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Stemless total shoulder arthroplasty for multiple epiphyseal dysplasia in a 52-year-old patient: a case report

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Multiple epiphyseal dysplasia (MED) is a rare genetic disorder with an incidence of 1 in 10,000 people.[4] In MED, the epiphyseal nucleus is the primary abnormality, whereas the vertebral bodies are nearly normal. Early-onset osteoarthritis has been reported in patients with MED with conservative or surgical treatment such as osteotomy in childhood and total joint replacement in adulthood.[16] Arthropathy secondary to MED is most common in load-bearing joints, and most reports of arthroplasty are of total hip arthroplasty.[12,13,19] Shoulder arthritis occurs in one-third of patients with MED and is mostly bilateral.[7]

Sewell et al reported that shoulder arthroplasty was effective at relieving pain, optimizing movement, and improving function for patients with skeletal dysplasia; however, compared with the general population, there was a higher complication rate and worse function.[17]

The upper arms of patients with MED are short, and stem insertion is expected to be difficult in total shoulder arthroplasty (TSA); therefore, we believe that stemless implants allow easier insertion of implants in patients with short upper arms in MED.

Stemless implants in TSA were first approved in Europe in 2004 and became available in Japan in 2018. There are many reports of good short-term postoperative outcomes for stemless implants.[6,18] The advantages of stemless implants include bone preservation,[1] shorter operative time, less intraoperative blood loss,[9] less stress shielding,[15] and easier implant removal during revision surgery.[6] There have been no reports on the outcomes of stemless shoulder arthroplasty in patients with MED. In this study, we report the results of a stemless arthroplasty for arthropathy secondary to MED.

Case Report

Patient background

The patient was a 52-year-old woman whose chief complaint was bilateral shoulder pain and limited range of motion. Her medical history included hypertension, diabetes, and dyslipidemia. She was diagnosed with MED in childhood. Around the age of 30 years, the patient developed polyarticular pain, which worsened especially in both shoulders by the age of 40 years. Her height was 128 cm, and shortening of the extremities was observed. This case was of a severe Fairbank type of MED, and secondary osteoarthritis of the shoulder due to MED was a hatchet-head type. The patient was able to walk with a cane at home and used a wheelchair outdoors. She had a surgical history of bilateral total hip arthroplasty. The left hip was replaced at the age of 36 years and the right hip at the age of 38 years. Examination of the right/left preoperative shoulder range of motion at the age of 52 years revealed that the forward active elevation was 100°/80°, the active external rotation with the elbow at the side was 10°/−30°, and the internal rotation to the posterior was the buttock/buttock (Fig. 1). The preoperative visual analog scale scores were 8 and 9 for the right and left shoulders, respectively. The Constant Score was 22/10 points. Preoperative radiography of both shoulders showed loss of articular cleft, deformity of the humeral head, and formation of bone spurs, and the humerus was shortened, with proximally varus deformity (Fig. 2, *A-D*). To evaluate humeral shortening, the entire length of

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Figure 1 Preoperative range of motion of forward elevation.

the humerus was radiographed with a scale preoperatively and compared with the average length for Japanese people. The length of her humerus (right/left) was 205 mm/206 mm (Fig. 2, *E* and *F*), which was shorter than the average length for women in Japan. The preoperative bone marrow density of the lumbar spine from L1 to L4 was 1.037 g/cm^2, which was not particularly low. Magnetic resonance imaging showed no cuff tears or loss of cartilage in the humeral head or glenoid (Fig. 3). The humerus was shortened, deformed, and adducted proximally; therefore, we thought that a conventional implant with a stem would be difficult to insert. We chose a stemless implant because of the high risk of postoperative fractures around the stem with short stem implants. In this case, the point of varus deformity of the humerus was approximately 60 mm from the osteotomy position, and since the distal end of the stem is very close to the position of varus deformity in a typical mini-stem, we determined that intraoperative rasping and distal stress concentration after stem placement were risk factors for fracture. Stemless implants were considered a good choice in terms of bone stock preservation, as the patient was relatively young. When she was 52 years old, TSA was performed for the left shoulder, which was the most symptomatic area. After 1 year of TSA on the left shoulder, TSA was performed on the right shoulder.

