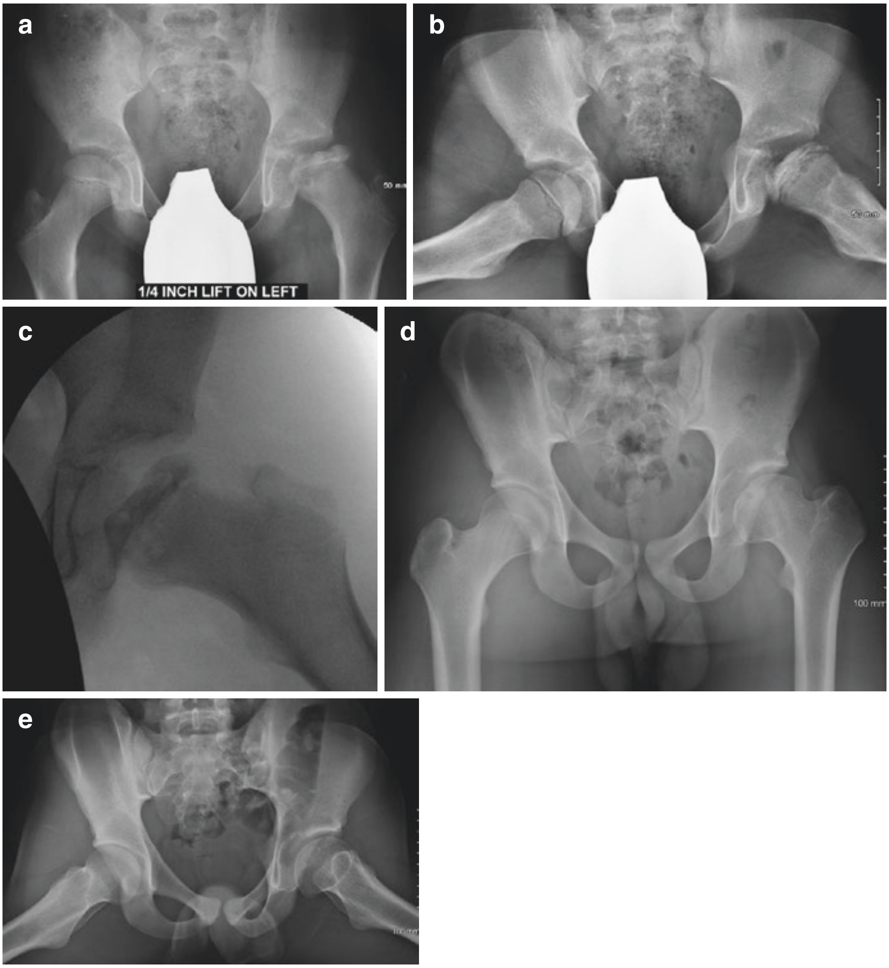


Fig. 3.8 (**a**, **b**) (Anterior-posterior pelvis **a** and frog-lateral **b** radiographs): There is acute early-stage II Perthes involving the left hip in an 8-year-old male patient; the right hip is normal. On exam there is a 30 ° left hip adduction contracture. (**c**) (Anterior-posterior fluoro image): The left femoral head is contained confirming marked improvement in passive left hip motion noted during the EAU. (**d**, **e**): Patient was treated with adductor tenotomy, abduction casting, and then A-frame bracing alternating with crutches and protective weight-bearing and range-of-motion protocol. Weight-bearing without crutches was resumed after 6 months of treatment. At follow-up at age 17, patient has excellent hip function as a sprinter on his high school track team. The Harris hip score is 100. Anterior-posterior (**d**) and frog-lateral (**e**) radiographs show a relatively aspherical but congruent hip joint. The femoral neck is relatively short. This is Stulberg type III hip

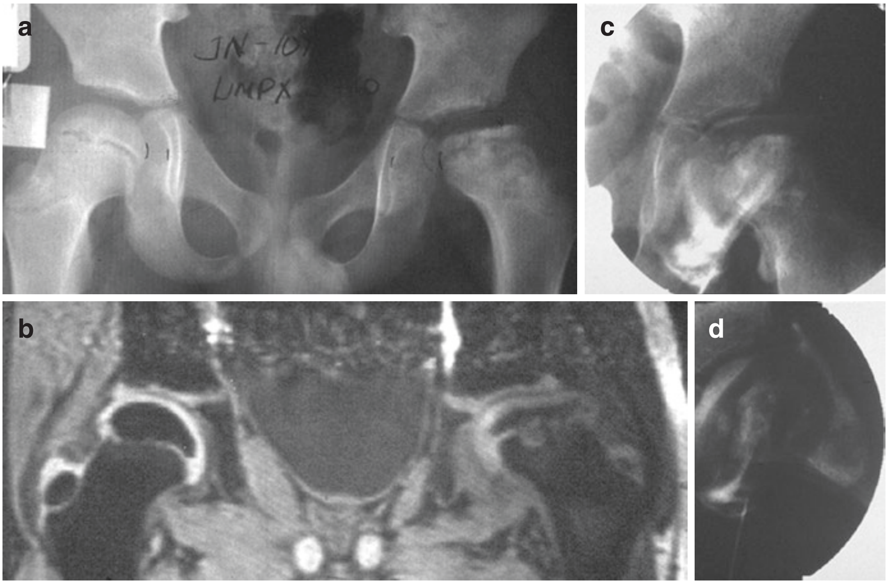


Fig. 3.9 (**a**, **b**) (Anterior-posterior pelvic radiographs **a** and MR images **b** of a 10-year-old male with 6-month history of limping and now left hip pain): The left femoral epiphysis in stage II is severely collapsed and subluxated; the right hip is normal. The MR image confirms the severe left femoral head deformity and hip joint subluxation. (**c**, **d**) (Arthrographic views taken during examination under anesthesia following open adductor tenotomy): The relatively aspherical femoral head is contained within the acetabulum. The lateral edge of cartilaginous femoral head lies lateral in (**c** anterior-posterior view in neutral abduction/adduction) and then medial (**d**, anterior-posterior view in abduction) to the lateral edge of the cartilaginous acetabulum. There is no femoral head distortion of the lateral labral chondral complex and/or rose thorn in the abducted position. Predictably, even for this 10-year-old patient, the femoral head can be physiologically contained with the adductor tenotomy abduction casting, bracing, protected weight-bearing, and range-of-motion protocol. (**e**) (Anterior-posterior radiograph of the left hip post application of Petrie cast): Note the femoral head is well contained within the acetabulum. (**f**, **g**) (Anterior-posterior pelvic radiograph 9 months after onset of treatment now with patient weight-bearing in the A-frame orthosis **g**): The hips are effectively contained; early healing of the left femoral head is present. (**h**–**i**) (Anterior-posterior left hip radiographs and corresponding MRU images 1½ years after onset of treatment): Notable pre- and posttreatment reconstitution of the femoral head is seen on both comparative anterior-posterior radiographs (**a**, **h**) and corresponding MR images (**b**, **i**). (**j**) (Anterior-posterior pelvic radiographic at age 15 + 9): Patient has excellent function of both hips. On exam there is a 1 cm leg length inequality, left leg being shorter than right. The range of right hip motion is 0–95 ° (extension-flexion) and abduction 35 °. The range of left hip motion is 0–90 ° (extension-flexion) and abduction 30 °. There is no reported anterior or lateral left impingement pain when assessing his motion. The left femoral epiphysis is fully healed and remodeled considerably, but aspherical as would be anticipated given the notable femoral head deformity at initial presentation. The left femoral head is relatively congruent with the remodeled acetabulum. This acetabulum is now desirably somewhat dysplastic which minimizes the potential for future femoral acetabular impingement. This is an aspherical but congruent hip, Stulberg III

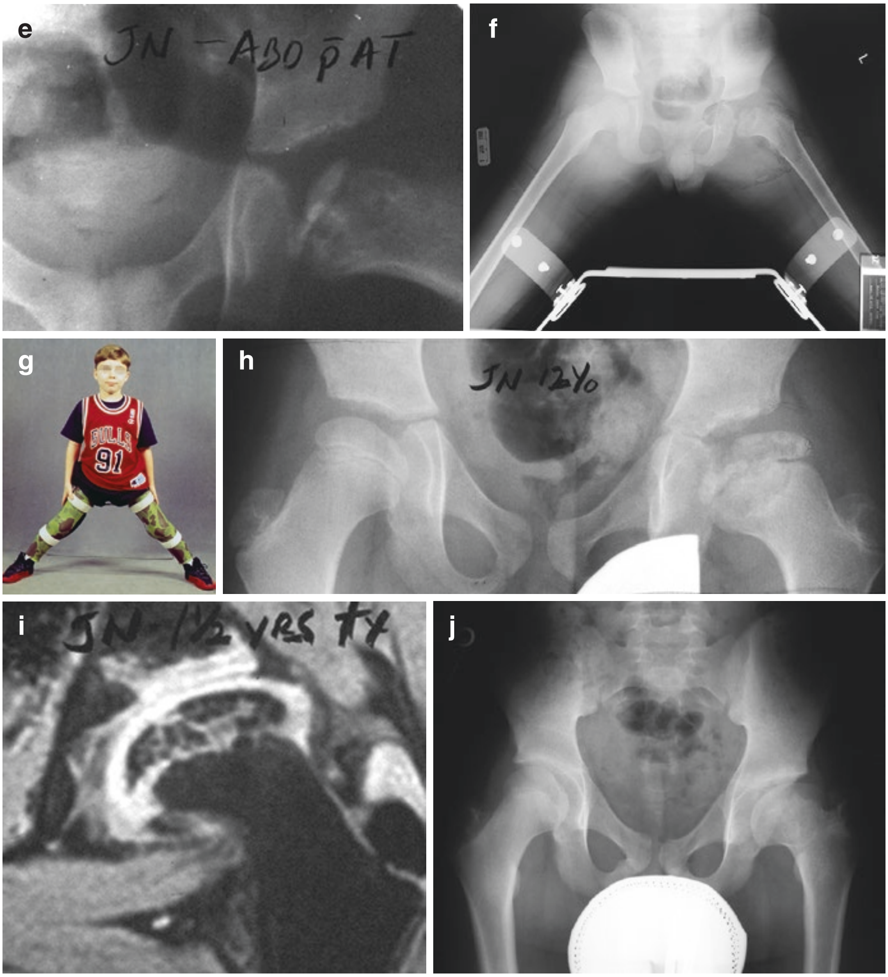


Fig. 3.9 (continued)

Response to and Outcome of Treating LCPD

The principal theory of optimal LCPD treatment is to utilize dynamic hip motion to maintain and promote femoroacetabular congruency. This requires containment of the femoral head in the acetabulum. In our protocol, containment is achieved with any combination of physical therapy, abduction casting/bracing, and tenotomy. Treatment is predicated by achieving passive hip abduction (resolution of hip

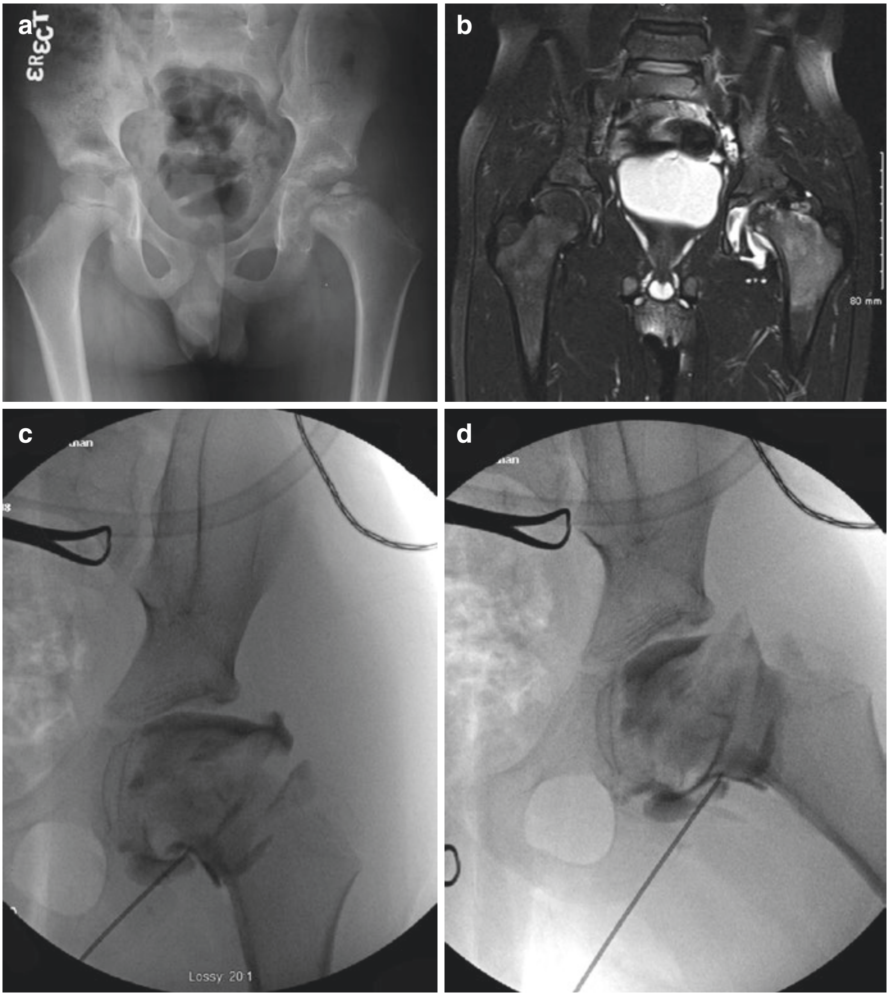


Fig. 3.10 (**a**–**e**) (Anterior-posterior radiograph **a** and corresponding MRI **b** of the pelvis of an 8-year-old male with previous observational treatment of acute Perthes disease of the left hip): There is stage II Perthes with notable extrusion deformity and subluxation of the femoral head. On fluoro spots from the intraoperative arthrogram, the femoral head surface is irregular with a lateral groove on the anterior-posterior distraction view (**c**) but containable on the abducted view (**d**). For this severe Perthes deformity, our dynamic containment treatment protocol would be the preferred approach. The adductor tenotomy, abduction casting, and A-frame splinting achieve reduction of the subluxated femoral head. Combined with ROM therapy, essential global femoral head and acetabular molding optimal occurs. Given the anticipated premature closure of the proximal femoral physis and relative trochanteric overgrowth, a lateral-based trochanteric apophysiodesis (**e**) as suggested by Stevens et al. was also performed at the time of the initial procedure [12]. (**f**–**i**): Favorable reconstitution of the femoral head is seen in follow-up anterior-posterior (**f**) and frog-lateral (**g**) radiographs taken 18 months after the initiation of containment treatment. At maturity the patient has excellent hip function and plays high school basketball. Harris hip score is 100. On anterior-posterior (**h**) and frog-leg lateral (**i**) views, the femoral head is aspherical with a relatively short femoral neck, a Stulberg type III outcome (note upsloping at the lateral acetabular rim on the anterior-posterior view; see text)

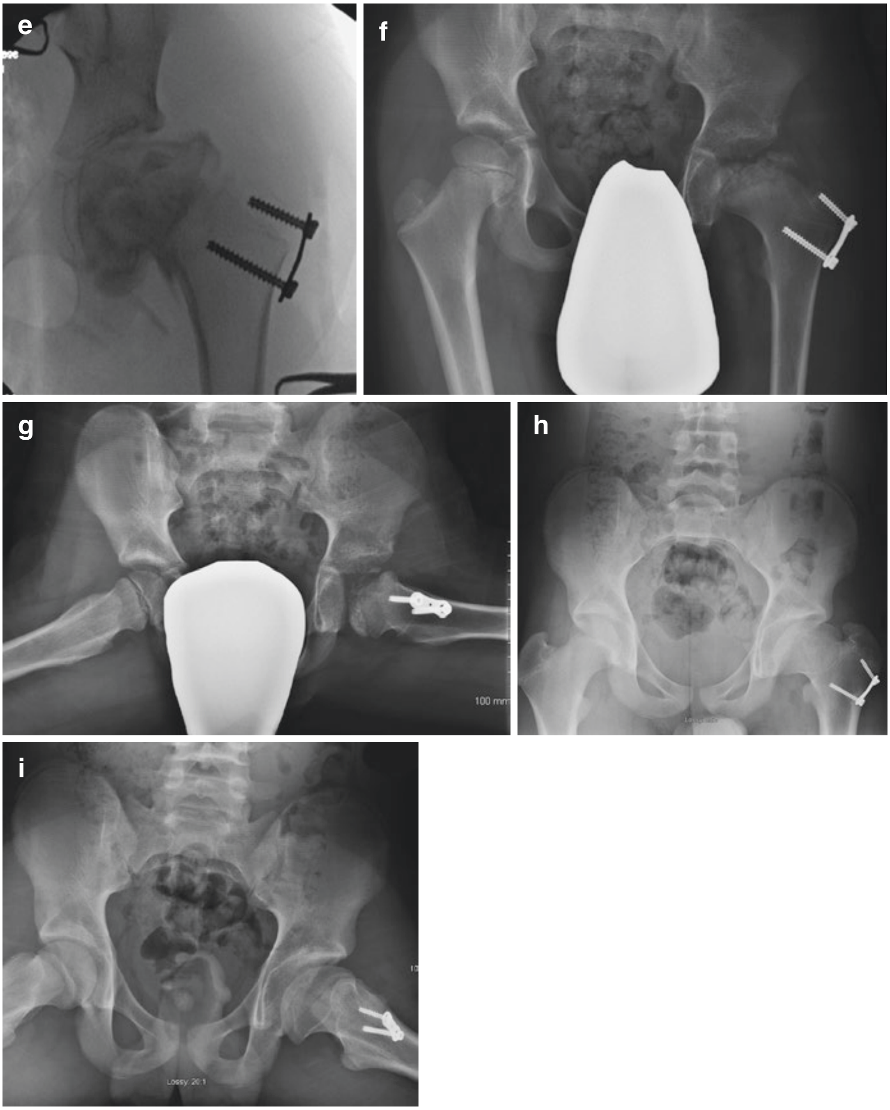


Fig. 3.10 (continued)

adductor contracture) which is serially assessed throughout the course of the disease allowing for alteration of treatment. The parents become effectively involved in carrying out the essential daily ROM exercise program. They observe improvement in passive abduction following the initial casting and bracing. Their direct involvement and observed success of ongoing treatment help assure compliance.

Our containment approach optimizes motion which potentiates relatively congruent femoral head/acetabular growth and remodeling. The outcomes at skeletal maturity have generally been very encouraging. Initially presenting less severe deformities, case examples (Figs. 3.6a–e and 3.7a–c) have near normal-appearing radiographic morphology at maturity, and there is little potential for late-occurring hip symptomology. In contrast case examples [8–10] all presented to us for treatment with severe, acute LCPD radiographic deformities. Severe deformities are more challenging. We have learned from our experience with all types of containment methods that predictably more severe deformities are best managed with our described comprehensive protocol of initially abduction casting and then bracing, ROM, and protected weight-bearing. As expected at maturity, there is variably both femoral neck shortening and high-riding trochanters. Femoral heads are aspherical, but more importantly the hip joints are relatively congruent. This is particularly so for Figs. 3.8d, e and 3.9j. A closer look at Fig. 3.10h does show an acetabular irregularity (a subtle lateral upslope). Seemingly, this occurred secondary to modeling of the acetabulum in response to the pre-existing cleft irregularity in the lateral femoral head (Fig. 3.10c, d). All three of these patients might in time experience the onset of problematic hip symptoms secondary to either femoral acetabular impingement or occasionally acetabular dysplasia that may in the future warrant consideration of performing joint preservation surgery [14]. For patients presenting with acute Perthes and severe deformities, we believe that femoral head/acetabular modeling effected by our treatment protocol which is focused on obtaining maximal ROM has optimized both current and long-term outcome for these patients.

Summary

Optimal treatment of LCPD requires two phases of treatment; containment and ROM hip modeling. In stage I our goal of intervention of acute LCPD is to aggressively achieve and maintain passive hip abduction so as to allow for optimal range-of-motion therapy to occur during the healing process. Beginning daily in stage II and continuing on through stage III, the patient's hip undergoes dynamic modeling through full ROM therapy. The authors maintain that treatment of the ubiquitous hip adduction contracture and continued dynamic modeling of hip cartilage through ROM therapy during stage II maximizes the geometric relationship of the femoral head and acetabulum resulting in favorable outcomes. Our abduction bracing/ROM treatment approach to acute Perthes potentiates achieving maximal functional hip motion at maturity for patients. In the process, development of femoral deformities is minimized, joint modeling and congruency maximized, and outcome optimized.

References

1. Trueta J. The normal vascular anatomy of the human femoral head during growth. J Bone Joint Surg Br. 1957;39-B(2):358–94.
2. Schoenecker JG, Podeszwaj DA. Developmental dysplasia of the hip. Orthop Knowl Update. 2011:1–15.
3. Leshikar HB, Schoenecker JG. Legg-Calve-Perthes disease. Orthop Knowl Update Pediatr. 2016;5:1–14.

4. Shah H, Siddesh ND, Joseph B. To what extent does remodeling of the proximal femur and the acetabulum occur between disease healing and skeletal maturity in Perthes disease? A radiological study. J Pediatr Orthop. 2008;28(7):711–6.
5. Froberg L, Christensen F, Pedersen NW, Overgaard S. The need for total hip arthroplasty in Perthes disease: a long-term study. Clin Orthop Relat Res. 2011;469(4):1134–40.
6. Larson AN, Sucato DJ, Herring JA, 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 Joint Surg Am. 2012;94(7):584–92.
7. Jaramillo D, Kasser JR, Villegas-Medina OL, Gaary E, Zurakowski D. Cartilaginous abnormalities and growth disturbances in Legg-Calvé-Perthes disease: evaluation with MR imaging. Radiology. 1995;197(3):767–73.
8. Holdsworth FW. Epiphyseal growth: speculations on the nature of Perthes's disease. Ann R Coll Surg Engl. 1966;39(1):1–16.
9. Catteral A, Pringle J, Byers PD, Fulford GE, Kemp HB, Dolman CL, et al. A review of the morphology of Perthes' disease. J Bone Joint Surg Br. 1982;64(3):269–75.
10. Ponseti IV, Maynard JA, Weinstein SL, Ippolito EG, Pous JG. Legg-Calve-Perthes disease. Histochemical and ultrastructural observation of the epiphyseal cartilage and physis. J Bone Joint Surg Am. 1983;65(6):797–807.
11. Rich MM, Schoenecker PL. Management of Legg Calve-Perthes disease using an A-frame orthosis and hip range of motion. A 25 year experience. J Pediatr Orthop. 2013;33(2):112–9.
12. Stevens PM, Anderson LA, Gilliland JM, Novais E. Guided growth of the trochanteric apophysis combined with soft tissue release for Legg-Calve-Perthes disease. Strategies Trauma Limb Reconstr. 2014;9:37–43.
13. Sankar W, Schoenecker P, Zaltz I, Miller P, Shore B. Determining hinge abduction in LCPD---Can we confidently make the diagnosis? J Pediatr Orthop. 2017;11:592.
14. Clohisy JC, Nepple JJ, Ross JR, Pashos G, Schoenecker PL. Does surgical hip dislocation and periacetabular osteotomy improve pain in patients with Perthes-like deformities and acetabular dysplasia? Clin Orthop Relat Res. 2015;473(4):13770–1377.

4

Prevention and Early Treatment of Legg-Calvé-Perthes Disease Utilizing Transphyseal Neck-Head Tunnelling (TNHT)

Nuno Craveiro Lopes

Introduction

The multifactorial origin of Legg-Calvé-Perthes disease (LCPD) is well documented [1–12]. The same is true for the existence of two or more ischemic episodes preceding the onset of the disease [2, 5, 12–20].

The first signs on the importance of multiple ischemic episodes were brought up in experimental research performed on dogs by Freeman [14] and on rabbits by Sanchis [18] and later by Bencano [21]. They were able to simulate the disease through repeated, total surgical section of the vascular network to the femoral head.

Vegter [22] using intra-articular injection of serum under pressure on the hip of a rabbit demonstrated the importance of the repeated fractional necrosis, provoked by the increase of intra-articular pressure on the onset of the disease, indicating that the probable cause of the deformity of the head is a reduction of its mechanical resistance due to superposing new ischemic episodes over a reconstruction process in progress.

Eyring [6], Soto-Hall [23], Kallio [24], Vegter [25] and Scoenecker [26] demonstrated that the in vivo positioning of lower limbs in prolonged and repeated medial rotation and extension is sufficient to increase the joint pressure on the hip and that it reached values superior to the systolic pressure when there was a simultaneous intra-articular effusion, even if small.

Mckibbin [11], Inoue [27] and Catterall [2] presented histological studies performed on femoral heads of patients with LCPD. These patients revealed images of necrosis superposed to zones of bone regeneration of a prior ischemic insult, confirming the theory of the repetitive ischemic episodes in the origin of LCPD.

After more than 30 years of experimental and clinical data analysis about the initial stage of LCPD [15, 28–32], the author verified that these episodes are

---

N. C. Lopes (✉)
Orthopedic Department, Portuguese Red Cross Hospital, Lisbon, Portugal

D. S. Feldman, D. Paley (eds.), *Legg-Calvé-Perthes Disease*,
https://doi.org/10.1007/978-1-0716-0854-8_4

symptomatic. Patients complain of transient pain and limping, and it is possible to detect those ischemic episodes with imaging techniques.

The same author has also found that those episodes represent an autonomous disease, which, if some other related factors are present, can evolve to LCPD. The author named this pre-LCPD situation ischemic disease of the growing hip (IDGH) [35–39].

The ultrasound has shown to be a valuable tool to evaluate the inflammatory state of a hip joint and is effective in demonstrating the existence of synovitis as well as measuring joint effusion and the thickness of the articular cartilage of the femoral head. Values greater than 6 mm of the anterior joint space and 3 mm of cartilage thickness are indicative of a joint with subacute or chronic synovitis [33, 52]. If this happens in the contralateral hip of a patient with LCPD, there is a risk that sooner or later the disease will develop there. As well, the magnetic resonance imaging is very specific and can demonstrate detailed marrow changes in the femoral head damaged by ischemia or bone necrosis [34].

The idea of improving the vascular conditions in the epiphyseal nucleus via the perforation of the femoral neck-head growth plate is not new. The effect of the surgical perforation of the growth plate has been studied experimentally by several authors in the past [8, 40–45].

Harris's research [46], using rabbits, confirmed histologically the penetration of cancellous bone with vascular structures through the perforations in the growth plate and a faster bone regeneration in the epiphyseal nucleus after bone necrosis provoked by surgical vascular section.

Siffert's research [45] showed histological preparation of femoral hips of rabbits. He observed the vascular and bone trabecular continuity between the metaphysis and the epiphysis after the perforation of the growth plate.

These researchers observed also that pinhole perforations did not have any negative repercussions on growth. No bone bridge appeared due to the closing of the borders of the growth cartilage, and perforations of small diameter also did not cause alterations of bone growth, despite the formation of a narrow bone bridge. However, the perforations of larger diameter caused a significant delay or a complete growth arrest through the formation of a rigid large bone bridge.

Within this context, the reinforcement of the arterial blood flow and of the venous drainage of the epiphyseal femoral head, independent from the retinacular network, could constitute a way of avoiding the repetition of ischemic episodes, thus preventing the onset of LCPD. If done early in the onset of the disease, it may promote a faster reconstructive process, with a better end result.

The perforation of the femoral neck-head growth plate was first performed by Ferguson [7], as a treatment for severe Legg-Calvé-Perthes disease in children. Those cases were done in an advanced fragmentation stage, in cases where the author had a poor response using traditional treatment methods. The technique of perforation of the neck-head growth plate had its advocates but was dropped when latter new techniques, such as femoral containment osteotomies, were developed by Soeur and De Racker [47] and other authors.

The growth plate drilling or tunneling is a different procedure than a core decompression, described by Ficat and Arlet [48] in the early treatment of avascular necrosis of the femoral head (AVN) in adults and adapted for older children with juvenile AVN by Price and Herrera-Soto [49]. Core decompression uses a large drill, generally with 8–10 mm of diameter to do a tunnel through the growth plate, to remove the necrotic bone from the femoral head, and bone graft is packed inside. When performed early, it can relieve pain, though epiphyseal height will not improve and complications, such as femur fracture, head blowout and growth arrest, may occur if done in young children.

Pathogenesis of LCPD: An Experimental Animal Model

To clarify the pathogenesis of LCPD, the author developed an animal model utilizing 27 White New Zealand rabbits, 7–8 weeks old [35, 36, 38]. This growth stage of the rabbit is equivalent to that of a 5–6-year-old child. This was performed to simulate the morphologic, radiological and histological aspects of LCPD. A non-invasive method was used with flexible splints on the legs of the rabbits in extension and medial rotation for 6 hours. This promoted an effusion and secondary collapse of the retinacular vessels [50]. The following day micro-trauma was produced on the right hip only, using a vibratory motor for a period of 30 seconds. This sequence was repeated twice a week.

The author observed that the prolonged and repeated positioning of the legs of the rabbit induced an intra-articular pressure level sufficient to produce an ischemic episode at the proximal femoral epiphysis. Most of the time, the first positioning had triggered a joint effusion. After this episode, the author observed a process of rapid revascularization of the preserved vascular canals with repositioning of myeloid and osteoid tissues by differentiation of endothelial cells in mesenchymal progenitor cells, without distortion of the epiphyseal structure or loss of its mechanical resistance. When a new ischemic episode was produced after the completion of the reparative process, all the sequence was repeated, again without loss of the mechanical resistance of the epiphysis (Fig. 4.1a). However, the repetition of an ischemic episode during the reconstruction of a previous episode leads to an alteration of the reparative response, with osteoclastic hyperactivity in the subchondral zone and formation of a distorted bone structure in the epiphysis by osteoblastic hyperactivity, which lead to a mechanically weak bone structure, similar to that described in LCPD as “woven bone”. In these conditions, the existence of trauma or micro-trauma on the right hip led to the appearance of a subchondral pathological fracture and collapse of the epiphysis, with necrosis and formation of a true bone sequestrum on that side only. This represents the initial stage of LCPD (Fig. 4.1b).

Our experimental research on the hips subject to repeated posture in medial rotation and extension showed us that the first signs of ischemia to the epiphyseal nucleus appeared after repeated positioning on both hips and signs of LCPD only on the hips subjected to micro-trauma. Those findings which are in accordance with the

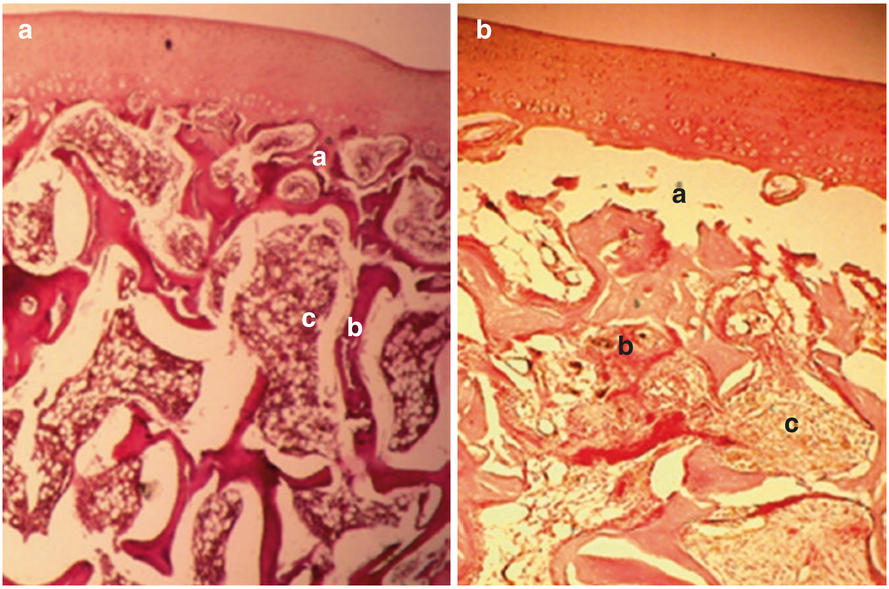


Fig. 4.1 (**a**) Experimental model 2 weeks – IDGH. Subchondral weakening (**a**), double-layer lamellae (**b**), normal myeloid tissue (**c**). (**b**) Experimental model 5 weeks – LCPD. Subchondral fracture (**a**), woven bone (**b**) and fibrous tissue proliferation (**c**)

other authors seem to confirm that the repeated ischemic events weaken the epiphyseal bone structure and that a trauma or micro-trauma is necessary to develop the subchondral fracture that begins LCPD [54].

The clinical data of the author confirms the repetitive ischemic episode theory, namely, that there is a period of fractional and sequential ischemic episodes that induce the epiphyseal fragility, before the onset of Perthes disease. Then if a trauma or micro-trauma occurs, subchondral fracture and epiphyseal collapse begin LCPD.

The conclusion of this experimental research suggests the existence of a pathologic entity, prior to LCPD, characterized by the existence of successive ischemic events at the level of the femoral proximal epiphysis that in certain circumstances can develop into LCPD. The author calls this entity ischemic disease of the growing hip (IDGH).

Effect of Transphyseal Neck-Head Tunnelling

Experimental Animal Model

The idea of supplementing the unstable epiphyseal blood flow through the growth plate is attractive but controversial.

To determine the effects of transphyseal neck-head tunnelling (TNHT) across the physeal growth plate, namely, the possible reinforcement of the epiphyseal blood flow and the repercussion on the growth of the proximal femur, the author has developed a second experimental study [38, 39] and later established a preventive protocol of LCPD based on the early detection of IDGH and its treatment by TNHT.

Forty-four White New Zealand rabbits, aged between 6 and 12 weeks, were used. Drills from 2 to 3.5 mm in diameter were used along with a specially made guide, to minimize trauma.

There were four study groups:

- Group I – 44 left hips. Not operated on, used as a control.
- Group II – 30 right hips. Drilling of <10% of the growth plate area. Single drilling with 2 and 2.5 mm drills, using as an entry point the trochanter crossing the neck-head growth plate through the epiphysis. The perforated area ranged in size from 6% to 10% of the total growth plate area, depending on the rabbit's age.
- Group III – 7 right hips. Drilling of >10% of the growth plate area. Single drilling with 3 and 3.5 mm drills, using the same technique. The perforated area ranged in size from 12% to 20% of the total growth plate area.
- Group IV – 7 right hips. Metaphyseal drilling. Single drilling with 2.5 mm drills on the metaphyseal area of the trochanter and neck of the femur, without reaching the growth plate.

The author was able to determine that the central perforation of an area less than 10% of the femoral neck-head growth plate area did not hinder growth. This conclusion is based on the direct and the radiological examinations and on the statistical evaluation of the numerous parameters measured. With the perforation we even noticed an increased sphericity of the femoral head.

The histological examination of these specimens showed normal growth plate thickness and organization, except immediately next to the perforated area.

In agreement with other authors [8, 22, 40], it was observed in the histological specimens that a small calibre perforation created a bone bridge whose tensile strength is inferior to the mechanical growth forces in the un-perforated portion of growth plate. This fact permits the progressive elongation of the thin bone bridge, with a dragging and thinning effect over the growth plate borders in the direction of the metaphysis. It was observed that the perforation of larger dimensions leads to a distortion of the proximal femur, such as a shorter neck with a prominent trochanter, a vertical tilting of the neck-head growth plate and flattening of the epiphyseal nucleus. These alterations were coupled with a decrease in the thickness of the growth plate and a marked structural disorganization. The author also noted that the resulting growth delay and deformity were bigger when the larger perforation was performed on younger rabbits (younger than 9 weeks of age), corresponding to a less than 6–7-year-old child [32, 51].

Besides the histological confirmation of the observations performed by other authors, it was demonstrated in this study that there was an accentuated and constant increase of small blood vessels in the epiphysis of the head of the femur after perforation of the growth plate, which was maintained until the end of the bone growth in

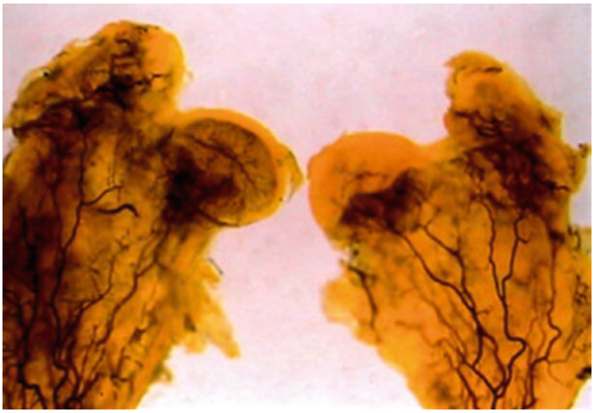


Fig. 4.2 Diaphanization of a specimen. Drilling at 4 weeks and sacrifice at 8 weeks. Right hip drilled, left hip control

rabbits (26 weeks). This result was not accomplished when only the metaphyseal zone was perforated.

This increase in epiphyseal blood circulation was due to the reinforcement of the vascular network formed through the perforation of the growth plate and not due to the hemodynamic changes on the retinacular vessels, which had not changed when compared with the control group.

The conclusion of this experimental research has shown that TNHT induces an increase of micro-vascularisation on the epiphysis due to the formation of a bone bridge thought the growth plate, and with vascular structures transversing from the metaphysis to the epiphysis and creating an anastomosis between the two networks (Fig. 4.2). It was shown that growth arrest or slowing due to the tunnelling procedure is a matter of dimension. When less than 10% of the area of the growth plate is drilled, the resistance of the bone bridge is much less than the growth force of the remaining growth plate, and growth continues normally. When the drilled area is bigger than 20%, the resistance of the bone bridge is higher than the growth force of the plate and there is an arrest. In between those two numbers, some slowing of growth occurs.

Surgical Technique of TNHT

The surgical technique for TNHT has been described in previous papers [28, 31, 39]. It is indicated for the treatment of IDGH, preventing the progression to LCPD, and for the treatment of LCPD at the stage of necrosis or early fragmentation. Using a 4–5 mm cannulated drill or trephine, a tunnel is drilled aimed to the centre of the ischemic or necrotic bone. The entry point is located at the lateral proximal metaphysis of the femur (Fig. 4.3), passing through the middle of the growth cartilage and stopping at the subchondral epiphysis. The patient is placed on a radiolucent table with the lower limbs draped free, using a C-arm always kept perpendicular to the operating table. The affected limb is positioned in medial rotation in order to place the neck of the femur parallel to the ground; a Kirschner wire is used as an aiming

Fig. 4.3 Procedure for TNHT with a 5 mm trephine

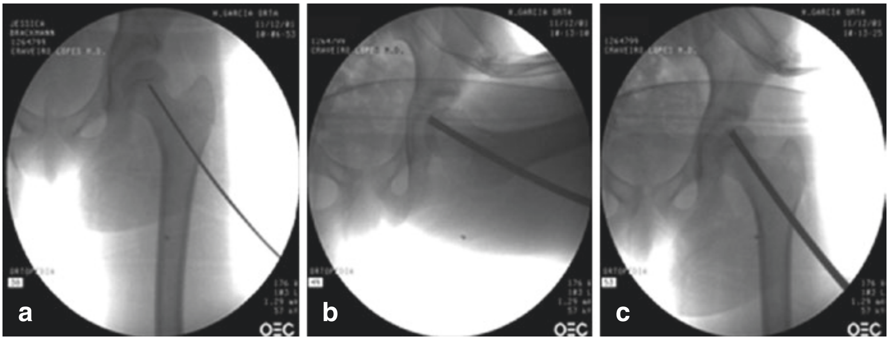


Fig. 4.4 Sequence of the procedure. (**a**) Wire guide positioning. (**b**) Introduction of the trephine over the wire guide reaching the subchondral bone. (**c**) Image control of the position of the trephine with flexion-abduction of the lower limb

device placing it over the skin on the anterior aspect of the hip to determine the proper level of the drilling procedure, as well as the proper entry point of the guide wire at the lateral side of proximal metaphysis of the femur. A stab wound is created for the introduction of a 2 mm guide wire; fluoroscopic control is recommended after about 2 cm of the guide wire has been inserted, using the C-arm on the anterior-posterior plane with the limb in neutral position, and on the lateral view by flexing and abducting the hip and flexing the knee at 90° (Fig. 4.4a). On the a-p view, the guide wire should aim to the transition of the middle and lateral 1/3 of the epiphysis and on the lateral view to the transition of the middle and anterior 1/3 of the epiphysis. After confirming proper positioning, the guide wire is further inserted and stopped just below the growth plate.

Then the 4–5 mm cannulated drill is introduced with a hand drill or with a low-speed pneumatic drill all the way to the subchondral bone of the epiphysis (Fig. 4.4b, c). If desired, a bone cylinder of the epiphyses can be obtained for research purposes, by replacing the cannulated drill with a trephine of the same diameter, right before starting drilling through the growth cartilage.

Fig. 4.5 Abduction-flexion orthosis used after TNHT for Perthes disease

Patients can walk 1 or 2 days after surgery, freely if done as a preventive procedure and using an abduction orthosis (Fig. 4.5) when done for the early treatment of Perthes disease.

Clinical Use of TNHT in Children with IDGH

As stated before, two or more ischemic episodes are necessary to induce Perthes disease. After more than 30 years of experimental and clinical data analysis about the initial stage of LCPD [15, 28–32], the author verified that these episodes are symptomatic. Patients complain of pain and transient limping, and it is possible to detect those ischemic episodes with imaging techniques. The same author has also found that those episodes represent an autonomous disease, which can evolve to LCPD.

It has been shown experimentally [39] and clinically [55] and confirmed by the author [28, 31] that the drilling of a tunnel representing less than 10% of the total area of the growth cartilage, as in TNHT, does not impair nor slow the growing of the growth plate. The performed tunnelling procedure represents less than 3% of the area of the growth plate in a child 5 or more years old. It has a wide margin of safety.

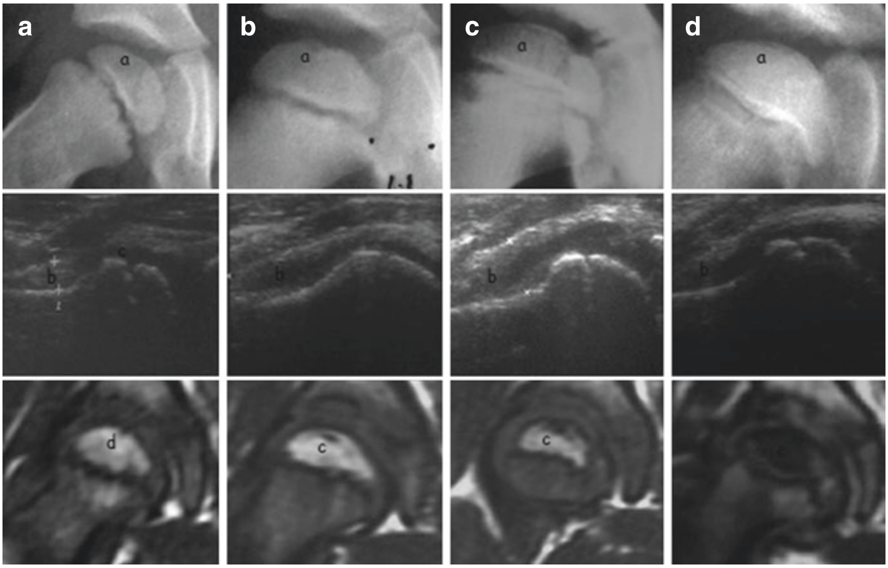


Fig. 4.6 (**a**) IDGH stage I. Irregular contour (**a**), synovitis (**b**), thickening of cartilage (**c**). No actual ischemia (**d**). (**b**) IDGH stage II. Irregular contour (**a**), synovitis and effusion (**b**), subchondral ischemia (**c**). (**c**) IDGH stage III. Porosis by bone weakening (**a**), synovitis and effusion (**b**), subchondral ischemia (**c**). (**d**) LCPD stage I. Dense collapsed bone with subchondral fracture (**a**), synovitis and effusion (**b**), extensive necrosis (**c**)

A TNHT procedure is applied if probability of evolution to LCPD is detected in a child 5 years old or more presenting an ischemic episode on the MRI (Fig. 4.6b, c) or a Perthes lesion on the necrotic or early fragmentation stage (Fig. 4.6d).

As a guide to the probability of evolution to LCPD, the authors had developed a classification of IDGH considering three stages based on the images of the x-rays, ultrasound and magnetic resonance [56].

Stage I (Fig. 4.6a) represents a hip where one can identify sequela of an old ischemic episode; it has a low probability of evolution to LCPD. *Stage II* (Fig. 4.6b) shows sequela of an old episode with a new ischemic episode in evolution; it is considered to have a moderate probability of evolution. Finally, *stage III* (Fig. 4.6c) presents signs of a recent episode with a new one in evolution. This situation is considered to have almost 100% probability to evolve to LCPD.

To identify the ischemic episodes and to detect the cases of IDGH at risk for evolution to LCPD, the authors have developed a screening protocol [56]. All paediatricians and general doctors of the area of influence of the hospital have been informed of the protocol and advised to send all children who present with hip, thigh or knee pain and limping for more than 1 day of evolution to the emergency unit.

From January 1993 to December 1995, before the implementation of the prevention protocol, the authors have observed 123 children, with ages between 3 and 12 years, presenting a painful syndrome of the hip. The authors did a second prospective comparative study between January 1996 and December 1998, after the implementation of the preventing protocol, including 110 children of the same age. The study protocol included a standard clinical, radiological and laboratorial examination to detect other diagnoses. Then an ultrasound screening was used to determine the existence of criteria of probability of IDGH. A second screening was applied using an MRI on those cases where US revealed signs of probability of IDGH, to detect an ischemic episode or the necrotic pattern of LCPD. In the case of a simple ischemic event, it is essential that the MRI examination be done in a temporal window of up to 4 weeks after the onset of the pain and limping episode, because the reparative process of the arterial and venous circulation within the vascular channels of the epiphysis is rapid and the MR image of subchondral ischemia disappears in 4 or 5 weeks.

The image data and histological examinations from 19 patients who presented signs and symptoms compatible with the existence of IDGH with risk of development of LCPD and 17 patients with LCPD in the initial stage were analysed. These patients were treated between 1995 and 2006 with TNHD and had samples for histological examination [56].

The analysis of the image data and histological examinations has shown that patients in whom IDGH was detected presented histological signals of a recent ischemic event on the osteoid and myeloid or recent ischemia over a remodelled anterior ischemic event (Fig. 4.7a). In all the cases where the bone samples showed profuse woven bone and total substitution of the myeloid by fibrous tissue, progressed into LCPD (Fig. 4.7b). Such biopsies were consistent with failure of immediate revascularization, as the vascular canals were collapsed.

On the first period, from January 1993 to December 1995, in 123 children, 32 cases (26%) had a diagnosis, including SCFE (3), septic arthritis (3), rheumatoid arthritis (3), trauma (2), LCPD (18) and other causes (3). On the remaining 91 cases, a first US screen has determined the existence of criteria of probability of IDGH in 43 cases. The other 48 cases were diagnosed as transient synovitis or unknown diagnosis. The second screening made with an MRI on those 43 cases detected an

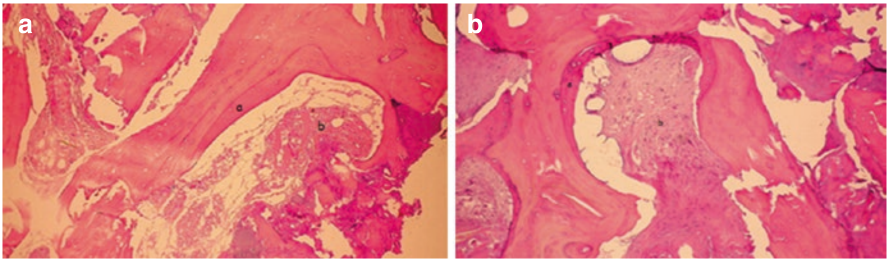


Fig. 4.7 (**a**) IDGH stage III. Ischemic event over a reparative process of a recent necrosis (**a**). Myeloid tissue begins to be substituted by fibrous tissue (**b**). (**b**) LCPD stage I. Woven bone (**a**), substitution of myeloid tissue by fibrous tissue (**b**)

ischemic episode in evolution in 6 of those patients (5% of all cases) who were diagnosed as IDGH stage II in 4 cases and stage III in 2 cases. So, on the first study period from 1993 to 1995 in 123 children, it detected 18 cases of LCPD and 6 cases of IDGH on stage II and III. On the second period, from January 1996 to December 1998, including 110 children, the author has diagnosed only five cases of LCPD and seven cases of ischemic disease stages II and III.

Epidemiological data from the area of influence of our unit, comparing the incidence of LCPD before and after the implementation of the IDGH detection protocol and LCPD prevention with TNHD, have shown a decrease of LCPD cases from 8.5/100.000 to 1.8/100.000 per year, on the population less than 16 years of age [56].

The current research indicates that the neck-head tunnelling could be essential in the prevention of Legg-Calvé-Perthes disease (LCPD) if used in the pre-radiological ischemic stage of the disease, characterized by transient hip pain and limping that can be detected through a subchondral cold spot on the MRI bone scan [5, 13, 17, 19, 20].

With regard to the contralateral hip, there are many studies demonstrating that it is not a normal hip. We described a significant number of irregularity, flattening or dimpling of the epiphyseal surface and smaller size of the epiphyses in the opposite hip in patients with unilateral disease [53, 67–69].

Although the bilateral disease rate was 17% in a review of all our patients and other statistics [54–56, 58], the author found in a review from 1992 to 2005 with a minimum follow-up of 7 years that in patients needing surgery for Perthes disease in one hip, the opposite hip was affected sooner or later in 32% of them [56].

It is also known that children with sequential onset of bilateral disease had a worse prognosis in the second affected hip [65, 66]. The increased risk of developing Perthes disease in the contralateral hip in those with unilateral disease is an important factor of concern for parents and caregivers because it means an extension of the need for orthopaedic care for many more years, a less favourable outcome in the second hip and bilateral disease with worse quality of life 6 during adult and young adult life.

After 30 years of experience with the TNHT procedure in patients with Perthes disease and later for prevention in stage II and III symptomatic ischemic disease, the author initiated its use for prevention of the opposite hip in unilateral disease. The criteria include the existence of signs of synovitis and thickening of the articular cartilage on the ultrasound, which correspond to IDGH stage I, in a child of 5 years of age or older who is proposed for surgical treatment to a hip with evolving Perthes disease (Fig. 4.8a–e).

The author has applied TNHD in 96 patients with the diagnosis of IDGH stages II and III; 55 of them have already reached the end of growth. For prevention of the contralateral hip in IDGH stage I, the procedure was applied in 28 cases. In both groups, none of the hips progressed to Perthes disease or showed growth disturbance of the proximal femur. No other complications as fractures, infections, etc. were reported on those cases (Fig. 4.9).

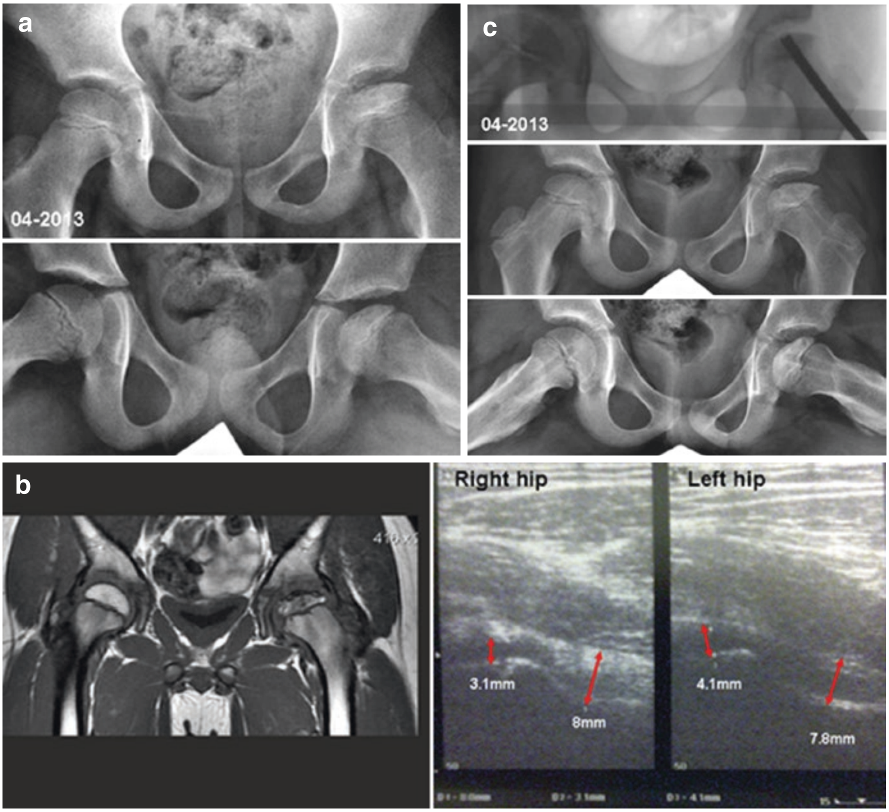


Fig. 4.8 (**a**) An 8-year-old boy, with a stage Elizabethtown 1b, Herring B lesion in the left hip. (**b**) The MRI shows 80% of involvement of the left hip and no actual ischemic event at the opposite hip, but the ultrasound showed synovial and articular cartilage thickening, signs that it was a pathologic hip too. (**c**) A bilateral drilling procedure was done to treat the Perthes disease at the left side and to prevent the evolution of the ischemic disease in the right side. At top, the intra-operative x-ray intensifier image of the drilling, and at the bottom, the immediate post-operative x-ray images. (**d**) X-ray images at 6 months after procedure with reossification of the subchondral zone and periphery of the lesion, with a mechanically resistant lateral column. The centre of the lesion is yet in reconstruction. In this stage, it is safe to discard the brace and begin normal life without impact activities. (**e**) Patient with 1.5 and 2.5 years of follow-up, at full remodelling with a Stulberg I result of the Perthes hip and normal growth of the right hip

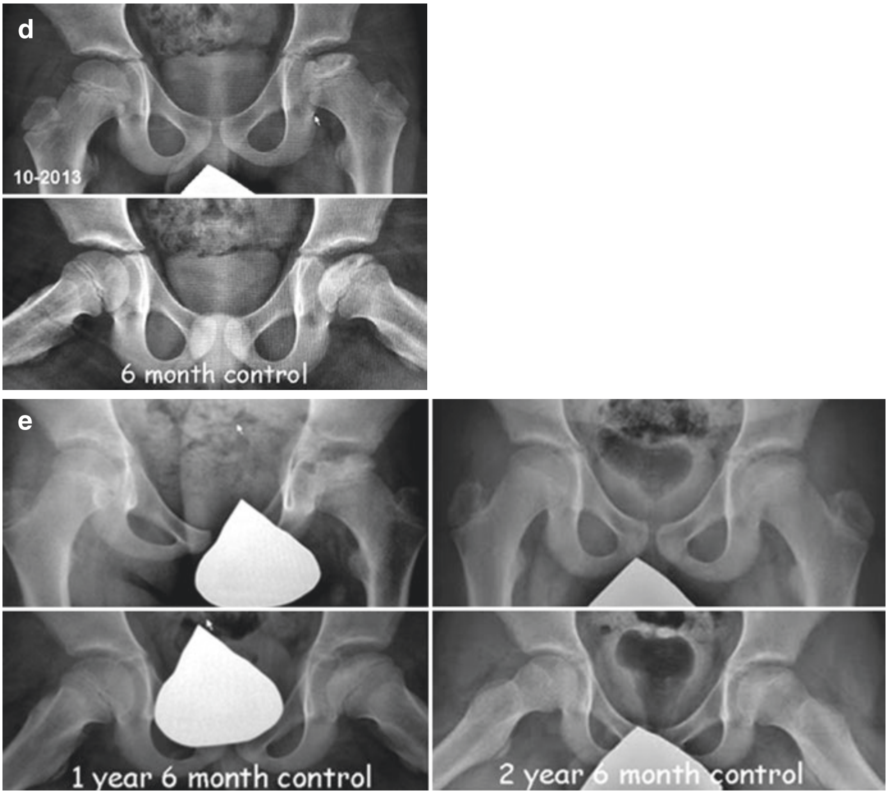


Fig. 4.8 (continued)

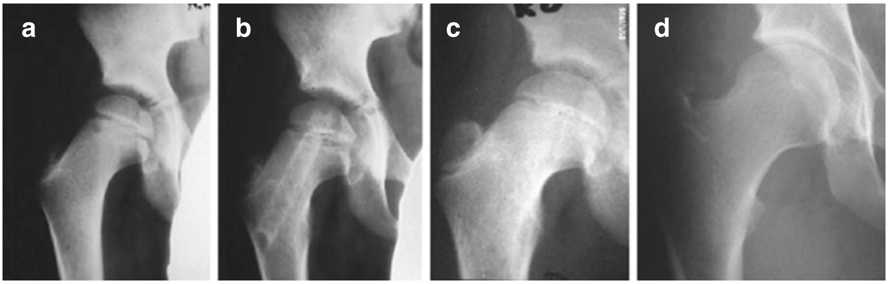


Fig. 4.9 Evolution of a case of IDGH after TNHD. (**a**) 10/1989 – 6 years old, IDGH stage III. (**b**) 11/1989 – 6 years old, 1 month after TNHD. (**c**) 9/1993 – 10 years old, 4 years after TNHD. (**d**) 04/2002 – 19 years old, 13 years after TNHD

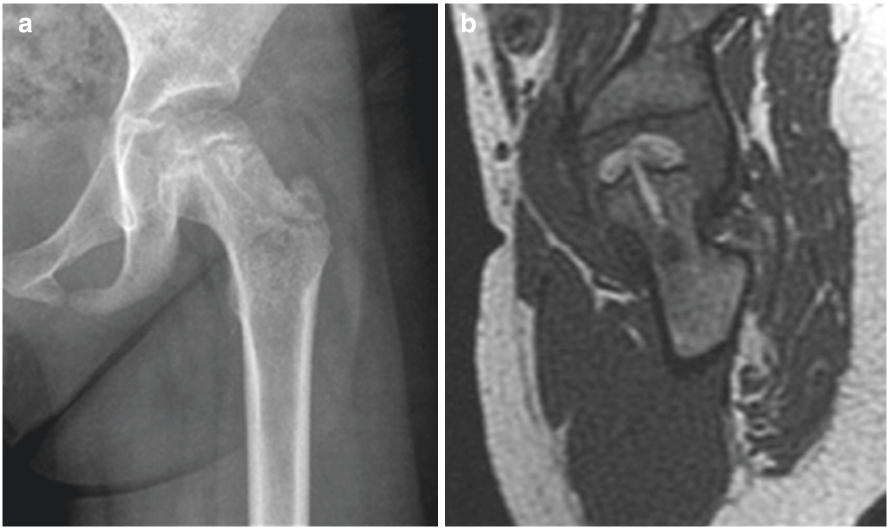


Fig. 4.10 (**a**) IDGH stage III. 7 months after TNHT. (**b**) Gadolinium-enhanced MRI. Vascular anastomosis metaphysis-epiphysis

Control of patency of the anastomosis using an MRI in T1 sequence with gadolinium confirmed that the anastomosis induced by the tunnelling procedure is a long-lasting one (Fig. 4.10a, b).

