

LONGITUDINAL PREAXIAL UPPER LIMB DEFICIENCY DISORDERS; GENETIC AND SURGICAL ASPECTS

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ABSTRACT

Introduction: Congenital Longitudinal radial deficiency, also called radial club hand or radial dysplasia, is an uncommon congenital anomaly characterized by various degrees of deficiency along the preaxial or radial side of the extremity. The severity of deformity ranges from mild thumb hypoplasia to complete absence of the radius. Longitudinal preaxial upper limb deficiency may occur in isolation but frequently is associated with other congenital malformations e.g. VACTERL, Holt-Oram, Thrombocytopenia with absent radius (TAR), Fanconi anemia. The management of congenital radial club hand has undergone significant changes. Numerous procedures and modifications have been developed to treat this condition. **Aim:** to discuss the genetic aspects, associated conditions, classification of these deformities and the management of such cases particularly the staged protocol for management of Bayne type IV and pollicization for hypoplastic thumb. **Methods:** 7 cases diagnosed clinically as congenital radial deficiency; 3 cases were classified as Bayne type IV congenital radial deficiency; two patients were classified as Bayne type N congenital radial deficiency while the remaining two cases were Bayne type 0. Five patients were boys, two were girls. The average age at the time of operative intervention was 12 months (range 10–15). All cases had complete physical examination, cardiac auscultation, whole skeletal radiographs, complete blood count with a peripheral blood smear and Cytogenetic studies including G-banding technique, DEB and MMC and chromosomal studies. we used a 2 stage approach for 3 cases of Bayne Type IV congenital radial deficiency, pre-centralization soft tissue distraction using light weight fibro-carbon ring external fixator (Ilizarov) followed by centralization procedure the remaining four cases of Bayne Type N and 0 deformities had hypoplastic thumbs which are further classified according to Buck-Gramcko modification of Blauth classification into: 3 cases of Grade IV (floating thumb) and one case of Grade V (total thumb aplasia) was treated by pollicization. **Results:** The preoperative workup showed that all these cases were isolated non syndromic unilateral radial deficiency without clinical or radiological evidence of associated anomalies or Fanconi's anemia. Improvements of wrist position parameters were noted associated with improvement in the measurements of radiographic construction lines during follow up evaluations for Bayne Type IV cases managed by the staged approach. Our results of thumb reconstruction by pollicization are uniformly good as regard to growth and mobility. Venous congestion was the most important pos-operative complication. **Conclusion:** by using the staged approach for type IV deformities, we can safely distract the taught soft tissues and avoid several of the difficulties often seen when centralization not proceeded by soft tissue distraction. Also, significant improvement in deformity with a more functional position of the hand can be achieved while minimizing overall complications. The aesthetic and functional results of pollicization are directly related to the transposed index digit and surrounding musculature. Pollicization of the index finger for congenital thumb hypoplasia is a well-accepted procedure with good outcome.

Key words: Longitudinal radial deficiency, radial club hand, soft-tissue distraction, centralization, thumb hypoplasia, pollicization.

INTRODUCTION

Congenital Longitudinal radial deficiency, also called radial club hand or radial dysplasia, is an uncommon congenital anomaly characterized by various degrees of deficiency along the preaxial or radial side of the extremity. The severity of deformity ranges from mild thumb hypoplasia to complete absence of the radius. Longitudinal preaxial upper limb deficiency may occur in isolation but frequently is associated with other congenital malformations e.g. VACTERL, Holt-

Oram, Thrombocytopenia with absent radius (TAR), Fanconi's anemia. ^[1]

Bayne and Klug classified radial deficiency into four types based on radiographic severity of the radius deficiency. ^[1] James and associates have modified Bayne's classification to include patients with deficiency of the thumb or carpus in the presence of a radius of normal length. This scheme is used in combination with the modified Blauth scheme for classifying thumb hypoplasia. ^[2]

Treatment indications vary with the severity of the deformity, the age of the patient and the associated anomalies. The objectives of treatment in radial club hand are to reduce the functional deficit incurred by a short or absent radius, a short ulna, an abnormal muscular anatomy, radial deviation of the wrist and hypoplastic thumb.^[3]

PATIENTS AND METHODS

This prospective case series was done in plastic surgery unit of Zagazig University Hospitals between December 2008 and December 2012. It was conducted on 7 consecutive patients of congenital radial deficiency. The study was approved by the local institutional review board, and written informed consent was obtained from the patients' parents.

