OPERATIVE TECHNIQUE

Short-term Effectiveness of a Modified Osteotomy for Total Elbow Joint Replacement in Patients of Hemophilic Elbow Arthritis with Severe Flexion Contracture Deformity: A Three-Cases Series Report

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Objective: To report the modified osteotomy and the short-term effectiveness of the total elbow joint replacement in patients of hemophilic elbow arthritis with severe flexion contracture deformity.

Methods: This study introduced the therapeutic approach of the total elbow joint replacement in patients of hemophilic elbow arthritis with severe flexion contracture deformity, and assessed the short-term effectiveness in three cases (three elbows) of end-stage hemophilic elbow arthritis admitted from October 2020 to December 2020. The included patients were all diagnosed with hemophilia A (factor VII deficiency), accompanied by severe bilateral elbow joint flexion contracture, which seriously affects daily life and requires surgical intervention. Clinical data and follow-up results were analyzed before total elbow arthroplasty and 1, 3, and 6 months postoperatively. Pre- and postoperative range of motion, pain score, and function score were compared, and intraoperative and postoperative complications are reported.

Results: All three patients were male, with an average age of 31 years. The main clinical manifestations were bilateral elbow arthritis with flexion contracture. Two of the patients underwent right elbow replacement, and one patient underwent left elbow replacement. All cases were followed up for 6 months postoperatively. No incision infection or ulnar nerve injury occurred. Postoperative triceps brachii muscle strength was slightly weakened compared with preoperative muscle strength. Average elbow flexion and extension range of motion was 60° (30°–100°) preoperatively and increased to 127° (110°–140°) postoperatively; rotational range of motion of the affected forearm was 47° (10°–85°) preoperatively and increased to 117° postoperatively. The mean visual analogue scale (VAS) was 6 (5–8) preoperatively and decreased to 3 (2–4) postoperatively. The mean MEPS score was 62 (55–75) and increased to 87 (80–95) postoperatively. During the follow-up, anteroposterior and lateral radiographs showed no signs of prosthesis loosening in the elbow.

Conclusions: For severe hemophilic elbow arthritis patients, the short-term treatment effect of total elbow replacement is good, following the strict adherence to the surgical indications and proper preparation for the perioperative period. The modified osteotomy can fully expose the visual field and reduce complications of ulnar nerve injury. The long-term effects need to be study future.

Key words: Elbow joint function; Hemophilia A; Hemophilic elbow arthritis; Pain score; Total elbow joint replacement

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Introduction

Hemophilia is an X-linked recessive inherited bleeding disorder and includes hemophilia A (factor VIII deficiency) and B (factor IX deficiency). A recent meta-analysis showed that the global prevalence of hemophilia A and B was 17.1 and 6.0 per 100,000 men, respectively. Most patients with hemophilia who have been observed have chronic pain and loss of function due to degenerative joint disease (hemophilic arthropathy) caused by repeated joint hematomas that seriously decreases the quality-of-life and increases the financial burden of patients.

As a serious complication of hemophilia, hemophilic arthropathy’s major feature is recurring spontaneous intra-articular bleeding, which causes joint structure damage and symptoms such as joint pain. Severe cases can lead to ultimately dysfunction and residual disability.

Currently, surgical treatments for hemophilic elbow arthritis include synovecctomy, radial head resection, and total elbow arthroplasty (TEA). The surgical characteristics of TEA in hemophilic elbow arthritis cases differ from those in other cases. Due to the early development of skeletal deformities, repeated bleeding and inflammation in the joints, the bone and joint structures may be damaged and result in anatomical variation, which may complicate surgery. Some studies report patients with severe end-stage hemophilic elbow arthritis undergoing total elbow arthroplasty (TEA).

