The outcome of total elbow arthroplasty in juvenile idiopathic arthritis (juvenile rheumatoid arthritis) patients

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Background: Elbow prosthetic replacement in patients with juvenile idiopathic arthritis (JIA) can be complicated and technically challenging. Thus, we sought to evaluate the clinical benefit and the prosthetic longevity of primary semiconstrained linked total elbow arthroplasty (TEA) performed to treat these patients.

Methods: Between 1983 and 2005, 29 elbows in 24 patients (20 women and 4 men) had been replaced because of JIA. The mean age was 37 years (range, 24-68 years). Because of underlying deformity, the implant contour was modified for 9 elbows (31%) and a customized implant was inserted in 5 elbows (17%). The mean follow-up duration was 10.5 years (range, 4.6-20.1 years).

Results: During the follow-up period, 8 elbows underwent reoperation, including 6 (21%) that underwent implant revision. At most recent follow-up, 22 elbows (76%) subjectively had a satisfactory overall functional result. The mean Mayo Elbow Performance Score was 78 points (range, 50-100 points), with 18 elbows graded as having an excellent or good result. Compared with preoperative range of motion, the mean extension-flexion arc improved from 65° ± 44° to 89° ± 35° (P = .01), mean flexion improved from 113° ± 23° to 126° ± 26° (P = .02), and mean extension improved from 48° ± 25° to 37° ± 26° (P = .08). By use of the Kaplan-Meier survivorship method, the rate of TEA survival from any revision was 96.4% (95% confidence interval, 89.8%-100%) and 79.9% (95% confidence interval, 65.1%-97.5%) at 5 years and 10 years, respectively.

Conclusion: Primary TEA for JIA patients is technically challenging and frequently requires implant modification or custom designs. These patients might have high complication and revision rates. However, most benefit from the intervention for a long term.

Level of evidence: Level IV, Case Series, Treatment Study.

Keywords: Juvenile idiopathic arthritis; total elbow arthroplasty; semiconstrained linked design; outcomes

This study was approved by the institutional review board at Mayo Clinic (study title, "Total Elbow Arthroplasty in Juvenile Rheumatoid Arthritis"; IRB No. 07-007468).

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least 6 weeks in the absence of a known etiology. These patterns include systemic arthritis, oligoarthritis (≤4 joints), and polyarthritis (>4 joints). The aggressive nature of the disease with multiple-joint involvements and soft-tissue damage may necessitate joint replacement during the early decades of a patient’s life. The elbow might be involved with the disease, and the damage can be extensive. Patients typically complain of persistent pain and progressive loss of motion. Total elbow arthroplasty (TEA) in these patients can be technically difficult and challenging because of unusual deformity, small anatomy, a narrowed medullary canal, severe stiffness, and soft-tissue contracture. TEA is not commonly performed for arthritis associated with JIA. It is usually indicated when the medical treatment fails to halt the disease progression and the patient has considerable pain and functional disability. Connor and Morrey reported on 24 elbows with JIA that underwent TEA with the use of linked and unlinked designs. To our knowledge, no other series in the English-language literature have described the outcome of TEA in JIA patients with a special emphasis on the linked design.

The purpose of this study was to determine the clinical results and implant longevity in patients who were treated with a primary semiconstrained linked TEA for end-stage JIA.

Materials and methods

We queried the joint registry at the Mayo Clinic and identified a total of 52 elbows that underwent TEA for end-stage JIA with the use of semiconstrained linked prostheses. We included in our study patients who underwent primary replacement and patients who had an established JIA diagnosis in their medical records. The diagnosis of the JIA had been made according to the formerly described principles. After we applied our inclusion and exclusion criteria, 29 elbows in 24 patients constituted our study cohort. A group of these patients have been reported on in the literature as part of other series for different research purposes. However, each of these patients was independently reassessed to specifically determine the clinical benefit and the survival rate in patients who underwent primary TEA using the linked design for end-stage JIA.

Patient characteristics, the presence of pain, and range of motion were obtained through patients’ records before and after the arthroplasty. The operative technique (implant design, fixation technique, and implant modification) and intraoperative range-of-motion assessment were recorded. Additional operations including revision with component exchange were also reviewed.