The average age at the time of operative intervention was 12 months (range 10–15). Five patients were boys, two were girls. Four right and three left extremities were involved. Three cases were classified as Bayne type IV congenital radial deficiency (figure 1); two patients were classified as Bayne type N congenital radial deficiency (figure 5)

while the remaining two cases were Bayne type 0 (table 1).

Preoperative workup was directed toward the detection for associated conditions -specially the most devastating associated condition "Fanconi's anemia" - which necessitates referral to pediatric sub-specialists, Genetic counseling and chromosomal studies. All cases had complete physical examination, cardiac auscultation, whole skeletal radiographs, complete blood count with a peripheral blood smear and Cytogenetic studies including G-banding technique, DEB and MMC and chromosomal studies.

Preoperative clinical and radiological assessment is carried out to determine the amount and type of deformity. Plain radiographs are obtained to evaluate the degree of radial aplasia and to assess associated abnormalities of the elbow, wrist, and hand in particularly the thumb. The degree of ulnar bow is calculated from the lateral radiograph as the angle between the proximal and distal ulna.

Table (1): Patients' Characteristics, Clinical Features and brief Operative Data

Patient	Sex	Side	Deformity	Associated Diagnosis	Pre-op Distraction	Definitive procedure	Age at Definitive procedure
1	Male	Left	Bayne Type IV	-	Ring external fixator at age of 11 months	centralization	13 months
2	Male	Right	Bayne Type IV	-	Ring external fixator at age of 13 months	centralization	15 months
3	Female	Left	Bayne Type IV	-	Ring external fixator at age of 10 months	centralization	12 months
4	Male	Left	Bayne Type N	-	-	pollicization	12 months
5	Female	Right	Bayne Type N	-	-	pollicization	10 months
6	Male	Right	Bayne Type 0	-	-	pollicization	12 months
7	Male	Left	Bayne Type 0	-	-	pollicization	14 months

correction of Bayne Type IV deformities was based on a staged protocol which starts on the initial visit as the parents were instructed to passively stretching of the taut radial structures, the wrist and elbow. This was followed by soft tissue distraction using light weight fibro-carbon ring external fixator (Ilizarov) (figure 2). On a later stage, wrist

centralization was done. After constructing an appropriate sized distraction (Ilizarov) unit, no acute correction at time of surgery was done. The family was instructed to begin distraction approximately 7 days after surgery at the rate of 1 mm per day, divided in two installments of 0.5 mm each. The rate was adjusted according to discomfort and

swelling of the hand. Physiotherapy including range-of motion exercises for the fingers and elbow

and dynamic slings to keep the fingers in extension was instituted.



Figure (1): Bayne type IV congenital radial deficiency (Left); the wrist is severely flexed radially due to the lack of structures on the radial side of the forearm and hand, X-ray (Right) shows complete absence of the radius and radial 2 rays.

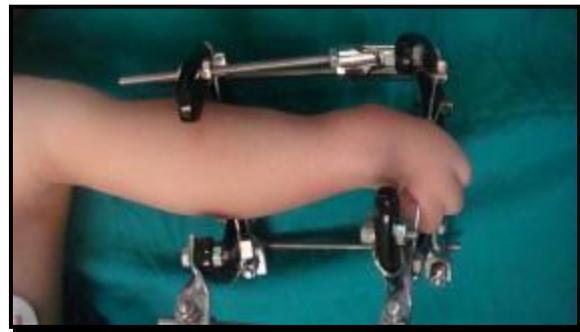


Figure (2): (Left) The fully constructed distraction unit is connected to the 2 rings and dual sloppy hinge is constructed on either end of a distraction rod, (Right) shows The final soft tissue distraction achieved by the previously constructed unit.

Approximately 6-8 weeks after achieving the desired position of the hand and wrist, with an attempt at slight overcorrection, the Ilizarov apparatus was removed and the extremity was placed in a long-arm cast with the wrist placed in the maximally corrected position. Alternatively, we removed the apparatus immediately prior to centralization.

Radiographic evaluations at regular intervals were done. Two measurements according to construction lines were recorded. The hand-forearm angle (HFA) which is the intersection angle between the long axis of the long finger metacarpal and the distal ulna and the hand-forearm position

(HFP) refers to the distance in millimeters between the base of the little finger metacarpal and the distal ulna.

