Total hip arthroplasty combined with subtrochanteric shortening osteotomy for Crowe type IV hip dysplasia in dwarfism: a case report

Mengxuan Yao and Huijie Li

Abstract
Dwarfism is a condition of extreme short stature. Total hip arthroplasty (THA) in patients with dwarfism is a very demanding procedure due to their specific joint deformity and small bone size, which increases the technical difficulty of the THA procedure in such patients with hip dysplasia. This current case report describes a 29-year-old female patient that was admitted due to shortening of the right lower limb, as compared with the contralateral side, which had been present for 18 years. She also complained of pain in the right hip that had been present for 2 months. The hip pain was aggravated by physical exertion, but relieved by rest. She had pituitary dwarfism without mental retardation or delayed sexual development and Crowe type IV dysplasia of the right hip. A THA combined with femoral shortening osteotomy was undertaken, which resolved the pain symptoms and improved her ability to undertake activities of daily living such as walking without pain. Dwarfism with hip dysplasia is a rare but extremely challenging problem that can be successfully treated with THA combined with femoral shortening osteotomy using an S-ROM stem.

Keywords
Dwarfism, developmental dysplasia of the hip, total hip arthroplasty, osteotomy

Date received: 1 November 2019; accepted: 2 April 2020

Introduction
Developmental dysplasia of the hip (DDH) can lead to gait abnormalities, limited abduction, reduced lower limb strength and an increase in the incidence of...
degenerative joint disease; and total hip arthroplasty (THA) is still required for patients with severe end-stage hip osteoarthritis.\textsuperscript{1,2} THA has been confirmed as an effective method to relieve pain, improve hip function and correct leg length in patients with this disease.\textsuperscript{3} However, patients with Crowe type IV DDH often have anatomical abnormalities of the hip including deformation of the femoral head, short and excessive anteversion of the femoral neck, narrowing of the femoral medullary cavity, acetabular dysplasia and reduced depth, insufficient bone stock above and posteriorly located greater trochanter and muscle contractures.\textsuperscript{4,5} Therefore, performing THA in such patients is rather technically demanding.\textsuperscript{6}

Total hip arthroplasty in patients with dwarfism is a very demanding procedure due to their specific joint deformity and small bone size, which increases the technical difficulty of THA application in such patients with hip dysplasia.\textsuperscript{7}

This current case report describes a patient with dwarfism with Crowe type IV DDH that underwent a posterolateral approach subtrochanteric shortening osteotomy, combined with total hip replacement with an S-ROM prosthesis. Good clinical results were achieved by applying that approach in this case.

Case report

A 29-year-old female was admitted to the Department of Osteopathy, The Third Hospital of Hebei Medical University, Shijiazhuang, Hebei Province, China in April 2018 due to shortening of the right lower limb, as compared with the contralateral side, which had been present for 18 years. She also complained of pain in the right hip that had been present for 2 months and the hip pain was aggravated by physical exertion, but pain relief occurred after rest. The patient was unable to walk more than one-block distance due to the severe pain and could not stand for more than half an hour. She had pituitary dwarfism without mental retardation or delayed sexual development. The Institutional Review Board of the Third Hospital of Hebei Medical University Ethics Committee requested that the authors seek consent from the patient for publication of this case report without full review and approval being required by the committee. Verbal consent was obtained from the patient by telephone interview for investigating and reporting her case. All necessary patient information was collected and anonymized.

The patient was of short stature, with a height of 133 cm and a weight of 37 kg. There was evidence of scoliosis (physiological curvature of the spine), the right lower limb was approximately 4 cm shorter than the left lower limb; and the right hip had reduced mobility, mainly in adduction and internal rotation. The Trendelenburg sign was positive on the right side and the Patrick test was also positive. X-ray and computed tomography examinations revealed Crowe type IV dysplasia of the right hip. The diagnosis at admission was made in the following order: (i) right hip dysplasia Crowe type IV; (ii) dwarfism.

