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Bilateral Total Hip Arthroplasty in a Patient With Achondroplasia: Challenges and Surgical Strategies

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ABSTRACT

This case report provides a history of a female patient with achondroplasia who underwent bilateral total hip arthroplasty (THA) using short femoral stem. On preoperative radiography, a severe bilateral hip osteoarthritis was noted, with deformity of the femoral metaphyses. After THA, the gait pattern improved significantly, the range of motion of both hips increased, and limb length discrepancy was corrected. There was also an improvement in Harris Hip Score value from 65 to 87. In patients with achondroplasia who develop end-stage hip osteoarthritis, there is a unique challenge for arthroplasty surgeons, including implant design, sizing, positioning, and soft-tissue balancing. We believe that the use of short femoral stems might represent an acceptable surgical strategy in the setting of complex changes in femoral anatomy.

Introduction

Achondroplasia is an autosomal dominant disorder that affects more than 250,000 individuals worldwide [1]. It is the most common form of short-stature skeletal dysplasia (dwarfism), with the most typical features being rhizomelia, midface hypoplasia, macrocephaly, and normal cognition [2].

Achondroplasia is caused by a gain-in-function mutation in the FGFR3 gene [3,4]. FGFR3 is a member of the receptor tyrosine kinase family. This protein is an essential regulator of chondrocyte proliferation within the growth plate [5]. A new, spontaneous mutation causes about 80% of cases of achondroplasia, and increasing paternal age is considered a significant factor influencing the high frequency of this condition [6].

Patients with achondroplasia may develop many medical complications during their lifespan, including obstructive and central sleep apnea, middle ear dysfunction, and cervicomedullary compression [2]. Patients with achondroplasia typically stand between 120 and 135 cm (4-4.5 ft) tall on average as adults [7]. Children and adolescents need to manage orthopaedic conditions such as leg bowing, lordosis, kyphosis of the spinal column, and spinal stenosis. All these factors lead to altered gait patterns and reduced walking distance [8].

Although achondroplasia is associated with numerous changes in the musculoskeletal system, primarily those affecting the spinal column and the knee joint, it is not considered a significant cause of premature hip osteoarthritis [9]. In patients with achondroplasia who develop end-stage hip osteoarthritis that requires total hip replacement, there is a unique challenge for arthroplasty surgeons, including implant design, sizing, positioning, and soft-tissue balancing [10]. Complex changes in pelvic and femoral anatomy significantly increase the risk of intraoperative complications. Long-term follow-up studies demonstrate a very high revision rate after total hip arthroplasty (THA) in patients with dwarfism [11-13].

This case report provides a history of a patient with achondroplasia who underwent bilateral THA using a metaphyseal fitting microplasty stem.

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Case history

A 31-year-old female patient with achondroplasia presented with complaints of pain in the right and left groin and walking difficulty. The first symptoms appeared in childhood, and the patient states that she had already undergone surgery on both hips 20 years ago in another hospital. Due to the lack of medical documentation from that period, it was impossible to establish the exact nature of the surgical treatment. Symptoms increased in severity over the last 3 years, interfering significantly with the performance of activities of daily living. The patient also had surgical correction of the equinus deformity of her right foot 15 years ago in our hospital.

It is worth mentioning that our patient previously consulted a spinal surgeon because of significant double-curved scoliosis of the thoracic and lumbar spine. A team of experienced spinal surgeons from our hospital decided not to pursue possible surgical treatment of scoliotic deformity until the completion of bilateral hip diagnostic evaluation and treatment.