The goal of centralization is to centralize the ulna, to stabilize the carpal bones and to correct angulation. We used dorsal S-shaped incision from the dorsum of the hand to the distal third of the forearm (figure 3). The fibrotic muscle mass attached to the carpus, including extensor carpi radialis and flexor carpi radialis, was dissected free. The extensor retinaculum is raised as a radially based flap. The wrist capsule is incised in a transverse direction over the ulnocarpal joint to expose the distal end of the ulna. The ulna is freed

from its soft tissue attachments distal to the physis, and the carpal bones are detached from the palmar capsule (figure 3). The carpus is then reduced onto the distal ulna. If reduction cannot be achieved, limited shaving of the carpus and distal ulna was done while avoiding injury to the growth plate in order to stabilize the carpus on the ulna. Then extensor carpi ulnaris (ECU) was advanced and shortened distally if redundancy was noted to

increase soft tissue tension, and if present, the radial wrist extensors are transferred into the extensor carpi ulnaris tendon. After confirming proper alignment of the carpus over the distal ulna, a Kirschner wire (figure 4) was placed in a retrograde fashion through the ulna and then passed antegrade into the index or long metacarpal (sometimes referred as radialization).



Figure (3): (Left) dorsal S-shaped incision was extended from the dorsum of the hand to the distal third of the forearm. (Right) The ulna is freed from its soft tissue attachments distal to the physis, and the carpal bones are detached from the palmar capsule. Limited shaving of the carpus and distal ulna was done while avoiding injury to the growth plate in order to stabilize the carpus on the ulna.



Figure (4): Kirschner wire was placed, the wrist capsule is imbricated and the wound is closed.



Figure (5): Skin incision; Buck-Gramcko design. (Right) dorsal view, (Left) palmar view.

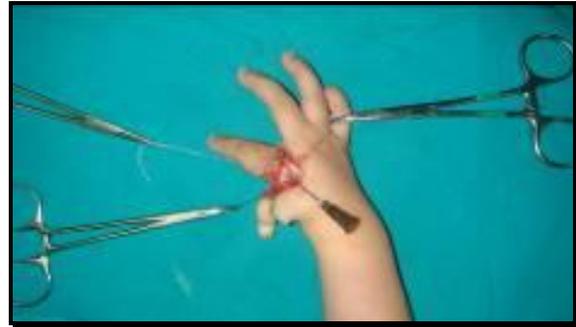


Figure (6): (Left) The first palmar and dorsal muscles are elevated from the index metacarpal and MP joints, released with a portion of the extensor hood and marked. (Right) The index finger is shortened by removal of the metacarpal bone while leaving its base and epiphysis intact.



Figure (7): (Left& Right) double-ended Kirschner wire is passed through the metacarpal head and into the proximal phalanx exit at the proximal phalanx. Note the index finger is positioned in 45 degrees of abduction and between 100 and 120 degrees of pronation.

The remaining 4 cases of Bayne Type N and 0 deformities had hypoplastic thumbs which are further classified according to Buck-Gramcko modification of Blauth classification into: 3 cases of Grade IV (floating thumb) and one case of Grade V

The procedure involved transfer of the index finger with its neurovascular pedicle, adjustment of the bones (shortening, rotation, and abduction), and muscular stabilization. We used the most popular skin incision developed by Buck-Gramcko (figure 5). After dissection, identification and isolation of the common digital vessels and nerve to the second web space, the proper digital artery to the long finger is ligated to allow tension-free pollicization of the index finger. 1st annular pulley is incised. Careful identification and isolation of the dorsal veins is a critical precaution. We do not shorten the extensor or flexor tendons. The first palmar and dorsal muscles are elevated from the index metacarpal and MP joints. The radial and ulnar lateral bands are defined for later transfer of the first dorsal and first palmar tendons, respectively (figure 6).

(total thumb aplasia) were treated by index finger pollicization. They were 3 boys and one girl; 2 right and 2 left) carried out by the same surgical team. Pollicization was done at a mean age of 12.5 months (table 1).

The index finger is shortened by removal of the metacarpal bone while leaving its base and epiphysis intact. Physeal ablation (epiphysiodesis) prevents excessive growth of the index pollicization. Placement of the index MP joint in hyperextension will prevent any additional CMC joint extension after pollicization. A double-ended Kirschner wire is passed through the metacarpal head and into the proximal phalanx. The index finger is manually shortened and rotated into position. The index finger is positioned in 45 degrees of abduction and between 100 and 120 degrees of pronation to recreate normal thumb alignment. The Kirschner wire is driven retrograde across the metacarpal base and into the carpus. The first web space is configured with skin devoid of suture (figure 7).