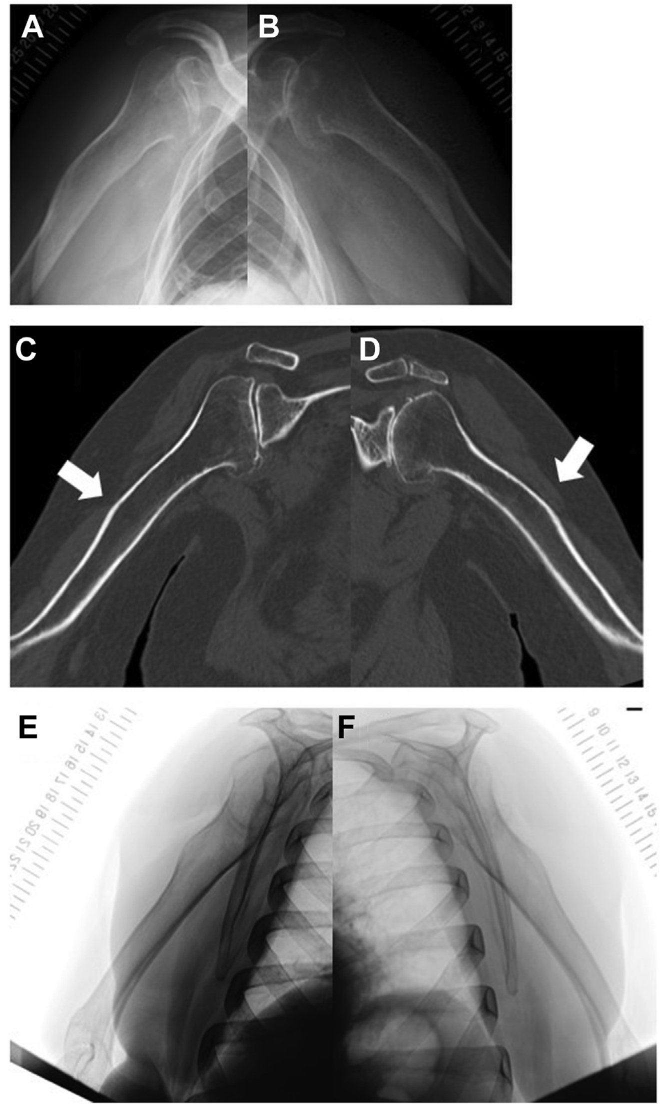

Figure 2 Preoperative plain radiography and computed tomography images. (**A**) AP view of the right shoulder. (**B**) Anterior posterior view of the left shoulder. (**C**) Coronal view of the right humerus. (**D**) Coronal view of the left humerus. *Arrows* show the varus point of the humerus. (**E**) Plain radiography of the entire length of the right humerus. (**F**) Plain radiography of the entire length of the left humerus.

Surgical technique

The patient underwent surgery under general anesthesia in the beach chair position (30° gap up). The deltopectoral approach was used, and the subscapularis tendon was detached by the peel-off technique. The proximal humerus was resected along native retroversion. At this point, the metaphyseal bone quality was evaluated by visual inspection, and the bone was compressed with the thumb. Bone qualities were sufficient; therefore, stemless implants were used as decided before the surgery. Additionally, the preoperative lumbar L1-L4 bone marrow density was normal, further confirming that stemless implants were appropriate. The humeral head was placed, the subscapularis tendon was reattached, securing the transosseous tunnels in the lesser tuberosity with a strong suture, and wound closure was performed in standard fashion. Intraoperative fixation of the humeral head and glenoid components was excellent in both shoulders. The humeral implants in both shoulders were the Comprehensive NANO stemless shoulder (Zimmer Biomet, Minato-ku, Tokyo, Japan). The humeral head was bio-modular, 44 × 15 mm on the right and 40 × 15 mm on the left. The glenoid implants in both shoulders were SMR (Lima, Shinagawa-ku, Tokyo, Japan). The implants used for the glenoid were both extra small, cementless glenoid 3 pegs on both the right and left shoulders. The glenoid was small, with a height of 30 mm and a width of 20 mm in this patient; thus, we used the SMR, which has a small variation in size.