Treatment options were discussed with the patient. The S-ROM prosthesis (DePuy Synthes Joint Reconstruction, Warsaw, IN, USA) was selected for use based on the patient’s narrow canal. A preoperative X-ray template was employed to develop a thorough acetabular reconstruction and femoral reconstruction plan. A posterolateral approach was selected. Meanwhile, the patient was given intravenous antibiotics to prevent infection (1 g cefazolin sodium intravenously once before surgery).

After satisfactory anaesthesia, the patient was placed in the left lateral decubitus position. The surgical site was routinely
disinfected with iodine alcohol, paved with a sterile sheet and operated on through a posterolateral approach. After dislocation of the hip joint, the femoral head was removed using an L-shaped osteotomy suitable for the S-ROM prosthesis. The acetabulum was exposed and the acetabular labrum was removed. Then, the residual round ligament tissue in the acetabular fossa was cleaned and the transverse ligament of acetabulum was exposed as the inferior edge of the acetabulum. Next, after identifying the true acetabular base, the acetabulum was reamed and widened and deepened, with an acetabular abduction angle of 40° and an anteversion angle of 15°. The file was ground to the subchondral bone and the acetabular prosthesis was placed after satisfactory oozing. A small cup size was used to obtain a bony coverage of the upper 70% of the cup. The shell was not augmented with screws and all acetabular components were placed medial to the original acetabular medial wall.

In the preparation of the femur, the proximal femur was first secured with a strap to prevent cortical splitting. Then, a canal opener was placed laterally on the posterior edge of the neck resection and used to open the pulp at the piriform sinus of the femur. Distal reaming was further initiated with an 8-mm drill, which was increased by 0.5 mm at a time until the cortex was palpated. No other reamer model was used after the cortex was palpated with the 8-mm drill. The proximal femoral taper was reamed with the appropriate instrumentation until the reamer body contacted the cortical bone of the proximal femur. A triangular reamer was used at the femoral moment until the cortex was sanded. Subtrochanteric femoral shortening transverse osteotomy was also required to facilitate easier hip reduction and to avoid excessive lengthening of the limb and possible stretching of the sciatic nerve. Approximately, from 1.0 to 1.5 cm below the lesser trochanter, a transverse osteotomy was performed using an oscillating saw perpendicular to the longitudinal axis of the femoral shaft. The degree of shortening was determined based on the preoperative radiographic measurements and intraoperative traction examinations. Then, the trial corresponding to the final conical reaming and triangular reaming were placed on the trial stem of the prosthesis, the trial sleeve was placed, then the trial femoral prosthesis was placed and the anteversion angle of the trial femoral prosthesis was adjusted to 20°. A prosthesis stability check was performed and a modular prosthesis was placed after satisfactory range of motion. The patient was implanted with a range of motion three-piece cementless titanium alloy femoral prosthesis. The size of the femoral head used was 22 mm and the size of the porous-coated shell was 44 mm.

Routine postoperative prophylaxis for infection (1 g cefazolin sodium intravenously four times a day for 1 day) and deep venous thrombosis of the lower extremities (10 mg rivaroxaban orally once a day for 1 week) was undertaken. On the 2nd day after the operation, the patient performed active hip flexion and extension exercises without weight bearing on the bed. In the early postoperative stage, the patient was forbidden to bear weight on the ground.

The preoperative Harris hip function score was 54 points. At 1, 3 and 6 months postoperatively, the Harris score was 39, 65 and 77 points, respectively. The postoperative hip function was improved. During the last follow-up, the patient’s lower limbs were of equal length; and the active and passive movements of the right hip were good, without discomfort. Moreover, the daily living activities were improved, with the patient displaying no difficulties when walking without an assistive device or when walking up and down the stairs. There was no claudication, no discomfort.
when sitting and no surgery-related complications. The radiographic results are presented in Figure 1.

