Review

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Shoulder arthroplasty for juvenile idiopathic arthritis

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Abstract

There is limited literature to guide shoulder surgeons in the management of juvenile idiopathic arthritis (JIA). We aim to help clinicians to formulate an approach to the surgical management of the condition through a review of the available literature on arthroplasty in JIA, general considerations when operating on patients with inflammatory arthropathy and recommendations based on the authors' experience. Four articles report formal data on arthroplasty in JIA with favourable improvements in post-operative pain and function scores after the long-term follow-up. Significant heterogeneity in treatment and a lack of standardisation in quantitative outcomes highlights the need for further larger scale and higher quality research. The aim of this study is to review the evidence and provide information on preoperative evaluation of surgical candidates, operative techniques, choice of implant design and to evaluate functional outcomes in patients who undergo shoulder arthroplasty.

Keywords

arthroplasty, hemiarthroplasty, JIA, JRA, juvenile idiopathic arthritis, shoulder

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Introduction

Epidemiology

Juvenile idiopathic arthritis (JIA) is the most common chronic rheumatic disease in childhood, with an overall incidence of 16 per 100,000. The average onset is 6 years, with peaks between 1 year and 4 years and between 9 years and 14 years. Girls are more affected than boys.[1,2] It can cause significant pain, disability and restriction in school and other activities in addition to an increased number of hospital visits.[3] Shoulder involvement tends to occur later in the course of pathogenesis with 15% incidence at 15 years.[4] Loss of abduction and internal rotation are often the first indications of significant shoulder involvement.[2,5]

Classification

JIA has been known in the past as juvenile rheumatoid arthritis and juvenile chronic arthritis each with their own classification systems for diagnosis. In 1995, the International League of Associations for Rheumatology formed a new classification system, subsequently revised in 2001, which consists of seven main categories of JIA in the hope of identifying potential differences in treatment response and prognosis. Oligoarthritis affects one to four joints during the first 6 months of disease. Polyarthritis affects five or more joints and can be separated into rheumatoid factor positive and rheumatoid factor negative. Psoriatic arthritis includes arthritis in addition to psoriasis or psoriatic features such as dactylitis. Enthesitis-related arthritis may include the presence of human leucocyte antigen (HLA)-B27 antigen while undifferentiated arthritis

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may fulfill no criteria or two or more of the above criteria.[6] Shoulder involvement is commonly seen in the polyarthritic form of JIA.[7]

Management

Management of JIA is based on medical interventions, physical and occupational therapy and psychosocial support.[8] Non-steroidal anti-inflammatory drugs (NSAIDs) have been the mainstay of treatment for decades.[8] However, the combination of NSAIDs and intra-articular corticosteroid injections are only partially effective at treating the symptoms and reducing long-term complications such as growth delay, erosive joint disease, persistently active disease and mortality. Disease-modifying anti-rheumatic drugs (DMARDs) have become an important component in the medical management in JIA patients, with increased remission rates and reduced long-term suffering.[9]

Methotrexate (MTX), a non-biologic drug, is recommended by the American College of Rheumatologists as the first-line treatment in JIA affecting five joints or more. In disease affecting less than five joints, it is recommended as second line if symptoms are not controlled with NSAIDs and/or glucocorticoid injections.[10] In disease not controlled by MTX treatment is escalated to biological DMARDs. These target and modulate specific components of the immune system. The mechanisms of action in biological DMARDs used for JIA include tumour necrosis factor alpha inhibitors such as adalimumab and etanercept, interleukin 1 receptor antagonists such as anakinra and interlueukin 6 receptor antibodies such as tocilizumab.[11]

For those patients who have failed medical management, surgical intervention is often indicated. Patients with aggressive polyarticular disease often require arthroplasty in young adulthood. In the shoulder, there are joint-preserving strategies, such as synovectomy, arthroscopic comprehensive arthroscopic management (CAM) procedure and interposition, in addition to salvage options such as arthroplasty, excision or arthrodesis. Arthroplasty options include resurfacing hemiarthroplasty (RHA) or stemmed hemiarthroplasty (SHA) or total shoulder arthroplasty (TSA).

