

## Management of Congenital Clasped Thumb in Patients with Distal Arthrogyriposis 3 Years Follow-up

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### ABSTRACT

**Background:** Congenital clasped thumb is a deficiency of active extension of the thumb with flexion and adduction deformity after the age of 3 months. In distal arthrogyriposis, congenital clasped thumb is characterized by global metacarpo-phalangeal (MCP) joint instability; severe narrow 1st web space and palmar contracture.

**Objective:** The present study aimed to describe patients' characteristics, and the results of treatment of congenital clasped thumb in distal arthrogyriposis.

**Patients and methods:** A total of 24 congenital clasped thumbs in 14 patients (9 boys and 5 girls) underwent release of tight first web space structures, Ghani flap, and MCP joint chondrodesis with or without flexor pollicis longus lengthening. There were 10 bilateral and 4 unilateral cases. The mean age of the patients was 4 years old (ranging from 3 to 5). **Results:** According to the parent parents of the participants, all patients were fulfilled with the thumb function and appearance. Union of the chondrodesis was achieved in 21 thumbs. There was statistically significant improvement of thumb abduction, rotation and opposition.

**Conclusions:** The 1st web space contracture release, Ghani flap and MCP chondrodesis is a good surgical option for management of congenital clasped thumb in distal arthrogyriposis.

**Keywords:** Congenital clasped thumb, Distal arthrogyriposis, Modified dorsal rotational flap.

### INTRODUCTION

Congenital clasped thumb by definition is a deficiency of active extension of the thumb with flexion and adduction deformity after the age of three months. The extension loss involves mainly the metacarpo-phalangeal (MCP) joint and sometimes, the interphalangeal (IP) joint<sup>(1)</sup>.

McCarroll<sup>(2)</sup> classified the clasped thumb according to the deformity flexibility into supple and complex types. Complex type is commonly associated with arthrogyriposis with the inability to correct the deformity passively. Arthrogyriposis or multiple joint contracture syndrome is a group of disorders characterized mainly by contractures of two or more joints in multiple body areas<sup>(3)</sup>. Distal arthrogyriposis is a subtype of arthrogyriposis that involves primarily the hands and feet. There are more than ten types of distal arthrogyriposis<sup>(4,5)</sup>.

Congenital clasped thumb in distal arthrogyriposis is characterized by global MCP instability, narrow first web space with deficient skin, and palmar contracture involving the skin and the subcutaneous tissues<sup>(6)</sup>. This study aims to describe patients' characteristics, and the outcome of management of congenital clasped thumb in distal arthrogyriposis.

### PATIENTS AND METHODS

A retrospective study was conducted on 24 clasped thumbs in 14 patients with distal arthrogyriposis (9 boys and 5 girls). All patients underwent release of tight first web space structures and MCP joint chondrodesis with or without flexor pollicis longus (FPL) lengthening, according to the technique described by Abdel-Ghani flap<sup>(7)</sup>.

Patients were operated in the period from March 2016 to March 2019. There were 10 bilateral and 4 unilateral cases. The patients' mean age was 4 years old (range from 3 to 5). The minimum follow-up period was 3 years. Supple type and complex clasped thumb patients rather than those with distal arthrogyriposis were excluded.

The following data were collected from the included patients:

**Medical history taking:** There were 5 (35.7%) cases with positive family history. Positive consanguinity was present in 10 (71.4%) cases. Drug intake during pregnancy was present in 3 (21.4%) cases. In 2 of them, contraceptive pills were taken and in the third case, the antibiotic was taken for fever treatment.

**Clinical examination:** It included evaluation of the web space narrowing, the severity of the deformity, and associated hand anomalies. Camptodactyly was present in 24 hands with different severity. There were 21 hands with mild to moderate camptodactyly, and only 3 were severe which necessitated surgical treatment. Ulnar drift hand was present in 17 (70.8%) hands. Wrist extension deformity was present in all hands but there was no associated radial deviation of the index in this study.

A pre-operative X-ray of the hand was done for all cases to identify the appearance of the ossific center of the base of the proximal phalanx of the thumb which was a precondition for surgical intervention.

