



ELBOW

Total elbow arthroplasty in bleeding disorders: an additional series of 8 cases



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Background: Total elbow arthroplasty (TEA) is a surgical option for an arthropathy secondary to a bleeding disorder. The literature consists of small case series. Our series provides further understanding into the outcomes of TEA in this population of patients.

Methods: Five patients underwent 8 primary TEAs for a bleeding disorder. Average age at time of surgery was 47 years. Four patients had hemophilia type A and 1 had von Willebrand disease. Clinical outcomes were evaluated with the Mayo Elbow Performance Score (MEPS) and the visual analog scale (VAS) for pain. Follow-up radiographs were evaluated for signs of loosening and infection.

Results: Revision surgery was performed in 3 TEAs. Two revisions were performed for aseptic loosening (104 and 118 months postoperatively). The third elbow underwent an excision arthroplasty for a deep infection 44 months postoperatively. Mean follow-up for the primary TEAs still in situ (5 elbows) was 114 months. The mean VAS score improved from 8 to 0 and MEPS from 35 to 95. The mean flexion arc improved from 70° to 100°, and rotation improved from 60° to 160°. Mean follow-up for the revised TEAs (3 elbows) was 94 months. The mean VAS score improved from 7 to 0 and the MEPS from 40 to 85. The mean flexion arc improved from 60° to 95°, and rotation improved from 70° to 160°.

Conclusions: Excellent clinical outcomes and an acceptable survival rate for TEAs, comparable with the nonhemorrhagic population, can be achieved in patients with bleeding disorders. Revision arthroplasty in this group of patients yields good clinical outcomes at medium-term follow-up.

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

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Arthropathy is a serious and disabling complication of hemophilia.^{1,8,31} The elbow is the second most frequently involved joint in hemophilic arthropathy.¹ A chronic synovitis occurs as a consequence of recurrent bleeds in the joint, which in turn leads to destruction of the cartilage.

A progressive contracture of the elbow develops because of the synovitis and cartilage damage.^{1,8,31}

Treatment options in the early stages of hemophilic arthropathy include medical (synoviorrhesis) or surgical synovectomy with or without excision of the radial head.^{3,6,10,15,25,29,30,33} Surgical options for an advanced arthropathy are arthrodesis and arthroplasty. The arthroplasty may be a soft tissue interposition, excision arthroplasty, or total elbow arthroplasty (TEA).^{3-5,12,16,33}

As experience in arthroplasty has grown, TEA has become a more favored procedure in patients with hemophilia. Most papers on TEA for patients with hemophilia describe only 1 to 3 cases, with the largest series consisting of 8 TEAs in 5 patients. All are retrospective reports.^{2,4,5,12,16-18,23,27,34}

The aim of this study was to evaluate the outcomes in our series of patients and to add these data to the small numbers reported in the literature.

Materials and methods

Patients

We obtained approval from the institutional ethical committee for a retrospective review of all patients who had undergone a TEA for arthropathy as a result of a bleeding disorder by a single surgeon (B.C.V.). Eight TEAs were performed in 5 patients (3 bilateral). All patients gave permission for their clinical data to be reported and were included in the review.

In all patients, the indication for TEA was significant pain and loss of function, with severe joint destruction on plain radiographs. The only female patient in our series had von Willebrand disease type III; the other 4 patients had severe hemophilia type A (factor VIII deficiency). The female patient with von Willebrand disease requires daily factor VIII and has a more severe form of bleeding disorder than those with hemophilia type A, who require only intermittent factor VIII. Two patients were human immunodeficiency virus (HIV) positive, 2 had hepatitis C, and 1 had both diseases. Before TEA, 1 patient had a synovectomy and removal of the radial head, and 1 patient had a release of the ulnar nerve. All patients had previous orthopedic operations of other joints, including bilateral total knee arthroplasties, bilateral ankle arthrodeses, shoulder hemiarthroplasty, and radial head excision (Table I).

