

**TOTAL HIP ARTHROPLASTY FOR A PATIENT WITH
ANGEL-SHAPED PHALANGO-EPIPHYSEAL DYSPLASIA
(ASPED)
A Case Report.**

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ABSTRACT

Angel-shaped phalango-epiphyseal dysplasia is characterized by the angel shape of the middle phalanx and severe coxarthrosis in adult life. This osteochondrodysplasia shows a further variety of heterogeneous multiple epiphyseal dysplasias. It also shows a late and dysplastic development of the femoral head that leads to osteoarthrotic changes with severe hip pain and gait disturbance. In this report, we show a 35-year-old female with Angel-shaped phalango-epiphyseal dysplasia that was treated by bilateral total hip arthroplasty. She has suffered from her coxalgia since she was 27 and since the age of 30 has been able to walk with the aid of crutches. The radiographs of her bilateral hip showed severe osteoarthrotic changes with a progressive disappearance of the joint space. Total hip arthroplasties were performed to treat the osteoarthrosis on the bilateral hip joint due to Angel-shaped phalango-epiphyseal dysplasia. One year after surgery, she is pain-free, and able to walk with a cane. Based on this case, we propose that total hip arthroplasty should be considered one of the treatments for the coxopathy in patients with Angel-shaped phalango-epiphyseal dysplasia.

Key Words: total hip arthroplasty, angel-shaped phalango-epiphyseal dysplasia, osteochondrodysplasia

INTRODUCTION

Osteochondrodysplasia is a heterogeneous disease of constitutional disorders caused by primary abnormalities of cartilage and bone development. "Angel-shaped phalango-epiphyseal dysplasia (ASPED)" (OMIM: 105835) is one type of osteochondrodysplasia characterized by angel-shaped middle phalanges and generalized epiphyseal dysplasia that disproportionately affects the middle phalanx and distal hip joint^{1,2)}. Only 11 cases have been reported previously. ASPED was first described as "hereditary peripheral dysostosis"¹⁾. Giedion renamed it ASPED in 1993, because former reports had overlooked the unique finger appearance of ASPED²⁾. The clinical phenotypes are a swan neck deformity of the fingers, abnormal dentation, and short stature, and

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short limbs. The radiological findings are consistent with the characteristic “angel-shaped phalanx (ASP)” of the middle phalanges, nonspecific brachydactyly, generalized mild hyperostosis of long bones, and epiphyseal changes, mainly in the hips^{3,4}). The most important radiological feature of the hip joint is the late and dysplastic development of both femoral heads, leading to severe osteoarthritis in the early thirties. Patients with debilitating hip pain due to severe coxopathy may require surgical intervention, but we have never seen case reports about such hip surgery for ASPED patients. In this report, we described the satisfactory result of total hip arthroplasty (THA) on one patient with severe arthrotic hip joints due to ASPED.

A CASE REPORT

The patient is a 35-year-old female. Her father is also seems to have ASPED. She first felt pain in her right hip when she was pregnant at the age of 27. After that, the pain gradually increased in both hip joints. When she first visited our hospital, she could not even walk a few steps. It was impossible for her to maintain a sitting position due to her restricted range of hip motion. Her respective height and weight were 139 cm and 41.0 kg. She displayed a characteristic features of short stature and short limbs with small hands, feet and fingers. Her skin was shrunken with many skin folds. Radiographs of her hands showed a distinctively marked shortening of the phalangeal bones indicative of so-called Angel-Shaped Phalango-Epiphyseal Dysplasia (ASPED) (Figure 1). She showed no mental retardation.

The ranges of motion in the left and right hip at the time of admission were, respectively 20° and 20° of flexion, and -5° and 12° of abduction. The Harris hip score was 5 points on both sides. The radiographs of both hip joints showed severe osteoarthritis (Figure 2). The joint spaces were not preserved at all, and femoral head deformity was noted.

To improve her daily quality of life, especially during evacuation, and to relieve the pain, we decided to perform THA on both hip joints. For young patients with coxopathy, joint preserving operations such as osteotomy are desirable to prevent or delay the need for THA. However, this patient was in the bilateral terminal stage of osteoarthritis and had seriously restricted range of motion. We considered THA as the most effective treatment to relieve her pain and improve her movement range. The first THA was carried out on the right hip joint (Figure 3a). The femoral head had protruded into the pelvis cavity and was fused to the acetabulum, causing fibrous ankylosis. It seemed difficult to dislocate the femoral head because of



Figure 1: Radiographic features of ASPED patient's hands. Characteristic “angel-shaped phalanx (ASP)” of the middle phalanges, nonspecific brachydactyly are shown.