At our institution, the follow-up was carried out at regular intervals. It was performed twice during the first year, at 2 years, at 5 years, and then every 5 years. Patients had been requested to complete a standardized elbow questionnaire and have radiographs obtained. The questionnaire included components of the standardized Mayo Elbow Performance Scoring system. The Mayo Elbow Performance Score (MEPS) was calculated at the most recent clinical evaluation using the questionnaire. The results were graded as excellent, good, fair, or poor according to this system.

Radiographs were assessed before the replacement and graded regarding the severity of joint destruction as follows: grade I, no radiographic changes except periarticular osteopenia; grade II, narrowing of the joint space with intact joint architecture; grade III, joint architecture alteration, whether mild (A) or moderate (B); grade IV, gross destruction of the joint; and grade V, ankylosis.

Radiographs were also evaluated immediately after the replacement for the cementing technique, which was considered adequate, marginal, or inadequate. The cementation was considered adequate when the radiolucent zone at the bone-cement interface was less than 1 mm wide and the cement extended past the tip of the prosthesis. It was considered marginal when the radiolucent zone at the bone-cement interface was 2 mm wide and the cement extended past the tip of the prosthesis or when the radiolucent zone at the bone-cement interface was less than 2 mm wide and the cement did not extend past the tip of the prosthesis. It was considered inadequate when the radiolucent zone at the bone-cement interface was 2 mm wide or greater and the cement did not extend past the tip of the prosthesis.

At most recent follow-up, anteroposterior radiographs were evaluated for component loosening and bushing wear. The component loosening was evaluated by comparing the immediate postoperative radiographs with the latest radiographs. The progressive radiolucency was graded as follows: none; type I, a radiolucent line less than 1 mm wide that involved less than 50% of the bone-cement interface; type II, a radiolucent line at least 1 mm wide that involved less than 50% of the bone-cement interface; type III, a radiolucent line more than 1 mm wide that involved at least 50% of the bone-cement interface; type IV, a radiolucent line more than 2 mm wide around the entire bone-cement interface; or type V, a radiolucent line more than 2 mm wide around the entire bone-cement interface with shifting of the component.

The bushing wear was evaluated on the most recent anteroposterior radiograph. Two intersecting lines were drawn: One was parallel to the yoke of the humeral component, and one was parallel to the medial or lateral articular surface of the ulnar component. An angle of intersection greater than 7° indicates changes in the bushings due to wear or plastic deformation. An angle greater than or equal to 10° indicates mild to moderate bushing wear.

Operative technique

The size and shape, as well as other anatomic deformities, make TEA a challenging procedure in patients with JIA. Preoperative planning is imperative. The surgeon should consider having specialized tools available, such as extra-small ulnar and humeral components, small cannulated flexible reamers, and large plate benders and diamond-tipped burs for implant modification. The humeral and ulnar intramedullary canals are characteristically narrow and may be obliterated by cortical bone especially the ulna (Fig. 1). The canal is carefully identified with a high-speed burr and entered using a flexible guide pin. If the canal is obliterated, a new canal must be created using a high-speed end-cutting burr. The canals are then sequentially reamed using small, flexible reamers placed over the guide pins. In patients with severe soft-tissue contracture or bony ankylosis, extra steps are required to gain adequate exposure and restore a reasonable range of motion. If there is bony ankylosis of the joint, a small osteotome or microsagittal saw may be needed to create a joint space in which to work (Fig. 2). Often, a plane of normal tissue can be identified either anterior or posterior, and then, the joint line is re-created using this as the starting point. If severe soft-tissue contracture
is present, complete anterior and posterior capsular releases are required. It is also necessary to release the common flexor and extensor attachments to the epicondyles. The humerus, but not the ulna, can be shortened to gain additional motion intraoperatively.\textsuperscript{16} The radial head can be resected to gain flexion if it impinges on the anterior humerus or flange of the humeral component. Appropriately sized trial components are carefully placed in the humeral and ulnar canals. If the trial components do not fit within the canal, the real, appropriately sized implants can be taken and modified in a stepwise manner. The components may require shortening, tapering of the tips, or bending to accommodate the deformity of their respective intramedullary canals. Once the joint is carefully reduced, it is taken through a gentle arc of motion. We typically evaluate the flexion and extension motion attained with gravity, in addition to gentle pressure.