In a control group of 20 patients diagnosed as having ischemic disease, not subjected to the tunnelling procedure, the author found that in 15 cases of stage II, 3 patients developed Perthes disease, a rate of 20%, and in 5 cases of stage III, all developed Perthes disease (Fig. 4.11a–d).

Clinical Use of TNHT in Children with Early LCPD

Tunnelling can also be useful in the early treatment of the LCPD, in children older than 5 years. The author's treatment protocol includes a transphyseal neck-head tunnelling of the femoral head, done the earliest possible in initial necrosis or initial fragmentation stages.

After TNHT, a hip flexion-abduction orthosis is used to protect the hip joint and lower the activity of the child.

The author's experience with TNHT on LCPD from 1971 to 2010 includes 138 cases. Of these cases, only 26 cases (17%) required contention surgery for collapsed hinge hips. At the beginning of our series, 13 patients underwent a femoral osteotomy. Later on, pelvic osteotomies were done in 3 cases, and on the latest period, ranging from 2000 to 2010, 10 cases of arthrodiastasis with a modified Ilizarov frame were performed [28, 30, 31, 56, 57, 59].

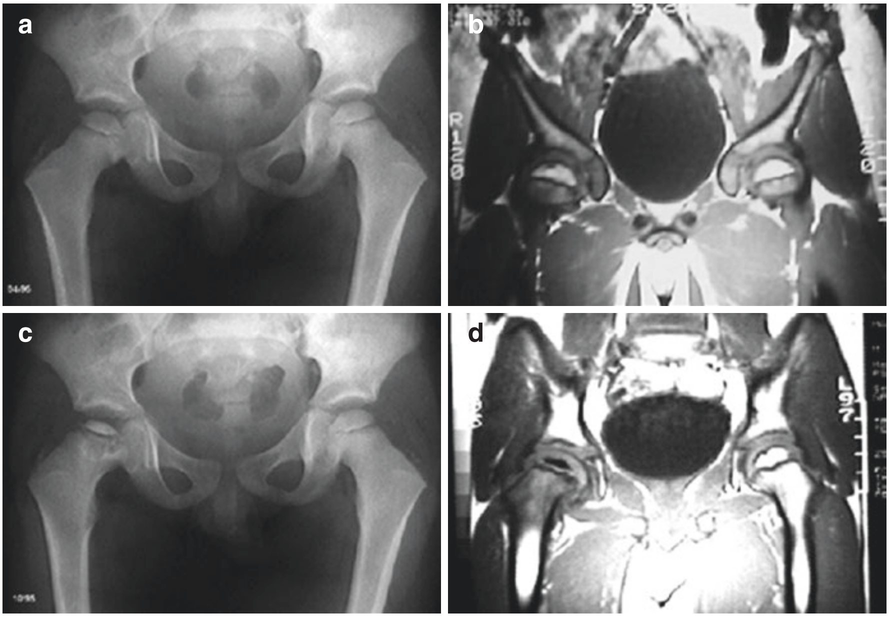


Fig. 4.11 (**a**) 04/1995 – IDGH stage III at right hip x-ray showing epiphyseal porosis and a wider medial space. (**b**) 04/1995 – MR image in T1W with low intensity subchondral ischemia signal. (**c**) 10/1995 – Lost from follow-up. 6 months later – LCPD stage I. (**d**) 10/1995 – LCPD stage I. Extensive necrosis

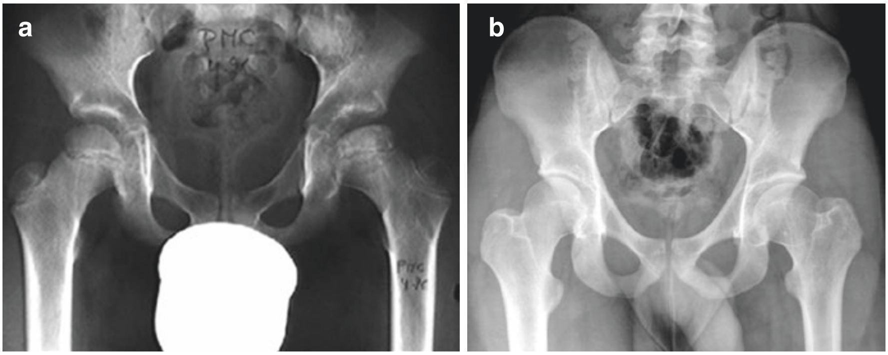


Fig. 4.12 (**a**) An 8-year-old boy, with a left Perthes disease, Herring B/C, treated with an early drilling procedure done in the necrotic stage. (**b**) At 10 years of follow-up, when he was 18 years old, he had a very good result with a Stulberg II hip

The author noted that the tunnelling procedure induces an acceleration of the natural history of the disease (Fig. 4.12a, b), shortening the time until the stage of reconstruction in 1/3 or half the time usually seen and increasing the arterial blood supply and venous drainage of the epiphysis, improving the conditions for a faster reconstruction with better end results and less patients needing invasive surgeries [31, 39]. This happens because it permits penetration of blood vessels to the centre of the necrotic zone and reabsorption (fragmentation) of the sequestrum by the cutting cones from inside to outside at the same time that the natural fragmentation process occurs from outside to inside [7]. When used with arthrodiastasis, TNHD allows a shorter and more effective external fixator usage, allowing on average 3–4 months until reconstruction is on the way and the fixator can be removed, unlike the average of 5–6 months established by other authors [60–64]. After the fragmentation stage is completed, TNHT is no longer useful.

Conclusions

The clinical study confirms the existence of a period of fractional and sequential ischemic episodes before the onset of LCPD. These ischemic events lead to weakening of the epiphyseal bone structure with woven bone formation. If a trauma or micro-trauma happens at that timing, then a pathologic subchondral fracture triggers the beginning of Legg-Calvé-Perthes disease.

The authors conclude that it is possible to identify the fractional and transient ischemic events that precede the beginning of Legg-Calvé-Perthes disease and to define a group of cases at risk of evolution to LCPD. On those cases, TNHT induces anastomosis of small blood vessels between metaphysis and epiphysis, thus supplementing the epiphyseal unstable supply, preventing new ischemic episodes and aborting the evolution to Perthes disease.

When the tunnelling procedure was used in cases with Perthes disease in evolution, the author found that it shortens the course of the disease to 1/3 or ½ and provides better vascularity to the femoral head with a shorter period of femoral head fragility and better end results, with only 17% of the patients needing invasive surgery.

Bibliography

1. Catterall A, et al. Legg-Calvé-Perthes disease. Current problems in orthopaedics. London: Churchill Livingstone; 1982.
2. Catterall A, et al. Perthes disease: is the epiphysial infarction complete ? A study of the morphology in two cases. J Bone Joint Surg. 1982;64B:276–81.
3. Chung SMK. The arterial supply of the developing proximal end of the human femur. J Bone Joint Surg. 1976;58A:961–70.
4. Crock HV. The blood supply of the lower limb bones in man. London: Livingstone Ltd; 1967.
5. Danigelis JA, et al. 99mTc-polyphosphate bone imaging in Legg-Perthes Disease. Radiology. 1975;115:407.
6. Eyring EJ, Murray WR. The effect of joint position on the pressure of intra-articular effusion. J Bone Joint Surg. 1964;46A:1235–41.
7. Ferguson AB, Howorth MB. Coxa plana and related conditions at hip. J Bone Joint Surg. 1934;16:781–803.
8. Ford LT, Key JA. A study of experimental trauma to the distal femoral epiphysis in rabbits. J Bone Joint Surg. 1956;38A:84–92.

9. Gage JR, Winter RB. Avascular necrosis of the capital femoral epiphysis as a complication of closed reduction of congenital dislocation of the hip. J Bone Joint Surg. 1972;54A:373–88.
10. Gore DR. Iatrogenic avascular necrosis of the hip in young children. A review of six cases. J Bone Joint Surg. 1974;56A:493–502.
11. Mckibbin B, Ralis Z. Pathological changes in a case of Perthes' disease. J Bone Joint Surg. 1974;56B:438–47.
12. Salter RB, et al. Legg-Calvé-Perthes disease. The prognostic significance of the subchondral fracture and a two-group classification of the femoral head involvement. J Bone Joint Surg. 1984;66A:479–89.
13. Calver R. Radionuclide scanning in the early diagnosis of Perthes disease. J Bone Joint Surg. 1981;63B:379.
14. Calvert PT, et al. Effects of vascular occlusion on the femoral head in growing rabbits. Acta Orthop Scand. 1984;55:526–30.
15. Craveiro Lopes N, Bettencourt P. Doença de Legg-Calvé-Perthes. Evolução Natural de 9 Parâmetros Radiológicos em Ancas Assintomáticas e Sintomáticas. Rev Ortop Traum IB. 1986;12P:153–64.
16. Freeman MAR, England JPS. Experimental infarction of the immature canine femoral head. Proc R Soc Med. 1969;62:431–3.
17. Mendonça J, et al. Detecção Cintigráfica Precoce da Doença de Perthes na Sinovite da Anca. Rev Ortop Traum IB. 1984;10P:153–61.
18. Paterson D, Savage JP. The nuclide bone scan in the diagnosis of Perthes' disease. Clin Orthop Relat Res. 1986;209:23–9.
19. Sanchis M, et al. Sinovitis Transitoria de la Cadera. Su Relacion con la Enfermedad de Perthes. Estudio Gammagráfico. Rev Esp Cir Osteoart. 1986;21:273.
20. Sutherland AD, et al. The diagnosis and management of Perthes disease. J Bone Joint Surg. 1980;62B:300.
21. Bencano AC, Rueda FSL, Padron JR. Enfermedad de Perthes: Mecanismo Patogenético Experimental. Rev Ortop Traum IB. 1989;33:428–33.
22. Vegter J. Fractional necrosis of the femoral head epiphysis after transient increase in joint pressure. An experimental study in juvenile rabbits. J Bone Joint Surg. 1987;69B:530–5.
23. Sotto-Hall R, et al. Variations in the intra-articular pressure of the hip joint in injury and disease. J Bone Joint Surg. 1964;46A:509–17.
24. Kallio P. Transient synovitis and Perthes disease is there an aetiological connection? J Bone Joint Surg. 1986;68B:808–11.
25. Vegter J. The influence of joint posture on intra-articular pressure. A study of transient synovitis and Perthes' disease. J Bone Joint Surg. 1987;69B:71–4.
26. Scoenecker PL, Bitz OM, Whiteside LA. The acute effect of position of immobilization on capital femoral epiphyseal blood flow. A quantitative study using the hydrogen washout technique. J Bone Joint Surg. 1978;60-A:899–904.
27. Inoue A, et al. The pathogenesis of Perthes disease. J Bone Joint Surg. 1976;58B:453–61.
28. Craveiro Lopes N, Bettencourt P. Doença de Legg-Calvé-Perthes. Novos Conceitos Diagnósticos e Terapéuticos. Rev Ortop Traum IB. 1985;11P:31–47.
29. Craveiro Lopes N, Bettencourt P. Resultados do tratamento conservador nas ancas com menos de 50% do núcleo lesado e sem sinais de cabeça em risco. Rev Ortop Traum IB. 1987;13P:35–40.
30. Craveiro Lopes N, Bettencourt P. Doença de Legg-Calvé-Perthes. Doença de Legg-Calvé-Perthes. Resultados do tratamento cirúrgico com osteotomia intertrocantérica de tríplo efeitos em ancas com sinais de "cabeça em risco". Rev Ortop Traum IB. 1987;13P:95–105.
31. Craveiro Lopes N, Bettencourt P. Doença de Legg-Calvé-Perthes. Resultado do tratamento cirúrgico com a tunelização transfisária cervico-cefálica nas ancas com mais de 50% de envolvimento e sem sinais de "cabeça em risco". Rev Ortop Traum IB. 1988;14P:39–46.
32. Craveiro Lopes N, Vasconcelos FP, Amaral S. Goniometria da anca normal. Estudo radiográfico na Criança Portuguesa. Rev Ortop Traum IB. 1989;15P:11–24.
33. Futami T, Kasahara Y, Suzuki S, Ushikubo S, Tsuchiya T. Ultrasonography in transient synovitis and early Perthes' disease. J Bone Joint Surg Br. 1991;73-B:635–9.

34. Bos CF, Bloem JL, Bloem RM. Sequential magnetic resonance imaging in Perthes' disease. J Bone Joint Surg Br. 1991;73-B:219–24.
35. Craveiro Lopes N. Modelo experimental básico para o estudo da patologia do crescimento na extremidade superior do fémur. Rev Port Ortop Traum. 1993;1(2):199–213.
36. Craveiro Lopes N. Legg-Calvé-Perthes disease after repeated extension-internal rotation posture of the hip followed by microtrauma. An experimental study in the growing rabbit. J Bone Joint Surg Br. 1993;(Supp II):75–164.
37. Craveiro Lopes N. Doença de Legg-Calvé-Perthes. Current concept review. Rev Port Ortop Traum. 1994;2(1):75–9.
38. Craveiro Lopes N. Etiopatogenia da doença de Legg-Calvé-Perthes. Modelo experimental no coelho White New Zealand em crescimento. Rev Port Ortop Traum. 1994;2(1):81–93.
39. Craveiro Lopes N. Estudo dos efeitos da tunelização transfisária cervico-cefálica do fémur como método para prevenir a doença de Legg-Calvé-Perthes. Modelo experimental no coelho White New Zealand em crescimento. Ver Port Ortop Traum. 1994;2(4):395–404.
40. Campbell CJ, et al. The effect produced in the cartilaginous epiphyseal plate of immature dogs by experimental surgical traumata. J Bone Joint Surg. 1959;41A:1221–42.
41. Haas SL. Restriction of bone growth by pins through the epiphyseal cartilaginous plate. J Bone Joint Surg. 1950;32A:338–43.
42. Imbert R. Pathologie Expérimentale de l'appareil de croissance des os longs. Marseille Chir. 1951;3:581–99.
43. Kleinberg S, Bochman J. The operative versus the manipulative treatment of slipped epiphysis. J Am Med Assoc. 1936;107(1):154.
44. Ollier L. Traité Expérimental et Clinique de la Régénération des Os et de la Production Artificielle du Tissu Osseux, vol. 1. Paris: Masson et Fils; 1867. p. 236–8, pp. 386–392.
45. Siffert RS. The effect of staples and longitudinal wires on epiphyseal growth. J Bone Joint Surg. 1956;38A:1077–88.
46. Harris WR, Hobson KW. Histological changes in experimentally displaced upper femoral epiphyses in rabbits. J Bone Joint Surg. 1956;38B:914–21.
47. Soeur R, de Racker C. L'aspect anatomopathologique de l'osteochondrite et les theories pathogéniques qui s'y rapportent. Acta Orthop Belg. 1952;18:57–102.
48. Arlet J, Ficat C. Ischemic necrosis of the femoral head. Treatment by core decompression. J Bone Joint Surg Am. 1990;72:151–2.
49. Herrera-Soto JA, Price CT. Core decompression for juvenile osteonecrosis. Orthop Clin North Am. 2011;42(3):429–36.
50. Borgsmiller WK, et al. The effect of hydrostatic pressure in the hip joint on proximal femoral epiphyseal and metaphyseal blood flow. Trans Orthop Res Soc. 1980;5:23.
51. Heikel HVA. On ossification and growth of certain bones of rabbit and man. Acta Orthop Scand. 1960;29(16):171–84.
52. Guille JT, Lipton GE, Szoke G, et al. Legg-Calve-Perthes disease in girls. A comparison of the results with those seen in boys. J Bone Joint Surg Am. 1998;80:1256–63.
53. Harrison MHM, Blakemore ME. A study of the "normal" hip in children with unilateral Perthes' disease. J Bone Joint Surg Br. 1980;62-B:31–6.
54. Salter RB, Thompson GH. Legg-Calve-Perthes disease. The prognostic significance of the subchondral fracture and a two-group classification of the femoral head involvement. J Bone Joint Surg Am. 1984;66A:479–89.
55. Peterson HA. Premature physeal parcial closure: assessment and treatment. J Bone Joint Surg Am. 1987;69B:154.
56. Craveiro Lopes N. Prevention of Legg-Calvé-Perthes disease. Transphyseal neck-head drilling. Rev Port Ortop Trau. 2011;19(III):213.
57. Dimitriou JK, Leonidou O, Pettas N. Acetabulum augmentation for Legg–Calve–Perthes disease. 12 children (14 hips) followed for 4 years. Acta Orthop Scand. 1997;68(Suppl 275):103–5.

58. Craveiro Lopes N, Bettencourt P. Doença de Legg-Calvé-Perthes. Evolução natural de 9 parâmetros radiológicos em ancas sintomáticas e assintomáticas. Rev Ortop Traum IB [P]. 1986;12(2):153–64.
59. Craveiro Lopes N. Treatment of Legg-Calve-Perthes disease using early transphyseal neck-head tunnelling of the femoral head combined with late hip arthrodiastasis with the Ilizarov apparatus. Comparative Study with Classic Intertrochanteric Osteotomy. Rev Port Ortop Trau. 2011;19(III):220.
60. Gallón LA, Lores CA, Zuluaga M, Pinta AF. Artrodiastasis en enfermedad de Perthes. Rev Col Ort Tra. 2007;21(3):146–53.
61. Joseph B, Mulpuri K, Varghese G. Perthes' disease in the adolescent. J Bone Joint Surg Br. 2001;83-B:715–20.
62. Maxwell SL, Lappin KJ, Kealey WD, Mcdowell BC, Cosgrove AP. Arthrodiastasis in Perthes' disease. Preliminary results. J Bone Joint Surg Br. 2004;86:244–50.
63. Segev E, Ezra E, Wientroub S. Treatment of severe late onset Perthes' disease with soft tissue release and articulated hip distraction: early results. J Pediatr Orthop. 2004;13(3):158–65.
64. Thacker M, Feldman D, Madan S, Straight J, Scher D. Hinged distraction of the adolescent arthritic hip. J Pediatr Orthop. 2005;25:178–82.
65. Wiig O, Huhnstock S, Terjesen T, . Pripp A H, Svenningsen S, The outcome and prognostic factors in children with bilateral Perthes' disease. A prospective study of 40 children with follow-up over five years. Bone Joint J 2016; 98-B:569–575.
66. Van Den Bogaert G, De Rosa E, Moens P, Fabry G, Dimeglio A. Bilateral Legg-Calvé-Perthes disease: different from unilateral disease? J Pediatr Orthop B. 1999;8(3):165–8.
67. Arie E, Johnson F, Harrison MHM. Femoral head shape in Perthes' disease: is the contralateral hip abnormal? Clin Orthop Relat Res. 1986;209:77–88.
68. Kandzierski G, Karski T, Kozlowski K. Capital femoral epiphysis and growth plate of the asymptomatic hip joint in unilateral Perthes disease. J Pediatr Orthop. 2003;12(6 Part B):380–6.
69. Kitoh H, Kitakoji T, Katoh M, Takamine Y. Delayed ossification of the proximal capital femoral epiphysis in Legg-Calve-Perthes' disease. J Bone Joint Surg Br. 2003;85(1):121–4.

5

Varus Derotational Osteotomy as Treatment of Legg-Calvé-Perthes Disease

Surya N. Mundluru and David S. Feldman

Introduction

The current accepted treatment options for Legg-Calvé-Perthes disease are based on the concept of containment. Containment can be achieved by positioning the extruding femoral head in a more covered position. This increase in coverage has been theorized to offset the disease progression, providing more favorable remodeling potential. The varus derotational osteotomy (VDRO) is one such treatment modality and has been widely accepted as a current mainstay of treatment for hip containment in Legg-Calvé-Perthes disease.

History

In 1929 Parker described using plaster abductions casts to position the entire epiphysis within the acetabulum (Fig. 5.1) [1]. The Parker "broomstick abduction cast" was one of the earliest known treatment options for Perthes. Later, many braces were created that mimicked the function of the original abduction casts. However, despite more wide adoption of abduction braces, patients would remain awkwardly constrained by the devices, negatively impacting patient compliance and adherence. It was not until 1965 when Axer described the first femoral osteotomy for the

---

S. N. Mundluru
McGovern Medical School at the University of Texas Health Science Center,
Houston, TX, USA

UT Physicians Orthopedics at Ironman Sports Medicine Institute – Texas Medical Center,
Houston, TX, USA

D. S. Feldman (✉)
Paley Orthopedic and Spine Institute, St. Mary's Medical Center, West Palm Beach, FL, USA
e-mail: dfeldman@paleyinstitute.org

D. S. Feldman, D. Paley (eds.), *Legg-Calvé-Perthes Disease*,
https://doi.org/10.1007/978-1-0716-0854-8_5

Fig. 5.1 Broomstick abduction cast by Parker, Harrison et al. [1]

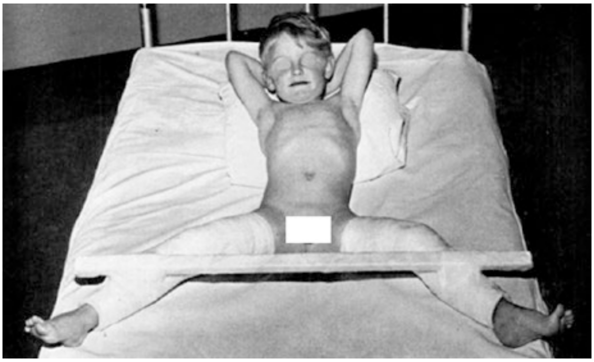


Table 5.1 Anatomical results and rate of regeneration of epiphysis of 11 patients after subtrochanteric osteotomy with derotation

Case number	Initial regeneration (months)	Final consolidation (months)
1	5	5
2	10[a]	34
3	10	16
4	3	8
5	4.5	10
6	3	16
7	9[a]	23
8	4	19
9	7	20
10	3	16
11	5	13
Average time	6	15.5[b]

From Axer et al. *JBJS* (1965)
[a] Subchondral defect
[b] Excludes case 7

treatment of Perthes' disease. His goal was to find "a less restricted method of treatment yielding similar or better results." The procedure was described as a varus subtrochanteric osteotomy with rotation, positioning the "plastic" epiphysis in the center of the joint space [2]. In his series of 12 patients from 1957 to 1963, 11 of the patients were noted to have "very good to good" outcomes with a mean remodeling time of 15.5 months (Tables 5.1 and 5.2). Only one patient was noted to have limitation of movement with a limp and a limb length discrepancy of 2.5 cm. All patients were immobilized for an average of 2–3 months in a plaster spica cast until radiographic signs of union. In 1976 Lloyd-Roberts and Catterall performed the first randomized control study looking at untreated Perthes' disease (total of 75 patients) in comparison to 48 patients treated with varus rotational osteotomy. They showed that the patients that had the best results after osteotomy were those that the operation was performed within 7 months of symptoms. They also found that patients treated at around the age of 6 years had better results compared to those treated at a much older age [3]. These early pioneering studies paved the way to further developments regarding operative management of Perthes' disease.

Table 5.2 Clinical results of 12 patients that had undergone a subtrochanteric osteotomy with derotation

Case number	Limp	Disparity (cm)	Limitation of movement
1	No	+1.0	Negligible
2	No	None	Negligible
3	No	−0.5 to −1.0	Negligible
4	Yes	−2.5	Considerable
5	No	None	Negligible
6	No	None	Negligible
7	No	None	Negligible
8	No[a]	−1.2	Negligible
9	No	None	Negligible
10	No	−0.5	Negligible
11	No	−0.5	Negligible
12	Slight	−1.5	Negligible

From Axer et al. JBJS, 1965

[a] Limps slightly when running

Biology

Axer had suggested that one of the effects of varus osteotomy was interruption of the destructive phase of Perthes' disease and a reduction in the duration of the disease [4]. Joseph further corroborated Axer's findings, demonstrating that in fact some children operated at stage I of the disease had an overall reduction in the duration of the disease primarily because of bypass of the stage of fragmentation. Even in children who did pass through the stage of fragmentation, the duration of the fragmentation phase was shorter than in children treated non-operatively. Additionally metaphyseal widening and epiphyseal extrusion, both of which tend to increase abruptly in non-operated children, are minimized in patients who underwent femoral osteotomy [5]. Heikkinen theorized that besides providing containment, the varus rotational osteotomy had a beneficial effect on the "disturbed venous drainage" of the affected hip in addition to decreasing the load against the involved head without restricting normal activity. He looked at 67 patients treated with the osteotomy and found that 74% of patients between the age of 5 and 9 had good results as opposed to only 40% of patients older than 9 years of age. He also performed venography on selected patients showing venous pooling and congestion in the affected hips was relieved by osteotomy improving femoral head blood flow (Fig. 5.2) [6]. However in 1992, Lee performed serial quantitative scintigraphic studies on 25 hips with Perthes' disease pre- and post-VDRO. The studies were performed preoperatively and at 2 weeks, 6 weeks, 6 months, 1 year, and 2 years postoperatively. He found that the vascularity of the femoral head decreased significantly at the 2-week initial visit; however, thereafter it steadily increased back to the preoperative level by 6 months with no statistically significant increase from preop at 2 years [7].

Fig. 5.2 Venogram of bilateral hips, right side unaffected and left side affected by Perthes' disease. Venous drainage was assessed after 6 ml of radio-opaque contrast material was injected into bilateral femoral necks. The affected side (left) shows increased reflux and extravasation into the diaphysis compared to the contralateral side. (Heikkinen et al. [6])

Techniques

Since Axer's original publication in 1965, the varus osteotomy has been adopted by surgeons and modified allowing for better surgical outcomes. Axer in 1980 published an update to his original paper. He described three variations. Excision of a medial based wedge with its reversal to obtain "varusation and containment" with anterior head containment by derotation. In the cases he was unable to achieve the desired degree of medial rotation of the proximal fragment, an excision of a posterior-based wedge to obtain head containment by extension was performed (Fig. 5.3). In young children (<6 years), an opening wedge technique was performed with a transverse division of the shaft in the subtrochanteric region, followed by pre-bending of the plate to the appropriate varus angle and application. He also decreased the postoperative immobilization and spica casting to only a total of 6 weeks [8]. Grant and Lehman described an innovative technique utilizing a cannulated blade plate for varus osteotomy. The system included cannulated chisels and cannulated one-piece plate that could be inserted over a guidewire (Fig. 5.4). They also recommended using a pretemplated/predetermined line known as the neck metaphyseal angle for determination of the position of the wire and blade placement to allow a reliable correction of the neck shaft angle (Fig. 5.5) [9, 10]. Joseph et al. described using a combination of a trochanteric epiphyseodesis and varus osteotomy (Fig. 5.6). There is no plaster immobilization utilized, but weight-bearing was avoided until mature bone was seen to cover the lateral part of the femoral epiphysis. In their study, they found a much larger proportion of children, who had this operation, had spherical femoral heads compared to a control group (62.5% vs 20%; $p < 0.001$). They performed the procedure on Catterall groups II–IV, children ages 7–12 years [11]. Ito et al. described using a

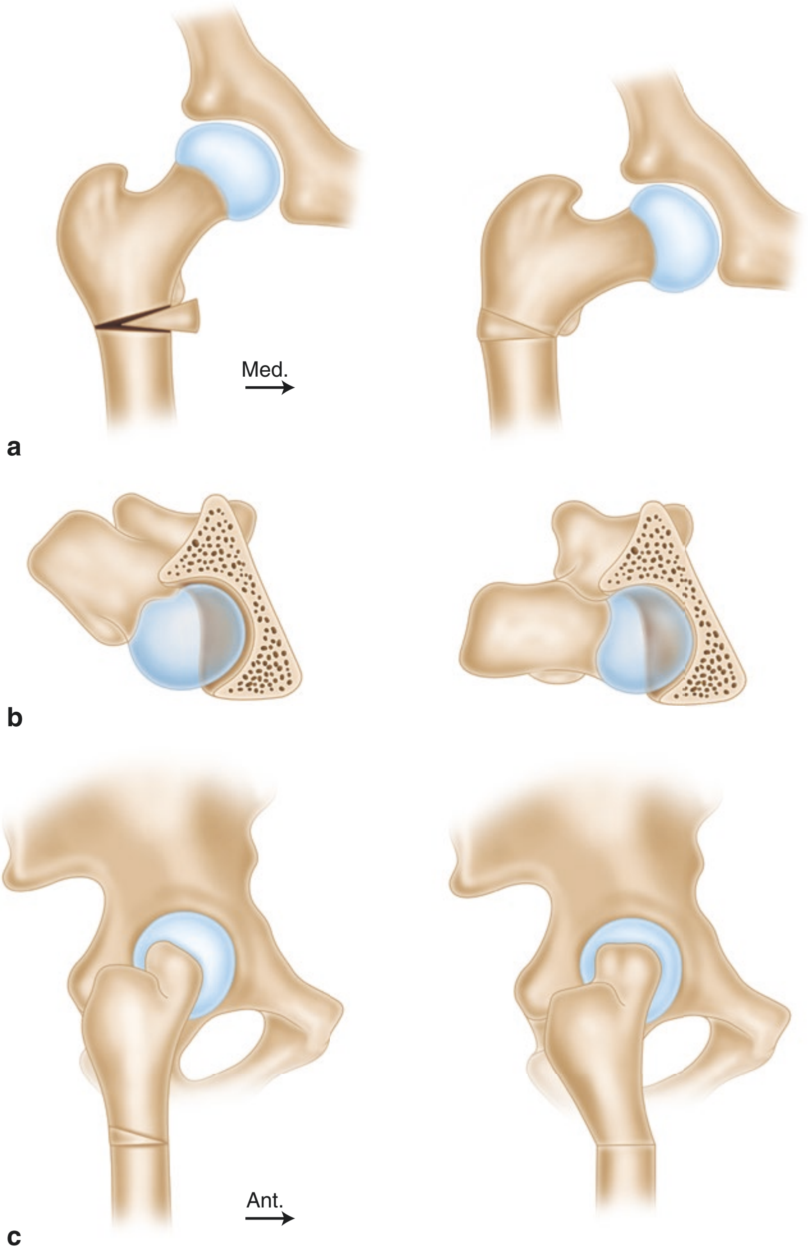
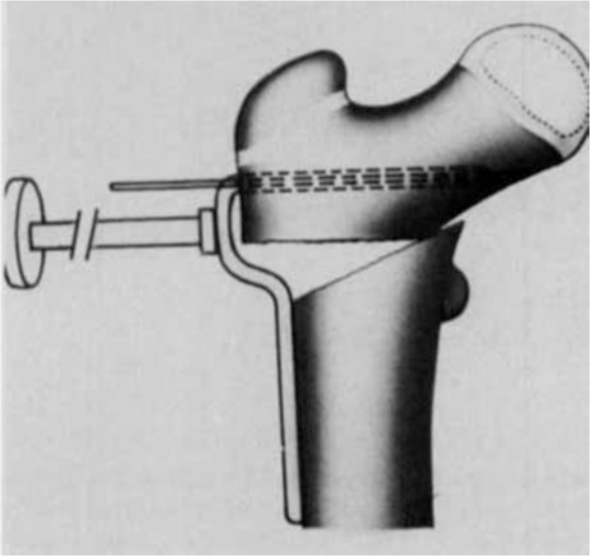
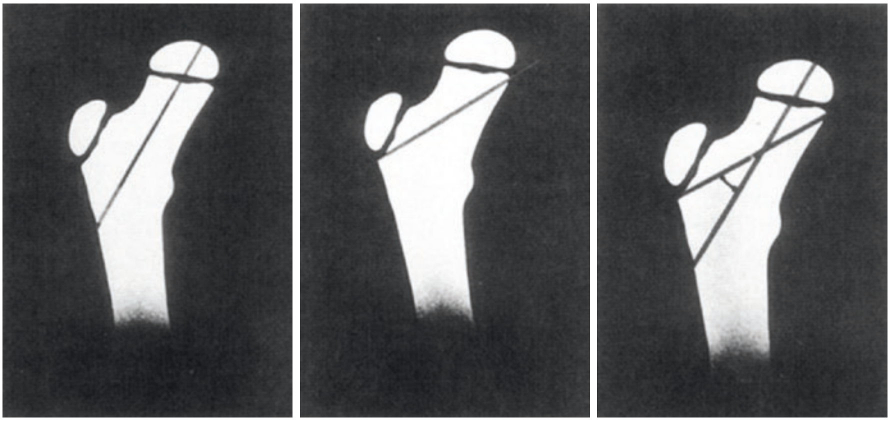


Fig. 5.3 Osteotomy techniques by Axer et al. [8]

Fig. 5.4 A new system, which included cannulated chisels and cannulated one-piece plate that can be inserted over a guidewire. (Grant, Lehman et al. [9])





Center Neck Axis

Trochanteric Metaphyseal Line

Neck Metaphyseal Angle

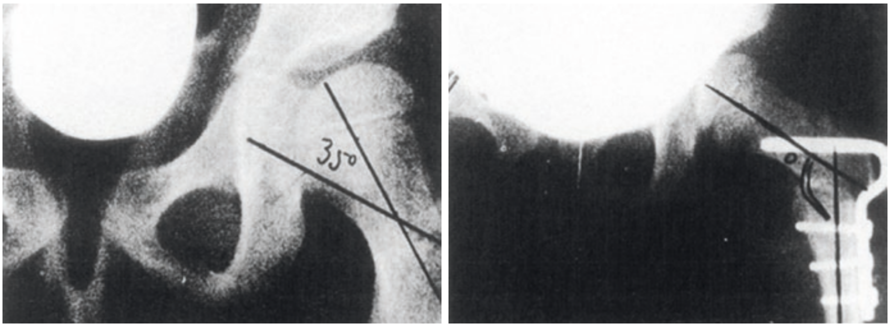


Fig. 5.5 Neck metaphyseal angle. (Grant et al. [10])

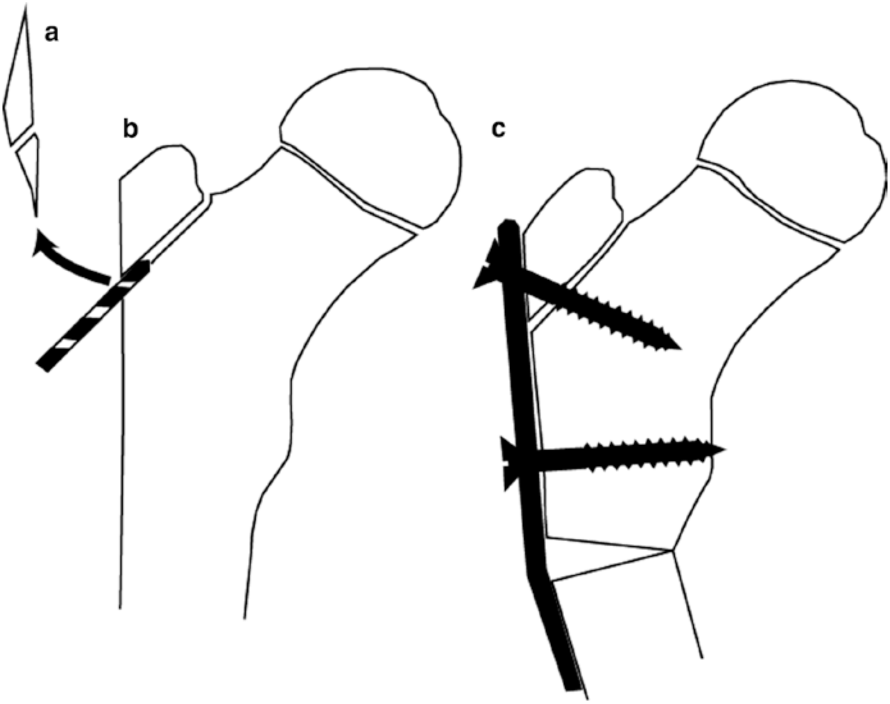


Fig. 5.6 Surgical technique that includes a trochanteric epiphyseodesis and varus osteotomy as described by Joseph et al. This method involves an osteotomy of the flare of the trochanter (**a**), then the growth plate of the trochanter is drilled (**b**), finally an open-wedge osteotomy is performed and a plate and screws are used to maintain a 20° varus angle (**c**)

three-dimensional corrective external fixator system for the varus and rotational correction (Fig. 5.7). In their series, 36 patients with Perthes' disease were treated with the technique for 3 months. All achieved union at a mean of 4 months from the original surgery. No malunions or device failures were noted [12]. Atsumi described a rotational open-wedge osteotomy for patients older than 7 years of age with Perthes' disease. In his technique a cannulated blade plate was used and partially inserted in an anteriorly shifted position. Next an intertrochanteric osteotomy was performed. After osteotomy the plate was further "seeded" to the appropriate depth. The device was then rotated posteriorly and fixed in line with the femoral shaft (Fig. 5.8). Using this technique on 19 patients with Perthes' disease Catterall stages III and IV, he noted an accelerated rate of repair of the fragmented epiphysis by 12 months. The mean age of the patient population was 7.8 years old [13]. In a muticenter prospective study, Kim assessed 52 patients treated with proximal femoral varus osteotomies for Perthes' disease. All patients were 6 years or older at the time of diagnosis (mean age 8 years). He found that greater varus angulation did not produce better preservation of the femoral head. Based on his findings, over- or under-"varusization" did not provide the most favorable outcomes. Best outcomes were noted in patients with initial lateral pillar B classification with only 10–15 ° of varus correction [14].

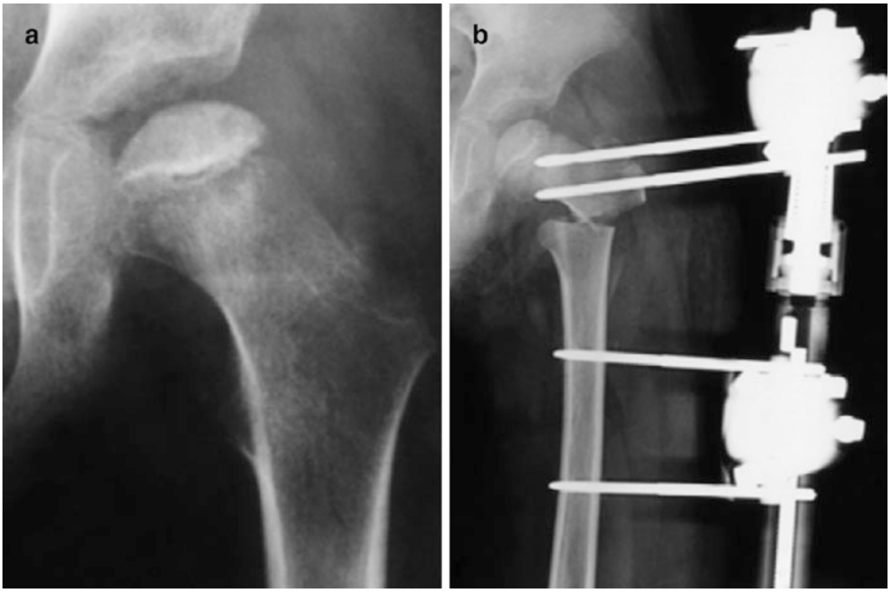


Fig. 5.7 Three-dimensional external fixator for treatment of Perthes' disease, Ito et al. [12]. (**a**) Preoperative AP radiograph demonstrating early stage Perthes. (**b**) After varus osteotomy with an external fixation

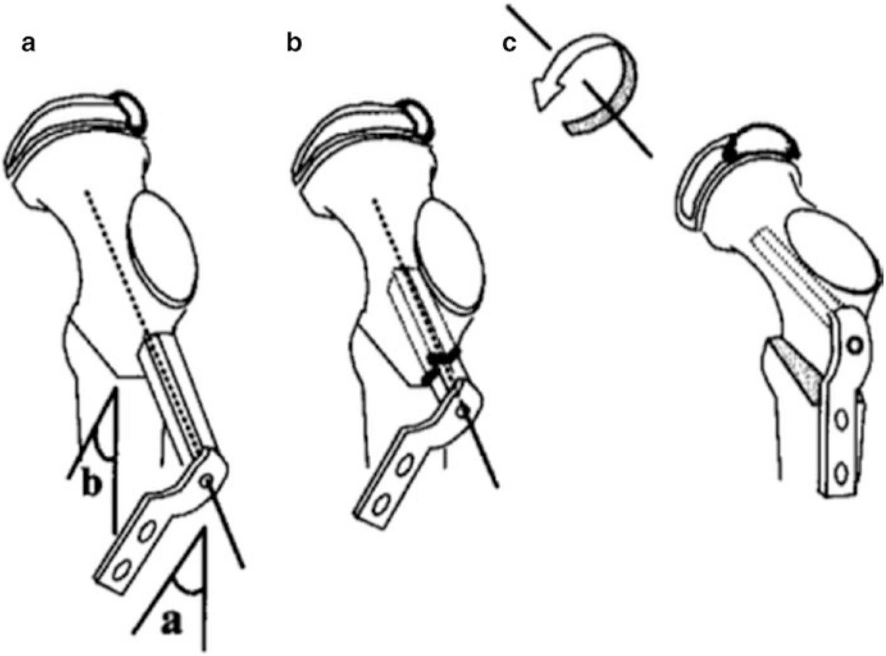


Fig. 5.8 Opening wedge intertrochanteric varus rotational osteotomy, Atsumi et al. [13]. (**a**) Planned angles of correction for varus derotational osteotomy. (**b**) Placement of blade plate to match preoperative plan. (**c**) After correction to varus and rotation

Time of Intervention

Though many have accepted the benefit of varus derotational osteotomy for the treatment of Perthes' disease, much controversy remains on the appropriate time and stage for maximal benefit from treatment. In 1980, Laurent looked at 78 patients Catterall groups II, III, and IV who underwent subtrochanteric varus derotational osteotomy as proposed by Axer. The average age of patients with the onset of symptoms was 6 years and 6 months. The average age at the time of osteotomy was 7 years and 8 months. He found that patients that had the osteotomy within 1 year of onset of symptoms and were Catterall stages II and III at intervention had the best outcomes (Fig. 5.9) [15]. Friedlander conducted a retrospective review of 116 patients with varus osteotomy for Herring group B and C disease. Radiographic outcome was assessed utilizing Stulberg's classification to grade residual deformity. Follow-up averaged 6 years and 9 months. Stulberg classes I and II (spherically congruent) were obtained in 86% of patients younger than 9 years old with Herring class B disease. Patients 9 years and older with Herring class B (Fig. 5.10) disease had spherically congruent results in 67% of cases. Patients with Herring class C disease 9 years and older had spherically congruent results in only 30% of cases. Patients 9 years or younger with Herring class C disease at time of intervention only had congruent results in 43% [16]. Joseph in 2003 evaluated the optimal timing for containment surgery for Perthes' disease. Ninety-seven patients who underwent the femoral osteotomy technique as described by him were included in the study. The mean age of patient population was 8 years of age. Using univariate and multivariate analysis, the most significant variable that had a bearing on outcome was timing of surgery. The chances of retaining a spherical femoral head were much higher in children operated on either the stage of avascular necrosis or in the early part of the fragmentation stage than in those operated later (Fig. 5.11). The goal of surgical intervention was effectively to bypass the stage of fragmentation by containment of the femoral head. By early intervention the integrity of the lateral pillar was maintained, the duration of the disease was reduced compared to prior controls, and the

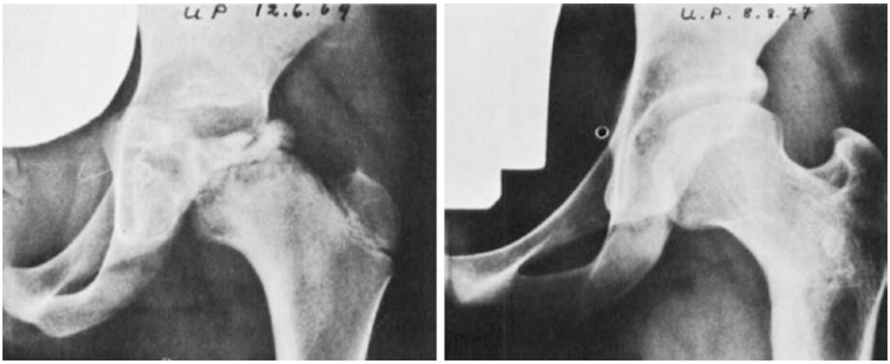


Fig. 5.9 (**a**) Preoperative xray demonstrating femoral head collapse and extrusion. (**b**) After varus osteotomy and healing there is a concentric femoral head, Laurent et al. [15]

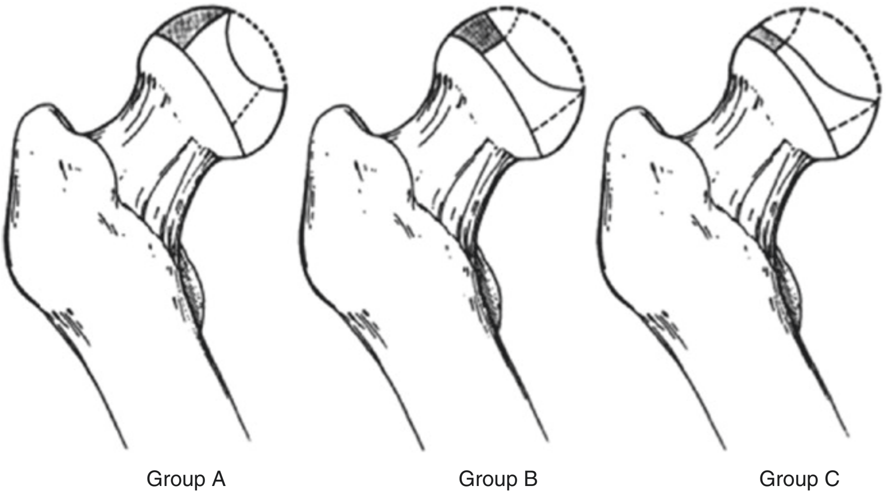


Fig. 5.10 An anteroposterior (AP) pelvis radiograph during the fragmentation phase was used to classify patients into group B (hips with >50% of the lateral pillar intact) (Fig. 5.2) and group C hips with <50% of the original height of the lateral pillar, Friedlander et al. [16]

adverse metaphyseal and acetabular changes that tend to appear most frequently in the stage of fragmentation did not develop [5].

Complications

As with any surgical intervention, varus derotational osteotomies are associated with potential complications. Complications described in the literature include premature epiphyseal closure, leg length discrepancy, greater trochanter overgrowth, abductor lurch, loss of motion, pain, and over-varusation.

Premature epiphyseal closure after VDRO was described by Barnes in 1980. He reviewed 22 patients with an average age of onset of 6 years, with Catterall group III and IV disease who developed premature closure compared to the contralateral hip. The patients had a follow-up at an average age of 14 years old and had an average leg length discrepancy range of 0.5–3.0 cm (average 2 cm). Other complication noted in the study included greater trochanter overgrowth in some patients. Decrease in abduction did correlate with greater trochanter overgrowth (impingement on the ilium); however, Trendelenburg sign, which was seen in seven patients, did not correlate. Overall 16 patients were pain free and active, while 6 had occasional pain after activity [17]. Leitch looked at 72 patients with Perthes' disease, 20 of which were treated with VDRO and 27 of which were treated with a Salter (innominate) osteotomy. Catterall grade at time of diagnosis was evenly spread across all groups (Grades II–IV). The study showed a leg length discrepancy of greater than 2 cm in a total of 6% of patients in the total population (13% of patients treated with VDRO and 9% of patients treated non-operatively) (Fig. 5.12).

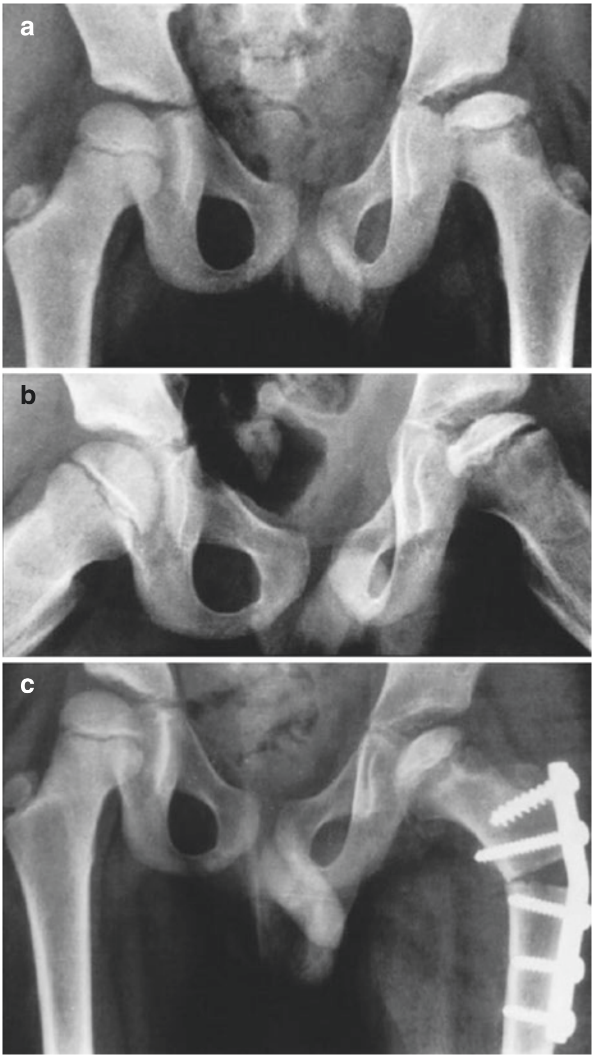


Fig. 5.11 (**a**) Preoperative xrays (**b**) Radiograph after osteotomy. (**c**) Radiograph after healing, Joseph et al. [5]

Greater trochanter overgrowth (determined by articulotrochanteric distance less than 5 mm on final AP pelvis radiograph) was noted in 26% of the patients treated with VDRO; however, interestingly 29% of patients treated non-operatively also had the phenomenon. Forty-three percent of the patients with greater trochanter overgrowth had a positive Trendelenburg sign. Weiner reviewed 79 patients who underwent VDRO. Patients were Catterall II, III, or IV at time of indication for surgery. Average age at surgery was 7.3 years, and average follow-up was 4.8 years. Fifteen patients developed “pitfalls” which included greater trochanter overgrowth (nine patients), persistent varus angulation less than 105 degrees (seven patients), shortening of the extremity greater than 1 inch (three patients), and premature

Fig. 5.12 Standing radiograph demonstrating limb length discrepancy after varus ostoeotomy, Leitch et al. *CORR* (1991)

physeal closure (nine patients). Of the patients that underwent greater trochanteric epiphysiodesis at initial surgery, only two patients (5.4%) developed a complication. In contrast in the patients operated with osteotomy without epiphysiodesis, 14 patients (28%) developed a complication. Sixteen patients had immediate postoperative varus inclination of less than 105 degrees, and seven of those (44%) developed a complication. Of the 63 patients that had postoperative varus of greater than 105 degrees, only 8 (13%) had a complication. So conclusions from this study showed that a neck shaft angle greater than 105 degrees and trochanter epiphysiodesis at initial surgery were less likely to be associated with long-term problems; however, no statistical analysis was performed in the study [18].

No discussion about femoral osteotomies would be complete without mentioning the Sugioka osteotomy [19]. If there is one third of the femoral head still intact and rotated between 20 and 30 degrees, the femoral head osteotomy has demonstrated excellent results beyond 10 years, in the hands of Dr. Sugioka [20]. These results have not been able to be reproduced in other western centers [21, 22]. Studies have claimed that there may be a difference in race in terms of outcome or perhaps it is the difficult nature of the procedure. At this time, this procedure is rarely performed outside of Japan.

Conclusions

The varus derotational osteotomy is an acceptable treatment option for Legg-Calvé-Perthes disease. Since its description by Axer in 1965, it has been a mainstay in the treatment algorithm. Since its inception it has undergone many iterations with a variety of techniques described to achieve the same goal: femoral head containment. By positioning an extruding femoral head in a more covered position, there is a theoretical benefit in accelerated healing, decrease in fragmentation of the epiphysis, and potential change in the vascular supply. Though the risks with this procedure are low, there is a potential for complications that may require adjunct procedures. The varus derotational osteotomy should be considered in the management of Perthes' disease and will remain a mainstay in treatment for a long time to come.

References

1. Harrison MH, Menon MP. Legg-Calvé-Perthes disease. The value of roentgenographic measurement in clinical practice with special reference to the broomstick plaster method. J Bone Joint Surg. 1966;48(7):1301–18.
2. Axer A. Subtrochanteric osteotomy in the treatment of Perthes' disease. J Bone Joint Surg Br. 1965;47:489.
3. Lloyd-Roberts GC, Catterall A. A controlled study of the indications for and the results of femoral osteotomy in Perthes' disease. J Bone Joint Surg Br. 1976;58:31.
4. Axer A, Schiller MG. The pathogenesis of the early deformity of the capital femoral epiphysis in Legg-Calve-Perthes syndrome (LCPS): an arthrographic study. Clin Orthop Relat Res. 1972;84:106.
5. 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;14(1):10–5.
6. Heikkinen E, Puranen J. Evaluation of femoral osteotomy in the treatment of Legg-Calvé-Perthes disease. Clin Orthop Relat Res. 1980;150:60–8.
7. Lee DY, Seong SC, Choi IH, Chung CY, Chang BS. Changes of blood flow of the femoral head after subtrochanteric osteotomy in Legg-Perthes' disease: a serial scintigraphic study. J Pediatr Orthop. 1992;12(6):731.
8. Axer A, Gershuni DH, Hendel D, Mirovski Y. Indications for femoral osteotomy in Legg-Calvé-Perthes disease. Clin Orthop Relat Res. 1980;150:78–87.
9. Grant AD, Lehman WB, Strongwater AM, Atar D. Cannulated blade plate for proximal femoral varus osteotomy. Clin Orthop Relat Res. 1990;259:111–3.
10. Grant AD, Strongwater A, Lehman W, Nelson J, Atar D. Neck metaphyseal angle guidance in proximal femoral varus osteotomy. J Pediatr Orthop. 1990;10(1):9–13.
11. Joseph B, Srinivas G, Thomas R. Management of Perthes disease of late onset in southern India. The evaluation of a surgical method. J Bone Joint Surg Br. 1996;78(4):625–30.
12. Ito H, Minami A, Suzuki K, Matsuno T. Three-dimensionally corrective external fixator system for proximal femoral osteotomy. J Pediatr Orthop. 2001;21(5):652–6.
13. Atsumi T, Yoshiwara S. Rotational open wedge osteotomy in a patient aged older than 7 years with Perthes' disease--a preliminary report. Arch Orthop Trauma Surg. 2002;122(6):346–9. https://doi.org/10.1007/s00402-002-0408-5.
14. Kim HK, da Cunha AM, Browne R, Kim HT, Herring JA. How much varus is optimal with proximal femoral osteotomy to preserve the femoral head in Legg-Calvé-Perthes disease? J Bone Joint Surg. 2011;93(4):341–7. https://doi.org/10.2106/JBJS.J.00830.

15. Laurent LE, Poussa M. Intertrochanteric varus osteotomy in the treatment of Perthes' disease. Clin Orthop Relat Res. 1980;150:73–7.
16. Friedlander JK, Weiner DS. Radiographic results of proximal femoral varus osteotomy in Legg-Calvé-Perthes disease. J Pediatr Orthop. 2000;20(5):566–71.
17. Barnes JM. Premature epiphysial closure in Perthes' disease. J Bone Joint Surg Br. 1980;62-B(4):432–7.
18. Weiner SD, Weiner DS, Riley PM. Pitfalls in treatment of Legg-Calvé-Perthes disease using proximal femoral varus osteotomy. J Pediatr Orthop. 1991;11(1):20–4.
19. Sugioka Y. Transtrochanteric rotational osteotomy in the treatment of idiopathic and steroid-induced femoral head necrosis, Perthes' disease, slipped capital femoral epiphysis, and osteoarthritis of the hip. Indications and results. Clin Orthop Relat Res. 1984;(184):12–23.
20. Inao S, Ando M, Gotoh E, Matsuno T. Minimum 10-year results of Sugioka's osteotomy for femoral head osteonecrosis. Clin Orthop Relat Res. 1999;368:141–8.
21. Dean MT, Cabanela ME. Transtrochanteric anterior rotational osteotomy for avascular necrosis of the femoral head. Long-term results. J Bone Joint Surg Br. 1993;75:597–601.
22. Rijnen WHC, Gardeniers JWM, Westrek BLM, Buma P, Schreurs BW. Sugioka's osteotomy for femoral-head necrosis in young Caucasians. Int Orthop. 2005;29:140–4.

Pelvic Osteotomies as Treatment for Legg-Calvé-Perthes Disease

6

Keith Bachmann and Vidyadhar Upasani

Introduction

This chapter will focus on pelvic osteotomies for the treatment of Legg-Calve-Perthes disease (LCPD). It is fitting that Salter [1] was the first to propose the concept of femoral head containment as the osteotomy he described for developmental dysplasia of the hip is the most commonly performed pelvic osteotomy for this patient population. Salter described the vascular insult, revascularization, and subchondral pathologic fracture that are observed in Perthes disease. In fact, the porcine model that Salter developed is still being used today to further understand this pathologic process and develop treatment strategies [2, 3].

