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ORIGINAL PAPER

Shoulder arthroplasty in patients with osteo-chondrodysplasias

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Abstract
Purpose Osteo-chondrodysplasias are a rare cause of limb malalignment, deformity and degenerative joint disease. Earlier in life, deformities may be managed with bony realignment and soft tissue releases; however, as degenerative changes progress, arthroplasty may be considered. There are limited reports examining shoulder arthroplasty in this population. This study aims to assess pain relief, function, and re-operation rate of shoulder arthroplasty in patients with osteo-chondrodysplasias.
Methods Between January 1984 and December 2012, 13 shoulders with end-stage arthritis secondary to osteo-chondrodysplasia underwent shoulder arthroplasty. Three were treated with hemiarthroplasty (HA), nine with anatomic total shoulder arthroplasty (TSA), and one with a reverse total shoulder arthroplasty (RSA). All shoulders were followed for two years or until reoperation (mean 7.9 years, range 2–25).
Results Shoulder arthroplasty significantly improved pain, elevation, external rotation, and internal rotation. All but one patient considered their shoulder to be better than pre-operatively; however, only two shoulders received an excellent Neer rating. Seven shoulders had satisfactory Neer ratings and four unsatisfactory. One TSA was converted to a RSA for aseptic glenoid loosening at 9.5 years (re-operation rate 8%).
Discussion Pain relief and improved function can be expected in patients with osteo-chondrodysplasias despite challenging anatomy. Unlike the only previous case series reporting a 31% revision rate at mean follow-up of seven years, our series shows the incidence of failure to be much lower.
Conclusions With the advent of smaller humeral components, the need for custom implants may not be necessary, and surgeons may intervene earlier and more confidently in this population.
Level of evidence: IV case series

Keywords Shoulder arthoplasty · Dysplasia · Skeletal dysplasia · Osteo-chondrodysplasia · Custom implant · Short stature

Introduction

Osteo-chondrodysplasias are a group of disorders which result in abnormal cartilage development leading to phenotypic skeletal abnormalities [1]. As a group, these disorders affect skeletal growth and alignment. As a result, they can also affect patient function. The phenotypic presentation is variable, with not all patients developing severe deformity. However, as a group, these patients tend to have limb malalignment, deformity and in some cases a predisposition to developing degenerative joint disease [2–4]. Early in life, deformities may be managed with bony realignment and soft tissue releases; however, as degenerative changes progress, arthroplasty may become the strongest treatment option [5, 6].

Shoulder arthroplasty can be challenging in this population for multiple reasons. Patients often have abnormal humeral anatomy which can limit prosthetic size options. Previous reports on this patient population have utilized custom prostheses to accommodate altered humeral anatomy in this population [7, 8]. Glenoid dysplasia may preclude the placement of a glenoid component or potentially place a resurfaced and realigned glenoid at increased risk of failure [9]. Patients

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may also have muscle hypotonia or joint contractures making surgical exposure difficult [2]. Concurrent lower extremity involvement may necessitate the use of gait aids, further placing a shoulder prosthesis at risk for failure. Previous studies have looked at lower extremity arthroplasty in this population; however, outcomes after shoulder arthroplasty in this population are limited to a few case reports and one small study [7, 8, 10–12]. With this study, we aim to assess pain relief, function, and survivorship of shoulder arthroplasty in patients with an underlying diagnosis of osteo-chondrodysplasia.

Methods

All human studies have been approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki. All persons gave their informed consent prior to their inclusion in the study. Between January 1984 and December 2012, 13 consecutive shoulders in nine patients with skeletal dysplasia were treated with shoulder arthroplasty (TSA) for end stage arthritis after failing non-operative management. Nine shoulders were treated with an anatomic TSA, three with a hemiarthroplasty (HA), and one with a reverse should arthroplasty (RSA). One patient had bilateral HA and two had bilateral TSA. One patient had a TSA on one side and an RSA on the other. All shoulders were followed clinically for a minimum of two years. Average follow up was 7.9 years (range, 2–20). Seven shoulders were evaluated in clinic at follow up, and six were followed remotely using a validated shoulder questionnaire [13]. The average height of patients undergoing arthroplasty was 145 cm (range 121–168). Average weight was 54 kg (range 47–72). Complete demographics with patient diagnoses are in Table 1.