Postoperative dressing and a long arm splint are crucial with the elbow flexed to greater than 100

degrees decreases the chance of inadvertent removal. The arm is elevated to promote venous drainage and is monitored overnight.

RESULTS

In our study, all the patients did not have associated malformations in other body organs or systems or Fanconi's anemia. Cases were all sporadic with no familial occurrence. No parental consanguinity was found. There wasn't any history of environmental exposure to the mothers during pregnancy. Exclusion of Fanconi's anemia was based on results of cytogenetic techniques and chromosomal studies (Giemsa staining and induction of chromosomal breakage by DEB at the National Research Center-Human Genetics and Human Genome Division - Cairo. None of the cases showed any aberration or the typical breaks characteristic for Fanconi's anemia i.e. isochromatid exchange giving triradius and quadradius figures on

DEB testing. They were all DEB –ve thus excluding a diagnosis of Fanconi anemia.

The results of soft tissue distraction and centralization for Bayne type IV deformities were followed through the removal of each device and after centralization (figure 8). The amount of distraction averaged 17 mm (range, 15–20 mm). The average duration of distraction was 55 days (range 46 to 62 days) (table 2). Wrist extension improved from an average of 25° of wrist flexion before surgery to an average of 16° of wrist extension after centralization for an average improvement of 41° (table 3). The resting wrist radial deviation posture averaged 100° before surgery and improved to an average of 26° after centralization (table 4). Before surgery the wrists averaged 73° of passive ulnar deviation and improved to 20° after centralization (table 5). The finger range of motion had recovered to almost preoperative baseline values.

Table (2): Distraction Achieved With Ring Fixator

Patient	Sex	Side	Distraction Duration (Days)	Distraction Achieved (mm)
1	Male	Left	62	20
2	Male	Right	46	15
3	Female	Left	56	17
Average			55	17

Table (3): Clinical Examination, Wrist extension (°)*

Patient	Sex	Side	Before Surgery	After Surgery
1	Male	Left	-45	10
2	Male	Right	0	25
3	Female	Left	-30	15
Average			-25	16

*Negative values denote flexion.

Table (4): Clinical Examination, Resting Radial Deviation (°)

Patient	Sex	Side	Before Surgery	After Surgery
1	Male	Left	105	35
2	Male	Right	95	20
3	Female	Left	100	25
Average			100	16

Table (5): Clinical Examination, Passive Ulnar Deviation (°)†

Patient	Sex	Side	Before Surgery	After Surgery
1	Male	Left	-85	-20
2	Male	Right	-90	-25
3	Female	Left	-45	-15
Average			-73	-20

†Negative values denote radial deviation.

We found that soft tissue distraction allowed the centralization to be performed without difficulty. There was no untoward affect of the distraction process on the radial soft tissues. In all cases the carpus was placed easily on the end of the ulna without tension and without the need for bony excision or carpal notching. All patients tolerated the distraction process well. One patient developed pin-track infections at the end of the distraction period which was managed conservatively by antibiotics and local dressings. There were no cases of neurovascular injury, wound infection, osteomyelitis, or growth plate damage. No patient lost any significant finger, wrist, or elbow mobility following completion of the treatment protocol.

Depending on radiographic examinations (figure 9) at an average follow-up of 14 months (range 12–16), the average preoperative hand-forearm angle of 96 degrees (range 86 to 108 degrees) improved by 78 degrees to a postoperative average of 18 degrees (range 14 to 25 degrees) (table 6). The average preoperative hand-forearm position of -18 mm (range -16 to -20 mm) improved by 34mm to a postoperative average of +16 mm (range +15 to +18 mm) (table 7).