In this same seminar [1], Salter also summarized the principal clinical aims for the treatment of LCPD. Avoiding late arthritis is the most significant goal, and he proposed three factors to consider: (1) children presenting before the age of 6 rarely develop femoral head deformity; (2) femoral head deformity was significant in terms of predicting long-term arthritis; and (3) femoral head deformity is preventable. Salter's principles for treatment thus include regaining full hip motion and then preventing or correcting extrusion of the femoral head (i.e., loss of containment). The final principle is to allow weight bearing of the contained femoral head to promote remodeling. With this Salter felt that the new goal for Perthes treatment should be containment and motion to allow for femoral head remodeling instead of old ideas of weight-relieving caliper among others. The best way to achieve this goal in his opinion was surgical treatment to provide a permanent containment of

---

K. Bachmann
Departments of Orthopaedic Surgery, and Public Health Sciences, University of Virginia,
Charlottesville, VA, USA

V. Upasani (✉)
Department of Orthopedic Surgery, Rady Children's Hospital San Diego,
San Diego, CA, USA
e-mail: vupasani@rchsd.org

D. S. Feldman, D. Paley (eds.), *Legg-Calvé-Perthes Disease*,
https://doi.org/10.1007/978-1-0716-0854-8_6

the femoral head. This containment would then not be reliant on patient compliance (once healing has occurred), and the remodeling can result in a spherical femoral head and symmetric hip motion.

Innominate Osteotomy

In 1961 Salter [4] described his innominate osteotomy for DDH. His principles include correcting the acetabulum to a position that maintains reduction and then elimination of the redundant capsular tissue superolaterally where the femoral head was resting prior to relocation. In 1965 [5] he presented on the use of the innominate osteotomy for Perthes disease at an Israeli Orthopaedic Society meeting (Fig. 6.1). This included discussion of a porcine model in which he tied off the epiphyseal vessels with suture ligation as well as joint insufflation maintaining pressure above 70 mmHg to recreate the ischemic necrosis of Perthes disease. Once this ischemic insult was removed, they found a “head within a head” formed. This new bone was deformed rather than the necrotic bone “softening”, flipping the paradigm. Salter also found in the pig model that the head would not deform with weight bearing if the leg was kept in flexion and abduction maintaining head containment. Salter’s recommendations in 1966 were innominate osteotomy for children over 6 years of age with full hip motion and no femoral head deformity.

Early studies reporting outcomes of Perthes disease suffered from inconsistency in terms of classification systems used for staging disease prior to treatment and for outcomes. Most studies at the time used Catterall’s classification [6] of femoral head involvement and measured outcomes using Mose concentric circles [7] method. The Mose concentric circles outcome involved plastic overlay with 2 mm concentric circles. If the femoral head was perfectly round on AP and lateral views, it was considered excellent or good outcome. If the sphericity was within 2 mm, it was a fair outcome, and sphericity with borders extending beyond 2 mm is classified as poor outcome.

In this context the early studies regarding Salter’s innominate osteotomy for Perthes disease appeared. Barer in 1978 [8] reported on 55 involved hips with 23 undergoing Salter innominate osteotomy. Inclusion criteria for surgery included age greater than 4 years, Catterall group 3 or 4 with consideration to group 2 in older patients. Of the 23 hips that underwent innominate osteotomy, 87% had a good or fair outcome based on Mose criteria with 75% good or fair outcome with ambulation in abduction or with a weight-relieving caliper. They recommended Salter osteotomy for more involved Perthes disease. In comparison with another popular early treatment method, Ingman in 1982 [9] compared Salter osteotomy to spica casting. In this initial study, they compared 41 innominate osteotomies to a group of 33 patients treated in a hip spica refined from 70 total treated in a hip spica at 30 degrees of abduction and 10 degrees of internal rotation. Their outcomes were rated clinically and radiographically. Clinical outcome was considered good if there was occasional minimal discomfort and minimal restriction of hip movement and fair if there was pain causing minor restriction of activity or more than 10 degrees

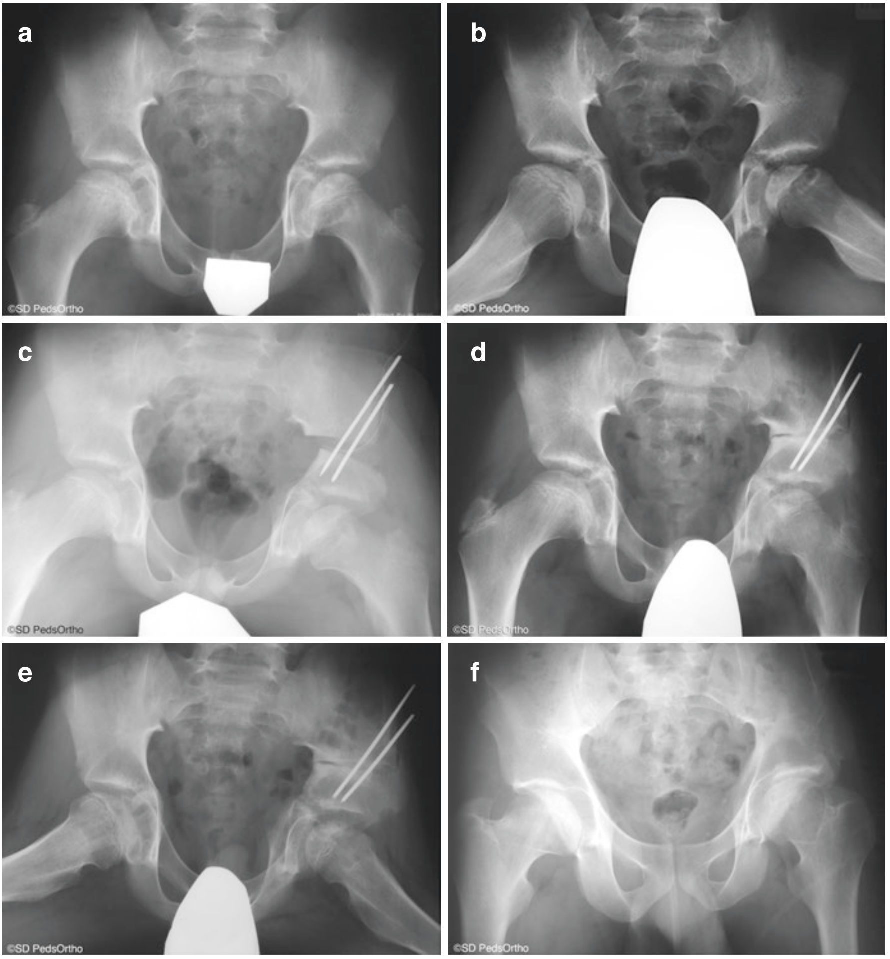


Fig. 6.1 Anterior-posterior (**a**) and frog-leg lateral (**b**) radiographs of a 6-year-old boy with left hip pain demonstrate early Perthes disease. Postoperative radiograph (**c**) demonstrates good femoral head coverage after a Salter innominate osteotomy. 3-month postoperative anterior-posterior (**d**) and frog-leg lateral (**e**) radiographs demonstrate appropriate healing of the iliac osteotomy. 8-year postoperative anterior-posterior (**f**) and frog-leg lateral radiographs demonstrate Stulberg II outcome with a small persistent osteochondral fragment visible on the lateral image

restriction of movement and no contracture other than up to 10 degrees of fixed hip flexion. Outcome was considered fair if there was disabling pain, fixed flexion deformity of more than 10 degrees or fixed deformity in any other plane, or limited hip range of motion less than 90 degrees. Radiologic assessment was rated good if there was no deviation more than 2 mm on AP or lateral view, a fair outcome if there was deviation between 2 and 4 mm on AP or lateral view, and poor outcome if there was deviation more than 4 mm. Their major findings were three poor clinical outcomes all in patients older than 8, two osteotomy patients, and one cast patient.

There were good and fair radiographic results in all patients less than age 6, with a slight lean toward better radiographic results for hip spica patients between ages 6 and 8 (30% good results osteotomy vs. 70% good in hip spica patients). Patients in both treatment groups older than age 8 had a 50% chance of poor outcome (Fig. 6.2). They found head-at-risk signs of lateral extrusion and lateral calcification to be of value in predicting poor outcomes in both groups. They did not find horizontal epiphysis or Gage's sign to be predictive of poor outcomes. The innominate osteotomy group did have a higher percentage of good outcomes in those patients with head-at-risk signs (43% vs. 25%). Despite these breakdowns showing efficacy of innominate osteotomy, they felt that their indications needed refinement as overall there was only a 38% good outcome in innominate osteotomy group compared to 51% in hip spica.

This same group then followed up their results 9 years later [10] with 27 patients including 11 with further follow-up from the original study. They rated clinical and radiographic outcomes with stricter radiographic criteria than their previous study.

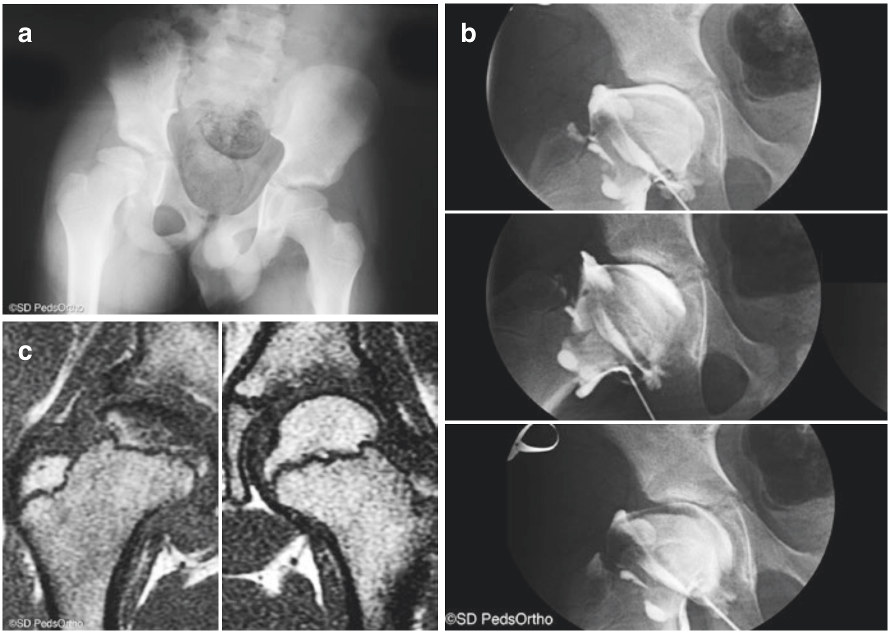


Fig. 6.2 Anterior-posterior (**a**) radiographs of an 8-year-old boy with right hip pain demonstrate early Perthes disease. Intraoperative arthrogram (**b**) demonstrates femoral head flattening with appropriate coverage on the middle abduction-internal rotation view. Pre-operative magnetic resonance images (**c**) demonstrate near complete loss of perfusion to the femoral epiphysis. Post-operative radiograph (**d**) demonstrates good femoral head coverage after a right Salter innominate osteotomy. 3-month post-operative anterior-posterior (**e**) radiograph demonstrate appropriate healing of the iliac osteotomy. 10-month post-operative anterior-posterior and frog-leg lateral (**f**) radiographs demonstrate repeat lateral extrusion. He underwent a right side femoral varus osteotomy and shelf procedure (**g**). 6-month post-operative anterior-posterior radiograph (**h**) demonstrates a well contained femoral head

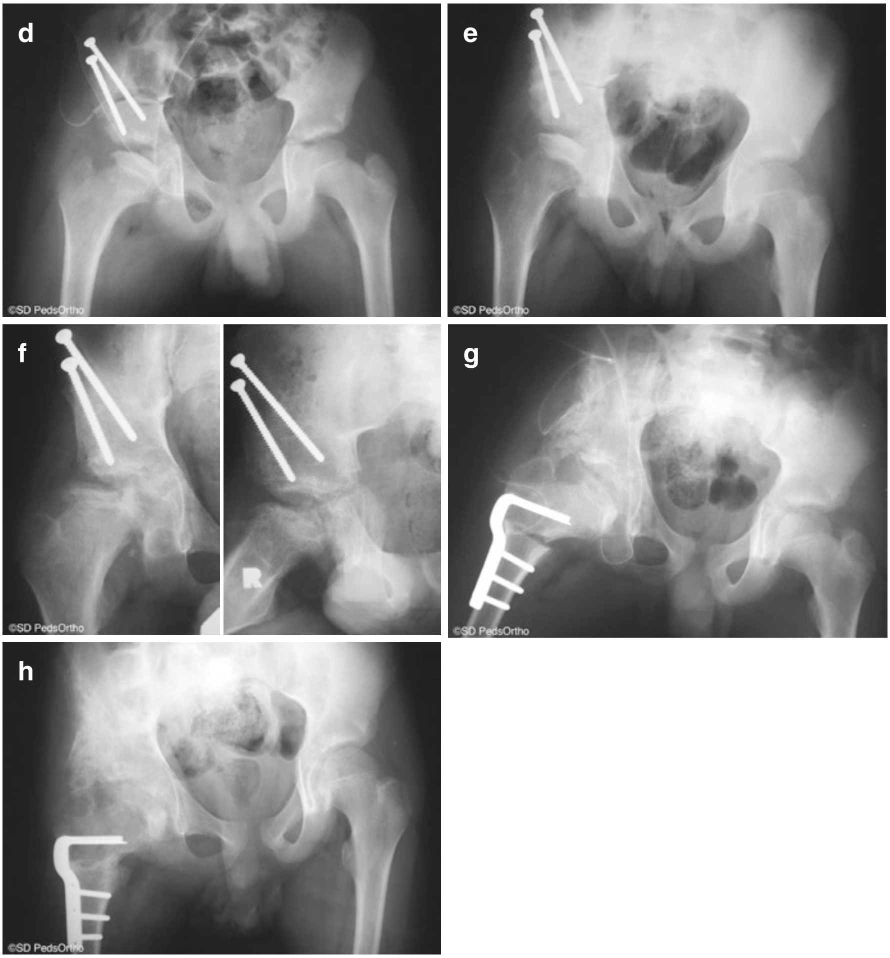


Fig. 6.2 (continued)

They used similar groups as others with good outcomes being spherical head on AP and lateral projections with the same radius as the contralateral hip. Fair outcome consisted of hips with less than 2 mm of deviation from a perfect sphere on either AP or lateral projection and poor if the deviation from a sphere was greater than 2 mm in either plane. Even with these stricter criteria, there were 56% good results and 96% good or fair results with the only poor result in a patient 6–8 years of age at the time of surgery. Importantly they also followed up 11 of the patients mentioned above from the 1982 [9] comparison study. Ten of these 11 patients showed radiographic improvement even with the stricter radiographic criteria.

Stevens, Williams, and Menelaus in 1981 [11] reviewed the results from Royal Children's Hospital, Australia. They treated a total of 300 children for LCP with 85 indicated for Salter osteotomy. They were able to follow up 70 of these patients with at least 2 years of follow-up. Their indications for surgery were older age and near-total head involvement. Average age at surgery was 6.6, and all patients were Catterall 3 or 4. In clinical follow-up 64 of 70 patients or parents were pleased with their result. 30 patients had a limp, 15 occasionally had pain, and 7 patients had hip stiffness necessitating admission for traction. These seven patients included the six who were dissatisfied with their outcome and one patient who was still pleased with their outcome. In review they found these patients either had hip irritability or femoral head deformity preoperatively. They found 20% of the remaining patients were lacking 10 degrees of internal rotation but found that there was no limitation; in fact most patients did not notice this. No patient had required further surgery.

For radiographic review Stevens et al. [11] measured femoral head coverage, head-to-teardrop distance, center-edge angle, and Mose estimates. No single radiographic measure was found to reflect progress or prognosis; therefore they used a modification of the Sundt criteria. This composite impression included good, spherical head well contained with no adaptive change and no increased head-to-teardrop distance, and fair, spherical head with enlargement or adaptive changes in the acetabulum with up to 20% of the head uncovered. A poor radiograph had a flat or irregular head, more than 20% of the head uncovered, an increase in head-to-teardrop distance, or moderate to marked acetabular changes. With these criteria their results were 54% good, 19% fair, and 27% poor. Twelve of the 19 patients in the poor group had minimal or no symptoms and remained active. They noticed trochanteric overgrowth significant enough to compromise an otherwise good result in five patients. Their final conclusion was to perform Salter's innominate osteotomy for a child over the age of 6 with the greater part of the femoral head affected when they begin developing femoral head uncovering. The patient must have almost full hip range of motion without irritability, abduction x-rays demonstrate containment of the head, and the patient must be within 8 months of initial presentation as these earlier osteotomy patients had better results.

Maxted and Jackson in 1985 [12] reviewed 36 children with complete femoral head involvement who underwent Salter innominate osteotomy. The children were aged 3–13 at the time of operation. Younger children (aged 3–6 years) fared better with eight out of nine patients with deformed femoral heads pre-op having circular heads at final healed follow-up. Three out of 10 patients aged 7–13 also saw femoral head shape recovery although clearly a smaller percentage than the younger patients. Thirty-three of 36 patients had a center-edge angle of 25 degrees or more at final healing, and 11 of 14 patients with a break in Shenton's line preoperatively had restoration of Shenton's line postoperative.

Salter's osteotomy was not the only innominate osteotomy being performed for Perthes' disease. In 1972 Canale et al. [13] from The State Hospital for Crippled Children in Elizabethtown, PA published their results with an innominate osteotomy utilizing a quadrangular graft instead of triangular as fashioned by Salter. Their results were compared to a nonoperative group with similar stage of disease and found improved outcomes in the surgical group if the patient was over age 7, the osteotomy was performed in stage 2 or early stage 3 of the disease, and surgery eliminated the need for long-term immobilization. In 2003 Yoon et al. [14]

described a modification of the osteotomy with a steeper angle down to the sciatic cut allowing stable fixation per their report with no need for internal fixation and only splints used postoperatively. They performed their osteotomy in 16 hips and used biodegradable screw fixation in 9 hips. They noted two patients with limited postoperative motion: one blamed on patient refusal to perform postoperative therapy and one with severe hip subluxation. Their mean center-edge angle improved from 19 degrees to 29 degrees with one patient less than 25 degrees. Epiphyseal extrusion also improved from mean 23% to mean 9.5%. They advocate their osteotomy due to no need for hardware removal, although based on more than half of their patients requiring screw fixation anyway this seems a questionable benefit.

All of the above studies hold one thing in common: they do not have a surgical reference or comparison group. Another alternative for surgical management involves femoral varus osteotomy to increase containment. In 1988 Sponseller, Desai, and Millis [15] published results from Boston Children's Hospital comparing patients with LCPD treated with either femoral varus or innominate osteotomy between 1968 and 1982. They begin by acknowledging the advantages and disadvantages which we will highlight here. The advantages of the femoral osteotomy are that the operation is done on the involved bone, provides better lateral coverage, and decreases the force across the joint. The disadvantages for the femoral osteotomy are that it may shorten the limb and may create excessive varus angulation leading to possible weakness of the abductors. The advantages of the innominate osteotomy are that it might compensate for femoral shortening due to femoral head flattening and provide better anterolateral coverage while the disadvantages are the technical difficulty of the procedure and that it might increase the trans-articular pressure. They evaluated the clinical and radiographic outcomes of 42 femoral and 49 innominate osteotomies and attempted to correlate preoperative risk factors for poor outcome. Overall they had ~70% good outcome in both groups and could find no statistically significant difference in clinical outcomes between the two. Radiographically the innominate osteotomy group had a more normal center-edge angle, femoral neck-shaft angle, and level of the greater trochanter when compared to the normal side. In terms of Stulberg and Mose grades however, there was no statistical difference.

Moberg [16] performed a similar comparison in 1997 with 16 femoral and 18 pelvic osteotomies over an average follow-up of 6 years. Their results clinically were similar between the two groups with five patients in each group experiencing occasional hip pain; one patient in the femoral osteotomy group and four in the pelvic osteotomy group walked with a slight limp, although Trendelenburg's sign was positive in three patients with femoral osteotomy and two patients after pelvic osteotomy. Average leg length discrepancy was 0.9 cm in the femoral osteotomy group and 0.8 cm in the pelvic osteotomy group. Radiographic results differed with a significantly better center-edge angle in the group after innominate osteotomy. There was no difference in the Mose circle assessment or the epiphyseal quotient of extrusion. Kitakoji in 2005 [17] also found a more anatomic proximal femur for innominate osteotomy group compared to femoral varus osteotomy group with more normal neck-shaft angle, center-edge angle, and articular-trochanteric distance all more normal in patients who had undergone innominate osteotomy. Clinically they found the pelvic osteotomy resulted in a better scar and for all of these reasons recommended innominate osteotomy as the surgery of choice if needed. At the end of Moberg's [16] discussion, they call for long-term prospective study examining natural history and results of treatment for Perthes disease.

Fortunately, Herring et al. [18] at Texas Scottish Rite had begun a prospective multicenter study on the effect of treatment on outcome. The Legg-Perthes study group had put this together and collected data beginning in 1984 with the report in 2004. Thirty-nine pediatric orthopedists contributed patients to the study, and the design allowed the treating surgeons to pick a treatment regimen and use that on all patients eliminating surgeon bias. The treatment regimens were no-treatment group, range-of-motion treatment including possible Petrie casting and adductor tenotomy, orthosis (Atlanta-Scottish Rite Abduction Orthosis), femoral osteotomy group, and innominate osteotomy group. Provisions were made for very mild Perthes patients to avoid treatment altogether if they maintained motion and remained asymptomatic. Patients initially were compared with Catterall classification on presentation, but eventually the lateral pillar classification was used to categorize disease severity, and Waldenstrom stage was noted as well as demographic factors. The study included patients age 6–12. Ultimately they were able to enroll 19 patients in the no-treatment group, 37 in the range-of-motion group, 129 in the brace group, 52 in the femoral osteotomy group, and 68 in the pelvic osteotomy group. These patients were followed to skeletal maturity and graded based on the Stulberg classification as a reliable index to predict long-term function.

The first results examined are the no-treatment group, the range-of-motion treatment group, and the brace treatment group. Finding no significant difference between these groups, they were combined into a nonoperative treatment group. There was also no statistically significant difference between the femoral and innominate osteotomy group, so these too were combined to increase the power. Comparing operative to nonoperative, there was a statistically significant difference in outcome using Stulberg classification with improved outcomes in the operative compared to the nonoperative hips [19] (61% Stulberg I or II outcomes in operative vs. 46% in nonoperative groups). Most of this difference was found for patients who presented with chronologic age greater than 8 at disease onset in lateral pillar classification B or B/C border. These patients still had overall poor outcomes; however, for example, patients older than 8 in B/C border group 73% in the operative group had Stulberg III outcomes with 60% in the nonoperative group having Stulberg IV or V outcomes. Other than these breakdowns, they found the strongest predictive factor for outcome was lateral pillar classification with all lateral pillar A patients having a Stulberg I or II outcome, 67% of lateral pillar B having the same outcome and only 28% and 13% of B/C border, and lateral pillar C having Stulberg I or II outcome. The second strongest predictor was then age with children presenting on or before the age of 8 having a 59% rate of good results and only 8% rate of poor results, while patients presenting after their 8th birthday had only a 39% rate of good outcomes with a 26% rate of poor outcomes.

The treatment recommendations coming out of Herring's study are closely followed by many. Lateral pillar group A patients tend to have an excellent prognosis and require no specific treatment. Lateral pillar group B patients with onset younger than age 8 also tend to do well not requiring specific treatment. Lateral pillar B/C border patients younger than age 8 do not do as well but did not benefit from surgery in this study. Patients who presented after their 8th birthday with lateral pillar group B or B/C border had significant improvement with either femoral or innominate osteotomy compared with nonoperative treatment. Unfortunately, patients in lateral pillar group C of any age have no evident improvement with surgical treatment and a very poor outcome. This begs more aggressive treatments for these patients.

The other major points made by Herring et al. were in part one of their two-part series [20] outlining the results of the long-term follow-up study. They noted that the Catterall classification suffers from wide variability and poor inter-rater reliability for preoperative grading, while the postoperative radiographic results suffered from a similar poor reliability as well as wide outcomes selection. Their first part paper highlighted lateral pillar classification as a reliable and reproducible option and the Stulberg grade as a reliable radiographic outcome measure.

Shelf/Chiari Pelvic Osteotomy

While Herring and colleagues studied innominate osteotomy and VDRO, other pelvic osteotomies have been proposed over the years to deal with the lateral subluxation in more involved Perthes disease. In 1990 Cahuzac [21] and colleagues reported their findings after Chiari osteotomy on 17 patients with Perthes disease. They advocated the Chiari osteotomy for older patients, with an average age of 8 years and 9 months at operation in their study. Center-edge angle improved from 13.6 to 38.4 and then regressed to 33.2 at follow-up. There was more loss of center-edge angle in patients younger than 8 at follow-up. There was also more loss in patients where the osteotomy site was more than 4 mm above the acetabular margin. When evaluated according to Mose criteria, 12 femoral heads were listed as regular, 4 as irregular, and 1 as very irregular. Again femoral heads that were irregular had a higher osteotomy.

Bennett and colleagues in 1991 [22] also reviewed 17 patients who had undergone 18 Chiari osteotomies for painful subluxated hips with Perthes disease. Of note, primary treatment was containment orthoses, and the mean time between presentation and surgery was 3 years in this study. As such, it is hard to review as a primary treatment option; however, in the setting of salvage, the clinical outcome was reported to be pain-free in the 13 patients reviewed clinically. Near-complete femoral head coverage was achieved radiographically; however, the postoperative motion was reported average 105 degrees flexion, 5 degrees internal rotation, 10 degrees external rotation, and 33 degrees of abduction. No patient was reported to have a femoral head meeting "good" criteria of Mose (less than 2 mm of spherical deviation), meaning no hip was better than Stulberg III.

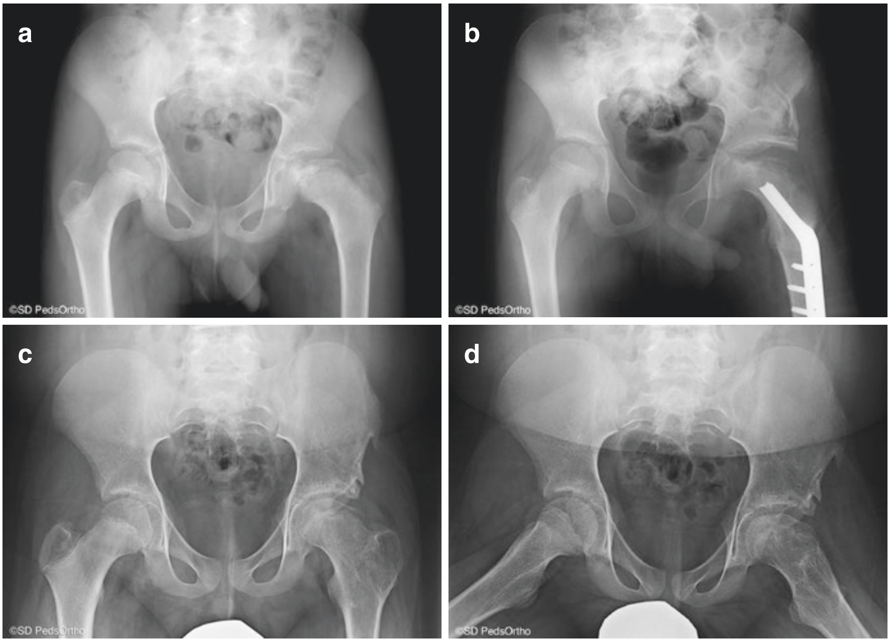


Fig. 6.3 Anterior-posterior radiograph (**a**) of an 11-year-old boy with left hip pain demonstrates late-stage Perthes disease. He underwent a left valgus femoral osteotomy with shelf acetabuloplasty (**b**). 3-year postoperative anterior-posterior (**c**) and frog-leg lateral (**d**) radiographs demonstrate appropriate healing of the osteotomies after removal of deep implants

Catterall as senior author in 1992 [23] highlighted another pelvic osteotomy again indicated for older more involved patients with poor natural history outcomes left untreated. Lateral shelf acetabuloplasty involves creating a trough above the lateral edge of the acetabulum so that strips of corticocancellous graft can be used to extend the acetabulum over the lateral edge providing increased size of the acetabulum (Fig. 6.3). Catterall followed 20 patients after shelf acetabuloplasty for an average of 22 months and compared to a group of 14 patients observed without surgical treatment. They found improved subluxation ratio from 1.36 to 1.19 at nearly 2-year postoperative follow-up for those treated with shelf acetabuloplasty, while the untreated group worsened from 1.36 to 1.66 in follow-up. Acetabular coverage (center-edge angle) also worsened in the nonoperative group, while he observed over coverage in the operative group. The femoral head size relative to the unaffected head was maintained (1.04 pre-op to 1.05 post-op) in the shelf group, while it continued to enlarge (1.09–1.25) in the nonoperative group. They reported no resorption of the shelf, and no patient had postoperative pain, while 45% suffered a reduction in abduction, it was always improved from pre-op. The post-shelf patients had 7 hips that were spherical and congruent, 11 hips aspherically congruent, and 2 hips were aspherical and incongruent. In the nonoperative group, limited abduction persisted in 57% of patients, two patients had minor discomfort, and one patient had moderate pain and underwent femoral osteotomy for hinge abduction.

Three additional patients reported mild disability from their hip. Only one patient had a spherical and congruent hip, seven were aspherically congruent, and six were aspherical and incongruent. The authors certainly demonstrated improvement over nonoperative treatment in late-onset Perthes.

Kuwajima et al. [24] employed a shelf in addition to Salter osteotomy to form an "augmented acetabuloplasty" and reported a comparison of Salter osteotomy alone in 2002. Their patient populations were closely matched in regard to age as this was the preferred treatment at two separate institutions. In older than 6 age group (still of the age range that does well), the augmented acetabuloplasty group showed a trend toward superiority that did not reach statistical significance. Radiographic measurements also portray resorption of the augmentation portion of the graft leading to a large increase in center-edge angle initially and then a large change at longer-term follow-up.

Combined VDRO with Innominate Osteotomy

The need for improved treatment for the older patient with total or near-total head involvement led to what has been termed "advanced containment" to avoid the lateral extrusion that results in a worse Stulberg grade at the end of treatment (Fig. 6.4). For some surgeons this took the form of combined femoral and pelvic osteotomies, while for others this means more control of the final position of the acetabulum provided by a triple pelvic osteotomy.

The initial report of combined femoral and pelvic osteotomies came out of Kansas with Olney and Asher in 1985 [25]. They reported on nine patients with Catterall group III or IV involvement and an average of 3.2 head-at-risk signs and average epiphyseal extrusion of 26% preoperatively. Their clinical outcomes were graded as seven good and two fair, and radiographic outcomes were four good, four fair, and one poor. They advocate this treatment for older children with advanced disease in whom single osteotomy would create too much varus (femoral osteotomy) or likely increase the joint pressure too much (pelvic osteotomy) if full containment were achieved.

Staheli, who had presented on this topic as early as 1980 at meetings, published his results in 1992 [26] on 14 patients with a mean age of 8 years and 4 months. Nine patients were Catterall III, and five were class IV with all patients having three or more head-at-risk signs. Average epiphyseal extrusion pre-op was 30%. Over a mean 8-year follow-up, 11 of 14 patients had a good clinical outcome. Seven hips were Stulberg II with another six Stulberg III and one Stulberg IV at follow-up, and epiphyseal extrusion was <20% in 13 of 14 hips. The authors advocate this treatment for the older child with severe femoral head involvement and head-at-risk signs.

Javid and Wedge in 2009 [27] summarized the effectiveness of the "double osteotomy" nicely. Their study consisted of 20 patients with disease onset over the age of 8 and predicted poor natural history left untreated. Six patients went on to Stulberg II, nine Stulberg III, and five Stulberg IV at a mean follow-up of 5-year and 5 months. Their conclusion based on these outcomes was that the radiographic

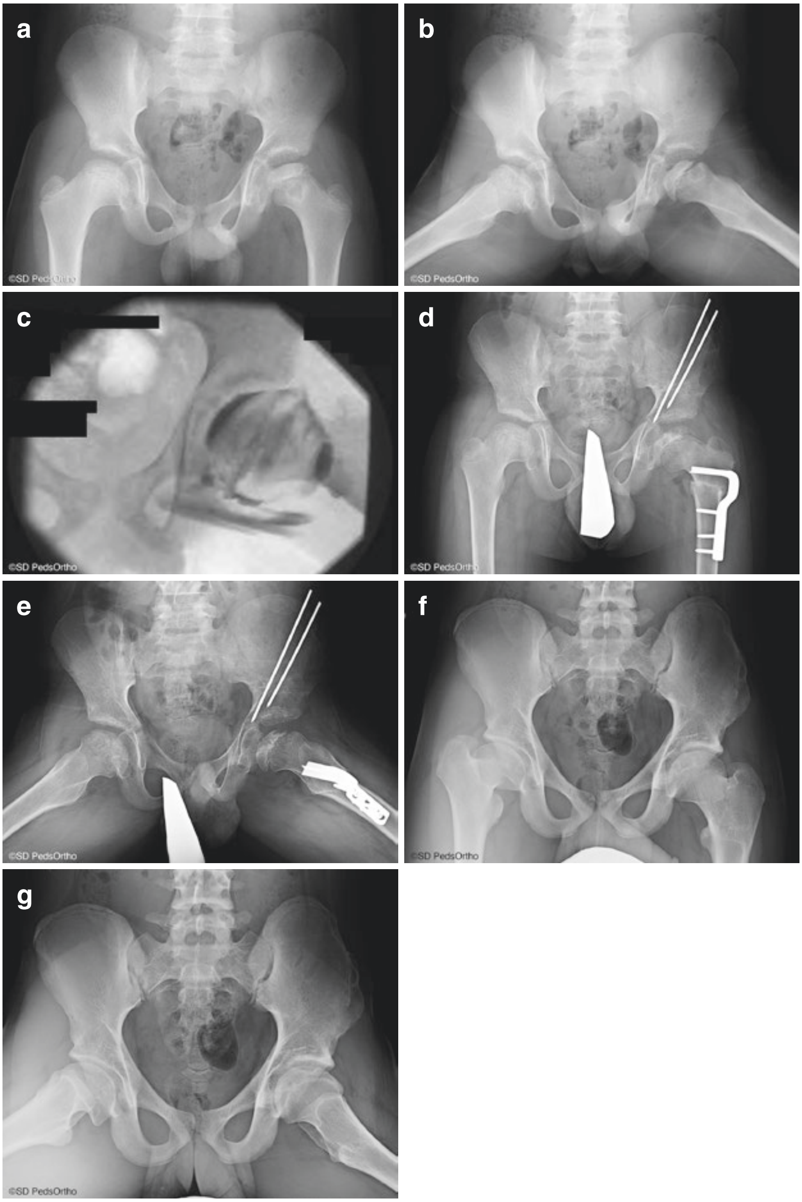


Fig. 6.4 Anterior-posterior (**a**) and frog-lateral (**b**) radiographs of a 9-year-old boy with left hip pain demonstrate early Perthes disease on the left and nearly resolved Perthes disease on the right which was originally diagnosed at age 6. Abduction-internal rotation arthrogram image (**c**) of the left hip demonstrates hinge abduction with impingement of the lateral femoral head and acetabulum. 3-month postoperative anterior-posterior (**d**) and frog-lateral (**e**) radiographs demonstrate appropriate healing of the Salter innominate and femoral varus derotation osteotomies. 5-year postoperative anterior-posterior (**f**) and frog-lateral (**g**) radiographs demonstrate a concentric Stulberg II left hip with coxa magna and breva deformity to the right hip after nonoperative management

outcome in comparison with the natural history in lateral pillar B/C border and C could be improved by converting likely poor results into fair results. This conclusion highlights the difficult patient group of the older Perthes patient.

Triple Innominate Osteotomy

Advocates of the triple pelvic osteotomy often cite computer modeling performed by Rab over time evaluating the containment achieved by osteotomies during positions of daily living as well as modeling of the femoral head changes produced using finite-element modeling. Rab found in 1981 [28] that attempting containment with proximal femoral varus or innominate osteotomy would improve lateral and anterior coverage of the femoral head with more anterior coverage provided by the innominate osteotomy. The epiphyseal compression forces were the same for normal, innominate osteotomy and femoral osteotomy, and there was an increase in medial shear force after femoral osteotomy up to two times body weight. In a finite-element analysis in 1982 [29], stress patterns in LCPD were analyzed demonstrating stress shielding for small necrotic defects (Catterall I or II) but collapse with larger necrotic defects perhaps explaining the relative success of younger patients as a larger portion of their femoral head remains cartilaginous, although they do no rule out remodeling potential.

In 2005 Rab [30] then modeled subluxation in early Perthes disease using a rigid body-spring method. He found that subluxation followed the direction of the necrotic defect mostly anterior. This subluxation began with a small necrotic segment simulating Catterall II changes. The subluxation was mostly anterior with very little lateral subluxation which they justified because the model of the defect was built that way and any lateral subluxation seen on exam may be related to differential loading or parallax on radiographic exam. In extremes of surgical correction (<110 femoral neck varus or 10 degrees or more of acetabular retroversion), the translational force converted to a medializing force.

Clinically providers were noticing extreme varus angulation would be required for adequate containment in more severe Perthes with a varus osteotomy. This provided concerns about limb length and limp. Similar extremes would be needed for acetabular reorientation, and that is not possible with the confines of the Salter osteotomy which led providers to triple pelvic osteotomy. In a report from 2002, Kumar et al. [31] reviewed 21 patients at an average age of 7 years and 7 months. Seventeen hips were Herring group C, and all of them had head-at-risk signs. The average follow-up was 2.5 years. No Stulberg grading was given, but acetabular head index improvement averaged 18%, and center-edge angle improved on average 22 degrees with the authors advocating more improvement with one procedure compared to combination surgeries.

In 2003 O’Connor et al. [32] reported a triple pelvic osteotomy performed through a single anterolateral skin incision. They followed this with a report on eight single incision triple osteotomies [33] over 6 years, although at follow-up none of the patients had reached skeletal maturity. They reported improved center-edge angles and advocated for triple osteotomy in this older (mostly 8- to 12-year olds with one 6-year old) and all with lateral pillar C classification.

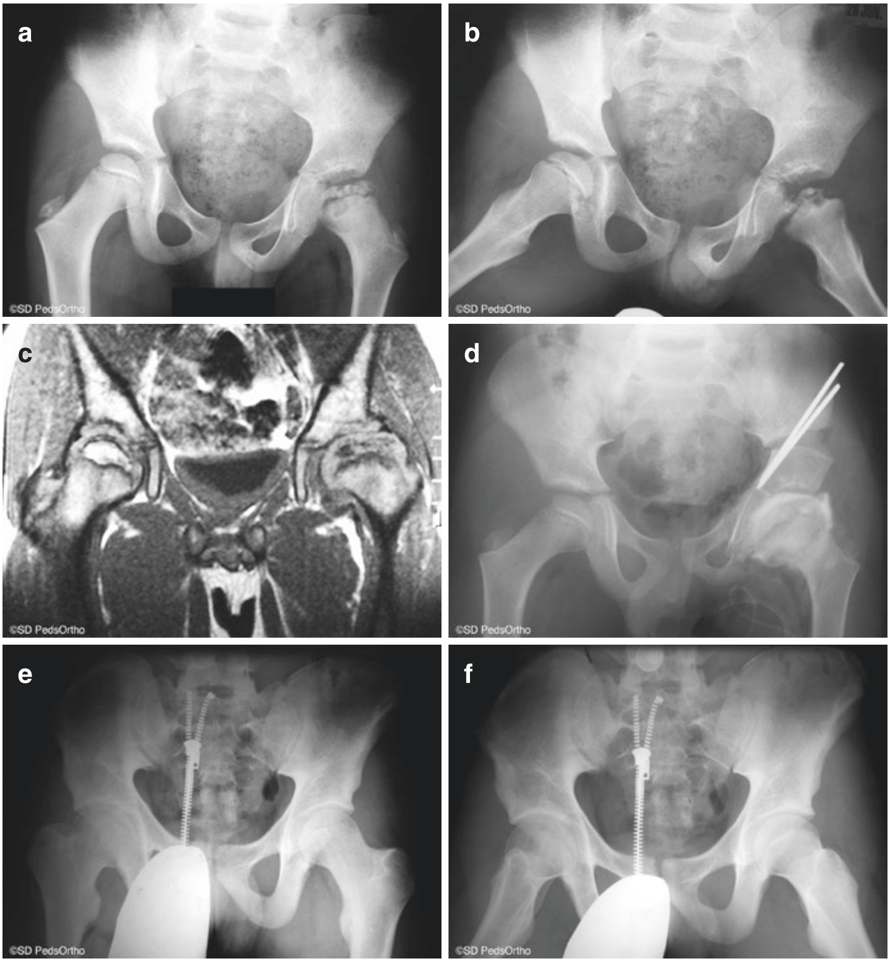


Fig. 6.5 Anterior-posterior (**a**) and frog-leg lateral (**b**) radiographs of a 6-year-old boy with left hip pain demonstrate late-stage severe Perthes disease. Preoperative MR image (**c**) demonstrates severe head involvement and flattening. Postoperative radiograph (**d**) demonstrates good femoral head coverage after a triple innominate osteotomy. 7-year postoperative anterior-posterior (**e**) and frog-leg lateral (**f**) radiographs demonstrate Stulberg I outcome

In another series of articles, Wenger et al. reported on their results first in 2010 [34] on 39 patients with a minimum of 3 years of follow-up (Fig. 6.5). Mean age at diagnosis was 7.5 years and mean age at surgery 8.3 years; their study included 21 hips lateral pillar B and 19 hips that were lateral pillar C. Clinically 35/40 hips were pain-free at follow-up improved from 40/40 with pain pre-op. 36 hips did not require additional surgery, and of these hips, 15 had a good outcome (Stulberg I or II), 17 had a fair outcome (Stulberg III), and 4 had a poor outcome (Stulberg IV or V). In addition to these patients, four underwent further surgery and were not evaluated for Stulberg outcome. None of the poor outcomes started as lateral pillar B, and patients over the age of 8 with lateral pillar C had a 50% chance of a poor outcome, reflecting other studies for this older age group. Center-edge angle increased from 25 to 42 on average over the cohort, and extrusion index decreased from 0.21 to 0.08 over the entire cohort.

The four patients in Wenger's series who required reoperation were older at diagnosis (average 9.5 years). One patient was still under covered and underwent shelf augmentation with varus femoral osteotomy, one patient had impingement and underwent cheilectomy, valgus femoral osteotomy, and shelf, and two patients had hinge abduction and underwent procedures to address the femoral head defects and subsequent knee alignment encouraging more knee varus.

In a follow-up study in 2012 [35], they wanted to assess changes in lateral coverage over time and followed 19 patients with 20 triple osteotomies who had sequential radiographs with blinded observers making readings including extrusion index, center-edge angle, acetabular roof angle, and Sharp's angle. They noted over minimum 3 years that all radiographic parameters changed with mean center-edge angle decreasing 6 degrees from 44 to 38, acetabular roof angle increased from −8 to −1, and Sharp's angle increased from 27 to 32 degrees. They found that no patient corrected to center-edge angle of 44 or less or an acetabular roof angle of −6 or higher (less coverage) ended with pincer morphology at minimum 3 years of follow-up. They did note that all 20 patients achieved the first goal of containment with the triple pelvic osteotomy, but 9 of the 20 had persistent pincer impingement at follow-up highlighting the freedom created by the three-part osteotomy and the double-edged sword this creates.

In the most recent paper out of his group, Wenger as senior author in 2015 [36] wanted to evaluate whether patients treated in the later stage after femoral head extrusion based on the modified Elizabethtown classification or the older (>8 years of age) lateral pillar classification C patients had any benefit from containment with triple osteotomy. They reviewed 54 patients with 56 hips who underwent triple pelvic osteotomy. The patients first undergo arthrogram with exam under anesthesia, adductor tenotomy, and Petrie casting to evaluate for motion and ensure the hip is containable as well as improve abduction prior to the triple osteotomy.

Although the results favor better outcomes for younger patients earlier in the disease process with less affected hips (lower Catterall grade, less involved lateral pillar classification), there were no statistically significant variables when evaluating an outcome of spherical femoral head. As mentioned there were trends: 74% of patients younger than age 8 had a spherical femoral head at final follow-up compared to 52% of patients older than age 8 with $p = 0.085$. Overall results were good with 64% Stulberg I or II, 25% Stulberg III, and six hips Stulberg IV or V. There were two hips with arthritic changes at follow-up with one patient considering total hip arthroplasty. Seven hips demonstrated radiographic findings of over coverage with a Tonnis angle <10 degrees, and multiple hips demonstrated acetabular retroversion based on presence of either positive cross over sign, ischial spine sign, or both signs. Eleven hips required revision during the follow-up period with 7 hips undergoing surgery for symptomatic impingement, 3 revisions for continued subluxation (Fig. 6.6) and 1 revision for a pubic nonunion. Given the ability to perform

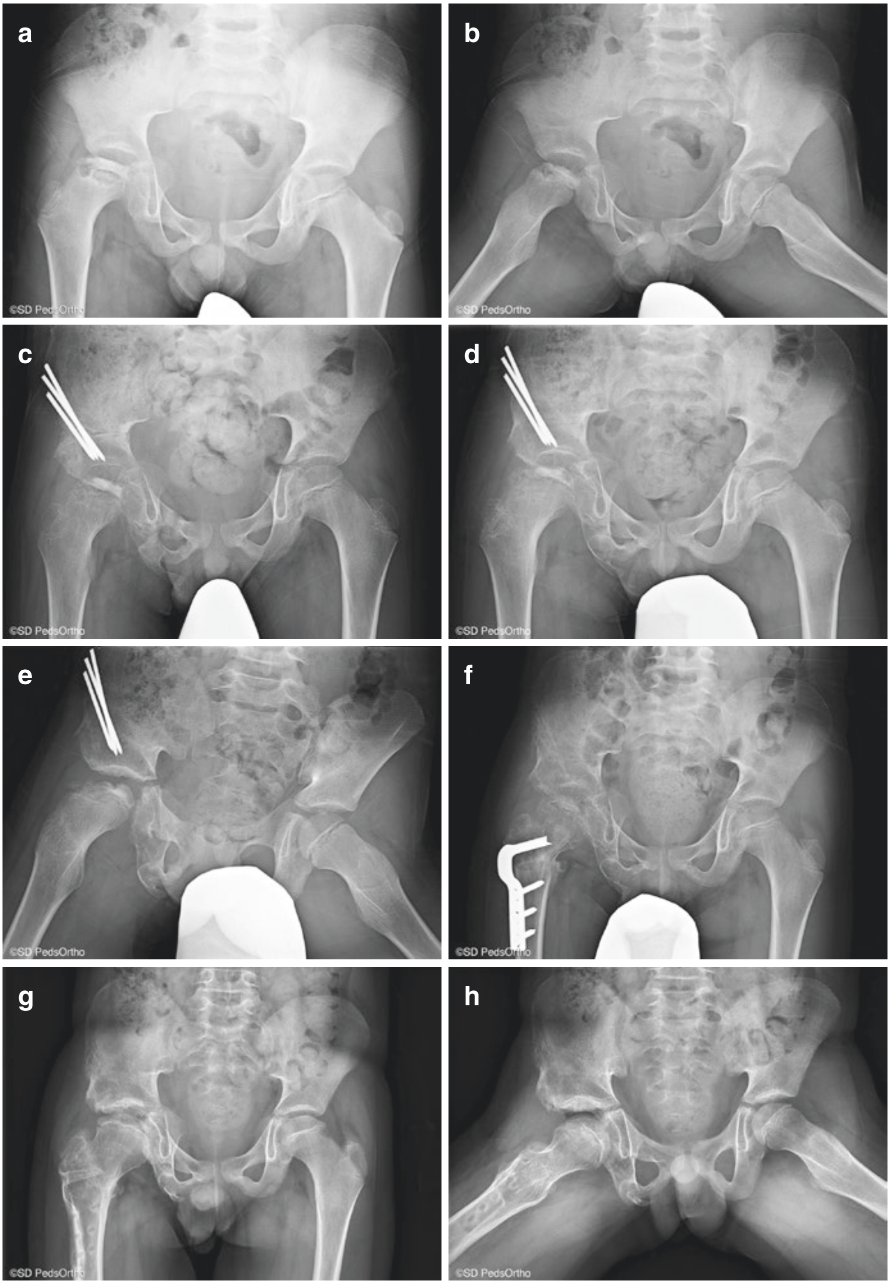


Fig. 6.6 Anterior-posterior (**a**) and frog-lateral (**b**) radiographs of a 7-year-old boy with right hip pain demonstrate severe Perthes disease with metaphyseal cysts. He recovered well from a triple innominate osteotomy (**c**). 8-month postoperative anterior-posterior (**d**) and frog-lateral (**e**) radiographs demonstrate repeat lateral extrusion. He underwent a right-side femoral varus osteotomy (**f**). 1-year postoperative anterior-posterior (**g**) and frog-lateral radiographs (**h**) demonstrate a well contained and concentric hip joint

"rim-trim" surgery to relieve this excessive impingement, the authors advocate triple innominate osteotomy as a good option for the more difficult Perthes patients: older, later in disease process, more involved femoral head as a more advanced containment method. Other options in this older patient population could also be a varus osteotomy in addition to the triple pelvic osteotomy or a Ganz type periacetabular osteotomy (Fig. 6.7).

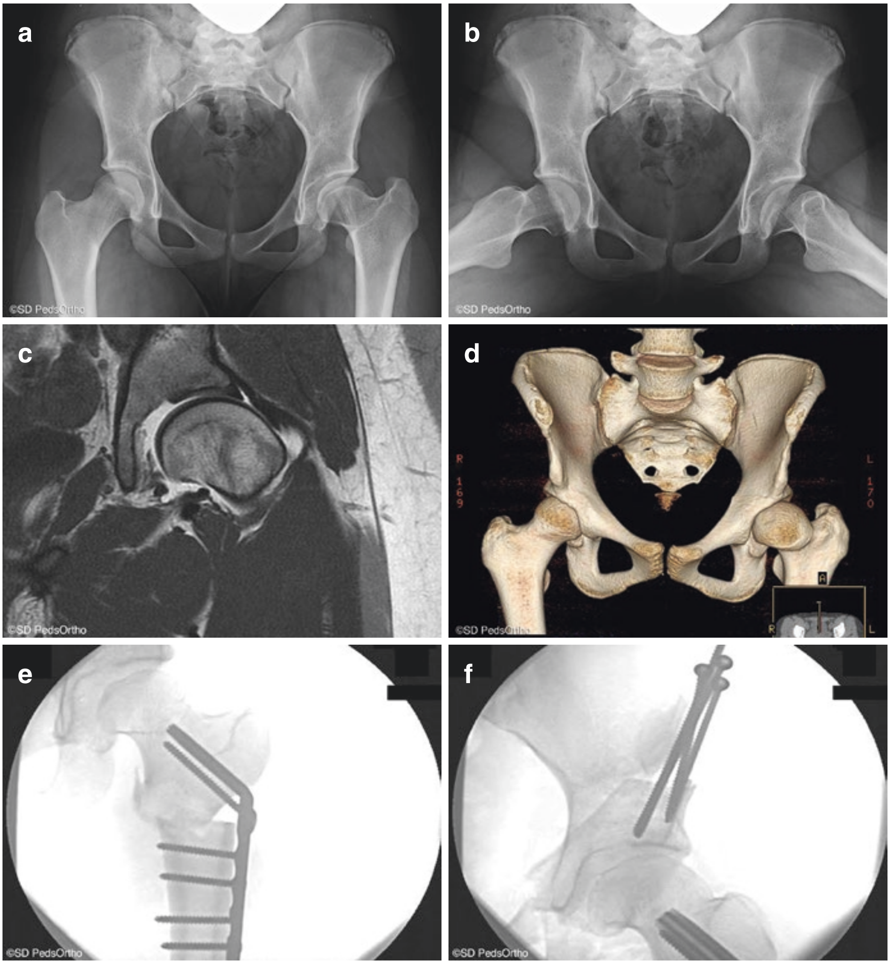


Fig. 6.7 Anterior-posterior (**a**) and frog-lateral (**b**) radiographs of a 15-year-old female with healed Perthes disease and resulting deformity including coxa magna, breva, and acetabular deficiency. A coronal MR image (**c**) demonstrates an intact labrum with decreased femoral head-neck offset and intact acetabular cartilage. The 3D CT image (**d**) demonstrates a femoral head-neck cam lesion and anterolateral acetabular undercoverage. Intraoperative fluoroscopy images were obtained during a femoral valgus osteotomy (**e**), periacetabular osteotomy, (**f**) and femoral head-neck osteochondroplasty (**g**). 2-year postoperative anterior-posterior (**h**) and frog-lateral (**i**) radiographs demonstrate appropriate healing of the femoral and pelvic osteotomies

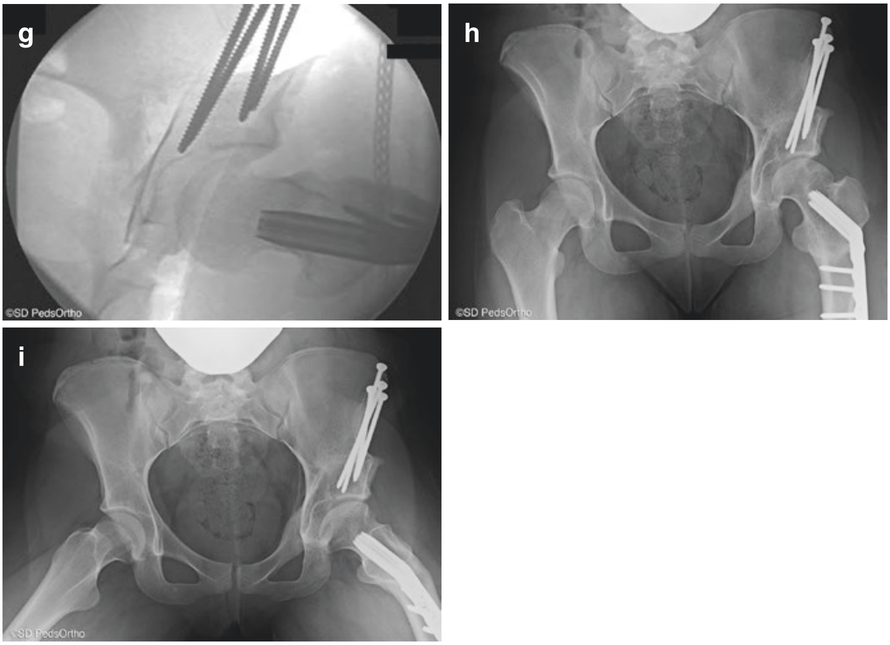


Fig. 6.7 (continued)

Conclusion

Work continues to further the treatment of Perthes disease. Young children with less involved femoral heads continue to have a good outcome, although monitoring and maintenance of motion are certainly required. For the younger patient with more femoral head involvement or the older patient with relatively mild disease, then simple containment with either innominate osteotomy as described by Salter or proximal femoral varus osteotomy can provide containment to allow the acetabulum to serve as a mold for remodeling of the newly formed bone to a hopefully spherical shape for best long-term outcome. Early studies showed older patients continued to have poor outcomes even with surgical interventions, and so surgeons keep searching for options to improve outcomes for these patients. Combining Salter with a varus derotational osteotomy as well as triple pelvic osteotomy is popular method of advanced containment. While the outcomes after these procedures are not perfect, they help improve on the natural history for these older children with more advanced disease with historically poor long-term outcomes.