At follow up, all shoulders were evaluated for pain, range of motion and patient satisfaction. Pain was evaluated on a five-point scale as described by Neer [14]. Subjective patient satisfaction was evaluated per patient responses of “much better”, “better”, “the same”, or “worse” than before the index shoulder arthroplasty. Active shoulder elevation in the scapular plane and external rotation were recorded in degrees, with internal rotation recorded as the highest vertebral segment reached by the thumb. Modified Neer ratings were calculated for each shoulder [14, 15]. Shoulders are graded as an excellent outcome when a patient is able to achieve active elevation of at least 140°, active external rotation of at least 45°, have “no” or “slight” pain and be subjectively satisfied with their shoulder. A satisfactory rating is given to shoulders with “moderate pain only with vigorous activity” or less, active elevation of at least 90°, external rotation of at least 20°, and patient satisfaction. Failure to meet the above active motion, moderate or severe pain, or subjective dissatisfaction results in an unsatisfactory modified Neer rating.

Ten shoulders were evaluated radiographically at an average of 5.8 years (range, 1–11) after arthroplasty. Three shoulders had radiographic follow-up of less than one year and were excluded from radiographic analysis. Pre-operative radiographs were available for all shoulders (Fig. 1). Radiographs were evaluated for pre-operative subluxation, joint space narrowing, and glenoid erosion. Subluxation was classified as none, mild (<25%), moderate (25–50%), or severe (>50%). Glenoid erosion was considered to be mild with wear to the subchondral plate, moderate with wear through the subchondral plate and severe with wear to or beyond the coracoid base. Post-operative radiographs were assessed for progressive cartilage loss and glenoid erosion (hemiarthroplasties), glenohumeral subluxation, periprosthetic lucencies and a shift in component position (Fig. 2). Periprosthetic lucencies were assessed using the method described by Sperling. If the prosthetic component was noted to shift in position compared immediate post-operative radiographs or a grade 4 or 5 radiolucency was present, the shoulder was considered to be “at risk” for failure [16].

Surgical technique

Shoulder arthroplasties were performed by three different fellowship trained shoulder surgeons over the study period. The deltopectoral approach was most common (14), with two shoulders undergoing an anteromedial approach due to limited external rotation intraoperatively. The subscapularis was tenotomized in 12 shoulders, with one undergoing a subscapularis peel. Humeral and glenoid deformities complicated the exposure in this group of patients. Aggressive capsular releases, including complete release of the humerus, were often required to obtain adequate exposure to both the deformed proximal humerus as well as the retroverted glenoid. Humeral bone grafting was required in three shoulders, both on the humeral side to supplement proximal fixation. Twelve of the 13 humeral components were placed in an uncemented fashion. Custom humeral components were required in two procedures, both of which were performed before the introduction of small humeral stem options. In both cases, the custom humeral component was shortened compared to standard implants to accommodate for the patient’s small stature (1) or bowed humeral deformity (1). Glenoid components were not routinely placed in the native glenoid retroversion, and instead surgeons attempted to eccentrically ream and partially correct patient retroversion.

Statistical analysis

Descriptive statistics are reported as mean (range) for continuous measures and number (percentage) for discrete variables. Re-operation for any cause was used to define implant survival. A Wilcoxon-rank-sum test was used to compare pre- verus post-operative changes in pain and range of motion. Radiographic