Discussion

Patients with dwarfism frequently undergo THA as a result of early joint degeneration. A large variety of disorders can lead to dwarfism, ranging from endocrine disorders such as hypothyroidism and growth hormone deficiency to genetic disorders causing skeletal dysplasia. Patients with dwarfism tend to undergo THA due to early joint degeneration and the influence of factors, including small prosthesis size, acetabular and femoral dysplasia, and poor bone quality, can pose surgical challenges. A previous study analysed 102 patients with dwarfism that underwent THA and found that THA undertaken in patients with dwarfism achieved comparable results with THA undertaken in a non-dwarfism population with regard to implant survivorship; however, there was a trend toward increased periprosthetic fractures and wear-related failures. Various methods have already been reported to overcome the problems caused by the small body of the patient. For example, a previous case report considered THA in patients with dwarfism to be a safe and effective procedure if accurate preoperative planning was performed. Customized prosthetic implants have also shown superiority in patients with dwarfism, but the biggest limitation or disadvantage associated with custom use is the prohibitive costs. A previous report described a case of total hip replacement performed in a patient with pseudoachondroplasia (height 122 cm and weight 46 kg) with a dysplastic acetabular roof. The authors chose a polyethylene cemented cup and a custom-made cemented stem with a 22-mm head, because they were not confident that a cementless cup could ensure the necessary structural stability. In this current case, a press fit modular S-ROM femoral prosthesis was selected because it has been used previously with satisfactory results. In our opinion, the modular nature of the component allowed for greater intraoperative variation and a more accurately achieved femoral offset, thus lowering the femoral fracture rate. No femoral fracture complications occurred during follow-up in the current case and the stem was in a good position with no signs of loosening. Another advantage was that the small-diameter straight stem was indicated for use in the small straight femoral canal in patients with DDH and also as an intramedullary fixation for osteotomies.

Surgical treatment becomes even more difficult in patients with dwarfism and DDH (Crowe type IV). The surgical treatment of DDH (Crowe type IV) is difficult in uncemented THA in adults due to the pathological anatomy of the hip. In most cases, femoral reduction can be difficult to achieve and implantation of the cup into the true acetabulum requires a femoral shortening osteotomy. Previous studies showed that THA combined with subtrochanteric femoral shortening osteotomy achieved good clinical results. Nevertheless, such arthroplasty is still technically demanding and has a higher complication rate than conventional primary THA. For example, a previous study found that the most common complications were the following: intraoperative femoral fracture, dislocation, delayed union or nonunion of the osteotomy site and neurovascular injury. Concomitant DDH in a patient with dwarfism is a rare occurrence. Therefore, the proper selection of the prosthesis and the use of a precise surgical technique are extremely important. Hence, the surgical application of THA, combined with subtrochanteric femoral shortening osteotomy, was selected in this current case. The follow-up of the present case showed good
Figure 1. Representative X-ray images of a 29-year-old female patient that was admitted to hospital due to shortening of the right lower limb, as compared with the contralateral side, which had been present for 18 years. She also complained of pain in the right hip that had been present for 2 months. The hip pain was aggravated by physical exertion, but relieved by rest. (a) A preoperative anteroposterior X-ray of the pelvis showed right acetabular dysplasia with high dislocation, thin cortical bone and a narrow medullary cavity. (b) A postoperative pelvic anteroposterior X-ray showed good alignment at the femoral shortening osteotomy and a good position for the prosthesis installation. (c) A postoperative pelvic anteroposterior X-ray at 6 months showed that the femoral shortening osteotomy had healed and the leg length had recovered well. After correction of the pelvic tilt, the cup reached its suboptimal position with an abduction angle of 58°.
clinical results for this patient that underwent THA with subtrochanteric femoral shortening osteotomy. There was also a study in which THA and subtrochanteric osteotomy were performed simultaneously to treat hip dysplasia in a patient with dwarfism. That case report describes the diagnosis of symptomatic Crowe type IV DDH in a patient with achondroplasia and highlights the difficulties of primary THA as a treatment option; and intraoperative radiographic imaging was advised to ensure proper prosthesis placement. However, the importance of prosthesis selection for clinical outcomes has been neglected in this current report, but it does provide a detailed description of the surgical techniques that were used and highlights the advantages of applying the S-ROM prosthesis, which adds to the limited clinical experience in this field.