Preoperative challenges

There are a number of challenges when considering shoulder arthroplasty for JIA patients. The perioperative management of DMARDs must be considered. Fibreoptic intubation should be considered with associated cervical spine disease and the potential risk of posterior atlanto-axial subluxation. The disease process stunts skeletal growth,[12] and as such 'off-the-shelf' implants may not be suitable for these patients. Furthermore, the relatively young age of patients undergoing arthroplasty provides a challenge for preserving bone and optimising tribology.

Shoulder JIA severely disrupts function in activities of daily living and potentially delays postoperative rehabilitation from hip and knee arthroplasty for the same disease. If there is elbow involvement also, then this effect is compounded for the patient. Clearly, the timing of surgical intervention for different joints must be carefully coordinated between orthopaedic subspecialties to avoid compromised rehabilitation for any procedure.

Need for review

There are few published articles on glenohumeral arthroplasty in JIA in the contemporary literature, with even fewer that have set parameters on when to operate or how to evaluate functional outcome postoperatively. The aim of this study is to review the evidence and provide information on preoperative evaluation of surgical candidates, operative techniques, choice of implant design and to evaluate functional outcomes in patients who undergo shoulder arthroplasty.

Materials and methods

Literature search

A search of the literature was conducted by two independent reviewers on PubMed, Medline, OVID, NICE and Cochrane Library to identify publications.

The key search terms used were Juvenile idiopathic Arthritis, JIA, Shoulder, Arthroplasty and Hemiarthroplasty. The literature search was carried out using an advanced search using combinations of the above keywords.

Criteria of inclusion

English language studies providing data on patients with a diagnosis of JIA who underwent SHA, RHA or TSA were included. No exclusion criteria were made regarding preoperative conditions, outcomes of surgery, age at the time of surgery or minimum length of follow-up. For publications on TSA or hemiarthroplasty which included a proportion of JIA patients, it was an inclusion requirement to state this proportion of JIA patients and to report on their outcomes independently.

Study selection

A total of 95 citations were screened. A total of 14 duplicate citations were removed. Fifty-seven publications were excluded by title after the initial search as they had no reference to (1) JIA, (2) shoulder joint or (3) total/hemiarthroplasty or surgical intervention.

The abstracts of 24 articles were screened; however, 11 articles were excluded as they were not eligible for review as there were no specific data on surgical outcomes. The full articles were retrieved for 13 publications and a further 9

Table 1. Demographic characteristics of included studies.

References	Year	Location	Age	Range	Gender M	Gender F	Patients (*n*)	Shoulder arthroplasties (*n*)	Arthroplasties followed-up (*n*)	Mean follow-up (years)	Range
Deshmukh	2005	Los Angeles/Boston, SA	60.3	17–85	58	209	16	16	16[a]	8.7	2.6–14.8
Bogoch	2007	Lausanne, Switzerland	33.1	25–53	3	5	10	13	11	9.1	4–15
Thomas	2004	Wexham Park, UK	32	25–50	1	8	13	15	9	6	4.9–7.1
Ibrahim	2018	Wexham Park, UK	36.4	19–49	2	9	11	14	14	10.4	5.8–13.9
						Total	50	58	50	8.6	

JIA: juvenile idiopathic arthritis.
[a]This assumes all 16 JIA cases were followed up as none were included in lost to follow-up data.

Table 2. Evidence.

References	Operative period	Location	Study type	Level of evidence[a]
Deshmukh	1974–1988	Los Angeles/Boston, USA	Retrospective	III
Bogoch	1986–1997	Lausanne, Switzerland	Retrospective	III
Thomas	1995–1999	Wexham Park, UK	Prospective	II
Ibrahim	2002–2010	Wexham Park, UK	Retrospective	III

[a]Oxford Centre for Evidence-Based Medicine 20011 Levels of evidence.

were excluded as they did not contain any patients with JIA. These nine articles were studies on shoulder arthroplasty in rheumatoid arthritis (RA) that either did not contain any JIA cases or matched the exclusion criteria. This review was thus concluded with four articles that presented surgical data on patients with a formal diagnosis of JIA.[13–15]

Data extraction

A spreadsheet was used to extract the data from each study that met the inclusion criteria. This included time period of study, type of study, the number or patients and individual shoulder cases (JIA), average age at surgery, male-to-female (M:F) ratio, hand dominance, indication for surgery, previous operations, rheumatoid factor status, time to surgery after diagnosis, cemented versus uncemented procedures, mean follow-up, complications and failures and functional outcome scores.