**Operative details**

All patients received semiconstrained linked Coonrad-Morrey prostheses (Zimmer, Warsaw, IN, USA). Implants of different versions, sizes, and humeral component surface finishes\textsuperscript{11} were used (Appendix). Four surgeons performed all procedures. One of the authors (B.F.M.) performed most of them (25 elbows). The mean operative time was 148 ± 54 minutes. The ulnar nerve was transposed in all elbows except 1 with gross deformity. The insertion of modified components was required in 9 elbows (31%) (7 ulnar and 3 humeral components). These were modified by shortening, tapering of the tips, or bending to allow passage down into the narrowed medullary canal. In addition, 5 elbows (17%) required the insertion of customized components (5 ulnar and 1 humeral component). The cementing fixation\textsuperscript{15} was adequate in 24 elbows (of 28 elbows) for the humeral component and 28 elbows (of 28 elbows) for the ulnar component. The surgeon elected to use uncemented fixation in 1 elbow with satisfactory press-fit fixation because of narrowed ulnar and humeral canals that could have resulted in bone fracture during preparation for cemented implants and instrumentation. In addition to the instrumentation, several procedures were performed, including radial head resection (8 elbows), coronoid osteotomy (2 elbows), and open reduction and internal fixation of the humerus (1 elbow) and the olecranon (1 elbow).

**Patient characteristics**

Twenty-nine elbows in 24 patients underwent primary TEA for end-stage JIA between 1983 and 2005. The dominant arm was affected in 13 patients (54%). The disease involvement was polyarticular in 17 patients (71%) and oligoarticular in 7 (29%). In 20 patients (24 elbows), other joints were involved in the operative arm (Appendix). Additional joints in the lower extremities were also affected by the disease in 12 patients (16 elbows). The use of an assistive device to perform activities of daily living was reported by 6 patients (10 elbows). The mean age at surgery was 37 years (range, 24-68 years). Surgery was performed in 24 elbows (83%) in 20 women and 5 elbows (17%) in 4 men. The patients mainly underwent the replacement to relieve their pain and improve their range of motion. In addition, 6 patients (7 elbows) had bony or fibrous ankylosis, 4 patients (4 elbows) had traumatic fractures, and 3 patients (4 elbows) had instability. Six patients (seven elbows) underwent a surgical intervention before the replacement (Appendix). The follow-up period was estimated from the replacement until the latest evaluation if the implant was not revised or until revision surgery with component exchange. The mean follow-up duration in this cohort was 10.5 years (range, 4.6-20.1 years). No patients in our study cohort were completely lost to follow-up.

**Statistical analysis**

The data are reported as mean ± standard deviation for continuous variables and count (percentage) for discrete variables unless otherwise specified. Values of n indicate the number of elbows. The baseline (preoperative) range-of-motion measurements were compared with their corresponding values measured at last follow-up using paired t tests. Flexion, extension, and the total arc of motion were compared among the baseline (preoperative), intraoperative, and last follow-up time points by use of 1-factor repeated-measures analysis of variance. Significant differences were analyzed further with the Ryan-Einot-Gabriel-
Analyses were performed with SAS software, version 9.2 (SAS Institute, Cary, NC, USA).

Results

Clinical outcomes

The MEPS results for all patients at a mean follow-up of 10.5 years (range, 4.6-20.1 years) were excellent in 8 elbows, good in 10, fair in 7, and poor in 4. The mean score was 78 points (range, 50-100 points). Most patients (18 elbows) sought the intervention because of pain and limited motion. The mean MEPS for these 18 elbows was 84 points (range, 50-100 points) (Table I). After the initial procedure, 6 elbows (21%) underwent a revision with component exchange (Appendix). No patient underwent resection arthroplasty with component removal. Of the 23 elbows that did not undergo a revision, the MEPS results at a mean follow-up of 11.3 years (range, 4.6-20.1 years) were excellent in 8 elbows, good in 10, and fair in 5. The mean score was 83 points (range, 60-100 points). Their results subjectively had been rated as satisfactory in 22 elbows (76%) and unsatisfactory in 1. Overall, all patients had a prosthetic elbow in place after management of all failures and all elbows except 1 (97%) had satisfactory results with the intervention at a mean clinical follow-up of 12.1 years (range, 4.6-20.2 years).