In a previous case report from Kamineni et al., one case exhibited sclerosis and occlusion of the humerus and ulnar medullary cavity during the operation, and a high-speed electric drill was required to recanalize the medullary cavity. In another case, ulnar medullary canal stenosis was found, which required special care during reaming to avoid drilling through cortical bone, and a smaller prosthesis was required. Although TEA were effective in pain relief and functional improvement of affected limbs, the high rate of postoperative complications still limited its application. Continuous damage to joints due to hemophilia, the second-stage revision rate is 12.5%–37.5%. Therefore, minimizing postoperative complications is an important means to effectively reduce secondary operations.

Hemophilic arthritis of the elbow resulted in severe bone structure destruction, hypertrophy of the radial head, subluxation of the radial and radioulnar joints, destruction of the ulnar joint, and articulation of the ulna and the inner side of the humerus trochlea. The proportion of ulnar nerve injury in elbow joint replacement is as high as 71.4%. Mild injury can recover spontaneously, while muscle atrophy occurs in severe cases, ulnar nerve release and transposition are required.

The aim in this study is to explore an optimal surgical method for reducing the ulnar nerve injury the end-stage elbow joint in patients with hemophilia, and assess the intraoperative bleeding, perioperative complications, and the early postoperative joint function of the patients.

Materials and Methods

Patients’ Information

All the patients underwent TEA from October 2020 to December 2020 in the Department of Orthopedics, The Third People’s Hospital of Shenzhen. Three patients were all male, aged 26–38 years (average, 31 years), and diagnosed with hemophilia A (factor VIII deficiency). All had bilateral elbow arthritis with flexion contracture: one case had right knee joint disease; one had bilateral hip, knee, and ankle disease; and one had bilateral knee disease without other diseases. Two cases underwent right elbow replacement, and one underwent left elbow replacement (Table 1). Preoperative imaging examinations showed that all elbow joints were severely deformed with obvious bone degradation.

This study was approved by the Ethical Committee of the Shenzhen Third People’s Hospital (2022-107-02); written informed consent was obtained from each participating patient.

Consent for publication of photos, and other identifying materials were obtained from all patients.

Coagulation Factor Pharmacokinetic Testing

Before surgery, we routinely test the pharmacokinetics of blood coagulation factors in patients to determine the half-life of blood coagulation factors in vivo. According to the pharmacokinetic characteristics, coagulation factors were infused regularly during the perioperative period to maintain a target concentration of coagulation factors. Coagulation factor dosage is based on the patient’s weight according to the formula: (target concentration-preoperative concentration) × body weight × 0.5. Blood coagulation factor concentration was measured immediately, 30 min, 9 h, 24 h, and 48 h after infusion of the coagulation factor, and the half-life was determined in vivo with simultaneous detection of coagulation factor inhibitors.

| Case | Gender | Age | Types of hemophilia | Surgical side | Comorbidity | Other joint involvement | Follow-up time (m) |
|------|--------|-----|---------------------|---------------|-------------|------------------------|-------------------|
| 1    | Male   | 38  | A                   | Right         | No          | Right knee             | 6                 |
| 2    | Male   | 26  | A                   | Right         | No          | Double hips, double knees, double ankles | 6                 |
| 3    | Male   | 29  | A                   | Left          | No          | Double knees           | 6                 |
**Prosthesis Selection**

The Morrey–Coonrad prosthesis (Zimmer Inc., Warsaw, IN, USA) were chosen, which is a semi-restrictive type (designed by Mayo Hospital and Zimmer Company in 1969 and successively improved in 1971, 1978, 1981, 1987, and 2000). Features of the prosthesis were 8° varus and valgus relaxation and 8° medial and lateral rotation relaxation; a bulge was added to the front of the lower end of the humeral prosthesis, where a bone block was implanted to enhance the fixation effect of the prosthesis and prevent the humeral stem from moving slightly backward or rotating axially.

**Surgical Methods**

**Surgical Position**

The patient was placed in the supine position, with the shoulder blades slightly elevated and the upper arms horizontal in front of the chest. After disinfection, a sterile airbag tourniquet was applied.