Postoperative management

After the surgery, the shoulder was immobilized using an abduction brace, Ultrasling (DJO LTD, Guildford, Surrey, United Kingdom), for 4 weeks. Eccentric deltoid, passive-assisted external rotation, abduction, and forward elevation were initiated on day 1. Active external rotation, abduction, and forward elevation were started 2 months postoperatively, and push-up movement from the wheelchair was started 4 months postoperatively.

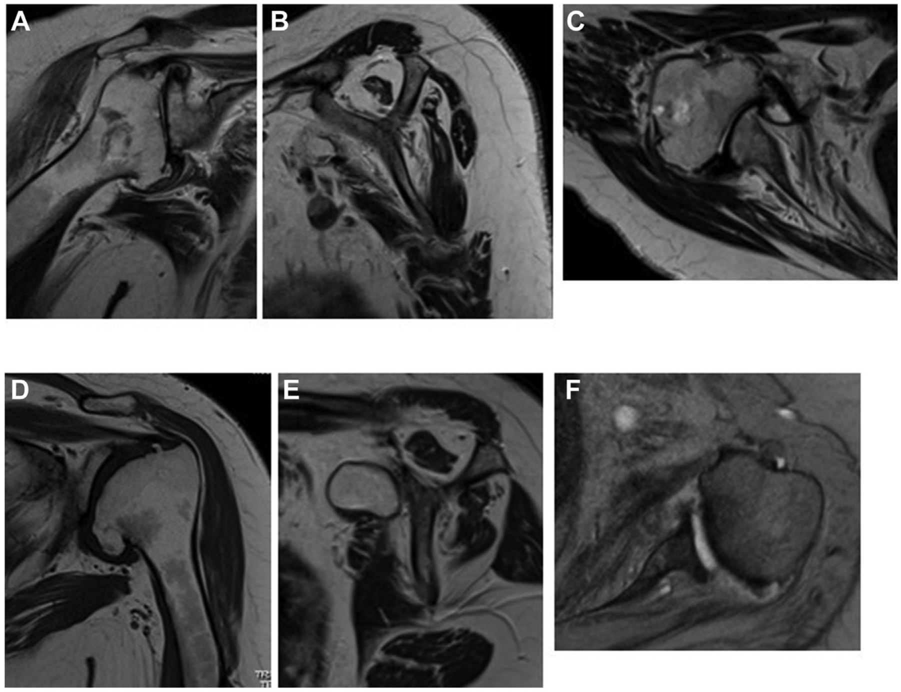

Figure 3 Preoperative T2-weighted magnetic resonance imaging. (**A**) Right shoulder coronal view. (**B**) Right shoulder sagittal view. (**C**) Right shoulder axial view. (**D**) Left shoulder coronal view. (**E**) Left shoulder sagittal view. (**F**) Left shoulder axial view.