Table 1 Patient demographics

Patient	Type of dysplasia	Age at surgery (years)	Gender	Operative side	Prosthesis type	Pre-op range of motion (forward elevation/ external rotation/ internal rotation)	Post-op range of motion (forward elevation/ external rotation/ internal rotation)	Post op complication
1	Uncharacterized congenital dysplasia	41	Female	Left	TSA - custom humeral stem	110/15/Lat Ileum	105/65/Sacrum	Neuropraxia (ulnar)
2	Achondroplasia	59	Female	Left	TSA - custom humeral stem	60/0/Abdomen	160/30/L5	
3	Achondroplasia	60	Female	Right	TSA	80/0/Abdomen	120/30/Sacrum	
4	Morquio's syndrome	43	Male	Right	TSA	20/0/Greater trochanter	140/30/L3	
5	Morquio's syndrome	43	Male	Left	HA	60/30/Lat Ileum	140/30/L3	
6	Multiple epiphyseal dysplasia	53	Female	Left	TSA	60/10/Sacrum	100/50/L5	
7	Spondyloepiphyseal dysplasia	18	Female	Left	HA	90/30/L5	40/10/Post Ileum	
8	Spondyloepiphyseal dysplasia	38	Female	Right	TSA	90/30/Sacrum	120/90/T12	
9	Spondyloepiphyseal dysplasia	41	Male	Left	TSA	100/60/Sacrum	180/90/L2	
10	Spondyloepiphyseal dysplasia	42	Male	Right	TSA	85/20/Sacrum	180/90/L2	Intraop fracture greater tuberosity
11[a]	Spondyloepiphyseal dysplasia	49	Female	Left	TSA	150/45/T12	Not Tested	
12	Stickler's syndrome	76	Female	Right	HA	80/20/Greater trochanter	85/25/PSIS	
13	Stickler's syndrome	83	Female	Left	RSA	80/20/Lat Ileum	140/25/PSIS	

[a] Revision

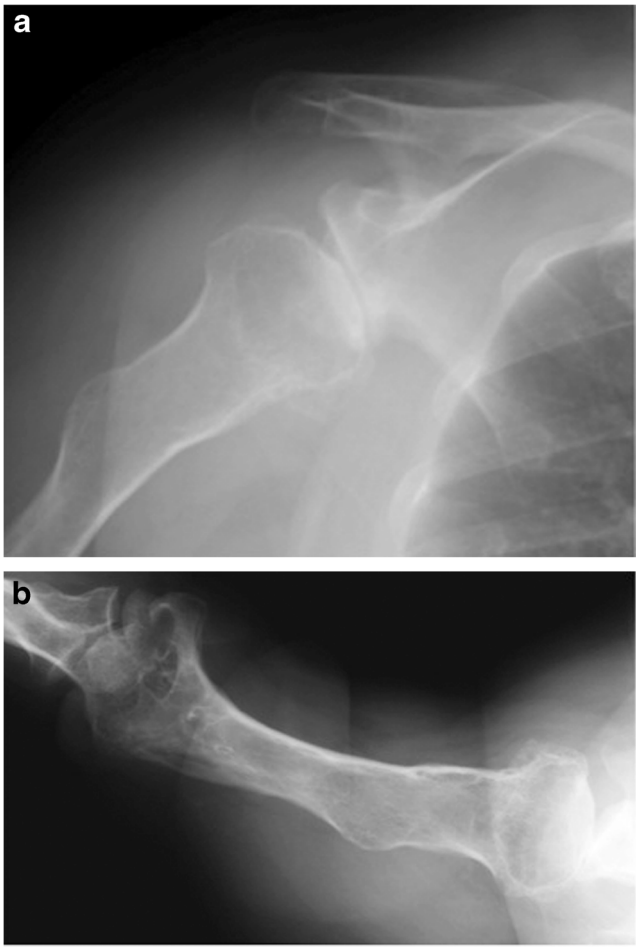

Fig. 1 **a,b** Pre-operative internal rotation (**a**) and axillary (**b**) views of a 58-year-old female with Achondroplasia and severe posterior subluxation

follow up of one year or until re-operation was assessed in a similar manner as outlined above. The alpha level for all tests was set at 0.05 for statistical significance.

Results

Significant pain improvements were noted from pre to post operatively, 4.6 to 1.8 ($p < 0.001$). Subjectively, 12 of 13 shoulders rated their outcomes as much better (8) or better (4) than prior to surgery. One patient, who underwent a revision, described their shoulder as being worse than before surgery. Objective improvements were noted in active elevation, external rotation and internal rotation. Active elevation improved from 82 to 126 ($p < 0.01$). External rotation improved from 22 to 47 ($p < 0.005$). Internal rotation improved from the sacrum preop to L4 postop ($p = 0.003$). In determining modified Neer ratings, only two shoulders achieved an excellent result. Seven shoulders achieved a satisfactory result, and four shoulders yielded an unsatisfactory result. Unsatisfactory results were due to pain (1), decreased range of motion (2), and reoperation (1).