Figure 2: Preoperative anteroposterior radiographs of both hips, showing marked femoral head deformity, severe osteoarthritis, and joint-space narrowing.

a contracture of the surrounding soft tissues. For the reasons mentioned above as well as our wish to preserve as much of her bone stock as possible, we planned to leave the femoral head in situ for THA. First, the femoral neck was cut in situ, and the femoral head was reamed from the side of the femoral neck. We used cement to achieve a firm bond between host bone and acetabular cup, and drilled through the joint interspace to make an anchor hole. Then, an all-polyethylene acetabular cup (Stryker, USA) was cemented into the reamed-out femoral head since the head itself was thought to be too stiff to achieve a firm bond. A femoral component (OMNIFIT C, Stryker, USA) was also implanted with cement. Four months later, the left THA was operated on in the same way. Surgical specimens were taken when drilling through the joint interspace. Macroscopic findings from those samples showed but fibrous tissue covering the joint surface instead of a cartilage layer. During the postoperative follow-up of the left THA, the progress of the bone union between the acetabulum and the preserved femoral head was radiologically observed (Figure 3b). One year and five months after the right THA and one



Figure 3a: Anteroposterior radiographs of the right hip just after THA, showing reconstruction with all-polyethylene acetabular component inserted onto the remaining femoral head.



Figure 3b: Anteroposterior radiographs of the right hip taken three months later, showing progress in the bone union between the acetabulum and the preserved femoral head.



Figure 4: Anteroposterior radiographs of both hips taken one year and five months after the right THA and one year after the left THA, showing completion of the bone union in the joint interspaces.

year after the left, her hip joints were pain-free, with a dramatic improvement in her daily activity. She is now able to walk for 1000 m with only a cane. The radiographs at that time reveal the bone union between the acetabulum and the remnant of the femoral head (Figure 4). The Harris hip score has improved up to 65 points on the both sides. Respective right and left hip flexion has improved to 60° and 60°, abducted to 5° and 2°.

DISCUSSION

The most significant clinical problem in patients with ASPED may be severe coxalgia due to osteoarthritis of the hip. Giedion report that all ASPED patients over thirty would developed disabling osteoarthritis of the hip, and should therefore undergo surgical treatment²⁾. For young patients with osteoarthritis of the hip, femoral osteotomy is procedures are the treatment of choice to prevent or delay the need for THA. However if these performed on a patient with a restricted range of motion, a deterioration in hip motion and function often follow. Moreover, in cases of severe bilateral arthrosis of the hip joint, poor results have been reported after such procedures⁵⁾. For the reasons mentioned above, these joint-preserving operations are not indicated for such a patient. Some osteoarthritis of the hip joint in ASPED is severe, bilateral and so serious that malfunction is caused by restricted joint motion. Rekeras *et al.* reported that patients with fused hips who were treated with THA experienced relief of pain, improved mobility, and decreased dependence on walking aids⁶⁾. Therefore THA should be consider as the first choice among surgical treatments to improve function. However, treatment with THA poses several problems for ASPED patients. First, the disabling osteoarthritis of the hip usually occurs at a younger age, so patients will have to undergo further surgery later in life. Second, patients with osteochondrodysplasia have a femoral deformity that makes it difficult to implant the prosthesis into the femoral medullary cavity.

The result of THA in patients with chondrodysplasia has been reported by Hunter⁷⁾ who reports that 28-hip replacements were performed on such patients with a mean satisfaction score of 6.14 points (out of a possible⁷⁾). That report concluded that hip replacement provides the one of the highest levels of satisfaction for patients with osteoarthritis of the hip joint due to chondrodysplasia. Huo *et al.* reported the clinical result of a customized femoral component in six patients with chondrodysplasia who had severe dysplasia of the hip⁸⁾. They mentioned that all those patients had excellent results after surgery.

After a review of the literature, we could not find any previous reports about THA in patients with ASPED. Here we report the first case of THA performed to treat osteoarthritis of the hip joints due to ASPED. Our patient showed fibrous ankylosis of the hip on the radiograph and experienced a restricted range of motion. After making anchor holes through the joint interspace, we cemented an acetabular cup into the left femoral head. Eventually the bone union between the acetabulum and the remnant of the femoral head was complete. After a bilateral THA, the patient was relieved of pain and satisfied with the improvement in activities of the abilities of daily living. Thus, we concluded that THA should be one of the options considered for the treatment of osteoarthritis of the hip joint in patients with ASPED.

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