Shoulder hemiarthroplasty in patients with juvenile idiopathic arthritis

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Replacement of the shoulder in juvenile idiopathic arthritis is not often performed and there have been no published series to date.

We present nine glenohumeral hemiarthroplasties in eight patients with systemic or polyarticular juvenile idiopathic arthritis. The mean follow-up was six years (59 to 89 months). The mean age at the time of surgery was 32 years. Surgery took place at a mean of 27 years after diagnosis. The results indicated excellent relief from pain. There was restoration of useful function which deteriorated with time, in part because of progression of the systemic disease in this severely affected group. No patient has required revision to date and there has been no radiological evidence of loosening or osteolysis around the implants.

We discuss the pathoanatomical challenges unique to this group. There was very little space for a prosthetic joint and, in some cases, bony deformity and the small size necessitated the use of custom-made implants.

Arthritis of the shoulder is rarely an early feature of juvenile idiopathic arthritis. Involvement of the hip and knee is more common and can be treated by arthroplasty.1-8 That of the shoulder is seen later in the course of ongoing systemic or polyarticular juvenile idiopathic arthritis with an incidence of 15% at 15 years from the onset of the disease.9

Persistent arthritis of the immature shoulder produces a maldeveloped proximal humerus and glenoid cavity (Fig. 1). Later in the course of the disease, erosion of bone and cartilage may cause medial migration and superior subluxation of the humeral head. Consequent dysfunction of the shoulder impairs basic daily activities such as toileting and the use of crutches or a stick, which may be required during rehabilitation after surgery on the hip or knee. If the elbows become involved, the function of the upper limb deteriorates further.

There are no published reports on glenohumeral arthroplasty in juvenile idiopathic arthritis in the current literature. In a report of total elbow arthroplasty in adults with juvenile idiopathic arthritis, Connor and Morrey10 highlighted the challenges imposed by the small size and variable shape of the distal humerus. They referred to the follow-up data at 12 years of Levene et al for shoulder arthroplasty in juvenile idiopathic arthritis, but this work has remained unpublished.

We have encountered unique difficulties in this group of patients which we report here and discuss potential strategies to accommodate the pathological anatomy.

Patients and Methods

Since 1995, we have replaced 15 consecutive shoulders in 13 patients with juvenile idiopathic arthritis as part of a prospective interventional cohort study. All the patients were under the joint care of a paediatric and adult rheumatology service as part of a multidisciplinary team including occupational and physiotherapists. There was no control group. In six arthroplasties, follow-up was less than two years and they are not reported here.

The criteria for inclusion in our study were patients with juvenile idiopathic arthritis undergoing replacement of the shoulder for arthritis at our institution. Those with adult-onset rheumatoid arthritis were not included. There were no cases of previous fracture, avascular necrosis or joint sepsis.

The indications for arthroplasty were pain and loss of function with end-stage destruction of the joint. Only severely affected patients, in whom medical management and physiotherapy had failed to control the symptoms, underwent surgery. Before 1995, such patients were generally managed non-operatively at our institution. One patient (case 2) had previously...
had double osteotomies of the glenoid and humeral neck, a procedure described by Benjamin.\textsuperscript{11}

There were eight patients (seven women, one man) in the series with nine hemiarthroplasties of the shoulder. All had been diagnosed with juvenile idiopathic arthritis before the age of ten years but were adults at the time of surgery (Table I). All are surviving and have systemic or polyarticular disease. They had undergone many operations on other joints in addition to their shoulder. All were noted to have involvement of the ipsilateral elbow at the time of surgery and some have subsequently undergone replacement of the elbow.

All the patients were assessed pre-operatively by one of the authors (AJP) with evaluation by the score of Constant and Murley.\textsuperscript{12} This is a validated 100-point shoulder assessment with variables of pain (15 points), activities of daily living (20), range of movement (40) and power (25). We reviewed each patient at one year after operation (AJP) and then twice a year when possible (AJP, ST, RAS), although some patients subsequently left the region and therefore were seen at less frequent intervals. Of the six patients with unilateral operations available for evaluation at the end of the current period of study, four had considerable arthritic involvement of the unreplaced joint which we scored as a crude form of control.

Plain anteroposterior, axillary and scapular Y-view lateral radiographs were taken pre-operatively with a radio-
opaque measurement disc of known diameter (Fig. 2). These films were sent to the manufacturer of the implant (Biomet Merck, Dordrecht, The Netherlands) and analysed against the geometry of a standard stem to establish whether and which standard prosthesis would be appropriate. In three cases, custom-made prostheses were designed from these films since standard implants were too large or otherwise inappropriate to fit the proximal humeral canal (Table II).