Table (6): Pre- and Postoperative Hand–Forearm Angle (HFA)

Sex	Side	Age at Soft tissue distraction (mo)	Age at centralization (mo)	Follo w up (mo)	Preop HFA (°)	Postop HFA (°)	Correction HFA (°)	
1	Male	Left	11	13	12	95	25	70
2	Male	Right	13	15	16	108	17	97
3	Female	Left	10	12	14	86	14	72
Average			11	13	14	96	18	79

Table (7): Pre- and Postoperative Hand–Forearm Position (HFP)

Patient	Sex	Side	Age at Soft tissue distraction (mo)	Age at centralization (mo)	Follow up (mo)	Preop HFP (mm)	Postop HFP (mm)	Correction HFP (mm)
1	Male	Left	11	13	12	-18	+15	33
2	Male	Right	13	15	16	-20	+16	36
3	Female	Left	10	12	14	-16	+18	34
Average			11	13	14	-18	+16	34

Our results of thumb reconstruction by pollicization are uniformly good. The results for sensitivity were satisfactory. Mobility was good, in fact, if mobility of the new MP was consistently good. Growth was good in all cases. The aesthetic and functional results of pollicization are directly related to the transposed index digit and surrounding musculature (figure 10). We found that a mobile index finger transferred to the thumb position will provide stability for grasp and mobility for pinch. In two cases with stiff index fingers, pollicization resulted in stable thumbs for gross grasp but there were deficiency in pinch.

There were 3 complications (table 8) in the postoperative period, including one case of marginal skin necrosis, 2 cases of venous congestion. There were no recognized complications related to arterial insufficiency, excessive bleeding, or nerve injury.

Table (8): Perioperative complications

Complication	No. of cases	%
Marginal skin necrosis	1	25
Venous congestion	2	50
Wound infection	0	0
Arterial insufficiency	0	0
Excessive bleeding	0	0
Nerve injury	0	0

Scar contracture of the web space along the incision was noted in 2 cases, resulting in adduction of the pollicized digit. In one case, the adduction was mild and did not require surgery. In the other case, function was sufficiently compromised necessitates surgical release by z-plasty which resulted in adequate correction. We were not able to accurately evaluate the functional efficiency of the pollicized digit because of the relatively short follow up duration and the young ages of the children.



Figure (8): (Left) Bayne type IV congenital radial deficiency, preoperative view, (Right) late follow up (13 months).



Figure (9): (Left): preoperative radiographic view of the deformity, showing hand-forearm angle of 86° and hand-forearm position of -16 mm, (Center& Right): Serial radiographic follow up views showing improvements of the previously mentioned measures.

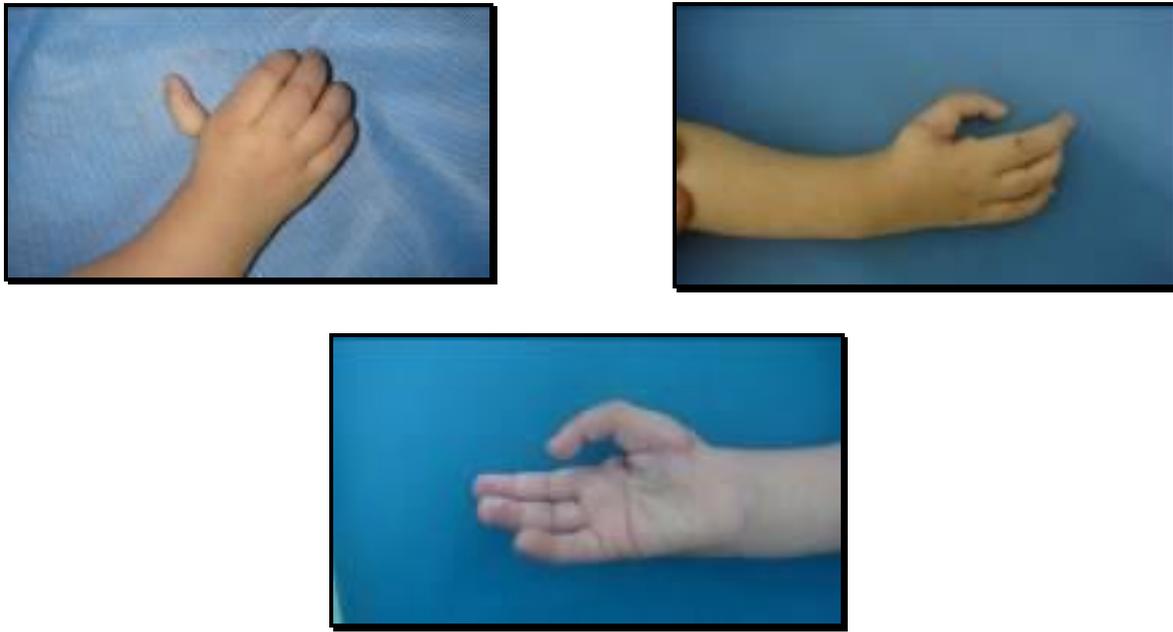


Figure (10): (Left) Hypoplastic thumb; Grade IV modified Blauth deformity (Center&Right) late follow up (11 months).

