



Arthroplasty for Congenital Hip Deformity

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Development dysplasia of the hip is complex problem encompassing a spectrum of anatomical abnormalities. Total hip arthroplasty (THA) has emerged as the gold standard for management of secondary osteoarthritis due to hip dysplasia. Unique acetabular and femoral anatomy presents many challenges to reconstructive surgeons. However, the goals of THA for these patients mirror the goals of THA for nondysplastic patients. Classification systems, such as the Crowe and Hartofilakidis systems, have been created to facilitate discussion and management of these unique challenges, and numerous techniques have been described to meet these demands, with the goal of recreating normal anatomy for these patients. On the acetabular side, surgeons aim to restore the native hip center will ensuring adequate acetabular implant coverage, while on the femoral side, they seek to restore normal femoral anteversion and provide equal leg lengths. This article details the various challenges reconstructive surgeons face, and the tools at their disposal to achieve the stated goals. When properly executed, THA for development dysplasia of the hip has shown excellent results, with no difference in outcomes for mildly dysplastic hips, though higher rates of mechanical failure for severely dysplastic hips.

Oper Tech Orthop 29:100724 © 2019 Elsevier Inc. All rights reserved.

KEYWORDS Hip arthroplasty, Hip deformity, Congenital hip, Developmental dysplasia, DDH

Introduction

Developmental dysplasia of the hip (DDH) is a common cause of hip dysfunction, and the primary cause of hip osteoarthritis in young patients. Of patients under 50 years old undergoing total hip arthroplasty (THA), 20% are due to secondary arthritis from DDH.¹ Reports suggest that 2.6% of all THAs are performed are due to DDH, at an average age of 42year old.^{2,3} The dysplastic hip presents a unique set of anatomical challenges during total hip reconstruction. Dysplastic acetabulum provide varying degrees of femoral head coverage, ranging from mild uncoverage to frank dislocation. Femoral abnormalities include excessive anteversion and valgus neck-shaft angles. These anatomical abnormalities result in abnormal contact stresses, predisposing patients to accelerated degenerative changes. While nonarthroplasty options, including femoral and periacetabular osteotomies, are useful

in young, active patients without marked osteoarthritis, for adults who present with symptomatic degenerative changes, THA remains the standard of care. Though the added technical complexity classically resulted in higher failure and revision rates, modern techniques have resulted in patient reported outcomes and functional results that are comparable to primary total hip arthroplasties.⁴⁻⁶

Case Presentation

A 26-year-old female presented to clinic, complaining of dull, aching right groin pain that worsened with activity and after long days. Reports the initial pain improves after the first few steps, and that she is able to walk >10 blocks. She occasionally uses a cane, but only as needed for comfort. She has participated in formal physical therapy which helped alleviate her low back and right thigh pain. For residual pain, she takes Nonsteroidal anti-inflammatory drugs (NSAIDs). Her past medical history is significant for right hip dysplasia and hypothyroidism. Her surgical history is significant for right hip anteromedial open reduction at 6 months, reduction of congenitally dislocated right patella at 1 year, and left distal

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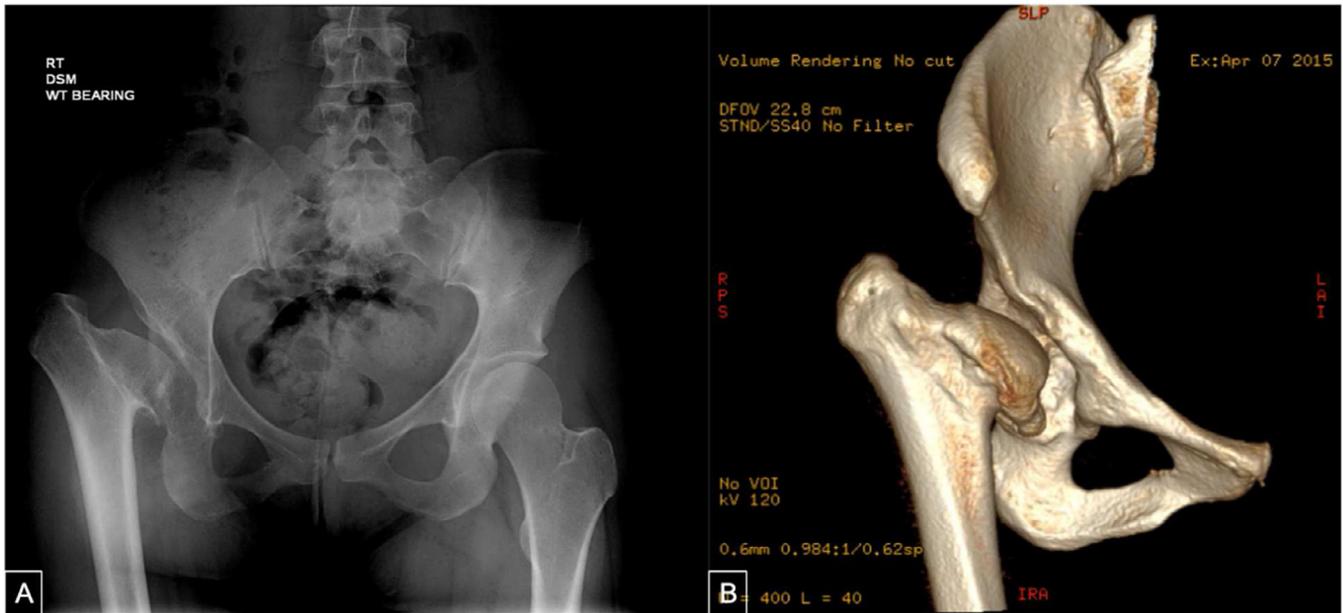