All the operations were performed by the senior author (MT) under general anaesthesia with an interscalene block. Because of cervical immobility or potential instability the use of endoscopic intubation was obligatory in some cases.

Anteroposterior and lateral films were taken post-operatively to assess the position of the stem and to exclude intra-operative fracture. These were repeated with an axillary view at each subsequent follow-up, determining specifically osteolysis, glenoid erosion and evidence of gross loosening. These films were reviewed by two of the authors (ST, RAS) independently of the operating surgeon.

Operative technique. A standard deltopectoral approach was used with the patient in a ‘beach-chair’ position. Intra-operative image intensification was used to visualise the proximal humerus as an aid to positioning the prosthesis and to reduce the risk of breaching the weak cortical bone.

All the patients underwent stemmed, modular hemiarthroplasty using small Biomodular implants (Biomet Merck). Three custom-made stems were coated with hydroxyapatite for cementless fixation. In four cases a standard hydroxyapatite-coated 7-mm stem was used without cement. In the two remaining cases, a standard, non-coated 6-mm stem was used which required fixation by cement. In all but one case the smallest available head diameter was used (Table II) with short neck lengths because of soft-tissue contracture.

Post-operatively a standard sling was applied with interscalene blockade by catheter for 24 to 72 hours. A Neer-type protocol was instituted for rehabilitation with intervals between phases set according to the model of fixation of the implant and the state of the soft-tissue repair.

Results

The mean follow-up was six years (59 to 89 months). One patient (case 1) was lost to follow-up after she moved away. She had good function when last reviewed at 35 months.

The Constant score increased in every patient at final follow-up (Tables III and IV). The scores of the four contralateral ‘control’ shoulders are

Table II. Operative details in the nine shoulders

<table>
<thead>
<tr>
<th>Case</th>
<th>Side</th>
<th>Date of surgery</th>
<th>Prosthesis</th>
<th>Cement</th>
<th>Cufft</th>
<th>Operative difficulties</th>
<th>Complications</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>L</td>
<td>14 Aug 96</td>
<td>Standard 6 mm stem 40/20 mm head</td>
<td>Y</td>
<td>N</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>2</td>
<td>R</td>
<td>05 Oct 95</td>
<td>Custom-made HA-coated 5 mm stem 40/15 mm head</td>
<td>N</td>
<td>S</td>
<td>Tear of long head of biceps on reduction, sutured</td>
<td>None</td>
</tr>
<tr>
<td>3</td>
<td>L</td>
<td>07 May 97</td>
<td>Standard 6 mm stem 40/15 mm head</td>
<td>Y</td>
<td>N</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>4</td>
<td>R</td>
<td>02 Nov 95</td>
<td>Standard HA-coated 7 mm stem 40/15 mm head</td>
<td>N</td>
<td>N</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>5</td>
<td>R</td>
<td>09 Oct 96</td>
<td>Custom-made HA-coated 5 mm stem 40/15 mm head</td>
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<td>N</td>
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</tr>
<tr>
<td>6</td>
<td>R</td>
<td>25 Nov 97</td>
<td>Standard HA-coated 7 mm stem 40/15 mm head</td>
<td>N</td>
<td>S</td>
<td>None</td>
<td>Brachial pain</td>
</tr>
<tr>
<td>7</td>
<td>L</td>
<td>25 Feb 98</td>
<td>Standard HA-coated 7 mm stem 40/15 mm head</td>
<td>N</td>
<td>M</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>8</td>
<td>R</td>
<td>18 Mar 98</td>
<td>Standard HA-coated 7 mm stem 44/17 mm head</td>
<td>N</td>
<td>N</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>9</td>
<td>R</td>
<td>02 Feb 99</td>
<td>Custom HA-coated 5 mm stem 40/15 mm N head</td>
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<td>None</td>
<td>None</td>
<td>None</td>
</tr>
</tbody>
</table>

* L, left; R, right
† M, massive tear (> 5 cm); N, normal; S, small tear (< 3 cm)
‡ ACJ, acromioclavicular joint