Selection or exclusion bias

All three studies were cohort studies with no comparison group, two retrospective and one prospective (Table 2). One study (Deshmukh), which had a minority of JIA patients, had a much higher average age and did not give a breakdown of the average age of JIA patients. The two JIA-only studies (Jolley and Thomas) had similar mean ages. The M:F ratios between the three studies varied (Table 1).

Four functional scoring systems were used; however, no single common system was used. These functional outcome systems include: disabilities of the arm, shoulder and hand (DASH) score, visual analogue scale (VAS), short form 36 (SF-36) and the constant score. The number of 'operated-on shoulders' lost to follow-up ranged from 6% to 15%; however, Deshmukh et al. do not specify how many specific shoulder patients were lost to follow-up, and of these how many were JIA shoulders. As such there appears to be minimal potential for selection or exclusion bias in the review.

Results

Demographic and pathological data

A total of 50 patients underwent 58 shoulder operations (Table 1). Fifty of the 58 were followed up. Deshmukh et al. operated on 16 JIA shoulders. They did not specify whether any of their numbers lost to follow-up had contained any JIA patients. As such it has been assumed that no JIA patients were lost to follow-up. Thomas et al. excluded six shoulders as their follow-up time was less than 2 years as part of their exclusion criteria. The total mean follow-up of the four studies was 8.6 years.

Table 2 demonstrates the operative time frame of the shoulder arthroplasties performed and the level of evidence of each of the articles according to the Oxford Centre for Evidence-Based Medicine 2011 Levels of evidence.

Thomas et al. reported their indications to operate. Clinically, pain and loss of function with end-stage destruction of joint. Radiographically, migration of the humeral head, bowed proximal diaphysis, overgrowth of the greater tuberosity, narrow intramedullary canal and erosion of the joint with a notched metaphysis. Ibrahim et al. had similar

Table 3. Pre and postoperative functional status.[a]

References	Forward flexion (°) preoperative	Post-operative	Internal rotation (level) preoperative	Post-operative	External rotation (°) preoperative	Post-operative
Deshmukh	57	80	L2-Sacrum	L2-Sacrum	12	25
Bogoch	52.3	89.1	Unrecorded baseline	Mean +3 vertebral levels	−8.6	30.5
Ibrahim	69	110	0[b]	2.5[b]	12	32

[a]Both Deshmukh and Jolles provide preopeartive DASH scores, but no follow-up data.
[b]Ibrahim et al. measure internal rotation (IR) according to predefined levels of thumb tip position, when trying to reach behind the back; 0 = lateral thigh, 1 = buttock, 2 = SI joint, 3 = midlumbar spine, 4 = T12, 5 = T6.

Table 4. Outcomes.[a]

References	VAS preoperative Mean (SD)	9-year follow-up mean (SD)	CS preoperative mean (SD)	5-year follow-up mean (SD)	SF-36	OSS preoperative mean	Follow-up mean
Deshmukh							
Bogoch	8 (1)	1.3 (1)			As described		
Thomas			11 (3.25)	50.1 (13)			
Ibrahim	9.0	0.64	15.2	57.0		11.8	36.0

VAS: visual analogue scale; CS: constant score; SF-36: short form 36; OSS: Oxford shoulder score.
[a]Both Deshmukh and Jolles provide preopeartive DASH scores, but no follow-up data.

criteria for surgery but also stipulated that patients had not had adequate satisfactory symptom control with nonsurgical management. Bogoch et al. described preoperative radiographic findings in a typical JIA shoulder: severe humeral and glenoid bone loss, medial migration of the humeral head to the base of the coracoid and ankylosis of the acromioclavicular joint.