Figure 1 A 26-year-old female with DDH. (a) Radiographs and (b) computed tomography (CT) scan reveal femoral head avascular necrosis with a shallow, anterosuperiorly deficient acetabulum.

femoral epiphysiodesis at 11 years. On exam, she is 5'3", 130 lbs, BMI 23.0. Her right leg is 2 cm shorter than her left, and she walks with a slight limp. Her right hip flexes to 110°, abducts to 20°, externally rotates to 60°, and adducts to 30°. She has 5/5 abductor motor strength and is otherwise neurovascularly intact. Radiographs and computed tomography scan reveal femoral head avascular necrosis with a shallow, anterosuperiorly deficient acetabulum (Fig. 1a-b).

Anatomy

Though each patient has unique anatomy, dysplastic hips share certain common abnormalities that become more pronounced with increasing severity of DDH. Dysplastic traits are typically described as acetabular or femoral. Dysplastic acetabula are characterized as anteverted, shallow, lateralized, and by anterolateral and superior bony deficiencies. Dysplastic femora also have excessive anteversion, with smaller femoral heads, shorter necks, narrower canals, less anterior bowing, and increased neck-shaft angles. A combination of these deficiencies, with small, lateralized femoral heads articulating with small acetabula, leads to a more lateral hip center of rotation and a smaller contact area. These increased contact pressures predispose patients to the early onset degenerative changes classically described in this population. Treating surgeons must also pay close attention to the soft tissues surrounding the hip joint. As DDH becomes more severe and the hip migrates further laterally and proximally, abductor mechanism weakens, hip capsule thickens, psoas tendon hypertrophies, and rectus, adductors, and hamstrings become chronically shortened, leading to classic gait imbalances. The femoral nerve often exists the pelvis more laterally and superiorly and must be paid attention intraoperatively.

The sciatic nerve can also be chronically shortened and must be carefully examined, with some studies reporting rates of sciatic nerve palsy as high as 13% following subsequent THA.⁷

Patient Examination

Though hip dysplasia typically presents as a painless limp, in children, young adults typically present with insidious onset, activity related groin or lateral hip pain, often with a leg length discrepancy and a limp. Progressive subluxation of the femoral head leads to proximal migration of the femur. This causes the abductor muscles to become more transversely oriented, and less efficient. Pain is typically reproduced with hip extension and external rotation. With progressive subluxation and the development of secondary degenerative changes, patients may also complain of catching and locking symptoms consistent with labral or chondral pathology.

Plain radiographs, anteroposterior (AP), lateral, and false-profile view of the affected hip and AP of the pelvis, are crucial for initial evaluation of hip pathology. In the absence of obvious structural abnormalities on radiographs, an MRI can help delineate further labral or chondral pathology. Computed tomography scans can also be very useful for better assessing structural abnormalities and can be very helpful for future surgical planning. Finally, arthroscopy, although controversial in the setting of pre-existing degenerative disease, can be an option to address loose bodies, labral, and chondral injuries, but often does little to mitigate the progression of degenerative changes. Some have even suggested that such intervention can destabilize the tenuous hip and accelerate the progression of degenerative changes.⁸