The physical examination started with gait analysis. We noted a severely reduced walking distance, and without assistive devices, ambulation was impossible. We found decreased walking stability during the stance phase in particular. We assigned this to the weakness of hip abductors, further confirmed by a positive Trendelenburg sign when the patient performed a single-leg stance test. The range of motion of both hip joints was significantly limited, with slightly better findings on the left side. The active flexion range of the right and left hip was 40° and 70°, respectively. The extension was complete, and abduction and adduction ranges were limited to 15°. Internal and external rotation was possible in the first 10° of the arc of motion. Despite the patient's complaint of a leg length discrepancy, where the right leg was shorter than the left, we found no differences in limb length when measuring from the anterior superior iliac spine to the medial malleolus. Spinal scoliosis and left-sided pelvic tilt caused apparent limb shortening in this case. We used a stadiometer to record the standing height, which measured 118 cm. We inspected 2 consolidated surgical scars on the lateral aspect of the right and left thighs. We also obtained the Harris Hip Score (HHS), which indicated severe functional limitation (HHS = 65).

The radiological assessment included an anteroposterior view of the pelvis and a direct lateral (cross-table) view of both hips (Fig. 1). We noted severe bilateral hip osteoarthritis (Kellgren-Lawrence grade 4), characterized by the collapse of the femoral head and neck and a deformity of the femoral metaphyses. We also discovered significant bowing in femoral diaphyses in both the coronal and sagittal planes. Osteosynthetic material (cerclage wires) was present in the proximal femora as a remnant of previous hip surgeries.

After discussing treatment modalities, possible outcomes, and potential complications of surgical treatment with the patient, our team decided to perform a 2-stage bilateral THA. We admitted the patient to the hospital 1 day prior to the planned surgery and obtained informed consent. We started antibiotic and deep venous thrombosis prophylactic therapy preoperatively, adhering to our hospital's protocol, which included administering a loading dose of first-generation cephalosporin 1 hour prior to incision and administering low-molecular-weight heparin 1 day before surgery, which we continued for 5 weeks after surgery [14]. We performed the preoperative templating using TraumaCad (ver. 2.0.1) software. After correction for image magnification, we selected the implant size for the Trilogy IT acetabular shell and Fitmore femoral stem (Zimmer Biomet, Warsaw, Indiana, USA) with extended offset options. Through templating, we also demonstrated that we could restore the right amount of femoral offset by using a stem with a neck-shaft angle of 140°.

After thorough preoperative preparation, we operated on the patient under general anesthesia. We decided to perform total right hip arthroplasty through a posterolateral approach. After dissecting subcutaneous soft tissues, we incised the gluteal and fascia lata and

