OPERATIVE TECHNIQUE

Treatment of Congenital Radioulnar Synostosis Using a Free Vascularized Fascia Lata Graft

Fan Bai, MD1,2, Shanlin Chen, MD1,2, Lu Liu, MD2, Dedi Tong, MD2, Pengcheng Li, MD2, Yanbo Rong, MD2, Jingheng Wu, MD2, Bo Liu, MD2, Yong Yang, MD2, Shufeng Wang1,2

1Peking University Fourth School of Clinical Medicine and 2Department of Hand Surgery, Beijing Ji Shui Tan Hospital, Beijing, China

Abstract

Objective: To describe the modified mobilization surgery technique that uses a free vascularized fascia lata graft as the interposition graft, and to evaluate the outcome of this procedure in treating congenital radioulnar synostosis (CRUS).

Methods: Eleven patients (eight boys and three girls with an average age of 6.0 years) were treated using this procedure between 2012 and 2017 in our institution. Five bilateral cases (four left forearms and one right forearm were treated), and six unilateral cases (three left forearms and three right forearms) were included. All 11 cases were treated with mobilization procedure with free vascularized fascia lata as the interposition graft, and were followed-up for an average of 2.2 years (range, 2–4 years). The parental satisfaction, postoperative ankylosis at proximal radioulnar joint, and active range of forearm rotation motion (measured by physical examination) were evaluated at the last follow-up.

Results: The average preoperative fixed pronation angle was 67.3° (range, 20°–90°). Ipsilateral thumb hypoplasia was noted in one case, and cleft palate and bilateral thumb hypoplasia were noted in one case; none of the patients had a family history of congenital radioulnar synostosis. Pronation and supination splints were used 3 days after the operation and were worn every night for 4–6 months postoperatively. Active and passive rehabilitation for elbow flexion and forearm rotation was initiated 4 weeks postoperatively. All patients were followed up for at least 2 years (average, 26 months; range, 24–48 months). The average forearm pronation range was 39° (range, 20°–60°), and the average forearm supination range was 33.2° (range, 10°–60°) at the latest follow-up. Re-ankylosis occurred in one case. An osseous bridge developed between the radius and ulna at the osteotomy site in one case. Radial nerve paralysis developed in two cases and spontaneously resolved 2 months later. Plate breakage was noted in one case 9 weeks postoperatively; however, union was achieved 7 months later.

Conclusion: Mobilization of proximal radioulnar synostosis using a free vascularized fascia lata graft as the interposition graft may prevent re-ankylosis and restore the forearm rotation function, making it a good option for the surgical treatment of CRUS.

Key words: Congenital radioulnar synostosis; Fascia lata; Forearm; Operative procedures; Osteotomy

Introduction

Congenital radioulnar synostosis (CRUS) is a relatively rare malformation in which there is an abnormal connection between the radius and the ulna due to an embryological failure of separation. It is more often bilateral than unilateral. The incidence of CRUS is generally sporadic; however, it may show an autosomal dominant inheritance pattern. At week 5 of gestation, the radius and ulna are...
joined together by a common perichondrium. By week 6, the cartilaginous anlage of the two bones gradually separates. Failure of this longitudinal segmentation and the persistence of the cartilaginous anlage between the radius and ulna during week 7 of development results in a persistent bridge of tissue. CRUS may develop concurrently with other deformities and may be associated with certain syndromes, such as amegakaryocytic thrombocytopenia, Carpenter syndrome, and Holt–Oram syndrome.

The most common presentation of CRUS is the fixation of one or both forearms in a position of pronation, a condition that has been present from birth. It was first reported by Sandifort in 1793, and afterwards only isolated case reports have been described, even after the advent of radiography that makes it much easier for doctors to make the definite diagnosis.

Patients with unilateral CRUS who have a fixed pronation angle of less than 30° can perform daily activities quite well through compensatory movements by the shoulder and wrist joints. Patients with bilateral CRUS or those with a fixed pronation angle of >60° may experience more severe limitations in daily activities, such as difficulty in maintaining personal hygiene and using utensils. Some patients may even be unable to write properly. Consequently, patients may develop psychosocial problems if they grow into adulthood with CRUS. In such severe cases, surgical intervention is recommended. Yammine et al. recommended surgical intervention for cases of hyper-pronation (≥90°) and bilateral synostosis. Simmons et al. stated that pronation angle >60° was an absolute indication for surgery. Furthermore, Simmons et al. and Ogino et al. suggested that fixed pronation of 15°–60° was a relative indication based on the associated functional deficits. We agree with Simmons’s opinion.