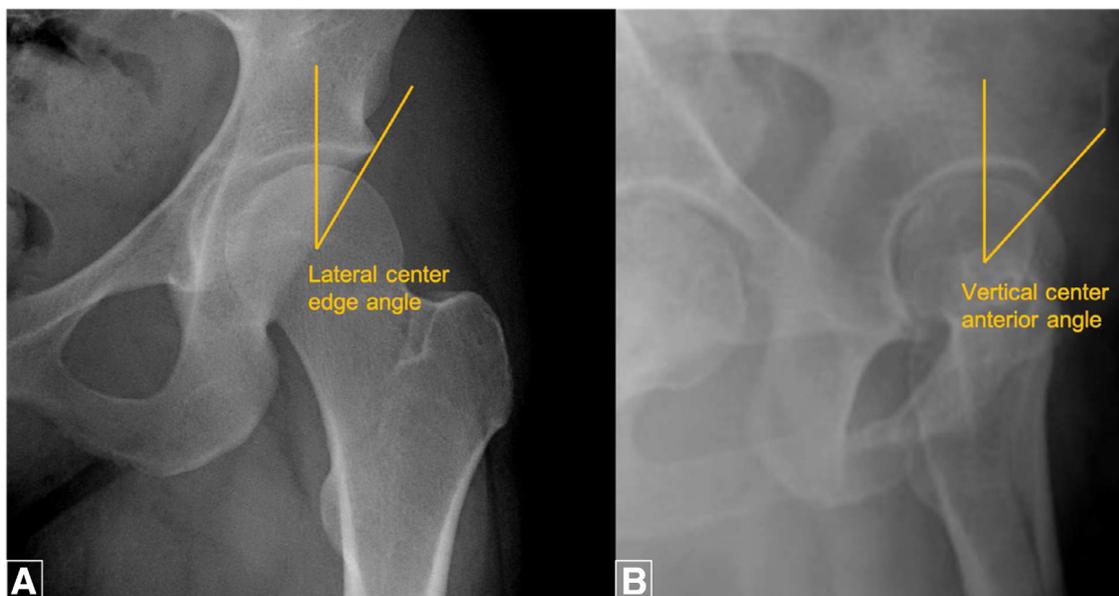


Figure 2 (a) Lateral center edge angle (CEA), on AP radiograph of the hip, a vertical line through the center of the femoral head and another through the center of the head and lateral edge of the acetabulum (Normal CEA is 25° - 40° , abnormal $<20^{\circ}$). (b) Vertical center anterior angle (VCA), on false profile view of the hip, a vertical line through the center of the femoral head and a line from the center of the femoral head to the anterior edge of the acetabulum (Normal VCA is 25°).

Diagnosis and Classification

Classification of dysplastic hips is based on the severity of the anatomical abnormality. These systems allow for patient assessment, guide surgeons with regard to operative planning, and allow for outcomes comparison following varied surgical techniques. The 2 most commonly used are the Crowe and Hartofilakidis classification systems. Both are based on AP radiographs of the pelvis, which allow for examination of antero-lateral acetabular coverage of the femoral head. A few other commonly cited measurements are based on radiographs, although not factored into the classification systems. On AP of the hip, a vertical line through the center of the femoral head and another through the center of the head and lateral edge of the acetabulum, create the lateral center edge angle (Fig. 2a). Normal center edge angle is 25° - 40° , with abnormal being $<20^{\circ}$. On false profile view of the hip, a similar angle is measured from a vertical line through the center of the femoral head and a line from the center of the femoral head to the anterior edge of the acetabulum, creating the vertical center anterior angle (Fig. 2b). Normal vertical center anterior angle is 25° . AP of hip also allows measurement of the femoral neck shaft angle, normal 120° - 135° .

Crowe et al created 4 categories based on AP radiographs of the pelvis, specifically based on proximal migration of the femoral head.⁹ On AP radiographs, the vertical distance between the interteardrop line and the junction between the femoral head and the medial edge of the femoral neck is measured (Fig. 3). The ratio between this distance and the vertical diameter of the dysplastic femoral head is the amount of subluxation. As such, if the vertical diameter of the femoral head is 4 cm and the head-neck junction to teardrop distance

is 2 cm, the hip is subluxated 50%. For severely deformed femoral heads, the predicted diameter of the femoral head is used, calculated as 20% of the distance from inferior most aspect of the ischial tuberosity and the superior most aspect of the iliac crest on true AP radiograph. Based on these measurements, Type 1: $<50\%$ subluxated, Type II: 50% - 75% subluxated, Type III: 75% - 100% subluxated, and Type IV: $>100\%$ subluxated (Fig. 4). This system is most commonly used to determine type of acetabular and femoral reconstruction during THA. The other classification that's commonly discussed but not commonly used clinically, is the Hartofilakidis classification.¹⁰ Type A: dysplasia, the femoral head remains within the acetabulum with minimal subluxation and there is some deficiency in the superior wall with a shallow acetabulum; Type B: low dysplasia, the femoral head creates a pseudoacetabulum superior to the true acetabulum with complete absence of the superior wall and a shallow acetabulum; Type C: high dysplasia, the femoral head is completely uncovered and has position superiorly and posteriorly, the true acetabulum is completely deficient and excessive anteversion may be present (Fig. 5).

Treatment

Nonarthroplasty

Though THA is the mainstay of surgical treatment for these patients, a few nonarthroplasty surgical options are worth mentioning. The most popular is the Ganz osteotomy, also known as the PAO, or periacetabular osteotomy. It is a redirection osteotomy indicated for skeletally mature patients