References

1. Salter RB. Legg-Perthes disease: the scientific basis for the methods of treatment and their indications. Clin Orthop Relat Res. 1980;150:8–11.
2. Babyn PS, Kim HKW, Gahunia HK, Lemaire C, Salter RB, Fornasier V, et al. MRI of the cartilaginous epiphysis of the femoral head in the piglet hip after ischemic damage. J Magn Reson Imaging. 1998;8(3):717–23.
3. Kim HK, Su PH, Qiu YS. Histopathologic changes in growth-plate cartilage following ischemic necrosis of the capital femoral epiphysis. An experimental investigation in immature pigs. J Bone Joint Surg Am. 2001;83–A(5):688–97.
4. Salter RB. Innominate osteotomy in the treatment of congenital dislocation and subluxation of the hip. J Bone Joint Surg Br. 1961;43–B(3):518–39.
5. Salter RB. Experimental and clinical aspects of Perthes' disease. J Bone Joint Surg Br. 1966;48B:393–4.
6. Catterall A. The natural history of Perthes' disease. J Bone Joint Surg Br. 1971;53–B(1):37–53.
7. Mose K. Methods of measuring in Legg-Calvé-Perthes disease with special regard to the prognosis. Clin Orthop Relat Res [Internet]. 1980;(150):103–9. Available from: http://www.ncbi.nlm.nih.gov/pubmed/7428206.
8. Barer M. Role of innominate osteotomy in the treatment of children with Legg–Perthes disease. Clin Orthop Relat Res. 1978;8(135):2–9.
9. Ingman AM, Paterson DC, Sutherland AD. A comparison between innominate osteotomy and hip spica in the treatment of Legg-Perthes' disease. Clin Orthop Relat Res. 1982;163(March):141–7.
10. Paterson DC, Leitch JM, Foster BK. Results of innominate osteotomy in the treatment of Legg-Calvé-Perthes disease. Clin Orthop Relat Res. 1991;266:96–103.
11. Stevens PM, Williams P, Menelaus M. Innominate osteotomy for Perthes' disease. J Pediatr Orthop. 1981;1(1):47–54.
12. Maxted M, Jackson R. Innominate osteotomy in Perthes' disease: A Radiological Survey of Results. J Bone Joint Surg. 1985;67–B(3):399–401.
13. Canale ST, D'Anca AF, Cotler JM, Snedden HE. Innominate osteotomy in Legg-Calvé-Perthes disease. J Bone Joint Surg Am. 1972;54(1):25–40.
14. Yoon TR, Rowe SM, Chung JY, Song EK, Mulyadi D, Anwar IB. A new innominate osteotomy in Perthes' disease. J Pediatr Orthop [Internet]. 2003;23(3):363–7. Available from: http://content.wkhealth.com/linkback/openurl?sid=WKPTLP:landingpage&an=01241398-200305000-00016.
15. Sponseller PD, Desai SS, Millis MB. Comparison of femoral and innominate osteotomies for the treatment of Legg-Calve-Perthes disease. J Bone Joint Surg. 1988;70–A(September):1131–9.
16. Moberg A, Hansson G, Kaniklides C. Results after femoral and innominate osteotomy in Legg-Calvé-Perthes disease. Clin Orthop Relat Res. 1997;334:257–64.
17. Kitakoji T, Hattori T, Kitoh H, Katoh M, Ishiguro N. Which is a better method for Perthes' disease: femoral varus or Salter osteotomy? Clin Orthop Relat Res [Internet]. 2005;(430):163–70. Available from: http://ovidsp.ovid.com/ovidweb.cgi?T=JS&PAGE=reference&D=med5&NEWS=N&AN=15662319.
18. Herring JA, Kim HT, Browne R. Legg-Calve-Perthes disease part II: prospective multicenter study of the effect of treatment on outcome. J Bone Joint Surg. 2004;86(10):2121–34.
19. Stulberg SD, Cooperman DR, Wallensten R. The natural history of Legg-Calve-Perthes disease. J Bone Joint Surg. 1981;63(7):1095–108.
20. Herring JA, Kim HT, Browne R. Legg-Calvé-Perthes disease part I: classification of radiographs with use of the modified lateral pillar and stulberg classifications. J Bone Joint Surg [Internet]. 2004;86–A(10):2103–20. Available from: http://linkinghub.elsevier.com/retrieve/pii/S0268089004000659.
21. Cahuzac JP, Onimus M, Trottmann F, Clement JL, Laurain JM, Lebarbier P. Chiari pelvic osteotomy in Perthes disease. J Pediatr Orthop. 1990;10(2):163–6.

22. Bennett JT, Mazurek RT, Cash JD. Chiari's osteotomy in the treatment of Perthes' disease. J Bone Joint Surg Br. 1991;73(2):225–8.
23. Willett K, Hudson I, Catterall A. Lateral shelf acetabuloplasty: an operation for older children with Perthes' disease. J Pediatr Orthop. 1992;12(5):563–8.
24. Kuwajima SS, Crawford AH, Ishida A, Roy DR, Filho JL, Milani C. Comparison between Salter's innominate osteotomy and augmented acetabuloplasty in the treatment of patients with severe Legg-Calve-Perthes disease. Analysis of 90 hips with special reference to roentgenographic sphericity and coverage of the femoral head. J Pediatr Orthop B. 2002;11(1):15–28.
25. Olney BW, Asher MA. Combined innominate and femoral osteotomy for the treatment of severe Legg-Calvé-Perthes disease. J Pediatr Orthop. 1985;5(6):645–51.
26. Crutcher JP, Staheli LT. Combined osteotomy as a salvage procedure for severe Legg-Calvé-Perthes disease. J Pediatr Orthop. 1992;12(2):151–6.
27. Javid M, Wedge JH. Radiographic results of combined Salter innominate and femoral osteotomy in Legg-Calve-Perthes disease in older children. J Child Orthop. 2009;3(3):229–34.
28. Rab GT. Containment of the hip: a theoretical comparison of osteotomies. Clin Orthop Relat Res. 1981;154(154):191–6.
29. Rab GT, DeNatale JS, Herrmann LR. Three-dimensional finite element analysis of Legg-Calve-Perthes disease. J Pediatr Orthop. 1982;2(1):39–44.
30. Rab GT. Theoretical study of subluxation in early Legg-Calvé-Perthes disease. J Pediatr Orthop. 2005;25(6):728–33.
31. Kumar D, Bache CE, O'Hara JN. Interlocking triple pelvic osteotomy in severe Legg-Calvé-Perthes disease. J Pediatr Orthop. 2002;22(4):464–70.
32. O'Connor PA, Mulhall KJ, Kearns SR, Sheehan E, McCormack D, Mullhall KJ. Triple pelvic osteotomy in Legg-Calvé-Perthes disease using a single anterolateral incision. J Pediatr Orthop B. 2003;12(6):387–9.
33. Conroy E, Sheehan E, O'Connor P, Connolly P, McCormack D. Triple pelvic osteotomy in Legg-Calve-Perthes disease using a single anterolateral incision: a 4-year review. J Pediatr Orthop B. 2010;19:323–6.
34. Wenger DR, Pring ME, Hosalkar HS, Caltoum CB, Lalonde FD, Bastrom TP. Advanced containment methods for Legg-Calvé-Perthes disease: results of triple pelvic osteotomy. J Pediatr Orthop. 2010;30(8):749–57.
35. Hosalkar H, Munhoz Da Cunha AL, Baldwin K, Ziebarth K, Wenger DR. Triple innominate osteotomy for Legg-Calve-Perthes disease in children: does the lateral coverage change with time? Clin Orthop Relat Res. 2012;470(9):2402–10.
36. Stepanovich M, Upasani VV, Bomar JD, Wenger DR. Advanced containment with triple innominate osteotomy in Legg-Calve-Perthes disease. J Pediatr Orthop. 2015;37:1.

Treatment of Legg-Calvé-Perthes Disease of the Hip by Joint Distraction

7

Dror Paley

Introduction

Management of Perthes disease remains controversial despite extensive literature exploring this subject. Obtaining and maintaining hip range of motion are the only principles of treatment that are universally agreed upon. Containment of the femoral head within the acetabulum is thought to have a beneficial role, especially in patients with more than 50% femoral head involvement [1]. Methods used to achieve containment include abduction bracing [2], femoral [3, 4] or innominate osteotomies [5], and shelf procedures [6, 7]. However, these methods are contraindicated when the degree of femoral head collapse and deformation prevent spherical hip motion [3]. Unloading of the hip was originally considered important in the treatment of Perthes disease [8]. Various methods, such as complete bed rest [8] and use of a Snyder sling [9], have been tried toward this end, but little evidence exists to show that these methods alter the natural history of the disease [10, 11]. The failure of unloading may be related to the misconception that non-weight bearing is equivalent to unloading. We now know that muscular forces on the non-weight bearing hip can apply one to two times the body weight. To truly remove all compressive forces from the hip, the muscular forces must be neutralized. This can be accomplished by hip joint distraction with an external fixator. Distraction of the hip also can reduce subluxation of the femoral head relative to the acetabulum.

Considering that the cartilage of the femoral head epiphysis actively proliferates into the uncovered and presumably unloaded lateral regions of the extruded femoral head [12], in 1989, I postulated that if the femoral head could be distracted back into the acetabulum, the epiphyseal cartilage might proliferate to fill the gap between the collapsed femoral head and the acetabulum. Furthermore, distraction would stretch out the contracted capsule and muscles around the hip and improved hip range of

---

D. Paley (✉)
Paley Orthopedic and Spine Institute, St. Mary's Medical Center, West Palm Beach, FL, USA
e-mail: dpaley@paleyinstitute.org

D. S. Feldman, D. Paley (eds.), *Legg-Calvé-Perthes Disease*,
https://doi.org/10.1007/978-1-0716-0854-8_7

motion could be expected. Finally, the repair process and neo-osteogenesis of the femoral head could proceed without risking femoral head collapse. Based on this theoretical rationale, I first applied hip joint distraction as a therapeutic approach to Perthes disease in 1989. Although arthrodiatasis of the hip had been used and applied for other pathologies such as chondrolysis [13], it had not been used during the resorption phase of Perthes disease prior to this time to the author's knowledge.

Surgical Procedure (Fig. 7.1 and 7.2)

The patient is positioned supine with no bump under the hip. The pelvis should remain level and not tilted toward one side or another. The entire forequarter of the limb, from midline anterior to midline posterior and from ribs to toes, was prepped and draped free.

- Step 1: *Arthrogram of hip joint.*

  Anteroposterior (AP), AP in 30 ° flexion, AP in maximum abduction, and frog lateral fluoroscopic views are obtained with arthrographic dye in place. These are used to assess degree of medial dye pool, hinge abduction, coxa magna, and maximum flattening of the femoral head in the 30 ° flexed view.
- Step 2: *Percutaneous adductor tenotomy.*

  The hip and knee are flexed and abducted, and the adductor longus tendon is palpated at the groin and percutaneously cut with a number 15 blade. The hip and knee are then extended and abducted, and the gracilis tendon is then palpated and percutaneously cut at the groin.
- Step 3: *Psoas tendon recession.*

  Make a 3–4 cm anterior groin line incision medial to the anterior inferior iliac spine. Feel the femoral artery pulse and stay lateral to it. Identify the medial border of the sartorius muscle and dissect deep and medial to it. The femoral nerve lies on the iliopsoas muscle at its anteromedial border. The nerve is identified and retracted medially. Dissect down the medial side of the iliopsoas muscle and on the undersurface of its medial border find the psoas tendon. Cut the tendon while leaving a continuous muscle bridge of the iliacus muscle.
- Step 4: *Insert a flexion extension axis pin into the femoral head.*

  A horizontal line of the pelvis is marked on the drapes, guided by the image intensifier (line across the top of both iliac crests or bottom of both ischial tuberosities). The affected lower limb is held with the patella forward, knee in extension, and hip in 15 degrees of abduction relative to the horizontal line of the pelvis. With the image intensifier and a wire, mark a line over the diaphysis of the femur and a point over the center of the acetabulum. Draw a line from the center of the acetabulum point, perpendicular to the diaphyseal femoral line. Place the image intensifier into the lateral view. The dye in the hip joint helps identify the circumference of the femoral head. Draw a line representing the equator of the femoral head in the sagittal plane on the lateral aspect of the hip. Insert a 2.5 mm Steinmann pin into the center of the femoral head from the intersection point of the AP line with the lateral line. This pin should be perpendicular to the shaft of

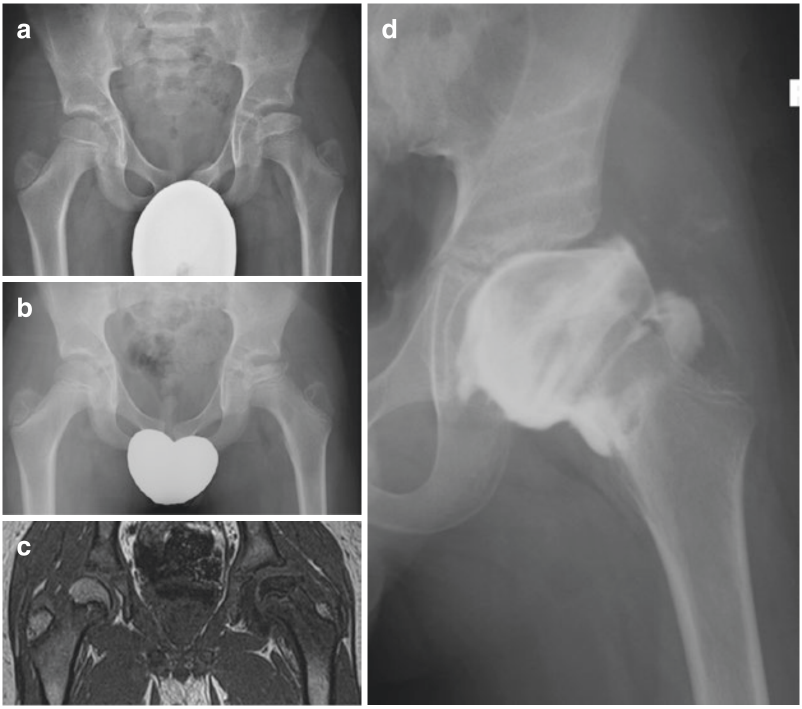


Fig. 7.1 (**a**) An 8-year and 10-month-old boy diagnosed with left-sided Perthes. There is already a slight increase in medial joint space and a small break in Shenton's line. The subchondral fracture can be seen. (**b**) Four months later the femoral head suddenly collapsed with increased pain and limitation of motion. There is more extrusion. (**c**) Pre-op MRI showing increased medial joint space, lateral extrusion and flattening of the femoral head. Note the marrow signal change in the femoral epiphysis compared to the opposite side. (**d**) Arthrogram left femoral head showing the flattening, extrusion, and medial dye pool. (**e**) Fixator placement starts with insertion of a center of rotation Steinmann pin into the femoral head (left). The pin is placed superior to the center of the head so that it ends after distraction in the center of the acetabulum. The unilateral external fixator is mounted on this pin through the cannulation in the hinge (left center). The pelvic pins are inserted at an oblique angle to the pelvis in order to abduct the hip (right center). A wire is inserted first followed by a cannulated drill bit followed by the threaded half pin. The arch is connected to the pins proximally. The hip is distracted acutely and the center of rotation pin removed and reinserted. Note that it lies in the center of the acetabulum and not the center of the femoral head which has been moved distally (right). (**f**) AP radiograph of both femurs and pelvis showing the abducted position of the left femur to the pelvis. There are four multiplanar pins in the pelvis and three in the femur. The rail is parallel to the femur shaft. The hip is overdistracted and Shenton's line is overreduced. The femoral head was moved medially and distally. (**g**) Only 2 months later the femoral head height is already increased and fills the distraction space created. Note the arch on the distal femur used to connect an anterior extension bar between the two arches. (**h**) Arthrogram of hip joint after removal of the external fixator. Note the rounding of the femoral head. (**i**) Femoral head immediately after removal. The lateral pillar is ossified, and the central portion, which has expanded, has some dead bone evident in its center (white sequestrum). (**j**) AP and frog lateral pelvis X-rays 4 years after distraction (age 13 years). The femoral head is now fully ossified. Both femoral head physes are about to close. The femoral head shape can be classified as a Stulberg Class 2. The acetabulum appears to be dysplastic due to the relative coxa magna. The patient has full range of motion of left hip and has no limp. ((C) Dror Paley. Used with Permission)

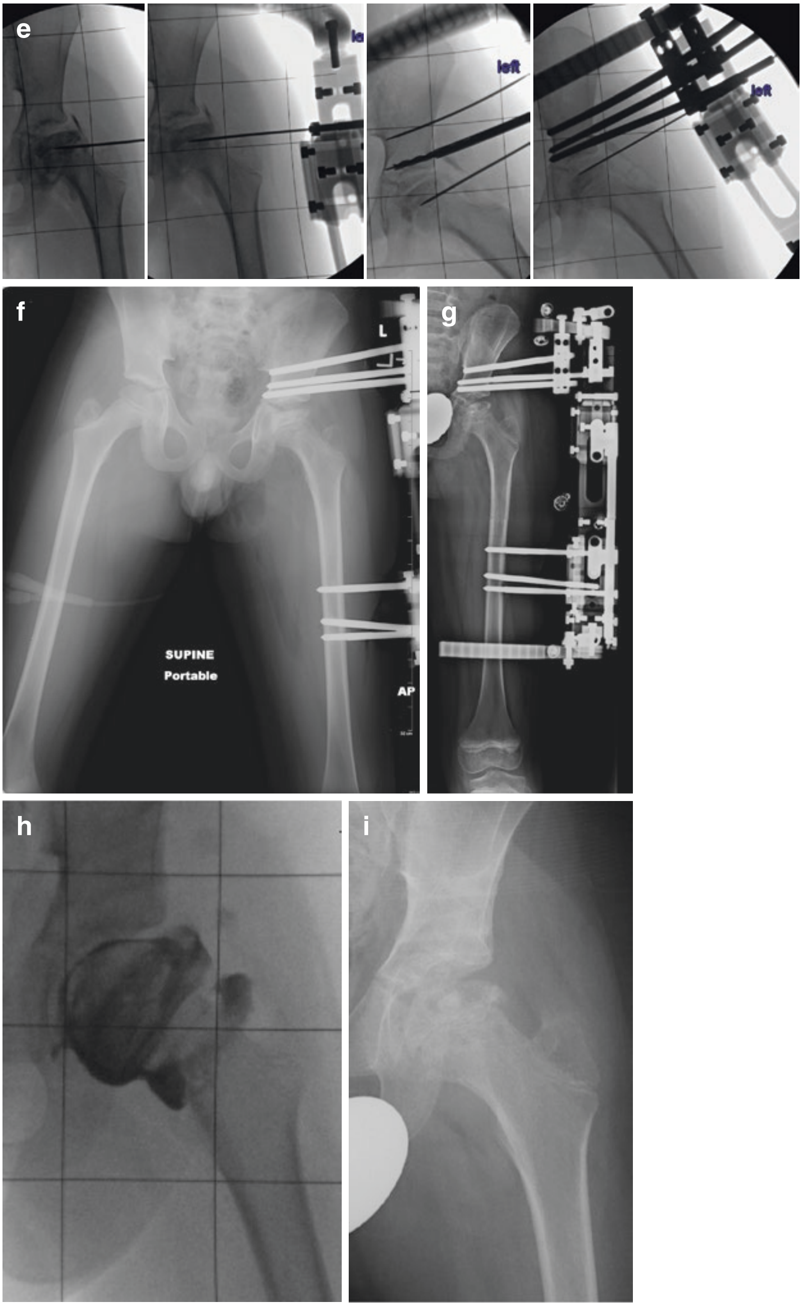


Fig. 7.1 (continued)

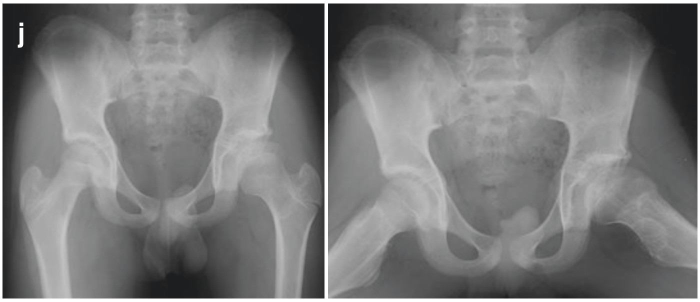


Fig. 7.1 (continued)

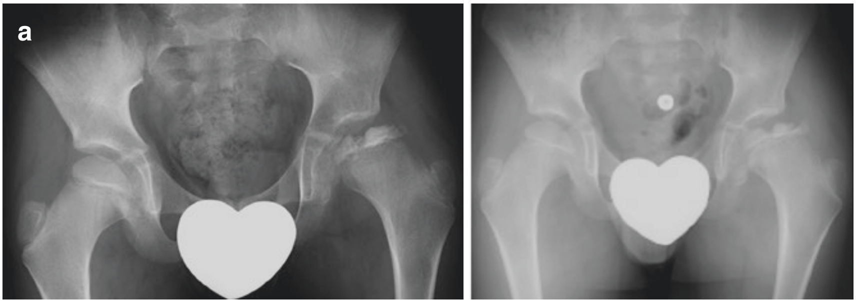


Fig. 7.2 (**a**) A 9-year-old boy with left hip Perthes. There is subluxation (break in Shenton's line and increased medial joint space) and extrusion. The epiphysis is decreased in height. (**b**) Three months later the hip is now fixed in adduction, which rapidly accelerates its deterioration. Hip motion was greatly reduced and pain increased. (**c**) Intraoperative fluoroscopic image showing hip distractor placement. Steinmann pin located at center of acetabulum. The pins in the pelvis are angled to hold the hip abducted. The flattening of the femoral head is outlined by the arthrographic dye. (**d**) Immediate postoperative AP radiographic view following application of hinged external fixation. The fixation of the hip holds it abducted relative to pelvis. Acute distraction of the joint with residual dye in the joint is seen. The femoral head has been shifted medially to eliminate the medial dye pool. There are four pins in the pelvis and four in the femur due to the larger size of the patient. Shenton's line is intentionally overcorrected. (**e**) AP radiograph of the hip 1 month after distraction already shows the rapid resorption of the femoral head. The necrotic bone is more evident due to this resorption. (**f**) Standing AP radiograph of both lower limbs in bilateral hip abduction brace a month after removal of the fixator. (**g**) AP (top) and frog lateral (bottom) radiographs 2 years after distraction (age 11 years). The femoral head is re-ossified. Shenton's line is intact. The femoral head appears enlarged and is likely ellipsoid in shape. The hip range of motion is almost the same as the opposite side with the exception of slight decrease in internal rotation. The patient has not pain or limp. (**h**) AP (left) and frog lateral (right) at age 14. The femoral head is fully ossified. The final femoral head shaped is classified as a Stulberg 2. There is no medial or superior shift. (**i**) Final hip range of motion is symmetric: upper left, abduction; upper right, flexion; lower left, internal rotation; lower right, external rotation. ((C) Dror Paley. Used with Permission)

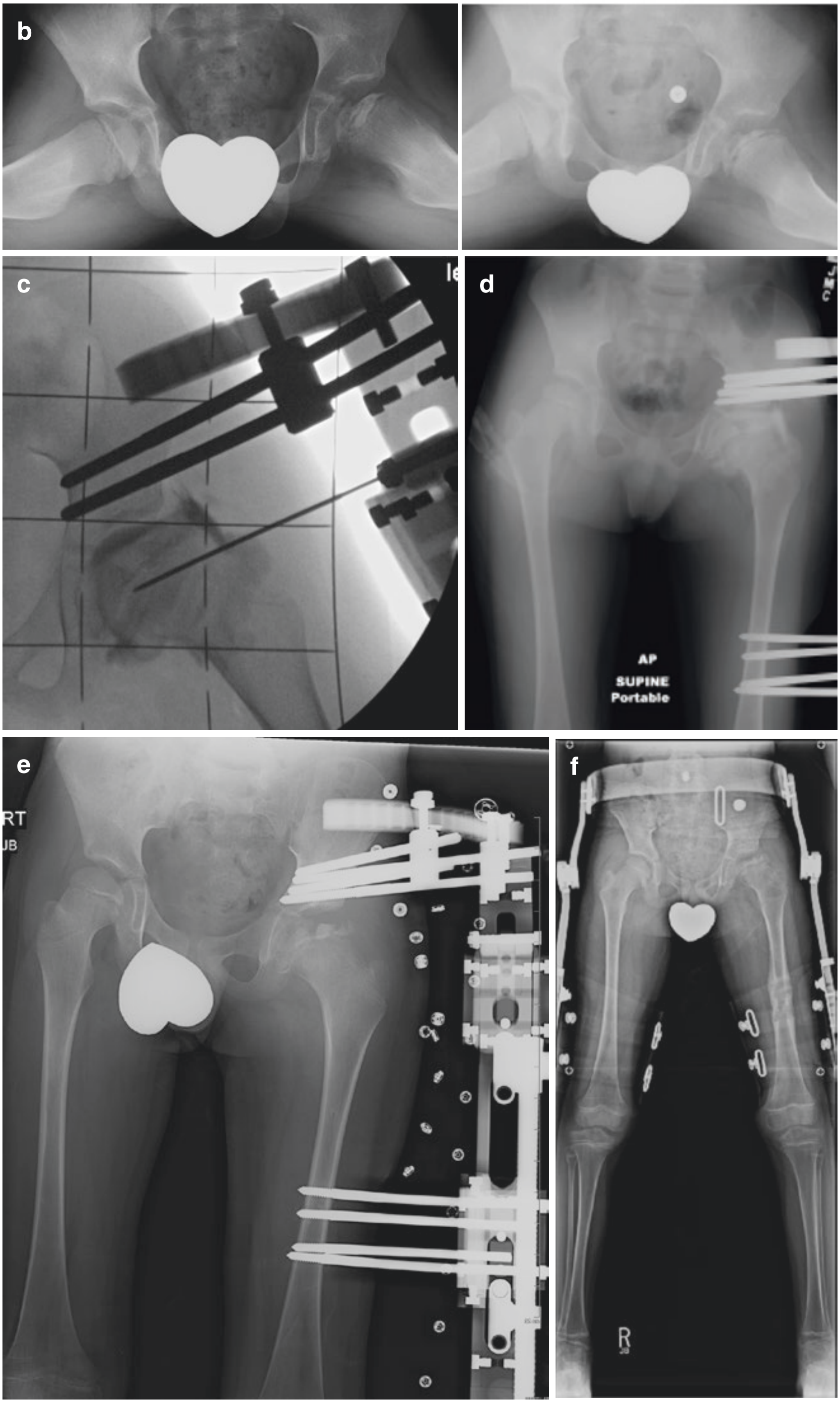


Fig. 7.2 (continued)

Fig. 7.2 (continued)

the femur, end in the center of the acetabulum and be in the midsaggital plane of the femoral head. Because the hip is usually proximally migrated, the center of rotation of the femoral head will be proximal to the center of the acetabulum. The axis pins should be centered on the acetabulum and is therefore more distal to the center of the femoral head.

- Step 5: *Preconstruct a hinged monolateral external fixator*

  Use one of the following: Orthofix, Biomet Zimmer, SN modular rail system (MRS), or Devise Orthopedics Drive Rail, and apply the cannulated hinge over the axis pin (please note that the Biomet Zimmer device was developed by Dr. David Feldman and the SN MRS and the Devise Drive Rail were developed by Dr. Paley). These three systems all offer the ability to distract above or below the hip hinge. The Orthofix device is limited to distracting only distal to the hinge.
- Step 6: *Insert the femoral frontal plane pins.*

  Adjust the distal clamp to the level desired on the femur (usually upper to middle third). Insert three frontal plane pins with the femur kept in the patella forward position. Leave room for lengthening on the distal fixator.
- Step 7: *Insert two pins into the anterolateral pelvis.*

  Roll the operating room table toward the opposite side so that the operative side is higher. Bring the image intensifier into the over the top position to take an oblique AP of the affected hip. Visualize a triangle in the supra-acetabular region. Drill a 1.8 mm wire into this triangle and then tap it in until a hollow sound from hitting a cortex is heard. If the wire is also in the correct alignment on the AP so that it is about 15 ° of abduction to the horizontal line of the pelvis, then advance the wire through the cortex. Use another wire to measure the length of the intraosseous part of the wire. Overdrill the wire with a 4.8 mm cannulated drill bit or in smaller children a 3.2 mm cannulated drill bit. Insert a hydroxyapatite-coated 4.5-mm- or 6-mm-diameter half pin, respectively. Repeat the same for a second pin either more proximal or more distal to the first.
- Step 8: Attach an arch to these first two pins so that the arch is in line with the rest of the fixator based on the constraints of the fixator.

  This arch will usually not be perpendicular to the pelvis due to the 15 ° abduction of the hip joint.
- Step 9: *Add two more pelvic pins.*

  Add one transverse and one oblique pin to the pelvic arch for a total of 4 pins in the pelvis. The transverse pin should be in the supra-acetabular region (most distal pelvic pin). The oblique pin should be between the transverse and the two anterior pins.
- Step 10: Test the hip motion. The hip should move easily in flexion-extension.
- Step 11: *Using the fixator distract the hip joint acutely.*

  Perform an acute distraction of the hip joint so that Shenton’s line is over reduced by up to 1 cm.
- Step 12: *Reduce lateral subluxation of the hip.*

  After distraction the femur is shifted medially in the distal pin clamp by loosening the set screws and pushing in the half pins. This reduces the medial dye pool space. If the femoral head distraction is not apparent perform more distraction after reduction.

- Step 13: Ins*ert a posterolateral pin in the femoral pin clamp.*

  In the MRS and Drive Rail, a posterolateral half pin is added to the AP clamp to achieve multiplanar fixation.
- Step 14: *Add a hip extension bar.*

  Add a removable distraction rod anteriorly between the pelvic arch and the distal femoral clamp. For easier attachment, a distal femoral arch can be added to attach the hip distraction rod more anteriorly.

Postoperative Management of Distraction Treatment for Perthes Disease

Physiotherapy is initiated after surgery, with emphasis on maintaining hip flexion and extension range of motion. The therapist must clearly understand that they are not to work on hip abduction, adduction, external rotation, or internal rotation because this would stress the external fixation pin-bone interface. The patient and therapist are taught to measure the true hip motion at the hip hinge rather than doing so clinically (i.e., between the thigh and the spine) (Fig. 7.3). The patient is taught how to perform flexion-extension exercises at home, supplementing the hour of daily physical therapy. Patients are allowed 50% weight bearing, while the external fixator is in place.

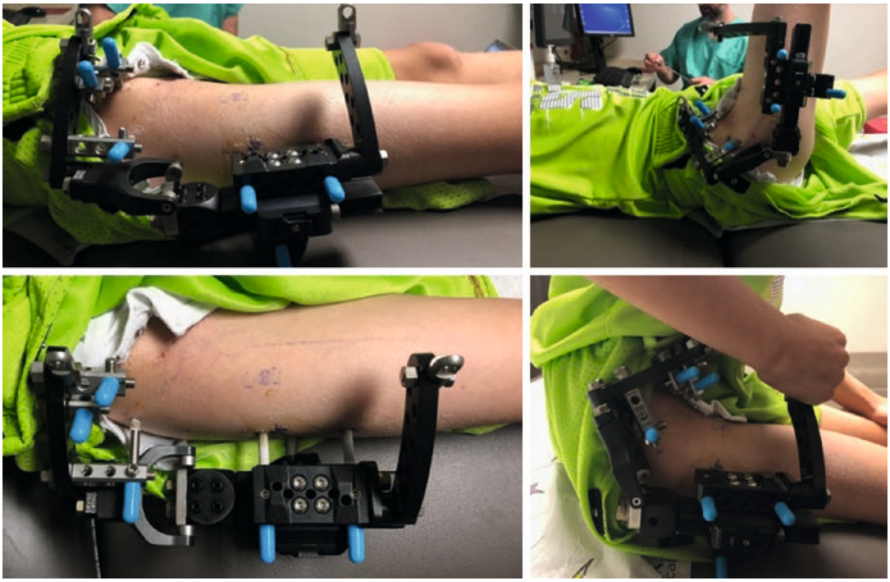


Fig. 7.3 Photographs of external fixator distraction of the hip for Perthes. The example is of a Drive Rail (Devise Orthopedics, Memphis, TN). Upper left, side view supine; lower left, front view supine; upper right, flexion of hip supine with fixator hinge flexion seen; lower right, flexion of hip sitting. ((C) Dror Paley. Used with Permission)

Physiotherapy is important for prevention and treatment of hip flexion contracture. Using the removable hip extension bar prevents this complication. The bar is removed for therapy and inserted at least half the time during the day and all night long. The fixator is left in place for 4 months in patients younger than 12 years and for 5 months in patients 12 years and older. This usually correlates with radiograph re-ossification of the lateral pillar.

Apparatus removal is performed under general anesthesia as an outpatient procedure. Because of the osteoporosis of the femoral head and neck, manipulation of the hip with the patient under anesthesia should not be performed after the removal to avoid fracture of the hip or femur. A bilateral abduction brace (pelvic band with bilateral thigh cuffs and hip hinges) set at 30 degrees of abduction per leg is applied after the removal and is used both day and night for 6 weeks. Resumption of full weight bearing begins on a gradual basis immediately after fixator removal, and full weight bearing without crutches is achieved in approximately 4 weeks after removal. After 6 weeks of full-time use, the abduction brace is used only at night for 6 months (Fig. 7.2f). Running, jumping, and participation in sports are not allowed for 1 year after treatment. Swimming, cycling, and walking are encouraged. The patient is taught a series of five stretches called the Paley Perthes exercises (Fig. 7.4). These should be performed twice daily until skeletal maturity.

Paley Perthes exercises (Fig. 7.4):

1. Wide abduction standing
2. Supine hip flexion
3. Prone internal rotation stretches
4. Prone external rotation stretches
5. Prone hyperextension of the hip

Results

Paley and Segev conducted a retrospective study of the first 16 consecutive patients (18 hips) treated by hip joint distraction between July 1989 and July 1999. Fourteen patients had Perthes disease, and two had avascular necrosis of the hip after slipped capital femoral epiphysis The patient group was comprised of four girls and 12 boys. Two patients had bilateral hip involvement and received the same treatment for both hips. One patient received repeat distraction treatment of the same hip. The mean patient age at the time of disease onset was 9.1 years (range, 6–14 years). The mean patient age at the time of surgery was 10.2 years (range, 6.5–15.6 years). All patients with Perthes disease had whole-head involvement, and the cases were classified as Catterall IV [3, 12–14] or depending on the date of initial presentation to the senior author. The two patients with slipped capital epiphysis experienced collapse of the femoral head resulting from avascular necrosis.

All patients in this series had marked proximal migration and subluxation, which are very poor prognostic factors for containment treatment. The surgical approach and treatment protocol for all patients treated by distraction included gradual

Fig. 7.4 Paley Perthes exercises: patients and parents are taught to do these exercises two to three times daily to obtain and maintain hip mobility. Standing wide abduction (left), maximum flexion (knee to chest) (top center), hip hyperextension (top right), maximum internal rotation (bottom center), maximum external rotation (bottom right). ((C) Dror Paley. Used with Permission)

distraction at half a mm per day until Shenton's line was over-reduced. The external fixator remained in place for 4 months. All the data were collected for clinical documentation in a prospective fashion by the treating author. All patients, while under general anesthesia, underwent intraoperative arthrography of the hip at the time of external fixator application and post-distraction arthrography of the hip at the time of fixator removal. Patients were examined every 6 months for the first 2 years and then annually for the remainder of the study period. Clinical observations were evaluated and recorded by the senior author at each follow-up visit and included subjective pain and activity levels, bilateral hip range of motion (flexion, fixed flexion deformity, abduction, adduction, prone internal and external rotation), knee range of motion, Trendelenburg test, clinical gait assessment, and anteroposterior plus frog leg view pelvic radiographs. The average time from surgery to most recent follow-up visit was 6.7 years (range, 3.5–13.4 years). The clinical evaluations and final follow-up radiographs were tabulated and analyzed.

Based on the total arc of hip range of motion, a clinical sphericity index was calculated to describe how close the hip motion was to being spherical. This index was calculated by dividing the total arc of motion in all three planes of motion (flexion-extension, abduction-adduction, and internal-external rotation) of the diseased hip by 270 degrees, which is the average normal total hip range of motion. The clinical sphericity index is expressed as a percentage of normal total range of motion. The hip was considered to move spherically if the index was greater than two-thirds (67%) of the normal range.

We also calculated the sphericity of the femoral head using measurements derived from pre- and post-distraction arthrograms. The ratios between the largest diameter of the femoral head divided by the lesser diameter (two times the lesser radius, perpendicular to the largest diameter and bisecting it in its middle) on the anteroposterior and lateral view arthrograms were added together and divided by 2 to calculate an index. A normal index for a spherical femoral head is 1 [15]. The closer the index is to 1, the more spherical is the head. The initial and final arthrogram ratios were compared.

The final follow-up radiographs, including those of patients who were not skeletally mature, were graded using the Stulberg [16] classification system. The following radiographic parameters were measured on the preoperative and final radiographs for the operated and normal hips: sharp acetabular angle, central edge angle, proximal migration of Shenton's line, and distance of the medial border of the femoral head from the tear drop. Closure of the proximal femoral physis on the normal side was noted and considered to be evidence of hip skeletal maturity. A premature closure of the diseased hip physis relative to the normal hip also was noted.

Fifteen patients had complained of varying degrees of pain before surgery. At final follow-up, only one patient complained of mild pain that did not require analgesics and did not interfere with daily activities. All patients returned to full school and/or work activities, including sports without limitation. All patients expressed satisfaction with the results and indicated vast improvement in their function compared with their pre-treatment abilities. Fifteen patients walked with a limp before the operation, compared with only one patient who walked with mild lurch gait at final follow-up. Fifteen patients had positive Trendelenburg sign before the operation, compared with only one with positive Trendelenburg sign at final follow-up. All patients had full ipsilateral knee range of motion before surgery and at final follow-up.

All our patients experienced marked limitation of motion on the affected side at presentation. At final follow-up, the mean flexion-extension arc of motion was 100 degrees (range, 90–130 degrees). The mean abduction-adduction arc of motion was 54 degrees (range, 25–75 degrees). The mean internal-external rotation arc of motion was 58 degrees (range, 0–90 degrees). The mean total hip arc of motion was 214 degrees (range, 115–285 degrees). The mean arc of motion for the treated hip was 79% of normal (range, 43–100%). At final follow-up, 16 of 18 hips that underwent hip joint distraction had their range of motion restored to at least two-thirds normal; two hips had a range of motion below functional range.

During distraction, early, rapid osteoporosis of the femoral head was consistently observed, revealing sclerotic dead bone. This was followed by gradual ossification of the lateral pillar, which usually was completed by 4 months. All patients except two underwent external fixator application after femoral head collapse and during the resorption phase. Two patients underwent application of the external fixator just after the initial subchondral fracture. In both of these cases, the femoral head re-collapsed after fixator removal and subsequently went through a resorption phase. One of these patients underwent a second distraction treatment, and complete success was achieved the second time.

At the most recent follow-up visit, nine patients had reached skeletal maturity as judged by closure of the femoral capital epiphysis in the normal hip. Three hips showed signs of premature physeal closure on the operated side. The mean preoperative Sharp acetabular angle was 45 degrees (range, 40–50 degrees) and at final follow-up was 44 degrees (range, 35–50 degrees). The mean preoperative center edge angle was 19 degrees (range, 0–30 degrees) and increased to 24 degrees (range, 15–35 degrees) postoperatively. The difference between pre- and postoperative Sharp acetabular angles was not significant ($P = 0.094$); the increase in the center edge angle after treatment was marginally significant ($P = 0.051$).

The mean proximal migration measured as a break in Shenton's line was 7 mm (range, 0–14 mm) preoperatively and improved to 2 mm (range, 0–12 mm) at the most recent follow-up visit. This difference was statistically significant ($P = 0.002$). The average distance from the medial femoral head to the teardrop was 13 mm preoperatively (range, 8–16 mm) compared with 11 mm (range, 6–18 mm) postoperatively, which was statistically significant ($P = 0.022$). The mean radiographic sphericity index improved from 1.29 (range, 1.1–1.6) at the time of frame application to 1.17 (range, 1.0–1.59) at the time of frame removal, which was statistically significant ($P = 0.001$). The Stulberg [17] classification based on the most recent radiographs was as follows: Class I, one hip; Class II, five hips; Class III, eight hips; and Class IV, four hips.

Complications

Most patients developed minor pin tract infections, which were successfully treated with oral antibiotics. The fixator on one patient had to be removed after only 2 months because of severe pin tract infection. This patient developed recurrent stiffness and subluxation of the hip after the first removal. After the second treatment, the patient was able to maintain a mobile hip with spherical hip motion.

One patient sustained a fractured neck of the femur caused by a fall on the day of fixator removal. The fracture was treated by screw fixation and healed uneventfully.

Two patients each underwent a second application of the fixator for contralateral Perthes disease at 3 years and 3 months and at 1 year and 4 months, respectively, after the index distraction treatment. One patient underwent treatment of Perthes disease shortly after a subchondral fracture of the hip. The course of treatment by distraction was uneventful. However, after fixator removal, the femoral head proceeded to undergo resorption, collapse, and subluxation. Reapplication of the external fixator a year later, during the maximum resorption phase, led to an excellent final result.

As an addendum to this study, I decided to review the radiographs and results of as many patients that could be located in 2009. This represented a 20 year follow-up on the earliest patient. Only 13 of the total hips and 11 of the total patients could be found. All of the Stulberg 4 cases were in the follow-up group. It is interesting to note that all of the Stulberg 4 cases had evidence of degenerative changes, while none of the Stulberg 1, 2 or 3 cases did. Only two of the four Stulberg 4 cases were symptomatic, while the others were not. Femoroacetabular impingement (FAI) was present in all of the Stulberg 3 and 4 cases reviewed. We were unaware of FAI when we first conducted this study. Some of the Stulberg 3 cases were subsequently treated by femoral head reduction osteotomy. The Stulberg grade did not change at final follow-up in 2009. The result grading also did not change since the two painful Stulberg 4 cases were the same symptomatic cases in the original study. It is clear that the four Stulberg 4 cases will all require a hip replacement. It is likely that the Stulberg 3 cases will require some treatment for FAI which could include hip arthroscopy or surgical dislocation of the hip with osteochondroplasty or femoral head reduction osteotomy [17].

Discussion

The natural history of Perthes disease and avascular necrosis of the hip joint is directly related to patient age at time of disease onset and amount of femoral head involvement [18–21]. Older age and whole femoral head involvement are poor prognostic factors [22–25]. Treatment by bed rest, non-weight bearing, and abduction orthosis is of limited value and is not well tolerated [1, 2, 8, 26, 27]. Range-of-motion exercises and various forms of surgical containment have constituted the mainstay of treatment for Perthes disease [28–30] that for children older than 6 years, any method of treatment offers a better prognosis than no treatment. Containment treatment in patients older than 11 years leads to only 40% satisfactory results [12, 14] compared with an overall age-independent success rate of 70–90% [18].

Stiffness, subluxation, and femoral head collapse are considered contraindications to surgical containment treatment. Therefore, the worst cases often are not treatable with containment. Abduction bracing is a nonsurgical containment treatment method. It is fraught with problems of noncompliance, especially in older children, and can lead to hip stiffness unless prescribed in conjunction with aggressive physical therapy [27]. Varus femoral osteotomy can achieve the greatest degree of femoral head containment [3]. The resulting coxa vara deformity may not remodel and therefore may produce a long-term limp due to abductor muscle dysfunction because the abductor lever arm and muscle tension are altered [31]. A pelvic osteotomy alone for containment is more limited in its amount of coverage [32, 33]. All these methods are contraindicated if the hip is stiff, especially if it cannot abduct sufficiently; these hips are suitable for a salvage procedure.

Both varus femoral and pelvic osteotomy methods distort the anatomy and have limited ability to change the shape of an already collapsed femoral head or to reduce subluxation [28].

The distraction we describe is not limited by hip stiffness, degree of femoral head deformity, or subluxation. Although distraction is performed with the hip in 15 degrees of abduction, the primary goal is not containment. The epiphyseal cartilage of the femoral head is not primarily damaged from the loss of circulation to the femoral head. Instead, it reacts by proliferating outside the acetabulum, leading to coxa magna and lateral ossification. The cartilage also proliferates medial to the femoral head when the femoral head has migrated laterally, and superiorly [15] proliferation or ossification is not observed superior to the femoral head, where it is in contact with the acetabulum. Because the femoral head cartilage seems to have the potential to grow in the unstressed regions inside and outside the acetabulum, I postulated that if the femoral head were pulled away from the acetabulum and kept there, the epiphyseal cartilage might proliferate into the acetabulum and fill the space created by the previous collapse. The acetabulum would act as a sort of mold for the femoral head. In many ways, this is similar to the theory behind containment. Pulling the femoral head down also would reduce the apparent subluxation of the hip, especially the break in Shenton's line. In cases in which collapse has not occurred or has not progressed to maximum, dead bone may be resorbed under the protection of the distractor. If the distractor remains in place long enough, new bone formation can replace removed bone, preventing collapse after fixator removal. Herring [14] noted that once the lateral pillar has re-ossified, no further collapse is to be expected. Therefore, we chose re-ossification of the lateral pillar as a satisfactory end point for fixator removal.

The radiographic findings obtained during distraction revealed very rapid progression of osteoporosis of the femoral head and neck. The dead bone could readily be distinguished from the live bone by its white sclerotic appearance; the remainder of the femoral head and neck appeared osteoporotic. At approximately 6–8 weeks after surgery, new ossification of the lateral pillar was observed. The lateral pillar was fully reconstituted by 4 months after initiation of the distraction treatment. In children older than 12 years, this took up to 5 months.

Mose [34] and Stulberg [16] showed that femoral head sphericity and congruency with the acetabulum are directly related to the long-term prognosis. Distraction leads to improved femoral head radiographic sphericity. Our results documented an average sphericity index improvement from 1.29 before treatment to 1.17 at frame removal, indicating increased roundness of the head and improved joint congruency. These findings were corroborated by the clinical range-of-motion results. All our patients experienced improved hip range of motion with distraction treatment. The clinical sphericity index increased, on average, to 79% at last follow-up. If we can assume that when something moves like a sphere, it must be shaped like a sphere, it can be said that most of these hips demonstrated spherical three-dimensional motion.

We also observed that distraction did not change the shape of the acetabulum, as evidenced by the lack of change in Sharp angle. The position of the femoral head in the acetabulum, as judged by the center head angle, did change. In 12 of 18 cases, sustained reduction of a previously subluxed femoral head occurred, as revealed by a reduction of Shenton's line and a decrease in lateral migration distance. This, too, is consistent with improved hip biomechanics and presumably improved longevity of the hip.

Clinically, the patients were active and had little if any gait abnormality, pain, or weakness after distraction treatment. At the most recent follow-up examinations, all except one of our patients was free from pain, limp, and Trendelenburg sign. All of our patients could walk normally and took part in normal daily activities, including sports, and were happy with their outcomes. Considering that 12 of 16 patients in this study were older than 8 years and that 7 were older than 10 years, the prognosis expected with conventional treatment would not be so favorable. Our overall results with distraction were 95% satisfactory based on pain and limp. Containment of the hip by femoral osteotomy, when performed in older patients with hip subluxation, may cause an "incongruent incongruency" situation and worsen the condition of the joint [35–37].

Distraction treatment of the hip has been termed *arthrodiastasis* and has been used for stiffness of the hip after trauma, chondrolysis, slipped capital femoral epiphysis, avascular necrosis, Perthes disease, and other conditions [38, 39]. Often combined with capsulectomy and arthrolysis, it has not been used as the primary treatment for Perthes disease [38] . One study showed unsatisfactory results of such an application that included use of an Ilizarov external fixator without a hinge [40]. The authors who presented that study have since adopted the hinge distraction method reported herein for the primary treatment of Perthes disease and have achieved vastly improved results Guarniero [41] presented the results of a comparative study of two groups of patients diagnosed with Perthes disease, treated by varus femoral osteotomy or hip joint distraction. They reported consistently good results for both groups of patients and noted that the femoral head underwent remodeling faster in the patients treated by hip joint distraction.

Segev who learned this technique during his fellowship with Paley reported on 16 of his own patients with Perthes treated by distraction. The average age was 12 years, which is a much older group of patients than most and therefore would be expected to have a very poor prognosis. All patients had improved range of motion and improved pain scores. This demonstrated improved prognosis over that expected for such an older group of patients [42–44].

Minimal interference with osseous architecture and relative simplicity of hip joint distraction combined with a low complication rate renders this treatment an attractive alternative for more advanced and later-onset cases of Perthes disease. According to Stulberg et al. [17], the most important prognostic factor that affects outcome is residual deformity of the femoral head, coupled with hip joint incongruity. Class I and II spherical hips are compatible with normal longevity of the hip; Class III and IV hips with aspherical congruency usually deteriorate during the sixth decade of life; and Class V hips with incongruity usually degenerate by the fourth decade. This series did not include any cases of incongruity (Class V). Six spherical hips (Class I and II) and 12 aspherical congruity hips (Class III and IV) were included. The long-term prognosis for these patients, therefore, is relatively good, considering that 8 of 18 hips were in patients who were older than 9 years at onset of disease.

In the author's series, we proceeded with treatment once stiffness, subluxation, and collapse were evident in the presence of whole-head involvement in all except two cases in which the treatment was performed immediately after subchondral fracture occurred. The femoral head went on to re-collapse after fixator removal in both patients. One of them underwent reapplication of the fixator and a second distraction treatment without tendon release more than 1 year after the first distraction treatment; a satisfactory result was achieved. Another patient also underwent a second distraction treatment. This patient was a boy who suffered severe deep soft tissue infection of the pelvic pin sites because of poor compliance and poor personal hygiene. For the second distraction treatment, he was treated at a pediatric rehabilitation center; no subsequent difficulty occurred at the pin sites, and an excellent result was achieved after the second treatment. The final results in both of these cases were as good as those achieved by the remainder of the patients after successful one-time treatment. Because distraction does not distort the anatomy, it can be reapplied if it fails the first time. In retrospect, both of the reapplications were avoidable (too early treatment in one case and poor home hygiene in the other). Based on our results, we conclude that immediately after subchondral fracture is too early to apply treatment. Treatment should not be implemented until femoral head resorption is evident, with or without subluxation and collapse. Ideally, the best timing for distraction is 1 day before collapse would occur. Of course such timing is impossible to determine. As such, distraction should be applied during the resorption phase, preferably prior to collapse. Once collapse occurs, the femoral head is misshapen. Distraction or not the femoral head cannot be returned perfectly to its precollapse shape. Applying distraction as soon as possible after collapse gives the best chance of returning it to as close as possible to its precollapse shape.

Although we did not have a control group at our institution and because most other clinical series would have considered many of the cases in this series to be too severe for conventional containment approaches, we think it is reasonable to conclude that hip joint distraction combined with adductor tenotomy and psoas recession leads to results that are as good as or better than the results of traditional containment treatment methods for patients with Perthes disease and for patients with avascular necrosis after slipped capital femoral epiphysis. In contrast the study previously referred to by Guarniero did have a control group of patients treated by varus osteotomy. The healing of the Perthes head involvement was twice as fast in the distraction group as in the varus osteotomy group. This finding was similar to the results observed in this study. A major advantage of hip joint distraction is that it is indicated even in cases in which marked stiffness, subluxation, or deformity of the femoral head is present and is not contraindicated for older children. Distraction treatment is particularly indicated for older children with more severe at-risk and poor prognostic signs. In conclusion, distraction treatment offers many theoretical and practical advantages over conventional containment treatment approaches and is a valuable addition to the armamentarium of the orthopedic surgeon who is faced with managing the difficult problem posed by Perthes disease.

Finally, although the sphericity of the femoral head is improved with distraction, once the femoral head is larger or misshapen into a more ellipsoid or saddle shape, it will ultimately suffer from femoroacetabular impingement. This will lead to pain, labral tear, and joint degeneration. Joint distraction is therefore not the definitive treatment to prevent arthritis of the hip. It is a good alternative to femoral and pelvic osteotomies and is this author's preferred method of treatment for whole-head involvement cases during the resorption phase, which lead to subluxation and collapse of the femoral head. Once the femoral head is fully ossified, and the final shape can be determined, the only treatment that can restore sphericity to the femoral head is the femoral head reduction osteotomy [17]. Therefore, hip distraction can be considered a temporizing method to treat Perthes at an earlier stage irrespective of age and prevent fixed subluxation and more severe deformation of the femoral head. The more definitive joint preservation treatment however is femoral head reshaping by surgical hip dislocation with osteochondroplasty or femoral head reduction osteotomy.

References

1. Kamhi E, Macewen GD. Treatment of Legg-Calve-Perthes disease: prognostic value of Catterall's classification. J Bone Joint Surg Am. 1975;57:651–4.
2. Meehan PL, Angel D, Nelson JM. The Scottish rite abduction orthosis for the treatment of Legg-Perthes disease: a radiographic analysis. J Bone Joint Surg Am. 1992;74:2–12.
3. Lloyd-Roberts GC, Catterall A, Salamon PB. A controlled study of the indications for and the results of femoral osteotomy in Perthes' disease. J Bone Joint Surg Br. 1976;58:31–6.
4. Axer A, Gershuni DH, Hendel D, Mirovski Y. Indications for femoral osteotomy in Legg-Calvé-Perthes disease. Clin Orthop Relat Res. 1980;150:78–87.
5. Salter RB. Role of innominate osteotomy in the treatment of congenital dislocation and subluxation of the hip in the older child. J Bone Joint Surg Am. 1966;48(7):1413–39.
6. Willett K, Hudson I, Catterall A. Lateral shelf acetabuloplasty: an operation for older children with Perthes' disease. J Pediatr Orthop. 1992;12(5):563–8.
7. Daly K, Bruce C, Catterall A. Lateral shelf acetabuloplasty in Perthes' disease. A review of the end of growth. J Bone Joint Surg Br. 1999;81(3):380–4.
8. Eaton GO. Long-term results of treatment in Coxa Plana: a follow-up study of eighty-eight patients. J Bone Joint Surg Am. 1967;48:1031–42.
9. Snyder CH. A sling for use in Legg-Perthes disease. J Bone Joint Surg Am. 1947;29(2):524–6.
10. Evans DL. Legg-Calvé-Perthes' disease; a study of late results. J Bone Joint Surg Br. 1958;40-B(2):168–81.

11. Evans DL, Lloyd-Roberts GC. Treatment in Legg-Calvé-Perthes' disease; a comparison of in-patient and out-patient methods. J Bone Joint Surg Br. 1958;40-B(2):182–9.
12. Catterall A. The natural history of Perthes' disease. J Bone Joint Surg Br. 1971;53:37–53.
13. Herring JA, Neustadt JB, Williams JJ, Early JS, Browne RH. The lateral pillar classification of Legg-Calve-Perthes disease. J Pediatr Orthop. 1992;12:143–50.
14. Salter RB, Thompson GH. Legg-Calvé-Perthes disease. The prognostic significance of the subchondral fracture and a two-group classification of the femoral head involvement. J Bone Joint Surg Am. 1984;66(4):479–89.
15. Bennett J, Stuecker R, Smith E, Winder C, Rice J. Arthrographic findings in Legg-Calvé-Perthes disease. J Pediatr Orthop B. 2002;11(2):110–6; Review.
16. Stulberg SD, Cooperman DR, Wallensten R. The natural history of Legg-Calve-Perthes disease. J Bone Joint Surg Am. 1981;63:1095–108.
17. Paley D. The treatment of femoral head deformity and Coxa Magna by the Ganz femoral head reduction osteotomy. Orthop Clin North Am. 2011;42(3):389–99, Viii. Epub 2011 May 14. Review.
18. Gower WE, Johnston RC. Legg-Perthes disease: long-term follow-up of thirty-six patients. J Bone Joint Surg Am. 1971;53:759–68.
19. Mcandrew MP, Weinstein SL. A long-term follow-up of Legg-Calve-Perthes disease. J Bone Joint Surg Am. 1984;66:860–9.
20. Yrjonen T. Long-term prognosis of Legg-Calve-Perthes disease: a meta-analysis. J Pediatr Orthop B. 1999;8:169–72.
21. Ippolito E, Tudisco C, Farsetti P. Long-term prognosis of Legg-Calve-Perthes disease developing during adolescence. J Pediatr Orthop. 1985;5:652–6.
22. Ippolito E, Tudisco C, Farsetti P. The long-term prognosis of unilateral Perthes' disease. J Bone Joint Surg Br. 1987;69:243–50.
23. Norlin R, Hammerby S, Tkaczuk H. The natural history of Perthes' disease. Int Orthop. 1991;15:13–6.
24. Mazda K, Pennecot GF, Zeller R, Taussig G. Perthes' disease after the age of twelve years: role of the remaining growth. J Bone Joint Surg Br. 1999;81:696–8.
25. Eyre-brook AL. Osteochondritis deformans coxa juvenilis or Perthes disease: the result of treatment by traction in recumbency. Br J Surg. 1936;24:166–82.
26. Kiepurska A. Late results of treatment in Perthes' disease by a functional method. Clin Orthop. 1991;272:76–81.
27. Martinez AG, Weinstein SL, Dietz FR. The weight-bearing abduction brace for the treatment of Legg-Perthes disease. J Bone Joint Surg Am. 1992;74:12–21.
28. Lack W, Feldner-Busztin H, Ritschl P, Ramach W. The results of surgical treatment for Perthes' disease. J Pediatr Orthop. 1989;9:197–204.
29. Bankes MJ, Catterall A, Hashemi-Nejad A. Valgus extension osteotomy for 'hinge abduction' in Perthes' disease: results at maturity and factors influencing the radiological outcome. J Bone Joint Surg Br. 2000;82:548–54.
30. Klisic PJ. Treatment of Perthes' disease in older children. J Bone Joint Surg Br. 1983;65:419–27.
31. Noonan KJ, Price CT, Kupiszewski SJ, Pyevich M. Results of femoral varus osteotomy in children older than 9 years of age with Perthes disease. J Pediatr Orthop. 2001;21(2):198–204.
32. Rowe SM, Jung ST, Cheon SY, Choi J, Kang KD, Kim KH. Outcome of cheilectomy in Legg-Calve-Perthes disease: minimum 25-year follow-up of five patients. J Pediatr Orthop. 2006;26(2):204–10.
33. Lee DS, Jung ST, Kim KH, Lee JJ. Prognostic value of modified lateral pillar classification in Legg-Calvé-Perthes disease. Clin Orthop Surg. 2009;1(4):222–9; Epub 25 Nov 2009.
34. Mose K. Methods of measuring in Legg-Calve-Perthes disease with special regard to the prognosis. Clin Orthop. 1980;150:103–9.
35. Lloyd-Roberts GC. Osteoarthritis of the hip: a study of the clinical pathology. J Bone Joint Surg Br. 1955;37-B(1):8–47.
36. Cooperman DR, Stulberg SD. Ambulatory containment treatment in Perthes' disease. Clin Orthop Relat Res. 1986;203:289–300.

37. Salter RB. Legg-Perthes disease: the scientific basis for the methods of treatment and their indications. Clin Orthop Relat Res. 1980;150:8–11.
38. Canadell J, Gonzales F, Barrios RH, Amillo S. Arthrodiastasis for stiff hips in young patients. Int Orthop. 1993;17:254–8.
39. Aldegheri R, Trivella G, Saleh M. Articulated distraction of the hip: conservative surgery for arthritis in young patients. Clin Orthop. 1994;301:94–101.
40. Kocaoglu M, Kilicoglu OI, Goksan SB, Cakmak M. Ilizarov fixator for treatment of Legg-Calvé-Perthes disease. J Pediatr Orthop B. 1999;8(4):276–81.
41. Guarniero R. Comparative study of two groups of patients diagnosed with Perthes disease, treated by varus femoral osteotomy or hip joint distraction. Presented at Perthes course, Sept 2006, Baltimore.
42. Segev E, Ezra E, Wientroub S, Yaniv M. Treatment of severe late onset Perthes' disease with soft tissue release and articulated hip distraction: early results. J Pediatr Orthop B. 2004;13(3):158–65.
43. Segev E. Treatment of severe late onset Perthes' disease with soft tissue release and articulated hip distraction. J Pediatr Orthop B. 2004;13(5):345.
44. Segev E. Correspondence: treatment of severe late onset Perthes disease with soft tissue release and articulated hip distraction (reply). J Pediatr Orthop B. 2008;17(1):55.