Thomas et al. reported that all their patients were diagnosed with JIA before the age of 10 years but had surgery as adults suffering from systemic or polyarticular disease. All had undergone multiple operations in other joints and were noted to have ipsilateral elbow involvement at the time of surgery.

Clinical outcomes

Bogoch et al. compared the preoperative SF-36 scores with the US population norms and found that in the JIA group they had significantly lower scores in the following domains: physical functioning, physical role, bodily pain, global health and vitality. In addition, they found smaller difference in social functioning systems, emotional role and mental health.

In addition, Bogoch et al. carried out their preoperative data on a post-operative recall basis and tried to quantify the original level of internal rotation according to vertebral level preoperatively and then quantified the number of levels increased or decreased according to the patient's own baseline (Table 3, mobility status).

Table 4 demonstrates the clinical outcomes recorded in each of the studies.

As per the initial description of the DASH score, healthy persons scores range from 3 to 6 on a 100-point scale, with a lower score representing a better function.[16] Of the three reviewed articles, Deshmukh et al. and Bogoch et al. published mean post-operative DASH scores of 51 and 44.7, respectively. This compares with the function of patients suffering from combined supraspinatus and infraspinatus rotator cuff tear requiring surgery.[17] The consensus between the Bogoch et al. and Thomas et al. patient populations were that patients reported relief of pain postoperatively, with a mean improvement in the VAS pain score by 6.7 and good relief from pain and restoration of useful function on the constant score (78% pain reduction, 33% improvement in function and 34% improvement in movement, respectively.)

Ibrahim et al. reported mean preoperative pain score of 9.0 out of 10, which improved to 0.64 at latest follow-up, with 8 out of 14 shoulders described as pain free. Constant–Murley scores (CMS) and Oxford shoulder scores (OSS) also improved significantly and a strong correlation was found between postoperative CMS and OSS scores. No shoulders had unsatisfactory outcomes in this article.

Bogoch et al. found improved range of movement of 39° of external rotation, which faired comparably with other shoulder arthroplasty outcomes, for example, 26° in glenohumeral joint osteoarthritis[18] and 35° in RA.[15] In addition, they reported poor SF-36 scores but their patients reported general satisfaction despite poor results.

Deshmukh et al. reported 320 consecutive cases of primary TSA over a period of 15 years. JIA patients formed

Table 5. Complications and revisions.

References	Arthroplasty	Complication	Age	Sex	Time	Outcome
Deshmukh	Total	Dislocation	39	F	1 day	Successful closed reduction
	Total	A	23	F	10.5 years	Cemented bipolar humerus, glenoid and humeral bulk allografts
Bogoch	Total and hemi	3 Intraoperative fractures	N/A	N/A	N/A	No additional fixation required – small cracks originating from exposed cortex of the resected osteopenia surface of the proximal medial humerus
	Total and hemi	Axillary nerve palsy	N/A	N/A	N/A	Spontaneous self-resolution
Thomas	Hemi	ACJ arthritis	26	F	9 months	Open excision of acromioclavicular joint
	Hemi	Persistent lateral forearm paraesthesia				Unable to self-toilet despite normal nerve conduction studies. MRI-C6 radiculopathy
Ibrahim	Resurfacing	Subacromial impingement			6 months	Arthroscopic subacromial decompression – resolved at 1 year
	Resurfacing	Subacromial impingement			6 months	Arthroscopic subacromial decompression – resolved at 1 year

the third largest cohort (5%) and JIA was considered as a majority diagnosis. They found that no diagnostic group behaved differently in terms of the type of prosthesis used (Neer II implant vs. non-Neer II implant prosthetic designs), survivorship, range of motion, strength, pain relief or DASH score.

Thomas et al. highlighted that postoperatively, there was a trend in worsening function over time, and that this was consistent with the disease pathology and outcome studies in patients diagnosed with JIA. Packham and Hall[19] demonstrated that those most at risk were those in the polyarticular or systemic JIA subgroup, going on to hypothesise that the 'burn out' phenomenon demonstrated in RA 'may not hold true' for JIA. Thomas et al. hypothesise that the decline in function outcome over time may be due to prosthetic loosening; however, there was no radiological evidence to prove this.

