Shoulder and Elbow Surgery in Juvenile Idiopathic Arthritis

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Abstract: Juvenile idiopathic arthritis (JIA) is a chronic inflammatory arthropathy that manifests itself prior to the age of sixteen years with symptoms lasting six weeks or longer. As JIA frequently affects the upper extremities, activities of daily living become compromised during the stages of development when young adults are striving for independence. Symptomatology includes ankylosing, pain and early growth plate closure. Patients with joint involvement prior to growth plate closure have the most destruction in terms of joint abnormality and surgical complexity. Medical management of JIA has allowed for better non-surgical management, yet, there is a continued need to understand the appropriate surgical intervention and order for the greatest functional gains. Comparative studies have shown that varied results as to whether the shoulder replacement should supersede the elbow replacement or should that be reversed or both joint replacements done simultaneously. Our experience found a more significant functional improvement after total elbow replacement due to the unpredictable nature from the shoulder replacement outcomes and an inability for patients to do simple tasks such as bringing a cup to their mouths or handling a toothbrush. The exception to this occurs if the ipsilateral shoulder joint is severely limited to the point that the stressors placed on the elbow due to compensation will lead to early loosening or failure of the elbow joint replacement. Various methods for performing joint replacement of the shoulder and elbow in the JIA population will be discussed. Soft tissue integrity including the functional status of the rotator cuff will be a consideration for which surgical procedure should be considered. Surgical approaches for the elbow present fewer options for improving pain and function in this patient population. Pre, peri and postoperative management is reviewed as careful attention to irregular bony dimensions and dysmorphic anatomy precludes the use of standard implants. Total shoulder and total elbow arthroplasty should be considered in the JIA population where pain and significant functional compromise are present. The order of procedures is dependent on multiple factors and expected outcomes. Educating patients on postoperative expectations over the lifespan is an important part of surgical management for patients with JIA.

Keywords: Juvenile idiopathic arthritis, JIA, Medical management of JIA, Elbow arthroplasty in JIA, Shoulder arthroplasty in JIA, Shoulder, Elbow, Upper extremity, Growth plate closure.

1. INTRODUCTION AND BACKGROUND

Juvenile idiopathic arthritis (JIA), formerly described as juvenile rheumatoid arthritis (JRA) in the United States and juvenile chronic arthritis (JCA) in Europe, is defined as a chronic inflammatory arthropathy with onset before the age of 16, persisting for 6 weeks or longer, and having no other etiologic explanation. Arthropathy has an incidence of approximately 10-15 per 100,000 worldwide [1, 2]. In the United States, recent estimates predict 300,000 children and adolescents are affected by the disease [3]. Upper extremity involvement is common and results in significant pain and debility. Unlike rheumatoid arthritis, JIA is usually associated with loss of motion, ankylosis, and growth abnormalities due to early growth plate closure. The loss of motion can have profound effects on the patient’s ability to function. Fortunately, advances in medical treatment have resulted in decreased surgical intervention in patients with JIA. While the rates of surgical intervention for JIA the surgical management of this patient cohort remains complex [4].

The International League of Associations for Rheumatology (ILAR) has defined 7 subcategories of JIA including: Systemic arthritis, oligoarthritis, polyarthritis with negative rheumatoid factor (RF), polyarthritis with positive RF, psoriatic arthritis, enthesitis related arthritis, and undefined arthritis [5]. In JIA, systemic arthritis is defined as arthritis of one or more joints with a fever of at least 2 weeks’ duration and the presence of evanescent erythematous rash, lymph node enlargement, hepato- or splenomegaly, or serositis. The oligoarthritis subgroup is defined by arthritis affecting 1 to 4 joints in the first 6 months of symptoms. Polyarthritis involves 5 or more joints and is further subdivided into those patients who are RF positive and RF negative. The psoriatic arthritis subgroup is characterized by arthritis with various findings of psoriasis or a first degree relative of psoriasis. Enthesitis related arthritis is characterized by arthritis and enthesitis with...
at least 2 of the following criteria: sacroiliac joint tenderness or inflammation in the lumbosacral spine, human leukocyte antigen B-27 (HLA-B27) antigen positivity, arthritis in a male over 6 years old, acute anterior uveitis, history of ankylosing spondylitis, sacroiliitis with inflammatory bowel disease, Reiter’s syndrome, or acute anterior uveitis in a first-degree relative. Lastly, the undifferentiated arthritis subgroup is considered of those patients who fulfill criteria in no category, or those who meet criteria for 2 or more categories.