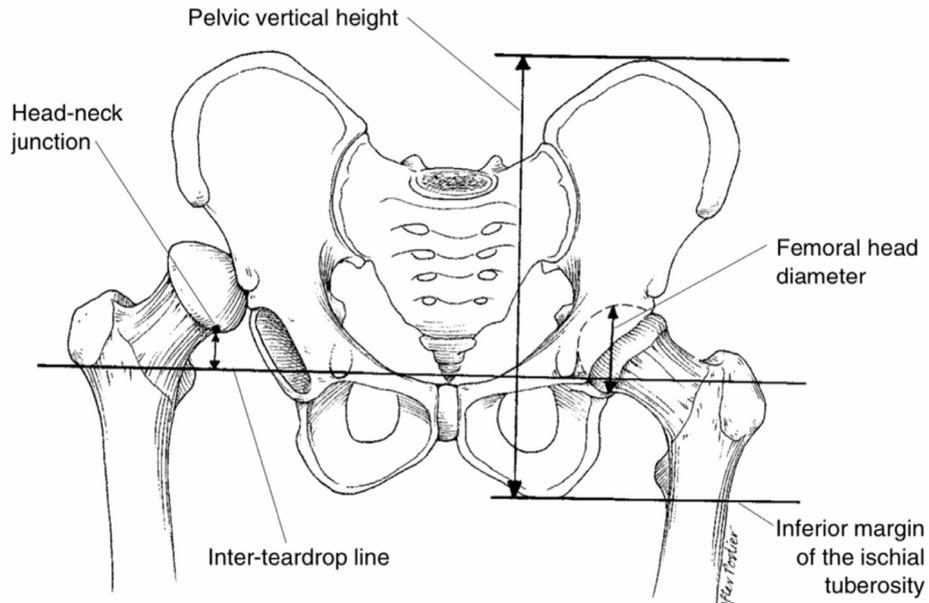


Figure 3 On AP radiograph schematic detailing measurements used for determination of Crowe classification.

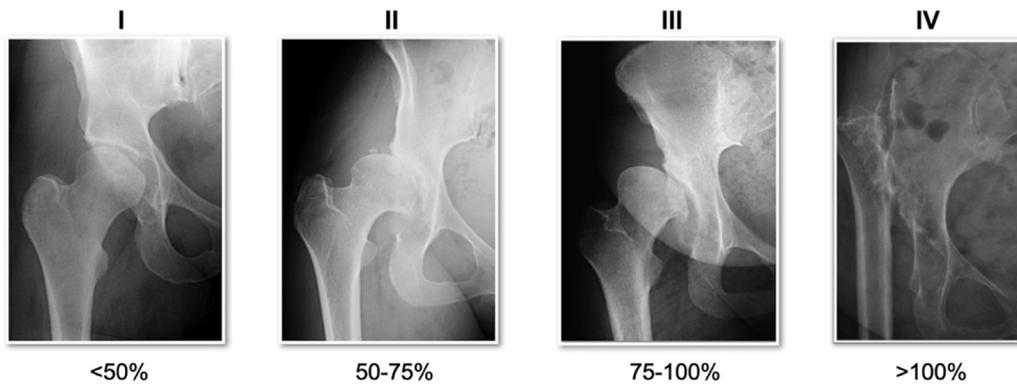


Figure 4 Crowe classification.

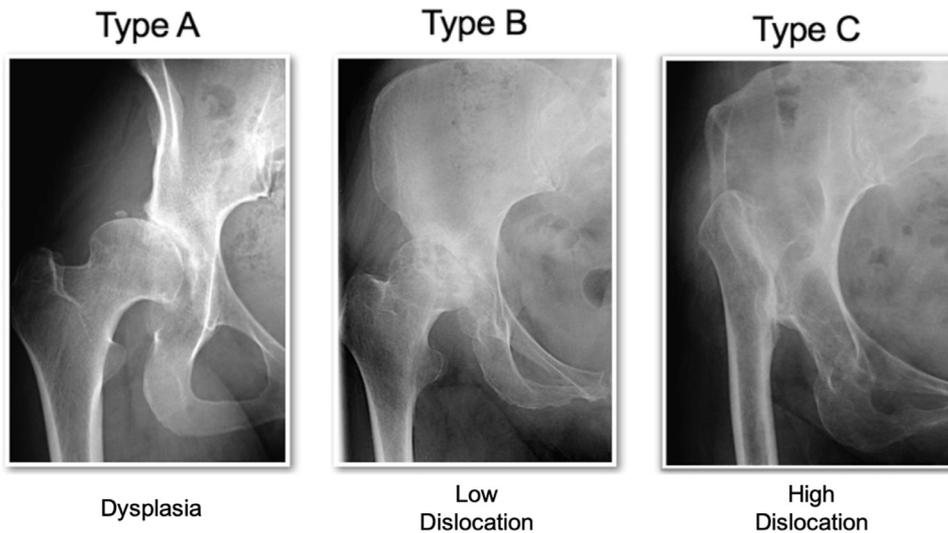


Figure 5 Hartofilakidis Classification.

with symptomatic dysplasia who have a concentrically reduced hip and no/minimal osteoarthritis. It involves making cuts through the pubis, ilium, and ischium and rotating the entire acetabulum. It maintains the native hyaline cartilage joint and preserves the posterior column which allows patients to weight bear, making it a good option for patients with symptomatic dysplasia without degenerative changes necessitating a THA. However, its technically very demanding, even in experienced hands, with a reported complication rate as high as 15%.¹¹ Though it can delay future THA, it can also complicate that operation when it eventually occurs by retroverting the native acetabulum.

Arthroplasty Surgical Technique

Given the difficulty and unreliable results that accompany pelvic osteotomies, the mainstay of surgical treatment for hip dysplasia with secondary osteoarthritis is THA. However, dysplastic hips require extra consideration given the aberrant anatomy.

