Original Article

Congenital Clasped Thumb

Management Issues with Congenital Flexion-Adduction Deformity of the Thumb (Congenital Clasped Thumb) A Single Center Experience

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ABSTRACT

Objective: To describe the pattern of extensor mechanism deformity and associated first web space contractures in patients with congenital clasped thumb and their outcomes.
Study Design: Retrospective case series.
Place and Duration of Study: This study was conducted at the Department of Plastic and Reconstructive Surgery, Liaquat National Hopsital, Karachi including data of the patients from 1997 to 2016.
Materials and Methods: 12 patients (20 thumbs) with clasped thumb are reported who underwent treatment. The series is divided into two groups according to the Weckesser classification. In the first group, the prominent pathological feature was hypoplasia /absence of the extensor tendons. The second group, the arthrogryphotic type, had hypoplasia/absence of extensor mechanism along with contracture of the intrinsic muscles of the thumb and shortening of the skin. Instability of the Metacarpo-phalangeal joint and adduction contracture of the first ray were found in 2 hands of the second group. Splinting only was adopted in 7 thumbs and surgical treatment was performed on 13 thumbs, with an average follow –up of 24 months.
Results: Conservative treatment is effective in type I cases when presented early. All patients were satisfied with the results of surgical treatment employed for type-II deformity.
Conclusion: Isolated clasped thumb deformities that lacked the extensor mechanism and failed to respond to conservative treatment with splinting, EIP tendon transfer and correction of first web space contracture with Z-plasty was found to be a successful and acceptable reconstructive option.
Key Words: Clapsed thumb, Weckesser classification, Splinting, Tendon transfer.

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INTRODUCTION

Congenital clasped thumb is progressive flexion and adduction deformity that is associated with a wide array of congenital anomalies, although it may also be present as an isolated abnormality[1]. This rare disease is characterized by deficiency of extensor pollicis brevis (EPB) or longus (EPL) or both and sometimes Abductor Pollicis longus (APL) due to functional or structural causes. The deformity is usually accompanied with a variable degree of narrowing and contracture of the first web space[4,5]. There is association of this disease with other generalized musculo-skeletal malformations, commonly; arthrogryposis, digitotalar dysmorphism and Freeman–Sheldon syndrome[6].

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The diagnosis is usually made if clasping persists and normal independent function of thumb does not develop by the age 3 to 4 months. It is often confused with trigger finger deformity because of fixed flexion found in thumb especially if the underlying pathology includes absence of EPL. Diagnosis can be made by passively correcting the flexion deformity which can be seen in case of congenital clasped thumb while difficult passive Interphalangeal joint extension and clicking sound is observed in trigger thumb.[7]
Several treatment options are available for congenital clasped thumb but choice depends on the type of deformity present. Conservative treatment with splinting has been successful but operative intervention is required for cases that developed web space contractures secondarily in neglected cases. Tendon transfers are useful in cases with deficient extensor mechanism[8].

MATERIALS AND METHODS

The study was conducted at department of Plastic and reconstructive surgery, Liaquat National Hopsital,

Karachi Including data of the patients from 1997 to 2016.

Inclusion Criteria:

- Irrespective of race and gender, patients age of 5 and 36 months were included in the study.
- Patients with type-I and type-II deformity with unilateral or bilateral hand involvement.

Exclusion Criteria:

- Patients with type-III and type-IV deformity
- Patients previously operated for same deformity.

The patients were enrolled by using a preformed following proforma after taking informed consent from parents or care taker of the patient by primary investigator:-

Proforma

A. Sr.No._____ B. Case Number:_____
C. Age: _____ (months)
D. Gender 1. Male 2. Female
E. Site: 1. Unilateral a. Right b. Left
2. Bilateral
F. Type of deformity: 1. Type-I 2. Type-II
G. Treatment: 1. Surgical 2. Non-surgical
H. Outcome: 1. Excellent 2. Good
3. Fair 4. Poor
I. Over all outcome: 1. Improved 2.Not improved

Data was entered and analyzed by using statistical software SPSS version 23.

They were exposed to thorough assessment, including recording of the family history, pregnancy history and full clinical assessment by experienced pediatrician and echocardiography was done when recommended to rule out associated anomalies.

To assess the type of deformity (according to Weckesser et al classification), every patient was subjected to detailed clinical and radiographic examination by an experienced hand surgeon.