### Surgical technique:

**Abdel-Ghani<sup>(7)</sup> flap** was marked by 2 lines; the first line was drawn along the first metacarpus and the second line was extended in a curved fashion from the 2<sup>nd</sup> metacarpus head to end at the 5<sup>th</sup> metacarpus nearly

at the level of the wrist. The flap was then continued by Z shaped incision at the palmar side of the web. The incision was first drawn (**Figure 1**).

Through that incision, the sequential release of the tight structures was performed (**Figure 2**). The palmar fascia, transverse head of adductor pollicis, and the first dorsal interosseous muscles were released to widen the first web space.

During the release of the adductor pollicis from the 3<sup>rd</sup> metacarpus, the index neurovascular bundle injury should be avoided. Global instability was evident in all cases after the release of the tight web space structures. The capsule of the 1<sup>st</sup> MCP joint was then incised, and the cartilage of the joint was cut till the identification of the subchondral bone of the head of the metacarpus and

the ossific center of the base of the proximal phalanx. A suitable size K- wire was then introduced through the long axis of the thumb to fix the MCP joint. This K-wire was advanced to the carpal bone after manipulation of the thumb in palmar abduction and extension. A second K-wire was then introduced from the first to the second metacarpus to keep the first web open and wide (**Figure 3**).

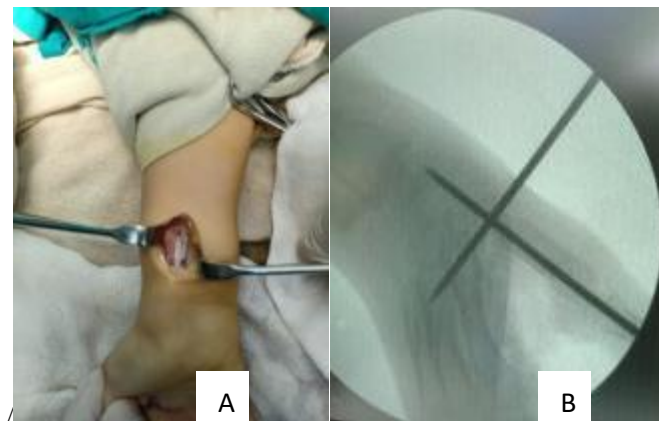
The intramuscular recession of the FPL was needed in 14 hands to correct the IP flexion deformity. The flap was then advanced and the skin was sutured. In all patients in this study, a full-thickness skin graft was required to augment the deficient skin at the ulnar side of the thumb (**Figure 4**). Above elbow thumb spica cast was finally applied.



**Figure (1):** Modified dorsal rotational flap with palmar Z extension.



**Figure (2):** After the opening of the Z incision and retraction of the Abdel Ghani flap.



**Figure (3):** A: Intramuscular recession of the FPL. B: Fixation of the MCP and the 1<sup>st</sup> web space by K wires.



**Figure (4):** Advancement and closure of the Abdel Ghani flap. A skin graft was used to augment the deficient skin of the thumb.

**Post-operative care and follow-up:**

Plain X-ray thumb posteroanterior and lateral views were done in the early postoperative period, and at the 6th week to detect the union of the chondrodesis of the 1st MCP joint. The k-wires and the cast were removed after the union of the chondrodesis of the 1st MCP joint.

**Ethical Approval:**

The study was authorized by Zagazig University's Ethical Institutional Review Board (IRB number: 9250/25-1-2021), and informed consent was signed by every child's parents. This study was executed according to the code of ethics of the World Medical Association (Declaration of Helsinki) for studies on humans.

**Statistical Analysis**

The collected data were introduced and statistically analyzed by utilizing the Statistical Package for Social Sciences (SPSS) version 22 for windows. Qualitative data were defined as numbers and percentages. Stuart-Maxell test was used for comparison between pre and postoperative categorical variables. Quantitative data were tested for normality by Kolmogorov-Smirnov test. Normal distribution of variables was described as mean and standard deviation (SD), and Wilcoxon signed ranks test was used for comparison between pre and postoperative quantitative variables. P value  $\leq 0.05$  was considered to be statistically significant.

