

Case Report

Radial club hand management with initial gradual distraction by Ilizarov

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ABSTRACT

Introduction: Radial club hand is a radial longitudinal deficiency, clinically characterized by shortening and bowing of the forearm and radial deviation of the wrist. Acute correction of the wrist during centralization became very difficult in severe cases. Ilizarov external fixator offers a solution through gradual distraction for safe and effective correction of severely stiff wrist radial deviation initial to ulnar centralization. We present a case of successful correction of unilateral radial club hand in one-year period of treatment.

Methods: We report a 5-year-old boy with unilateral radial club hand. He had shortened and bowed ulna, severe stiffness of wrist radial deviation which failed to be stretched with serial casting, and lack of hand function. Ilizarov external fixation was used for gradual distraction at 1-2 mm per day until the wrist looked straight. The procedure was followed by centralization as well as ulnar osteotomy for correcting ulnar bowed and fixed with K-Wire through carpus and second metacarpal for three months.

Results: In the one year follow up, the ulnar was still centralized with no neurovascular impairment and there was improved hand function. The original technique for severe shortening of the concavity structures requires extensive soft-tissue release during the centralization procedure. It is associated with carpal resection, leading to further shortening of an already short forearm. Ilizarov external fixation is used to make the stretching process by gradual distraction of radial aspect safer and more effective before ulnar centralization.

Conclusion: Gradual distraction by Ilizarov external fixator is a good option for safe and effective stretching of severe stiffness of wrist radial deviation prior to ulnar centralization.

ABSTRAK

Pendahuluan: Radial club hand adalah suatu kelainan longitudinal pada radius yang ditandai dengan pemendekan serta pembengkokan lengan bawah serta deviasi sendi pergelangan. Koreksi akut pada sendi pergelangan akan menjadi sangat sulit di beberapa kasus. Fiksasi eksternal Ilizarov menawarkan solusi melalui distraksi gradual dalam penatalaksanaan deviasi radius dengan kekakuan pergelangan berat sebelum prosedur sentralisasi ulna. Kami menyajikan keberhasilan koreksi kasus pada radial club hand unilateral dalam setahun penatalaksanaan.

Metode: Kami melaporkan pasien laki-laki berusia 5 tahun dengan radial club hand yang disertai masalah pemendekan dan pembengkokan ulna, kekakuan berat pada pergelangan yang gagal diregangkan dengan serial casting, dan berkurangnya fungsi tangan. Fiksasi eksternal Ilizarov digunakan untuk distraksi gradual 1-2 mm per hari hingga pergelangan terlihat lurus. Prosedur ini diikuti oleh sentralisasi dan osteotomi ulna untuk memperbaiki pembengkokan dan difiksasi menggunakan K-wire melalui carpus dan metacarpal ke-2 selama 3 bulan.

Hasil: Setelah 1 tahun, ulna masih tersentralisasi tanpa gangguan neurovascular dan fungsi tangan. Teknik yang umum digunakan umumnya memerlukan prosedur jaringan lunak yang ekstensif sehingga sering berakhir dengan reseksi karpal, yang menyebabkan pemendekan lebih jauh lagi. Fiksasi eksternal Ilizarov digunakan untuk memastikan peregangan bertahap radius lebih aman dan efektif, sebelum prosedur sentralisasi ulna.

Kesimpulan: Distraksi gradual menggunakan fiksasi eksternal Ilizarov merupakan opsi yang baik dan efektif untuk peregangan wrist radial deviation dengan kekakuan berat, sebelum dilakukan sentralisasi ulna.

Keywords: Radial club hand, severe stiffness, wrist radial deviation, gradual distraction, Ilizarov, external fixation

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INTRODUCTION

Radial club hand is a severe radial longitudinal deficiency, a congenital deformity of the upper extremity with a spectrum of hand and forearm anomalies ranging from mild radial hypoplasia to complete absence of the radius,¹ clinically characterized by shortening and bowing of the forearm and radial deviation of the wrist.² It is a relatively rare deformity with an incidence of 0.5 per 10,000 live births.³

Centralization is one of the approved options for treating severe radial club hand.⁴ However, acute correction of the wrist during centralization become very difficult in severe cases.⁵ Manual soft tissue stretching, even with serial casting, often turn into complications and incomplete correction. By performing extensive soft tissue release, shortened forearm will become even worse. Ilizarov external fixator offers a solution through gradual distraction for safe and effective correction of severely stiff wrist radial deviation initial to ulnar centralization. We presented a case of successful correction of unilateral radial club hand in one year treatment. This paper represents our experience in treating it using gradual distraction by Ilizarov external fixator.

METHODS

We report a 5-years-old boy with unilateral radial club hand. There was radial inclination of the hand, associated with palmar flexion-pronation. Along with severe esthetic problem, radial club hand caused functional deficits because of short forearm, unstable wrist and reduced extrinsic tendon course (see Figure 1). There was no family history of congenital anomalies. Previously, the patient had never undergone any procedure other than casting for six times.