Operational Definitions: The deformed thumbs were classified according to Weckesser et al classification in to 4 types:

Type 1	Supple and normal sized thumb with absent or hypoplastic extensor mechanism. The thumb could be passively abducted and extended against the resistance of thumb flexors
Type 2	Complex with additional findings of joint contracture, first web space contracture, Thenar muscles and collateral ligament abnormality. The thumb could not be passively extended and abducted.
Type 3	Hypoplastic Thumb with deficient tendons and muscles.
Type 4	Clasped thumbs associated with Arthrogryposis and its associated syndromes. The extensor mechanism may have little or no abnormality.

Simplified Gilbert's Chart

Function of thumb	Excellent	Good	Fair	Poor
Abduction	40◦ - 45◦	30◦ - 40◦	10◦ - 30◦	< 10◦
Opposition	With little	With ring	With middle	none

Treatment Protocols: All procedures were performed by the same hand surgeon.

- **Non-Operative Treatment:**

Full time splinting of thumb in extension and abduction for 6 months with plaster cast changed every 6 weeks to allow for growth of the hand. This was followed by night splinting for further 6 months after achieving active extension of the thumb. [Figure 2: Non-operative: splinting of thumb in extension and abduction]

- **Operative treatment:**

Reconstructive options for every case were different based on the underlying deformity found, i.e. degree of narrowing of the first web, stability of the metacarpophalangeal (MP) joint and muscle deficiency.[Figure 3: Operative release of webspace with z-plasty5]

The surgical treatment mainly aimed at widening the narrowing of the first web space, including the deep tight structures. The web space narrowing was released in almost all case by a simple Z-plasty except in one where a dorsal rotational advancement flap was used due to severe web space narrowing. Through the skin incision designed to widen the web space, the dissection was deepened to the underlying fascia over the thenar muscles, protecting the flexor tendons and the neurovascular bundle to the index finger. The tight structures were identified and released. The origin of adductor pollicis muscle was released from the third metacarpus in one patient. The thumb was then manipulated into extension and abduction after achieving full release and held in position by 2 crossed k-wires across the web space. One wire passed longitudinally through the thumb, transfixing and holding all joints in extension and wide abduction. A second k-wire passed transversely through the metacarpal, transfixing it to the index metacarpal.

Active extension of the first MP joint was restored by tendon transfer. The preferred tendon was EIP (Extensor indicis) and if that was found to be deficient, a slip of FDS (Flexor digitorum superficialis) was used and transferred to the remnants of vestigial extensor mechanism. EIP tendon was found through a mini incision at second MCP joint from the dorsum of the hand and tenotomized, pulled to wrist level, and then transferred to thumb with a subcutaneous tunnel and attached to the proximal phalanx base.

- **Post-Operative Management:**

An above-elbow splint was applied immediately. The k-wires were removed after 6 weeks of surgery. The

position of thumb in extension was maintained in a night splint for at least 6 months post-operatively, active use of thumb was encouraged during day time.
The assessment of results was done using the following criteria:

Cosmetic appearance: First web space was deepened by a simple z-plasty improving the appearance of hand.

- The modified dorsal rotational advancement flap allowed maximal degree of widening of the web space

Function of Thumb:

- Simplified Gilbert's method was used to assess the functional outcome.

RESULTS

12 patients were studied, out of which 4 (33.3%) had unilateral and 8 (66.7%) had bilateral thumb involvement. Hence, a total of 20 clasped thumbs (subjects) were included. There were 7(58%) males [3 (25%) unilateral, 4 (33.3%) bilateral] and 5(41%) females [1 (8.3%) unilateral, 4 (33.3%) bilateral]. Table Mean age was 17.9 (± 10.02) months (range: 5 to 36 months) at presentation. All cases presented with the main finding of lack of active extension of the thumb.
20 thumbs were treated. Non-operative treatment was carried out in 7 (35%) thumbs [5 (25%) type-I, 2 (10%) type-II] who presented before the age of 12 months. Operative treatment was done in 13 (65%) thumbs, [all type-II] who presented later than 12 months of age or which had not responded to non-operative treatment.
There was improvement in all the operated thumbs. Outcome was excellent in 5 (25%) and good in 6 (30%) thumbs, and were able to pick up a pen, a key and grasp a ball with full active extension at all joints of thumb.
Non-operative treatment was successful in all type-I thumbs i.e. 5 (25%), outcome was excellent in 3 and good in 2 thumbs. They were able to pick up a pen, a key and grasp a ball with full active extension at all joints of thumb. The outcome was poor all type-II thumbs i.e. 2 (10%) [1 fair, 1 poor), requiring surgical intervention later. Figure 1.