Two kinds of surgical methods exist: derotational osteotomy, which was developed to derotate the forearm to a more functional position; and mobilizing procedures, which were designed to reconstruct the proximal radioulnar joint (PRUJ). Theoretically, a mobilization procedure that involves the removal of the synostosis and reconstruction of the PRUJ is a good treatment option. However, high postoperative re-ankylosis rates have been reported; hence, only a few surgeons prefer this method. Derotational osteotomy was first introduced by Green et al., and this procedure could be undertaken at the level of the site of synostosis or distal to it—radius osteotomy alone or radius/ulna osteotomy simultaneously at various levels. Because it is easier to perform and could consistently provide predictable results, derotational osteotomy has become the main weapon in the armamentarium of surgeons. However, derotational osteotomy changes only the position of the forearm and cannot restore the active range of rotation of the forearm. Because of this inherent drawback, surgeons have continued to search for a better mobilization technique. In 1997, Kanaya et al. used a free vascularized adipofascial graft harvested from the arm as an interposition graft combined with radial osteotomy and reported good outcomes with respect to preventing postoperative re-ankylosis. His work reignited the enthusiasm of using a mobilization procedure, and the pedicled vascularized fascial graft harvested from the upper extremity has been designed and reported, which could provide a similar result to Kanaya’s method.

Compared with a free vascularized adipofascial graft harvested from the arm, a free vascularized fascia lata graft is much thicker and may be a better option that could prevent the reoccurrence of synostosis. Furthermore, the anatomy of the fascia lata is quite consistent; it shares the same blood supply with the anterior lateral thigh flap, the anatomy of its nutrient vessel is constant, and its pedicle is long and easy to harvest. Besides, with fascia lata being the interposition graft, the operation time could be shortened as one team separate the synostosis and another harvest the fascia lata at the same time.

Since 2012, we modified Kanaya’s technique by using a free vascularized fascia lata graft as an interposition graft. The purpose of this study was as following: (i) to introduce the details of this modified mobilization technique; and (ii) to report the postoperative outcomes after 2 years’ follow-up.

Methods

This case series was approved by the ethical review board of our institution, and all guardians of the patients provided informed consent. This study was carried out in accordance with the provisions of the Declaration of Helsinki. Inclusion criteria: (i) CRUS patient diagnosed through radiological image; (ii) treated with mobilization procedure using free vascularized fascia lata as the interposition graft; (iii) with a follow-up of at least 2 years. Exclusion criteria: (i) ipsilateral concomitant anomalies of upper limb; (ii) history of ipsilateral forearm surgery for diseases except from CRUS.

Surgical Procedure

Anesthesia and Position

General anesthesia was provided, and the patient was placed in the supine position, with the forearm placed on a well-padded hand table. Two teams, a forearm team and a fascia lata graft team, worked simultaneously.

Approach and Exposure of Forearm

The area of synostosis was confirmed using fluoroscopy and marked on the skin. Two incisions were designed at dorsal and volar part of forearm, respectively.

Dorsal incision: a posterior longitudinal slightly curve incision was made from the lateral epicondyle to a point 1 cm distal to the distal margin of the synostosis. The deep fascia was separated, and the attachment of the anconeus was exposed between the olecranon and proximal part of the ulna. The anconeus was elevated proximally, the periosteum was separated and elevated transversely, and the synostosis was exposed (Figure 1A). An 18/21-G needle was used to mark the distal edge of the synostosis, and the area of
synostosis was confirmed again using fluoroscopy. The dorsal capsule of the PRUJ was incised partially, and the radial head was exposed.

Volar incision: The modified Henry approach was used\(^{12}\), and the incision started at the level of the cubital crease and along the anterior edge of the brachioradialis and the extensor carpi radialis muscles to the distal part of the forearm. The distal edge of the incision was dependent on the planned osteotomy site. Therefore, the distal edge was 1 cm distal to the synostosis if osteotomy was performed at the radial neck and 2 cm distal to the apex of the pronator curvature if osteotomy was performed at the apex of the pronator curvature due to a more severe deformity of the pronator curvature.