While there are many subtypes of 7 subgroups of JIA, for the orthopedic surgeon, understanding the 3 generalized presentations of JIA has clinical merit. There are three simplified presentations; the presence of arthritis with systemic features, oligoarticular arthritis, and polyarticular arthritis. The degree of shoulder pathology in JIA is variable, and correlates to these simplified presentations. Systemically manifested JIA shows a high prevalence of shoulder symptoms, present in up to 80% of patients. [6] Shoulder dysfunction in the polyarticular form is also common, observed in approximately 50% of patients [6]. In the oligoarticular form, there is minimal shoulder involvement. When the shoulder is affected, upwards of 95% of patients will have bilateral dysfunction [6]. Of the polyarticular presentation, RF positive patients typically have more aggressive disease.

Patients that develop involvement of the joints prior to growth plate closure have the most severe abnormalities and deformities. In the shoulder, there is flattening of the humeral head, a dysplastic glenoid cavity, and significantly contracted rotator cuff tendons and associated joint capsule [7, 8]. Often the humeral head erodes the glenoid medially to the level of the coracoid process. The elbow usually presents with a flexion contracture and frequently has range of motion (ROM) under 90°, and sometimes complete ankylosis. The bony abnormalities include thin intramedullary canals with deformity of the proximal ulna and often dislocation of the radial head.

Shoulder or elbow first?

There have been several studies that discuss the treatment of patients with inflammatory arthritis with ipsilateral involvement of both the shoulder and the elbow. Neer et al. advocated that shoulder arthroplasty should be performed first, whereas Friedman and Ewald found better functional improvement if elbow arthroplasty was performed prior to shoulder arthroplasty [9, 10]. Gill et al. found there was no difference between the first stage of shoulder and elbow arthroplasty first. The authors found most functional improvement occurred after the second surgery was performed [11]. Further studies by Rozen and Nagels, and Vrettos et al. reported that simultaneous ipsilateral shoulder and elbow replacement can be performed safely, which is a cost-effective alternative, and may facilitate rehabilitation [12, 13]. Our experience shows that the most functional improvement is from elbow replacement, as shoulder replacement can be unpredictable in patients with inflammatory arthritis due to attenuation or prolonged contracture and mobilization of the rotator cuff tendons. Thus, when the shoulder and elbow are equally painful, we prefer to perform the elbow arthroplasty first. There is a critical exception, however. If both the elbow and shoulder are equally symptomatic, and the patient’s shoulder motion is severely limited, we will perform the shoulder arthroplasty first. The theory behind this management is that if an elbow replacement is performed with an ipsilateral stiff shoulder, the patient will attempt to rotate through the elbow prosthesis, leading to early implant loosening and failure. In this scenario, we prefer to perform the shoulder arthroplasty first, followed by a quickly staged elbow replacement.

2. SHOULDER PROCEDURES FOR JIA

2.1. Shoulder Synovectomy

Synovectomy of the shoulder for JIA patients is rarely performed as most patients have dry synovitis and capsular contractures, and thus would not benefit from the intervention. If performed, the effects are transient with high symptom recurrence. The patient with wet synovitis, inflammation as diagnosed via ultrasound or magnetic resonance imaging (MRI), and mechanical symptoms may benefit from an arthroscopic synovectomy [14]. Further, some authors have suggested the importance of timing of arthroscopic synovectomy. Toledo et al. reported on arthroscopic synovectomy of several joints, including the shoulder. The authors advocated for early intervention in monoarticular or oligoarticular arthritis, after first-line medical therapies had failed. The authors found a sustained response after arthroscopic synovectomy and postulated that the removal of hypertrophic synovium and resultant inflammation led to complete remission in one-third of their cohort. Open synovectomy is not typically performed in this patient population.

2.2. Shoulder Hemiarthroplasty

Patients with JIA often undergo shoulder hemiarthroplasty as opposed to total shoulder arthroplasty due to the unique technical difficulties associated with total shoulder arthroplasty in this population. Patients with JIA have markedly contracted joint capsules, commonly with debilitating internal rotation and adduction limitations. As such, these patients require extensive soft tissue release and subscapularis mobilization for exposure and glenohumeral dislocation. There is often significant erosion of the glenoid, with bone loss to the level of the coracoid process and notching of the humerus from native impingement. In these instances, instrumenting a glenoid component is challenging. The outcomes after shoulder hemiarthroplasty in the JIA patient, however, are favorable. Pain relief is usually obtained but ROM frequently does not improve. A short humeral stem prosthesis or humeral resurfacing is recommended to avoid interference with the stem of an elbow replacement if needed, as well as the creation of a stress riser. Short humeral stems may also abrogate the tendency to place the implant in varus from the bowed metadiaphysis in this patient population. To this end, there is a role for custom-made implants in the patient with severely dysplastic anatomy. Any humeral replacement may still cause erosion of the glenoid and continued pain.
2.3. Stemmed Shoulder Hemiarthroplasty