Approach

Although some suggest that the algorithm for selecting surgical approach is based on the degree of dysplasia, more often than not it based on surgeon preference. Some have argued that for predominately dysplastic acetabula, the anterior approach provide superior exposure and should be utilized. Conversely, if the predominate concern is shortening in a severely dysplastic case and need improved access to the femur, or if better visualization of the sciatic nerve is desired, the posterior approach may be better equipped. The direct lateral (Hardinge) has also been described for severely dysplastic cases, as it can provide elongated exposure to the femur and allow for shortening.

Acetabular Reconstruction

The ultimate goal of acetabular reconstruction in any THA is restoration of the native hip center with complete acetabular implant coverage. In severely dysplastic hips, this represents a fine balancing act, as femoral heads that are proximally subluxated and lateralized cannot achieve both parameters. Though no good studies could be found, expert opinion suggests that 30%-50% of lateral uncovering of the acetabular component is acceptable.^{12,13} Lack of support of the acetabular implant increases stresses at the bone-implant interface, increasing the risk of mechanical failure. In revision settings, stable trails with 50% acetabular coverage are often acceptable, and this seems to translate to the dysplastic hip reconstruction. Thankfully, surgeons have at their disposal numerous tools to achieve these goals. Clinically, the complexity of acetabular management is dictated by the Crowe classification system.

For mildly dysplastic, Crowe I hips, the acetabular component can usually be placed in the true acetabulum without augmentation. There is typically sufficient coverage such that the acetabular component can be medialized without the risk of protrusio.

Crowe II and III hips present the greatest challenge for surgeons. With significant subluxation of the femoral head without frank dislocation and continued weight bearing, patients developed significant superolateral acetabular bony deficiency prevents adequate coverage for standard cups, occasionally necessitating the use of special components such as extra small cups (38-50 mm), metal augments, and autografts. In some cases, sufficient lateral coverage of the socket can be obtained at or near the normal anatomical position by deepening the socket with reaming to the medial wall. When this is not feasible, 3 alternative methods of reconstruction can be used.

The first is acetabular augmentation with superolateral metal augments or bone grafting (Fig. 6). Augments in

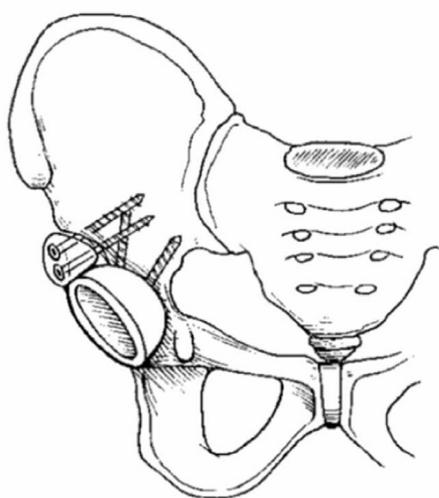


Figure 6 Acetabular augmentation, schematic, and radiographs.

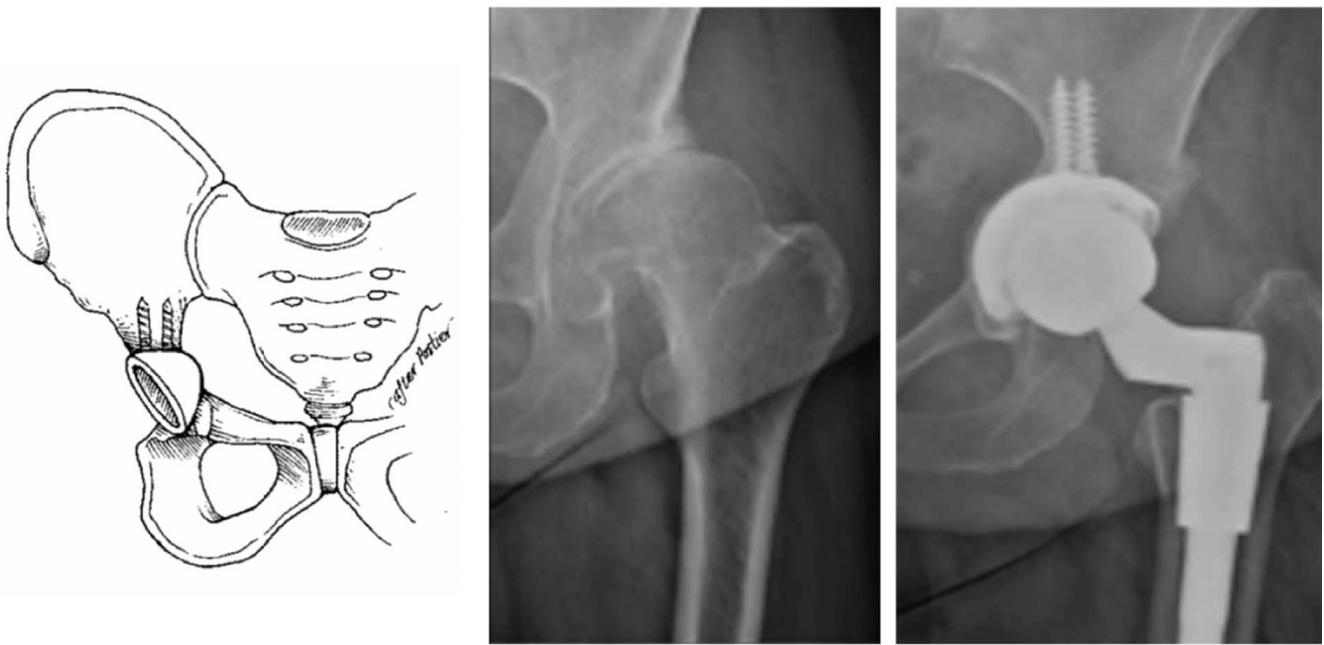


Figure 7 Cotyloplasty.