**Surgical Incision and Exposure**

A Bryan–Morrey posterior elbow median incision of approximately 15 cm was used, beginning with nerve-free rubber strip traction protection. The Sharpey fibers on the olecranon process were excised, and the inner side of the triceps brachii was peeled off along the periosteum of the olecranon to the lateral side, using special attention to maintain its continuity. The medial and lateral collateral ligaments were resected, lateral side, using special attention to maintain its continuity. The medial and lateral condyles of the distal humerus and the olecranon fossa of elbow fl

**Handling the Distal Humerus**

The distal humerus (Fig. 1(A)). Pay attention to the muscular branches of the ulnar nerve that innervate the flexor carpi ulnaris muscle. The intraoperative pulling must be gentle.

**Handling the Proximal Ulna and Radius and Medullary Cavity**

The slender bone of the olecranon of the ulna was removed according to the degree of dislocation and contracture, generally about 2–4 cm. After the installation of the prosthesis, the insertion of triceps brachii was firmly fixed to the proximal end of the ulna. Steel wire and No. 5 Aixibang wire were used to open the ulnar bone marrow cavity with a drill bit, and then the ulnar bone marrow cavity was enlarged with a grinding drill bit and a pulp cavity file (Fig. 1). Pay attention to the Angle of reaming so as to not penetrate the medullary cortex or fracture the cortex. The medullary cavity of three patients in this group was very narrow, and it was necessary to enlarge the medullary cavity repeatedly with a grinding drill until the smallest ulna prosthesis model could be installed. The radial head was obviously hyperplasia and coarse deformation. After the prosthesis was installed, the joint was flexed and extended, and there were obvious impacts and protrusions. The radial head was not removed.

**Release Contracture Tissue in Front of Elbow Joint**

After the initial installation of the joint prosthesis, the elbow joint was straightened, and the anterior elbow soft tissue with tension and contracture was released. In this group, the skin and soft tissue contracture in front of the elbow joint seriously affected the straightening, and the elbow joint could be basically straightened after the release of the Z-shaped skin flap and the release of the anterior elbow fascia and contracture tissue (a difference of 10°; Fig. 1(B)).

**Fitting Joint Prostheses and Closing Wounds**

Thoroughly rinse the ulna and humeral medullary cavity, fix the Zimmer semi-restricted elbow joint prosthesis with bone cement, trim the prosthesis in the anterior position of the distal humerus, trim the bone graft, and use the triceps insertion point and tendon part to use the No. 5 Aixon Bond suture thread. It is firmly fixed with the proximal end of the ulna, and no avulsion of the flexor and extension joints is the standard (Fig. 1(C)). The free ulnar nerve was moved forward and fixed in the subcutaneous soft tissue. The tourniquet was loosened, the bleeding was fully stopped, the incision was sutured directly without the drainage tube, and the incision was fixed with a straight plaster and elastic bandage.

**Postoperative Rehabilitation**

A second-generation cephalosporin antibiotic was used before and after the operation to prevent infection. After the operation, ice was applied to the wound for 20 min at 4-h intervals. The dressing was changed within 24 h, the wound was inspected for oozing and subcutaneous stasis, and patients were checked for neurological symptoms. On the
first postoperative day, patients were encouraged to start active and passive exercises of fist and elbow joint extension and flexion. Within one week after the operation, fix the affected elbow in a straight position with a plaster while resting at night. During the hospitalization period, the rehabilitation therapist helped the patient to practice daily elbow extension every day under oral analgesics or nerve block anesthesia. The patients were instructed to go to the outpatient clinic regularly for rehabilitation training every month after being discharged. Elbow extension resistance exercises were started at 6 weeks postoperatively. It was recommended to limit weight bearing of the affected limb to 3–4 kg. Based on Arnold and Hilgartner stag IV, the imaging stages of hemophilia arthritis were all stage V (end-stage elbow arthritis; Fig. 2). Elbow joint flexion, extension, and rotational range of motion were recorded preoperatively at 1, 3, and 6 months postoperatively. The visual analogue scale (VAS) was used to assess elbow pain, and the Mayo elbow performance score (MEPS) was used to assess elbow joint function.