Thomas et al. reported a series of 9 hemiarthroplasties in 8 patients with systemic or polyarticular JIA. [8] Hemiarthroplasty was performed with small Biomodular implants (Biomet Merck, Dordrecht, Netherlands). Of the 9 patients, 3 required custom implants due to the morphology of the humeral canal. The patients had excellent pain relief and noted improvement in function, but the functional improvement deteriorated over time [8]. The authors of this study utilized preoperative radiographs with a radiopaque marker for implant sizing. The authors, however, recommended that all patients should undergo a preoperative computed tomography (CT) scan for templating, noting that the differences in coronal and sagittal canal diameters are difficult to assess on plain radiographs. Further, the CT may be used to facilitate the creation of custom-made prostheses. The necessity for axial imaging is further underscored by the prevalence of corticosteroid-induced osteoporosis and thin cortical bone to safely house the implant. In the analysis of the rotator cuff, the authors found the rotator tendons intact but attenuated. There was marked capsular contracture, extensive adhesions, and fibrous ankylosis requiring release and prohibiting the placement of a glenoid component without overstuffing the joint. Medial humeral head migration and native glenoid erosion occurred throughout the study. Lastly, the authors recommended the avoidance of cemented implants when possible, given the likelihood of revision with arthroplasty performed at such a young age with expected disease progression.

Jolles et al. reported on the outcomes of 11 shoulder hemiarthroplasties for polyarticular JIA with a mean follow-up of 9 years. Stemmed, modular humeral components were utilized to address the unique pathoanatomy present in JIA. There was no reference to the specific implant system included in the manuscript. The prostheses were uncemented when possible, performed in 73% of patients. Patients reported significant pain reduction measured by visual analogue scale (VAS) scoring, from 8 to 1.3. The authors reported significant gains in postoperative ROM in all planes measured (forward flexion, external rotation and internal rotation). Functional outcome scores were measured by the Disabilities of the Arm, Shoulder and Hand (DASH) score and the Short Form Survey (SF-36) score. Postoperative SF-36 scores remained below societal norms, and DASH scoring averaged 47.2. Despite these modest functional outcomes, the authors noted that this profoundly disabled population reported positive results postoperatively. The authors concurred with Thomas et al. with regard to the technical difficulties associated with the procedure. In particular, implantation of the humeral prosthesis was complicated by the presence of severe osteoporosis, cortical thinning, large areas of bone loss, and the overall diminutive size of the humerus. Life-long contracture and inability to intraoperatively mobilize the soft-tissue envelope was cited as a contraindication to glenoid resurfacing in all but one case. Further, the authors noted ankylosis of the acromioclavicular joint which was treated with distal clavicle excision to increase scapulothoracic motion.

2.4. Shoulder Resurfacing Hemiarthroplasty

Ibrahim et al. reported on 14 stemless, resurfacing hemiarthroplasties utilized for 11 patients with JIA [7]. The authors utilized non-cemented Copeland Mark-3 prostheses (Biomet Orthopedics, Warsaw, IN, US). Two patients required extra small custom-made prostheses to address proximal humeral anatomy. All patients had excellent pain relief, and 6 of the 14 cases had very good or excellent functional outcomes. Range of motion was improved in 11 of the 14 patients, with greatest improvements noted in forward flexion (69° preoperatively vs. 110° postoperatively). Significant gains in internal and external rotation were noted, though only external rotation improvement was clinically significant (12° vs. 32°). Four patients underwent biceps tenodesis and all had intact but attenuated rotator cuffs upon visual inspection. The authors noted the advantage of using the resurfacing prosthesis in avoiding the interaction of a total elbow replacement as well as fewer issues with the proximal humeral fit with abnormal geometry. The resurfacing was noted to be bone preserving, allowing theoretically easier revision when necessary. The humeral resurfacing, however, was noted to have worse outcomes with moderate or severe humeral head erosion. In this scenario, the authors utilized autograft obtained from the distal clavicle and femoral head allograft when there was significant humeral head erosion.

2.5. Total Shoulder Arthroplasty

Of the Jolles et al. cohort, 3 shoulders underwent total shoulder arthroplasty, though only a single patient was available for clinical follow up. The patient had an excellent outcome, with significant pain relief on VAS scoring, and significant improvements in ROM. Postoperative ROM was improved by 90 degrees in forward flexion, 45 degrees in external rotation, and 8 spinal levels of internal rotation. Functional outcome scores remained tempered. These outcomes must be interpreted with the consideration that this patient likely had less disease burden, without severe contractures and bony loss which often precludes glenoid resurfacing.

