Limb shortening osteotomy in a patient with achondroplasia and leg length difference after total hip arthroplasty

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What to Learn from this Article?

Sometimes atypical conditions present to us like limb length difference in achondroplasia. Such situations need to be individualized and customized decisions need to be taken keeping in mind patients preference and demands

Abstract

Introduction: Achondroplasia is the most common reason for disproportionate short stature. Normally, orthopedic limb lengthening procedures must be discussed in the course of this genetic disorder and have been successful in numerous achondroplastic patients in the past. In some cases, the disease may lead to leg length differences with need for surgical correction.

Case Report: We report a case of achondroplastic dysplastic coxarthrosis with symptomatic leg length difference after bilateral total hip arthroplasty in a 52-year-old female patient, in which a distal femoral shortening osteotomy was successfully performed.

Conclusion: Femoral shortening osteotomy is very uncommon in patients with achondroplasia. We conclude, however, that in rare cases it can be indicated and provide the advantage of shorter operation time, less perioperative complications and faster recovery compared to leg lengthening procedures.

Keywords: Achondroplasia, dysplastic coxarthrosis, limb shortening, distal femur osteotomy.

Author's Photo Gallery





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Introduction

Achondroplasia is a skeletal dysplasia and the most common form of short limb dwarfism in humans with a prevalence between one in 10 000 and one in 30 000 newborns [1]. In over 95 percent of cases, it is caused by a point mutation in the gene for fibroblast growth factor receptor 3, leading to multiple effects on organ systems with various clinical manifestations. The most striking one involves cartilage function with a detrimental effect on longitudinal bone growth [2]. While orthopedic limb lengthening usually is considered in achondroplastic dwarfs and has been successfully performed to a varying extent in the past [3], indication for shortening procedures of the extremities in these patients is very rare.

Here, we present the case of an achondroplastic patient suffering from symptomatic leg length discrepancy after bilateral total hip replacement surgery in the course of dysplastic coxarthrosis. In contrast to common belief, femoral shortening osteotomy was performed in order to accomplish leg length equalization as well as symptom relief.

Case Report

A 52-year-old female achondroplasia patient (body height 130 centimeters) was referred for orthopaedic treatment with a symptomatic leg length difference after bilateral total hip arthroplasty. Her medical history contained psoriasis and atopic dermatitis, as well as several orthopedic operations in the course of her underlying disease. During childhood and adolescence, she had undergone several surgical procedures due to recurrent bilateral dislocation of the patella including

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bilateral varus osteotomy of the distal femur. Apart from orthopedic issues, no other complications from achondroplasia were apparent.

Dysplastic cox arthrosis affecting both hip joints had been treated with bilateral hip replacement surgery. The first prosthesis was implanted on the right hip without any perioperative complications eight years prior to presentation at our clinic. Hip replacement surgery on the left side was carried out six years after total hip arthroplasty on the right side. A postoperative overlength of five centimeters of the left femur resulted whereas prior to the procedure the leg length had been balanced out.

At our clinic, the patient presented with a limping gait pattern and complained about constant pain in the left hip joint as well as intense and motion dependent lower back pain. Activities of daily living were possible only with extreme difficulty. A heel elevation of three centimeters had been tried temporarily on the right side, but did not prove to be sufficient for the patient. Both hip implants were found to be stable with no radiological signs of loosening. A postoperative overlength of the left femur (70 centimeters versus 65 centimeters) was apparent while the tibiae were of approximately equal length with both leg axes in slight (normo)-varus positions.

The patient had become well adapted to her short stature



Fig 3(a),(b):
Postoperative
result after distal
femur shortening
osteotomy. A
limited contact
(LC) 4.5/5.0 mm Tplate was used for
o s t e o t o m y
refixation.