DISCUSSION

Longitudinal radial deficiency, also called congenital radial club hand or radial dysplasia is characterized by various degrees of deficiency along the preaxial or radial side of the extremity. They can range from mild thumb hypoplasia to complete absence of the radius. The incidence of this deformity is estimated to be 1 in 30,000 to 1 in 100,000 live births. It is bilateral in 38-58% of cases. Longitudinal radial deficiency usually occurs sporadically with no known cause. About 40% of patients with unilateral club hand and 27% with bilateral club hand have associated congenital anomalies involving cardiac, genitourinary, skeletal, and hematopoietic system.^[1]

Goldfarb et al. noted that even with the isolated RLD, 40% of unilateral RLD patients and 77% of bilateral RLD patients had an associated abnormality.^[4]

Unfortunately, there were no accurate reports about the actual incidence in Egypt. In our study, all the patients had isolated radial deficiency without associated malformations in other body organs or systems or Fanconi's anemia. All cases were sporadic with no familial occurrence. No parental consanguinity or history of environmental exposure to the mothers during pregnancy. This isn't in agreement with the report about this deformity.

Surgical reconstruction of radial club hand is a formidable task. The goal of treatment of radial

club hand is a functional and cosmetically acceptable upper limb. The principles of treatment are to correct the radial deviation of the wrist, balance the wrist on the forearm and maintain some wrist and full finger range of motion, and allow growth of forearm.^[5]

The centralization procedure seeks to balance the carpus on the distal ulna; however, this can be difficult to accomplish in patients with a severe preoperative deformity. Stretching and splinting have been recommended to stretch the tight radial-sided structures but this often is insufficient to allow a tension free alignment of the carpus on the distal ulna at the time of centralization.^[4]

Several authors have described the concept of soft tissue distraction preceding centralization for congenital radial deficiency. Most of these reports mention unilateral or simple transfixation external fixators. Others used the Kessler and monolateral external fixators for soft tissue distraction.^[6]

In our study, we considered strategic two stage correction for stage IV radial aplasia starting with pre-centralization soft tissue distraction using light weight fibro-carbon external ring (Ilizarov) fixator followed by centralization. By using this staged approach, we safely distracted the taught soft tissues and avoided several of the problems associated with reconstruction of the congenital radial club hand deformity that are often seen when centralization is not preceded by distraction.

The results of our study showed that soft tissue distraction and centralization in 3 patients with an average follow-up of 14 months showed significant improvement from a preoperative average total hand-forearm angulation of 96 degrees to 18 degrees at the latest follow-up examination. This was in agreement with other studies evaluating this approach.

Sabharwal et al. [6] concluded that by using pre-centralization soft tissue distraction with the Ilizarov external fixator, significant correction of both angulation and translation of the hand unit over the distal ulna could be achieved while maintaining adequate bone stock and neurovascular function.

To determine whether this technique decreases the risk for recurrence, it is necessary to follow up these patients until skeletal maturity. A longer follow-up duration is required for assessing the incidence and severity of recurrent deformity. In this study we presented early data on this technique.

Long-term follow-up studies report problems with recurrence and stiffness. Despite numerous technical modifications to preserve alignment, some recurrence of the radial deficiency is universal. The wrists tend to be either flexible and deviated or stiff and straight. Centralization has been shown to improve the appearance of the extremity but has not been proved to enhance function. Currently, long-term maintenance of the carpus on the end of the ulna without sacrificing wrist mobility or stunting forearm growth remains a daunting task. [7]

Pollicization of the index finger to reconstruct an absent or severely hypoplastic thumb is the crowning achievement of 20th century congenital hand surgery. The technique is the result of contributions from many surgeons over the past 100 years. [8]

In our study, we preferred to do pollicization at around the age of one year (average 12.5 months).