8

Comparison Between Distraction and Femoral Varus Osteotomy in the Treatment of Legg-Calvé-Perthes Disease

Roberto Guarniero and José Roberto Bevilacqua Guarniero

Introduction

The goal of treatment in Legg-Calvé-Perthes disease (LCPD) is to prevent deformity of the femoral head and henceforth prevent degenerative changes of the hip in adult life, which could manifest itself at an early age [1–3].

The basic principle of treatment is the prevention of abnormal forces acting on the femoral head during its vulnerable phases of revascularization and bone healing.

The four basic goals of treatment are:

1. To eliminate hip irritability
2. To restore and maintain a good range of hip motion
3. To prevent femoral head extrusion and subluxation
4. To obtain a femoral head as spherical as possible when healed

The selected method depends upon several factors:

(a) Clinical symptoms and physical findings
(b) Radiographic femoral head appearance
(c) Parents' ability to carry out treatment

It is possible to conclude that the main focus of the treatment of LCPD is to allow repair of the femoral head under optimal biomechanical conditions. When surgical treatment is indicated, it should interfere as little as possible with the child's psychological and physical development.

R. Guarniero (✉) · J. R. B. Guarniero
Ortopedia e Traumatologia, das Clínicas HCFMUSP Universidade de Sao Paulo,
Sao Paulo, SP, Brazil

D. S. Feldman, D. Paley (eds.), *Legg-Calvé-Perthes Disease*,
https://doi.org/10.1007/978-1-0716-0854-8_8

The contemporary concept of treatment is containment of the head deeply within the acetabulum. This may, during the phase of softening, allow the anterolateral segment of the head to be molded by the socket and prevented from bulging outwards, as discussed by Lloyd-Roberts and Fixsen [4].

Either the surgical procedure of osteotomy of the pelvis or femur can be used to obtain femoral head containment. The osteotomy can be a femoral varus osteotomy or the innominate Salter osteotomy of the iliac bone with similar results [5, 6].

Varus osteotomy has been used in this hospital since 1965. In three previous studies done at our institution, Cordeiro [7, 8] and Guarniero et al. [9, 10] analyzed the role of intertrochanteric osteotomy of the femur in the treatment of 52 cases of LCPD showing 56.6% of good and fair results and few complications. The main conclusion of these studies is that the age of the patient influenced the results. Patients operated upon the age of 9 years or older had poor radiologic and clinical results. The intertrochanteric varus osteotomy is a good method of treatment for the ages of 4–9 years old. The varus osteotomy may avoid or delay the appearance of osteoarthritis of the hip [9, 10].

The choice of the type of osteotomy should take into account several factors, among them, the patient's age and the surgeon's experience. Many papers do not show significant difference in the results among the two types of osteotomy, femoral varus or acetabular. Furthermore, according to Joseph et al. [11], femoral osteotomy seems to be more reliable.

Trying to improve the outcome in LCPD, we started using the arthrodiastasis of the hip with an external monolateral fixator, creating negative pressure over the avascular femoral head in the active phase of the disease [12]. According to Paley [13] cartilage in LCPD is very bioactive and tends to proliferate in unloaded regions of the hip joint, and the distraction obtained with the external fixator takes the load off the femoral head, creating a space for the proliferation of the cartilage and shaping itself to the acetabular cavity.

Arthrodiastasis of the hip is a relatively new method of treatment. It does not change the anatomy of the joint. It provides a way to bypass loading forces of the joint which negates the harmful effect of stress on articular surfaces thereby promoting healing of the areas of necrosis.

We have been applying this method since 1992 as an alternative to femoral osteotomy in surgical treatment for LCPD [12].

Femoral Varus Osteotomy

Indication

The main indication for a femoral osteotomy is the tendency for lateralization of the femoral head and when the proximal femoral epiphysis is seen well contained in the abduction on the radiographic image. In this situation, containment can be achieved by varus intertrochanteric osteotomy with or without rotation as described by Axer [14].

According to Chung [15] varus osteotomy is used in patients with LCPD to contain the femoral head by rotating it medially so the articular margin falls within the acetabulum lip and the osteotomy redistributes pressure by shifting the weight-bearing surface to the lateral, usually intact, femoral head margin.

Varus osteotomy alone is contraindicated if the femoral head is severely subluxated and the lateral margin of the head cannot be contained within the lateral acetabular lip. The procedure is also contraindicated in severe coxa magna in which coxa vara is already present.

Using the Catterall [16] classification, the indication for surgery is mainly reserved for cases in groups III and IV, "at risk," in older children, generally over 8 years old.

The indication for surgical treatment in the younger child is based on the presence of the so-called radiographical "risk factors."

Surgical Technique Considerations

The surgical technique follows three main points:

1. Intertrochanteric varus osteotomy
2. Medial displacement of the femoral shaft
3. Bone fixation with a metallic plate and screws

The varus angulation should be no less than 110 ° but should allow containment of the femoral head epiphysis.

The osteotomy is usually held securely with threaded screws and a side plate or blade plate. Cordeiro [7, 8] described the fixation with Ottolenghi's plate (Figs. 8.1, 8.2, 8.3, 8.4, and 8.5).

After operation the patient is kept in a cast, a 1½ hip spica, for 6–8 weeks.

After cast removal the patient remains another 6 weeks in touch weight bearing with crutches.

The patient must be examined every 4 weeks after the operation.

Advantages of Femoral Varus Osteotomy

- The proximal femoral osteotomy decompresses the head.
- The surgeons usually are very well familiarized with the technique.

Disadvantages of Femoral Varus Osteotomy

- This type of osteotomy requires internal fixation and subsequent plate removal.
- In some cases is observed a slight shortening of the operated leg (femur).

Fig. 8.1 Patient aged 8 years. Catterall III

Hip Distraction: Arthrodiastasis

Hip distraction is a relatively new method for the treatment of Legg-Calvè-Perthes disease (LCPD). The technique has been used in this institution for LCPD since 1995, and the results are very promising [12, 17, 18] .

We agree with Hosny [19] that joint distraction attempts to neutralize muscular and weight-bearing forces on the femoral epiphysis, induce neovascularization, and prevent head deformation. Another advantage of distraction is that it does not change the anatomy of the proximal femur in patients with LCPD. Hip distraction seems to be a valid treatment option for LCPD patients where poor results are expected from conventional treatment.

Fig. 8.2 Cordeiro's technique for femoral varus osteotomy

Fig. 8.3 Same patient. Long-term result

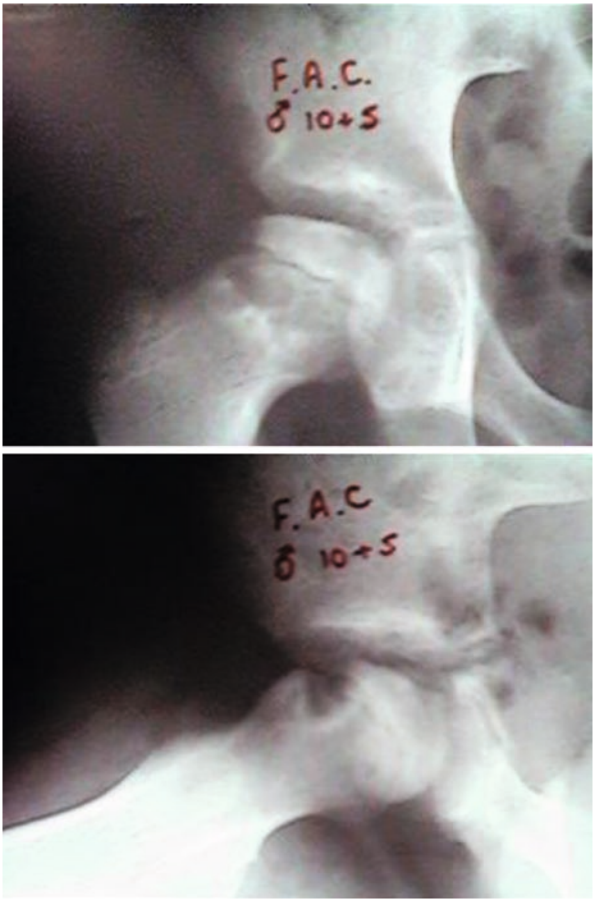


Fig. 8.4 Patient male 10 years and 5 months of age. Catterall III

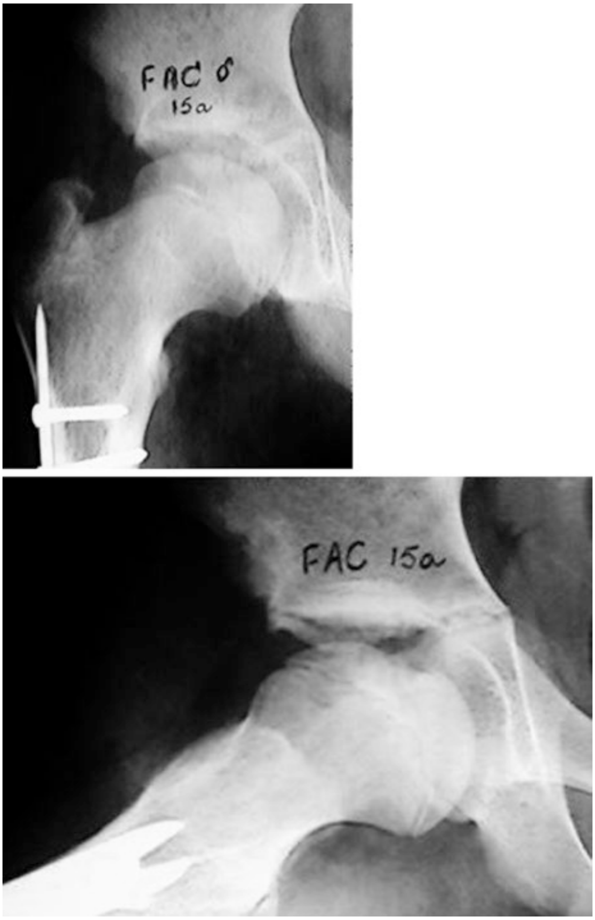


Fig. 8.5 Five years after operation. Good result

Our protocol for hip distraction is unilateral disease, active phase of the disease (necrosis or fragmentation), age of the patient above 6 years, Catterall groups III and IV, and hip subluxation under 50% [12].

The distraction can be done with a monolateral external fixator (Fig. 8.6) or with a circular framed Ilizarov's fixator (Fig. 8.7). In Figs. 8.8, 8.9, 8.10, 8.11, 8.12, 8.13, 8.14, and 8.15 are illustrated the main points of the surgical procedure. Note in Figs. 8.8, 8.9, and 8.15 is an important step of the procedure – the correction of the Shenton's line.

Fig. 8.6 Example of distraction with a monolateral external fixator. The arrow points to "new bone formation" just after 1 month of distraction

Fig. 8.7 Example of distraction with an Ilizarov's circular external fixator

Fig. 8.8 Shenton's line just before distraction

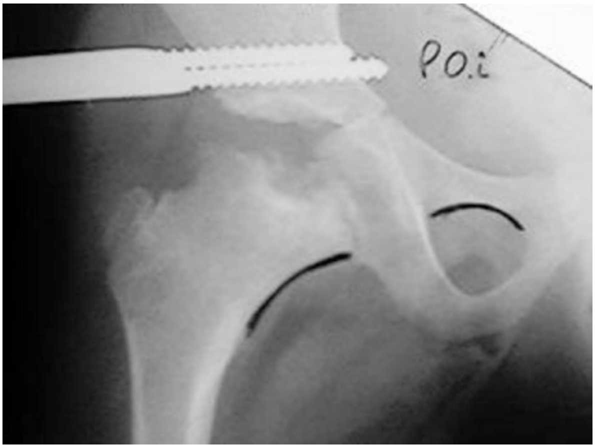


Fig. 8.9 Immediate postoperative. Correction of the Shenton's line

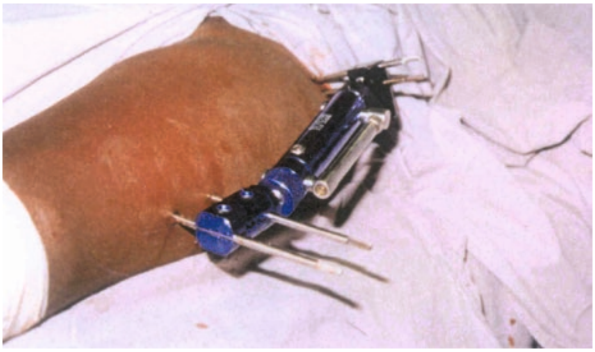


Fig. 8.10 External monolateral fixator in place


[redacted]

Fig. 8.11 The procedure is controlled by image intensifier

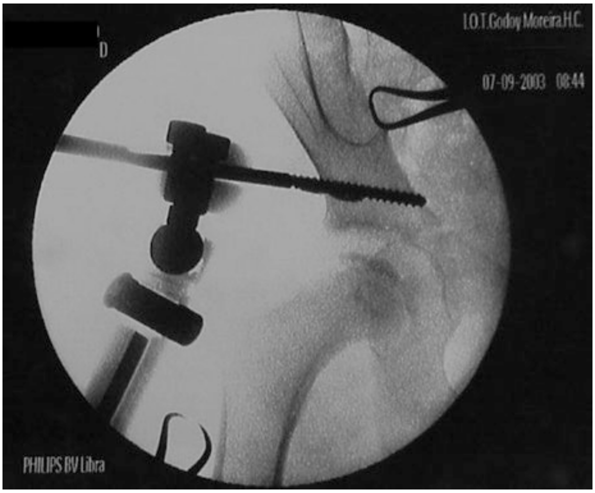
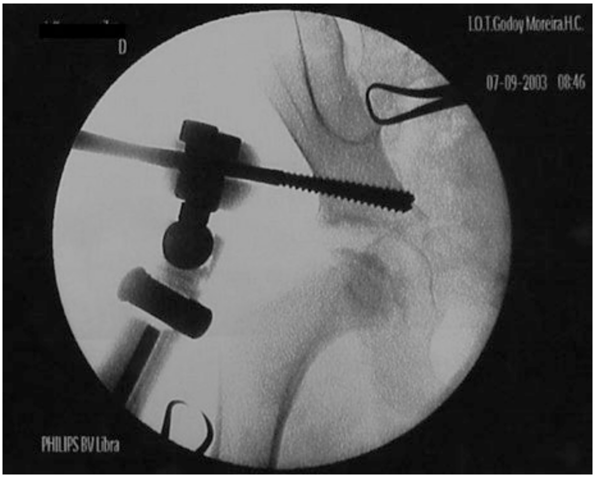

[redacted]

Fig. 8.12 Positioning the proximal pair of Schanz screws

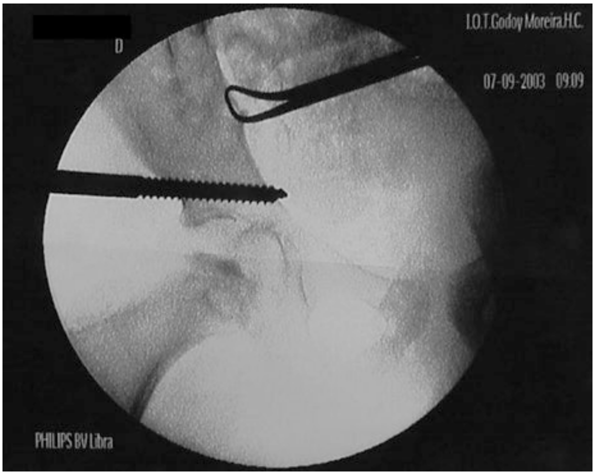
Comparison Between Distraction and Femoral Varus Osteotomy

Between 1995 and 1998, a prospective comparative study was done in this service aiming to evaluate the time period of femoral head reossification (remodeling) between two groups of patients (“FVO” and “distraction”) with LCPD submitted to surgical treatment. In the “FVO group,” 18 patients were submitted to femoral varus osteotomy; and in “distraction group,” 18 patients were submitted to hip distraction with a monolateral external fixator.



[redacted]

Fig. 8.13 Beginning of the distraction



[redacted]

Fig. 8.14 The end of the distraction

This study was approved by the Institutional Review Board on Ethics and Research of this university. The indication for surgical treatment was exactly the same in both groups: all patients aged 6 years old or above, presenting unilateral hip involvement with pain and decrease in the joint motion. The patients were in Catterall groups III or IV. In Table 8.1 is shown the result concerning the time period of femoral head reossification.

The time period for new bone formation has been shortened with the usage of distraction with a monolateral external fixator. The average period with distraction was 3.16 months and with femoral varus osteotomy was 7.16 months. In Table 8.2 is shown the evaluation concerning Stulberg's classification, at skeletal maturity.

From this study we concluded that distraction is a viable alternative in the treatment of LCPD and tends toward better healing and radiographic outcome than VDO.

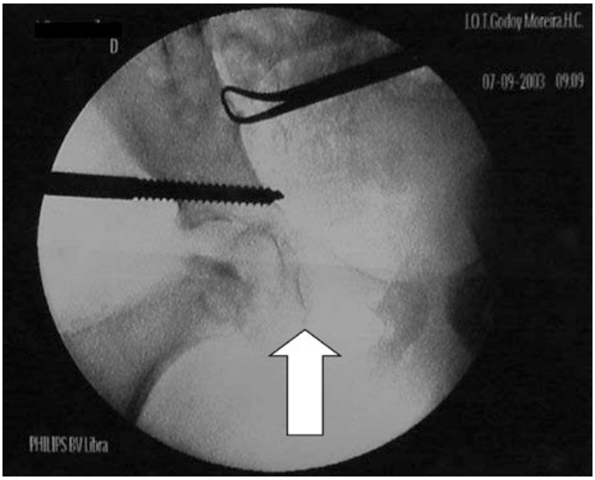

[redacted]

Fig. 8.15 Note the overcorrection of the Shenton’s line (arrow)

Table 8.1 Time period for femoral head reossification (months)

Time period (months)	FVO group	Distraction group
Minimum	4	2
Maximum	11	4.5
Average	7.16	3.16

Table 8.2 Stulberg’s classification at skeletal maturity of both groups

Group	FVO	Distraction	Total
I	2	1	3
II	5	11	16
III	8	1	9
IV	3	4	7
V	0	1	1
Total	18	18	36

References

1. Kim HKW. Legg-Calvé-Perthes disease:etiology, pathogenesis and biology. J Pediatr Orthop. 2011;31:S141–6.
2. Wenger DR, Pandya NK. A brief history of Legg-Calvé-Perthes disease. J Pediatr Orthop. 2011;31:S130–6.
3. Herring JA. Legg-Calvé-Perthes disease at 100: a review of evidence based treatment. J Pediatr Orthop. 2011;31:S137–40.
4. Lloyd-Roberts GC, Fixsen J. Orthopaedics in infancy and childhood. London: Butterworth-Heinemann; 1990. p. 157.
5. Wiig O, Terjesen T, Svenningsen S. Prognostic factors and outcome of treatment in Perthes’ disease. J Bone Joint Surg Br. 2008;90-B(10):1364–71.
6. Herring JA. JBJS A. 2004;86:2121–34.
7. Cordeiro EN. Femoral osteotomy in Legg-Calvé-Perthes disease. Clin Orthop. 1980;150:69–72.

8. Cordeiro EN. Osteotomia varizante intertrocantérica do fêmur na moléstia de Perthes. São Paulo: Tese Fac Med Univ; 1972.
9. Guarniero R, Ishikawa MT, Luzo CAM, Montenegro NB, Godoy RM Jr. Resultados da osteotomia femoral varizante no tratamento da doença de Legg-Calvé-Perthes. Clinics. 1997;52:132–5.
10. Guarniero R, Adachi PP, Gouveia LAM. The results of femoral varus osteotomy for the treatment of Legg-Calvé-Perthes disease. Mapfre Med. 1995;6(Suppl III):139–40.
11. 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;14(1):10–5.
12. Luzo CA, Guarniero R, Montenegro NB, de Godoy Junior RM. Initial experience of use of an articulated external fixator in treating Legg-Calvé-Perthes disease by means of arthrodiastasis during the active phase of the disease. Rev Bras Ortop. 2016;51:337–45.
13. Paley D. Joint distraction for hip pathology. In: Paley D, Herzenberg J, Tetsworth K, Bhave A, editors. Principles of deformity correction course. Baltimore: Maryland Center for Limb Lengthening and Reconstruction, 1999.
14. Axer A. Subtrochanteric osteotomy in the treatment of Perthes' disease. J Bone Joint Surg Br. 1965;47:489–99.
15. Chung SMK. Hip disorders in infants and children. Philadelphia: Lea & Febiger; 1981.
16. Catterall A. The natural history of Perthes' disease. J Bone Joint Surg Br. 1971;53:37–52.
17. Luzo CAM. Artrodiastase com fixador externo unilateral no tratamento da doença de Legg-Calvé-Perthes [tese]. São Paulo: Faculdade de Medicina, Universidade de São Paulo; 1998.
18. Guarniero R, de Godoy Junior RM, Montenegro NB, Luzo CAM, Kiss MHB. Artrodiastase para o tratamento da rigidez do quadril na artrite reumatoide juvenil (ARJ) – resultados preliminares. Rev Bras Ortop. 2010;45(suppl 1):15–8.
19. Hosny GA. Articulated distraction. Orthop Clin N Am. 2011;42:361–4.

9

Femoral Head Reduction Osteotomy for Coxa Magna and Nonspherical Femoral Head for Legg-Calvé-Perthes Disease

Dror Paley

Femoral head sphericity is the key to joint preservation and longevity in Perthes disease. There is no controversy that a spherical femoral head has the best prognosis after Perthes disease [1, 2]. The Stulberg classification correlates well with natural history after Perthes disease [1]. Stulberg Class I and II are both spherical femoral heads. Stulberg Class III, IV, and V are all aspherical elongated femoral heads all referred to as coxa magna but also described as ellipsoid, mushroom shaped, saddle shaped, flattened, etc. Although a Class III has a better prognosis than a Class IV or V, they all lead to pain, femoroacetabular impingement, labral tears, degenerative changes to the femoral head and acetabulum, and loss of range of motion and limp. Most have symptoms as early as the teen age years, although radiographic degenerative changes may not manifest in Class III for two or three decades. The goal of containment and distraction treatment by brace/osteotomy or external fixation respectively is to prevent or reverse loss of sphericity of the femoral head. Once it is present at the end of treatment, it leads to damage to the labrum and joint cartilage. Until recently, restoring the sphericity of the femoral head was not an option. Cheilectomy [3] and valgus osteotomy [4] were considered salvage procedures which produced short term benefits.

More recently, safe surgical dislocation combined with relative neck lengthening and trochanteric transfer have made it possible to improve reshaping techniques of the deformed femoral head [5, 6]. Osteochondroplasty of the anterolateral femoral head may be indicated, especially if the remainder of the femoral head is spherical and lacks degenerative changes. Unfortunately, the majority of cases of deformed femoral heads have degenerative changes corresponding to contact with the rim of the acetabulum, while the more anterolateral extruded femoral head has well preserved excellent hyaline cartilage covering it. In such cases osteochondroplasty

---

D. Paley (✉)
Paley Orthopedic and Spine Institute, St. Mary's Medical Center, West Palm Beach, FL, USA
e-mail: dpaley@paleyinstitute.org

D. S. Feldman, D. Paley (eds.), *Legg-Calvé-Perthes Disease*,
https://doi.org/10.1007/978-1-0716-0854-8_9

would resect the healthy cartilage while preserving the degenerated cartilage. This may explain why 'cheilectomy' usually failed for Perthes disease [3].

In 2001, Reinhold Ganz from Bern, Switzerland, performed an intra-articular osteotomy of the central third of the femoral head and reduced the lateral cartilage to the medial preserved portion of the femoral head [5]. The lateral segment of the femoral head was mobilized on its vascular pedicle. The vascular pedicle to the medial femoral head was also preserved. This technique is ideal for the femoral head deformity following Perthes. As mentioned above, the lateral part of the femoral head usually has well-preserved cartilage because it never enters the acetabulum due to the enlarged femoral head. Meanwhile the adjacent cartilage that is impinging on the acetabular rim is usually damaged and is therefore ideal to be resected. The sooner this procedure is performed, the less degeneration there is to the femoral head and acetabulum. For this reason the ideal age to perform this procedure is before secondary changes occur, such as age 9–16 years. This chapter will examine the improving and evolving techniques of femoral head reduction osteotomy (FHRO) and their indications and results [7–10].

Goals of Treatment

- Convert an aspherical shaped femoral head to a spherical shaped femoral head.
- Eliminate any residual femoroacetabular impingement by osteochondroplasty.
- Reduction of coxa magna to normal sized femoral head.
- Change from coxa breva to a relatively longer femoral neck.
- Advance the overgrown greater trochanter.
- Identify and repair labral tear.
- Restore normal femoral head coverage.

Treatment Strategy

- Determine preoperatively whether the elongation of the femoral head is perpendicular to the femoral neck or not (Fig. 9.1). If perpendicular then a Ganz-type (Fig. 9.2a) FHRO is preferred, while if it is more horizontal, then the Paley type is preferred (Fig. 9.2b)
- Determine preoperatively whether this is a type A or type B (Fig. 9.3a, b). Type A requires a uniplanar reduction with parallel bone cuts, while type B requires a biplanar osteotomy with convergent bone cuts (wedge resection).
- Measure the opposite normal shaped femoral head and acetabulum and measure the ipsilateral femoral head and acetabulum.
- Determine the amount of reduction needed on AP and lateral views.
- Plan the amount of transfer needed to move the greater trochanter laterally and distally so that the tip of the greater trochanter is at the level of the center of the femoral head and is twice the radius of the femoral head away from the center of the joint.

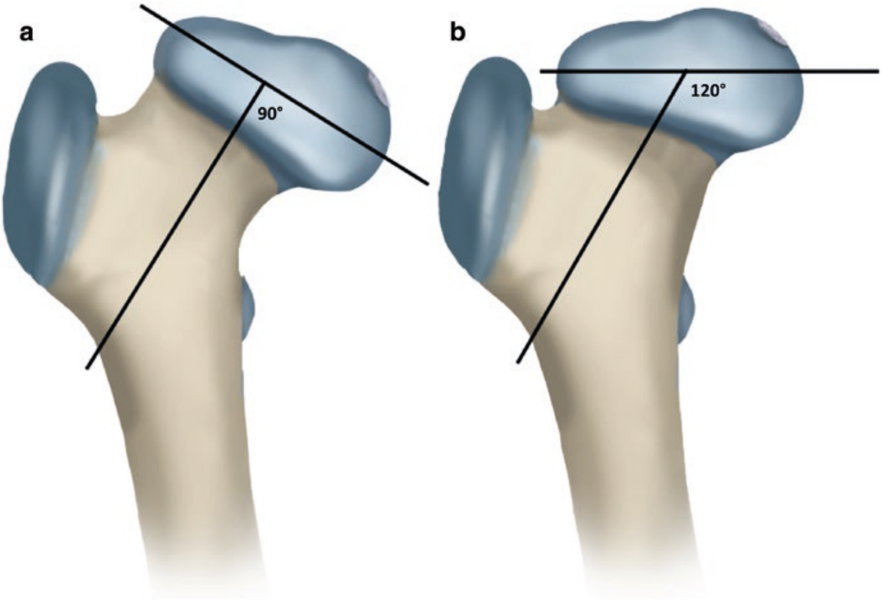


Fig. 9.1 Illustration of Type 1 elongated femoral head with elongation perpendicular to femoral neck (left) vs. Type 2 elongated femoral head with elongation not perpendicular to femoral neck. ((C) Dror Paley. Used with permission)

Surgical Details of Femoral Head Reduction Osteotomy

- Step 1: Anesthesia, positioning and draping: An epidural is placed by the anesthesia service with a catheter running up the back on the nonoperative side. A urinary catheter is inserted. The patient should be placed on their side on a beanbag on a radiolucent table in the lateral decubitus position with the operative side up. The ipsilateral arm should be appropriately padded and supported on an armrest. An axillary roll is utilized for the arm on the opposite side, and there is soft protection of boney prominences including the peroneal nerve as it crosses the fibular neck on the leg that is down. The entire operative side should be prepped and draped free from the nipple to the toes. The drapes should extend from the mid buttocks to the scrotal/labial-thigh fold. The lower limb should be completely free of the drapes. A sterile pocket should be created using drapes or a special pouch and fixed to the anterior side of the patient in line with the hip. Place a bump under the operative knee to abduct the operative hip.
- Step 2: Incision to and through fascia lata: An approximately 15-cm-long incision centered on the greater trochanter (GT) is made to and through the fascia lata (Fig. 9.4a). The fascia is separated from the gluteus maximus (GMax) muscle anteriorly to reflect this muscle posteriorly (Fig. 9.4b, c). This avoids splitting and denervating the anterior portion of this muscle.

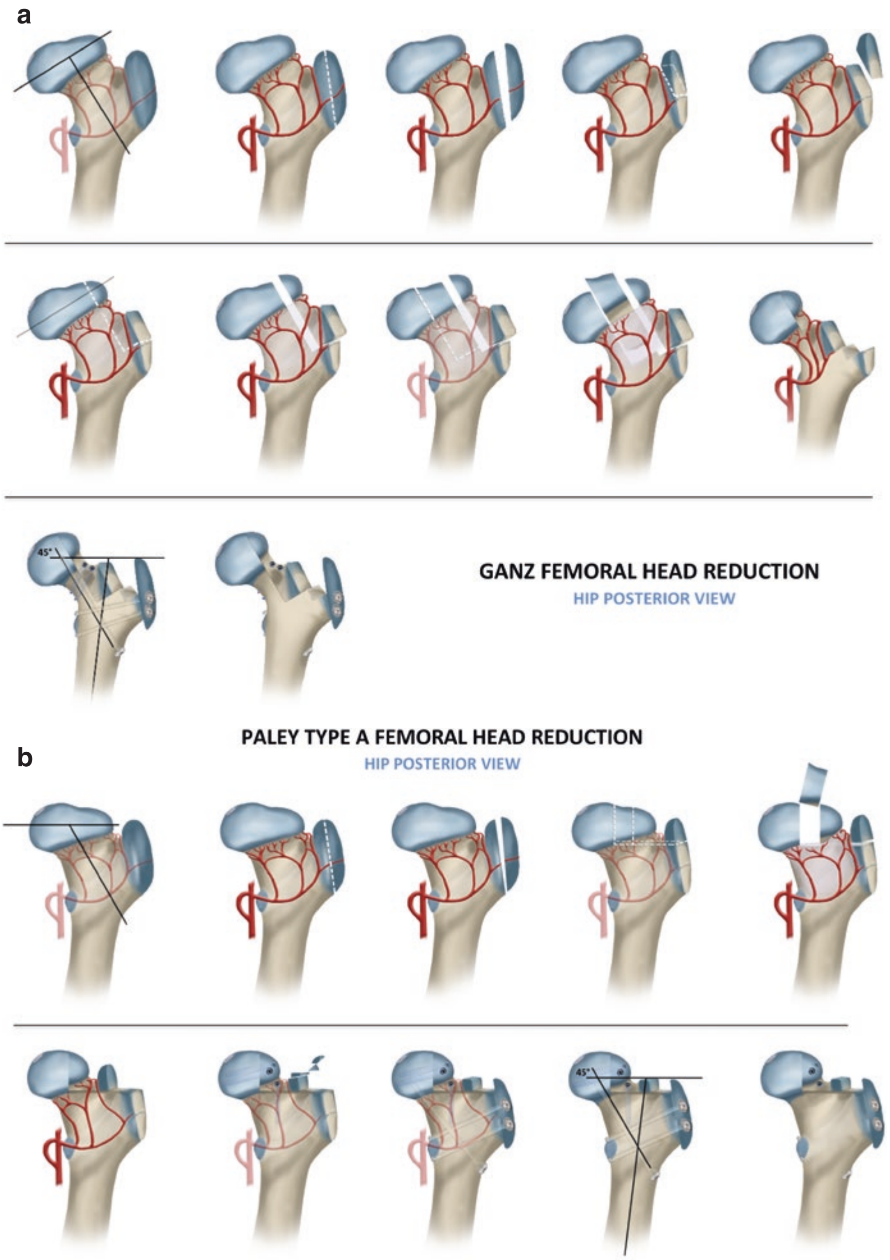
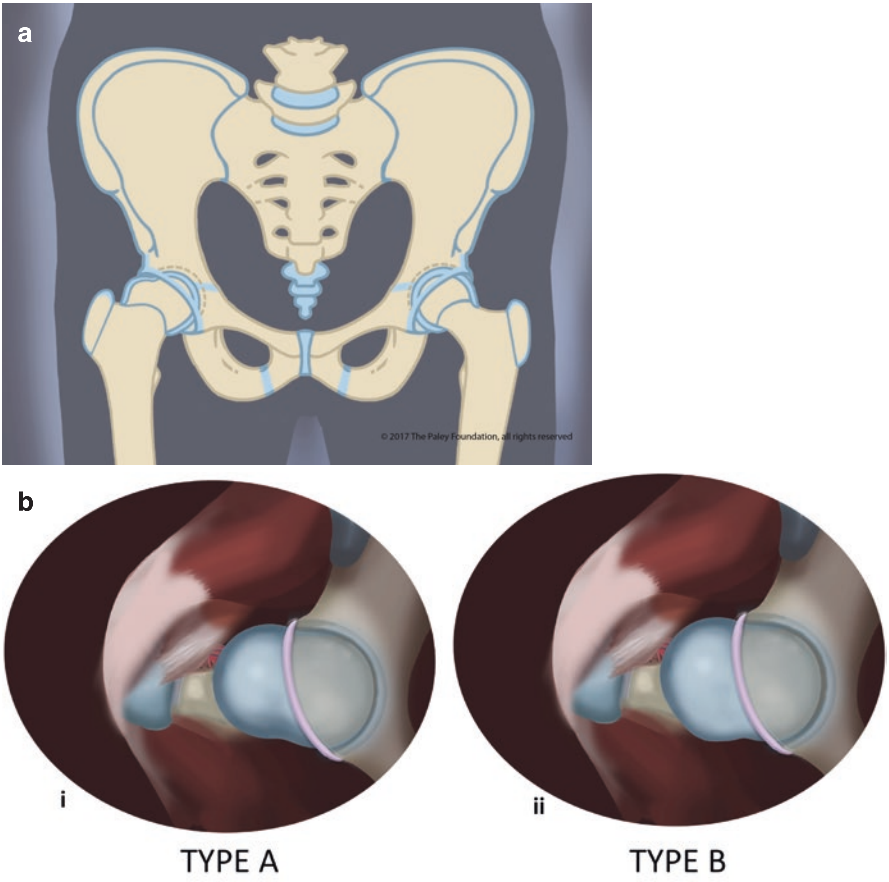


Fig. 9.2 (**a**) Ganz-type FHRO with cuts parallel to the femoral neck applied to Type 1 and femoral head elongations. (**b**) Paley-type FHRO with cuts perpendicular to the femoral head applied to Type 2 femoral head elongations. ((C) Dror Paley. Used with permission)

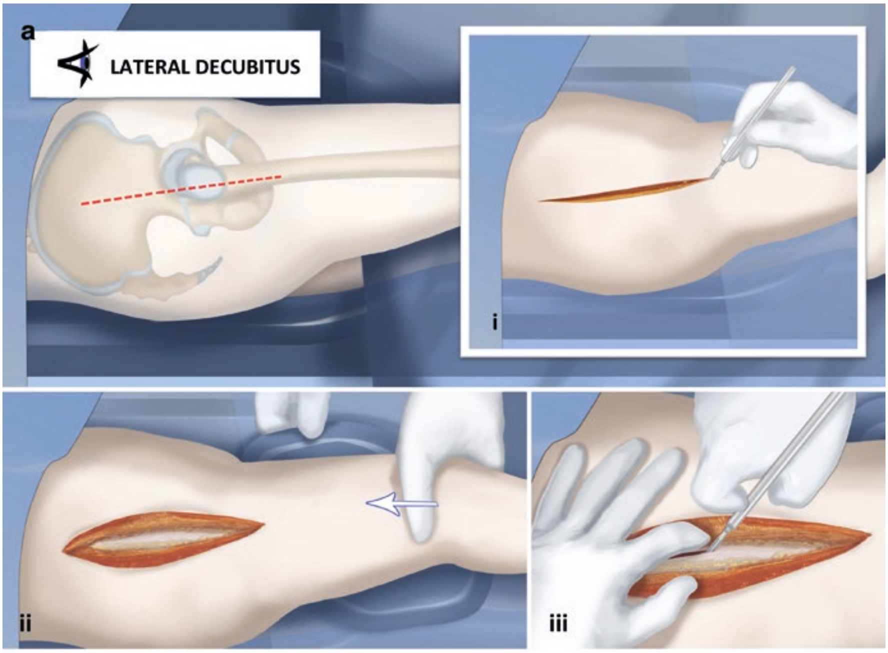


© 2017 The Paley Foundation, all rights reserved

Fig. 9.3 (**a**) Illustration of saddle shaped right coxa magna, coxa breva with overgrown greater trochanter. The saddle part of the femoral head is where the femoroacetabular impingement occurs. (**b**) Superior view of (i) Type A coxa magna with lateral elongation of femoral head and (ii) Type B coxa magna with lateral elongation with anterior protrusion. ((C) Dror Paley. Used with permission)

- Step 3: Trochanteric bursa and piriformis muscle: Enter the trochanteric bursa (Fig. 9.4c) and identify the inferior edge of the piriformis tendon and the posterior edge of the gluteus medius muscle (Fig. 9.5a). The posterior border of the gluteus medius is very distinct. The terminal branch of the inferior gluteal artery (which anastomoses with the medial femoral circumflex artery) runs along the inferior border of the piriformis muscle. Also identify the junction between the gluteus maximus tendon and the posterior edge of the vastus lateralis.

- Step 4: Retract gluteus medius and minimus muscles: Identify the junction of the gluteus medius and its underlying gluteus minimus and the superior border of the piriformis muscle (Fig. 9.5b). The gluteus medius is more superficial than the deeper piriformis. The piriformis is in a more superficial layer than the gluteus minimus. The border of these three muscles interdigitate with each other such that the posterior border of the gluteus medius slightly overlies the superior border of the piriformis which slightly overlies the posterior border of the gluteus minimus. Separate and retract distally the superior border of the piriformis from the posterior borders of the two glutei. Undermine the gluteus minimus to find the interval between this muscle and the underlying capsule. Insert a Z retractor into this interval to lift the glutei away from the capsule (Fig. 9.5c).
- Step 5: Elevate and retract the vastus lateralis: Using a cautery elevate the posterior border of the vastus lateralis off the femur. This is just anterior to the GMax tendon. Insert a Hohmann elevator to elevate this muscle (Fig. 9.5c).
- Step 6: Cauterize the posterior border of the GT: Connect a virtual line between the elevated vastus and the elevated glutei. Use a cautery to mark this line along



© 2017 The Paley Foundation, all rights reserved

Fig. 9.4 (**a**) Lateral decubitus position with midlateral incision (i). The incision goes straight through skin, subcutaneous fat (ii), and fascia lata (iii). (**b**) The fascia is split longitudinally the entire length of the incision (iv, v) and then separated anteriorly from the underlying gluteus maximus (GMAX). (**c**) The gluteus maximus is reflected posteriorly from its anterior edge so as not to split and denervate it (viii, ix). The trochanteric bursa is opened (x) to expose the greater trochanter (xi). ((C) Dror Paley. Used with permission)

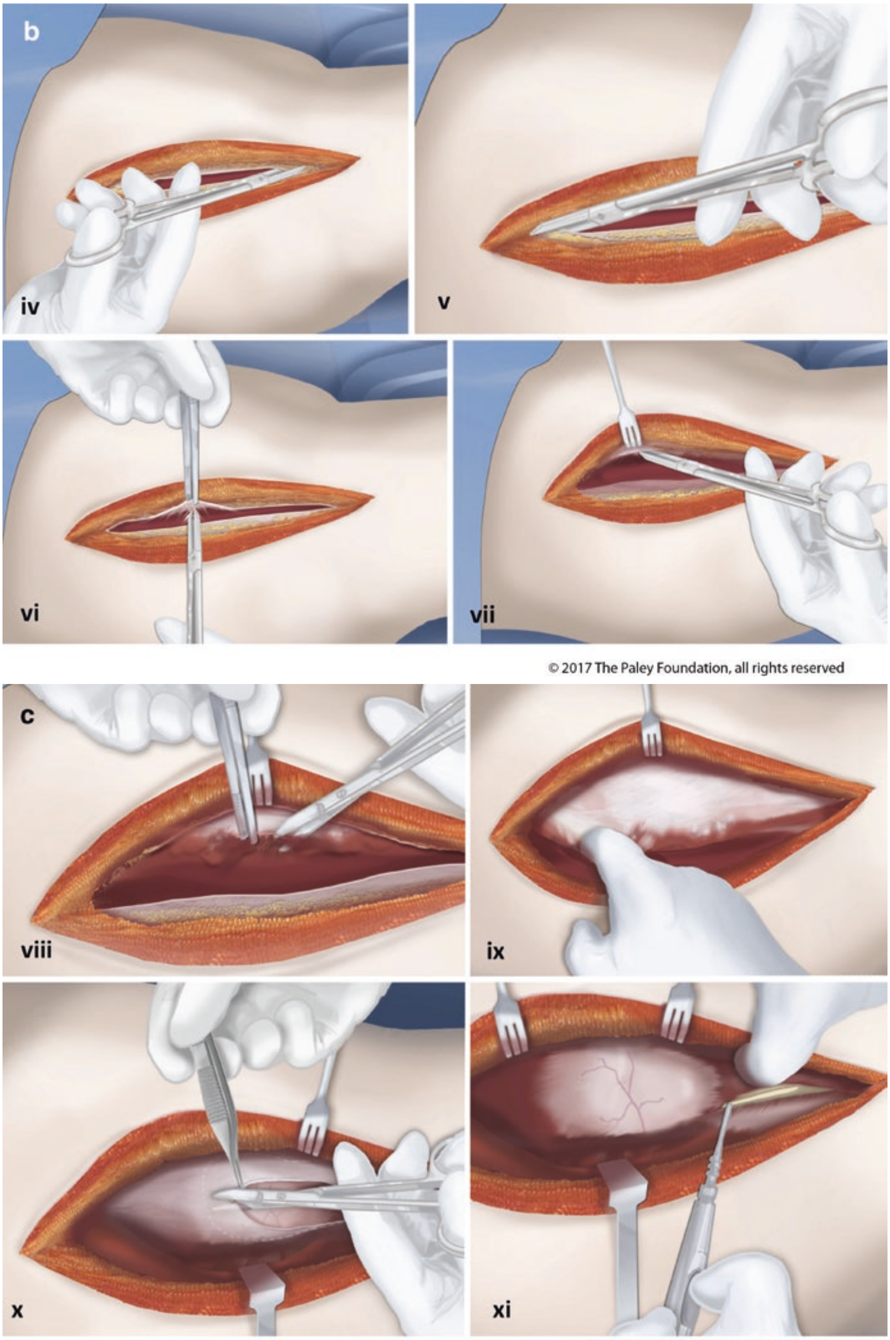


Fig. 9.4 (continued)

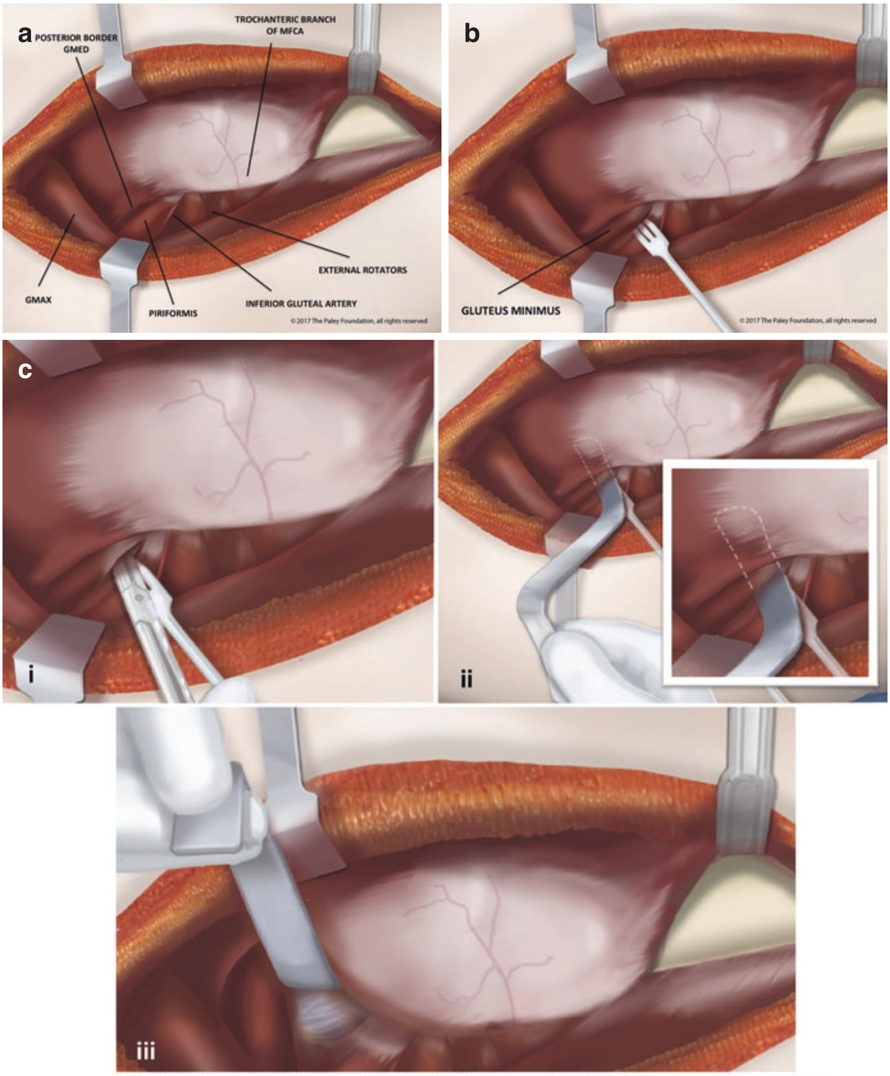


Fig. 9.5 (**a**) The posterior border of the gluteus medius (GMED) is very distinct. The inferior gluteal artery helps define the inferior border of the piriformis muscle. The trochanteric branch of the medial femoral circumflex artery (MFCA) is readily seen on the greater trochanter. The gluteus maximus tendon is seen inserting into the femur. The Vastus lateralis muscle is lifted off of the femur. (**b**) The gluteus minimus can be seen by reflecting the piriformis muscle distally. The GMED is slightly superficial to the Piriformis which is slightly superficial to the gluteus minimus. (**c**) The space under the gluteus minimus is dissected (i) so that a Z retractor can be inserted beneath it overtops the capsule (ii). The Z retractor is used to expose the underlying capsule (iii). A Hohmann elevator is lifting the Vastus Lateralis off the femur. ((C) Dror Paley. Used with permission)

the trochanteric ridge. This serves to cauterize the trochanteric vessels which are branches of the medial femoral circumflex vessels (Fig. 9.6a, b).

- Step 7: GT osteotomy: Use a saw to cut from posterior to anterior along the trochanteric ridge (Fig. 9.6c). The thickness of the lateral portion of the GT should be approximately 1.5 cm (Fig. 9.6b). The distal end of the cut should not penetrate the lateral femoral cortex to avoid subtrochanteric notching. The plane of this cut should be in line with the true lateral (maximum profile) of the GT notwithstanding the rotation at the knee.
- Step 8: Cut the soft tissues around the GT: The soft tissues around the GT should be cut with a sharp serrated curved scissors. At the proximal end, insert one leg of the scissors under the gluteal flap and the other end in the osteotomy (Fig. 9.6d). Orient the curve of the scissors concave toward the femur, to allow them to conform to the hip capsule and go around the corner anteriorly (Fig. 9.6e). Make

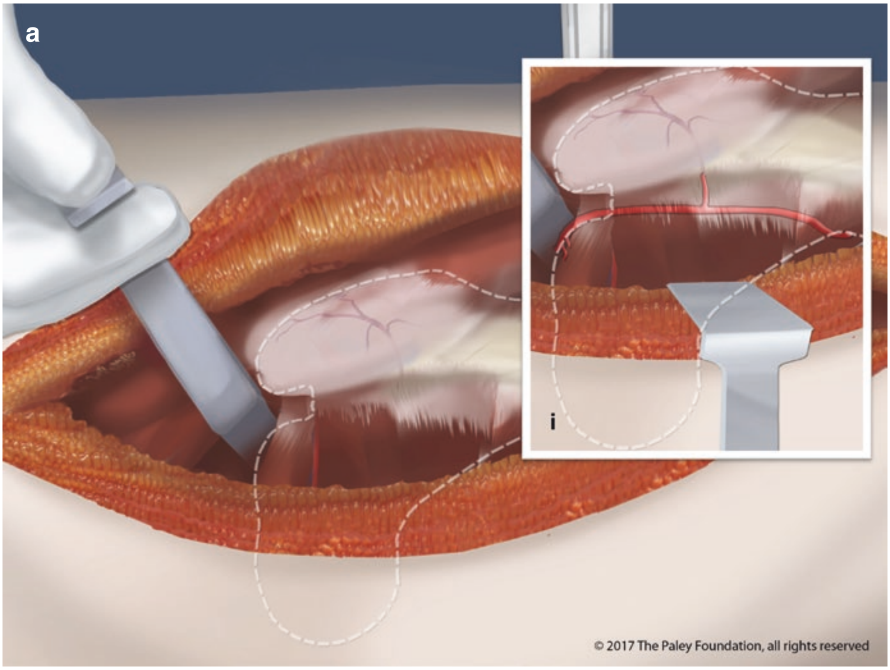
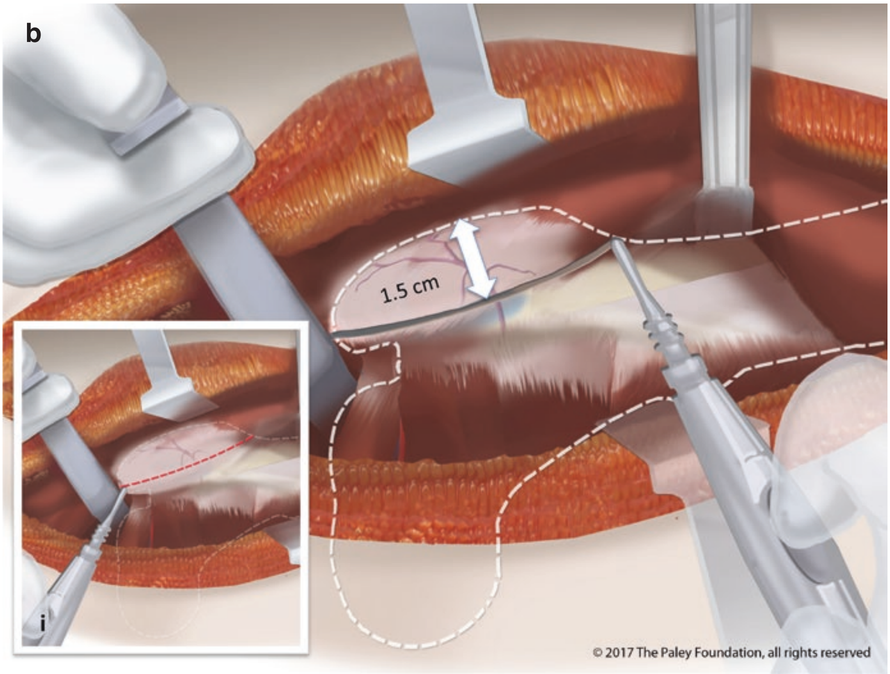


Fig. 9.6 (**a**) A lateral view of the greater trochanter is shown with the external rotator muscles inserting medial to the posterior trochanteric ridge. Underlying these running along the periosteum is the ascending branch of the MFCA (i) as it courses toward the piriformis fossa and anastomoses with the inferior gluteal artery. (**b**) The line for the trochanteric osteotomy is marked with a cautery. This line is defined along the trochanteric ridge between the insertion of the vastus lateralis distally and the Z retractor proximally. The trochanteric artery is intentionally cauterized. The thickness of the trochanteric segment is 1.5 cm. (**c**) Osteotomy of the greater trochanter is carried out with an oscillating saw. (**d**) The mobile trochanter is mobilized by cutting the soft tissues on its superior, medial, and inferior sides. At the upper end any tendinous extension from the piriformis is released. (**e**) The superomedial side of the trochanter is released. (**f**) The inferomedial side of the trochanter is released. ((C) Dror Paley. Used with permission)



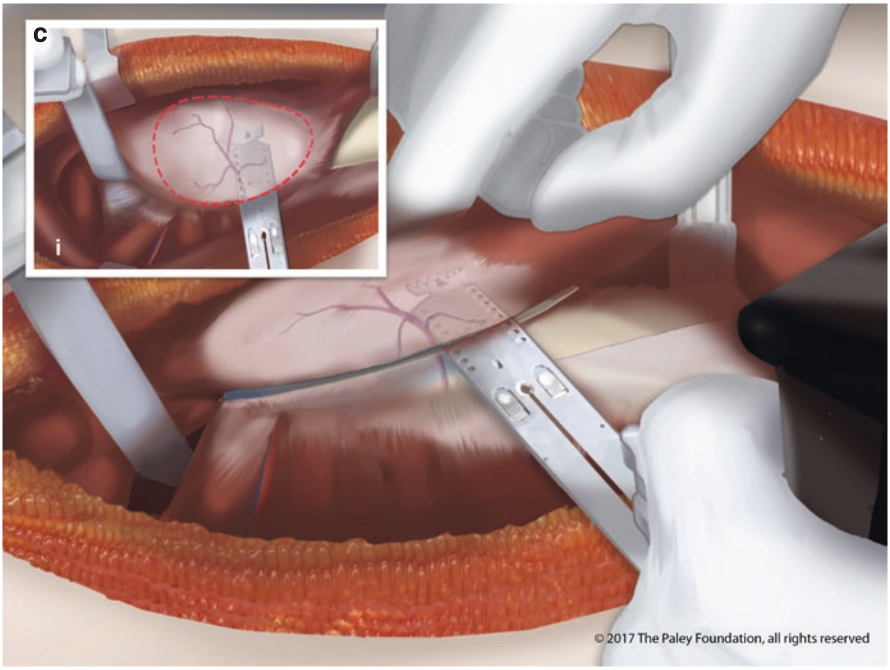


Fig. 9.6 (continued)

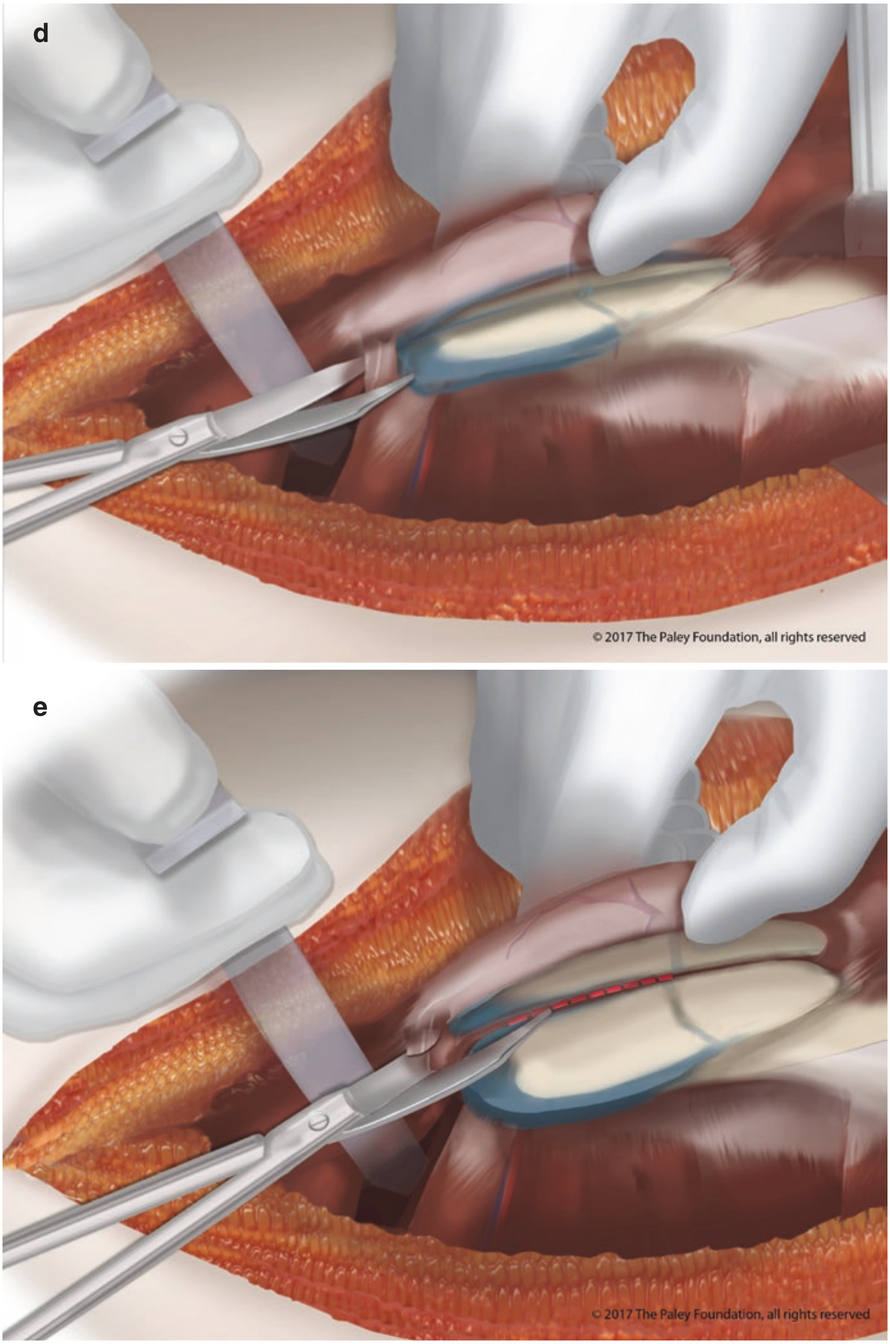


Fig. 9.6 (continued)

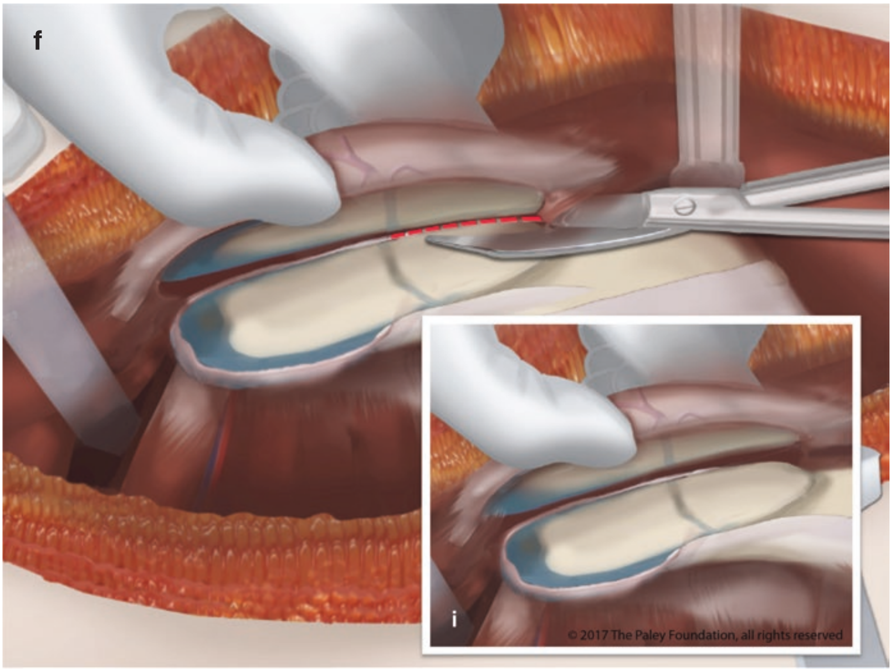


Fig. 9.6 (continued)

sure that the piriformis muscle remains posterior to the scissors. The piriformis tendon should remain connected to the stable portion of the GT with no more than a third of it still attached to the mobile GT (Fig. 9.6d). Release the part of the piriformis tendon connected to the mobile GT. Distally, do the same placing one leg of the scissors between the femur and the vastus and the other leg inside the osteotomy (Fig. 9.6f). The two cuts will converge on each other and free the GT osteotomy so it can be "flipped."