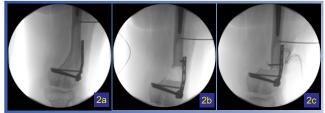


Fig 2: Intraoperative c-arm images: measuring of plate position (a) osteotomy (b) and plate fixation (c)

during lifetime and did not feel any strain for increasing her body height. Now she was eager to cope with everyday life again - as she had been before her second total hip arthroplasty. The possible therapeutic options were evaluated and discussed with the patient. We advised for right leg lengthening. The patient strongly opposed and refused the operation, mentioning friends with short stature history who underwent lengthening procedures. She did not want to "suffer the same complications" and lengthening treatment schedule.

Informed consent for a distal femoral shortening osteotomy of the left lower limb was obtained. Shortening of the distal femur by five cm was performed using a Low Compression Plate with angle stability (LCP DF, Synthes, Solothurn, Switzerland) for osteotomy fixation. There were no perioperative complications. In the postoperative course, the patient was instructed in 15 kilograms weight bearing and was mobilized with full range of motion. The patient was discharged in good medical condition with clean wound conditions four days after surgery. At the follow-up six weeks later, the patient presented with a balanced leg length. Three month after surgery the osteotomy was healed and the patient was allowed full weight bearing. Walking without crutches was reached four months after the operation.

One year after surgery, the control radiograph showed a fully consolidated osteotomy. In the gait analysis, the gait pattern was normal showing no signs of instability and the patient managed walking on uneven ground without difficulty. The straight leg raise test was performed without any signs of a muscular deficit. During the chair rising test, neither assistance nor support by the upper extremity was needed. Active bending of the knee joint was performed with good muscular strength. The patient was entirely satisfied with the postoperative result.

Discussion

Lower limb shortening procedures are rare in achondroplasia. We present a patient with limited quality of life due to symptomatic leg length difference after several orthopaedic operations, including bilateral total hip replacement. To equalize the leg length difference we



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ig 4: Radiograph one yea after surgery showing a complete union of the astentomy

conducted a femoral shortening osteotomy.

Generally, the operative leg length equalization includes techniques such as shortening, lengthening or growth plate closure (epiphysiodesis) during growth procedures. In achondroplasia most common are limb-lengthening procedures, thus allowing patients to achieve a body height within the normal range of their population. Several techniques to obtain this objective have been developed, i.e. intramedullary leg lengthening [4], external fixator extraction distractors [5], or combinations of those [6]. However, in achondroplastic patients limb lengthening is usually performed during childhood or adolescence [2], and can be associated with significant time exposure and morbidity [7]. Bone lengthening of 20 to 30 percent is possible, and up to 50 percent and more is reported [3]. At any rate, multiple surgical procedures and a comparably long period of treatment are required. This also may possibly involve complications ranging from pain and pin tract or deep infections to postoperative contractures and joint stiffness [7], all of which can increase disability and significantly delay rehabilitation.

In the case presented, the 52-year-old patient was well adapted to her life situation and body height. She refused to undergo right leg lengthening. Even if right femoral lengthening of five centimeters were technically possible and conducted successfully, the resulting total body height of 135 centimeters versus 130 centimeters would presumably not have had a significant impact on the patient's ability to cope with everyday tasks. The patient

had been searching the internet before consultation, which made her aware of the potential complications of leg lengthening compared to shortening. She therefore preferred a distal shortening osteotomy of the left femur to limb lengthening procedures with the advantage of faster recovery and reduction of postoperative pain and complications.

Conclusion

Femoral shortening osteotomy has proven successful in the treatment of a number of developmental dyslasias [8,9], but is a very uncommon procedure in patients with achondroplasia. However, in our case presented here, left femoral shortening was indicated and successfully performed. We conclude that, if rare, there might exist similar conditions in other patients who could benefit from such surgery with the advantage of shorter operation time in a single procedure, less perioperative complications and faster recovery and rehabilitation compared to limb lengthening.

Clinical Message

Finding the appropriate operation for achondroplastic patients suffering from unequal limb length may be challenging. Although rare, limb-shortening procedures may be indicated in certain patients. A detailed and critical evaluation is essential

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