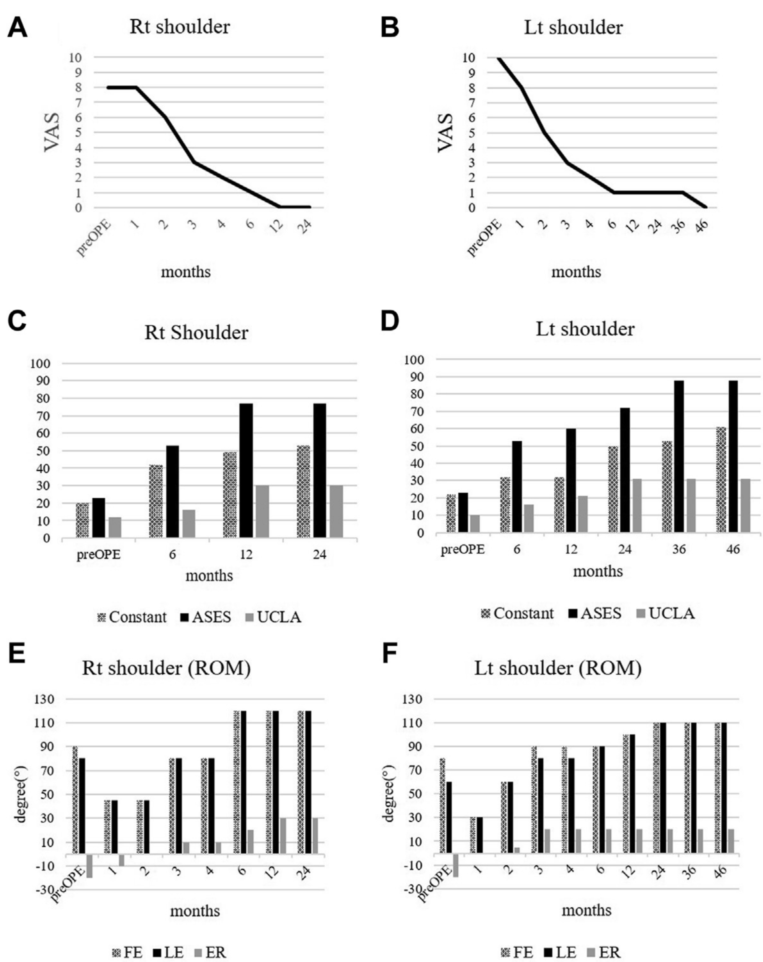

Figure 4 Postoperative clinical outcomes. (**A**, **B**) Visual analog scale score of both shoulders. (**C**, **D**) Constant, ASES, and UCLA scores of both shoulders. (**E**, **F**) Range of motion of both shoulders. *Rt*, right; *VAS*, visual analog scale; *ASES*, American Shoulder and Elbow Surgeons; *ROM*, range of motion; *Lt*, left; *FE*, forward elevation; *LE*, lateral elevation; *ER*, external rotation.