general can help provide cup stability allowing for anatomical cup positioning. Additionally, autologous bone grafting provide coverage and increase bone stock which may be critical should revision surgery be necessary.¹⁴ Autograft can easily be harvested from the patient's native femoral head, and with sufficient coverage the native hip center can be restored, recreating native hip biomechanics. However, bone grafting increases the technical complexity of the case and may be complicated by nonunion, graft resorption, and ultimately graft failure with cup loosening.^{15,16}

The second option is medialization of the native acetabulum (Fig. 7) First described by Hartofilakidis in 1996, cotyloplasty involves perforation of the medial wall of the shallow acetabulum by controlled overreaming.¹⁰ Prior descriptions involved osteotomes and T handle curettes, while modern techniques rely on undersized reamers, deliberately fracturing through the medial wall and creating purposeful protrusion.^{17,18} Care must be taken to ream through the medial wall till the medial periosteum is encountered. This technique involves using undersized reamers and careful reaming through the medial wall until the medial periosteum is seen. Successful medialization allows surgeons to implant a porous coated press fit component and obviates the need for bone graft augments. While obvious advantages of this technique are attributable to restoration of the native hip center, loss of medial bone can make revision surgery more difficult and increases the risk of catastrophic failure with cup migration into the pelvis.⁴

The third, and least desirable option, is reconstruction of the acetabulum in a superior location (high hip center) (Fig. 8) Placing the cup in a high pseudoacetabulum can increase implant coverage, obviating the need for autograft and thus the associated complications, while inherently improving biologic fixation of the component. Given improved bone stock, it also allows surgeons to continue

using cementless cups. Additionally, high hip centers in severely dysplastic hips circumvent the need for concomitant femoral shortening osteotomies. However, high hip centers are not anatomical and thus have unfavorable biomechanics. The abductors are disadvantaged, with some studies reporting nearly 30% reduction in the lever arm, the implant experiences higher contact stresses, and thus increased rates of cup loosening have been reported, although others have found that with appropriate medialization, loosening rates are no different.^{19,20} Additionally, higher hip centers are associated with increased rates of anterior impingement of femoral component on anterior inferior iliac spine in flexion, and the ischial tuberosity in extension. Given the morphology of the pelvis, surgeons must be careful to avoid concurrent lateralization of the hip center as it becomes more superior, as that too increases lever arm and joint reactive forces. Studies have shown that every millimeter of lateral displacement of the acetabular cup relative to the native hip center increases the hip load by 0.7%, and for every millimeter of proximal displacement, there is a 0.1% increase in hip load.²¹ Surgeons should try to restore normal hip center whenever possible. However, if superior location is deemed necessary, it is imperative that the surgeon avoid lateralization of the hip center.

Interestingly, Crowe IV hips are characterized by the complete dislocation of the femoral head. Though the acetabulum is typically underdeveloped and shallow, anterosuperior acetabular deficiencies are not usually a problem since the femoral head has not been articulating with the native socket. Good acetabular bone stock allows surgeons to place the acetabular component at the native hip center with traditional reaming techniques and without the use of autograft. Given chronic disuse of the native acetabulum, surgeons may need an extra small acetabular component. The bone can also be abnormally soft, so care must be taken not to overream.

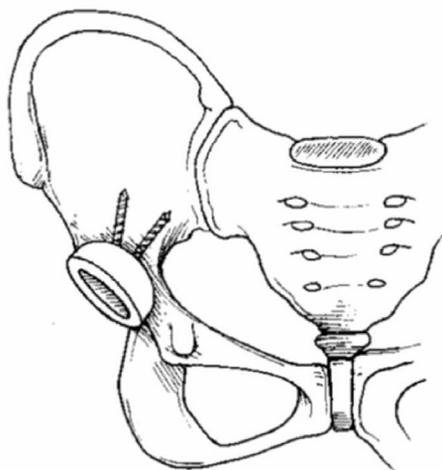


Figure 8 High hip center.

Femoral Reconstruction

The crux of femoral reconstruction is to recreate normal femoral anatomy. Dysplastic femurs classically have excess anteversion of the femoral head, excess coxa valga, small and misshapen medullary canals that are narrower and straighter, and posteriorly located greater trochanters. Clinically, determination of femoral component anteversion and the amount of femoral shortening required to prevent excess leg lengthening follows acetabular reconstruction.

For Crowe I/II hips, when there is minimal change to hip center of rotation, femoral length is not an issue. Without the need for shortening osteotomies, implant choice is based on bone quality, femoral geometry, and of course, surgeon preference. Four implant options have been well described for this purpose: nonmodular proximally porous-coated metaphyseal filling, cemented DDH, extensively porous-coated, and modular femoral implants.⁴

Nonmodular proximally porous-coated metaphyseal filling implants come in a few varieties, both single and dual taper designs, and are commonly used implants in primary, nondysplastic hips. As such, these are reserved for mild anatomical abnormalities.