- Step 9: Insert 2 Hohmann elevators: Flex the hip and remove the bump from under the knee and allow the leg to adduct. Bluntly dissect under the muscle flap to the anterior inferior iliac spine (AIIS). Place a Hohmann elevator around the AIIS with care to stay between bone and muscle on the inside of the pelvis (Fig. 9.7a). Use a Cobb elevator to sweep the gluteus minimus muscle off of the capsule and lateral wall of the ilium. Impact a Hohmann elevator through the lateral wall of the ilium to retract the glutei proximally (Fig. 9.7b). These two elevators, which are 90 ° to each other, stay in place for the duration of the procedure.
- Step10: Z Capsulotomy: Make a Z-shaped capsulotomy. The intermediate limb of the Z (lateral to medial) is located at 11 o'clock for a left hip and 1 o'clock for a right hip with 12 o'clock being the top of the hip joint (Fig. 9.7c). Take care not to cut the labrum (Fig. 9.7d). Make one limb of the Z anterolateral and one limb

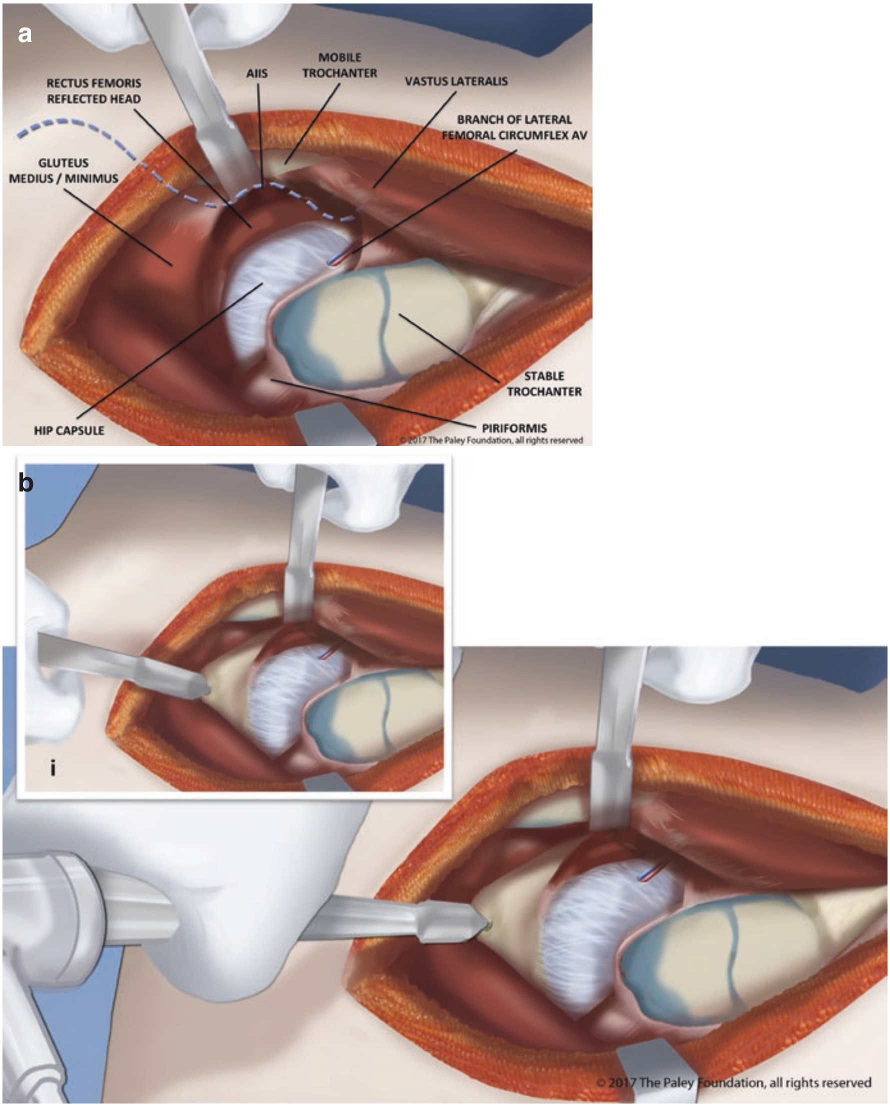


Fig. 9.7 (**a**) The hip is flexed and a Hohmann elevator is inserted around the anterior inferior iliac spine (AIIS). The branch of the lateral femoral circumflex artery should be spared if possible. (**b**) The gluteus minimus is elevated off of the capsule and lateral ilium. A Hohmann elevator is impacted into the lateral ilium to help retract the soft tissues. The two Hohmann's are 90 ° to each other and should be retracted separately with separate hands in two different directions for best exposure. (**c**) The Z-shaped capsulotomy has the distal arm lateral and the proximal arm medial. It is shown for right and left hips. (**d**) The anterior part of the capsulotomy. (**e**) After the entire capsulotomy is completed, the hip can start to be dislocated and the ligamentum teres cut with curved right angle scissors (i). ((C) Dror Paley. Used with permission)

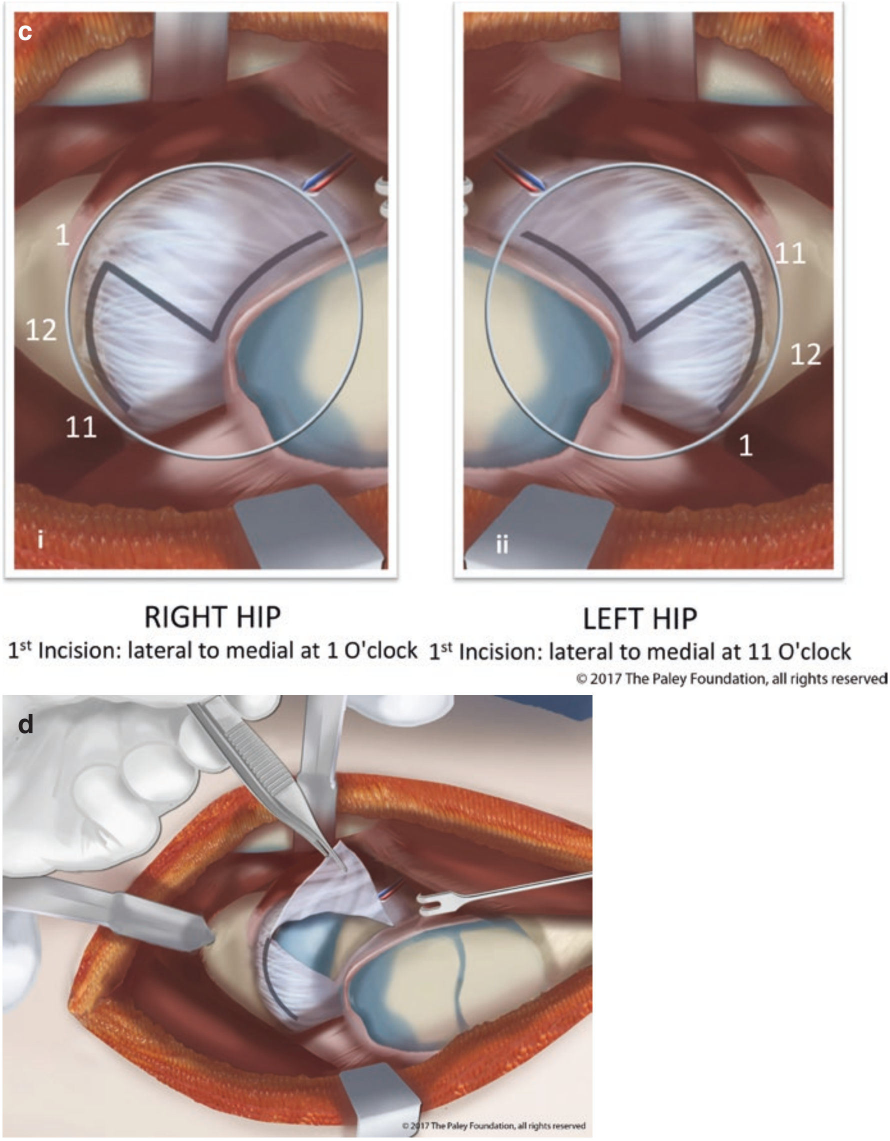


Fig. 9.7 (continued)

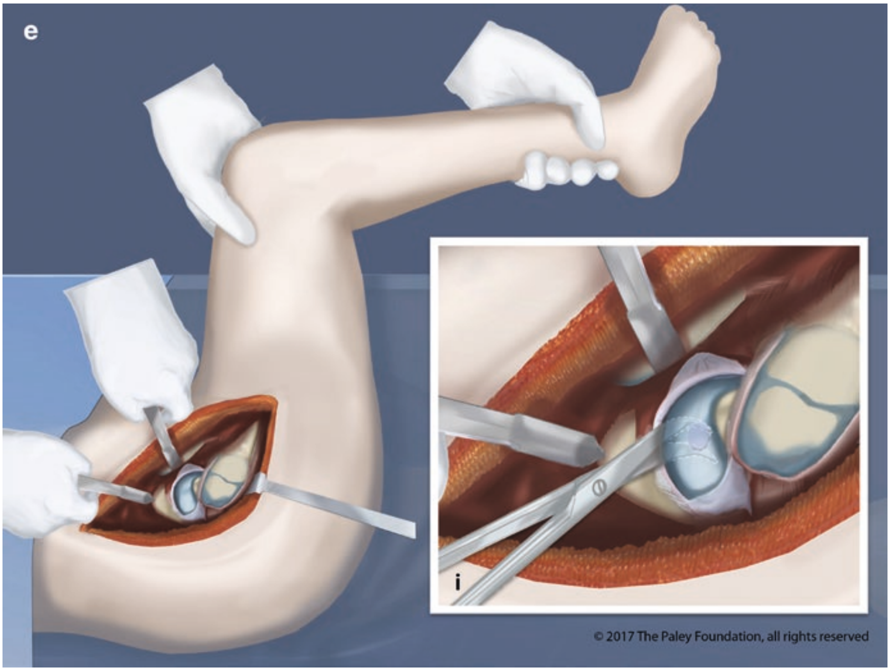


Fig. 9.7 (continued)

posterior-medial. Leave a cuff of capsule at both limbs in order to sew into and also to preserve the relationship of the capsule to the labrum.

- Step 11: Cut ligamentum teres: To be able to dislocate the hip cut the ligamentum teres with a long curved scissors (e.g., Jorgenson's) (Fig. 9.7e).
- Step 12: Dislocate the hip: Flex and externally rotate the hip to dislocate it (Fig. 9.8).
- Step 13: Resect the ligamentum teres: resect the remnant of the ligamentum both in the acetabulum and from the femoral head. Clean out the cotyloid notch from soft tissues (Fig. 9.8).
- Step 14: Inspect and repair the labrum: Inspect the labrum with a blunt hook to look for a partial or complete labral tear. If the labrum is torn resect and repair according to the pattern of the tear. Use suture anchors in the acetabular rim to anchor the labrum back to bone
- Step 15: Measure the size of the acetabulum: Use spherical acetabular sizers to measure the diameter of the acetabulum (Fig. 9.9a).

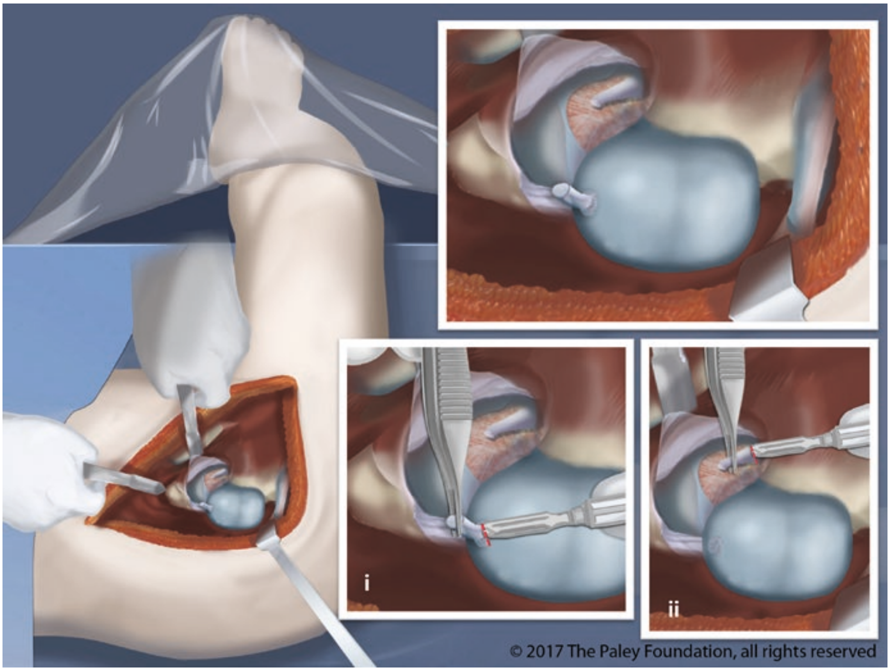


Fig. 9.8 The leg is crossed over the opposite leg and dropped into a sterile bag. The hip is now dislocated. The ligamentum teres and any pulvinar can be excised. The labrum can be probed for tears. ((C) Dror Paley. Used with permission)

- Step 16: Size the femoral head: Use femoral head spherical templates to assess the sphericity of different parts of the femoral head (Fig. 9.9b, c), starting with a template corresponding to the acetabular template. The femoral head can be sized for its curvature in different planes. Mark the medial femoral head where the sizer leaves the round. Do the same for the lateral portion of the femoral head. Using this method the femoral head can be mapped for its sphericity.
- Step 17: Retinacular flap: Reduce the hip back into joint. Do a subperiosteal dissection of the posterior femur starting at the level of the lesser trochanter and working proximally up to the level of the piriformis fossa (Fig. 9.10). The retinacular vessels are elevated with the periosteum.
- Step 18: Mark the vertical osteotomy lines: Redislocate the hip and mark the osteotomy lines. There are two osteotomy patterns: Ganz-parallel to the femoral neck and (Fig. 9.2a); Paley (Fig. 9.2b)- perpendicular to the femoral head. Furthermore there are two types of osteotomy pairings: (A) parallel cuts (Fig. 9.11a) and (B) non-parallel cuts (Fig. 9.11b). These are illustrated for the Paley type but not for the Ganz type since the former is the more common deformity pattern. For both A and B types the final diameter of the femoral head is the same and should correspond to the template diameter of the acetabulum.

- Step 19: Mark the horizontal osteotomy line: Mark this line distal enough to include the anterior part of the piriformis fossa but not too deep to narrow the femoral neck too much (Fig. 9.11a, b).
- Step 20: Make the vertical osteotomies (Figs. 9.12a–e and 9.13a–e): Use a thin saw to make the horizontal cut first and then make the vertical cuts. Do not cut all the way to the posterior head/neck. After making most of the cut with the saw, complete the posterior part of the osteotomy with an osteotome. Crack the back of the osteotomy to avoid disrupting any retinacular vessels that may be attached.
- Step 21: Remove the intercalary segment: Remove the middle segment created by the osteotomies (Fig. 9.12e and 9.13e).
- Step 22: Mobilize the lateral segment: Mobilize the lateral segment by lifting it from anterior to posterior hinging on the retinacular flap (Fig. 9.14a). Carefully strip the retinaculum off of the neck to remain on the mobile lateral segment. This allows it to move from lateral to medial to close the defect without tethering or tearing the retinaculum (Fig. 9.14b).

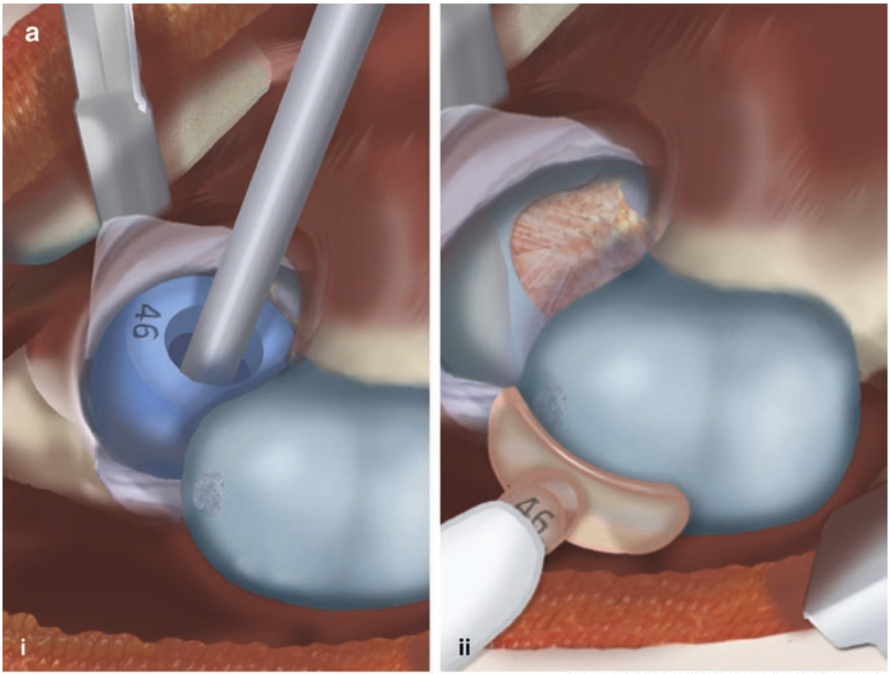
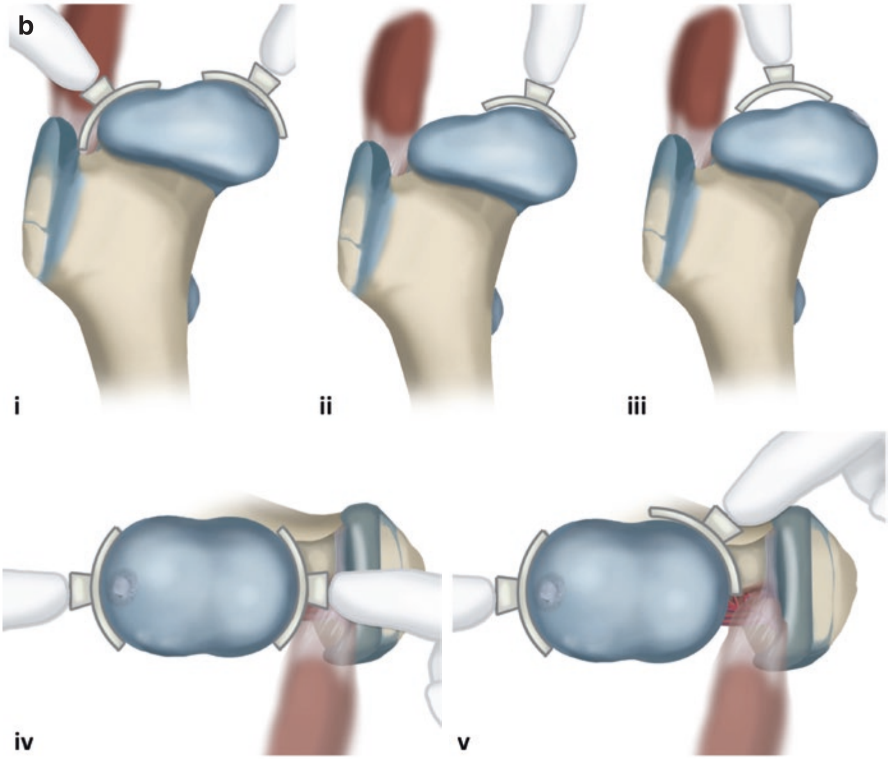


Fig. 9.9 (**a**) The acetabulum is sized (i). A femoral head template of the same size as the acetabulum (in this case 46 mm) is used to check the sphericity of the medial femoral head. (**b**) The femoral head is template along its medial and lateral sides as well as along its medial and lateral superior surfaces. It is important to note where the femoral head leaves the round. This is shown for Type A. (**c**) The same is done for a Type B case. Note the anterior bump and incongruity. ((C) Dror Paley. Used with permission)



TYPE A

Fig. 9.9 (continued)

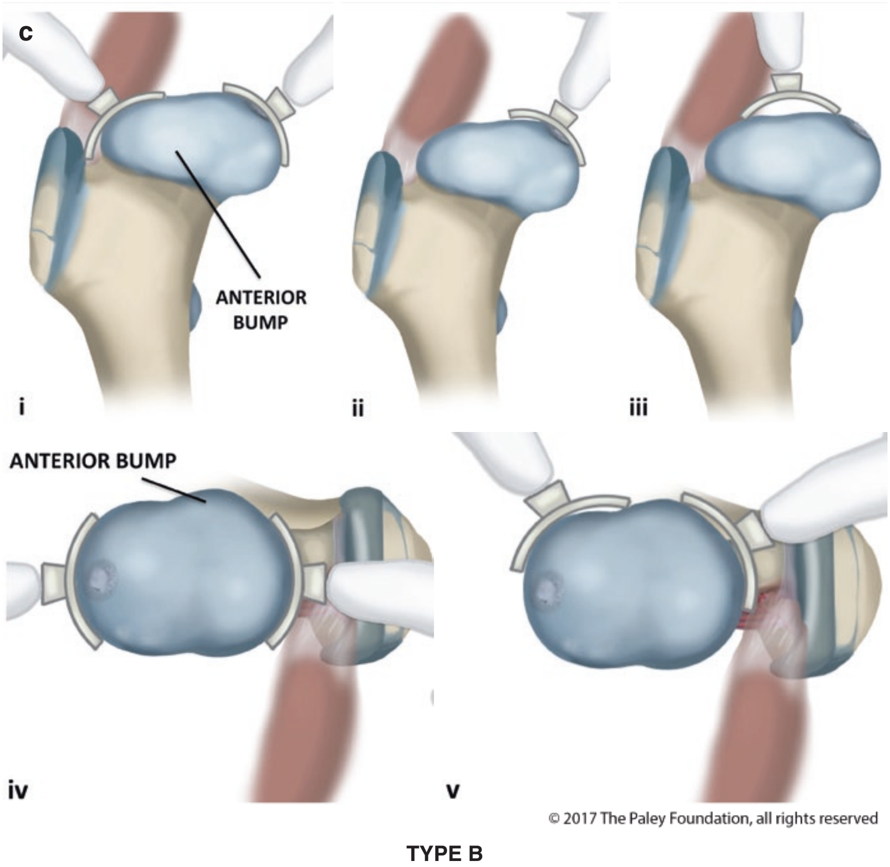


Fig. 9.9 (continued)

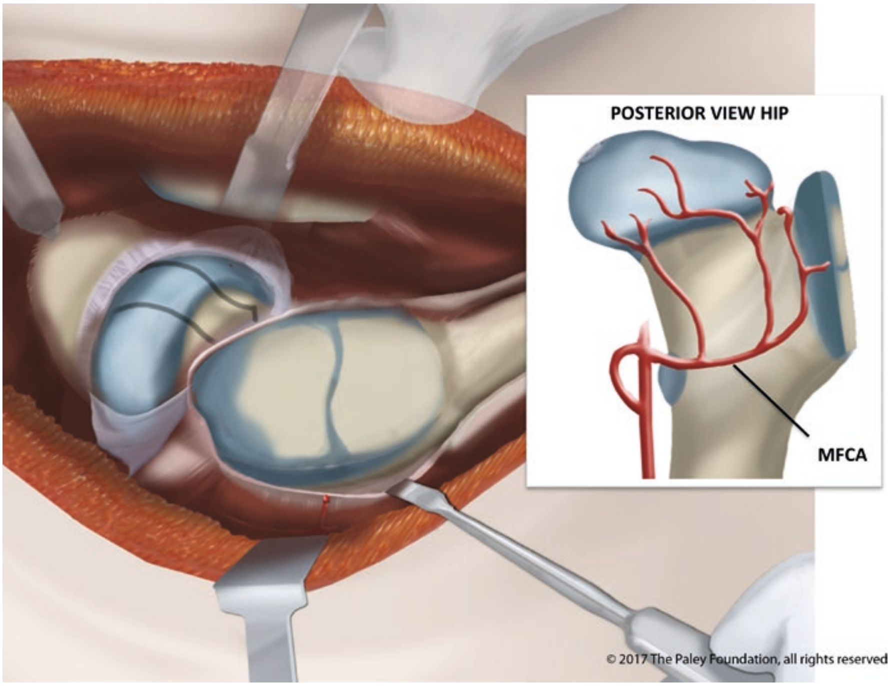


Fig. 9.10 The hip is reduced into joint again and the periosteum at the back of the femur is stripped carefully and extended proximally to help separate the retinaculum from the lateral femur. This moves the MFCA away from the lateral femur. ((C) Dror Paley. Used with permission)

- Step 23: Reduce the femoral head: Adjust the lateral segment so that the posterior congruity is as perfect as possible (Fig. 9.14b). If there is a step, make sure it is anterior.
- Step 24: Fix the femoral head with headless screws: Fix the segments with two parallel k-wires and then measure and drill over these and insert two parallel variable pitch headless screws(Fig. 9.15a). The variable pitch thread compresses the intra-articular osteotomy line.
- Step 25: Insert a third headless screw perpendicular to the osteotomy at the base of the lateral fragment: Drill a wire, cannulated drill, measure, and then insert a headless screw (Fig. 9.15b).
- Step 26: Osteochondroplasty: Perform an osteochondroplasty on any incongruous anterior aspects of the femoral head to avoid impingement and to eliminate any incongruity from the reduction.
- Step 27: Insert prophylactic screw for femoral neck: Drill a guide wire from the fovea retrograde into the femoral neck through the lateral femoral cortex. Measure and insert an antegrade cannulated screw up the femoral neck (Fig. 9.15c, d).

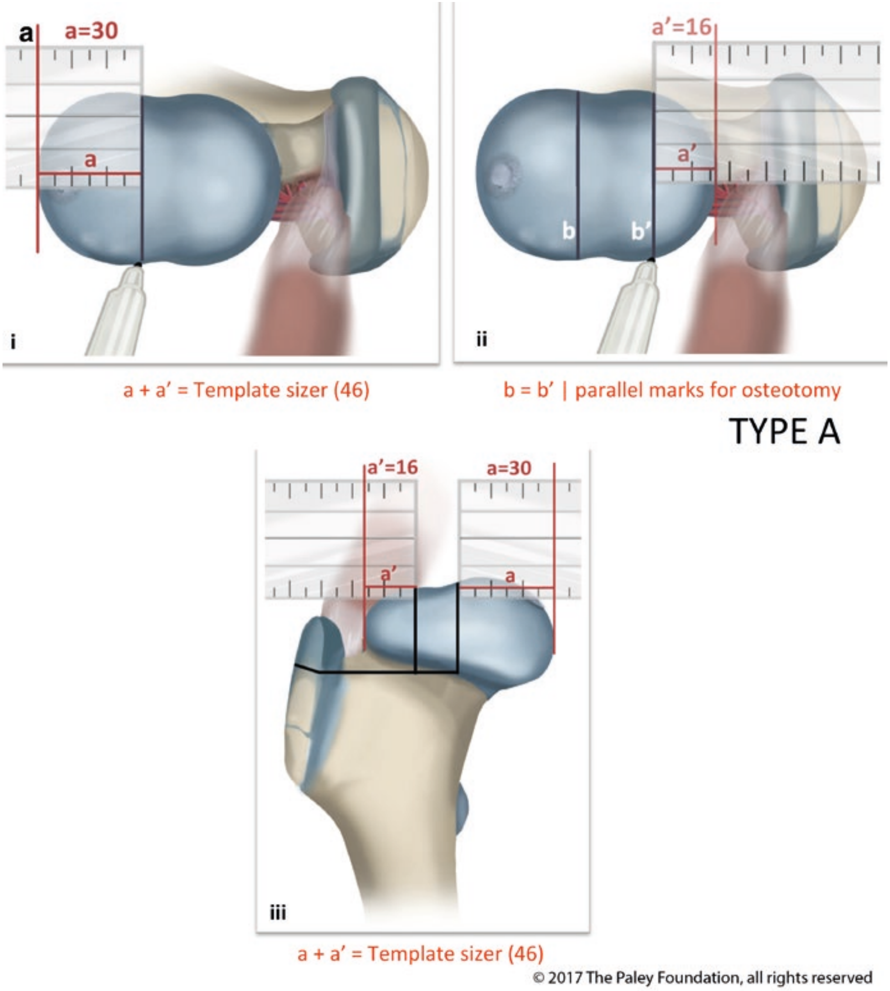


Fig. 9.11 (**a**) The femoral head is marked at the borders of where it leaves the round based on the femoral head spherical templates. Since the medial part measures 30 mm and since the total diameter of the reduced femoral head is 46 mm, then the lateral segment should be 16 mm. Since the coxa magna is Type A, the planned osteotomy lines are parallel. The baseline runs just below the ridge of the femoral head and distal to the anterior piriformis fossa. It is still superior to the most posterior piriformis fossa. The stable trochanter will be part of the lateral fragment. (**b**) For Type B the two lines are convergent posteriorly. The dimensions of the medial and lateral parts of the femoral head are the same as for Type A and add up to a 46 mm diameter femoral head. ((C) Dror Paley. Used with permission)

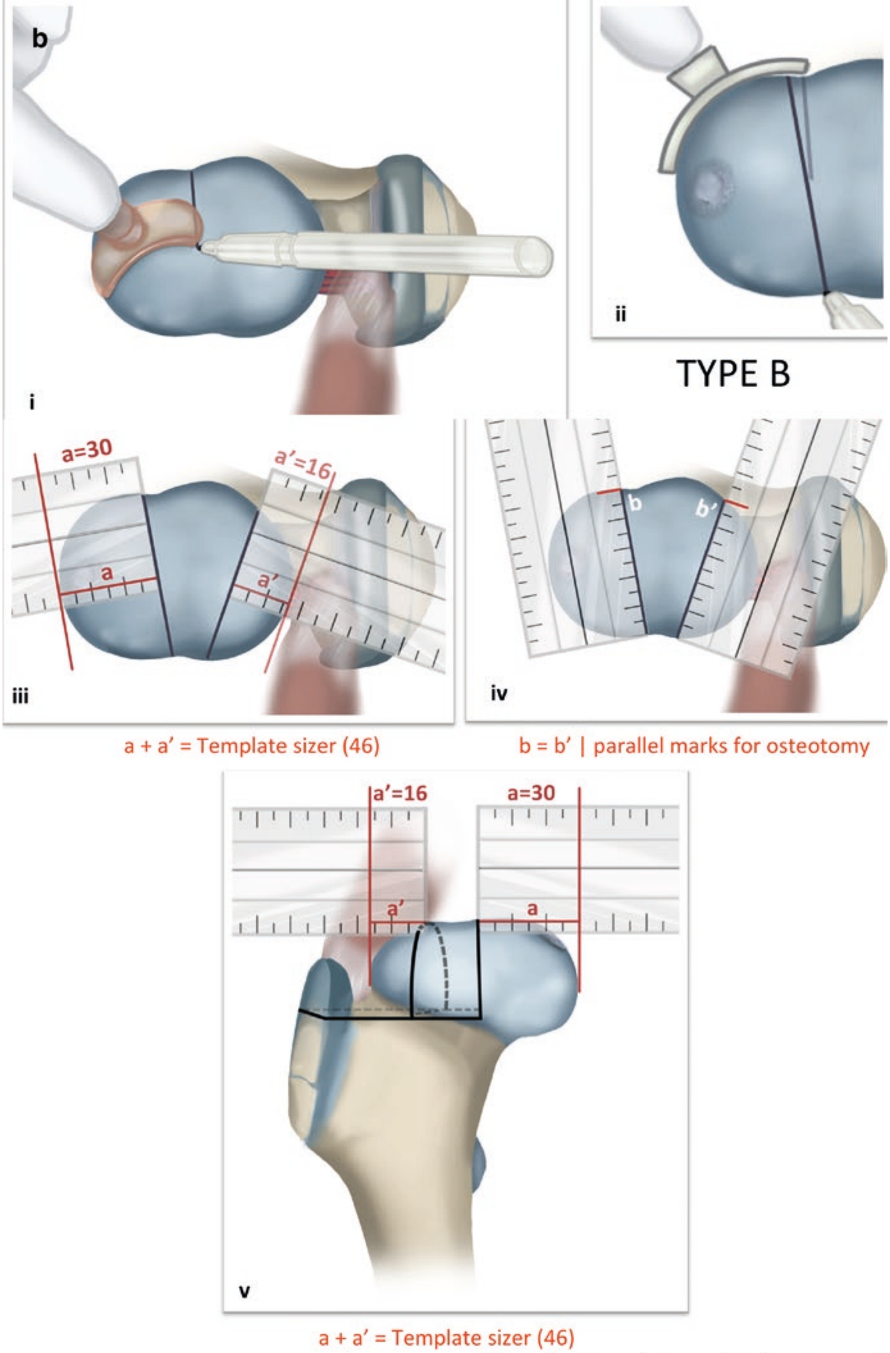
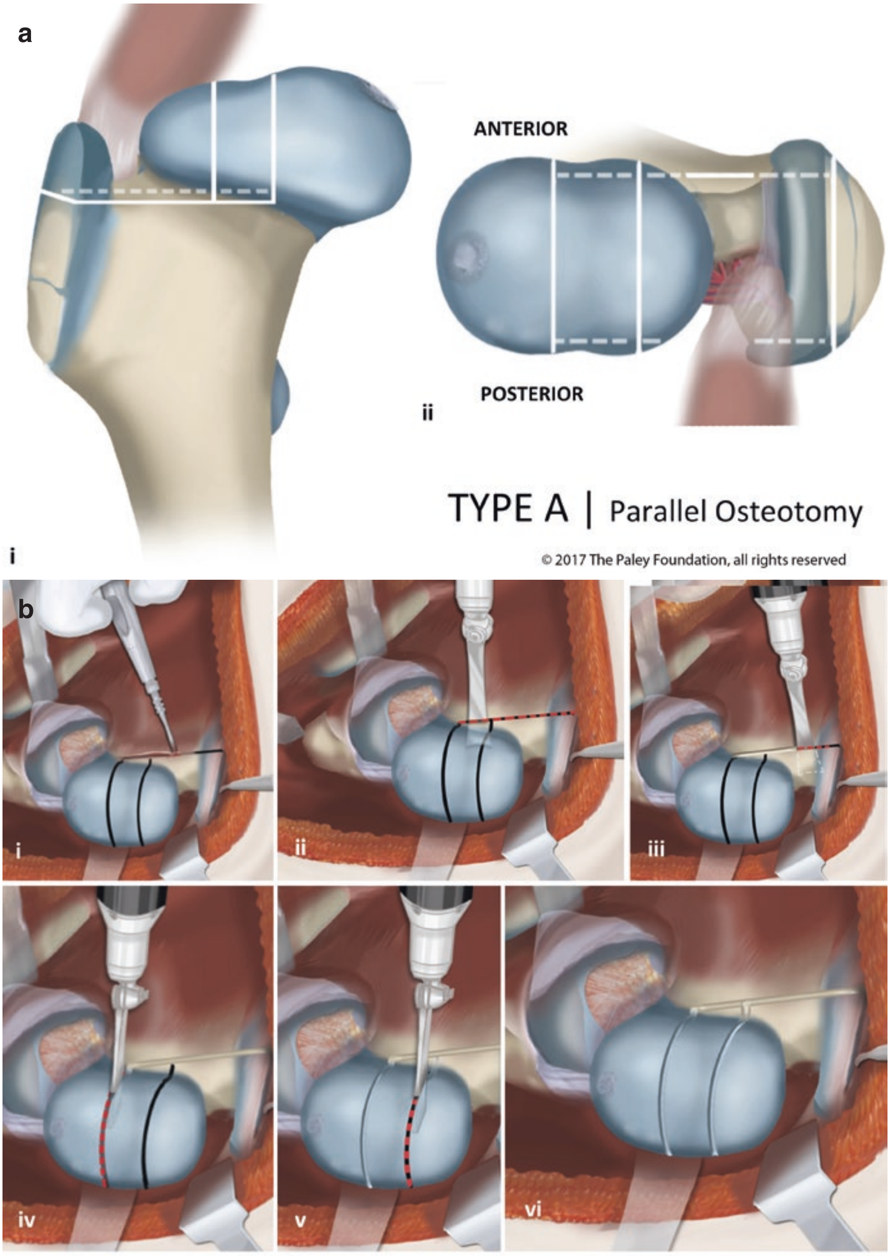


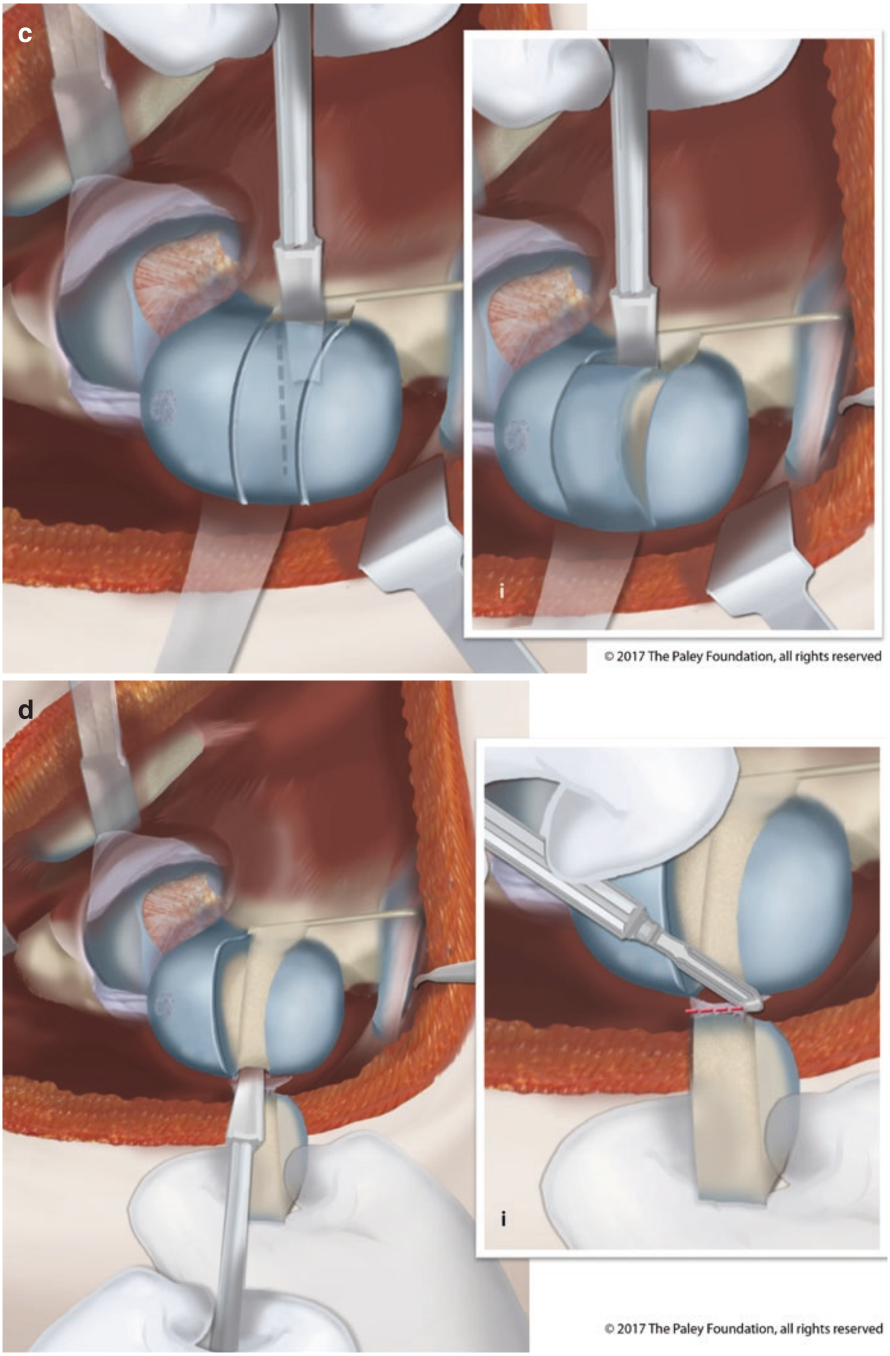
Fig. 9.11 (continued)



© 2017 The Paley Foundation, all rights reserved

© 2017 The Paley Foundation, all rights reserved

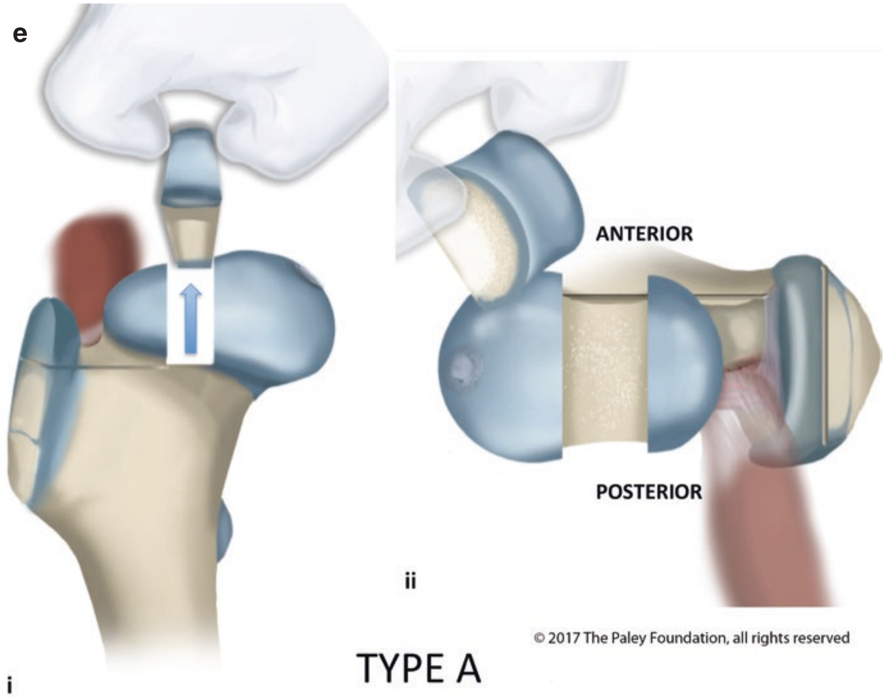
Fig. 9.12 (**a**) The osteotomy lines are outlined in white; parallel cuts. (**b**) The cautery is used to mark the anterior baseline cut (i). A very thin saw is then used to make the horizontal baseline cut (ii, iii) and then the two parallel vertical cuts (iv–vi). (**c**) An osteotome is used to complete the posterior aspect of the osteotomies and to pry out the intercalary segment (i). (**d**) The fragment is sharply dissected off of any posterior retinacular tissues. (**e**) The intercalary middle segment is removed. ((C) Dror Paley. Used with permission)



© 2017 The Paley Foundation, all rights reserved

© 2017 The Paley Foundation, all rights reserved

Fig. 9.12 (continued)



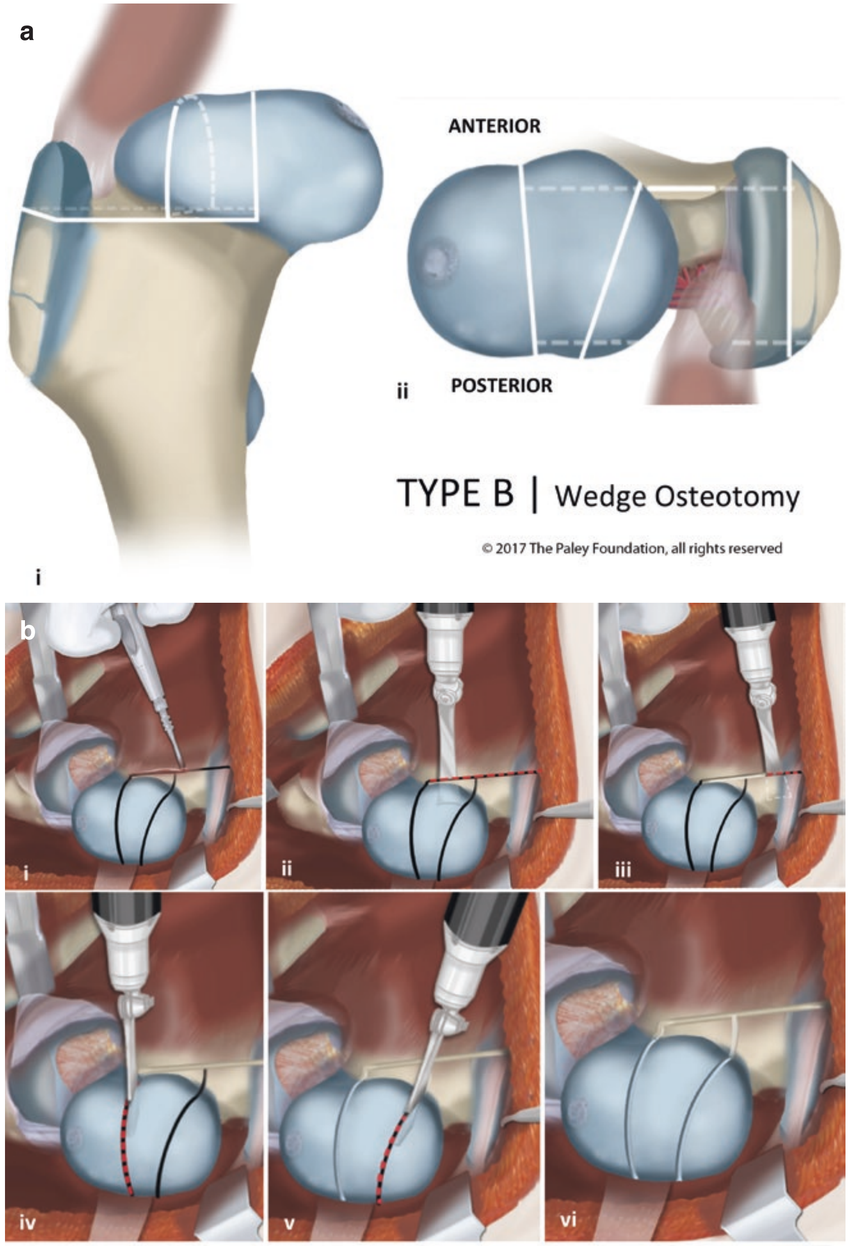
© 2017 The Paley Foundation, all rights reserved

Fig. 9.12 (continued)

- Step 28: Resect the stable trochanter: Reduce the femoral head into joint. In this position, resect the stable trochanter with great care not to damage the retinacular flap (Fig. 9.16a, b).
- Step 29: Capsular repair: Repair the Z capsulotomy taking up some of the redundancy to prevent subluxation (Fig. 9.17a).
- Step 30: Transfer the mobile trochanter: Fix the mobile trochanter into place more distal and lateral than before. Aim to lower the tip of the greater trochanter to the level of the center of the femoral head. Insert two 3.2 mm drill bits and fix the trochanter at the desired level (Fig. 9.17b). Use another same length drill bit, to measure the length of each screw and then replace each drill bit with a 4.5 mm screw with washer (Fig. 9.17c).

Postoperative Care

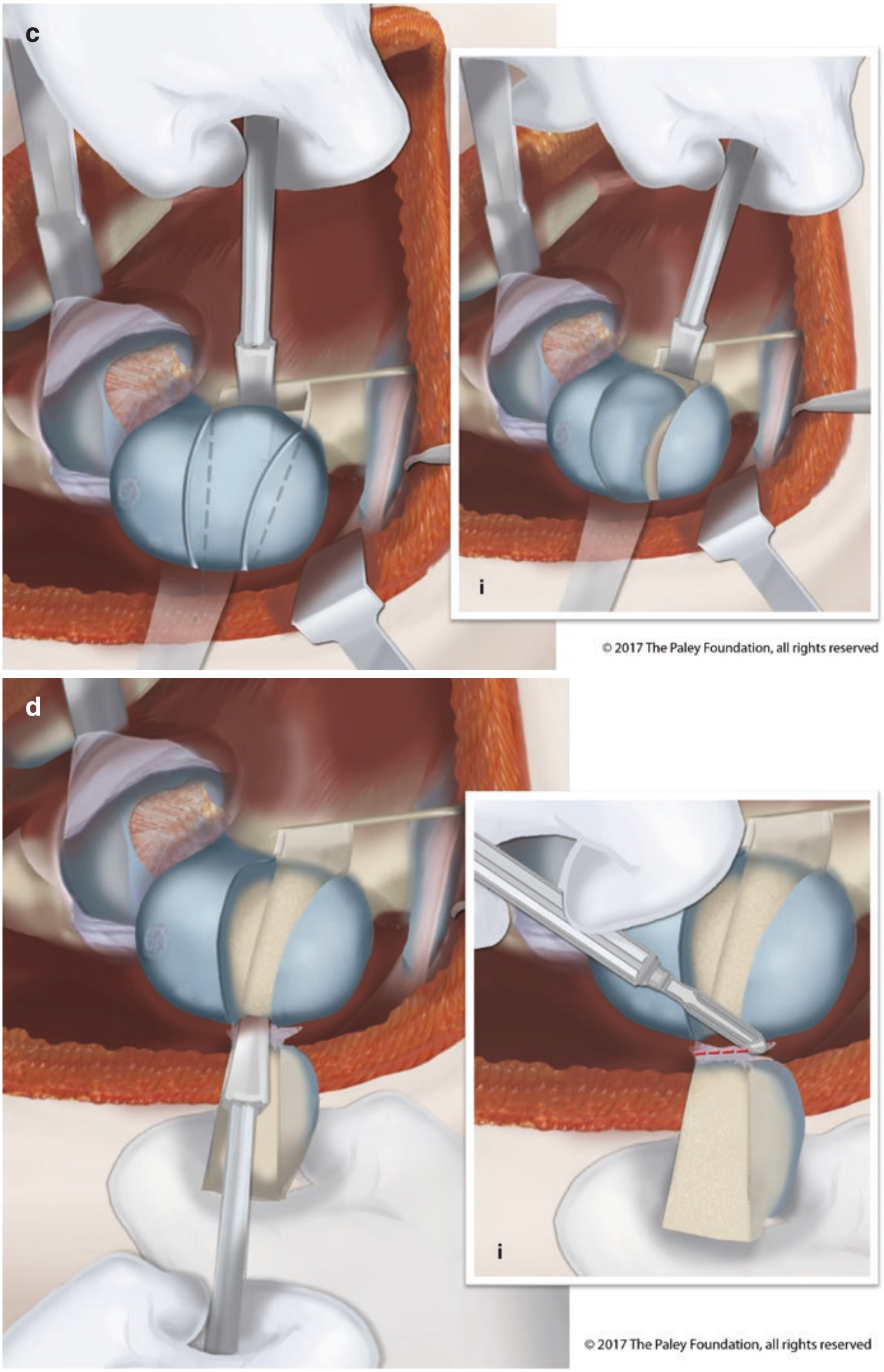
Place the operative leg on a CPM machine in the recovery room. The patient stays on CPM for 20 hours a day for about a week. After that they use CPM half the day on and half the day off and all night long for five more weeks. When ambulating the patient uses a unilateral hip 5 ° hip abduction brace. Physical therapy is instituted allowing only passive hip abduction and flexion. No external rotation or adduction



© 2017 The Paley Foundation, all rights reserved

© 2017 The Paley Foundation, all rights reserved

Fig. 9.13 (**a**) The osteotomy lines are outlined in white; nonparallel cuts. (**b**) The cautery is used to mark the anterior baseline cut (i). A very thin saw is then used to make the horizontal baseline cut (ii, iii) and then the two nonparallel vertical cuts (iv–vi). (**c**) An osteotome is used to complete the posterior aspect of the osteotomies and to pry out the intercalary segment (i). (**d**) The fragment is sharply dissected off of any posterior retinacular tissues. (**e**) The intercalary wedge shaped middle segment is removed. This removes the anterior bump with it. ((C) Dror Paley. Used with permission)



© 2017 The Paley Foundation, all rights reserved

© 2017 The Paley Foundation, all rights reserved

Fig. 9.13 (continued)

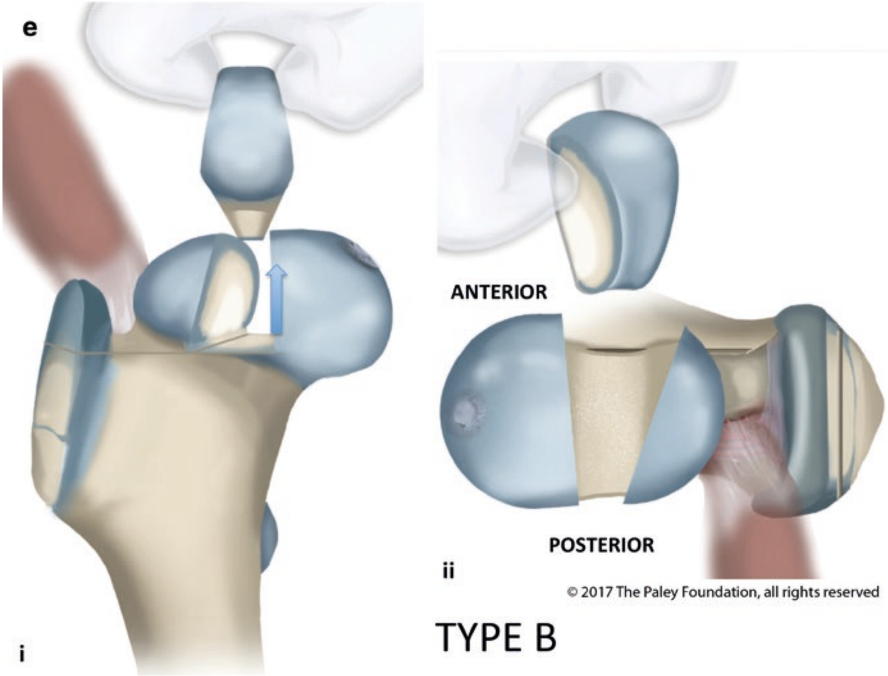


Fig. 9.13 (continued)

past the midline is permitted for 6 weeks. The patient is allowed touch down weight bearing for 12 weeks using a walker or crutches. Active assisted and active range of motion is permitted between 6 and 12 weeks. Muscle strengthening and full weight bearing without crutches begins after 12 weeks.