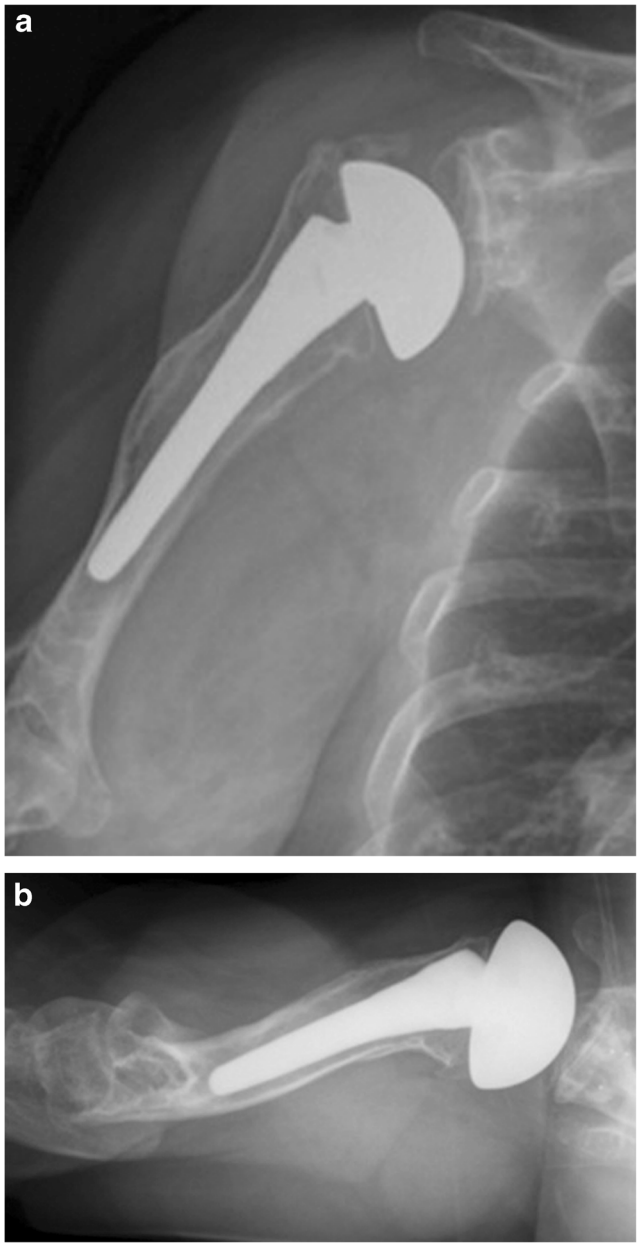

Fig. 2 **a,b** Post-operative radiographs of the same patient with Achondroplasia after undergoing an anatomic total shoulder replacement

Complications occurred in two shoulders. One shoulder developed post-operative ulnar nerve paresthesias which resolved by the time of her one-year follow-up appointment. One shoulder developed aseptic loosening of the glenoid component, which ultimately was revised to a RSA at 9.5 years post operatively.

Pre-operative radiographs were available for all shoulders. Dysmorphic features were noted in all shoulders and included a combination of proximal humeral bowing, abnormal epiphyseal alignment, altered humeral/glenoid version. Moderate

subluxation was present in three shoulders (superior-1, posterior-1, and inferior-1). Pre-operative glenoid erosion was present in 11 shoulders. This was graded as severe central (1), severe posterior (1), severe inferior (1), central (1), mild central (6), and mild posterior (1). Post-operative radiographs were available for ten shoulders (six TSA, three HA, one RSA). Glenoid erosion had progressed in all three HA, with two being graded as severe. Severe superior subluxation was present in one HA, but all TSA remained well centred in relation to the glenoid. No humeral stems were identified as having radiolucent lines. Four glenoid components had lucencies and were grade 1 (1 shoulder), grade 3 (1 shoulder), grade 4 (1 shoulder), grade 5 (1 shoulder). Two glenoid components had shifted in position when compared to immediate post op radiographs. Of these shifted components, one also had a grade 5 lucent line, leaving two glenoids "at risk" at final follow-up. One TSA was revised as outlined previously, and one continued to live with a painful shoulder but did not elect to pursue revision surgery. The single RSA had a stable component without loosening or notching at two-year follow up.