Cemented DDH stems are narrower with minimal metaphyseal flare, and depending on implant, low short necks. They allow orientation of the stem independent of a patient's native anteversion, providing increased flexibility in dysplastic femora.

Extensively porous-coated implants have small metaphyseal cross sections and are able to bypass metaphyseal abnormalities and gain distal fixation, allowing them to compensate for abnormal anteversion. However, they cannot be used for very small femoral canals due to the risk of fatigue failure with very-small-diameter porous coated stems. The Wagner stem was originally designed in 1987 specifically for patients with severe proximal femoral bone loss. With its circular cross-sectional design, it too allows stem

orientation independent of the patient's native version. Narrow stem diameters (<12 mm) are often required.

Modular femoral implants allow metaphyseal fill with uncemented, porous coated implants while still allowing surgeons to independently modulate stem version. These implants allow the mix and match of metaphyseal shape and diaphyseal size. The Depuy S-ROM is a commonly used implant for this purpose, providing very narrow stem diameters from 8 to 12 mm.

For Crowe III/IV hips, marked change to the hip center of rotation often requires a concomitant femoral shortening osteotomy. Primary reasons for femoral shortening include reducing the hip prosthesis and avoiding lengthening of the sciatic nerve, greater than 3-4 cm lengthening is associated with sciatic nerve injury.^{7,22} Shortening helps overcome soft tissue contractures, particularly from the rectus, but also from the hamstrings and adductors facilitating hip reduction. Appropriate shortening also avoids residual leg length discrepancies following arthroplasty. Clinically, even for these severely dysplastic hips, surgeons should first attempt reduction with a short neck on the femoral trial. If this reduces then no further need for shortening. However, if the hip reduces but there is loss of sciatic nerve signaling, then shortening is required. Obviously if the hip is not reducible, then shortening is required. For these dysplastic cases, somatosensory evoked potential monitoring is crucial for sciatic nerve monitoring.

Though a few different techniques have been described, the most reliable and widely used is the shortening subtrochanteric osteotomy. These preserve the metaphyseal area, which is important for implant fixation, and allows simultaneous correction of femoral angulation and anteversion.

As with standard THA, the femoral neck cut is made 1 cm proximal to the lesser trochanter. The femur is prepared by reaming before making any osteotomy cuts, and often the proximal femur piece fragment can be broached or reamed to prepare for the chosen implant. In cases with severe