**RESULTS**

In this study, we used the same evaluation parameters as **Abdel Ghani et al.** (6) depending on both Gilbert's grading system and the parent satisfaction. There was statistically significant improvement of thumb abduction, rotation and opposition, with good to excellent results in 24, 22 and 19 cases respectively. Results are presented in **tables 1 and 2**. Union of the chondrodesis was achieved in 21 thumbs. Three thumbs with fair instability were present due to nonunion. This instability did not necessitate revision of the chondrodesis. The parents of all patients were fulfilled with both the function and the appearance of the thumb. There was postoperative necrosis of the edge of the flap in 10 hands which resolved and resulted in no complications at the final follow-up.

**Table (1): Assessment of outcome of surgery of studied patients.**

Variable	Preoperative (N=24)		Postoperative (N=24)		Test	P-value
	No.	%	No.	%		
<b>Abduction</b>						
Mean $\pm$ SD	12.04 $\pm$ 8.01		40.20 $\pm$ 4.39		-4.289 <sup>a</sup>	<0.001
Median (Range)	9 (0 – 28)		41 (32 – 45)			
<b>Rotation</b>						
Mean $\pm$ SD	71.79 $\pm$ 9.69		105.33 $\pm$ 10.65		-4.292 <sup>a</sup>	<0.001
Median (Range)	70 (60 – 88)		105 (88 – 120)			
<b>Opposition</b>						
With little	0	0%	12	50%	23.000 <sup>b</sup>	<0.001
With ring	0	0%	7	29.2%		
With middle	0	0%	4	16.7%		
None	24	100%	1	4.2%		
<b>Stability</b>						
Very stable	0	0%	14	58.3%	24.000 <sup>b</sup>	<0.001
Stable	0	0%	7	29.2%		
Mild instability	0	0%	3	12.5%		
High instability	24	100%	0	0%		

a: Wilcoxon signed ranks test; b: Stuart-Maxell test; P-value<0.05 is significant.

**Table (2): Pre-operative and post-operative position and thumb function according to Gilbert’s grading system.**

Variable	Preoperative (N=24)		Postoperative (N=24)		Test <sup>b</sup>	P-value
	No.	%	No.	%		
<b>Abduction</b>						
Excellent	0	0%	14	58.3%	24.000	<0.001
Good	0	0%	10	41.7%		
Fair	11	45.8%	0	0%		
Poor	13	54.2%	0	0%		
<b>Rotation</b>						
Excellent	0	0%	12	50%	22.219	<0.001
Good	0	0%	10	41.7%		
Fair	6	25%	2	8.3%		
Poor	18	75%	0	0%		
<b>Opposition</b>						
Excellent	0	0%	12	50%	23.000	<0.001
Good	0	0%	7	29.2%		
Fair	0	0%	4	16.7%		
Poor	24	100%	1	4.2%		
<b>Stability</b>						
Excellent	0	0%	14	58.3%	24.000	<0.001
Good	0	0%	7	29.2%		
Fair	0	0%	3	12.5%		
Poor	24	100%	0	0%		

b: Stuart-Maxell test; P-value<0.05 is significant.

**Figure 5** represents the follow-up of one of our cases.



**Figure (5):** A: Four years old girl with distal arthrogyriposis had a right-sided congenital clapsed thumb. B, C: 6 months postoperative follow-up with wide 1<sup>st</sup> web space. D to F: Three years of clinical follow-up with some retraction of the edge of the Ghani flap and adequate opposition of the thumb with the little finger. G: Follow-up X-ray posteroanterior view shows union of the chondrodesis of the 1<sup>st</sup> MCP joint.

## DISCUSSION

Surgical correction of the clasped thumb in distal arthrogryposis involves mainly the release of the contracted web space structures, augmentation of the web skin deficiency, and MCP joint chondrodesis.

**Abdel Ghani** <sup>(7)</sup> modified the Buck-Gramcho dorsal rotational flap <sup>(8)</sup>. Ghani flap is characterized by a wide rectangular end compared to the narrow tipped apex of the Buck-Gramcho flap. Also, the Ghani flap can reach far enough to the mid-palmar level. This allows wide release of the first web and avoids web re-narrowing complications after flap retraction.