**Results**

A total of three cases were included in this study, they were all male hemophilia of A type without comorbidity. The surgical side was located right in two patients and left in one. All three patients were followed-up 6 months. During the follow-up period, the patient’s elbow pain and function significantly improved, and no complications occurred.
**Functional Improvement**

The triceps brachii muscle strength after operation was slightly weakened compared with that before operation. The average VAS score decreased from 6 preoperatively to 3 postoperatively, and MEPS increased from 62 to 87. In addition, pain and function of the affected elbow significantly improved.

After an average follow-up of 6 months, average elbow flexion and extension range of motion increased from 60° preoperatively to 127° postoperatively, and the average rotational range of motion increased from 47° preoperatively to 117° postoperatively. Flexion and extension range of motion significantly improved postoperatively compared with that preoperatively, although the rotational range of motion tended to improve postoperatively compared with that preoperatively.

During the follow-up, radiographs of the affected elbow were acquired, and there were no signs of prosthesis loosening.

**Use of Coagulation Factors during the Total Elbow Arthroplasty Perioperative Period**

According to the results of the pharmacokinetic test (Table 2), coagulation factor VII was infused regularly during the perioperative period, and the coagulation factor concentration was maintained at 100% on the day of surgery, 60%–80% for 1–3 days after surgery, 40%–60% for 4–6 days, and 30%–50% for 1–2 weeks postoperatively.

**Complications**

There were three cases of humeral internal and external epicondylar fractures that occurred during the operation. No incision infection or ulnar nerve injury occurred after operation.

**Case Descriptions**

**Case 1**

Before the operation, the patient’s elbow was extended −60°, flexed 105°, pronation 20° and supination 40°, and after the operation, the patient’s elbow extended 0°, flexed 120°, pronation 75° and supination 80° (Fig. 3).

It can be seen during the operation that a large amount of bone hyperplasia and a large amount of dark yellow synovial hyperplasia were observed on the medial right ulna. There was clear synovial tissue and hyperosteogeny. We performed an osteotomy at the distal end of the right humerus and the proximal end of the ulna, installed a prosthesis of XS size, and fixed it with bone cement. The inspection showed that the right elbow joint could be straightened up to 0°. The incision was washed, and the joint capsule was sutured. The right ulnar nerve was subcutaneously anterior, and the incision was sutured (Fig. 4).

**Case 2**

Before the operation, the patient’s elbow was extended −75°, flexed 130°, pronation and supination 0°, and after the operation, the patient’s elbow extended −5°, flexed 90°, pronation 40° and supination 20°.

During the operation, the elbow joint was observed in subluxation, the articular surface of the internal and external condyles of the humerus was severely worn, the surrounding bone hyperplasia formed a false joint, and the surface was dark brown with fibrous tissue adhesion and yellow-brown synovial hyperplasia.

**Case 3**

Before the operation, the patient’s elbow was extended −40° and flexed 140°, and after the operation, the patient’s elbow extended 0° and flexed 140°.

It can be seen during the operation that a large amount of bone hyperplasia and a large amount of dark yellow synovial hyperplasia on the inner side of the left ulna. There was clear synovial tissue and hyperosteogeny. The left humerus and the proximal ulna underwent osteotomy, a prosthesis of XS size was installed, and it was fixed using bone cement. The inspection showed that the left elbow joint could be straightened up to 0°, after which the incision was washed and the joint capsule was sutured. The left ulnar nerve was subcutaneously anterior, and the incision was sutured.