The aponeurosis of the biceps brachii was incised, and the biceps brachii was retracted to expose the neurovascular bundles (Figure 1B). The medial nerve, radial artery, ulnar artery, brachial artery, and origin of the posterior interosseous artery was retracted gently. Generally, a transverse branch of the brachial artery that is distal to the elbow joint and runs toward the radial side could be exposed and used as the recipient artery. The radial nerve was retracted laterally and carefully protected. The attachment of the biceps brachii was elevated, and the biceps brachii was freed proximally until it was contractible when pulled distally. The attachment of the brachialis was preserved. The bellies of the brachioradialis and the extensor carpi radialis muscles were retracted radially and dorsally, and the attachment of the supinator and pronator teres were exposed.

Remove of Synostosis and Osteotomy on Radial Shaft
The synostosis and synchondrosis were separated with a high-speed burr and seen using a dorsal approach. The radial nerve and vital structures at the palmer side were protected. The hypoplastic radial head was carefully trimmed, if necessary, taking care not to expose the nucleus of the epiphysis. Trapezoid osteotomy was performed on the radial shaft between the supinator and pronator teres muscles. The shape of the trapezoid was designed according to the degree of dislocation and deviation of the radial head. Computer-assisted designs were always very useful. The radius was fixed with plates and screws. In cases with a high degree of radial head
deviation and wider pronator curvature angle, the apex of “the radial arch” was chosen as the osteotomy site. The range of passive rotation of the forearm was increased after internal fixation. If supination of the forearm was still not satisfactory, the interosseous membrane and the attachment of the pronator quadratus could be released until the range of supination was >60° (Figure 1C–E).

Harvest the Fascia Lata
Before the operation, Doppler ultrasonography was performed at the midpoint of a line between the anterior superior iliac spine and the lateral edge of the patella to track and mark the perforator vessels. The incision was designed according to the location of the main perforator vessels. The surgical technique was similar to the technique used in harvesting an anterior lateral thigh flap. The area of the fascia lata harvested was approximately 5 cm × 8 cm (Figure 1F, G).

Reposition of Tendon Attachments and Interposition of Fascia Lata
The tendinous attachment of the biceps brachii was guided to the dorsal side and sutured to the radial tuberosity using the bone-through method. The anconeus was guided to the volar side and sutured with local fascia tissue. The fascia lata graft was interposed into the interval after removing the synostosis and sutured with the adjacent fascia and periosteal tissue. The graft was unfolded so that the space could be fully interposed (Figures 1H and 2). Arterial anastomosis was then performed by either an end-to-end or an end-to-side approach.

Postoperative Treatment
Postoperatively, an above-the-elbow splint was applied to keep the elbow flexed at 90° and the forearm supinated at 80°. Three days later, a second splint was used to keep the forearm pronated at 80°. Two splints were worn every other day alternatively. At 4 weeks postoperatively, range of motion exercises were initiated both actively and passively, and splints were worn every other night alternatively. The splints were worn for at least 4 months.

Results
General Characteristics
Eleven patients with CRUS (eight males and three females) with an average age of 6.0 (range, 1.5–96.5) years were treated at our department between November 2012 and December 2017. All 11 cases were followed up for an average of 2.2 years (range, 2–4 years). Five bilateral cases (four left forearms and one right forearm) and six unilateral cases (three left forearms and three right forearms) were included. Based on the Cleary and Omer classification⁶, there was one type 2 case, nine type 3 cases, and one type 4 case (Figure 3). Ipsilateral thumb hypoplasia was found in one case; bilateral thumb hypoplasia and cleft palate were also observed in one case. There was no relevant family history, and the average preoperative pronation angle was 67.3° (range, 20°–90°).

Radiographic Findings
The space of the PRUJ could be seen clearly in nine patients, without any signs of recurrence of synostosis. Re-ankylosis occurred in one case. An oblique bone bridge was observed between the radius and the ulna at the osteotomy site in another case. The bone bridge was resected when the plate was removed 9 months after PRUJ reconstruction.