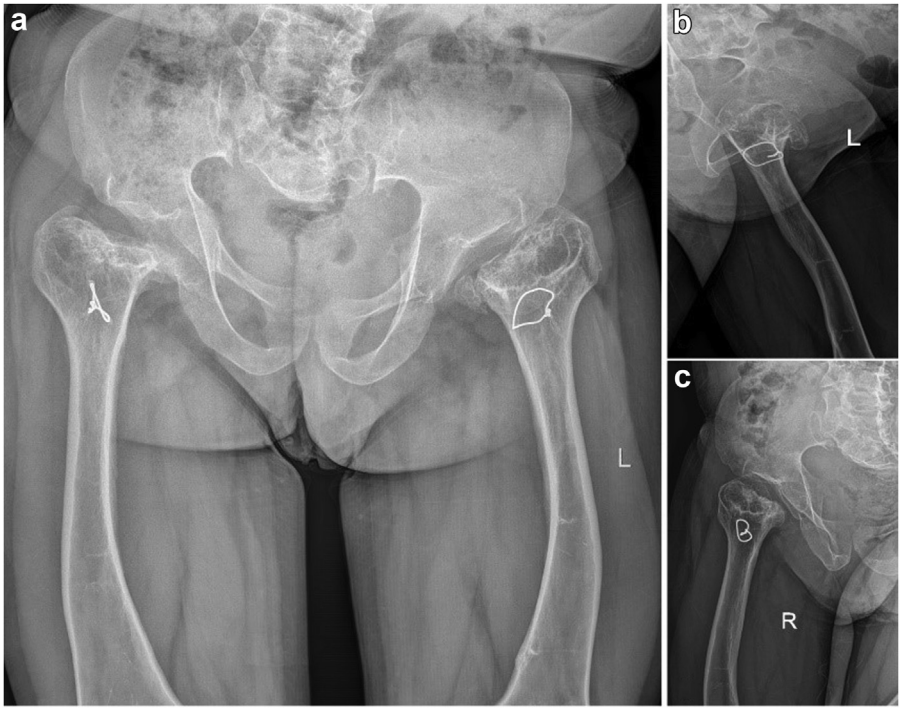

Figure 1. (a) Anteroposterior pelvis view of young woman with achondroplasia and hip degeneration (preoperative radiography from 2019). (b) Lateral X-ray of the left hip and (c) right hip.

split the gluteus maximus fibers. We identified, dissected, and tagged the short external rotators for later repair. We made a T-shaped incision in the posterior capsule of the hip joint and repeated the same tagging procedure. Due to the presence of fibrotic scar tissue, a significant release of capsular attachments was required to allow for hip luxation. We marked the osteotomy line based on the preoperative templates, cleaned up the acetabulum, and removed the fibrotic tissue and pulvinar.

We prepared the acetabulum using acetabular reamers, each one increasing in size by 1 mm. We implanted the uncemented acetabular shell and reinforced the primary fixation with 1 cancellous screw. We placed a polyethylene liner and continued with the preparation of the femoral canal. After completion of femoral rasping, we inserted a probe into the medullary canal to check for signs of possible fracture and detected a small crack in the posteromedial cortex of the proximal femur. Because of this, we decided to perform a prophylactic wire cerclage of the proximal femur before implanting the femoral stem. The hip was reduced and the stability of the prosthetic system was verified through a full range of motion. Although preoperative templating predicted the use of femoral stem with neck-shaft angle of 140°, it could not provide adequate stability of the left hip prosthesis when tested intraoperatively. Therefore, we opted to use a stem with a lower neck-shaft angle of 137°, which resulted in a greater horizontal femoral offset. After implanting the final components (femoral stem and ceramic head), we abundantly irrigated the hip joint, repaired the capsule and external rotators, and sutured the wound in a standard fashion.

The rehabilitation started on the first postoperative day following the special regime. The first 2 weeks allowed partial weight bearing, followed by a gradual transition to full weight bearing. The patient ambulated using assistive devices (a walker at first, then 2 underarm crutches). The physiotherapists started isokinetic exercises that targeted hip flexors and abductors to enhance muscle strength and stability of the prosthetic system. The inpatient rehabilitation facility successfully completed the rehabilitation process.

Three days after the surgery, the patient was discharged from the hospital. We continued the outpatient clinical assessment and scheduled control examinations every 2 weeks. The patient was satisfied with the surgical results, prompting plans to perform the total replacement of the left hip 3 months after the right hip surgery. We took a full-body-length X-ray with a radiographic ruler to more precisely assess the lower limb length discrepancy that resulted from the THA of the right hip (Fig. 2).

We performed the same preoperative preparation and operative procedure for the left hip. During the preparation of the acetabulum, we found the subchondral bone of poor quality, probably due to the long-term loss of physiologic load-bearing forces. Despite all precautionary measures, we noted a small degree of iatrogenic acetabular protrusion at the time of surgery. This finding prompted us to perform acetabuloplasty of the medial wall with cancellous bone allograft. Unlike the right hip arthroplasty, this time we used a femoral stem with a neck-shaft angle of 140°, according to the preoperative templating. One month after the left hip arthroplasty, we took a control pelvis X-ray (Fig. 3a).

The patient had no complaints regarding hip surgeries; the gait pattern significantly improved, the range of motion of both hips increased, and the limb length discrepancy was corrected. There was also an improvement in the HHS value (HHS = 87). The follow-up continued for the next 3 years, with periodic controls in 6-month intervals (Fig. 3b). There were no complications observed during follow-up. Finally, written informed consent was obtained from the patient prior to submission of this article.