Surgical procedure

All patients received a bolus of 1500 mg cefazolin 30 minutes before skin incision; this was continued for 24 hours. We used a posterior approach under tourniquet control. For the revision cases, the old scar was used. A triceps split was used in 6 cases, a Campbell approach in 1, and a triceps-sparing approach in 1. The ulnar nerve was identified in all patients and released in situ without transposition.

A synovectomy and contracture release were performed. When the radial head was still present, it was excised. After preparation of humerus and ulna, the components were cemented with

gentamicin-impregnated cement. Superficial and deep drains were routinely used and removed within 24 hours. The wound was closed in layers with a continuous subcutaneous suture for skin closure to reduce the risk of bleeding and the need for factor VIII supplementation for removal of sutures. A plaster backslab was applied for 2 weeks in the patients who had an unlinked prosthesis, followed by a plaster splint, which was worn at night for a further 4 weeks. A Robert Jones bandage was applied for 10 to 14 days in the linked prosthesis, after which the elbow was left free. The female patient had an ipsilateral hemiarthroplasty of the shoulder performed at the same sitting as the TEA.

Hemophilia management

In all cases, a hematologist and a nurse with hemophilia training were involved in the perioperative care. Depending on the patient's profile and the clotting status, either a repeated bolus or a continuous infusion of factor VIII was administered. The female patient with von Willebrand disease also received factor VIII. Clotting factor titers were measured preoperatively, intraoperatively, and postoperatively. The patients were all well versed in the management of their condition and capable of managing their factor VIII requirements themselves with telephonic consultation from the hematology team.

Evaluation of clinical and radiologic outcomes

No patients were lost to follow-up. All patients were assessed at follow-up by the senior author (B.C.V.). The mean follow-up of the primary replacements (5 elbows) was 114 months (31-142). The mean follow-up of the revised elbows (3 elbows) was 94 months (24-160). The visual analog scale (VAS) score, Mayo Elbow Performance Score (MEPS), and range of movement were recorded and plain radiographs taken.²¹ The MEPS of patient number 4 (operated on in 1991) was derived from the data recorded in his clinical notes as this score was described only in 1993.²¹ Radiographic evaluation of the cement mantle was graded according to Morrey's criteria.²²

Statistical analysis

Values were described as median (interquartile range) as of a non-normal distribution. The preoperative and postoperative flexion, extension, arc of flexion, and rotation and the VAS score for pain and MEPS were compared by the Wilcoxon signed rank test, calculating exact *P* values. *P* values < .05 were considered statistically significant. All data were analyzed in SPSS 17.0 (SPSS Inc., Chicago, IL, USA).

Because of the low number of revision cases (3), the Wilcoxon signed rank test was not performed in this group.

Results

Mean age at primary surgery was 47 (32-63) years; the other characteristics of the 5 patients (8 TEAs) are shown in Table I. All TEAs, except the primary surgery in patient number 4, were performed by the senior author (B.C.V.).

Table I Clinical characteristics of all patients

Patient	Sex	Disease	HIV	Hep C	Age (years)	Follow-up (months)	Prosthesis	Revision due to	Revision prosthesis	Time to revision (months)	Other operations
1	M	Hem A	+	+	37	142	Kudo	N/A	N/A	N/A	TKA, AA
2	M	Hem A	+	—	57 (left)	31	Discovery	N/A	N/A	N/A	BAA
					54 (right)	89	Coonrad/Morrey	N/A	N/A	N/A	
3	F	vWD	—	+	63 (left)	44	Coonrad/Morrey	N/A	N/A	N/A	AA, TKA, HSA
					59 (right)	89	Coonrad/Morrey	N/A	N/A	N/A	
4	M	Hem A	—	+	31	267	Souter	Loosening	GSB III	104	AA, RHE
5	M	Hem A	+	—	35 (left)	154	Kudo	Deep infection	Excision	44	BAA, BTKA
					35 (right)	155	Kudo	Loosening	Discovery	118	

Hem A, hemophilia type A; *vWD*, von Willebrand disease; *Hep C*, hepatitis C; *HIV*, human immunodeficiency virus; *N/A*, not applicable; *TKA*, total knee arthroplasty; *AA*, ankle arthrodesis; *BTKA*, bilateral total knee arthroplasty; *BAA*, bilateral ankle arthrodesis; *RHE*, radial head excision; *HSA*, hemi shoulder arthroplasty.