Bone Union and Subjective/Objective Function Evaluation
The radius of 10 cases achieved bone union successfully, and the average union time was 3 months. The average postoperative forearm pronation and supination ranges at the last follow-up were 39° (range, 20°–60°) and 33.2° (range, 10°–60°), respectively. The satisfaction score was evaluated by the parents of six patients. The average score was 4.2, with 5 as “excellent” and 1 as “poor” (Figure 4).

Complications
Plate breakage and questionable nonunion occurred in one case, which was classified as Cleary and Omer type 4¹. This patient underwent fixation with a plate used for metacarpal fractures; definite union was confirmed when the plate was removed 7 months after the initial operation. Radial nerve palsy developed in two cases but resolved spontaneously 2 months postoperatively.
Discussion

Vascularized (free or pedicle) autologous fascia tissue could prevent re-ankylosis, and this has been verified by several studies in addition to Kanaya’s study\textsuperscript{13–15}. Theoretically, using a thicker fascia tissue, such as the fascia lata, could yield better results and prevent re-ankylosis. The fascia lata shares the same blood supply with an anterior lateral thigh flap, the anatomy of the nutrient vessel is constant, and its pedicle is long and easy to harvest\textsuperscript{16}. In addition, two surgical teams can work at the same time on the forearm and the thigh; this may lead to a shorter operation time compared to the time required for the procedure described by Kanaya \textit{et al.}\textsuperscript{16}.

Lesser Trauma and Shorter Operation Duration

The shorter operation duration often means lesser trauma to the patient. In this study, 11 patients were treated with technique mentioned above, and the procedure was performed by two teams of surgeons simultaneously to achieve a minimum operative duration.

Better Donor Site Appearance and Good Post-Operative Function

This technique use fascia lata as interposition, and no skin graft was harvest at thigh. Carefully closing the incision with plastic surgical technique, the patient can have a more cosmetic acceptable appearance at donor site. Also, the postoperative forearm function can be satisfying, as shown in this study. With an average active rotation range of 72.2° (39° for supination and 33.2° for pronation) after at least 2 years of follow-up, the patients can perform most of their daily activities. And for patients’ parents, most of them were satisfied with the outcome of this procedure.
Tricks and Pitfalls During Treatment
The most annoying complication of this technique is the re-ankylosing of the PRUJ, which is the same as previous studies. Though the fascia lata should have a better result in protecting the articular space theoretically and this study shows the same result, there is always a risk of graft necrosis in all the free vascularized tissue transplantation techniques. An experienced team is the guarantee of a successful operation. If necessary, a monitor skin flap is helpful.

The soft tissue contracture is another annoying problem in CRUS patients. So, appropriate release of aponeuroses and ligaments can be helpful to achieve a better forearm rotation range. The postoperative rehabilitation should be taken into a very important position during the treatment to enhance the effect that soft tissue release brings. Even after soft tissue release and rehabilitation, the forearm soft tissue can still become contractual due to the innate development of CRUS, and a steady follow-up should be maintained. Besides, the compartment syndrome should be noticed at the donor site: after harvesting the fascia lata, leave the residual fascia and do not try to fix it.

Further Problems to Be Solved
The crucial point of any remobilizing procedure is to avoid postoperative re-ankylosis. However, for severe deformities, many secondary problems may occur along with synostosis, such as radial head deformation and dislocation, absence of supinatory curvature, increased pronator curvature, inner rotation of the radius and ulna, and dislocation of the distal radioulnar joint\(^\text{13}\). As mentioned above, the soft tissue of individuals with CRUS can also be abnormal with shortened interosseous membranes, absent annular ligaments, and hypoplasia of the supinator and pronator teres muscles. The release of soft tissue can be essential to some extent.

Limitations in this Study
There are still some limitations in this study. First, the reconstruction of humeroradial joint was still not ideal, especially in patients with a posteriorly dislocated radial head. Second, most of the patients in this study had type 3 CRUS, and there was only one patient with type 2 CRUS and one patient with type 4 CRUS; thus, we did not separate the cases into further groups according to the Cleary and Omer classification. Moreover, whether there is difference in postoperative range of motion between different types was uncertain. To achieve better outcomes in future research and clinical works, computer-assisted designs should be used to preoperatively evaluate bony and soft tissue deformities and assist with the operation, thereby further improving the outcome of the procedure. A controlled trial study will also be essential in order to compare the outcomes of derotational osteotomy and different mobilization techniques.

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