Table No.1: Patients ratio with regard to unilateral and bilateral.

	Unilateral	Bilateral	Total
Male	3 (25%)	4 (33.3%)	7 (58.3%)
Female	1 (8.3%)	4 (33.3%)	5 (41.7%)
Total	4 (33.3%)	8 (66.7%)	12 (100.%)

According to our results, out of all type-II thumbs that were treated surgically, the failure rate was around 15%. While all type –II thumbs treated with splinting only showed no improvement.

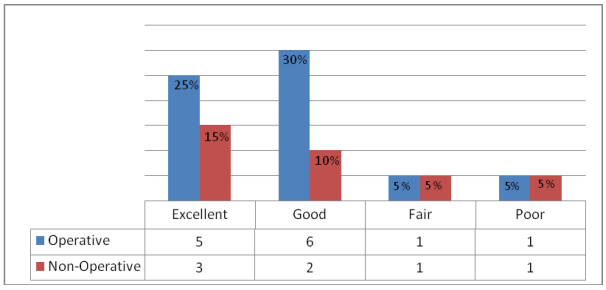

Figure No.1: Patients operative and non-operative results.

Figure No. 2: Non-operative: splinting of thumb in extension and abduction

Figure No.3: Operative release of webspace with z-plasty

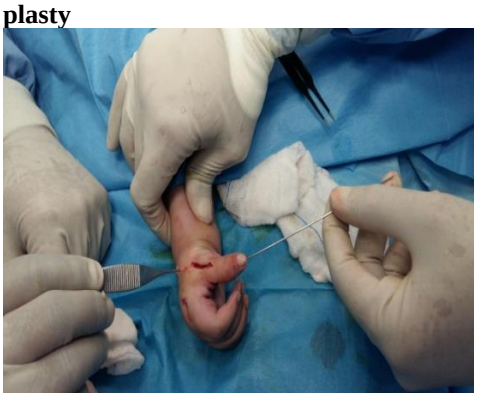

Figure No.4: EIP tendon re-routed for transfer

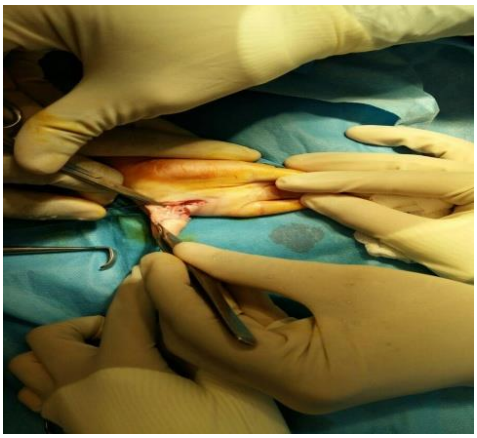

Figure No.5: EIP attached to proximal phalynx base and thumb in abduction & extension

DISCUSSION

As early as 1846, Tamplin described a case of adducted thumb deformity. This deformity is sometimes referred to as Thumb- Clutched hand, but a better term is a congenital clasped thumb.[1,3,5]

The congenital clasped thumb is associated with various well defined syndromes, it may also present as an isolated abnormality. With regards to the possible predisposing factors, the incidence of positive consanguinity ranges from 10% - 60%and positive family history is 32.5%, as reported by Tsuyuguchi et al and Ghani et al. the high incidence of bilateral cases strongly suggest the genetic predisposition as an important causative factor and this is in accordance with the literature. However, the exact cause remains known to date. Similar findings were noted in our study with positive consanguinity and family history in about one-fourth of our cases.[3,9]

With regards to the gender predilection of the disease, a male predominance was seen in our study which is similar to the ratio of 2.5:1 as reported by Ghani et al and Lin et al. however, it differs from the 1 : 1 ratio reported by Tsuyuguchi et al[10]. This difference may be due to the fact that clasped thumb is associated with various syndromes that have varied modes of inheritance and also to the small number of reports in the literature that document the gender predisposition of the affected cases.