Functional outcome

Table II shows all clinical outcome parameters of the 5 primary TEAs still in situ. Table III shows the clinical outcome parameters of the 3 revision cases.

In the primary cases, the range of movement improved significantly; the median flexion arc improved from 70° to 100°, and the median arc of rotation improved from 60° to 160°. Preoperative pain, measured with the VAS, was severe in 4 elbows (8) and moderate in 1 elbow (7). This improved significantly to a median of 0 at the most recent follow-up. The median MEPS improved from 35 to 95 ($P = .04$). All patients were very satisfied and stated that they would have the operation again.

In the revision cases, the median flexion arc improved from 60° to 95°, arc of rotation improved from 70° to 160°, and MEPS improved from 40 to 85. The VAS score improved from a median of 7 to 0 at most recent follow-up.

Prosthesis survival

The prostheses used included the Kudo type 4 (Biomet Inc., Warsaw, IN, USA), Coonrad/Morrey (Zimmer, Warsaw, IN, USA), Souter-Strathclyde (Stryker Howmedica Osteonics, Kalamazoo, MI, USA), Discovery (Biomet Inc.), and GSB III (Zimmer) as displayed in Table I.

Revision surgery was performed in 3 elbows (2 patients). The first patient (HIV positive, hepatitis C negative) developed a deep infection after a bleed following a fall 44 months after insertion of a Kudo prosthesis. He was treated with an excision arthroplasty and remains very satisfied at a follow-up of 110 months after removal of the prosthesis with a VAS score of 0 and a MEPS of 85. In the same patient, the opposite elbow was revised to a Discovery prosthesis 10 years after the primary TEA because of aseptic loosening of the ulnar component of a Kudo replacement. His VAS score for pain was 0 and the MEPS was 100 at follow-up of 37 months after revision. The third

Table II Preoperative and postoperative outcome of primary elbows, still in situ

Patient	Flexion arc		Rotation arc		VAS for pain		MEPS	
	Pre	Post	Pre	Post	Pre	Post	Pre	Post
1	80	120	120	160	8	0	35	100
2	70	100	60	160	8	0	35	95
	70	100	60	160	8	2	40	80
3	80	95	20	100	8	0	35	95
	60	90	0	160	7	0	50	95
Median	70	100	60	160	8	0	35	95
IQR	20	22.5	30	0	0.5	1	10	10
P value	.025		.042		.039		.042	

VAS, visual analog scale; *MEPS*, Mayo Elbow Performance Score; *pre*, preoperatively; *post*, postoperatively; *IQR*, interquartile range. *P* values are calculated by Wilcoxon signed rank test.

revision case was a hepatitis C-positive patient with aseptic loosening of the humeral component of a Souter-Strathclyde prosthesis after 9 years, revised to a GSB III prosthesis. At a follow-up of 163 months after the revision TEA, the patient remains very satisfied with a VAS score for pain of 2 and a MEPS of 80 (Figs. 1 and 2).

Radiologic follow-up

In both the primary and revision elbows, there were no progressive radiolucent lines on follow-up radiographs.

Discussion

The aim of this study was to evaluate the outcomes of our series of TEAs implanted for arthropathy due to bleeding disorders. This series demonstrated good clinical outcomes with significant improvement in the arc of both flexion and

Table III Preoperative and postoperative outcome of revised elbows

Patient	Flexion arc		Rotation arc		VAS for pain		MEPS	
	Pre	Post	Pre	Post	Pre	Post	Pre	Post
4	100	95	120	120	7	2	65	80
5	60	85	70	170	7	0	40	85
	50	105	70	160	9	0	20	100
Median	60	95	70	160	7	0	40	85
IQR	50	20	50	50	2	2	45	20

VAS, visual analog scale; MEPS, Mayo Elbow Performance Score; pre, preoperatively; post, postoperatively; IQR, interquartile range.

rotation, low VAS score for pain, and high MEPS. Revision surgery was necessary in 3 cases (37.5%) at an average of 7.4 years after TEA because of aseptic loosening in 2 cases and late deep infection in 1 patient. Subsequently, all 3 patients had a clinical outcome that was comparable to the outcomes of the unrevised cases.