**Discussion**

**Main Findings**

This study reported a modified osteotomy in patients with hemophilic elbow arthritis undergoing TEA and found that intraoperative osteotomy of medial and lateral condylar muscle insertion of the humerus and triceps muscle insertion of the proximal ulna can fully expose the visual field and reduce complications of ulnar nerve injury. The surgery method is safe and effectual. Postoperative triceps brachii muscle strength was slightly weakened compared with preoperative

| TABLE 2 Pharmacokinetic results of coagulation factor VII |
|----------------------------------------------------------|
| Coagulation factor activity (%) | 0 h | 0.5 h | 9 h | 24 h | 48 h |
|-------------------------------|-----|-------|-----|------|------|
| Case | Weight (kg) | Inhibitor | Infusion of coagulation factor dose (IU) |     |      |     |     |     |
| 1    | 67            | (–)   | 2500 | 2    | 88   | 59  | 37  | 20  |
| 2    | 50.5          | (–)   | 2000 | 1    | 52   | 34  | 18  | 2   |
| 3    | 52            | (–)   | 2000 | 2    | 66   | 40  | 23  | 9   |
muscle strength. Average elbow flexion and extension range of motion, rotational range of motion of the affected forearm were enhanced. The mean VAS was decreased postoperatively; the mean MEPS score was increased. During the follow-up, no signs of prosthesis loosening was observed in the elbow.

**Surgical Characteristics of TEA**

The surgical characteristics of TEA in hemophilic elbow arthritis cases are different from other cases. Due to the early development of skeletal deformities, repeated bleeding and inflammation in the joints, the bone and joint structures may be damaged and cause anatomical variation, which may bring difficulties to surgery. In the case report of Kamineni et al., one case was found to be sclerosis with occlusion of the humerus and ulnar medullary cavity during operation, so a high-speed electric drill was required to recanalize the medullary cavity. In the other case, ulnar medullary canal stenosis was found, which required special care during reaming to avoid drilling through cortical bone, and a smaller prosthesis was required. The three cases reported in this article all had different degrees of elbow flexion deformity before operation. During the operation, we found that in addition to the very narrow ulnar cavity that requires repeated reaming, the ulnar olecranon and the distal humerus will collide with the distal humerus due to long-term flexion deformity and arthritis caused by the disorder of the humeral-ulnar joint relationship. If the olecranon is

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**Fig. 3** Comparison of elbow joint function before and after surgery, showing that before the operation, the patient’s elbow flexed 105° (A), extended –60° (B), pronation 20° (C) and supination 40° (D); after the operation, the patient’s flexed 120° (E), elbow extended 0° (F), pronation 75° (G) and supination 80° (H)

**Fig. 4** The X-ray of elbow joint of case 1 at 6 months after operation, showing the X-ray of the elbow joint of Case 1 at 6 months after operation (A: anteroposterior view; B: lateral view)
Postoperative complications include bleed-
Chapman-Sheath reported all cases had complications after operation. It is In the follow-up of
sis must be enlarged by a written test drill to expand the proximal end of the ulna is closed or narrowed, the prosthe-
No. 1 tendon thread. (iii) Because the medullary canal at the level above the olecranon fossa of the humerus ulna. The middle of the oste-
tomy was repaired in u-shape and the humerus prostheses was inserted to achieve stable rotation of the prostheses. The insertion point of triceps brachii tendon at the proximal ulna was subperiosteal dissection with thin layer of bone. The olecranon about 1.5–2 cm was removed as required. The elbow joint could be completely extended and bent after the test model was installed.

Postoperative Complications
According to reports, the incidence of complications after TEA is 12.5%–42.9%, and the second-stage revision rate is 12.5%–37.5%10 Postoperative complications include bleeding, deep infection, prosthesis loosening and ulnar nerve damage. Chapman-Sheath et al.9 reported five cases out of seven elbows which underwent TEA treatment. The average follow-up was 42 months after operation. There was one case each of ulnar nerve palsy, deep infection and axillary vein thrombosis. The complication rate was 42.9%. In the case series of six cases with seven elbows such as Marshall Brooks et al.,10 all cases had complications after operation. It is worth mentioning that because the ulnar nerve was not pre-
positioned in the initial operation, five elbows out of seven elbows were postoperative There are varying degrees of ulnar nerve damage. The best result was the seven cases out of eight elbow reported by Sorbie et al.15 In the follow-up of more than 5 years, only one case required revision because of osteolysis. The three cases reported in this article suffered from obvious elbow pain or movement limitation before TEA, and X-ray showed severe damage to the elbow joint. In addition, all three cases of elbow had different degrees of flexion deformity, and the patients also had requirements for the appearance of their limbs. TEA combined elbow joint replacement or hemophilia elbow joint replacement was performed. The average follow-up period was 6 months, and the pain and function of the elbow were significantly improved. No complications occurred. The long-term effect of the operation still needs to be observed.