Early intervention is favored by many authors to promote early remodeling of the new carpometacarpal articulation and promote intrinsic muscle hypertrophy. They suggested that if mobility, sensitivity and brain integration were generally satisfactory, strength and appearance are two elements to improve. [9]

One of the technical variables is the skin incision. Many types of incision have been proposed. Our technique used both dorsal and volar flaps, as described by Buck-Gramcko. [10] We have been satisfied with this technique because it offers

the ability to create a satisfactory first web space and realistic-appearing thumb.

In our technique, we didn't shorten the flexors or adjust the extensors. Many authors believe that the spontaneous adaptation of the flexor tendons and extensors throughout growth is excellent, but some surgeons prefer to adjust the flexors and more commonly extensors. [9]

Although it is important to monitor patients carefully in the immediate postoperative period for wound infection, impaired arterial inflow, abnormal sensation, and venous congestion, it is also necessary to continue to observe these patients over the long term and consider additional surgeries to maximize function if there is any deficit. [11]

In our case series, there were 3 postoperative complications; including one case of marginal skin necrosis, 2 cases of venous congestion, and 2 suboptimal outcomes; scar contracture of the web space along the incision was noted in one case and redundant skin along the ulnar aspect of the first web space complicated one pollicization. The most notable perioperative complication in this series was venous congestion, which occurred in 2 cases.

Egloff and Verdan reported that 3 of their 30 pollicizations (10%) required scar revision for contracture. [12]

Goldfarb et al. reported the most frequently noted postoperative functional deficits in their patients were poor active opposition and thumb extension. They reported 15 thumbs had poor extension and required tendon shortening or epiphyseal arthrodesis at a subsequent procedure. [11]

CONCLUSION

Staged approach for type IV congenital radial club hand is an effective, reliable and feasible approach than serial casting or unilateral/dual-bar fixators. A significant improvement in deformity with a more functional position of the hand can be achieved while minimizing overall complications.

Pollicization of the index finger for congenital thumb hypoplasia is a well-accepted procedure with good outcome. The aesthetic and functional results of pollicization are directly related to the transposed index digit and surrounding musculature.

RECOMMENDATIONS

A longer follow-up study is required for assessing the incidence and severity of recurrent deformity after soft tissue distraction and centralization for type IV deformities until skeletal maturity as we presented early data on this technique in this study.

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النقص الكعبري الطولي؛ الوجهة الجينية والتدخل الجراحي

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يعد النقص الطولي الكعبري (العجز ما قبل محوري) أو ما يعرف عادة باليد المضربية من العيوب الخلقية للطرف العلوي التي تشتمل على مجموعة متنوعة من التشوهات والتي تتراوح ما بين تشوه نقص نسيجي طفيف بأصبع الإبهام إلى النقص الكامل لعظمة الكعبرة. ويبدو من المهم معرفة الأسباب التي ينجم عنها مثل هذه العيوب الخلقية، وكذلك تصنيفها بشكل دقيق، ومعرفة العيوب الخلقية الأخرى المتزامنة معها، وذلك لرسم سياسة العلاج المثلى.

يجب أن يؤخذ في الاعتبار العيوب الجسمية العديدة أو المتلازمات الخلقية التي قد تصاحب النقص الكعبري، عند تقييم هذه الحالات وتجهيزها للتدخل الجراحي حسب أولويات وخطورة العيوب المصاحبة

أجريت هذه الدراسة على سبعة مرضى على التوالي- يعانون من تشوهات بالطرف العلوي شخصت كنقص كعبري طولي- من المترددين على وحدة جراحة التجميل والإصلاح بمستشفيات جامعة الزقازيق في الفترة من ديسمبر ٢٠٠٨ وحتى ديسمبر ٢٠١٢: ثلاث حالات منهم كانت تشوهات من الدرجة الرابعة؛ و اثنين من الدرجة N، في حين كانت الحالتين المتبقيتين من الدرجة ٠. وكان متوسط العمر وقت التدخل الجراحي ١٢ شهراً (١٠-١٥ شهراً) (خمس حالات من الذكور واثنان من الإناث).

وأظهرت إجراءات الفحص الشاملة المنهجية لهذه الحالات أنها كانت عيوب مفردة في جهة واحدة غير مصاحبة بأي عيوب خلقية أخرى أو متلازمات مرضية أو فقر الدم الفانكوني. واستند استبعاد فقر الدم الفانكوني على نتائج الدراسات الوراثة الجينية والدراسات الكروموسومية من خلال اختبارات (DEB و MMC) والتي تم إجراؤها بالمركز القومي للبحوث - قسم الوراثة البشرية بالقاهرة.