Table III. Details of the Constant scores

<table>
<thead>
<tr>
<th>Case</th>
<th>Pain</th>
<th>Function</th>
<th>Movement</th>
<th>Power</th>
<th>Total</th>
<th>Control‡</th>
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<tr>
<td>1</td>
<td>Pre-op†</td>
<td>0</td>
<td>0</td>
<td>8</td>
<td>0</td>
<td>8</td>
</tr>
<tr>
<td>2</td>
<td>35 mths FU†</td>
<td>15</td>
<td>10</td>
<td>42</td>
<td>15</td>
<td>82</td>
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<tr>
<td>3</td>
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<td>0</td>
<td>0</td>
<td>6</td>
<td>0</td>
<td>6</td>
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<tr>
<td>4</td>
<td>89 mths FU</td>
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<td>5</td>
<td>32</td>
<td>9</td>
<td>61</td>
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<tr>
<td>5</td>
<td>Pre-op</td>
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<td>0</td>
<td>10</td>
<td>0</td>
<td>10</td>
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<tr>
<td>6</td>
<td>88 mths FU</td>
<td>5</td>
<td>5</td>
<td>19</td>
<td>3</td>
<td>32</td>
</tr>
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<td>Pre-op</td>
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<td>0</td>
<td>12</td>
<td>0</td>
<td>12</td>
</tr>
<tr>
<td>8</td>
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<td>12</td>
<td>5</td>
<td>24</td>
<td>5</td>
<td>46</td>
</tr>
<tr>
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<td>Pre-op</td>
<td>0</td>
<td>0</td>
<td>14</td>
<td>0</td>
<td>14</td>
</tr>
<tr>
<td>10</td>
<td>44 mths FU</td>
<td>10</td>
<td>6</td>
<td>17</td>
<td>8</td>
<td>41</td>
</tr>
<tr>
<td>11</td>
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<td>0</td>
<td>0</td>
<td>14</td>
<td>0</td>
<td>14</td>
</tr>
<tr>
<td>12</td>
<td>66 mths FU</td>
<td>9</td>
<td>7</td>
<td>10</td>
<td>4</td>
<td>30</td>
</tr>
<tr>
<td>13</td>
<td>Pre-op</td>
<td>5</td>
<td>4</td>
<td>8</td>
<td>0</td>
<td>17</td>
</tr>
<tr>
<td>14</td>
<td>60 mths FU</td>
<td>10</td>
<td>8</td>
<td>28</td>
<td>7</td>
<td>53</td>
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<tr>
<td>15</td>
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<td>0</td>
<td>0</td>
<td>14</td>
<td>0</td>
<td>14</td>
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<tr>
<td>16</td>
<td>59 mths FU</td>
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<td>6</td>
<td>22</td>
<td>10</td>
<td>53</td>
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<tr>
<td>17</td>
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<td>0</td>
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<td>4</td>
<td>0</td>
<td>4</td>
</tr>
<tr>
<td>18</td>
<td>44 mths FU</td>
<td>14</td>
<td>12</td>
<td>20</td>
<td>7</td>
<td>53</td>
</tr>
</tbody>
</table>

* lost to follow-up at 35 months
† pre-op, pre-operative; FU, final follow-up
‡ score for contralateral non-operated but involved shoulder
also shown in Table III. The higher scores in the operated shoulders were mainly due to better relief from pain. No patient has required revision to date and there has been no case of deep infection.

**Radiological review.** Figure 3 shows the position of the standard and custom-made implants. In some patients the stem was positioned in slight varus (Fig. 3b). There was no case of periprosthetic fracture at or following surgery. In no patient was there radiological evidence of periprosthetic osteolysis or progressive radiolucency. No other signs of loosening have been seen to date and the bone stock of the proximal humerus has remained well preserved as on the post-operative radiographs. On clinical grounds no additional imaging other than plain radiography has been indicated.

Some erosion of the non-resurfaced glenoid was evident in a few patients, but this was not symptomatic. Progressive, superior migration of the implant has not been encountered to date.

**Complications.** One patient (case 5) developed symptoms of arthritis of the acromioclavicular joint, which was held to be exceptional in this group of patients, some nine months after operation. She underwent open excision of the acromioclavicular joint with excellent early relief of symptoms.

Another (case 6) complained of persistent paraesthesiae and pain affecting the lateral aspect of her forearm which did not respond to simple analgesia and physiotherapy. She remains unable to self-toilet effectively because of a poor range of movement and her function has deteriorated with time. Peripheral nerve-conduction studies were comparable with those of the contralateral arm and within normal limits. MRI of her cervical spine showed degenerative changes consistent with a C6 radiculopathy, but she has declined further intervention. There have been no other significant complications to date.

**Discussion**

This is a small series of patients with no unoperated control group other than the contralateral shoulder of four patients with significant arthritic involvement. Our methodology is otherwise reasonable. Given the paucity of published information on shoulder hemiarthroplasty in patients with juvenile idiopathic arthritis, we have performed this procedure as part of a prospective study using the same type of prosthesis and validated scoring methods. Assessment was performed by observers independent of the operating surgeon.