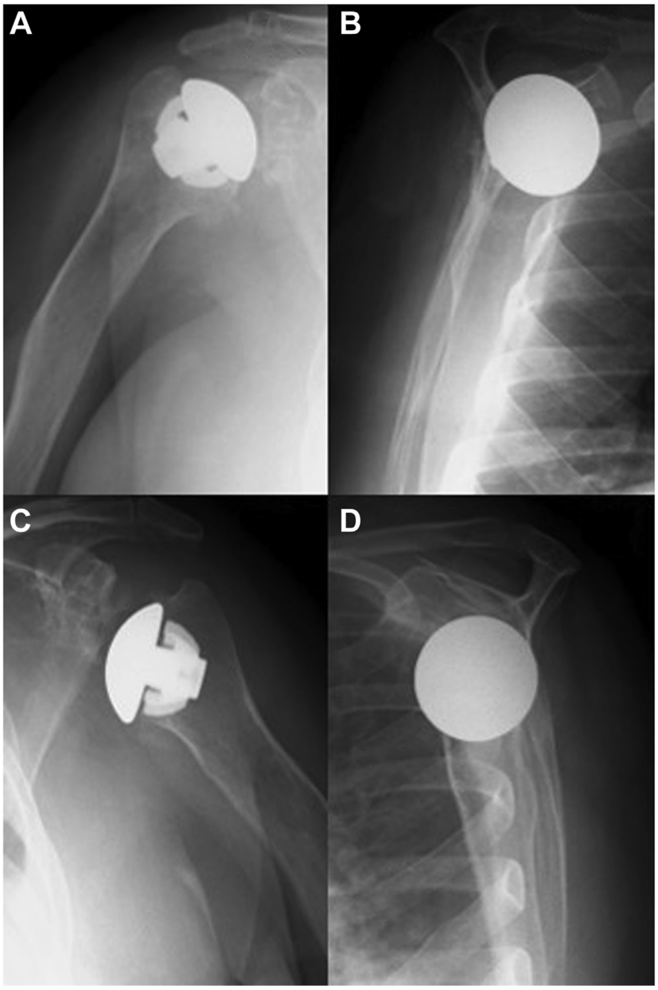

Figure 5 Plain radiographs immediately after surgery. (**A**) Anterior posterior (AP) view of the right shoulder. (**B**) Scapula Y view of the right shoulder. (**C**) AP view of the left shoulder. (**D**) Scapula Y view of the left shoulder.

Results

The visual analog scale score of the left shoulder was 10 before the surgery, which improved to 5 at 2 months, 2 at 4 months, 1 at 1 year, and 0 at 3 years 10 months, postoperatively (Fig. 4, *A* and *B*). The Constant score was 22 preoperatively and improved to 32 at 6 months, 32 at 1 year, 50 at 2 years, and 61 at 3 years 10 months postoperatively. The American Shoulder and Elbow Surgeons and UCLA scores of the left shoulder also showed gradual improvement after surgery (Fig. 4, *C* and *D*). The active range of motion was 80° of flexion, 60° of abduction, −20° of external rotation, and buttock level of internal rotation preoperatively but improved to 90° of flexion, 90° of abduction, 20° of external rotation, and L1 level of internal rotation at 6 months postoperatively, to 100° of flexion, 100° of abduction, 20° of external rotation, and L1 level of internal rotation at 1 year postoperatively, and to 110° of flexion and abduction, 20° of external rotation, and T12 level of internal rotation at 3 years and 10 months postoperatively (Fig. 4, *E* and *F*). The visual analog scale score, clinical score, and range of motion of the right shoulder showed similar improvement as shown in Fig. 4. Postoperative radiography showed no abnormal findings in either shoulder, including dislocation of the implants, and no obvious complications developed (Figs. 5 and 6). The total follow-up period was 2 years for the right shoulder and 3 years and 10 months for the left shoulder. The patient's range of motion at last follow-up is shown in Fig. 7.