3. ELBOW PROCEDURES FOR JIA

3.1. Elbow Synovectomy

Elbow synovectomy is infrequently performed in patients with JIA despite having a high incidence of symptoms. Improved medical management, including the use of local glucocorticosteroids, has further decreased the need for surgical intervention. The support for elbow synovectomy for JIA is lent from promising results in wrist and knee synovectomy, or elbow synovectomy outcomes from the adult rheumatoid arthritis population. To the authors’ knowledge, only a single publication exists on elbow synovectomy in patients with JIA. Mäenpää et al. reported on 24 synovectomies performed in 19 patients. [15] Larsen grade was used to evaluate the degree of joint destruction, and most patients had early changes (Larsen grade 0-2), accounting for 96% of patients. These patients underwent open synovectomy via a posterolateral approach. There were no significant changes in ROM postoperatively or functional outcome. The
survival rate defined by need for re-synovectomy or total elbow replacement was 84% at 5 years. Complete pain relief occurred in 44% of patients and the subjective outcome was good to excellent in 72% of patients. The radial head was excised in 4 instances (16.7% of patients). The authors’ view of elbow synovectomy is that it may be useful in patients with bulky, inflammatory synovitis recalcitrant to medical treatments. The procedure should be performed in the patient without considerable joint destruction (Larsen grades 0-2). The decision on whether to perform the procedure as an arthroscopic or open procedure is debatable. The theoretic advantages of arthroscopic synovectomy are less local trauma, faster recovery, and improved ROM at the exchange of a more technically demanding operation in a small joint. If the radial head should be removed, open elbow synovectomy is the best option.

3.2. Interposition Arthroplasty
Interposition arthroplasty may be considered for patients with early arthritis secondary to JIA as an option for improving pain while preserving bone stock. Interposition material includes fascia lata autograft, dermal autograft, Gelfoam (Pfizer, New York, NY, USA) and Achilles tendon autograft. Fernandez-Palazzi et al. reported on 12 elbows that underwent fascial arthroplasty, dermal arthroplasty, and Gelfoam arthroplasty between the ages of 10 and 19 [16]. Only 4 of these were performed for JIA, and they were followed from 6 to 23 years postoperatively. The surgical technique utilized was a semicircular incision from medial to lateral condyle over the ulna. The olecranon was exposed and osteotomized at the joint line. The elbow joint is then lysed, with resection of the distal end of the humerus, proximal ulna and radial head if necessary. The graft was then interposed and sutured to the anterior capsule. Postoperatively, improvement of ROM spanned 60° to 130° in the cohort. Functional results were not reported in this series, but results were graded as excellent in 1 patient, good in 1 patient, and fair in 2 patients. Taken together, interposition arthroplasty may be considered as a temporizing treatment to preserve bone stock in early stage JIA of the elbow.

3.3. Total Elbow Arthroplasty
Total elbow arthroplasty is the mainstay of management in patients with JIA who have late-stage joint destruction and have exhausted nonoperative treatment modalities. Several small series have been published on total elbow arthroplasty in this unique population. Connor and Morrey first reported their experience with 19 patients with JIA, accounting for 24 elbows. Eighteen arthroplasties were performed with semiconstrained Coonrad-Morrey prostheses (Zimmer, Warsaw, IN, USA), and 6 were unconstrained, resurfacing Capitellocondylar prostheses (Johnson and Johnson, New Brunswick, NJ, USA) [17]. The indication for total elbow arthroplasty was pain in 83% of patients, and ankylosis in 17% of patients. Average follow-up was 7.4 years. Pain relief was excellent, with 96% of patients having no or mild pain. Range of motion postoperatively was limited, increasing on average from 63° to 90°. The pain relief and increase in ROM did convey significant improvements in functional outcomes, however. There was a high rate of complications noted in 50% of patients. Early complications included perioperative death, intraoperative olecranon fracture, perioperative subluxation of the prosthesis, avulsion of the extensor mechanism, persistent stiffness, and wound drainage. Late complications included aseptic loosening, instability and worn bushings. Of the 23 elbows that were followed for at least 2 years, 12 had an excellent result, 8 had a good result, and 3 had a poor result.