Coagulation Factor Replacement Therapy
Special attention should be paid to the replacement therapy of coagulation factors during the perioperative period of TEA. The maintenance of coagulation factor concentration is conducive to the safe operation of the operation, and at the same time can reduce the risk of bleeding, infection and prosthesis loosening during the perioperative period. In a small number of patients with hemophilia, there are coagula-
tion factor inhibitors. Once the inhibitors appear, it will be difficult to maintain the concentration of coagulation factors during the perioperative period, which will cause great diffic-
ulna was changed, that is, the internal and external condyles of the humerus were not retained, and the medullary cavity was slotted after horizontal transverse osteotomy above the olecranon fossa of the humerus ulna. The middle of the oste-
tomy was repaired in u-shape and the humerus prostheses was inserted to achieve stable rotation of the prostheses. The insertion point of triceps brachii tendon at the proximal ulna was subperiosteal dissection with thin layer of bone. The olecranon about 1.5–2 cm was removed as required. The elbow joint could be completely extended and bent after the test model was installed.

Modified Surgery Methods
The characteristics of previous hemophilic elbow joint replacement: (i) subperiosteal stripping of the insertion points of the medial and lateral condyle flexors and common extensor tendons of the humerus, transverse osteotomy at the upper edge of the olecranon fossa, and humerus prosthesis installed to remove the flexor and common extensor tendons. The insertion points were firmly sutured to the medial and lateral myofascia respectively. (ii) The triceps brachialis is subperiosteal stripped. After the ulnar prosthesis is installed, two bone holes are made on the back of the proximal ulna, and it is firmly sutured and fixed with Aixibang No. 1 tendon thread. (iii) Because the medullary canal at the proximal end of the ulna is closed or narrowed, the prostheses must be enlarged by a written test drill to expand the medullary canal.

In this study, the surgical method changed the osteote-
tomy method of the humerus and ulna according to the pathological characteristics of the elbow joint in hemophilia. That is, without retaining the medial and lateral condyles of the humerus, the osteotomy was performed transversely at the level above the olecranon fossa of the humerus, and then the medullary cavity was slotted and the osteotomy was performed. The middle part is trimmed into a U shape and the humerus prosthesis is inserted to achieve rotational stability of the prosthesis. The triceps tendon insertion at the proximal end of the ulna is subperiosteal stripped with a thin layer of bone, and the olecranon of about 1.5–2 cm is removed as needed.

According to the pathological characteristics of hemophiliac elbow joint, the osteotomy method of humerus and
period. Frequent coagulation factor input may stimulate the production of coagulation factor antibodies in the body.\(^\text{16}\) Therefore, for patients who do not have inhibitors in the body, maintaining the coagulation factor concentration during the perioperative period is beneficial to reduce the risk of postoperative bleeding and infection; on the other hand, the incidence of postoperative complications is reduced, which is conducive to reducing the number of operations. It is also of great significance to reduce the production of inhibitors. There are no inhibitors in our cases. Before surgery, we routinely test the pharmacokinetics of blood coagulation factors in patients to determine the half-life of blood coagulation factors \textit{in vivo}. According to the pharmacokinetic characteristics of coagulation factors, coagulation factors were infused regularly during the perioperative period to maintain the target concentration of coagulation factors during the perioperative period. No complications such as bleeding and infection occurred after the operation.