Postoperative Considerations

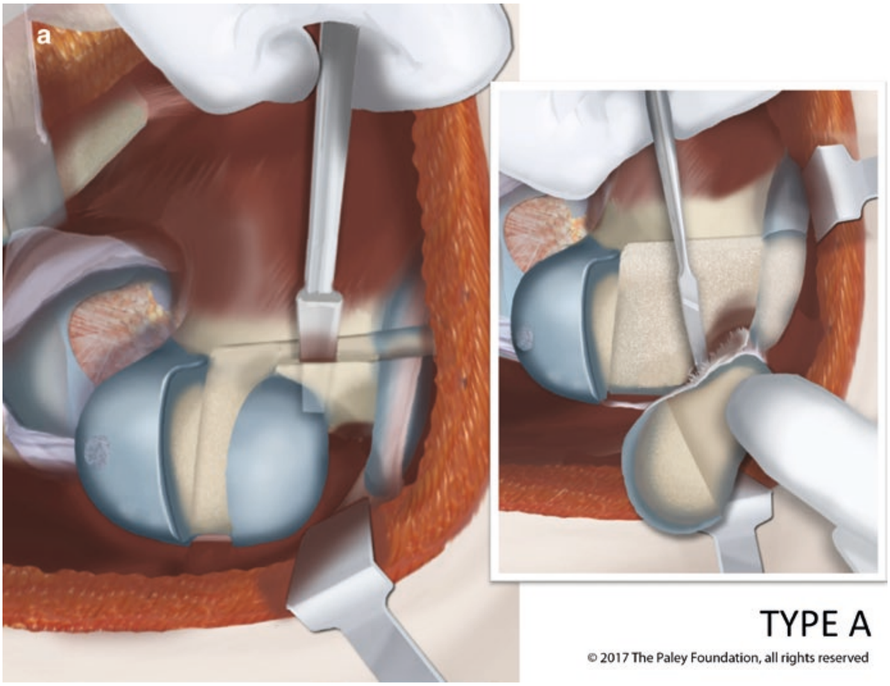
After healing of the FHRO, the hip should be assessed for the need for a periacetabular osteotomy (PAO). The need for a PAO may be obvious or subtle after the FHRO. It is not recommended to do the PAO at the time of the FHRO unless there is gross instability. In most cases the PAO is performed six or more months after the FHRO.

Leg length discrepancy can be treated by contralateral femoral epiphysiodesis in skeletally immature patients. Lengthening using an implantable lengthening rod is another option preferably after skeletal maturity.

Case Examples

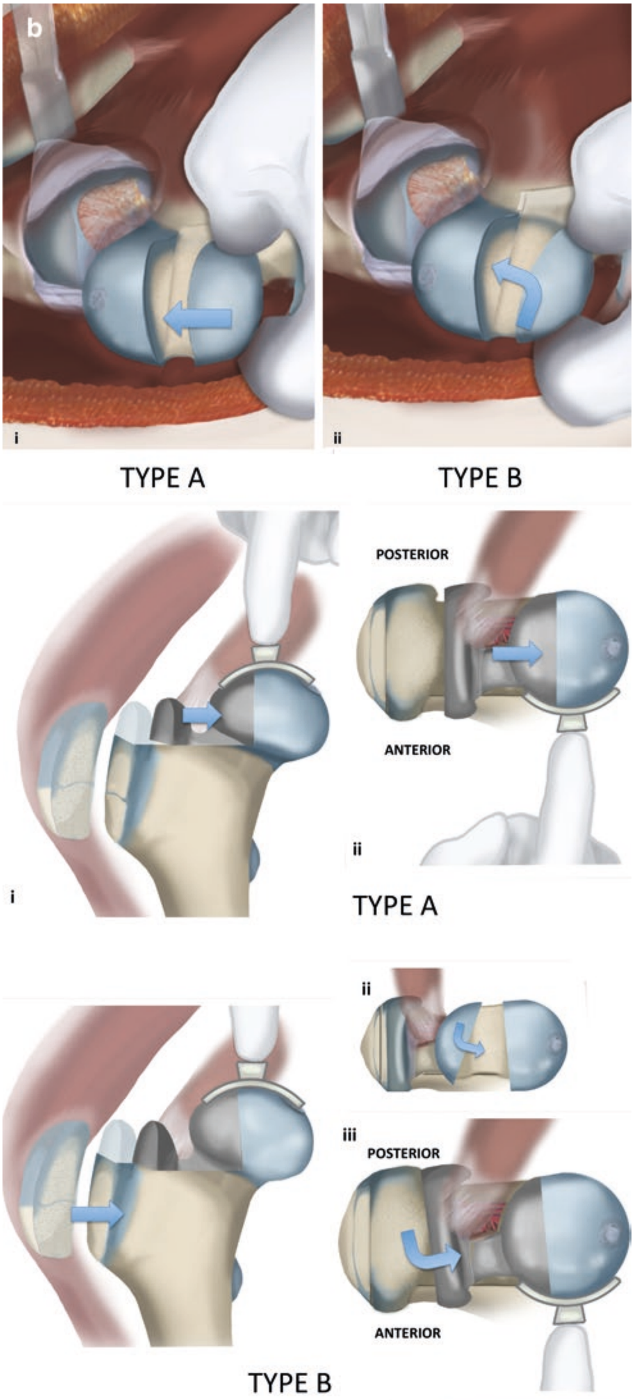
Patient 1 (Fig. 9.18): An 11-year-old girl with history of left sided Perthes disease since age 6. Previously treated by range of motion but no bracing or surgery. Pre-op range of motion was 0–90 ° flexion, no internal rotation and 30 ° external rotation, abduction 10 ° with pain. She had a limp with obvious lurch to the left.

Femoral head was elongated horizontally, not perpendicular to the femoral neck. A Ganz-type FHRO was performed as one of the author's first such operations in 2007. Osteochondroplasty was also carried out. The femoral head impingement was easily seen. The cartilage outside the acetabulum was well preserved, and all of the degenerative changes corresponded to impingement with the acetabular rim. After the FHRO the reduced hip did not impinge. Her femoral head shape changed from a Stulberg class IV to Stulberg Class I after surgery. Postoperative radiographs did not deteriorate even 10 years after the original surgery. While currently a Paley type



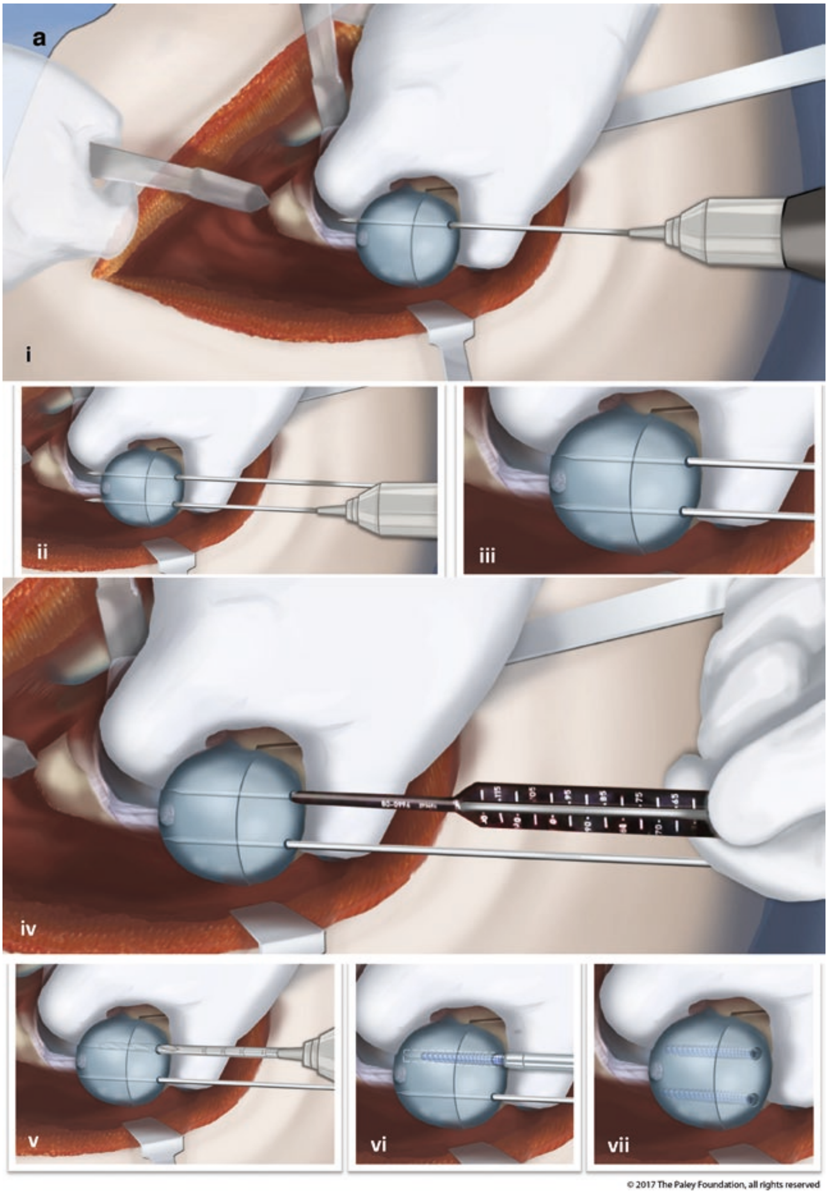
© 2017 The Paley Foundation, all rights reserved

Fig. 9.14 (**a**) The lateral segment is mobilized posteriorly to connect this dissection with the original retinacular flap dissection. (**b**) The untethered lateral fragment can now be moved medially without putting tension on or tearing the retinacular flap. The trochanter moves medially with the segment which maintains the stability of the retinacular flap. This is shown for both Type A (upper) and Type B (lower). ((C) Dror Paley. Used with permission)



© 2017 The Paley Foundation, all rights reserved

Fig. 9.14 (continued)



© 2017 The Paley Foundation, all rights reserved

Fig. 9.15 (**a**) The lateral mobile femoral head is reduced to the stable medial femoral head. Priority should be given to matching the posterior aspects of the two parts since the anterior part can be shaved off with the osteochondroplasty method. Two wires are inserted in parallel to each other and perpendicular to the osteotomy line. After measuring and drilling, two variable pitch headless screws are inserted into the femoral head from lateral to medial. (**b**) A third headless screw is drilled and inserted perpendicular to the baseline osteotomy. (**c**) A wire is drilled from the fovea down the femoral neck and out the lateral side of the femur. A hole is drilled for a cannulated screw with a cannulated drill. (**d**) A cannulated screw is inserted to prevent fracture of the femoral neck. ((C) Dror Paley. Used with permission)

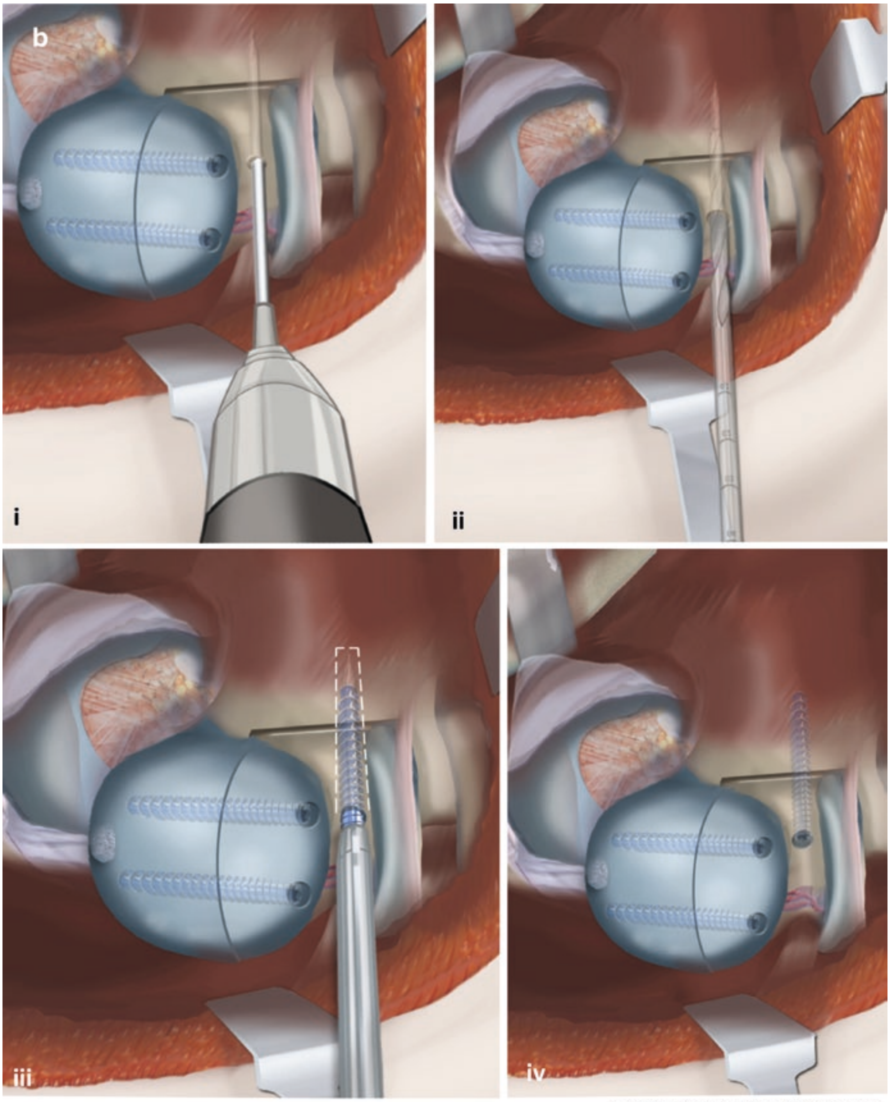


Fig. 9.15 (continued)

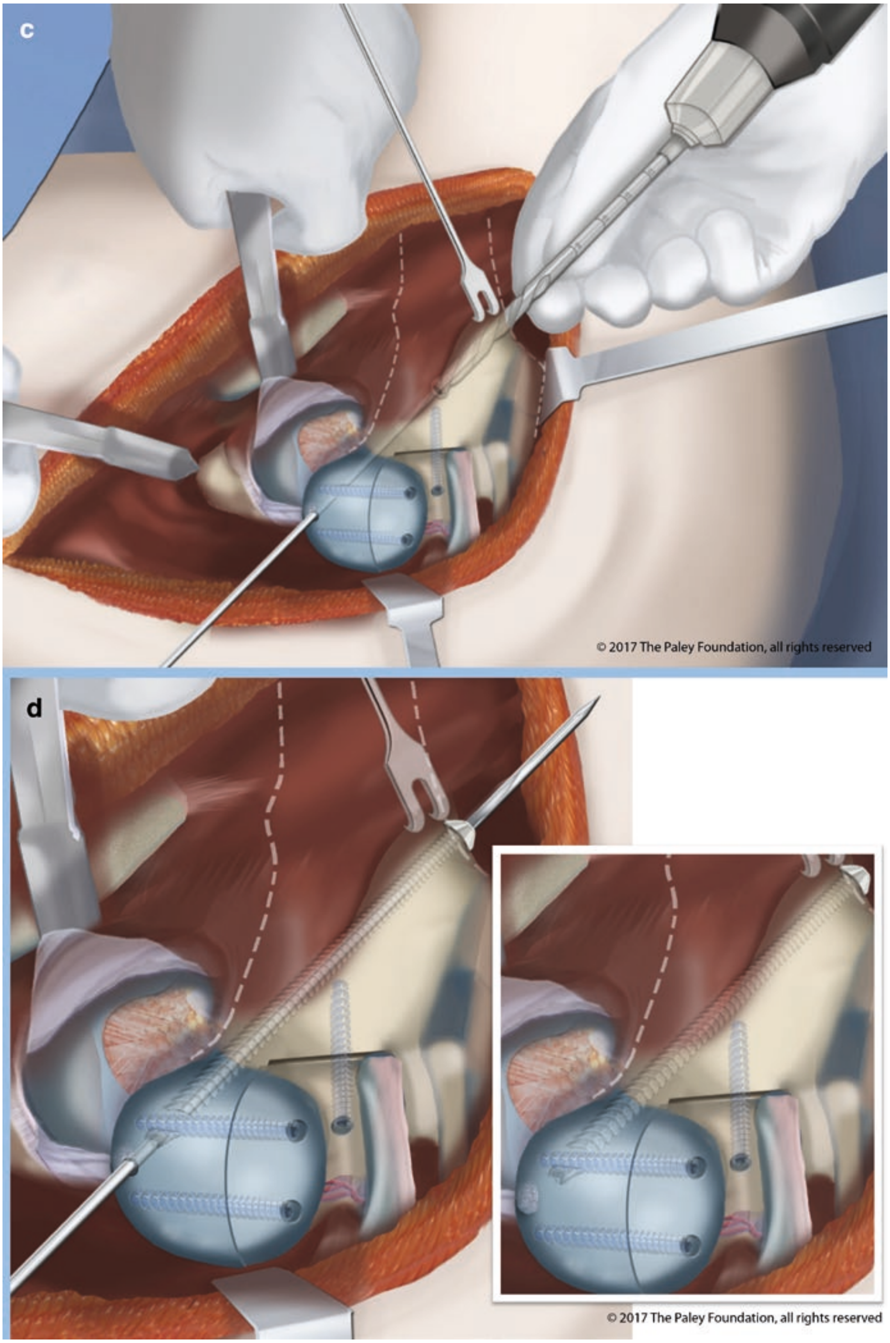


Fig. 9.15 (continued)

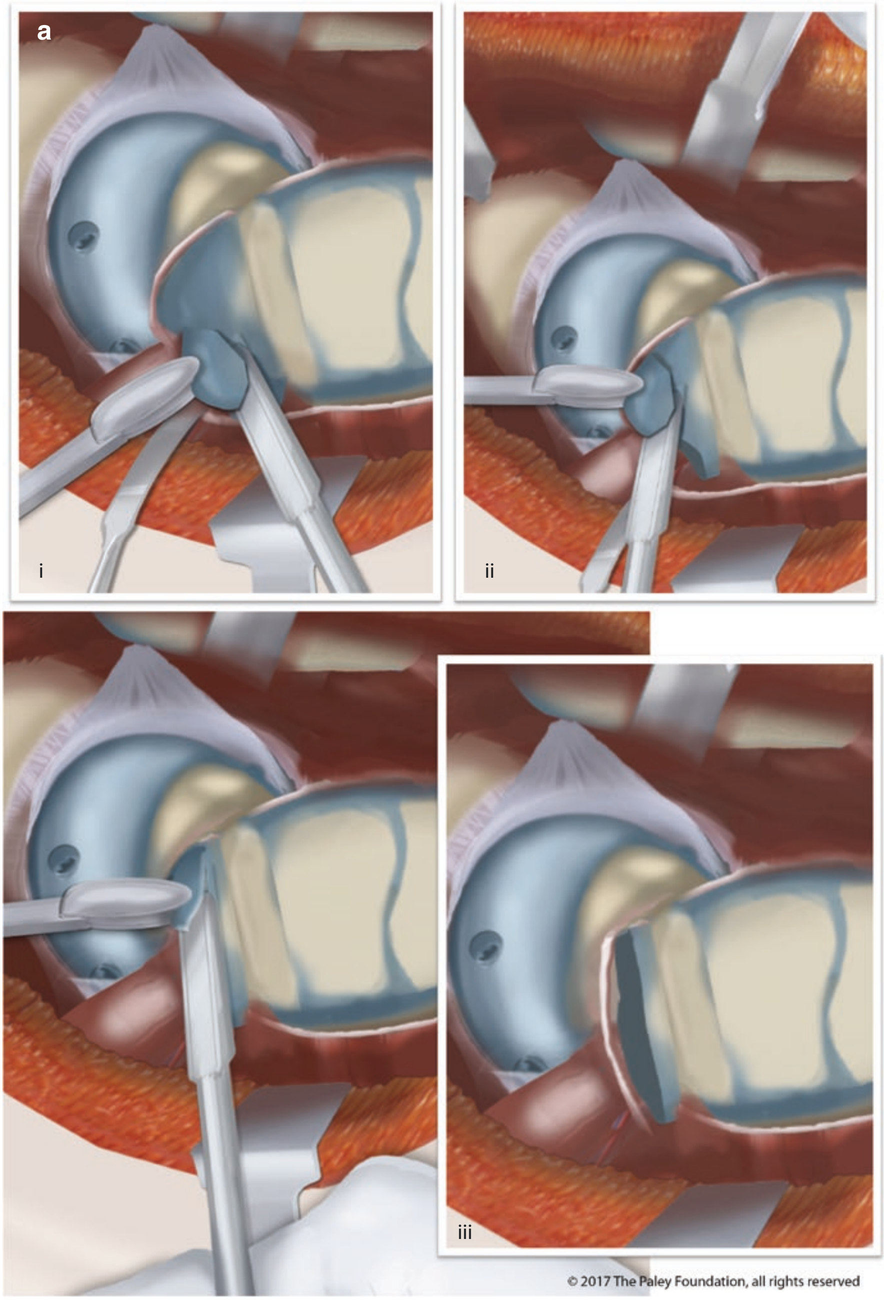


Fig. 9.16 (**a**) The femoral head is reduced into joint. The stable trochanter is excised carefully so as not to injure the retinacular vessels. (**b**) A superior view shows the femoral head mostly covered by the acetabulum after the FHRO. ((C) Dror Paley. Used with permission)

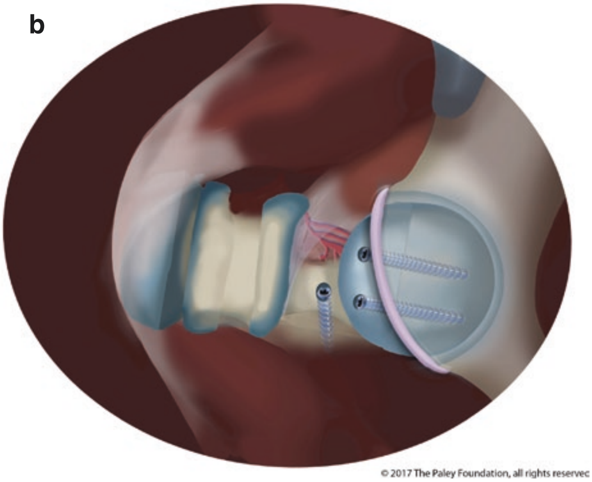


Fig. 9.16 (continued)

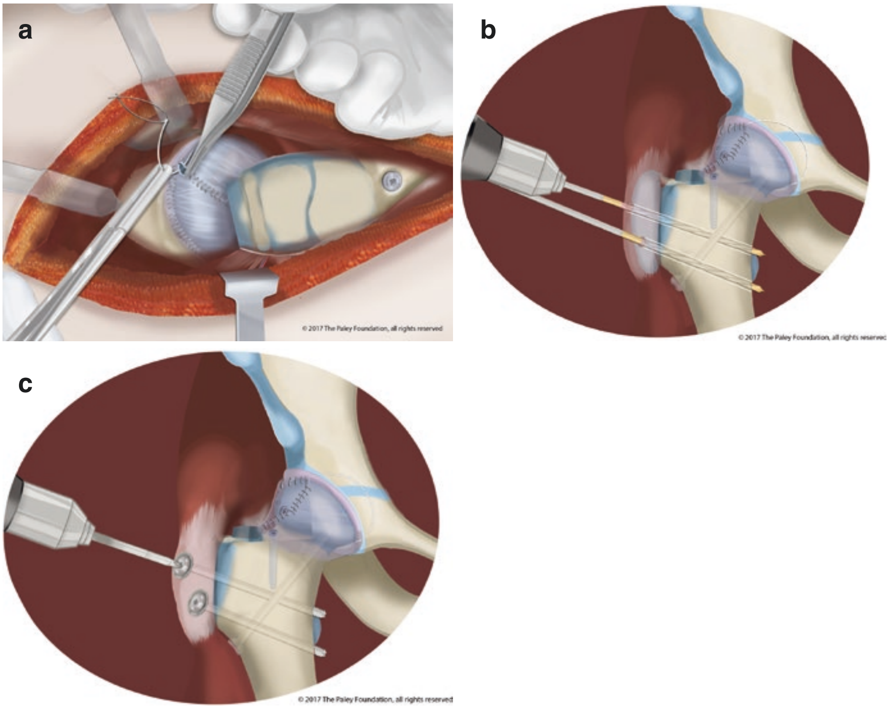


Fig. 9.17 (**a**) The capsule is closed and any redundant capsule is advanced to maintain joint stability. (**b**) The greater trochanter is advanced laterally and distally and then fixed with two 3.2 mm drill bits. The tip of the greater trochanter should be at the level of the center of the femoral head. (**c**) The drill bits are replaced with two 4.5 mm screws with washers. ((C) Dror Paley. Used with permission)

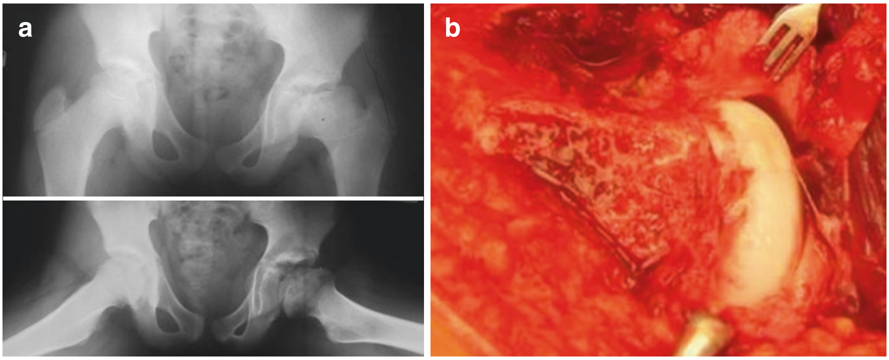


Fig. 9.18 (**a**) AP and frog lateral radiographs of pelvis of an 11-year-old girl with left coxa magna, coxa breva, coxa vara, and overgrown trochanter sequelae from previous Perthes. The elongation of the femoral head is not perpendicular to the femoral neck. (**b**) Intraoperative photograph after trochanteric osteotomy and capsulotomy but before dislocation. Note the amount of the femoral head that sits permanently outside the acetabulum. This lateral cartilage appears pristine. (**c**) Intraoperative photograph showing the indentation of the femoral head created by the labrum and acetabular rim. This produces the saddle shape. (**d**) Intraoperative photograph at time of dislocation showing that despite being only 11 years old, degenerative changes already exist in the region corresponding to the concavity of the femoral head. (**e**) Intraoperative photograph of the dislocated femoral head from a superior view. The enlarged lateral portion is too big to enter the acetabulum. (**f**) Intraoperative photograph showing anterior profile view. The femoral head has a saddle shape. The medial part is spherical. The lateral part is also round. The concave central part is misshapen and had degenerative changes. Performing a "cheilectomy" would remove the lateral part of the femoral head and leave the depressed, degenerative central part in a weight bearing location. (**g**) Intraoperative photograph showing the Ganz-type lateral osteotomy parallel to the femoral neck but at an oblique angle to the femoral head. The entire piriformis fossa is contained with the lateral segment. The femoral head template shows where the medial part leaves the round and defines where to make the medial osteotomy. (**h**) Intraoperative photograph showing two reference wires inserted to guide the medial cut (left superior wire) and the resection of the intercalary segment (right lateral wire). (**i**) Intraoperative photograph showing the resection of the intercalary segment after two parallel osteotomies (A type) also parallel to the femoral neck. Notice the narrowing of the femoral neck over a long length. (**j**) Intraoperative photograph showing advancement of the lateral mobile femoral head to the medial stable femoral head. The prominent anterolateral incongruity of the femoral head neck junction will be resected with an osteochondroplasty. (**k**) Intraoperative photograph after the reduction from a superior end on view. (**l**) Intraoperative photograph showing anterior profile view of the femoral head after the reduction and osteochondroplasty. The femoral head sphericity has been restored. (**m**) Postoperative AP pelvis radiographs after FHRO. The femoral head is well contained. The lateral portion of the femoral head sits under the dome of the acetabulum. The femoral head appears round on both AP and lateral views and would be classified as a Stulberg Class 1 femoral head. (**n**) Clinical photographs 1 year after surgery showing restoration of full and symmetric range of motion of both hips. (**o**) AP and frog lateral radiographs of the pelvis 10 years after FHRO. The joint space is well maintained. The femoral head is perfectly round. The remnant of stable trochanter was never resected and remains like an exostosis on the femoral neck. She underwent a derotation osteotomy by another surgeon 1 year prior. It is notable that the acetabulum is dysplastic although she remains completely asymptomatic. ((C) Dror Paley. Used with permission)

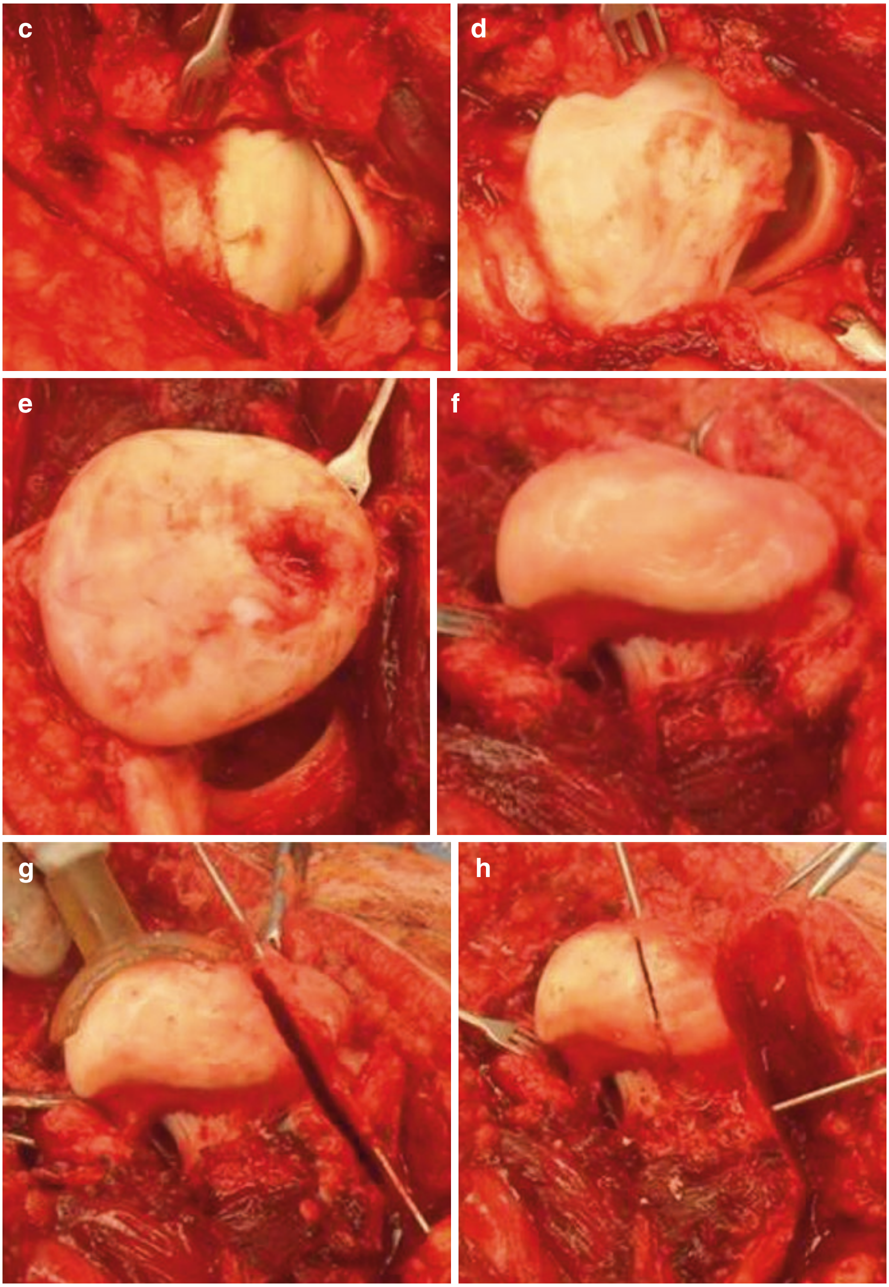


Fig. 9.18 (continued)

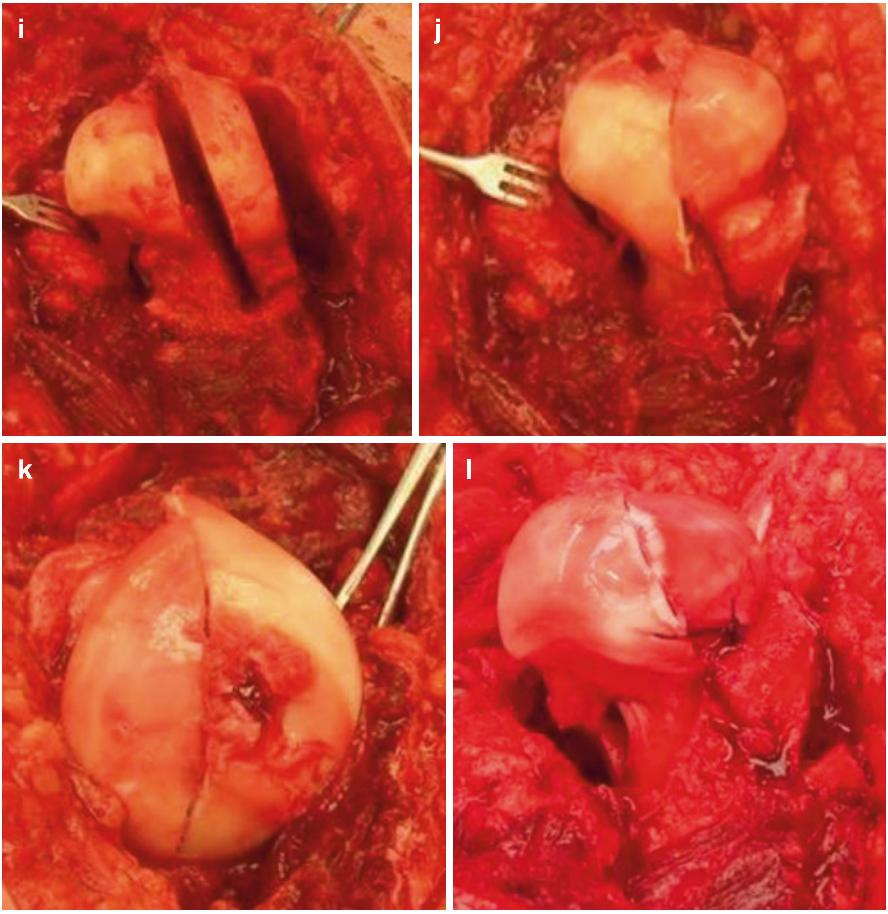


Fig. 9.18 (continued)

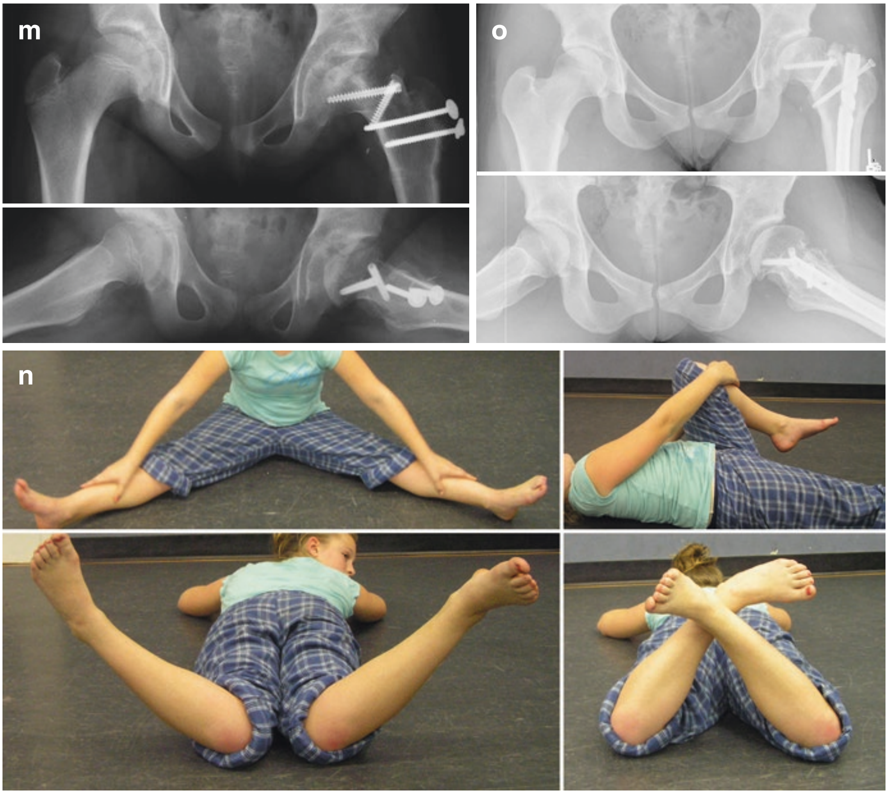


Fig. 9.18 (continued)

of FHRO would be performed based on the pathoanatomy, the result from the Ganz type is excellent. She did undergo a derotation osteotomy of the femur for external femoral torsion. She has some lack of coverage that may require a PAO in the future. Arguably she should already have had one except that she is completely asymptomatic.

Patient 2 (Fig. 9.19): A 15-year-old girl with history of left-sided Perthes disease since age 8, treated nonoperatively with rest, casting, and physical therapy. She developed left hip pain a year prior to presentation. The pain was brought on by activity and prolonged sitting. She had a mild lurch to the left, which got worse as

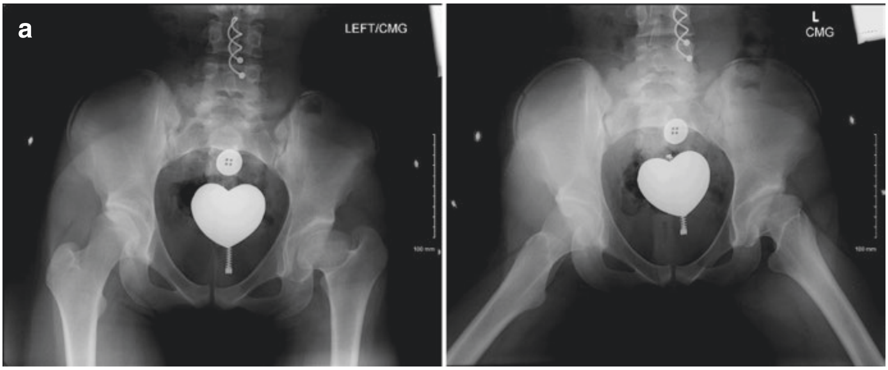


Fig. 9.19 (**a**) Preoperative AP and frog lateral radiographs of 15 year old girl with left coxa magna, coxa breva, and overgrown trochanter sequelae from previous Perthes. (**b**) 3D CT AP and lat views showing left Coxa magna and uncovered, elongated femoral head. The elongation is not perpendicular to the femoral neck. (**c**) Postoperative AP radiograph showing healed Paley-type A femoral head reduction osteotomy with level of greater trochanter restored and relative neck lengthening. The femoral head coverage is inadequate and a PAO is required. (**d**) Final AP radiograph after FHRO and Ganz-type PAO. There is excellent coverage of the femoral head. (**e**) AP femur radiograph after implantable limb lengthening to equalize limb lengths. The femoral head remains round, located, well covered, and the hip shows excellent joint space with no degeneration. (**f**) Intraoperative photograph showing superior view of femoral head after dislocation. The diameter of the femoral head is measured with a caliper. This is a Type A femoral head with minimal to no cartilage degeneration. (**g**) Intraoperative photograph showing acetabular sizer in place (blue). The profile view of the femoral head shows the saddle shape. (**h**) Intraoperative photograph showing femoral spherical template corresponding to acetabular size measured. The lateral edge of the template is where the femoral head leaves the round. A mark should be made in line with this edge. (**i**) Intraoperative photograph showing the line and a caliper measuring the width of the medial femoral head at the point where the femoral head leaves the round based on a laterally placed femoral head template. (**j**) Intraoperative photograph showing lateral part of femoral head measured for the remainder of the diameter of the femoral head after the planned reduction. This width is marked. (**k**) Intraoperative photograph showing the femoral head marked with two parallel lines for osteotomy resection of the central portion of the femoral head (A type). (**l**) Intraoperative photograph from anterior view of the femoral head showing the baseline osteotomy perpendicular to the two parallel femoral head osteotomy lines. These femoral head lines are perpendicular to the femoral head joint surface and not parallel to the femoral neck. The baseline is extends laterally to exit into the trochanteric osteotomy. (**m**) Intraoperative photograph showing the femoral head osteotomy is being made with a thin sagittal saw which is being cooled with water. (**n**) Intraoperative photograph showing the lateral fragment reduced to the medial fragment and being fixed with a headless variable pitch screw. Note that the posterior reduction is prioritized. The guidewires for the headless screws are perpendicular to the intra-articular osteotomy in order to offer maximum interfragmentary compression. (**o**) Intraoperative photograph showing the reduced femoral head. The two headless screws are buried under the cartilage. The posterior femoral head is very congruous and spherical. There is mild anterior incongruity. The lateral femoral protrudes anteriorly beyond the medial femoral head. (**p**) Intraoperative photograph showing the anterior femoral head incongruity. The spherical template rests on the posterior femoral head and demonstrates the step off relative to the anterior femoral head. (**q**) Intraoperative photograph of spherical templating of posterior femoral head at the osteotomy. (**r**) Intraoperative photographs after osteochondroplasty resection of the incongruous anterolateral part of the femoral head. ((C) Dror Paley. Used with permission)

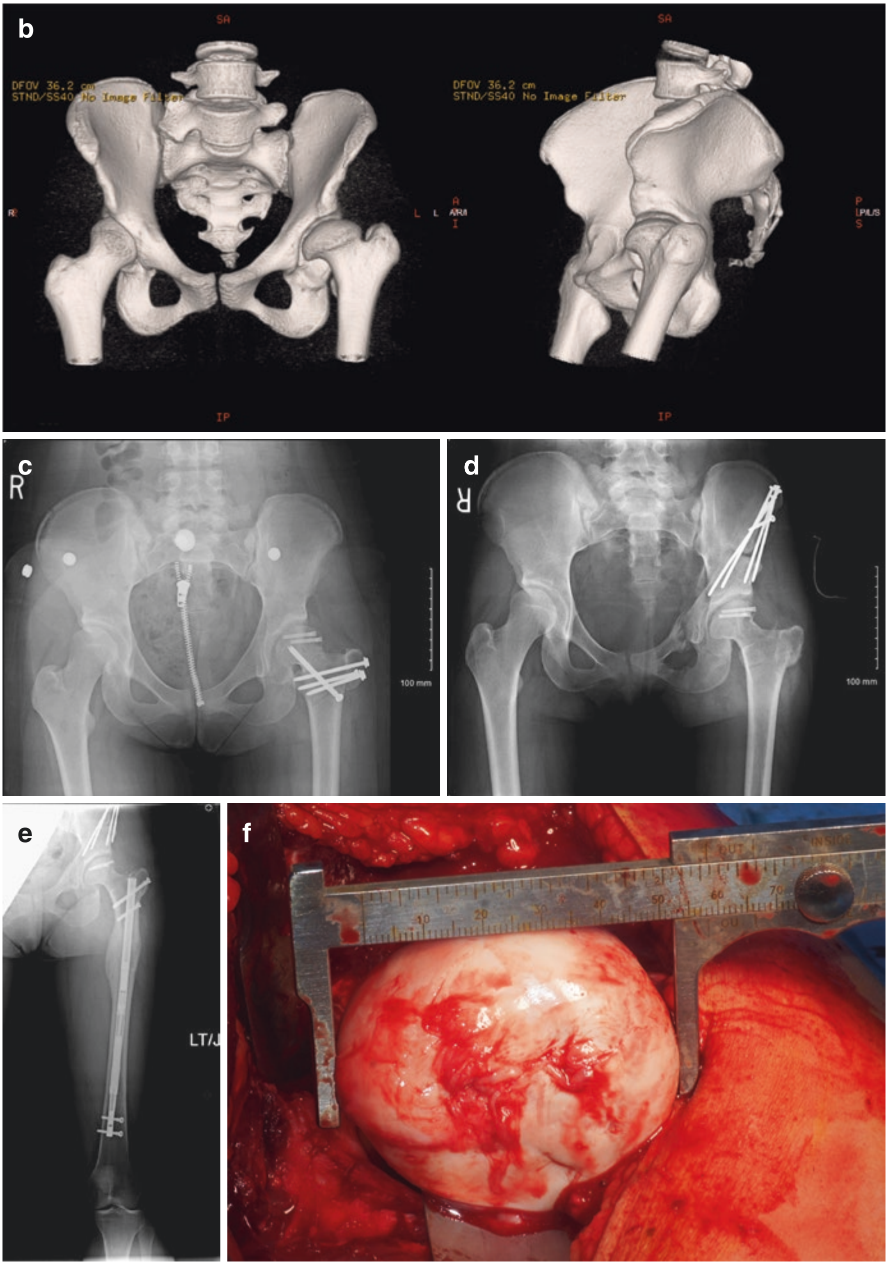


Fig. 9.19 (continued)

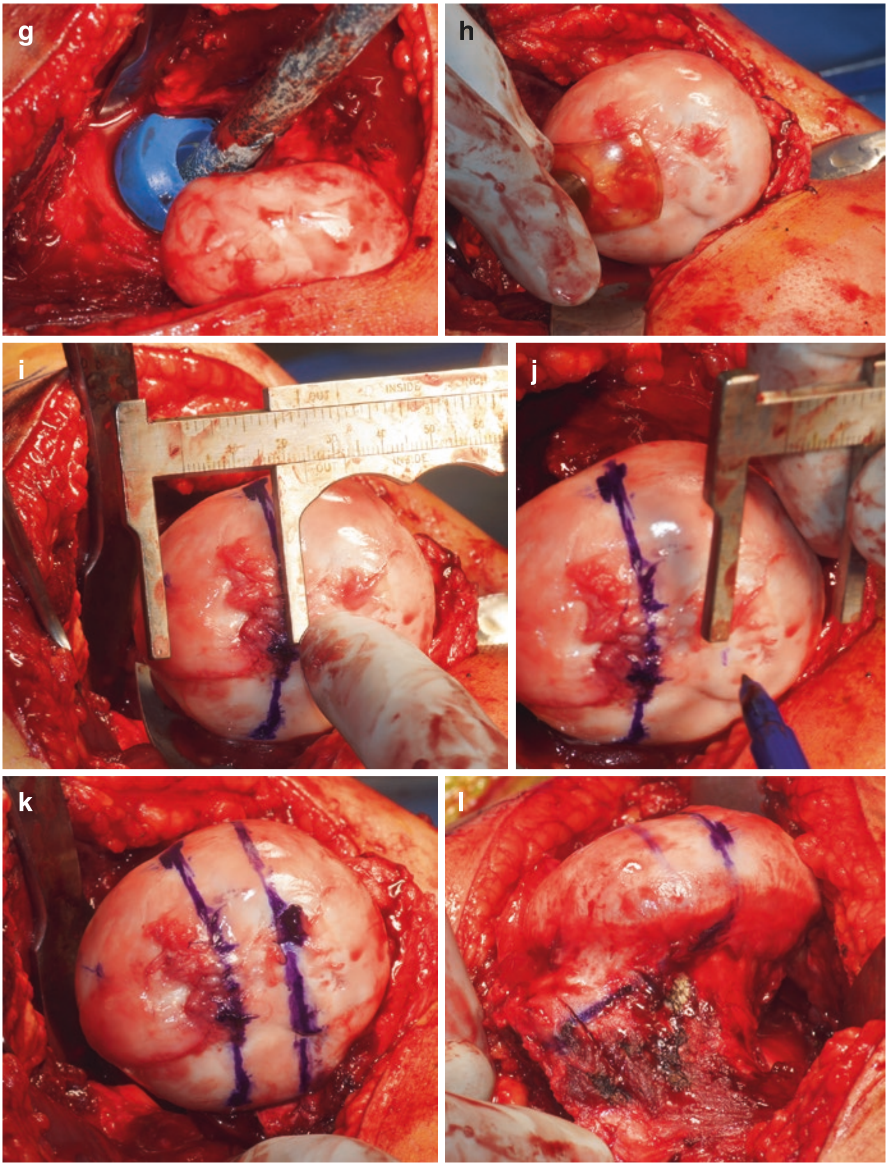


Fig. 9.19 (continued)

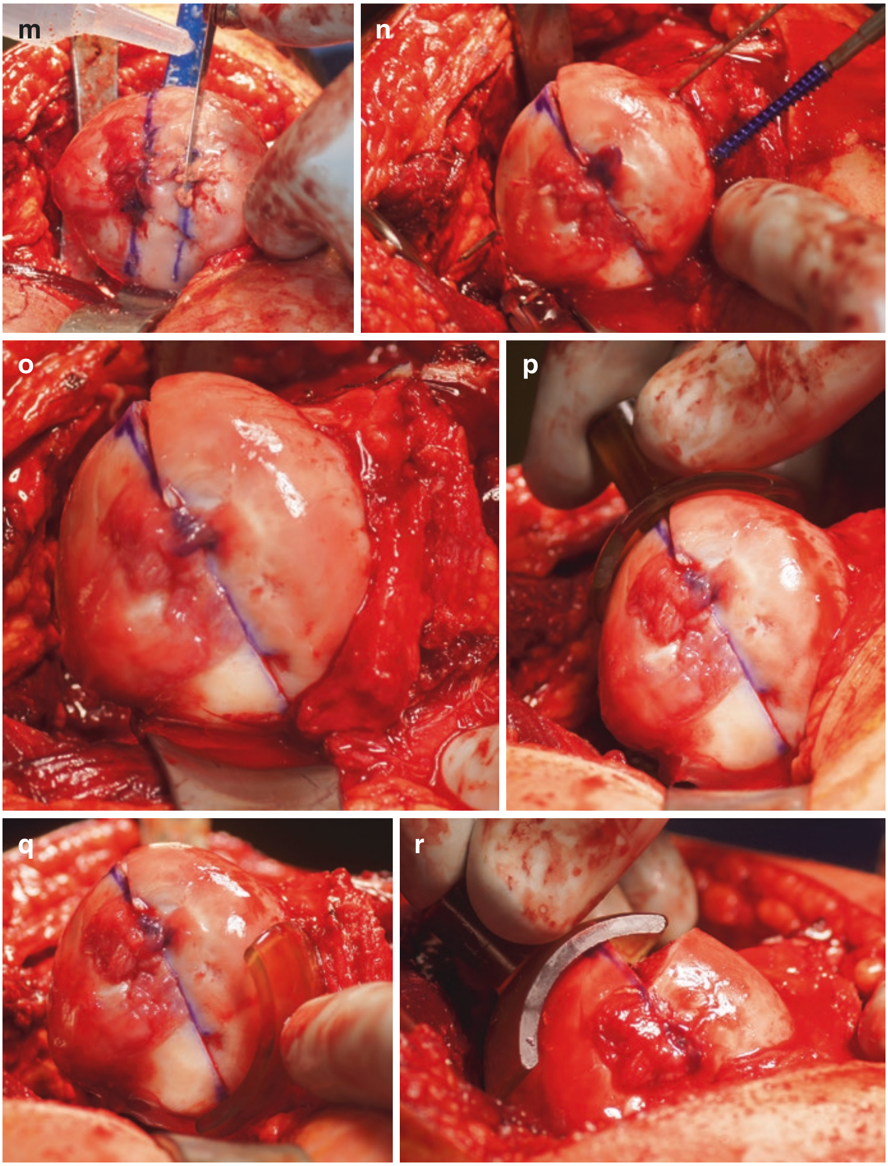


Fig. 9.19 (continued)

the day progressed. On examination she had pain on flexion with internal rotation of the hip. The hip range of motion was flexion 0–90 °, internal rotation 5 °, external rotation 45 °, adduction 20 °, and abduction 15 °.

The femoral head was elongated horizontally such that the perpendicular to this axis is not in line with the femoral neck. A Paley-type FHRO was performed. Intraoperative photographs show the step-by-step measurements confirming this was a type A. Two parallel cuts were made. An anterolateral osteochondroplasty eliminated any incongruity from the reduction. Her femoral head would be classified as a Stulberg Class III before surgery and Stulberg Class I after the FHRO. The femoral head was insufficiently covered due to previous acetabular remodeling. A PAO was performed 6 months after the FHRO. The patient currently has no limp or pin in the hip and has equal and symmetric full hip range of motion to the opposite side. Final radiographs 2 years after FHRO confirm good reduction, coverage, and joint space maintenance of the hip joint.

Patient 3 (Fig. 9.20): A 14-year-old boy with history of right-sided Perthes since age 11, treated nonoperatively by range of motion exercises. He became significantly disabled by the progressive collapse and misshapen femoral head. He had a significant antalgic lurch to the right. On examination his hip flexion was 0–90 ° with obligatory external rotation. He had 45 ° prone hip internal rotation and 10 ° prone hip external rotation. His hip abduction was 15 °. He had a leg length discrepancy.

Intraoperative photographs confirm that there is ample degeneration in the central portion but that the cartilage on the lateral rim is well preserved. The elongation of the femoral head is not perpendicular to the neck, and there is a large anterolateral bump which makes this a type B. This lends itself to a wedge resection of the anterolateral bump with nonparallel cuts restoring the femoral head sphericity. Postoperative radiographs demonstrate the transformation from a Stulberg Class V to a Stulberg Class I femoral head.

Four years after his Paley-type FHRO, he underwent lengthening with an implantable limb lengthening device to equalize his limb lengths. Ten years after the FHRO, he has no pain in his right hip and walks and runs with no limp. He has equal and full range of motion to his opposite hip. His hip abduction and external rotation movements increased significantly after surgery. His hip no longer has obligatory external rotation with flexion. He is showing no signs of any deterioration in the joint clinically or radiographically.

Pearls and Pitfalls

1. Prognosis improves with better preserved femoral and acetabular cartilage. Degenerative changes in the non-resected areas theoretically worsen the prognosis.
2. The younger the patient (age 9–16 years) often the better the outcome.
3. Preoperative planning of the size of the wedge or rectangle to be taken is essential in aiding operative execution.

4. The retinacular flap is a key component to the operation and should be performed in stages as described in the operative discussion.
5. The femoral neck screw protect against fracture of the femoral neck.
6. The capsule at the end of the reduction is capacious and should be advanced to prevent lateral subluxation.
7. Be certain the trochanter is well fixed. In older/larger patients, three screws are preferred.