اعتمدنا في هذه الدراسة على تقنية إصلاح تشوهات الدرجة الرابعة (لثلاث حالات) على مرحلتين منفصلتين، وتهدف المرحلة الأولى لفرد وتليين الأنسجة الرخوة باستخدام مثبت خارجي حلقي (اليزاروف) ذو وزن خفيف من ألياف الكربون ثم إجراء عملية تثبيت الرسغ في وضع مركزي في المرحلة الثانية. وقد حصل تحسن ملحوظ في وضع الرسغ مصحوباً بتحسين المعايير القياسية في الأشعات السينية أثناء المتابعة الدورية المنتظمة لهؤلاء المرضى.

يحتاج تقييم دور إصلاح هذه التشوهات على مرحلتين في تقليل نسبة إرتدادها إلى متابعة هؤلاء المرضى حتى مرحلة النضج الهيكلي للطرف العلوي، غير أننا قدمنا في هذه الدراسة نتائج أولية لهذه التقنية. وعلى الرغم من ذلك فيمكننا القول أنه من خلال استخدام تقنية إصلاح تشوهات النقص الكعبري الدرجة الرابعة في مرحلتين، يسهل تعديل وتقويم الساعد والمعصم دون مواجهة صعوبات تذكر، مقارنة بإجراء التثبيت المركزي للرسغ غير مسبوق بفرد وتليين الأنسجة الرخوة، بما يثبت فعاليته وإمكانية تنفيذه بأمان، حيث تحقق باستخدام هذه التقنية تحسن كبير في التشوه مع تخفيض المضاعفات بشكل عام.

تظهر الدراسات التي أجريت في هذا المجال من خلال متابعة دورية على المدى الطويل للحالات التي أجريت لها جراحات، ارتداد التشوه بنسب متفاوتة مع احتمال حدوث تيبس بمفصل الرسغ والأصابع. وخلصت هذه الدراسات أن جراحة تمركز الرسغ وعظمة الزند غالباً ما تؤدي لتحسن الشكل المظهري للطرف العلوي دون حدوث تحسن وظيفي ملحوظ.

إن نقص تنسج الإبهام وتشوّهاته والتي تحدث بدرجات متفاوتة هو في أغلب الأحيان أحد أشكال النقص الكعبري الطولي. ويصنف هذا التشوه إلى خمس درجات، كل منها له الطريقة المناسبة في الإصلاح. يعتبر وجود أو عدم وجود مفصل رسغ-مشطي بحالة جيدة هو العامل الحاسم في الإختيار ما بين إبقاء الأصبع وإصلاحه أو اجتنائه وأبهمة أصعب السبابة.

أجريت للأربع حالات المتبقية (اثنان من نوع N واثنان من نوع ٠) والتي كان الإبهام بها يعاني من نقص نسيجي من الدرجة الرابعة أو الغياب الكامل لأصبع الإبهام (الدرجة الخامسة) عملية إعادة بناء الإبهام بواسطة أبهمة أصعب السبابة. كانت النتائج جيدة بشكل عام من ناحية النمو وقابلية الأصبع للحركة. ومن خلال هذه البحث، ظهر أن الإحتقان الوريدي من أهم المضاعفات التي قد تحدث بعد الجراحة.

ويمكننا استنتاج الارتباط المباشر بين النتائج الوظيفية والتحسين في الشكل الجمالي والمظهري لليد من جهة وحالة أصعب السبابة والعضلات المحيطة بها من جهة أخرى. أصبحت تقنية أبهمة أصعب السبابة لإعادة إعمار الإبهام من الجراحات ذات الصبب الذائع و النتائج الجيدة. ومع ذلك، فمن المهم للجراحين، والمرضى، ومقدمي الرعاية للأطفال المصابون بمثل هذه التشوهات، أن يكونوا على بينة من احتمال حدوث مضاعفات ما بعد الجراحة، أو أن يواجهوا نتائج دون المستوى الأمثل، والتي قد تتطلب عمليات جراحية لاحقة. ومن الضروري أيضاً التأكيد على مواصلة متابعة هؤلاء المرضى على المدى الطويل والنظر في جراحات إضافية لتحقيق أقصى قدر من المثالية الوظيفية إذا ظهرت دلائل عجز بالأصبع الذي تم أبهّمته.