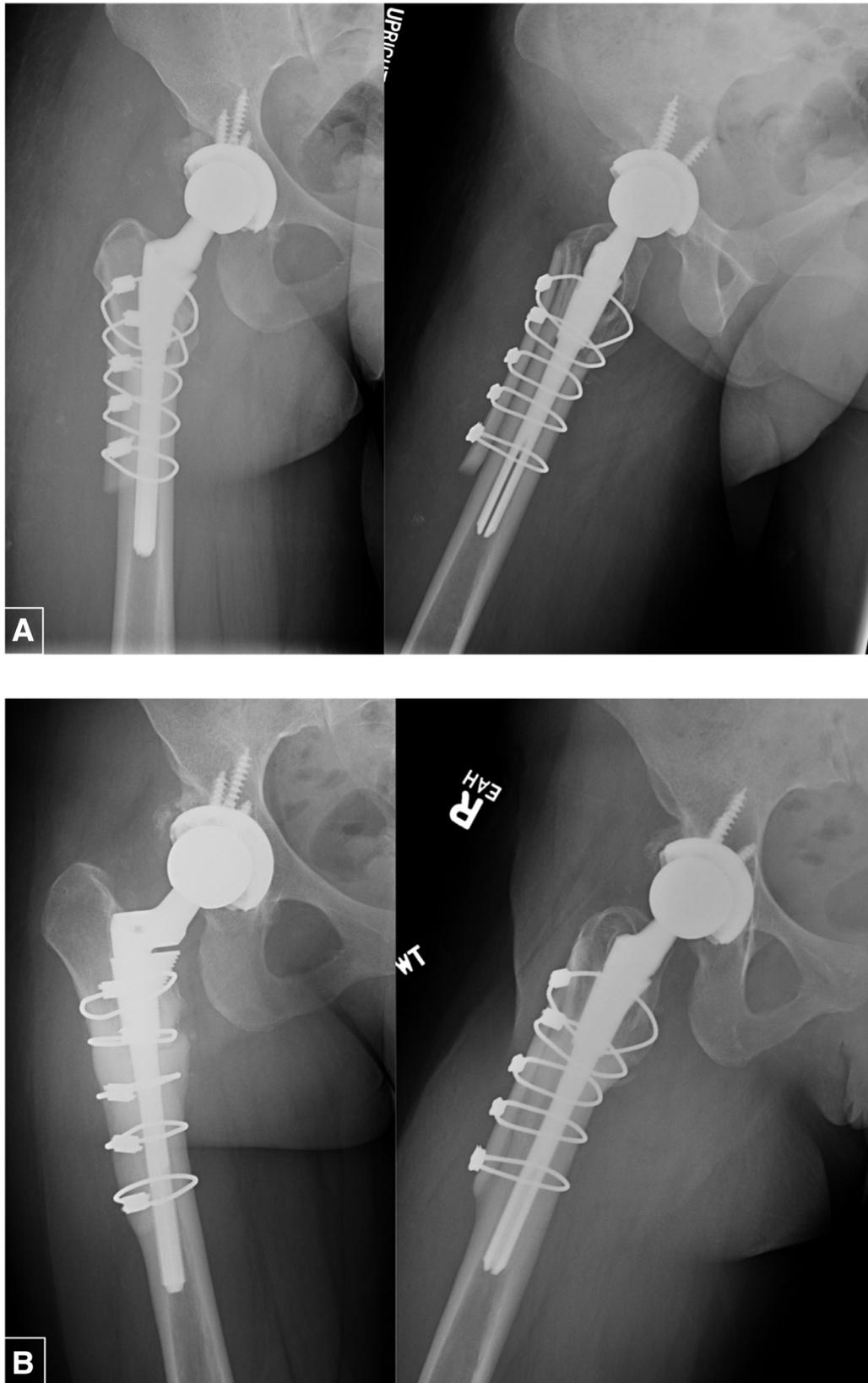


Figure 9 A 26-year-old female with DDH status post RTHA with acetabular medialization and subtrochanteric shortening osteotomy. (a) Immediate postop, cortical strut graft cerclage wires. (b) Two years post-op, radiographs show excellent healing.

femoral deformity, definitive preparation of the distal segment must follow the osteotomy in order to get proper implant orientation and version. The second femoral osteotomy should be based on the preoperative template that estimates the length of shortening required to facilitate hip reduction. This is confirmed intraoperatively, after the femoral trial is placed in the proximal segment, the implant is reduced, and the amount of overlap between proximal and distal segments present once leg lengths are made equal can be determined. The osteotomized diaphyseal segment is saved as it provides robust autograft for osteotomy augmentation. Once leg lengths have been confirmed, the hip and osteotomy site can all be reduced, with trial components in place. Free hand trimming of bone ends may be required to achieve proper fit between the osteotomy cuts. Many osteotomy cuts have been described, including transverse, oblique, z-shaped, and chevron cuts.^{23,24} Each cut has its own advantages and disadvantages, providing differing degrees of rotational control. The transverse cut is the simplest and provides the most flexibility. Transverse osteotomy sites can be easily adjusted if more shortening is required, but surgeons must take care to properly rotate the distal segment as malrotation is most likely with this simple osteotomy.²⁵ Of note, prophylactic cabling of the distal segment has been described to prevent iatrogenic fracture of these dysplastic femora. Once the hip and femur are reduced with the final implant in place, the previously resected femoral shaft can be split and used as onlay autograft.²⁶ If this bone is of particular poor quality, surgeons can use cortical strut grafts or augment their fixation with a hook plate. Cerclage cables are used as necessary to secure these augments, although care should be taken to avoid unnecessary soft tissue disruption.

Postoperative Care

Postoperative care is similar to that for standard THA. In addition to the typical anticoagulation and pain control regimen, early mobility remains a tenant of postoperative care. For mildly dysplastic hips that do not require a femoral osteotomy, patients are weight bearing as tolerated immediately postop. If shortening osteotomy is required, sufficient time must be given to allow for union, typically 6-8 weeks of touch down weight bearing.

Outcomes

For patients with developmental hip dysplasia, THA significantly improves Harris hip scores and remains the gold standard of treatment for patients with secondary arthritis. Thillemann et al in their analysis of the Danish Hip Arthroplasty Registry found that for Crowe I and II hips, long-term outcomes were no different between patients with this mild dysplasia and those without hip dysplasia.² Of the nearly 54,000 patients included in their review, over 2000 had osteoarthritis secondary to dysplasia, finding no difference in the risk of revision from 0.5 to 12 years. However, patients with Crowe III or IV dysplasia were twice as likely to sustain early dislocation. Similarly, Engesaeter et al in their review of

Norwegian Arthroplasty Register data that included nearly 60,000 primary THA and over 7000 THA for dysplasia, found that at 15-year follow-up, THA revision was 1.5-2× more likely in patients with dysplasia.⁶

Case Resolution

Our 26-year-old female with Crowe III dysplasia and femoral head AVN underwent THA with excellent results. Watson Jones approach was used, and the cup was placed in the native hip center with medialization. She was fitted with a 46 mm cup with 3 screws, achieving 70% cup coverage with native acetabulum. A subtrochanteric femoral shortening osteotomy was used to prevent overlengthening, and the femur was fitted with an S-ROM with a small proximal sleeve, but standard stem and neck. A cortical strut graft was also placed posteriorly to augment the osteotomy fixation, with 5 cerclage wires (Fig. 9a). At 2 years post-op, she had equal leg lengths, full painless hip range of motion, with excellent healing on follow-up radiographs (Fig. 9b).

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