Ilizarov external fixator was applied to the patient for gradual postoperative deformity correction. Firstly, two pins were inserted into the proximal and distal metaphysis. A 1 cm skin incision was made over entry site to insert each pin. Moreover, a track was created by careful blunt dissection down to the cortex of the ulna. Using a drill guard to protect the soft tissues, a pin was then drilled through both cortices of the ulna but only advanced slightly through the deeper cortex. The tip of the pin then could be located by palpation and a track safely through the skin incision using blunt dissection. A drill guard was placed over the tip of the pin so that the

pin could be advanced safely through the skin incision and soft tissues, protecting the important neurovascular structures, which may have an abnormal anatomical location in a radial club hand. The left thumb was also amputated.



Figure 1. Clinical appearance of patient's left radial club hand (left) and plain x-ray before surgery (right).

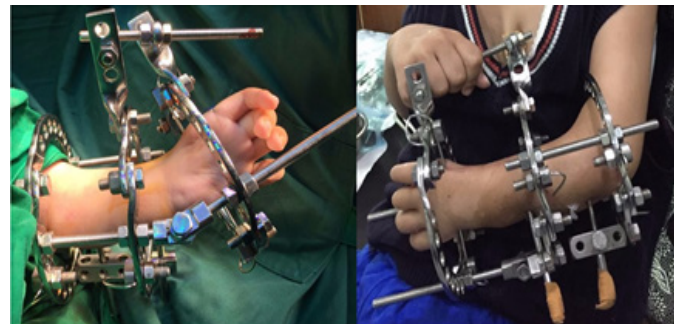


Figure 2. Post-operative (left) and during gradual correction by Ilizarov external fixator (right).



Figure 3. Physical examination showed deformity, lack of hand function, joint stiffness and radial deviation contracture.

In the proximal and distal metaphysis, two pins were passed through the ulna at an angle of approximately 90° to each other. The ulna was divided at the level of the distal metaphysis. One full fixator ring was then applied to each pair of pins and the rings linked using connecting rods with hinges included.

After the procedure, we performed osteotomy and preserved a periosteal sleeve as advocated by Ilizarov

(1990). No distraction was performed during the surgery. The patient was admitted for 24 to 48 hours for pain medication and to instruct the parents on pin site care as well as elbow and finger exercises. Lengthening was commenced approximately one week after application of the device at a rate of 1 mm per day in four increments. The patient was followed-up at monthly intervals subsequently to assess progress, checking in particular for evidence of pin track infection, loss of motion in the elbow, wrist or digits, and pain. At this point, the wrist already looked straight but still unstable on the radial side (see Figure 3).

The problems consisted of shortening and bowing of ulna, severe stiffness of wrist radial deviation, which failed to be stretched with serial casting, and lack of hand function. Ilizarov external fixation was used for gradual distraction at 1-2 mm per day for three weeks (see Figure 2). The distraction was done to get a satisfactory forearm length and correcting the deformity until the wrist looked straight. The fixator was not changed for several days before centralization procedure, allowing the soft tissue to relax. The fixator was removed afterwards. In one year follow up, ulnar was still centralized, no neurovascular impairment and hand function was improved. Particularly, the patient could start to grasp objects, although not fully.



Figure 4. Centralization with ulnar osteotomy.



Figure 5. One year follow up after gradual correction by Ilizarov external fixator.

RESULTS

Severe radial club hand is hard to treat. The main objective is to stabilize the carpus in alignment with the forearm in order to improve not only the esthetics but also the flexor digitorum strength. Variable treatment methods have been offered for treating radial club hand. Some treatments have proven to achieve long-lasting alignment of the hand. Those treatments replace the radius by vascularized epiphysis transfer from a toe, radialization positioning of the carpus facing the distal ulna, or centralization.⁶

The original technique for severe shortening of the concave structures requires extensive soft-tissue release during the centralization procedure. It is associated with carpal resection for creating a notch to be the house of the ulnar head, leading to further shortening of the existing forearm.⁶

On the other hand, inadequate soft tissue release may result in distal ulnar bowing, intercarpal fusions, carpometacarpal angulations, and recurrence of deformity. Correction by percutaneous techniques is becoming more popular. Unilateral external fixators have been used for pre-centralization soft tissue stretching, but they cannot correct both the radial deviation and flexion deformities.

The Ilizarov external fixation is a better option than other unilateral external fixators with more comprehensive and nearly physiologic distraction in addition to ulnar lengthening. For patients with unilateral RLD, the deformity is prominent. In bilateral cases, short forearms may impair the function of hands. The Ilizarov external fixator can manage both problems.

Ilizarov external fixation is used for making the stretching process of the radial aspect safer and more effective prior to ulnar centralization. Supportive casting after centralization is an easy and effective method to obtain soft tissue stretching instead of extensive release of the soft tissue. Moreover, it helps to put the limb in a correct position since the beginning so that the hand function will be developed in correct position as well. It will make surgical treatment less extensive.

Summary

Gradual distraction by Ilizarov external fixator is a good

option for safe and effective stretching of severe stiffness of wrist radial deviation prior to ulnar centralization and can avoid an extensive surgical procedure that endangers the important structures. However, surgeon knowledge of detailed technique and patient compliance are needed for achieving good outcome.

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