When we comparing the most recent range-of-motion assessment with the preoperative assessment, the mean extension-flexion arc improved from 65° ± 44° to 89° ± 35° postoperatively (P = .01), mean flexion improved from 113° ± 23° to 126° ± 26° postoperatively (P = .02), and mean extension improved from 48° ± 25° to 37° ± 26° postoperatively (P = .08) (Table II). In a subset of 7 elbows with ankylosis or near ankylosis before the replacement (extension-flexion arc ≤20°), the mean extension-flexion arc improved from 9° ± 9° to 67° ± 46° postoperatively, mean flexion improved from 86° ± 11° to 116° ± 39° postoperatively, and mean extension improved from 77° ± 11° to 49° ± 24° postoperatively. In all patients, most of the improvement occurred early after the replacement. The intraoperative mean extension-flexion arc was 121° ± 22°, from 13° ± 15° of extension to 133° ± 17° of flexion (n = 25). Compared with the intraoperative range-of-motion evaluation, the most recent extension-flexion arc lost a mean of 30° ± 35° (P < .001), extension lost a mean of 23° ± 30° (P < .001), and flexion lost a mean of 7° ± 24° (P = .15).

Reoperations and complications

A complication that required an additional operation occurred in 8 elbows in 7 patients. Of these, 6 elbows (21%) in 5 patients underwent a revision with component exchange (Appendix). No patient underwent resection arthroplasty, and all had a prosthetic elbow in place after the revision surgery. The rate of survival from any reoperation was 93% (95% confidence interval [CI], 84%-100%) and 72.6% (95% CI, 56.5%-92.5%) at 5 years and 10 years, respectively. The rate of survival from any revision with component exchange was 96.4% (95% CI, 89.8%-100%) and 79.9% (95% CI, 65.1%-97.5%) at 5 years and 10 years, respectively.

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**Table I** Outcomes based on presentation and patient needs before intervention

<table>
<thead>
<tr>
<th></th>
<th>Pain and limited motion</th>
<th>Ankylosis and severe contracture</th>
<th>Pain and gross instability</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of elbows</td>
<td>18</td>
<td>7</td>
<td>4</td>
</tr>
<tr>
<td>Revision No. of elbows</td>
<td>2</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>MEPS grade No. of elbows</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Excellent</td>
<td>7</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Good</td>
<td>8</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Fair</td>
<td>2</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>Poor</td>
<td>1</td>
<td>1</td>
<td>2</td>
</tr>
</tbody>
</table>

*Includes patients who had bony ankylosis or fibrous ankylosis with extension-flexion arc less than 20°.

†Includes any revision with component exchange.

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**Table II** Range-of-motion details

<table>
<thead>
<tr>
<th>Motion</th>
<th>Preoperative (°)</th>
<th>Latest follow-up (°)</th>
<th>Change (°)</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Extension (N = 28)</td>
<td>48 (25)</td>
<td>37 (26)</td>
<td>−11 (31)</td>
<td>.08</td>
</tr>
<tr>
<td>Flexion (N = 28)</td>
<td>113 (23)</td>
<td>126 (26)</td>
<td>12 (28)</td>
<td>.02</td>
</tr>
<tr>
<td>Extension-flexion arc (N = 28)</td>
<td>65 (44)</td>
<td>89 (35)</td>
<td>24 (46)</td>
<td>.01</td>
</tr>
<tr>
<td>Pronation (N = 22)</td>
<td>40 (32)</td>
<td>69 (27)</td>
<td>29 (41)</td>
<td>.003</td>
</tr>
<tr>
<td>Supination (N = 22)</td>
<td>29 (33)</td>
<td>40 (33)</td>
<td>11 (47)</td>
<td>.29</td>
</tr>
<tr>
<td>Pronation-supination arc (N = 22)</td>
<td>69 (55)</td>
<td>109 (49)</td>
<td>40 (73)</td>
<td>.02</td>
</tr>
</tbody>
</table>

Data are expressed as mean (standard deviation).
The chronicity nature of the disease with arthritis and gross joint damage could affect the quality of bone stock hosting the prosthesis. However, no detected difference was observed in rates of survivorship from revision at 5 years and 10 years between the groups based on the radiographic grading before the replacement \( P = .45 \). This could be because of the small number of events in our series (Table IV).

### Radiographic outcomes

Radiographs were available at a minimum of 2 years after surgery (mean, 10.3 years; range, 2.3-20.1 years) for 23 elbows (79%). For the remaining 6 elbows, a completed standardized elbow questionnaire was available but the patients did not provide radiographs at their latest evaluation. Radiographs of these elbows were obtained between a few days and 1.1 year after surgery. The mean clinical follow-up period with completed questionnaires for these 6 elbows was 8.3 years (range, 4.6-13.8 years). We did completely fail to contact any of the patients in our study during follow-up.