Cases of patients with dwarfism and Crowe type IV DDH are rare. The follow-up of the current case showed good clinical results for a patient that underwent THA with subtrochanteric femoral shortening osteotomy using an S-ROM stem. At 6 months after the operation, the Harris score reached 77 points. The hip function was improved and the leg length was restored. However, long-term follow-up observation is needed. This current case provides insights for clinicians treating patients with such a rare diagnosis.

Declaration of conflicting interest
The authors declare that there are no conflicts of interest.

Funding
This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

ORCID iD
Huijie Li https://orcid.org/0000-0002-6726-9923

References
1. Dezateux C and Rosendahl K. Developmental dysplasia of the hip. Lancet 2007; 369: 1541–1552.
2. Aronsson DD, Goldberg MJ, Kling TF, et al. Developmental dysplasia of the hip. Pediatrics 1994; 94: 201–208.
3. Wang D, Li LL, Wang HY, et al. Long-term results of cementless total hip arthroplasty with subtrochanteric shortening osteotomy in Crowe type-IV developmental dysplasia. J Arthroplasty 2017; 32: 1211–1219.
4. Liu S, Zuo J, Li Z, et al. Study of three-dimensional morphology of the proximal femur in developmental adult dysplasia of the hip suggests that the on-shelf modular prosthesis may not be an ideal choice for patients with Crowe type IV hips. Int Orthop 2017; 41: 707–713.
5. Necas L, Hrubina M, Melisik M, et al. Cementless hip arthroplasty and transverse shortening femoral osteotomy with the S-ROM stem for Crowe type IV developmental dysplasia. Eur J Orthop Surg Traumatol 2019; 29: 1025–1033.
6. Zeng WN, Liu JL, Wang FY, et al. Total hip arthroplasty for patients with Crowe type IV developmental dysplasia of the hip: Ten years results. Int J Surg 2017; 42: 17–21.
7. De Fine M, Traina F, Palmonari M, et al. Total hip arthroplasty in dwarfism. A case report. Chir Organi Mov 2008; 92: 67–69.
8. Modi RM, Kheir MM, Tan TL, et al. Survivorship and complications of total hip arthroplasty in patients with dwarfism. Hip Int 2017; 27: 460–464.
9. Osagie L, Figgie M and Bostrom M. Custom total hip arthroplasty in skeletal dysplasia. Int Orthop 2012; 36: 527–531.
10. Memminger M, Banci L and Meoli A. Bilateral Total Hip Replacement in Dwarfism With a Custom Laser-Printed Trabecular Acetabular Shell. Orthopedics 2019; 42: e477–e479.
11. Wirtz DC, Birnbaum K, Siebert CH, et al. Bilateral total hip replacement in
pseudoachondroplasia. *Acta Orthop Belg* 2000; 66: 405–408.

12. Tamegai H, Otani T, Fujii H, et al. A modified S-ROM stem in primary total hip arthroplasty for developmental dysplasia of the hip. *J Arthroplasty* 2013; 28: 1741–1745.

13. Moura DL and Figueiredo A. High congenital hip dislocation in adults – arthroplasty and functional results. *Rev Bras Ortop* 2018; 53: 226–235.

14. Zagra L, Bianchi L, Mondini A, et al. Oblique femoral shortening osteotomy in total hip arthroplasty for high dislocation in patients with hip dysplasia. *Int Orthop* 2015; 39: 1797–1802.

15. Çatma MF, Ünlü S, Öztürk A, et al. Femoral shortening osteotomy in total hip arthroplasty for severe dysplasia: a comparison of two fixation techniques. *Int Orthop* 2016; 40: 2271–2276.

16. Li L, Yu M, Yang C, et al. Total hip arthroplasty (S-ROM stem) and subtrochanteric osteotomy for Crowe type IV developmental dysplasia of the hip. *Indian J Orthop* 2016; 50: 195–200.

17. Mu W, Yang D, Xu B, et al. Midterm outcome of cementless total hip arthroplasty in Crowe IV-Hartofilakidis Type III developmental dysplasia of the hip. *J Arthroplasty* 2016; 31: 668–675.

18. Malcolm TL, Phan DL and Schwarzkopf R. Concomitant achondroplasia and developmental dysplasia of the hip. *Arthroplast Today* 2015; 1: 111–115.