Complications and component failure

In the 4 studies reviewed, 10 complications were reported with only 1 requiring revision surgery (Table 5). Deshmukh et al. reported a dislocation of a TSA in a JIA patient occurring day 1 post-operatively, which was successfully treated with closed reduction. The authors stated that prosthetic positioning was not a cause of the instability. They also reported a case of humeral and glenoid loosening of a TSA 10.5 years post-operatively, which was managed with a cemented bipolar humerus and glenoid and humeral bulk allografts. No comment was made on the follow-up or the success of this treatment regime. Joules et al. reported a 23% rate of intraoperative fractures of the proximal medial humeral cortex which did not require additional fixation. In addition, they reported a case of axillary nerve palsy which spontaneously resolved. No information was given about the length of time it took to resolve. Thomas et al. reported two complications with shoulder hemiarthroplasties. The first was acromioclavicular joint arthritis which occurred 9 months postoperatively, requiring open excision of the acromioclavicular joint. The second was persistent lateral forearm paraesthesia that restricted the patient's self-toileting function. However, it is questionable whether this is attributable to either the disease process or the shoulder hemiarthroplasty itself as the patient had normal conduction studies and a magnetic resonance imaging (MRI) scan revealing C6 radiculopathy. Ibrahim et al. reported impingement syndrome symptoms in two patients who underwent arthroscopic subacromial decompression and improved, after initially not responding to nonsurgical intervention. Ibrahim et al. have followed-up patients radiographically also and report evidence of glenoid erosion in 5 of 14 shoulders, less than 5 mm in each case and not associated with a poorer clinical outcome.

Discussion

Limitations

The literature surrounding arthroplasty in JIA is limited. The articles we identified are limited in quality and report on heterogenous treatments with differing outcome measures. Only four publications met the inclusion criteria for shoulder arthroplasty in JIA, with specific breakdown of their data. In total, only 58 shoulders were operated on and 50 followed-up. Clearly drawing any significant conclusion, especially quantitative, is impossible. We were unable to perform a systematic review given this paucity of quality data.

We have based a set of recommendations around our own experience and that of the published authors featured. This cannot be taken to be based on high-quality evidence but, we hope, is a pragmatic guide for clinicians facing the operative management of JIA.

Preoperative cessation of medication

There has been significant debate regarding the effect of DMARD use on the risk of perioperative complications in adult RA patients undergoing elective orthopaedic procedures, with evidence suggesting that some novel DMARDs such as leflunomide can infer an increased risk of surgical site infection.[20,21]

Without robust data on JIA, we can only use recommendations for arthroplasty in RA as a guide. These recommendations however are often inconsistent; the American College of Rheumatology recommends withholding tumour necrosis factor (TNFα) inhibitors for at least one week prior to surgery,[22,23] while the British Society recommends withholding therapy for three to five times the half-life of the drug,[23] and the Canadian Rheumatology Association recommends withholding therapy for two half-lives of the drug.[24] Susan M Goodman, rheumatologist at the Hospital for Special Surgery, New York City, published guidelines on perioperative management of biologics and DMARDs with a focus on total joint arthroplasty.[25] More recently, Goodman and the American College of Rheumatology have collaborated with the American Association of Hip and Knee Surgeons to produce a comprehensive guideline on perioperative management of DMARDs for hip and knee arthroplasty in populations including those with JIA.[26]

The authors would recommend this, in conjunction with liaison with the patient's treating rheumatologist, as a guide for perioperative drug management.

Anaesthetic considerations

JIA patients are at risk of atlanto-axial subluxation.[27] This has consequences for intubation and positioning of the head. Again, in the absence of robust literature pertaining to JIA, it is reasonable to mirror practice in RA patients. In a review of perioperative management of RA patients published in the *Journal of the Association of Anaesthetists of Great Britain and Northern Ireland*, several recommendations were made on the issue.[28]

While it remains uncertain whether perioperative management is changed by diagnosis, atlanto-axial subluxation can be detected on plain flexion/extension radiographs[29] and sub-clinical subluxation on preoperative MRI.

Neck flexion and extension should be assessed and documented preoperatively, with the aim of avoiding these extents intraoperatively.