Subgroup analysis

The nine TSAs were compared to the three HAs to assess for differences in pain and range of motion. No significant differences were noted in pre-operative pain or range of motion ($p > 0.05$). Post-operative external rotation was greater in the TSA group (59 versus 22, $p = 0.04$). There were no other statistically significant differences between the groups in regards to preoperative and post-operative pain and range of motion ($p > 0.05$).

Discussion

Reports on shoulder arthroplasty for the treatment of osteochondrodysplasias remain limited [7, 8, 12]. The largest series was published recently and examined 13 shoulders in ten patients [12]. Compared with their series, we were able to corroborate a significant reduction in pain. Both groups studied also demonstrated improved range of motion; however, follow-up motion in elevation (126 versus 84) and external rotation (47 versus 24) were greater in this study. This may be in part due to the greater restriction of motion pre-operatively in Sewell's series. Additionally, the re-operation rate in this series (8%) does not support the concern raised by the previous series where 31% of the shoulders required reoperation at a similar follow-up of seven years. No common failure mode was identified across these two studies. Instead, common failure modes of specific implants were seen. However, with distorted anatomy, surgeons should be cautious to appropriately balance the shoulder intraoperatively when considering anatomic arthroplasty to prevent post operative instability in patients with pre-operative bony abnormalities and concurrent soft tissue contractures.

The rarity of these diagnoses make the study of this population difficult. Our study is limited to a small group of patients over a 28-year period. Three surgeons performed all operations, possibly limiting the generalizability of this procedure over a broader population of surgeons. We acknowledge that both implants and techniques have changed over this time, where some early patients would likely have been treated using a reverse shoulder prosthesis today. The study length did not allow for all patients to be assessed with ASES or other modern validated shoulder scores which would have better represented patient outcomes. The Neer rating remains limited by their objective requirements, allowing a patient who is satisfied with their shoulder to be graded as unsatisfactory due to range of motion parameters. Additionally, some patients elected to forego radiographs at follow-up due to out of pocket costs, leaving patients represented by this study with greater clinical follow-up than radiographic follow-up. Therefore, more significant radiographic wear may have been present at the time clinical follow-up was obtained.

With the introduction of smaller humeral stems and increased modularity, custom implants are less likely to be required. The retrospective nature of this study in addition to the small population increases the opportunity for type 2 beta error or garnering a meaningful complication/revision rate. Given the limited outcome data available and in introduction of newer, more conforming implants, the study remains important.

Like the previous series, shoulder arthroplasty in patients with osteochondrodysplasias can be expected to provide improved pain and range of motion [12]. Restoration or improvement elevation is extremely important in this population, which is often short in stature and needs the ability to use the arm overhead. Ninety-two percent of shoulders in our study rated their shoulder as much better or better than before surgery despite 31% having an unsatisfactory modified Neer rating. With the advent of new implants, complications such as intraoperative fractures may be lessened. Those patients with severe glenoid dysplasia remain challenging secondary to poor glenoid bone stock [9]. In such patients, reverse TSA may provide an option for more stable glenoid fixation. Ultimately, shoulder arthroplasty is a reliable intervention leading to improved patient outcomes in the setting of osteochondrosyplasias. Surgeons may be able to intervene earlier more confidently in this population which can be expected to have significantly improved pain and range of motion.

Conclusion

Patients with osteo-chondrodysplasias treated with shoulder arthroplasty represent a challenge due to soft tissue contractures

and altered glenohumeral anatomy. Shoulder arthroplasy provides patients pain relief and improved function despite this challenging anatomy. With the advent of smaller humeral components, the need for custom implants may not be necessary, and surgeons may intervene earlier and more confidently in this population when arthroplasty is indicated.

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The study was performed in accordance with the ethical standards in the 1964 Declaration of Helsinki.

Conflict of interest The authors declare that they have no conflict of interest.

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Ethical approval This study was approved by the IRB.

Informed consent Informed consent was obtained from all individual participants included in the study.

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