The results suggest that hemiarthroplasty of the shoulder may be effective in patients with severe, end-stage juvenile idiopathic arthritis of the shoulder. Large improvements in the Constant score after surgery indicate good relief from pain and restoration of useful function. There was deterioration of function with time, attributable in part to the progression of the disease in other joints and organ systems. One patient (case 7), for example, underwent ipsilateral replacement of the elbow between the first and second follow-up with an increase in her Constant score of more than 20 points during this period. This increase was largely in the functional components of the score which are affected by the function of the ipsilateral elbow. When the contralateral, involved shoulder has been scored as a control, the results for the replaced shoulder are better (Table III), mainly because of greater relief from pain.

A trend towards worsening function is consistent with outcome studies in adult patients with juvenile idiopathic arthritis. Packham and Hall demonstrated significant ongoing activity of the disease over a prolonged period with the highest risk of severe functional limitation in the polyarticular and systemic subgroups. They suggested that the concept of disease ‘burn-out’, recognised in rheumatoid arthritis, may not hold true for the severe forms of juvenile idiopathic arthritis.

The observed functional deterioration in our series may indicate loosening of the humeral implant, but there were no radiological signs to suggest this. Alternatively, ongoing erosion of the non-resurfaced glenoid is a recognised cause of pain after hemiarthroplasty. However, relief from pain
was sustained throughout the study, militating against symptomatic erosion as an important factor.

One patient (case 6) has had an unsatisfactory result with a poor range of movement and forearm dysaesthesia. She relates her symptoms to the interscalene block, but an intra-operative nerve injury would be a more likely cause. Despite clear signs of a C6 radiculopathy on MRI she has refused to consider further surgery.

Technical difficulties relating to the small size and variable shape of the affected joints in juvenile idiopathic arthritis have been well documented at the hip, knee and elbow. These are compounded by osteoporosis, which may be steroid-related, with egg-shell-thin cortical bone and fragile vascularity. Generally, favourable results and a fairly low rate of revision are attributed to the low levels of activity and functional demands of these patients.

We encountered specific difficulties at the shoulder and recommend routine intra-operative image intensification. The rotator cuff was usually attenuated but intact; there was always marked capsular contracture and extensive intra-articular adhesions, sometimes with complete fibrous ankylosis. Extensive intra- and extra-articular release was always necessary. The small, eroded humeral head migrated medially in some cases with notching of the proximal shaft (Fig. 1). A bowed metaphysis and proximal diaphysis were commonplace which can force the stem into varus. Differences in the diameter of the canal in the sagittal and coronal planes were not always fully appreciated on the pre-operative plain radiographs, but can have a profound effect on version of the stem.

There is very little space for restoration or imposition of glenohumeral offset in these shoulders. Thus, even with small components and an eroded glenoid, it would have been technically difficult to insert a glenoid component without overstuffing the joint.

Dysplastic joint anatomy, porous bone and unpredictable function of the rotator cuff all predispose to early failure of a prosthetic glenoid in these patients. Although there is evidence of improved relief from pain with glenoid resurfacing in osteoarthritis, there is a high early incidence of radiolucent lines surrounding the glenoid component in rheumatoid shoulders with a subsequent rate of loosening of up to 40% at eight years. We believe that hemiarthroplasty has been the appropriate procedure in our series, affording excellent relief from pain and adequate closure of the soft-tissue envelope.

We strongly advocate the use of detailed pre-operative radiographs for measurement, both to predict potential anatomical problems and for the design of custom-made implants when necessary. Spiral CT with three-dimensional image reconstructions has been used in other joints undergoing replacement for juvenile idiopathic arthritis. It may be a useful adjunct in the shoulder, but has not been used. Custom-made prostheses are expensive but we advocate a low threshold for using them. Replacement of the elbow may be required in the future and this may be compromised by a stem of standard length in these patients. Similarly, bone stock for revision surgery is compromised by a stem of standard length relative to these small and weakened bones. For these reasons we have avoided the use of cement whenever possible.

We conclude that patients with end-stage, painful involvement of the shoulder in juvenile idiopathic arthritis may benefit from glenohumeral replacement. Overall function may be limited by progressive systemic disease in adjacent joints but relief from pain remains durable in the medium term. We have not had to revise any of these prostheses to date and it remains to be seen whether revision will be impossible. This must be borne in mind when considering surgery in such a young group of patients and we will continue to follow these patients closely.

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References