In anterior atlanto-axial subluxation a large donut ring head support large enough to accommodate the occiput can prevent anterior movement of the head and C1.[30]

Furthermore, the Association of Anaesthetists of Great Britain & Ireland (AAGBI) authors consider fibre-optic intubation to be the appropriate and safe option in RA patients with an anticipated difficult airway or known cervical instability.

Preoperative planning

Operative choices. Prior to joint replacement, joint-preserving strategies can be employed in patients not responding to disease modifying anti-rheumatic drugs. Arthroscopic debridement and synovectomy is recognised in the treatment of RA.[31] Ovregard et al. suggested a 3-year follow-up period to evaluate the effect of synovectomy in children.[32,33] Restorative, reparative and reconstructive techniques are options for the young patient with osteoarthritis. These include microfracture and osteochondral transport alongside a strict rehabilitation programme to stimulate healing response.[34,35] The CAM procedure includes glenohumeral chondroplasty, removal of loose bodies, humeral osteoplasty, osteophyte resection, capsular release, subacromial decompression, axillary nerve neurolysis and biceps tenodesis. This has been shown to reduce pain and improve function in young active with severe glenohumeral joint osteoarthritis with 85% 2-year survivorship.[36] Burkhead and Hutton demonstrated good functional outcomes with autologous fascia lata biologic glenoid resurfacing and hemiarthroplasty[33] although these results have yet to be replicated. In the case of severe articular destruction and failed medical and failed non-arthroplasty techniques, hemiarthroplasty and total shoulder replacement are the next options.

Arthroplasty. Hemiarthroplasty is traditionally preferred in younger patients to avoid the complications of glenoid loosening associated with TSA.[37] In addition, it is more bone preserving and pre-empts future revisions. However, hemiarthroplasty has come under scrutiny with studies reporting better long-term outcomes with TSA, even in young patients, thought to be secondary to glenoid bone erosion from metallic components.[38,39] RHA has the benefit of preserving bone stock and also obviates the concerns of unusual bony anatomy such as diaphyseal shape and size that is a challenge when using stemmed implants. Humeral head erosion can be a concern, however, and allograft may be needed.

Various stemless hemiarthroplasty designs are currently undergoing clinical trials.[40] These may confer an advantage for JIA patients with stunted skeletal growth and humeral shaft deformity. They may also facilitate further revision surgery with mitigated bone loss and also circumvent the concerns of humeral head erosion when utilising RHA.

JIA patients may require multiple revisions throughout their lifetime. The need for bone preservations must be weighed against better reported functional outcomes in TSA. The authors would recommend either a stemless implant or a stemmed implant that is part of a platform system to allow for revision. With regards total shoulder replacement, all polyethelene glenoid compenents have been shown to have superior survivorship in the average patient,[41] however, severe bone loss can make revision

difficult,[42] making metal backed glenoid components an option for the JIA patient.

Deshmukh et al. used six different components which included Neer II design, Kirschner II design, Gristina, Dana, Cofield and Michael-Reese. However, there is no detail regarding which component was used for the 16 JIA cases out of the 320 shoulder operations performed. Variation in practice reflects the fact that there is currently no consensus on an ideal implant. Thomas et al. recommend a low threshold for the use of custom-made uncemented short stemmed prosthesis, as this population of patients are at risk of further elbow surgery and revision surgery and the use of standard prosthesis with cement on limited bone stock can be potentially disastrous. Furthermore 'off the shelf' implants may not fit the skeletally small JIA patient. Regardless of implant choice the authors would advocate preoperative templating to ensure that the smallest sizes fit the patient.

Following success in wrist and hand arthroplasty, pyrocarbon RHA has been postulated as an effective alternative, with potentially improved tribology and reduction in revision rates. However, after 2-year follow-up of a prospective multi-centre study, clinical scores and implant survival remain comparable to hemiarthroplasty and inferior to TSA.[43]

Intraoperative nuances

Operative technical difficulties expressed by Bogoch et al. and Thomas et al. were consistent in the JIA population. First, soft tissue contractures were so severe they had to be released to allow for surgical access. Small, osteoporotic and deformed bone with fragile vascularity, owing to previous decades of medical treatment possibly steroid related, made component fixation and prosthetic sizing difficult. Hence, Thomas et al. recommended intraoperative image intensification. They also noted that the rotator cuff was often intact but attenuated. Uncemented prostheses were most often put in due to a characteristically narrow intramedullary canal, however, this was also technically challenging due to thin cortices.