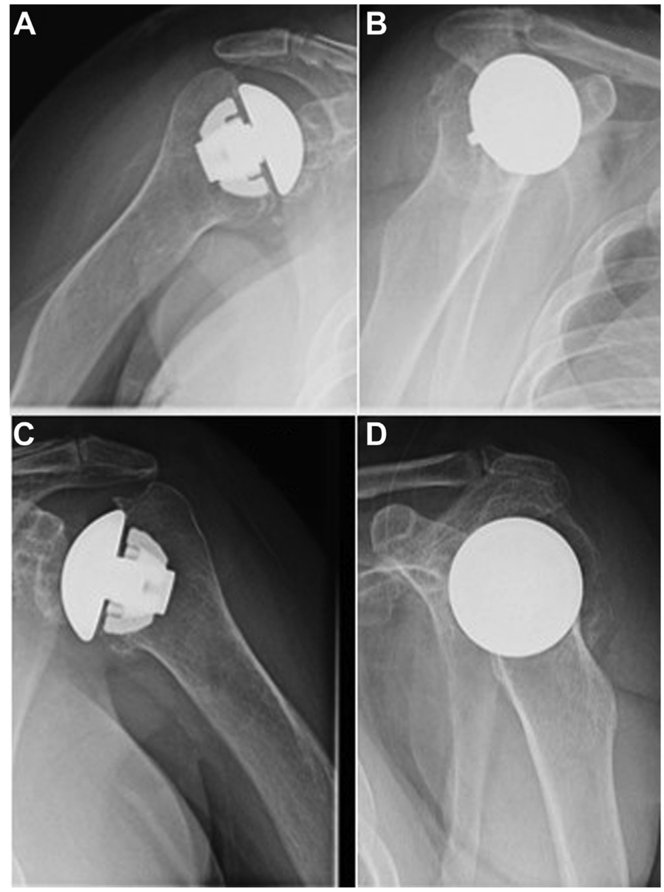

Figure 6 Plain radiographs at the last follow-up period. (**A**) Anterior posterior (AP) view of the right shoulder. (**B**) Scapula Y view of the right shoulder. (**C**) AP view of the left shoulder. (**D**) Scapula Y view of the left shoulder.

Discussion

There are 2 types of MED: the severe Fairbank type and the mild Ribbing type. The Ribbing type is more common, in which limb shortening is mild or normal. In the Fairbank type, dysplasia of the tubular epiphyses results in limb shortening and short stature.[7] Roland classified shoulder arthropathy secondary to MED into 2 types: the minor epiphyseal abnormality type and the hatchet-head type. In the hatchet-head type, osteoarthritis develops earlier, resulting in severe contracture of the shoulder joint. This type is characterized by an enlarged epiphyseal area, a strong deformity of the head, and a curvature of the diaphysis. The glenoid is also often hypoplastic, resulting in poor joint conformity and limited range of motion at an earlier stage.[7] In this case, in addition to the deformity of the shoulder joint, bilateral upper arm curvature was also observed and contractures were strong; thus, both shoulders met the criteria to be classified as the hatchet-head type. For stemless TSA, numerous reports show that postoperative range of motion and clinical scores are not different from those of conventional TSA[2,10] and that there is no significant difference in reoperation rates.[14] In a study of 40 hemi-shoulder arthroplasty and 35 TSA cases with stemless implants with an average follow-up of 126 months, Magosch et al reported that no cases of stemless implant loosening occurred, indicating that there were no

Figure 7 Range of motion of the patient at last follow-up.

problems with the fixation of the stemless implants.[11] The clinical advantages of stemless implants include bone preservation,[6] shorter operative time,[9] less intraoperative blood loss,[1] less stress shielding,[15] and easier implant removal during revision surgery.[6] Razfar et al performed a finite element analysis to simulate the loading stress on the humeral bone, comparing the loading stress on the humerus at the normal bone, standard stem, short stem, and stemless TSA models. The stresses on the proximal cortical bone were reduced in the standard and short stems, and the stresses on the distal cortical bone and distal tip of the implant were the same as those in the normal model. Stemless implants, on the other hand, showed almost the same stresses on the proximal cortical bone as those in the normal model.[15] In this case, the stemless implant was chosen due to concerns about stress concentration in the curvature; a stemless TSA for shoulder arthropathy secondary to MED was performed with good albeit short-term results. Fixation of the stemless implant depends on the bone quality of the proximal humerus.[5] There are several reports of methods showing bone quality by pushing the osteotomy surface with the surgeon's fingertips. This method of evaluating bone quality is subjective and depends on the surgeon's judgment.[3] Levin et al proposed a threshold for bone density using the deltoid tuberosity index and proximal humerus Hounsfield units on preoperative radiographic and computed tomography studies to allow for the preoperative determination of sufficient bone volume for stemless TSA and showed that lower deltoid tuberosity index values and bone density in the proximal humerus with Hounsfield unit values were associated with the need to switch from stemless to short stem humeral fixation in the primary TSA.[8] Further research, including the accumulation of bone density data, is needed to determine preoperative bone quality in the proximal humerus.

Conclusion

To the best of our knowledge, this is the first description of stemless TSA for arthropathy secondary to MED, which showed good short-term results. Stemless implants may be useful in cases of shoulder arthropathy secondary to MED, where stem insertion is difficult.

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