Shoulder and Elbow Tips and Tricks: Loss of Elbow Flexion in Congenital Arthrogryposis Treated with a Bipolar Latissimus Transfer

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Introduction

Arthrogryposis multiplex congenita is a rare disorder characterized by the presence of multiple congenital limb contractures. These contractures present most often as deficits in passive and active range of motion [ROM] with coexistent structural and/or functional abnormalities of surrounding soft tissue anatomy[1]. There are many etiologies for this presentation however the clinical presentation is the end product of decreased fetal movement leading to multiple joint contractures in utero with increased collagen proliferation, the fibrotic replacement of muscle and a marked thickening of joint capsules[2,3].

It has been reported in the literature previously that up to 72% of patients with arthrogryposis have upper extremity involvement with 25% of them involving the elbow. Other studies report a 59%–92% elbow involvement[3-5]. Of those with elbow involvement the most common presenting deformity is extension contracture of the elbow, absent skin creases and atrophy of the involved limb[1-5]. The elbow joint is thought to be the most critical for allowing the performance of activities of daily living which involve the upper extremity including self-feeding, self-care of the face and hair and independent toileting[6]. As such, restoration of biceps dysfunction is critical in improving the quality of life in arthrogrypotic patients with elbow involvement and decreased ability to care for themselves.

Though non-operative measures consisting of range of motion exercises and splinting are effective management for the majority of children when these measures fail there are several surgical techniques available to restore active elbow flexion in these patients[7]. Most discussed are the Steindler flexorplasty, a triceps-to-biceps transfer, pectoralis major transfer, and the latissimus dorsi transfer.

We chose to use a bipolar latissimus transfer as it has a large, mobile and robust vascular pedicle with minimal donor site morbidity. The purpose of this report is to present a case of a 20-year-old man with congenital arthrogryposis and elbow extension contracture treated with a bipolar latissimus transfer to restore active elbow flexion to the right upper extremity. 7 months out form surgery the patient achieved active ROM of the elbow up to 117 degrees and reports being pain-free a majority of the time.

Statement of Informed Consent

The patient presented in this report was informed that data concerning the case would be submitted for publication and provided consent.

Case Report

A 20-year-old male with congenital arthrogryposis presents to the office with right elbow flexion weakness 0/5. His wrists are bilaterally flexed and ulnar deviated with attendant restrictions in ROM. He had previously been able to use his left upper extremity for all self-care including washing hair, brushing teeth and shaving, but his ROM has regressed in that arm as well. His passive right elbow arc of rotation is from 0 to 110 degrees. An MRI of the right latissimus was obtained to determine its suitability for transfer. Results showed minimal intramuscular fat without significant muscular atrophy. To achieve active elbow range of motion the patient elected to perform a right upper extremity pedicle latissimus dorsi myocutaneous flap transfer.

The patient was placed in a lateral decubitus position. An oblique skin paddle was designed and then incised over the center of the right latissimus dorsi muscle. Skin and subcutaneous tissue were divided, and the anterior border of the latissimus was identified. The entire muscle was taken with dissection performed to its insertion on the humerus (Figure 1). The insertion was detached with the tendinous portion of the latissimus intact with the pedicle transfer. The circumflex scapular vessel and serratus branches were taken down to isolate the muscle on its neurovascular pedicle which included the thoracodorsal nerve, artery and vena comitans

Figure 1. Development of latissimus skin paddle and dissection.

(Figure 2). When the muscle was completely isolated a tunnel was placed between the posterior chest wall and the anterior axilla for the neurovascular pedicle. Checkpoint nerve stimulator was used to confirm intraoperative contraction.

A deltopectoral approach was then utilized proximally. Distally at the elbow a Z-incision was made to prevent contracture and allow exposure of the ulna. Proximally the conjoint tendon and proximal and distal aspects of the pectoralis major muscles were isolated. We tunneled underneath the pectoralis major staying close to its insertion point to identify the remnant long head of the biceps tendon. The tendinous portion of the latissimus was placed through this tunnel with capacious space ensuring no pressure would be put on the pedicle as it came anterior to the chest wall. Distally the best location for the transfer was identified to be laterally at the subcutaneous border of the ulna. Suture anchors were placed in the ulna for distal attachment. The proximal portion of the tendon was reattached to the tip of the coracoid with a suture anchor (Figure 3). This anchor was then double loaded, and the tendon was reefed back onto the tip of the coracoid for a second attachment. It was reinforced with additional sutures sewn along the conjoint tendon and the graft allowing for multiple points of fixation.

Figure 2. Latissimus isolated on its neurovascular pedicle; tunnel development.

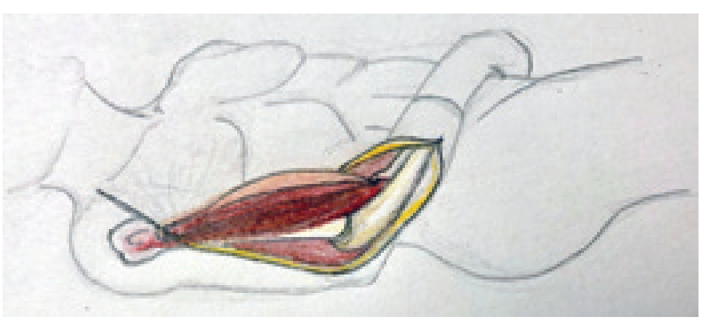

Figure 3. Distal attachments to ulna, proximal attachments to coracoid.

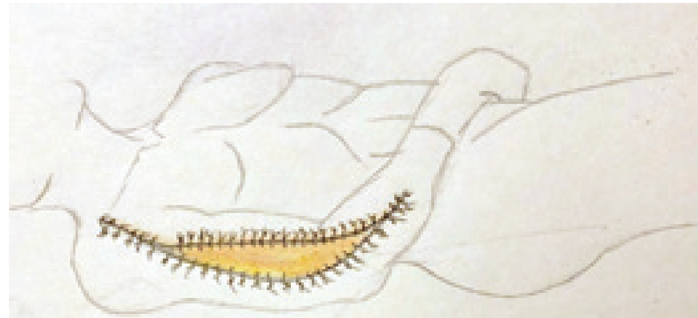

Figure 4. Closure of extended deltopectoral and distal z-shaped incisions.

The latissimus had been marked in-situ 5 cm apart to create proper tensioning. This was recreated when the muscle was transferred anteriorly. Extended deltopectoral approach and distal z-shaped incisions were closed (Figure 4).

The patient was taking to the intensive care unit for flap monitoring with his right upper extremity immobilized in a posterior slab at 90 degrees of flexion. At his post-operative day two visit the flap was warm, and well-perfused with grossly intact elbow flexion. For the first four weeks only passive ROM from 90-130 degrees was permitted at which point active physical therapy began. At approximately three weeks post-op the patient developed a mild cellulitis at the donor site which resolved without issue with oral antibiotics. By three months post-op the patient was able to volitionally flex the right elbow.

At most recent follow-up 7 months post-op the patient was able to achieve full elbow extension and 117 degrees of active flexion. Passive ROM was up to 130 degrees. On the Manual Muscle Testing Grading system his strength in the right upper extremity was 4-/5 in flexion.

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