**Improvement Postoperation**

Although the incidence of complications after TEA for hemophilic elbow arthritis is relatively high, the results of postoperative functional recovery reported in most case series are relatively satisfactory. At present, there are very few reports on TEA for hemophilic elbow arthritis at home and abroad, and only a few small sample case series are reported. In this article, the average VAS score decreased from 6 points before surgery to 3 points after surgery, and MEPS increased from 62 points before surgery to 87 points after surgery. The pain and function of the affected elbow were significantly improved. The results are consistent with the existing case series. According to 25 cases, rheumatoid arthritis is the most common indication for TEA, and TEAs have more complications than most other joint replacement surgeries.\(^\text{17,18}\) As for the elbow joint flexion and extension and rotation range of motion, the better results reported in foreign countries include the five cases of four elbow case series reported by Kamineni et al.\(^\text{6}\) (including one case of prosthesis removal due to deep infection in 2 months after surgery). After an average follow-up of 10.2 years, the average flexion and extension range of the affected elbow increased from 38.75° preoperatively to 88.75° postoperatively, and the average rotational range increased from 58°preoperatively to 145° postoperatively. Vochtelo et al.\(^\text{19}\) reported five cases of eight elbows. The average follow-up was 114 months after the operation. The median flexion and extension range of the affected elbow increased from 70° preoperatively to 100° after the operation. The first 60° increased to 160° after the operation. Chapman-Sheath et al.\(^\text{9}\) reported five cases of seven elbows. The average follow-up was 42 months after the operation. The average flexion and extension range of the affected elbow increased from 50° preoperatively to 93.5° postoperatively, and the average rotational range increased from 33° preoperatively to 115° after operation. The worst results were reported by Ernstbrunner et al.\(^\text{20}\) in eight cases with 11 elbows. The average follow-up was 9.1 years after operation. The average flexion and extension range of the affected elbow increased from 71° before operation to 81° after operation, and the average rotation range was from 130° before operation increased to 156° after surgery. In our case series, after an average follow-up of 6 months, the average elbow flexion and extension activity increased from 60° preoperatively to 127° postoperatively, and the average rotational activity increased from 47° preoperatively to 117° postoperatively. The flexion and extension range of motion was significantly improved compared with that before the operation, and although the rotation range of motion was significantly improved compared with that before the operation, the difference was not significant (\(P = 0.07\)). Compared with the previous case series, our case is more prominent in the improvement of flexion and extension activities, with an average of 67°. The reasons may include the following aspects. First of all, the average age of our cases is relatively young, the patients have higher requirements for the appearance and function of the affected elbow, and they have better compliance. They can follow the doctor’s instructions for rehabilitation after the operation. Second, we routinely equip a rehabilitation therapist to assist patients in rehabilitation training after the operation, which also promotes the functional recovery of the affected elbow. Finally, during the operation, we found that the anatomical deformity of the affected elbow caused the impact of the elbow joint in the extension position, and the olecranon and the distal humerus were osteotomy, and the soft tissue contracture of Case 2 was released. We believe that this is the most important reason for obtaining greater flexion and extension activity after surgery.

**Limitations**

This study shows that for severe hemophilic elbow arthritis cases, under the premise of strictly grasping the surgical indications and fully preparing for the perioperative period, a total elbow joint replacement surgery can obtain a good therapeutic effect. There were some limitations in this study. First, the sample size is small and the sample size needs to be expanded in the future to continue research. Second, the current follow-up time is relatively short, and it is necessary to continue follow-up to further evaluate the efficacy.

**Conclusion**

This study reported a modified osteotomy in patients with hemophilic elbow arthritis undergoing TEA and found that intraoperative osteotomy of medial and lateral condylar muscle insertion of the humerus and triceps muscle insertion of the proximal ulna can fully expose the visual field and reduce complications of ulnar nerve injury. These findings suggested that for severe hemophilic elbow arthritis cases, under the premise of strictly grasping the surgical indications and fully preparing for the perioperative period, TEA with modified osteotomy can result in a good therapeutic effect.
References