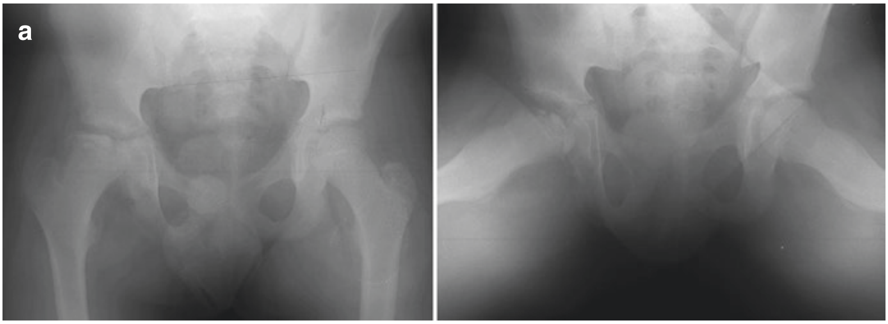


Fig. 9.20 (**a**) AP and frog lateral radiographs of the pelvis at age 11 years, showing Perthes disease affecting the right hip. There is whole head involvement and early collapse with a break in Shenton's line. There is already coxa magna with extrusion of the femoral head (proximal and lateral migration) and the greater trochanter already appears to be overgrowing and the femoral neck is already shorter than the opposite side. (**b**) Preoperative photographs showing hip range of motion. There is excellent flexion and extension but limited external rotation. There is no photograph of his hip abduction but it is limited. (**c**) Preoperative AP radiograph (left) compared to postoperative AP radiograph (right) after Paley-type FHRO. The femoral head went from a Stulberg Class 5 to a Stulberg Class 1. (**d**) Preoperative frog lateral radiograph (left) compared to postoperative frog lateral radiograph (right) after Paley-type FHRO. The femoral head went from a Stulberg Class 5 to a Stulberg Class 1. (**e**) Intraoperative photograph showing superior view of Type B femoral head. Note the large anterolateral bump. There is already significant damage to the central femoral head cartilage. Note that the lateral femoral head has excellent cartilage. (**f**) Intraoperative photograph showing superior view of the wedge shaped osteotomy resecting the anterior bump. (**g**) Intraoperative osteotomy showing superior view with the wedge removed. The wedge contains the most damaged cartilage. (**h**) Intraoperative photograph showing superior view of the osteotomy wedge closed, creating a spherical femoral head. (**i**) Intraoperative photograph from anterior view. The spherical template fits perfectly on the femoral head. Notice that the osteotomy line is perpendicular to the femoral head and not parallel to the femoral neck (Paley type). Correspondingly, the baseline osteotomy is perpendicular to the femoral head osteotomy and in line with the femoral head reduction. (**j**) Postoperative hip range of motion 10 years after FHRO surgery and 6 years after leg lengthening surgery showing excellent range of motion with improved hip abduction and external rotation. The range of motion is now symmetric to the opposite side. Note there is no lurch or Trendelenburg when in single leg stance (left). (**k**, **l**) Ten-year follow-up AP (**k**) and frog lateral (**l**) pelvis radiographs show excellent preservation of the femoral head shape and location with no evidence of any degenerative changes in the joint. Arguably, there is some relative dysplasia of the acetabulum and it would have been reasonable to do a PAO. The patient remains asymptomatic which is why he has not undergone further surgery. (**m**) Ten-year follow-up standing orthoroentgenogram demonstrating equal leg lengths and normal alignment. ((C) Dror Paley. Used with permission)

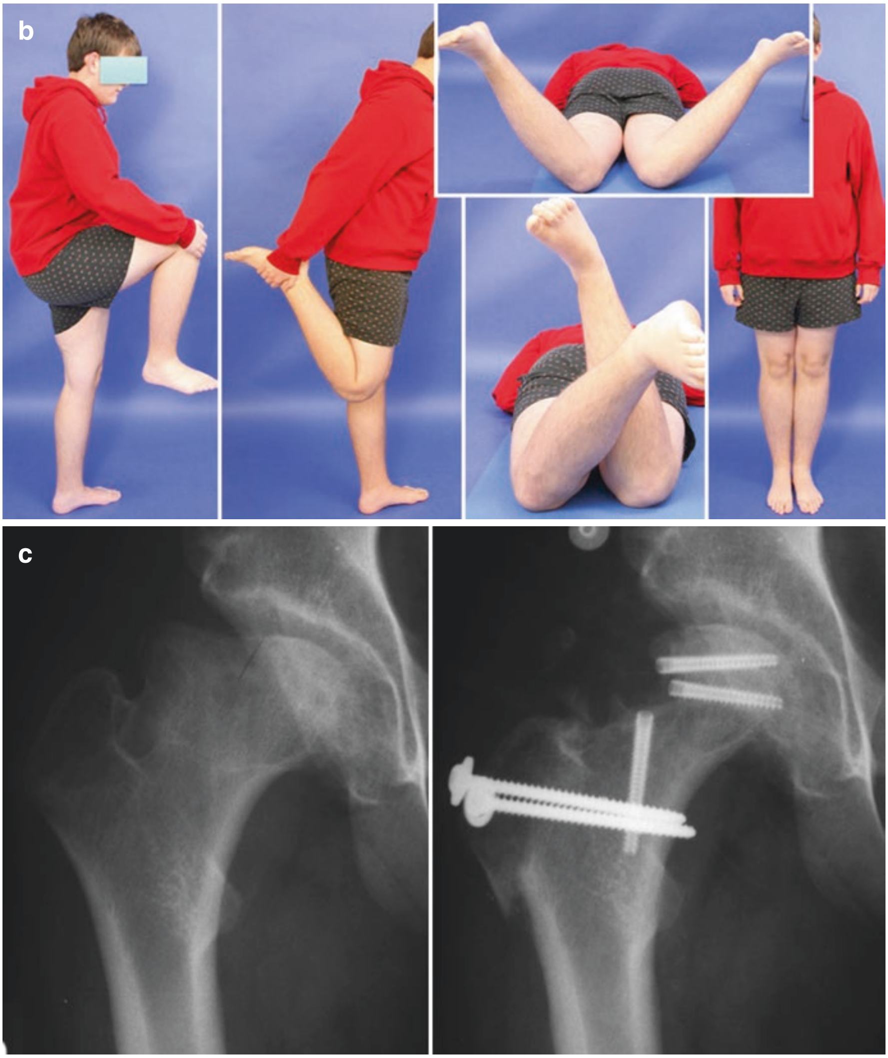


Fig. 9.20 (continued)

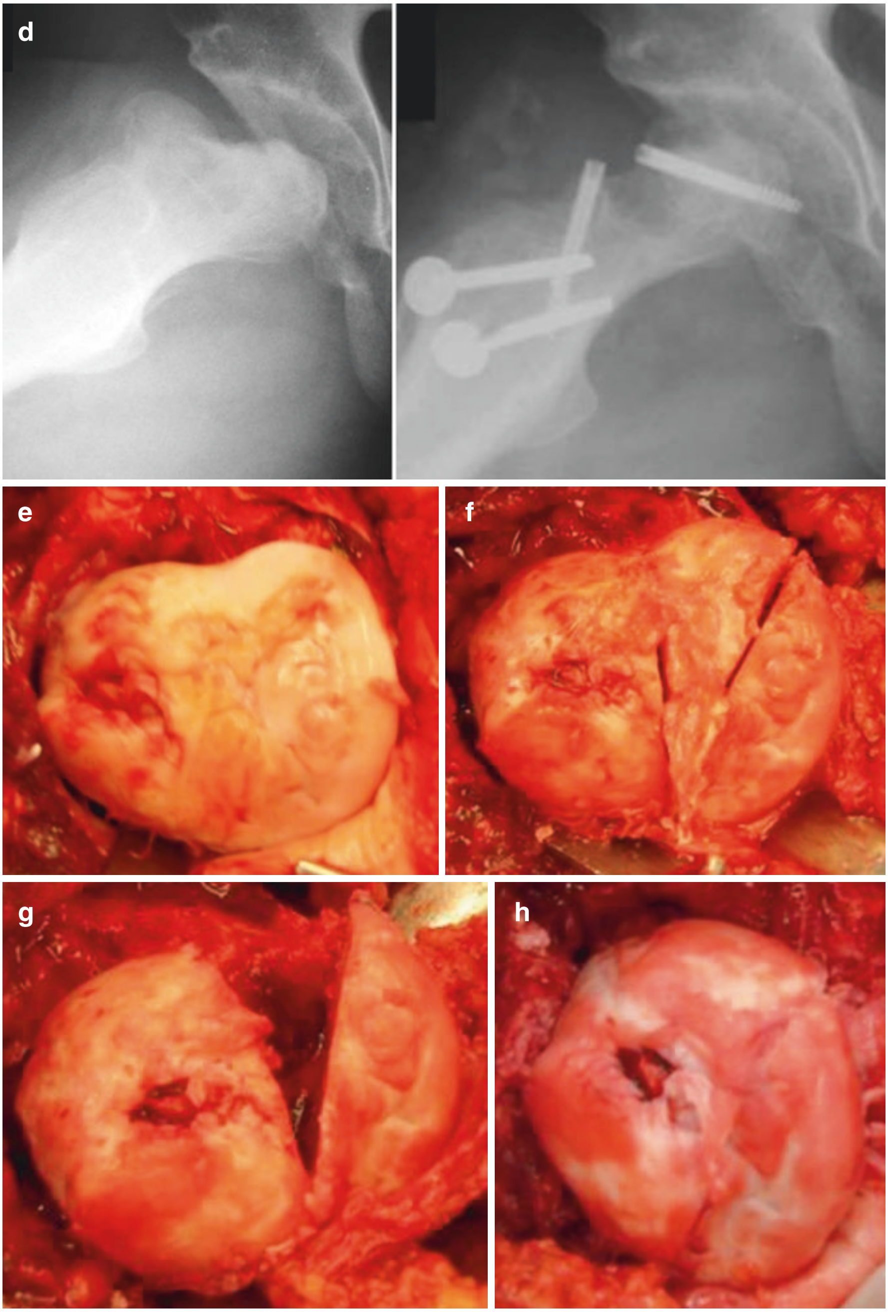


Fig. 9.20 (continued)

Fig. 9.20 (continued)

8. The presence of an open physis is not a contraindication due to risk of avascular necrosis as once thought. It is important to close the physis to avoid a bifid femoral head from developing.
9. Both the Ganz and Paley types of FHRO narrow the femoral neck and run the risk of fracture. It is therefore important to use a prophylactic screw to protect the femoral neck. The Ganz type leads to a long segment of narrowed femoral neck thus increasing the lever arm on the narrowed region compared to the Paley type.

Outcome of Ganz- and Paley-Type FHRO

Ganz (2009) [5] reported the results of the first 11 patients treated with the Ganz FHRO: The patients ranged in age from 9 to 15 years. In three hips, postoperative instability was treated by a femoral varus osteotomy in two and a periacetabular osteotomy (PAO) in one to stabilize the hip. Subsequently the next three cases were treated with PAO at the time of the FHRO. Ganz et al. (2010) [11] reported on their first 14 FHROs since 2001 (presumably including the 11 patients previously mentioned). Eight hips also had a PAO at the same time as the index procedure, while three had a PAO performed at a later date. In one case a varus intertrochanteric osteotomy was performed to treat subluxation, and in another a Colonna was performed at the same time. Therefore, only 1 of their 14 cases did not have an additional procedure. None of the 14 cases developed avascular necrosis. No other details on this group were reported.

Seibenrock et al. (2014) [9] published a series of 11 hips treated by the Ganz FHRO. The Ganz osteotomy was used both in cases with elongation of the femoral head perpendicular to the femoral neck and not perpendicular to the femoral neck. The mean age was 13 years 6 months (range 7–23 years). Follow-up was a mean of 5 years (range 1–10). The mean extrusion index decreased from an average of 47–21% after FHRO. Ten of the hips were rated as Stulberg Class II in 5, Class III in 4, and Class IV in 1. The Merle d'Aubigne score did not improve from prep to postop. No hip developed AVN. Five of 11 hips had concomitant pelvic osteotomy surgery for coverage. Another five hips had a triple pelvic osteotomy an average of 2.3 years after the FHRO. There was an improvement in pain score after surgery.

Paley (2011) [8] reported on 20 cases of Ganz FHRO performed for cases with femoral head elongation perpendicular and not perpendicular to the femoral neck. The mean age was 14 years (range 10–23). Fourteen (70%) of the 20 had improved range of motion, no pain or minimal to no limp and were considered good or excellent (satisfactory) results. Six (30%) of the 20 had significant pain, limp and or stiffness and were considered fair or poor (unsatisfactory) results. The follow-up at the time of the report was 2.7 years (range 1–5). Unpublished updated follow-up on the satisfactory group has shown no deterioration with a mean follow-up of 9 years with range from 7 to 11 years. Only five cases in this study had a pelvic osteotomy, all of which were in the satisfactory group. Several appear to be uncovered and would benefit from a coverage procedure (Fig. 9.18).

Paley et al. reviewed a second series of FHRO performed between 2010 and 2016 that were no included in the first series (unpublished). There were 25 hips in 24 patients in this second series. During the time of the first series, the author did not recognize the difference between the Ganz and the Paley type of osteotomy. The Paley type of osteotomy evolved out of the Ganz type until it became evident that the two osteotomies were quite distinct and that they each had different indications. In the second series, the author recognized this distinction and applied the Ganz osteotomy selectively when the femoral head elongation was perpendicular to the femoral neck. Similarly, the Paley type of osteotomy was applied when the femoral head elongation was not perpendicular to the femoral neck. There were 19 Paley and 6 Ganz FHRO performed. The mean follow-up was 3 years (range 1–6 years). The Harris hip score improved from 57 to 84 in the Ganz group and 65 to 87 in the Paley group. Sphericity as defined as mm away from a perfect Mose circle diameter on AP/LAT view went from 12/9 mm pre-op on Ganz to 8/7 and 12/11 on Paley to 6/6. The extrusion index went from 31% pre-op in Ganz to 13.3% postop vs 34.3% pre-op in Paley to 18.7% post-op. Using the previous satisfactory and unsatisfactory result score from Paley 2011, the current results showed 4 satisfactory and 2 unsatisfactory for Ganz vs 16 satisfactory vs 3 unsatisfactory for Paley. There were two cases of AVN both in Ganz osteotomies.

These results suggest that tailoring the FHRO to always be perpendicular to the femoral head elongation axis may be preferable to always making the osteotomy parallel to the femoral neck. Critical examination of some of the author's as well as Siebenrock's [9] Ganz-type FHRO when used in cases with elongation not perpendicular to the femoral neck demonstrates less than ideal reshaping of the femoral head. Furthermore, making the osteotomy parallel to the femoral neck is more limiting in the amount that can be reduced. Osteotomy perpendicular to the femoral head elongation axis increases the amount that can be reduced safely.

A third technique of femoral head reduction osteotomy was published by Burian et al. (2013) [10]. They performed an anteromedial wedge resection of the femoral head. They dislocated the hip through an anterior approach without performing an osteotomy of the greater trochanter. They then created a wedge with its apex distal and medial in the calcar region and with its base superiorly in the femoral head. This allows them to advance the more mobile medial segment of the femoral head toward the lateral side. No relative neck lengthening or distalization of the overgrown greater trochanter is possible with this approach. This method does offer two significant advantages over the Ganz and Paley FHRO. First it avoids dissection in the region of the piriformis fossa, thus reducing the risk of avascular necrosis. Second, in cases with elongation of the femoral head in a more medial direction, which have a large overhang medially that is not well centered on the femoral neck, both the Ganz and the Paley FHRO are difficult to perform. This last situation may be the ideal indication for the Burian FHRO.

Summary

Surgical dislocation of the femoral head allows for intra-articular osteotomy of the femoral head and neck to reshape the femoral head and try and restore its sphericity. This can be accomplished by simple resection of parts of the periphery of the femoral head and neck (osteochondroplasty) or by more complex resection of intercalary parts of the femoral head and neck (femoral head reduction osteotomy). Due to the different patterns of deformity of the femoral head, the orientation of the osteotomy will change accordingly. Three types of FHRO have been developed by different surgeons, each of which has its own indications. An understanding of these indications as they relate to femoral head geometry allows the surgeon to base the choice of osteotomy on pathoanatomy.

Acknowledgments The author wishes to thank Pamela Boullier Ross who illustrated all of the illustrations used in this manuscript. The author would also like to thank the Paley Foundation for giving permission to Springer to reproduce these figures in this book chapter.

References

1. Stulberg SD, Cooperman DR, Wallensten R. The natural history of Legg-Calve-Perthes disease. J Bone Joint Surg Am. 1981;63:1095–108.
2. Rowe SM, Moon ES, Song EK, et al. The correlation between coxa magna and final outcome in Legg-Calve-Perthes disease. J Pediatr Orthop. 2005;25:22–7.
3. Rowe SM, Jung ST, Cheon SY, Choi J, Do Kang K, Kim KH. Outcome of cheilectomy in Legg-Calve-Perthes disease. Minimum 25-year follow-up of five patients. J Pediatr Orthop. 2006;26(2):204–10.
4. Paley D. Surgery for residual deformity in adolescents. Orthop Clin North Am. 2012;43(3):317–28.
5. Ganz R, Huff TW, Leunig M. Extended retinacular soft tissue flap for intraarticular hip surgery: surgical technique, indications, and results of its application. Instr Course Lect. 2009;58:241–55.
6. Leunig M, Ganz R. Relative neck lengthening and intracapital osteotomy for severe Perthes and Perthes-like deformities. Bull NYU Hosp Jt Dis. 2011;69(Suppl 1):S62–7.
7. Paley D. Intra-articular osteotomies of the hip, knee, and ankle. Oper Tech Orthop. 2011;21:184–96.
8. Paley D. The treatment of femoral head deformity and coxa magna by the ganz femoral head reduction osteotomy. Orthop Clin N Am. 2011;42(3):389–99.
9. Siebenrock KA, Anwander H, Zurmuhle CA, Tannast M, Slongo T, Steppacher SD. Head reduction osteotomy with additional containment surgery improves sphericity and containment and reduces pain in Legg-Calve-Perthes disease. Clin Orthop Relat Res. 2015;473(4):1274–83.
10. Burian M, Dungl P, Chomiak J, Ostadal M, Kubes R, Schwarz O. Operative treatment for saddle-shaped femoral heads [Czech]. Acta Chir Orthop Traumatol Cech. 2016;83(4):247–53.
11. Ganz R, Horowitz K, Leunig M. Algorithm for combined femoral and periacetabular osteotomies in complex hip deformities. Clin Orthop Relat Res. 2010;468(12):3168–80.

Biologic and Pharmacologic Treatment of Legg-Calvé-Perthes Disease

10

Neeraj M. Patel and David S. Feldman

Introduction

Legg-Calve-Perthes disease (LCPD) is typically considered an idiopathic avascular necrosis of the pediatric femoral head. There has been a considerable debate regarding its etiology. Hypotheses include vascular, thrombophilic, inflammatory, and environmental sources [1–7]. While the inciting insult remains unclear, previous authors have described the pathophysiologic stages of LCPD at both the microscopic and macroscopic levels.

As elucidated in the previous chapters, the treatment of LCPD traditionally involves either observation with activity modification or surgery, depending largely on the patient's age and stage of the disease [8]. The role of biologic or pharmaceutical treatment in LCPD has not been established. Such medical management could offer a new strategy or an adjuvant treatment by which to alter the course of the disorder, especially for those patients who have not yet developed advanced disease. While the etiology of LCPD has not yet been determined, the known aspects of its pathophysiology provide opportunities for biologic intervention. Specifically, modulation of bone resorption, bone formation, and inflammation carry the potential to alter disease progression.

N. M. Patel
Ann & Robert H. Lurie Children's Hospital of Chicago, Chicago, IL, USA

D. S. Feldman (✉)
Paley Orthopedic and Spine Institute, St. Mary's Medical Center, West Palm Beach, FL, USA
e-mail: dfeldman@paleyinstitute.org

D. S. Feldman, D. Paley (eds.), *Legg-Calvé-Perthes Disease*,
https://doi.org/10.1007/978-1-0716-0854-8_10

Bone Resorption

The fragmentation stage of LCPD is marked by a period of increased bone resorption. This manifests radiographically as lucencies within the femoral head and can last between 3 and 12 months before re-ossification begins. During this stage, an imbalance between bone resorption and formation results in a predominance of osteoclasts and replacement of necrotic bone with fibrovascular granulation tissue [9]. The femoral head is thus left vulnerable to further damage with repeated loading.

Investigators have sought to target bone resorption in order to alter the pathologic repair process and prevent future deformity. One method by which to accomplish this goal is modulation of the RANK (receptor activator of NF-κβ) and RANKL (RANK ligand) pathway. Interaction between RANK and RANKL plays a key role in osteoclast regulation. RANKL is secreted by osteoblasts and binds to the RANK receptor on osteoclast progenitors and mature osteoclasts to stimulate bone resorption. In regulation of the process, osteoprotegerin (OPG) binds to RANKL to prevent its interaction with RANK, thereby inhibiting osteoclast differentiation and activation (Fig. 10.1) [10–13]. The RANK/RANKL/OPG system therefore provides a potential therapeutic target against bone resorption in LCPD.

RANKL inhibitors such as denosumab have been studied for treatment of postmenopausal osteoporosis and may prevent resorption in LCPD [14]. Human studies

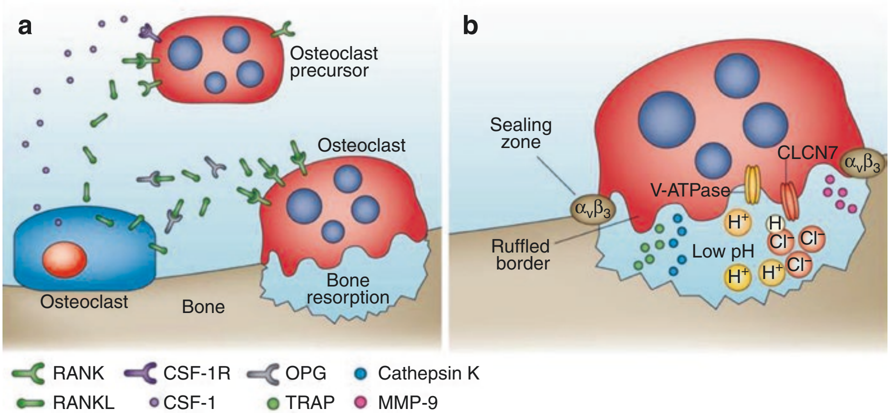


Fig. 10.1 Depiction of interactions between RANK, RANKL, and OPG. (**a**) Binding of RANK to RANKL stimulates osteoclast activation. OPG binds to RANKL to inhibit this pathway. (**b**) The process of bone resorption involves chloride channels and vacuolar adenosine triphosphatase, among other factors. These may serve as therapeutic targets for the treatment of LCPD in the future. Abbreviations: $av\beta_3$ $av\beta_3$ integrin, CLCN7 chloride channel 7 (H^+Cl^- exchange transporter 7), CSF-1 macrophage colony-stimulating factor 1, CSF-1R macrophage colony-stimulating factor 1 receptor, MMP-9 matrix metalloproteinase 9, OPG osteoprotegerin, RANK receptor activator of nuclear factor κB, RANKL receptor activator of nuclear factor κB ligand, TRAP tartrate-resistant acid phosphatase, V-ATPase vacuolar-type H^+-ATPase. (Adapted from Adamopoulos and Mellins [13])

are currently lacking, but Kim et al. evaluated the effect of exogenous OPG in 18 male piglets that underwent induced infarction of the femoral head. Radiographic analysis during the reparative phase of the disease showed improved femoral head morphology in animals treated with OPG compared to controls. The experimental group also had a reduction in the number of osteoclasts and better trabecular framework than controls. There was no effect on long-bone growth [15].

Bisphosphonates also modulate bone resorption and present another potential option in LCPD. These drugs are effective in several adult conditions and show early promise in post-traumatic avascular necrosis in children [16–19]. They are currently the most widely studied inhibitor of osteoclastic resorption in LCPD, though most research has been performed in animal models. Nitrogen-containing bisphosphonates, the most recent generation of these drugs, inhibit farnesyl pyrophosphatase, an enzyme in the HMG-CoA (3-hydroxy-3-methylglutaryl-coenzyme A) reductase pathway [20]. This inhibits microtubule formation in the ruffled borders of osteoclasts and induces apoptosis of these cells, thereby decreasing bone resorption [21].

In a systematic review of the literature, Young et al. found early promise in bisphosphonate use, though only a few articles met their inclusion criteria. Three Level IV clinical studies involved bisphosphonate administration in children. While none of these included patients with LCPD- the etiology of osteonecrosis was post-traumatic in one and malignancy-related in the others, subjective improvements were seen in pain and gait in 24 of 29 individuals. Of eight animal studies analyzed in their review, the authors found that seven found decreased femoral head deformity and six reported improved trabecular structure in subjects treated with bisphosphonates [22]. Another review of the literature specific to LCPD reports similar findings, primarily in animal studies [23]. Therefore, while there is a lack of human research in bisphosphonate use for LCPD, these drugs show early promise in experimental investigations.

Little et al. studied the effect of zoledronate on the spontaneously hypertensive rat, a previously established model of LCPD. One hundred twenty rats were divided into three groups: monthly saline administration, weekly zoledronate, or monthly zoledronate. Fifteen weeks later, bone mineral density was 18% higher with weekly zoledronate and 21% higher with monthly administration. “Flat” femoral heads, as calculated by epiphyseal quotient, were found in 32% of the controls but in only 12% of those rats given zoledronate weekly and 3% of those receiving the drug monthly. However, a 2.3–2.6% decrease in femoral length was also found in animals that received zoledronate [24], questioning the impact of bisphosphonates on growth.

Kim and colleagues stratified 24 piglets into three groups: a control group that received saline, another that received prophylactic ibandronate (a bisphosphonate) prior to induction of ischemic necrosis and a third that received ibandronate after ischemia. At 8 weeks, subjects receiving ibandronate pre- and post-ischemia had better-preserved femoral head morphology and trabecular framework (Fig. 10.2). Bisphosphonate use resulted in long-bone growth inhibition in this study as well, again raising the question of optimal dosage and delivery of the drug [25]. However,

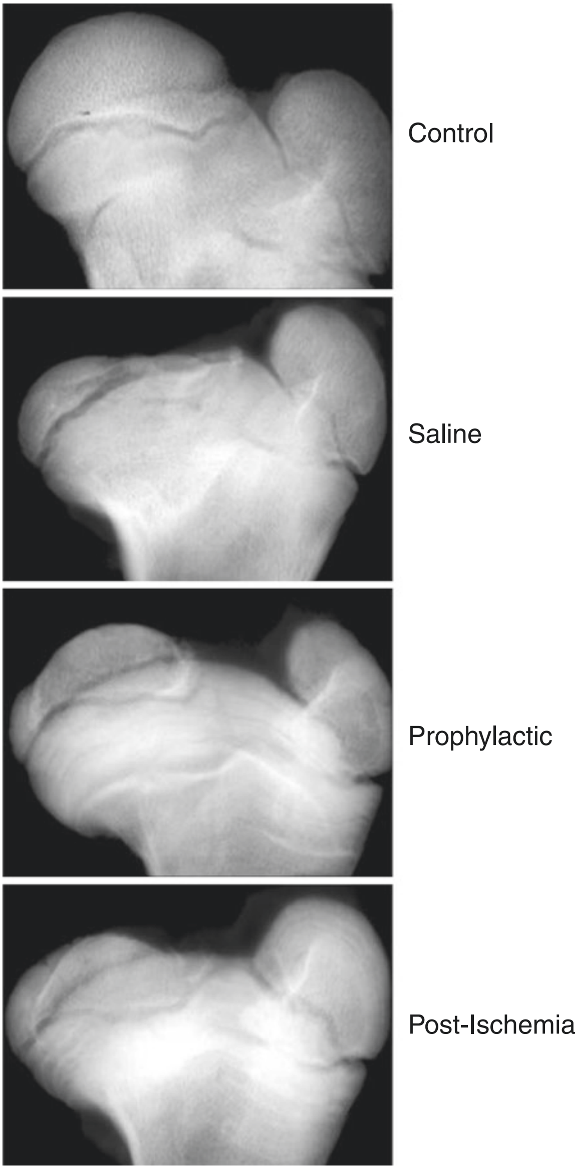


Fig. 10.2 Representative images from the work of Kim et al. demonstrating femoral head morphology in piglets that received saline, prophylactic ibandronate, or post-ischemic ibandronate. A normal, unaffected hip is shown as a control. (Adapted from Kim et al. [25])

the previously referenced systematic review reported that while decreased long bone growth was seen in six experimental studies with high-dose bisphosphonates in fast-growing animals, such growth inhibition has not been seen in human research [22].

The same authors then evaluated the local bioavailability of parenteral ibandronate in 15 piglets at various stages of femoral head revascularization and repair. Drug levels were similar to controls when there was no revascularization but increased significantly in subjects undergoing revascularization. Therefore, the optimal timing of bisphosphonate administration might be when revascularization has begun, as it may not be accessible to a completely infarcted head. Clinically, this could potentially be confirmed by bone scan or perfusion magnetic resonance imaging [26].

While the above research is promising, the adverse effects and optimal dosing of bisphosphonates in LCPD remain unclear. Högler et al. studied the short-term safety profile of parenteral zoledronate in 34 children with various musculoskeletal disorders. They found that the frequencies of post-infusion flu-like symptoms, hypocalcemia, and hypophosphatemia were all above 75%. There were no renal effects after three infusions. Dosing was between 0.02 and 0.025 mg/kg in 26 patients and 0.05 mg/kg in the other 8. The authors report that side effects were typically strongest after the first infusion compared to subsequent administrations. The electrolyte abnormalities did not cause symptoms in this cohort, and side effects were easily managed. The authors suggest that a first dose less than 0.02 mg/kg may be prudent and that premedication and calcium supplementation should be considered [27]. Along these lines, Munns and colleagues evaluated 63 children who received intravenous zoledronate for a variety of conditions. When compared to higher doses, an initial dose of 0.0125 mg/kg reduced the rate and magnitude of hypocalcemia but not the incidence of flu-like symptoms [28].

In one of the only studies that include children with LCPD treated with bisphosphonates, Johannesen et al. evaluated the systemic effects of zoledronate. Their cohort included 17 patients with LCPD and 20 with post-traumatic avascular necrosis, all of whom were treated for at least 12 months. Children received an initial intravenous dose of 0.0125 mg/kg, followed by subsequent infusions of 0.025 mg/kg. Markers of bone turnover were decreased and parathyroid hormone levels increased during the first 12 months, but subsequently stabilized. Bone modeling was reduced, but there were no fractures, spondylolisthesis, or osteonecrosis of the jaw. Importantly, age-adjusted bone mineral density in the lumbar spine and total body increased significantly, more so in the LCPD group than in post-traumatic patients. Given this significant increase with concomitant decrease in bone modeling, the authors were unable to provide a firm safety profile of zoledronate in children with LCPD [29].

Given the concerns about the safety profile of systemic bisphosphonates, Aya-ay et al. sought to evaluate the effect of ibandronate injected directly into the femoral heads of 27 piglets. The drug was almost completely localized to the femoral head 48 hours later. At 3 weeks, 50% of the original dose was retained in the femoral head with 30% remaining after 7 weeks. Radiographic analysis showed that subjects receiving intraosseous ibandronate showed better preservation of the femoral head than those injected with saline. The authors suggest that while it would be more invasive than systemic administration, intraosseous bisphosphonates could provide a safer, more localized treatment option. Furthermore, while the bioavailability of systemic bisphosphonates is dependent on the vascularity of the femoral head, vascular status does not impact local injection [30].

Although human studies of bisphosphonates in LCPD are scarce and the safety profile is just beginning to be defined, experimental studies show early promise. There is currently a randomized clinical trial in progress in Australia comparing parenteral zoledronate to standard LCPD treatments (Clinical Trial Registration ACTRN12610000407099). Such research will help elucidate the efficacy and safety of bisphosphonate use in LCPD.

Bone Formation

While many experimental models have modulated bone resorption in the biologic treatment of LCPD, upregulation of bone formation is another potential therapeutic target. This method could be especially important because while bisphosphonate treatment preserved the trabecular framework of the femoral head and prevented further deformity, it was also associated with a lack of bone turnover and remodeling [25]. Along these lines, authors have hypothesized that the addition of anabolic medications could achieve even better preservation of the femoral head with improved remodeling.

Several authors have evaluated bone morphogenic proteins (BMPs) in animal studies. BMPs are osteoinductive proteins produced by osteoprogenitor cells and osteoblasts to promote differentiation of mesenchymal cells into osteoblasts [31–34]. Specifically, BMPs activate a transmembrane serine/threonine kinase receptor, leading to activation of intracellular signaling molecules, Smads, that regulate the transcription of targeted genes [35]. This ultimately results in new bone formation. While BMPs have been studied in the context of spine fusion and fracture nonunion, their role in LCPD is unclear [31, 36–40].

In an experiment that was the first of its kind, Vandermeer et al. studied the impact of combined ibandronate and BMP-2 on the osteonecrotic femoral heads of immature pigs. These agents were injected directly into the femoral head. There were four experimental groups: normal hips, saline injection, ibandronate alone, and ibandronate with BMP-2. Subjects receiving ibandronate and BMP-2 had significantly better femoral head morphology and greater trabecular volume, thickness, and number. These hips also had decreased osteoclast number and larger osteoblast surface value than controls (Fig. 10.3). Of note, the subjects in this group were additionally noted to have heterotopic ossification within the joint capsule. The authors report that while this finding could be due to the dosage or injection technique, further investigation is required [41].

The same center subsequently administered BMP-2 and ibandronate during non-weight bearing (NWB) treatment of ischemic femoral head osteonecrosis in 18 piglets. The authors postulated that while NWB treatment is protective in LCPD, it promotes bone resorption without bone formation. They therefore hypothesized that administration of ibandronate and BMP-2 could modulate bone resorption and formation during NWB. Compared to the NWB-only and BMP-2 with NWB groups, subjects receiving both ibandronate and BMP-2 while remaining NWB had less

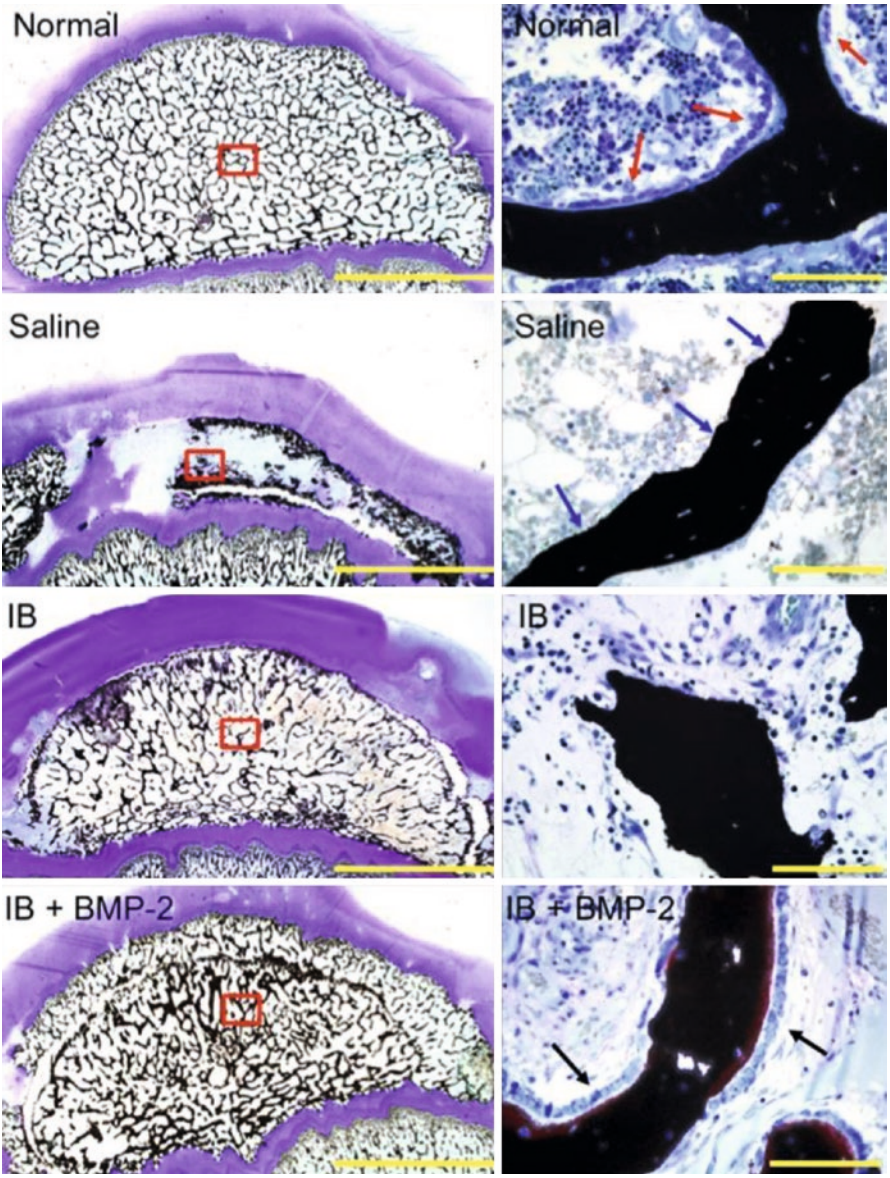


Fig. 10.3 Vandermeer et al. compared the histology of normal femoral heads with those receiving saline, ibandronate (IB), or IB with BMP-2. On the left column, low-magnification views demonstrate improved trabecular preservation and sphericity in the treatment groups. Of note, the IB + BMP-2 group has a "head within a head" appearance with thickened central trabeculae. The right column shows osteoblasts along the trabecular surface in the control group (red arrows) but absence of these cells in the saline group (blue arrows) and IB-only group. The IB + BMP-2 subjects show osteoblasts lining the trabeculae (black arrows), similar to controls. (Adapted from Vandermeer et al. [41])

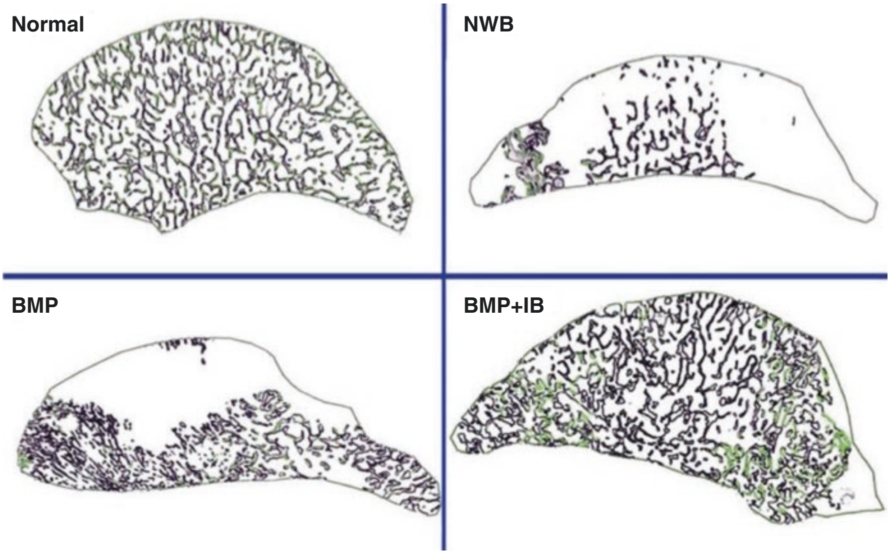


Fig. 10.4 Histomorphometric images from the work of Kim et al. show improved trabecular framework (black outline) with increased calcein-labeled surfaces (green outline, indicating new bone formation) in animals receiving both BMP-2 and ibandronate (IB) during non-weight bearing (NWB) compared to NWB alone and BMP-2 alone. (Adapted from Kim et al. [42])

femoral head deformation and higher bone volume. These animals also had lower osteoclast number and increased bone formation (Fig. 10.4). Four subjects that received both medications had capsular heterotopic ossification, while none of the BMP-only animals did [42]. Of note, the authors halved the dose of BMP-2 compared to their previous study and also changed the method and rate of infusion. However, the reason behind the presence of heterotopic ossification in the dual-medication group and its absence in the BMP-only group remains unclear. Another group of investigators used a sucrose acetate isobutyrate carrier (rather than saline) to deliver these agents and suggested that modification of this system could result in less heterotopic ossification in future studies [43]. Therefore, administration of BMP-2 and bisphosphonates appears to have some efficacy during NWB treatment of LCPD, but heterotopic ossification remains a concern.

Previous studies have described the mechanism by which statin medications increase BMP-2 expression [44–46]. Along these lines, Zou et al. evaluated the effect of a locally injected bisphosphonate (clodronate) with subsequent simvastatin injection in a porcine model. Compared to animals receiving only one of these medications or neither drug, those who received clodronate with simvastatin showed no gross radiographic collapse and superior trabecular framework. Heterotopic ossification was not seen in any subjects. The authors suggest that the synergy between an initial anti-resorptive burst followed by sustained osteogenic stimulation may provide a biological approach for the treatment of LCPD [47]. Specifically, sustained delivery of a statin might allow for better control of side effects than the use of a BMP.

Another drug that has been studied in rat model LCPD is strontium [48]. Strontium activates the calcium receptors on the osteoclasts and osteoblasts through the OPG/RANKL system. Shown to have a positive effect on postmenopausal osteoporosis [49], study is needed on whether direct injection into the avascular femoral head or given systemically in human subjects is safe and can help prevent collapse.

While research on the modulation of bone formation in LCPD is less robust than that of bisphosphonate use, early animal studies show promise for new techniques in the treatment of LCPD. Specifically, concurrent or staged administration of bisphosphonates with either a BMP or statin may provide improved bone formation and remodeling compared to bisphosphonates alone. However, heterotopic ossification remains a concern and further study is needed prior to human use.

Inflammation

The fragmentation phase of LCPD has been found to be a period of increased inflammation. Little and Kim initially postulated that inflammatory factors such as tumor necrosis factor-alpha (TNF-α) and interleukins (IL) may play a role in the inhibition of bone formation. Specifically, they cite the ability of these cytokines to block osteoblast differentiation in diseases like rheumatoid arthritis [50]. If LCPD involves a similar pathophysiology, these inflammatory mediators could serve as a therapeutic target.

Kim, Kamiya, and associates later found that LCPD results in chronic hip synovitis and increased intra-articular IL-6. Serial MRI was performed on 28 children and synovial fluid was taken from 13. Compared to the unaffected hip, MRI showed a fivefold increase in synovial fluid volume at the initial visit and threefold increase at latest follow-up. IL-6 levels were significantly increased in LCPD, but not IL-1 or TNF-α [49]. Of note, IL-6 has previously been shown to stimulate osteoclasts while inhibiting osteoblasts [51–54]. The same authors subsequently showed that IL-6 production depends on hypoxia-inducible factor-1 (HIF-1) and that tocilizumab – an IL-6 receptor blocker – decreased the presence of inflammatory cytokines in an animal model [55]. This links the hypoxia caused by ischemia with inflammatory changes and proposed another therapeutic avenue in LCPD (Fig. 10.5). Therefore, while inflammation has not been studied as extensively as bone resorption and formation in LCPD, it may have important pathophysiologic implications.

Future Directions

A great deal of research is still necessary prior to the use of biologic treatments in LCPD. Early studies are promising, but the overwhelming majority were performed utilizing animal models. Bisphosphonates are the most widely studied medication at present, and a randomized clinical trial is currently in progress in Australia. Further study of RANKL inhibitors as well as agents like cathepsin K inhibitors and

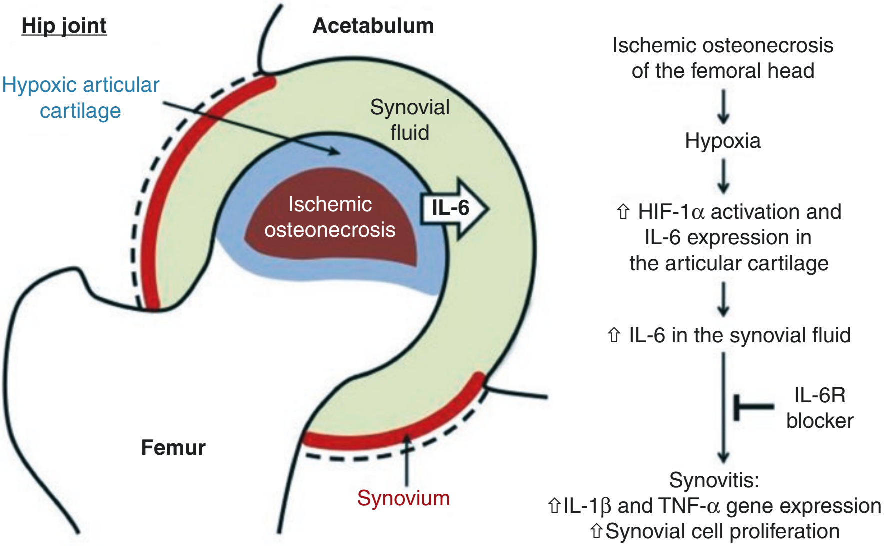


Fig. 10.5 Schematic overview of the link between hypoxia and inflammation, as proposed by Yamaguchi et al. Ischemia results in hypoxia, which stimulates HIF-1 production by chondrocytes. This leads to increased IL-6 production, which is released into the synovial fluid. The eventual result is proliferation of synovial cells, IL-1β, and TNF-α. Tocilizumab, an IL-6 receptor blocker, can dampen this response. (Adapted from Yamaguchi et al. [55])

inhibitors of osteoclastic chloride channels or vacuolar adenosine triphosphatase may provide additional options for modulation of bone resorption (Fig. 10.1) [13, 56–58]. Anabolic medications, such as BMPs and statins, also carry potential, but the optimal dosing, delivery systems, efficacy, and safety must be better defined.

Tocilizumab, an IL-6 inhibitor, was previously shown to be effective and safe in adult rheumatoid arthritis and idiopathic juvenile arthritis [59–63]. According to recent experimental research, it may eventually be an option for Perthes disease. Future work will likely evaluate the effect of IL-6 inhibition on bone remodeling, as this cytokine is known to stimulate osteoclasts and inhibit osteoblasts. As previously noted, Yamaguchi et al. elucidated the link between hypoxia (increased HIF-1) and inflammation (increased IL-6) [53]. Other studies have also correlated HIF-1 stimulation with increased production of vascular endothelial growth factor (VEGF) and expression of Sox9, a transcription factor that upregulates chondrocyte differentiation [64–66]. These all serve as possible lines of future study.

Other etiologies of Perthes disease – including vascular pathology, thrombophilia, and abnormal lipid metabolism – are less established [1–3, 5–7]. Further molecular-level study of these theories may lead to additional therapeutic targets in the future. In the meantime, evaluation of human efficacy and safety of agents that modify bone resorption, bone formation, or inflammation appears to be the next logical step in investigation.

Conclusion

While pharmaceuticals have not traditionally been a part of the therapeutic algorithm in Perthes disease, recent animal and laboratory studies show great promise for biological treatment of this disorder. Modulators of bone resorption, such as bisphosphonates and RANKL inhibitors, are the most studied agents and are starting to be evaluated in children. Anabolic agents like BMPs and statins may eventually provide a strong adjunct to bisphosphonates, but delivery techniques and dosage must be perfected prior to human application. Finally, the regulation of inflammatory cytokines has the potential to impact the pathophysiology and symptomatology of LCPD. Further research on these and other agents may ultimately provide biologic options for the treatment of Perthes disease.

References

1. Aksoy MC, Aksoy DY, Haznedaroglu IC, Sayinalp N, Kirazli S, Alpaslan M. Thrombomodulin and GFC levels in Legg-Calve-Perthes disease. Hematology (Amsterdam, Netherlands). 2008;13(6):324–8.
2. Hresko MT, McDougall PA, Gorlin JB, Vamvakas EC, Kasser JR, Neufeld EJ. Prospective reevaluation of the association between thrombotic diathesis and Legg-Perthes disease. J Bone Joint Surg Am. 2002;84-a(9):1613–8.
3. Ismayilov V, Aksoy DY, Sayinalp N, Haznedaroglu IC, Aksoy MC. Increased soluble selectins as a reflection of activated platelets and endothelium in Legg-Calve-Perthes disease. J Pediatr Hematol Oncol. 2014;36(7):e410–1.
4. Kim HK. Pathophysiology and new strategies for the treatment of Legg-Calve-Perthes disease. J Bone Joint Surg Am. 2012;94(7):659–69.
5. Liu R, Fan L, Yin L, et al. Comparative study of serum proteomes in Legg-Calve-Perthes disease. BMC Musculoskelet Disord. 2015;16:281.
6. Vosmaer A, Pereira RR, Koenderman JS, Rosendaal FR, Cannegieter SC. Coagulation abnormalities in Legg-Calve-Perthes disease. J Bone Joint Surg Am. 2010;92(1):121–8.
7. Woratanarat P, Thaveeratitharm C, Woratanarat T, Angsanuntsukh C, Attia J, Thakkinstian A. Meta-analysis of hypercoagulability genetic polymorphisms in Perthes disease. J Orthop Res. 2014;32(1):1–7.
8. Chaudhry S, Phillips D, Feldman D. Legg-Calve-Perthes disease: an overview with recent literature. Bull Hosp Joint Dis (2013). 2014;72(1):18–27.
9. Kim HK, Su PH. Development of flattening and apparent fragmentation following ischemic necrosis of the capital femoral epiphysis in a piglet model. J Bone Joint Surg Am. 2002;84-a(8):1329–34.
10. Boyce BF, Xing L. Biology of RANK, RANKL, and osteoprotegerin. Arthritis Res Ther. 2007;9(Suppl 1):S1.
11. Boyle WJ, Simonet WS, Lacey DL. Osteoclast differentiation and activation. Nature. 2003;423(6937):337–42.
12. Lacey DL, Timms E, Tan HL, et al. Osteoprotegerin ligand is a cytokine that regulates osteoclast differentiation and activation. Cell. 1998;93(2):165–76.
13. Adamopoulos IE, Mellins ED. Alternative pathways of osteoclastogenesis in inflammatory arthritis. Nat Rev Rheumatol. 2015;11(3):189–94.
14. Cummings SR, San Martin J, McClung MR, et al. Denosumab for prevention of fractures in postmenopausal women with osteoporosis. N Engl J Med. 2009;361(8):756–65.

15. Kim HK, Morgan-Bagley S, Kostenuik P. RANKL inhibition: a novel strategy to decrease femoral head deformity after ischemic osteonecrosis. J Bone Miner Res Off J Am Soc Bone Miner Res. 2006;21(12):1946–54.
16. Agarwala S, Jain D, Joshi VR, Sule A. Efficacy of alendronate, a bisphosphonate, in the treatment of AVN of the hip. A prospective open-label study. Rheumatology (Oxford, England). 2005;44(3):352–9.
17. Ramachandran M, Ward K, Brown RR, Munns CF, Cowell CT, Little DG. Intravenous bisphosphonate therapy for traumatic osteonecrosis of the femoral head in adolescents. J Bone Joint Surg Am. 2007;89(8):1727–34.
18. Lai KA, Shen WJ, Yang CY, Shao CJ, Hsu JT, Lin RM. The use of alendronate to prevent early collapse of the femoral head in patients with nontraumatic osteonecrosis. A randomized clinical study. J Bone Joint Surg Am. 2005;87(10):2155–9.
19. Nishii T, Sugano N, Miki H, Hashimoto J, Yoshikawa H. Does alendronate prevent collapse in osteonecrosis of the femoral head? Clin Orthop Relat Res. 2006;443:273–9.
20. Russell RG, Xia Z, Dunford JE, et al. Bisphosphonates: an update on mechanisms of action and how these relate to clinical efficacy. Ann N Y Acad Sci. 2007;1117:209–57.
21. Fleisch H. Bisphosphonates: mechanisms of action. Endocr Rev. 1998;19(1):80–100.
22. Young ML, Little DG, Kim HK. Evidence for using bisphosphonate to treat Legg-Calve-Perthes disease. Clin Orthop Relat Res. 2012;470(9):2462–75.
23. Little DG, Kim HK. Potential for bisphosphonate treatment in Legg-Calve-Perthes disease. J Pediatr Orthop. 2011;31(2 Suppl):S182–8.
24. Little DG, McDonald M, Sharpe IT, Peat R, Williams P, McEvoy T. Zoledronic acid improves femoral head sphericity in a rat model of Perthes disease. J Orthop Res. 2005;23(4):862–8.
25. Kim HK, Randall TS, Bian H, Jenkins J, Garces A, Bauss F. Ibandronate for prevention of femoral head deformity after ischemic necrosis of the capital femoral epiphysis in immature pigs. J Bone Joint Surg Am. 2005;87(3):550–7.
26. Kim HK, Sanders M, Athavale S, Bian H, Bauss F. Local bioavailability and distribution of systemically (parenterally) administered ibandronate in the infarcted femoral head. Bone. 2006;39(1):205–12.
27. Hogler W, Yap F, Little D, Ambler G, McQuade M, Cowell CT. Short-term safety assessment in the use of intravenous zoledronic acid in children. J Pediatr. 2004;145(5):701–4.
28. Munns CF, Rajab MH, Hong J, et al. Acute phase response and mineral status following low dose intravenous zoledronic acid in children. Bone. 2007;41(3):366–70.
29. Johannesen J, Briody J, McQuade M, Little DG, Cowell CT, Munns CF. Systemic effects of zoledronic acid in children with traumatic femoral head avascular necrosis and Legg-Calve-Perthes disease. Bone. 2009;45(5):898–902.
30. Aya-ay J, Athavale S, Morgan-Bagley S, Bian H, Bauss F, Kim HK. Retention, distribution, and effects of intraosseously administered ibandronate in the infarcted femoral head. J Bone Miner Res Off J Am Soc Bone Miner Res. 2007;22(1):93–100.
31. Schmitt JM, Hwang K, Winn SR, Hollinger JO. Bone morphogenetic proteins: an update on basic biology and clinical relevance. J Orthop Res. 1999;17(2):269–78.
32. Giannoudis PV, Einhorn TA. Bone morphogenetic proteins in musculoskeletal medicine. Injury. 2009;40(Suppl 3):S1–3.
33. Itoh K, Udagawa N, Katagiri T, et al. Bone morphogenetic protein 2 stimulates osteoclast differentiation and survival supported by receptor activator of nuclear factor-kappaB ligand. Endocrinology. 2001;142(8):3656–62.
34. Kaneko H, Arakawa T, Mano H, et al. Direct stimulation of osteoclastic bone resorption by bone morphogenetic protein (BMP)-2 and expression of BMP receptors in mature osteoclasts. Bone. 2000;27(4):479–86.
35. Nishimura R, Hata K, Ikeda F, et al. The role of Smads in BMP signaling. Front Biosci. 2003;8:s275–84.
36. Crandall DG, Revella J, Patterson J, Huish E, Chang M, McLemore R. Transforaminal lumbar interbody fusion with rhBMP-2 in spinal deformity, spondylolisthesis, and degenerative

disease--part 2: BMP dosage-related complications and long-term outcomes in 509 patients. Spine. 2013;38(13):1137–45.
37. Garrison KR, Shemilt I, Donell S, et al. Bone morphogenetic protein (BMP) for fracture healing in adults. Cochrane Database Syst Rev. 2010;(6):Cd006950.
38. Mont MA, Ragland PS, Biggins B, et al. Use of bone morphogenetic proteins for musculoskeletal applications. An overview. J Bone Joint Surg Am. 2004;86-A(Suppl 2):41–55.
39. Paul JC, Lonner BS, Vira S, Kaye ID, Errico TJ. Use of recombinant bone morphogenetic protein is associated with reduced risk of reoperation after spine fusion for adult spinal deformity. Spine. 2016;41(1):E15–21.
40. Valentin-Opran A, Wozney J, Csimma C, Lilly L, Riedel GE. Clinical evaluation of recombinant human bone morphogenetic protein-2. Clin Orthop Relat Res. 2002;395:110–20.
41. Vandermeer JS, Kamiya N, Aya-ay J, Garces A, Browne R, Kim HK. Local administration of ibandronate and bone morphogenetic protein-2 after ischemic osteonecrosis of the immature femoral head: a combined therapy that stimulates bone formation and decreases femoral head deformity. J Bone Joint Surg Am. 2011;93(10):905–13.
42. Kim HK, Aruwajoye O, Du J, Kamiya N. Local administration of bone morphogenetic protein-2 and bisphosphonate during non-weight-bearing treatment of ischemic osteonecrosis of the femoral head: an experimental investigation in immature pigs. J Bone Joint Surg Am. 2014;96(18):1515–24.
43. Cheng TL, Murphy CM, Cantrill LC, et al. Local delivery of recombinant human bone morphogenetic proteins and bisphosphonate via sucrose acetate isobutyrate can prevent femoral head collapse in Legg-Calve-Perthes disease: a pilot study in pigs. Int Orthop. 2014;38(7):1527–33.
44. Jeon JH, Piepgrass WT, Lin YL, Thomas MV, Puleo DA. Localized intermittent delivery of simvastatin hydroxyacid stimulates bone formation in rats. J Periodontol. 2008;79(8):1457–64.
45. Jeon JH, Puleo DA. Formulations for intermittent release of parathyroid hormone (1-34) and local enhancement of osteoblast activities. Pharm Dev Technol. 2008;13(6):505–12.
46. Mundy G, Garrett R, Harris S, et al. Stimulation of bone formation in vitro and in rodents by statins. Science (New York, NY). 1999;286(5446):1946–9.
47. Zou Y, Fisher PD, Horstmann JK, Talwalkar V, Milbrandt TA, Puleo DA. Synergistic local drug delivery in a piglet model of ischemic osteonecrosis: a preliminary study. J Pediatr Orthop B. 2015;24(6):483–92.
48. Chen Y-P, et al. Effectiveness of strontium ranelate in the treatment of rat model of Legg–Calve–Perthes disease. Ind J Orthop. 2018;52:380.
49. Horák P, Skácelová M, Kazi A. Role of Strontium Ranelate in the therapy of osteoporosis. J Rheum Dis Treat. 2017;3:050.
50. Little DG, Kim HK. Future biologic treatments for Perthes disease. Orthop Clin North Am. 2011;42(3):423–7.. viii
51. Kamiya N, Yamaguchi R, Adapala NS, et al. Legg-Calve-Perthes disease produces chronic hip synovitis and elevation of interleukin-6 in the synovial fluid. J Bone Miner Res Off J Am Soc Bone Miner Res. 2015;30(6):1009–13.
52. De Benedetti F, Rucci N, Del Fattore A, et al. Impaired skeletal development in interleukin-6-transgenic mice: a model for the impact of chronic inflammation on the growing skeletal system. Arthritis Rheum. 2006;54(11):3551–63.
53. Kaneshiro S, Ebina K, Shi K, et al. IL-6 negatively regulates osteoblast differentiation through the SHP2/MEK2 and SHP2/Akt2 pathways in vitro. J Bone Miner Metab. 2014;32(4):378–92.
54. Tamura T, Udagawa N, Takahashi N, et al. Soluble interleukin-6 receptor triggers osteoclast formation by interleukin 6. Proc Natl Acad Sci U S A. 1993;90(24):11924–8.
55. Yamaguchi R, Kamiya N, Adapala NS, Drissi H, Kim HK. HIF-1-dependent IL-6 activation in articular chondrocytes initiating synovitis in femoral head ischemic osteonecrosis. J Bone Joint Surg Am. 2016;98(13):1122–31.
56. Reid IR. Anti-resorptive therapies for osteoporosis. Semin Cell Dev Biol. 2008;19(5):473–8.
57. Schaller S, Henriksen K, Sveigaard C, et al. The chloride channel inhibitor NS3736 [corrected] prevents bone resorption in ovariectomized rats without changing bone formation. J Bone Miner Res Off J Am Soc Bone Miner Res. 2004;19(7):1144–53.

58. Sorensen MG, Henriksen K, Neutzsky-Wulff AV, Dziegiel MH, Karsdal MA. Diphyllin, a novel and naturally potent V-ATPase inhibitor, abrogates acidification of the osteoclastic resorption lacunae and bone resorption. J Bone Miner Res Off J Am Soc Bone Miner Res. 2007;22(10):1640–8.
59. Brunner HI, Ruperto N, Zuber Z, et al. Efficacy and safety of tocilizumab in patients with polyarticular-course juvenile idiopathic arthritis: results from a phase 3, randomised, double-blind withdrawal trial. Ann Rheum Dis. 2015;74(6):1110–7.
60. Burmester GR, Rubbert-Roth A, Cantagrel A, et al. A randomised, double-blind, parallel-group study of the safety and efficacy of subcutaneous tocilizumab versus intravenous tocilizumab in combination with traditional disease-modifying antirheumatic drugs in patients with moderate to severe rheumatoid arthritis (SUMMACTA study). Ann Rheum Dis. 2014;73(1):69–74.
61. De Benedetti F, Brunner HI, Ruperto N, et al. Randomized trial of tocilizumab in systemic juvenile idiopathic arthritis. N Engl J Med. 2012;367(25):2385–95.
62. Maini RN, Taylor PC, Szechinski J, et al. Double-blind randomized controlled clinical trial of the interleukin-6 receptor antagonist, tocilizumab, in European patients with rheumatoid arthritis who had an incomplete response to methotrexate. Arthritis Rheum. 2006;54(9):2817–29.
63. Yokota S, Imagawa T, Mori M, et al. Efficacy and safety of tocilizumab in patients with systemic-onset juvenile idiopathic arthritis: a randomised, double-blind, placebo-controlled, withdrawal phase III trial. Lancet (London, England). 2008;371(9617):998–1006.
64. Kim HK, Bian H, Aya-ay J, Garces A, Morgan EF, Gilbert SR. Hypoxia and HIF-1alpha expression in the epiphyseal cartilage following ischemic injury to the immature femoral head. Bone. 2009;45(2):280–8.
65. Zhang C, Li Y, Cornelia R, Swisher S, Kim H. Regulation of VEGF expression by HIF-1alpha in the femoral head cartilage following ischemia osteonecrosis. Sci Rep. 2012;2:650.
66. Zhang C, Yang F, Cornelia R, Tang W, Swisher S, Kim H. Hypoxia-inducible factor-1 is a positive regulator of Sox9 activity in femoral head osteonecrosis. Bone. 2011;48(3):507–13.

Index

D. S. Feldman, D. Paley (eds.), *Legg-Calvé-Perthes Disease*,
https://doi.org/10.1007/978-1-0716-0854-8