In all four of our reviewed articles severe preoperative limitations of forward elevation and internal rotation contractions were found. Bogoch also reported joint stiffness which differs from a typical case of RA. Ankylosis of the acromioclavicular joint (with subsequently decreased scapulothoracic motion) was common, with movement inhibition being worse than patients post glenohumeral fusion. Distal joint deformities magnified the upper limb disability.

Both Bogoch et al. and Thomas et al. used a deltopectoral approach. Thomas et al. described their operative technique using a 'beach-chair'position. They describe stepwise capsular release with torque limitation on severely osteoporotic bone, along with further release of the inferior capsule, intra-articular adhesions and the subacromial space. A Z-lengthening procedure was performed for those found to have subscapularis tendon and anterior shoulder capsule fusion. Bogoch et al. found the rotator cuff to be generally intact, though thin. The supraspinatus tendon was present but severely thinned, immobile and apparently nonfunctioning. In all patients, they performed subscapularis tendon and circumferential capsular releases.

Poor quality soft tissues and contracted subscapularis require a 270° release. Z lengthening is appropriate if this release does not reach the footprint in 30° of external rotation. This can be reinforced with a mesh, dermal matrix or pectoralis major transfer.

Bogoch et al. often found a 'dry joint' – no synovitis; 'with the proximal humerus medially migrated under the coracoid process because of glenoid bone loss, medial and anterior, as well as extreme osteoporosis'. In 10 of 13 patients they excised the distal clavicle to increase scapulothoracic motion when spontaneous acromioclavicular ankylosis had occurred. They found that multiple soft tissue releases did not offer any lateralisation of the humerus and furthermore that excision of the distal clavicle in acromioclavicular joint (ACJ) ankylosis patients had no effect on the final outcome either. They also did not advocate acromioplasty due concerns over proximal migration. Ibrahim et al. also do not advocate routine acromioplasty or ACJ excision at the index procedure.

Advanced glenoid bone resorption combined with severe contracture of the capsuloligamentous soft tissues prevented consideration of the glenoid reconstruction with bone grafts and glenoid prosthetic components in most Bogoch et al. cases. This was supported by Thomas et al. who felt that the unpredictable function of the rotator cuff predisposed to early failure of a glenoid prosthetic component, based on evidence suggesting a 40% rate of glenoid loosening in rheumatic arthritis total shoulder replacement.[44,45] Ibrahim et al. suggest routine cross-sectional imaging to determine patients in whom there may be sufficient glenoid bone stock to safely implant a glenoid component.

All our reviewed articles reported extreme osteopenia, cortical thinning and severe bone loss both at the humeral head and the glenoid, resorbed anteriorly and medially to the base of the coracoid. As such the humerus tended to rest in a medial position with a rigid soft tissue envelope due to contractures. Thomas et al. also reported that this caused notching of the proximal humeral shaft.

In the most severe cases, piecemeal excision of the humeral head was performed to allow for safe dislocation. In situ neck cut should be considered with difficult delivery of the humeral head. Using image intensification, stemmed, modular hemiarthroplasty using small biomodular implants (Biomet Merck) were inserted, of which three were custom-made stems coated with hydroxyapatite for cementless fixation. Bogoch et al. used modular humeral components also, with four requiring extra small prosthetic components.

Thomas et al. felt that the bowed metaphysis and proximal diaphysis commonly forced the stem into varus. In addition, they noted that the disparity between the preoperative sagittal and coronal intramedullary diameters were often not fully appreciated but could have a profound effect on stem version. As such, computer tomography image reconstructions as recommended by Bogoch et al. should be advocated in all JIA shoulder arthroplasty cases. This was a problem that Ibrahim et al. mitigated with the use of the RHA and is one of the reasons they cite for using this particular implant in their series. Another perceived benefit they mention is negating the concerns over the 'vacant segment' stress riser between the tips of ipsilateral shoulder and elbow prostheses in patients with inflammatory arthropathies. Fractures in this area are challenging to treat.