Discussion

Achondroplasia is associated with a diverse range of abnormalities of the musculoskeletal anatomy, affecting both the axial and appendicular skeleton [15]. Some of the hip-specific changes include pelvic distortion, which is characterized by the squaring of the iliac wings and broadening of the pelvic cavity, resulting in a “champagne-glass” appearance on the pelvis X-ray. Other changes encompass a horizontal acetabulum, characterized by a decreased beta angle, overcoverage of the femoral head, proximal femoral metaphyseal flaring with a short and broad femoral neck, and an increased anterior bow of the femoral shaft [12,16]. It is stipulated that all the factors mentioned above, together with hyperlordosis of

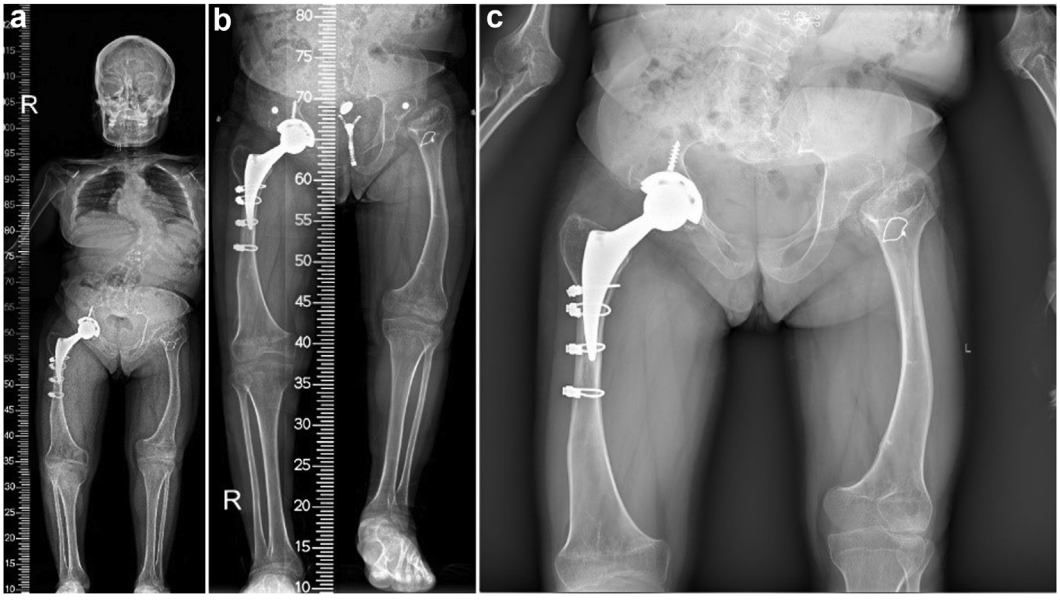

Figure 2. First postoperative control, 1 month after the surgery of right hip. (a) Anteroposterior X-ray of the whole skeleton, (b) anteroposterior X-ray of the pelvis and lower extremities, and (c) anteroposterior X-ray of the pelvis and upper legs.

