

## Case report

## Bilateral simultaneous total hip replacement in Achondroplasia

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## ABSTRACT

We present a case of bilateral simultaneous total hip replacement in a very short statured (height 112 cm) 45 years old male patient with Achondroplasia and disabling osteoarthritis of both the hips. We describe the difficulties encountered and provide a methodical approach for such cases.

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## 1. Introduction

Achondroplasia is a genetic condition that results in an abnormal short stature. Early onset of hip osteoarthritis is one of the main clinical problems in Achondroplasia. Total hip replacement remains the mainstay of treatment to restore the mobility and quality of life of such patients with disabling arthritis of hip, however there is a paucity of literature<sup>1,2</sup> explaining the surgical treatment plan for these patients. These patients need special implants to compensate for the distorted geometry of the joint and the bone size. We performed bilateral simultaneous total hip replacement in an adult male with achondroplasia using special implants with modular stem and the patient is doing well at the latest follow up.

## 2. Case report

A 45 years old male presented with severe pain in both the hips for the past one and a half year. The pain was gradually progressive and the patient could barely walk a distance of half a kilometer. His daily activities were severely hampered. Patient's was 112 cm in height with his lower limb measuring 54 cm (Fig. 1) and weighed 58 kg.

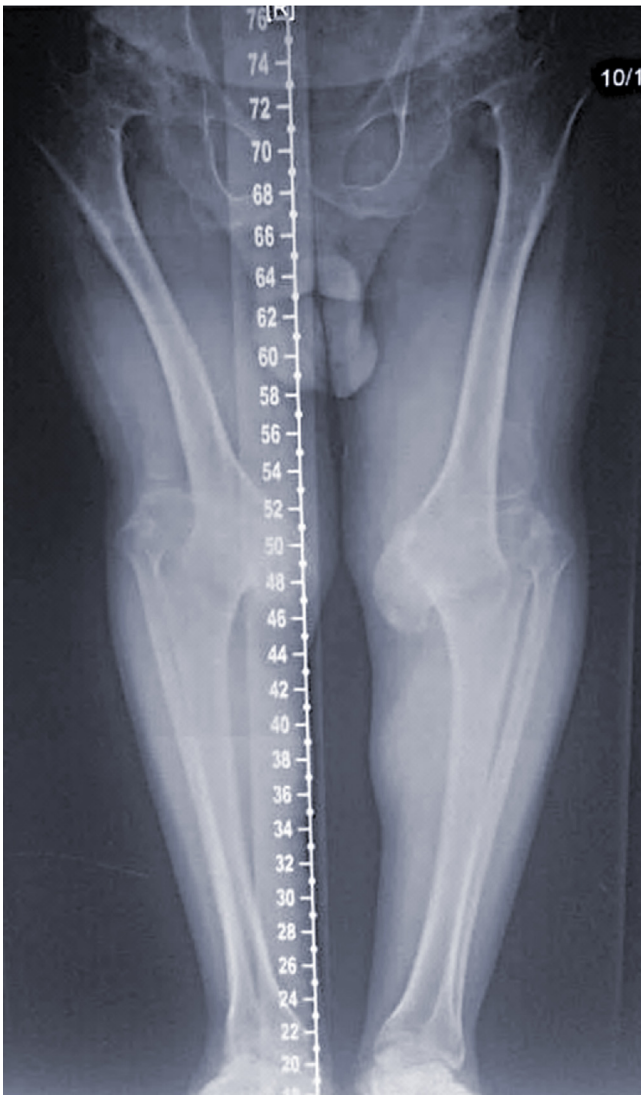
He was a diagnosed case of Achondroplasia having characteristic phenotypical features and a disproportionate short stature. However no neurological involvement or mental retardation was noted. On examination, movements in both the hips joints were

restricted and painful especially abduction and rotation. On the right side there was a fixed flexion deformity of 20° with jog of movements and adduction deformity of 30°. While the left hip had a fixed flexion deformity of 15° with jog of movements and an adduction deformity of 20° and a true shortening of 2 cm. On radiographic evaluation (Figs. 2 and 3) the acetabulum was shallow and the femur was distorted.

In view of his disability due to advanced arthritis of hip, he was planned for bilateral Total Hip Replacement. He was counseled accordingly and an informed consent was taken. Surgery of the right hip was performed in lateral position through the posterior approach followed by the left hip. Extensive soft tissue release was carried out because of the severe deformity. The hip was dislocated cautiously followed by acetabular reaming. A porous metal cup (Regenerex® Biomet) was used along with a highly crossed linked E- Polyethylene liner. In view of the distorted femoral anatomy a special stem (S-ROM, DePuy Orthopaedics Inc., Warsaw, Ind) was required (Fig. 4). A bilateral adductor tenotomy was carried out to release the tight adductors and ease the reduction. The total blood loss was 600 ml and he was transfused with one unit of packed RBCs. Postoperatively the patient was monitored in High Dependency Unit for 24 h and prophylactic antibiotics were started. Low Molecular Weight Heparin was instituted for Deep Vein Thrombosis prophylaxis. Early in bed mobilization was started on 1st post operative day. Drains were removed on 2nd post operative day and analgesics were administered round the clock for the first three days. Full weight bearing mobilization with a walker was done on the 5th post operative day. Hip strengthening and gait training exercises were given in a phased manner. He was discharged after removing the sutures on 14th post operative day.

Postoperatively after 2 years of follow up there was no limb length discrepancy and patient could perform his activities of daily

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**Fig. 1.** X-ray scannogram of both the lower limbs showing the arthritic hips and deformed knees as well..

living without assistance and analgesics. Radiographic analysis showed no signs of loosening or implant malposition (**Fig. 5**)

### 3. Discussion

Achondroplasia is the most common form of dwarfism in humans.<sup>3</sup> This condition is characterized by a height that is less than expected for that age. The maximum shortening is seen in the humerus and forearm bones followed by the femur. These patients develop deformities of the skeletal system and joint pathologies like dysplastic hip and secondary osteoarthritis. Total Hip Arthroplasty is the treatment of choice in those individuals who are crippled and have restricted activities of daily life.

Total hip replacement in these patients is a demanding procedure because of the risk of high rate of complications such as dislocation, implant malpositioning, periprosthetic fractures and early aseptic loosening.<sup>2</sup> There may be one or more comorbidities that may increase the risk of the surgery. As the size of the bone is reduced and the bone morphology is deformed, an exact preoperative evaluation with geometric analysis including CT scan of the proximal femur and acetabulum is necessary.



**Fig. 2.** AP and lateral radiographs of the right hip with thigh showing the distorted femur.

Posterior approach was employed in the surgery in order to gain better exposure of the femoral head and the acetabulum. The implants used were of special characteristics to suit the anatomy as well as the quality of the bone. The Regenerex cup combines a clinically proven material with technology that provides optimum pore structure for bone fixation, initial stability, strength and flexibility. The coating is biocompatible that maintains a strong construct without increasing the stiffness of the implant.<sup>4</sup> This helps in early mobilization of the patient.

The most daunting challenge was of femoral preparation and its component as the femoral anatomy was distorted with narrow medullary canal and excessive anteversion. The stem with modular component was the best option to allow easier control of anteversion.

We used stem-sleeve modular implant in the form of S-ROM modular hip system as it was the most familiar modular implant to



**Fig. 3.** AP and lateral radiographs of the left hip with thigh showing the distorted femur.

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