Thomas et al. noted in two patients that there were a tear of the long head of biceps on reduction and they were repaired with sutures. Long head of biceps should be kept intact if possible, but if found to be obstructing or torn can be tenodesed to pectoralis major.

Bogoch et al. demonstrated a typical case of JIA shoulder in which they used hemiarthroplasty as limited glenoid bone stock restricted the use of a glenoid component and the periarticular soft tissue contractures meant that small modular prosthetic heads had to be used, even after capsular release. Thomas et al. reported that some shoulders after hemiarthroplasty had stems fixed in a slight varus position. They reported no periprosthetic fractures at or following surgery, no evidence of periprosthetic osteolysis or progressive radiolucency, no signs of loosening or loss of proximal humeral bone stock. They did note that some erosion of the non-surfaced glenoid had occurred in some patients radiographically, but they did not exhibit symptoms.

Postoperative analgesia and rehabilitation

Both Deshmukh et al. and Bogoch did not disclose their post-operative analgesia regimen, whereas Thomas et al. used an interscalene blockade with a catheter for 24–72 h. A Neer-type protocol was used with intervals between phases set according to the model of fixation of the implant and the state of the soft tissue repair. A recent prospective RCT comparing a single bolus interscalene block with liposomal bupivacaine and continuous interscalene nerve block demonstrated liposomal bupivacaine and single bolus block to provide excellent pain relief at much lower cost and risk of complication than continuous nerve block.[46]

Note on outcome scores

The first core outcome set for JIA was reported in 1997 by Giannini et al., which included six overall variables: (1) physician global assessment of overall disease activity, (2) parent or patient global assessment of overall well-being, (3) functional ability, (4) number of joints with active arthritis, (5) number of joints with limited range of motion and (6) erythrocyte sedimentation rate (ESR).[47] More recently, an Outcome Measures in Rheumatology (OMERACT) JIA Core Set Working Group has been formed to update this to include more on patient- and parent-reported outcome measures.[48]

The authors advocate standardisation for the measurement of functional outcomes to allow greater comparison between publications and to compare the differences in implants used and surgical technique. There appears that there is no single functional scoring system that evaluates all outcomes, and as such maybe an agreed set of multiple systems should be used in conjunction with each other.

It is important to be aware of smallest detectable changes (SDCs) when analysing outcome scores. Van Kampen et al. demonstrated the SDC of the simple shoulder test (SST) was 2.8, DASH 16.3, *Quick*DASH 17.1 and OSS 6.0. The minimal important change score for the SST was 2.2, DASH 12.4, *Quick*DASH 13.4 and OSS 6.0.[49] It is additionally important, in patient-reported outcome measures, to be wary of ceiling and floor effects, when 15% or more of the individuals within a sample report the best or the worst level of the score.[50,51]

Conclusion

There are few descriptions of functional outcome or operative technique for JIA shoulder arthroplasty within the literature. Furthermore, patients with JIA have often had procedures performed on other joints prior to their shoulder. We have reviewed four published series between the years of 2004 and 2018.

Among the few papers that have been published on the topic, the operative challenges and principles of surgical management appear consistent.

We have outlined some of the challenges and considerations for shoulder surgeons approaching the patient with JIA.

There appear to be more shoulder arthroplasties done worldwide than that seen at first glance; they are often disclosed as 'others' or under the umbrella term of 'inflammatory arthritis'. Further effort must be made to publish their data separately to allow for better evaluation of surgical and functional outcome in this niche population.

The data suggest that shoulder arthroplasty in end-stage patients with JIA allows for patient satisfaction from pain relief and offers some improvement from functional outcome, with low risk of complication. Longer term data, with larger numbers and prospective controlled trials and agreed upon consistent functional scoring systems may offer better insight to this topic. This review concludes that carefully considered approach to arthroplasty, in close coordination with rheumatologists and anaesthetists, is a valuable treatment option for end-stage severe shoulder JIA.

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