It is a syndrome characterized by loss of active extension of thumb due to absence of the extensor mechanism i.e. EPB / EPL alone or both[1,5]. Several Classification systems exist to categorize congenital clasped thumb. The first classification system however was designed by Weckesser et al in 1968 and in 1985 Tsuyugushi et al classified congenital clasped thumb in to three types. McCarroll and Tsuyugushi's classifications are amongst the most commonly used [2]. Weckesser, Reed and Heiple called it a syndrome and divided it into 4 groups. In Group 1, the thumb was deficient in extension only[2,10]. In Group 2, flexion contractures accompany deficient extension. In Group 3, the thumb is hypoplastic with deficient tendons and muscles. Group 4 consists of the few remaining cases that do not fit in first 3 groups (i.e. Arthrogryposis and its associated syndromes like MASA). Literature supports that group 2 cases appear three times as frequently as group 1, while group 3 and 4 are five times less frequent than group 2. McCarroll simply categorized this deformity into flexible and complex types.

Diagnosis is usually difficult during early weeks of life, an infant frequently clutches the thumb and releases it intermittently in spontaneous motion. However, by 3 or 4 months of life, the normal child ceases to clasp his thumb under his fingers. If clasping persists and normal independent action of thumb does not develop, the syndrome of congenital clasped thumb is present. For the reason of fixated deformity of the thumb, this condition is usually confused with trigger thumb[11]. Ruland and Slake reported that flexion deformity in clasped thumb syndrome may be confused with trigger thumb and thus may lead to unnecessary releasing surgeries. This approach aggravates symptoms instead of relieving them[7].

Characteristically, the first metacarpal is held in adduction and proximal phalanx becomes flexed and partially subluxated[5]. Ulnar deviation of the rest of the hand is another common feature. The thumb flexes at the interphalangeal joint when the Extensor Pollicis Longus is absent. The thumb flexes at MP joint and drops to the palm when the Extensor Pollicis Brevis becomes ineffective. When the Abductor Pollicis Longus fails to function, the whole thumb lies against the palm and touches the ulnar border of the hand[1,12,13].

Treatment of congenital clasped thumb depends on the disease stage, age at presentation and associated pathologies. The treatment protocol employed in our series was similar to that followed by Weckesser et al and McCarroll who divided the clasped thumb into two types (supple and complex) The supple deformity was treated with splinting initially with operative intervention in terms of tendon transfer used for cases that failed to respond to conservative management[2,3]. The fixed flexion deformity of the complex type was treated with reconstruction of web space contracture, lax ligaments and hypoplastic or absent extensor mechanism of thumb as appropriate.

There may be no universal criteria for the evaluation of the results due to the inability in assessing the thumb function at that young age. Some authors used the degree of active extension of first MP joint as the reference for evaluation[4,8-14]. Tsuyuguchi et al added the degree of active radial abduction of the Trapeziometacarpal joint to their system of evaluation. Lipskeir and Weizenbluth added width of the first web space to their scoring system and mentioned that active extension of the first MP joint is the most important factor. For our study simplified Gilbert's method was used that focuses on the degree of abduction and opposition of the thumb[1-4, 8, 11, 19, 20].

In this study we noted that the underlying pathology in each case was different and the severity of the disease

with the appropriate treatment offered was dependent mainly on the age of the patient at the time of presentation. In our study, type 1 cases treated with splinting achieved excellent to good results in 5 out of the 7 cases with poor outcome in only 2 thumbs. Almost all type 2 cases given the option of surgical intervention in terms of Tendon transfer showed excellent results with only 2 cases required repeat intervention. The results of our study are consistent with those seen in the literature. Tsuyuguchi et al reported that good results were achieved with type 1 and type 2 cases with conservative methods. In type 2 and type 3, cases in whom conservative treatment is ineffective surgical treatment produces good results[3,9,14]. Ghani et al, Lin et al and Medina et al reported that conservative treatment was effective for patients under 1 – 2 years of age but patients who have skin contractures, absence or weakness of extensor mechanism of thumb and patients with severe hypoplasia or agenesis of thumb, yield better results with surgery.

CONCLUSION

Congenital clasped thumb is a rare but progressive flexion-adduction deformity. If diagnosed timely, proper planning of the treatment according to the degree of severity and type of deformity can restore hand to its full functional capability.

Author's Contribution:

Concept & Design of Study:	Maryam Noor, Mirza Shehab Afzal Beg
Drafting:	Sheeraz ur Rahman, Batool Urooj Rajput
Data Analysis:	Sheeraz ur Rahman, Batool Urooj Rajput
Revisiting Critically:	Maryam Noor, Mirza Shehab Afzal Beg
Final Approval of version:	Maryam Noor, Mirza Shehab Afzal Beg

Conflict of Interest: The study has no conflict of interest to declare by any author.

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