Of the 23 elbows with radiographs at a minimum of 2 years, 15 showed absent lucency or a minor lucency grade (none, type I, or type II) for both components and 5 for either the ulnar (1 elbow) or humeral (4 elbows) component (Fig. 3). Three elbows had major lucency (type IV or V) around the ulnar component and one around the humeral component. These required revision with component exchange. The radiographs of the 23 elbows were also evaluated for bushing wear. Sixteen elbows did not show evidence of wear with an angle of intersection less than or equal to 7° (Appendix). Three elbows had mild to moderate wear changes for the bushings with an angle of 10° or greater at the yoke. Of the 3 elbows, 2 had undergone revision including 1 in which isolated bushing exchange was performed.

### Discussion

JIA is a debilitating multifactorial disease with a prevalence between 16 and 150 per 100,000 in developed countries. The course of the disease is variable, and patients may still experience the active phase into adulthood. The treatment of the condition essentially is medical but eventually progresses to surgical intervention. The young age of patients at presentation is a concern because of the lack of reliable options to the replacement. In our series, 7 elbows underwent surgical intervention before the replacement that did not correlate with implant revision. Another concern in this group is related to the revision rate and prosthesis durability. Celli and Morrey reported TEA results in patients aged 40 years or younger: 21 TEAs were carried out in JIA patients with a mean follow-up period of 91 months. In 2 of the 21 elbows (10%), an additional operation for triceps insufficiency was performed. The MEPS outcomes were excellent in 67% (14 of 21 elbows), good in 29% (6 of 21 elbows), and fair in 5% (1 of 21 elbows). The mean score was 91 points.

Several characteristics of JIA patients are important for the surgeon to consider. The systemic effects of the disease often lead to small bone architecture, severe joint contracture, and/or ankylosis. Peden and Morrey stated that...
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Connor and Morrey reported outcomes of 24 TEAs (linked and unlinked designs) in JIA patients. Of the 24 elbows, 12 (50%) had 13 complications, including 1 perioperative death and 3 major component exchanges. For the 23 elbows with a mean follow-up period of 7.4 years (range, 2-14 years), the MEPS outcomes were excellent in 52% (12 of 23 elbows), good in 35% (8 of 23 elbows), and poor in 13% (3 of 23 elbows). The mean score was 90 points. The mean postoperative extension-flexion arc gained 27° compared with the preoperative assessment. The mean postoperative extension-flexion arc was 90°, from 35° of extension to 125° of flexion. Dennis et al reported the results of 21 TEAs (capitello-condylar prostheses) at a mean follow-up of 57 months (range, 13-104 months). TEA was carried out in a subset of 6 elbows in JIA patients. On the basis of the scoring system designed by Inglis and Pellicci, the mean score was 91.7 points after the replacement (total of 100 points). Dennis et al reported minor improvement in motion excluding extension. Motion improved by 6.7° for flexion, 17.8° for extension, 3.3° for pronation, and 8.3° for supination.

A number of weaknesses are present in our study. The foremost include the retrospective nature of the study design, small sample size or number of patients in our study, and lack of complete data in some patients. We describe patient outcomes using the standardized MEPS, and this does not account for the extent of disease severity and distribution. However, a major component of the MEPS does assess daily function. Thus, this may not be sufficient to provide complete details about the value of the intervention. We also report the implant longevity and its survival from revision as an endpoint. The 10-year survivorship rate and its corresponding wide CI are in part due to the small number of patients at risk at the 10-year time point. Lastly, we do not report patient activity levels before and after the procedure. The strength of our study is the use of a single design and the midterm to long-term follow-up with an accepted assessment tool, the MEPS, and a standard measure of survival.

Conclusions

Patients with end-stage JIA undergoing primary total elbow replacement with the use of the linked design have substantial improvement in satisfaction and function. The arthritis and the joint architecture pose considerable technical challenges. Thus, careful preoperative planning and execution, especially of canal preparation, is essential. Complications can be problematic and may be encountered. Nevertheless, satisfactory clinical outcomes and implant durability also can be obtained. More than 75% of these patients have considerable and sustained functional gains.

Disclaimer

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Supplementary data

Supplementary data associated with this article can be found, in the online version, at http://dx.doi.org/10.1016/j.jse.2014.03.012

References