Connor and Morrey concluded several aspects unique to JIA that complicate total elbow arthroplasty; the humeral and ulnar canal are dysmorphic and often have a completely obliterated intramedullary canal, there is extensive joint destruction, and the elbows have a unique predilection for stiffness as compared to other inflammatory arthropathies. As such, the authors advocated for extensive preoperative templating to ensure the fit of implants. The authors also cautioned that proud implants may change the center of rotation of the elbow joint, thus further contributing to stiffness in this patient population. Lastly, the authors recommended cementation of implants to avoid early aseptic loosening.

The largest cohort of JIA total elbow arthroplasty was reported by Baghdadi et al. [18] Twenty-four patients underwent 29 total elbow arthroplasties for JIA with the semiconstrained Coonrad-Morrey prosthesis, followed for over 10 years. Nearly 50% of the prostheses necessitated modification by shortening, tapering, or bending for passage into the intramedullary canal, or required custom-made implants. All prostheses were cemented, if possible. Eight elbows underwent reoperation, including 6 that underwent implant revision. Seventy-six percent of patients reported a satisfactory functional result, and 62% of elbows were graded as good or excellent. The mean flexion arc improved from 65° to 89°. Survivorship was 96% at 5 years and 79% at 10 years. The authors highlighted the need for implant modification or custom-made implants for the deformities associated with JIA. Specialized tools such as extra-small ulnar and humeral components, cannulated flexible reamers, large plate benders and diamond-tipped burrs can be used for implant modification. They also advocated for 2cm of humeral shortening to improve ROM in the ankylosed elbow.

Several publications have specifically addressed the challenges of complete ankylosis secondary to JIA. Figgie et al. reported on 16 patients with 19 total elbow arthroplasties for complete ankylosis of the elbow [19]. In the study, 8 patients held a diagnosis of JIA. The authors found the arc of motion increased on average from 0° to 71° in the JIA patients. Despite the incomplete restoration of arc of motion, these patients reported significant functional gains. The Hospital for Special Surgery Elbow Score improved on average 62 points, and 80% of patients reported good or excellent outcomes. Mansat and Morrey reported on 2 elbows which were nearly ankylosed from JIA [20]. Both patients saw improved ROM in flexion-extension arcs, from 20° to 95°, and 20° to 105°, respectively. Both were satisfied with the operation and reported one good and one excellent outcome.

Cross et al. reported on the results of custom, noncemented total elbow arthroplasties for young patients with inflammatory arthritides [21]. Fourteen total elbow arthroplasties were performed in 10 patients. Six patients and 10 elbows carried a
diagnosis of JIA. The custom implants were fabricated by Osteonics (Allendale, NJ, USA) to have a porous coating, metaphyseal fit. The mean follow-up was 18 years. Range of motion improved from 50° to 115° postoperatively. Functional outcome scores were significantly improved as measured by the Mayo Elbow Performance score (35 vs. 91). Four patients underwent bushing revision at 8, 8, 22 and 22 years postoperatively. One deep infection was noted requiring implant removal. Remarkably, at final radiographic follow-up, all patients had fully ingrown prostheses with no evidence of loosening or loss of fixation.

Total elbow arthroplasty in the JIA population is technically challenging. Implants must frequently be modified or custom-made to accommodate dysmorphic and stenosed anatomy. The authors recommend preoperative CT scans be performed to check implant fit and custom necessity. Linked prostheses are preferable due to the extensive soft tissue release that is often necessary in the stiff or ankylosed JIA elbow. Improvements in ROM are modest and are superseded by significant functional gains after this intervention. Cementless fixation may be good option in the future for these patients in order to improve durability and long-term outcome.

CONCLUSION

Treatment of the upper extremity in patients with JIA is challenging due to their complex boney deformities and soft-tissue contractures. The age of the patient at time of surgery is also a significant consideration, as patients with JIA undergo arthroplasty in the third and fourth decade of life. Ipsilateral shoulder and elbow involvement are common in JIA. In this cohort, the most painful joint should be addressed first. If the ipsilateral shoulder and elbow are equally symptomatic, we prefer to perform elbow procedures first unless shoulder stiffness would preclude function. Synovectomy is rarely performed in patients with JIA unless they have bulky, wet synovitis that is not responsive to medical treatment. Interposition arthroplasty may be a consideration for young patients with elbow involvement. Shoulder arthroplasty is technically challenging and often, hemiarthroplasty is performed, as the glenoid cannot be resurfaced either due to boney erosion or the contractures of the capsule and subscapularis. Total elbow arthroplasty is also technically challenging, requiring extensive soft tissue release, implant modification or custom implants. Both shoulder and elbow arthroplasty can provide significant pain relief and functional improvement for the JIA population, but ROM gains are marginal.

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CONFLICT OF INTEREST

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