**Anbarasan et al.** <sup>(10)</sup> reported the results of abductor pollicis brevis rerouting in congenital clasped thumb in arthrogryposis multiplex congenita. They used a polystyrene cone for radiographic assessment of the first web space <sup>(9)</sup>. The abductor pollicis brevis was released, rerouted and anchored to the ulnar aspect of the MCP joint at the adductor pollicis insertion with the thumb in pronated and abducted position. They reported that the rerouted tendon acts as a static tenodesis of the MCP joint in extended position. However, they had 2 cases of MCP instability that required revision by chondrodesis <sup>(10)</sup>.

In their case report, **Kim et al.** <sup>(11)</sup> discussed a case of bilateral congenital clasped thumb in which the left thumb had global instability as detected by stress test and stress radiograph that was treated with double-breasting procedure and had good 2 years follow-up. However, according to **Abdel Ghani et al.** <sup>(12)</sup> chondrodesis is the best way to achieve stability in cases of MCP global instability.

Global instability of the 1<sup>st</sup> MCP joint is a very common finding in congenital clasped thumb in arthrogryposis. MCP chondrodesis was performed in all cases in this study to correct this flexed unstable joint. Shortening of the thumb that resulted from the chondrodesis alleviated the necessity to release the contracted skin at the flexor aspect of the thumb and also alleviated the necessity for flexor pollicis longus lengthening for 10 thumbs. Chondrodesis also obviated the need for tendon transfer.

Care should be taken during chondrodesis to prevent injuring the physis of the proximal phalanx. The appearance of the ossific nucleus of the proximal phalanx can be considered a prerequisite for surgery. We had no case of proximal phalanx growth arrest in this study.

To our knowledge, little was written about the patients' characteristics of congenital clasped thumb, especially in patients with distal arthrogryposis. In this study, male to female ratio was 1.8:1 compared to 2.5:1 in **Abdel Ghani et al.** <sup>(12)</sup> and **Lin et al.** <sup>(13)</sup> studies. Positive consanguinity was present in 10 (71.4%) cases compared to 57.5 % in **Abdel Ghani et al.** <sup>(12)</sup>. History of drug intake during pregnancy was present in 3 (21.4%) cases compared to 32.5% in **Abdel Ghani et al.** <sup>(12)</sup> reports. As regards the associated hand

anomalies, camptodactyly was present in all hands compared to 50% of cases in **Abdel Ghani et al.** <sup>(12)</sup>. Ulnar drift hand was present in 17 (70.8%) hands compared to 9 (12.3%) hands in **Abdel Ghani et al.** <sup>(12)</sup>. In their study, **Abdel Ghani et al.** <sup>(12)</sup> mentioned 1 hand with radial deviation of the index finger. Differences in patients' characteristics may be related to the fact that **Abdel Ghani and collaborators** <sup>(12)</sup> studied the characteristics of all types of congenital clasped thumb patients. However this study was concerned with complex congenital clasped thumb in patients with distal arthrogryposis.

In the current study, there was a statistically significant postoperative improvement in thumb abduction, rotation, opposition, and stability. According to Gilbert's grading system, there were 14 (58.3%) excellent and 10 (41.7%) good abduction ranges compared to 57.97%, 36.23% in **Abdel Ghani et al.** <sup>(6)</sup>. The thumb rotation in this study was excellent in 50%, good in 41.66% and fair in 8.33% of cases compared to 43.47%, 50.72%, and 5.8%, respectively in **Abdel Ghani et al.** <sup>(6)</sup>. Opposition to the little finger was obtained in 50% of the hands in this study, 29.16% of the hands had opposition to the ring finger, 16.66% had opposition to the middle finger and 4.16% of the hands had no opposition, compared to 28.9%, 27.5%, 21.7% and 21.7% respectively in **Abdel Ghani et al.** <sup>(6)</sup>.

**Limitations of the study:** The small sample size, short follow-up period and the absence of a control group to compare with are the limitations of this study.

## CONCLUSION

Congenital clasped thumb in distal arthrogryposis is a complex deformity that can be managed adequately by the release of the tight 1<sup>st</sup> web space structures, Ghani flap and 1<sup>st</sup> MCP joint chondrodesis.

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