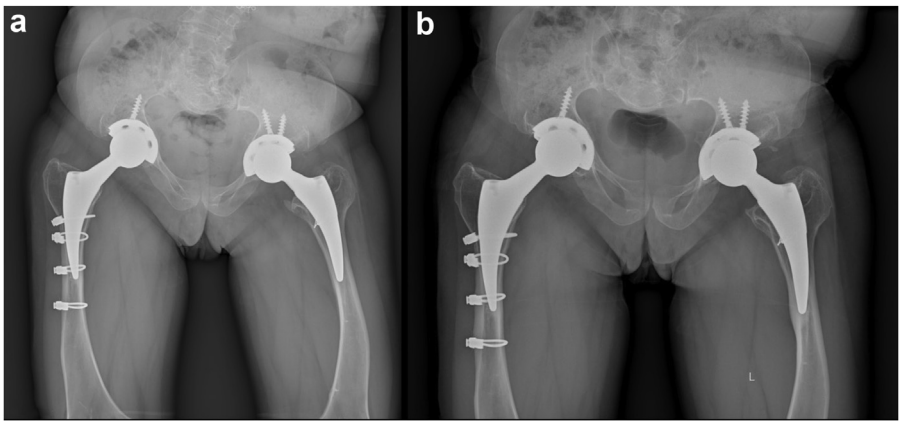

Figure 3. (a) Postoperative standing anteroposterior X-ray of the pelvis at the 3 months after the surgery of left hip, and (b) at 3 years postoperative visit after bilateral arthroplasty. All components were well seated, without evidence of loosening, fracture, or wear.

the lumbosacral spine, change in muscle tone resulting from spinal stenosis, generalized ligamentous laxity, and overall lower limb malalignment, can cause nonphysiologic load distribution and altered biomechanics of the hip joint [10]. This can hasten the development of the degenerative joint disease, further aggravating the clinical picture of patients with achondroplasia.

Previous studies concluded that THA represents a safe and viable treatment modality for patients with skeletal dysplasia and short stature [12,13]. However, it is worth mentioning that orthopaedic surgeons encounter many challenges when considering such a procedure. First, it is necessary to define strict criteria for selecting patients who will benefit from hip replacement surgery. Patients with achondroplasia often experience a crippled gait that has a multifactorial origin [15]. Therefore, before embarking on the hip replacement pathway, it is advisable to discuss with the patient the expected degree of functional improvement. Due to the complex changes in pelvic and femoral anatomy, it is of utmost importance to perform thorough preoperative planning, which includes an X-ray of the pelvis and thigh from multiple views and a computed tomography scan [17]. The small size of the acetabulum, deformity of the femoral metaphysis, and bowing of the femoral shaft often preclude the use of standard, off-the-shelf implants. Small-diameter femoral heads significantly increase the risk of joint instability and dislocation due to reduced head-neck ratio and jump distance [18]. This underlines the necessity of precise acetabular cup placement, respecting Lewinnek safe zones with adequate anteversion and inclination angles [19]. Additionally, the degree of spinopelvic mobility, which is frequently affected in individuals with achondroplasia, influences and changes the position of the acetabular component [20].

The preparation of the femoral canal and the choice of femoral stem represent one of the most daunting challenges when performing total hip replacement in patients with achondroplasia. Because of varying degrees of proximal femur deformity, bowing of the femoral shaft, and narrowing of the femoral canal, it is challenging to achieve adequate position, size, and initial stability using standard implants [21]. Several studies demonstrated an increased risk of intraoperative iatrogenic periprosthetic fractures, which lead to prolonged surgery time, delayed postoperative recovery, and higher incidence of mid-term and long-term complications, such as nonunion, infection, and implant loosening [22,23]. The literature has devised and described different surgical strategies for femoral preparation in the setting of distorted bone morphology. Gautam et al. reported a case of bilateral total hip replacement in a patient with achondroplasia, using the modular S-ROM (DePuy Orthopaedics Inc., Warsaw, Indiana, USA) cementless stem, which allows for independent adjustment of anteversion, height, and offset [11]. Malcolm et al. performed a subtrochanteric femoral shortening osteotomy on a patient with concomitant achondroplasia and developmental dysplasia of the hip, using the same modular implant [12]. Femoral osteotomy is also advised in the setting of increased femoral anteversion and anterior bowing to correct angular and rotational deformities [24]. Other authors described the use of custom-made femoral stems prepared in computer-assisted manufacturing processes [25].