Synovectomy is the first option for hemophilic arthropathy of the elbow, especially when damage of the cartilage is limited. It is the most frequently performed surgical procedure in patients with hemophilia, and results are reasonable. This can be performed medically (synoviorrhesis) or surgically (synovectomy).^{6,7,11,14,15,20,28-30,35} A Silastic interposition arthroplasty elbow has been described with acceptable results after a follow-up of 5 years.^{3,33}

With severe destruction of the elbow joint, a TEA is indicated. Most previously reported series include 1 to 3 cases,^{2,4,5,12,13,16-19,23,34} with the 3 largest series having 5 to 8 cases.^{5,12,17} Therefore, our series adds to the sparse literature.

Chapman-Sheath et al⁵ described 7 TEAs (6 patients) with a mean follow-up of 42 months. They found an improvement in flexion from 97° to 122° and in rotation from 33° to 115°. Both the preoperative and postoperative values were higher in our series; the amount of improvement was equivalent. Pain improved from severe in all elbows to no pain in 5 elbows and mild pain in 2. They reported one revision due to a late infection.

Marshall Brooks et al¹⁷ demonstrated outcomes in 7 TEAs (6 patients) with a mean follow-up of 118 months. Range of motion improved in 5 of the 7 TEAs, with mean flexion improving from 111° to 120° and mean extension improving from 44° to 37°. There was one excision arthroplasty for deep infection. Finally, Kamineni et al¹² presented 8 TEAs with a follow-up of 70 months. MEPS improved from 24 to 90. There were 2 cases of deep infection, 1 managed with excision arthroplasty and the second with retention of the implants after débridement. One TEA showed signs of aseptic loosening but had not been revised. Our series compares favorably with these 3 series, demonstrating similar improvement in function and



Figure 1 Anteroposterior radiograph of patient 4, 14 years after revision TEA.



Figure 2 Lateral radiograph of patient 4, 14 years after revision TEA.

pain relief. Furthermore, in these series, infection was most often associated with a significant bleed as occurred in our 1 case of sepsis. The length of follow-up in our series is equivalent to that of Marshall Brook's series but longer than that of the other 2 papers.^{5,12,17} This may account for the higher number of revisions in our series. The 2 revisions for aseptic loosening were performed at 9 and 10 years, a longer interval than the mean length of follow-up of those in the series of Chapman-Sheath and Kamineni.^{5,12}

One of our patients underwent an ipsilateral shoulder replacement at the same sitting. Ipsilateral shoulder and elbow replacement has previously been reported in a series of patients with rheumatoid arthritis.⁹ In that series, it was found to be safe and had the advantage of a single anesthetic. Ipsilateral surgery also facilitates rehabilitation of the whole upper limb and may decrease costs. Performing a concurrent shoulder and elbow replacement in a hemophilic is a major saving in the cost of factor VIII. This technique has been described in a case report on a hemophilic patient by Phillips et al.^{9,23,32,36}

These patients are perceived to be at higher risk of postoperative complications, not only because of their hemophilia but also because they frequently have a concomitant HIV or hepatitis C infection.^{19,24,26} Three of our patients were HIV positive, all of them contracting the disease in the early 1980s.

The main limitations of our series, similar to other papers dealing with this topic, are the small number of patients and the study's retrospective nature.

Conclusion

Our data confirm the findings in the literature that TEA for hemophilic arthropathy is a good surgical option with excellent clinical results and an acceptable survival rate. Furthermore, revision surgery in this group of patients results in good clinical outcomes with relative longevity (3 and 13 years in our patients).

This paper also highlights the medical and cost-saving benefits of performing an ipsilateral shoulder replacement at the same sitting as a TEA. Finally, the life expectancy of HIV-positive patients has improved dramatically because of antiretroviral therapy. They can now live relatively normal lives and should be treated as one would treat any other patient.

Disclaimer

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