1. Dale TM, Saucedo JM, Rodríguez-Merchan EC. Total elbow arthroplasty in haemophilia. Haemophilia. 2018;24(4):548–56.
2. Iorio A, Stonebraker JS, Chambost H, Makris M, Coffin D, Herr C, et al. Establishing the prevalence and prevalence at birth of hemophilia in males: a meta-analytic approach using national registries. Ann Intern Med. 2019;171(8):540–6.
3. Mannucci PM, Tuddenham EG. The hemophilias—from royal genes to gene therapy. N Engl J Med. 2001;344(23):1773–9.
4. Arnold WD, Hilgartner MW. Hemophilic arthropathy. Current concepts of pathogenesis and management. J Bone Joint Surg Am. 1977;59(3):287–305.
5. Utukuri MM, Goddard NJ. Haemophilic arthropathy of the elbow. Haemophilia. 2005;11(6):565–70.
6. Kamineni S, Adams RA, O’Driscoll SW, Morrey BF. Hemophilic arthropathy of the elbow treated by total elbow replacement. A case series. J Bone Joint Surg Am. 2004;86(3):584–9.
7. Rodríguez-Merchan EC. The role of orthopaedic surgery in haemophilia: current rationale, indications and results. EFORT Open Rev. 2019;4(5):165–73.
8. Ishiguro N, Yasuo S, Takamatu S, Iwata H. Hemophilic arthropathy of the elbow. J Pediatr Orthop. 1995;15(6):821–5.
9. Chapman-Sheath PJ, Gianfranco P, Carr AJ. Arthroplasty of the elbow in haemophilia. J Bone Joint Surg Br. 2003;85(8):1138–40.
10. Marshall Brooks M, Tobase P, Karp S, Francis D, Fogarty PF. Outcomes in total elbow arthroplasty in patients with haemophilia at the University of California, San Francisco: a retrospective review. Haemophilia. 2011;17(1):118–23.
11. Song SJ, Bae JK, Park CH, Yoo MC, Bae EK, Kim KI. Mid-term outcomes and complications of total knee arthroplasty in haemophilic arthropathy: a review of consecutive 131 knees between 2006 and 2015 in a single institute. Haemophilia. 2018;24(2):299–306.
12. Ernstbrunner L, Hingsammer A, Catanzaro S, Sutter R, Brand B, Wieser K, et al. Long-term results of total knee arthroplasty in haemophilic patients: an 18-year follow-up. Knee Surg Sports Traumatol Arthrose. 2017;25(11):3431–8.
13. Pasta G, Annunziata S, Polizzi A, Calogna L, Jannelli E, Minen A, et al. The progression of hemophilic arthropathy: the role of biomarkers. Int J Mol Sci. 2020;21(19):7292.
14. Beeton K, Rodríguez-Merchan EC, Altbee J. Total joint arthroplasty in haemophilia. Haemophilia. 2000;6(5):474–81.
15. Sorbie C, Saunders G, Carson P, Hopman WM, Olney SJ, Sorbie J. Long-term effectiveness of Sorbie-QUESTOR elbow arthroplasty: single surgeon’s series of 15 years. Orthopedics. 2011;34(9):e561–9.
16. Dale TM, Saucedo JM, Rodríguez-Merchan EC. Hemophilic arthropathy of the elbow: prophylaxis, imaging, and the role of invasive management. J Shoulder Elbow Surg. 2015;24(10):1669–78.
17. Ikävalko M, Lehto MU, Repo A, Kautiainen H, Hämäläinen M. The Souter-Strathclyde elbow arthroplasty. A clinical and radiological study of 525 consecutive cases. J Bone Joint Surg Br. 2002;84(1):77–82.
18. van der Lugt JC, Rozing PM. Systematic review of primary total elbow prostheses used for the rheumatoid elbow. Clin Rheumatol. 2004;23(4):291–8.
19. Vochteloo AJ, Roche SJ, Dachs RP, Vrettos BC. Total elbow arthroplasty in bleeding disorders: an additional series of 8 cases. J Shoulder Elbow Surg. 2015;24(5):773–8.
20. Ernstbrunner L, Hingsammer A, Imam MA, Sutter R, Brand B, Meyer DC, et al. Long-term results of total elbow arthroplasty in patients with hemophilia. J Shoulder Elbow Surg. 2018;27(1):126–32.