Current controversies and future considerations

Studies that analyzed the complication rates and long-term survival of THA in patients with dwarfism and skeletal dysplasia showed conflicting results. Chiavetta et al. conducted a comparison between the outcomes of 62 total hip replacements, which were performed on 32 patients with dwarfism, and the control group, which included all other THA patients [13]. They found a significantly higher rate of revision procedure in the dwarfism group (29%), with the most common reasons for revision including aseptic loosening of the femoral component. Moore et al. observed a greater infection rate at 90 days following the THA and increased wear-related complications at 5-year intervals [10]. They proposed an intriguing explanation for the occurrence of accelerated polyethylene wear. They postulate that the small size of prosthesis components, combined with abnormally distributed joint reaction forces, leads to a decreased contact area with regions of high-stress concentration. Other studies conducted by Modi et al. and Osagie et al. did not see a statistically significant difference in survival rates of THA in patients with short stature compared to the general population [19,25]. Oba et al., in their study, analyzed 68 THAs performed in 49 patients with dwarfism with a follow-up duration of 10 years [26]. They demonstrated promising results, with long-term survival of 94.1%. Researchers attributed the higher failure rate in earlier studies to the more frequent use of cemented femoral stems [27]. Finally, it is crucial to consider that patients with short stature who undergo THA tend to be younger than the average THA patient population [28]. This fact underscores the importance of optimizing surgical technique to improve the longevity of the prosthesis and decrease the risk and number of revision procedures.

To the best of our knowledge, this is the first case report that provides a history of the patient with achondroplasia who underwent THA using a metaphyseal fitting microplasty stem. This cementless stem belongs to the group of neck-resecting short femoral stems [29]. Some authors refer to this stem as an

"intermediate-type stem", as it is a shortened version of standard femoral stems [30]. The basic concept of short stems is that they can provide more physiologic transfer of load to the metaphyseal region, thus slowing the development of proximal bone atrophy due to stress shielding [31]. There are additional advantages of microplasty stems. First, these stems reduce bone removal during femoral preparation, thereby increasing the bone stock available for future revision procedures. Second, these stems represent a family of short femoral stems with different neck-shaft angle values that enable the surgeon to modify the femoral offset and achieve proper soft-tissue tension. Finally, many patients with achondroplasia suffer from knee osteoarthritis and significant knee instability in the coronal plane, which necessitates constrained knee prosthetic designs with long femoral stems [15]. In this context, a previous THA with a short stem will facilitate the implantation of such knee prostheses without any technical challenges.

Summary

THA in individuals with achondroplasia represents a viable, albeit very demanding, procedure from the technical point of view. Further investigations are crucial to precisely delineate the pathophysiological background of hip osteoarthritis in achondroplasia. We believe that the use of short femoral stems might represent an acceptable surgical strategy in the setting of complex changes in femoral anatomy.

Key Points

- A thorough diagnostic workup, including clinical assessment and radiographic series, should be performed prior to surgical treatment, as well as detailed preoperative templating, to delineate the specifics of surgical strategy.
- Special care should be taken when performing a surgery given the increased risk of intraoperative complications due to distorted hip anatomy.
- It is necessary to establish a multidisciplinary approach, including spinal surgeons and physical and occupational therapists, to maximize the postoperative outcomes and enable the patient to attain a high degree of functional independence.

Conflicts of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

For full disclosure statements refer to https://doi.org/10.1016/j.artd.2025.101654.

Informed patient consent

The author(s) confirm that written informed consent has been obtained from the involved patient(s) or if appropriate from the parent, guardian, power of attorney of the involved patient(s); and, they have given approval for this information to be published in this case report (series).

CRediT authorship contribution statement

Danilo Jeremic: Validation, Investigation. **Jelena Nesovic Ostojic:** Conceptualization. **Branislav Krivokapic:** Validation, Investigation. **Zoran Bascarevic:** Supervision. **Nikola Zarkovic:** Writing – original draft. **Nemanja Slavkovic:** Writing – review & editing.

Data availability statement

All datasets presented in this study are included in the article.

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