

Original Article

Congenital hand anomalies in Upper Egypt

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ABSTRACT

Background: Congenital hand anomalies are numerous and markedly variant. Their significance is attributed to the frequent occurrence and their serious social, psychological and functional impacts on patient's life. **Patients and Methods:** This is a follow-up study of 64 patients with hand anomalies of variable severity. All patients were presented to Plastic Surgery Department of Sohag University Hospital in a period of 24 months. **Results:** This study revealed that failure of differentiation and duplication deformities were the most frequent, with polydactyly was the most common anomaly encountered. The mean age of presentation was 6 years and female to male ratio was 1.46:1. Hand anomalies were either isolated, associated with other anomalies or part of a syndrome. **Conclusion:** Incidence of congenital hand anomalies in Upper Egypt is difficult to be estimated due to social and cultural concepts, lack of education, poor registration and deficient medical survey. Management of hand anomalies should be individualised, carefully planned and started as early as possible to achieve the best outcome.

KEY WORDS

Congenital; hand anomalies; plastic surgery; Sohag; Upper Egypt

INTRODUCTION

The human hand is essential for interaction with the surrounding environment, so its functional integrity is critical for individual's social life and daily activities.^[1] Congenital hand anomalies rank the second most frequent malformation affecting the human body.^[2] They are encountered in 0.1–0.2% of all newborns.^[3] males are more commonly affected with polydactyly and syndactyly are the most common anomalies.^[4] The aetiology is obscure for most cases of which some occur sporadically whereas

others have the clear genetic background. Congenital hand anomalies may be isolated, component of a syndrome or may occur in the form of a sequence.^[5]

Several classification systems were designed. The most widely accepted classification system is that designed by Swanson and modified by the International Federation of Societies for Surgery of the Hand and the Japanese Society for Surgery of the Hand [Table 1]. It is based on the morphological variations related to developmental

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Table 1: Abbreviated International Federation of Societies for Surgery of the Hand Swanson classification of congenital hand anomalies

Category	Subcategory	Diagnosis
Failure of formation	Transverse	Proximal forearm
	Longitudinal	Digital absence
	Central	Symbrachydactyly
		Longitudinal radial deficiency
		Longitudinal ulnar deficiency
Failure of differentiation		Cleft hand
	Soft tissue	Simple syndactyly
	Skeletal	Camptodactyly
	Tumorous	Congenital trigger digit
		Radioulnar synostosis
		Metacarpal synostosis
		Phalangeal synostosis
		Symphalangia
		Clinodactyly
		Triphalangeal thumb
		Haemangiomas
Osteochondromas		
Fibrous dysplasia		
Duplication		Ulnar polydactyly
		Thumb duplication
Overgrowth		Macrodactyly
		Hemihypertrophy
Undergrowth		Brachydactyly
		Brachysyndactyly
Constriction band syndrome		With oedema
		Without oedema
Generalised abnormalities and syndromes		Marfan syndrome
		Skeletal dysplasia
		Dwarfisim

defects and has the advantages of being simple and practical.^[3]

Achieving the best functional outcome with improving hand appearance as possible are the main goals of management of hand anomalies.^[6] The management is best achieved by a multidisciplinary team including, a hand surgeon, a paediatrician, a physiotherapist, a psychotherapist and a geneticist to get desirable results.^[7] Early effective psychotherapy of parents helps in improving their reaction with their child's anomaly and reduces their fear and sense of guilt. The management plan should be carefully designed, started early and must be completed before school age to avoid the negative social impacts on the child.^[3] The recommended age of reconstruction is between 1 and 2 years old, however, earlier surgical intervention is indicated for certain anomalies that may result in progressive deformity such as radial dysplasia or syndactyly between digits of unequal lengths.^[6] Post-operative physiotherapy is of paramount importance to achieve a good function.

PATIENTS AND METHODS

This study was achieved from October 2012 to November 2014 and included all patients with hand anomalies presented to the outpatient clinic of Plastic Surgery Department of Sohag University Hospital from different governorates of Upper Egypt.

A detailed history was taken and complete physical examination with necessary investigations (plain X-ray, routine pre-operative investigations; blood grouping, blood picture, prothrombin time and concentration) were done. The management plan was individually designed regarding patient's age, type and severity of deformity present, residual hand function and appearance of the affected hand. The plan and possible complications were discussed with patients or their parents.

Different surgical procedures were done including excision of accessory digits, separation of syndactylised digits using different techniques with or without grafting, correction of camptodactyly through several procedures and correction of cleft hand through defect excision and web reconstruction.

Follow-up was arranged for all cases at regular visits (every week in the 1st months then every 2 weeks for the following 2 months, then monthly visits with minimum follow-up of 3 months). Physiotherapy for needed cases was done up to 6 months at the rehabilitation clinic of Rheumatology Department of Sohag University Hospital.

RESULTS

The study included 64 patients, 26 males and 38 females. Patients' age ranged from 2 months up to 37 years old with mean presentation age was 6-year-old. Thirty-five patients (55%) had positive consanguinity. The family history of similar anomalies was found in 20 patients and family history of other anomalies was found in 6 patients. Forty-three patients had isolated hand anomaly/anomalies, whereas associated anomalies were detected in 21 patients of which 8 patients had different syndromes [Tables 2 and 3].

Failure of differentiation anomalies was detected in 33 patients whereas duplication was found in 25 patients [Table 4].

Polydactyly was the most common anomaly detected; it was found in 25 patients of whom 18 patients were

females and 7 patients were males. Ulnar polydactyly was more frequent than radial polydactyly [Table 5]; detected in 19 out of 25 patients. For patients with radial polydactyly, Wassel Type II was the most common type; detected in 4 out of 6 patients, the other two patients had Wassel Type IV.

Sixteen patients with ulnar polydactyly were operated with excision of the accessory digit [Figure 1]. No complications were detected on follow-up. Three patients with radial polydactyly were operated of whom 2 patients had a dominant ulnar digit, and one patient had dominant radial digit which was preserved [Figure 2].

Syndactyly was the second most common anomaly. It was detected in 24 patients (36%); 15 females and 9 males [Table 6].

Table 2: Distribution of hand anomalies of the study

Hand anomaly/ anomalies	Isolated	With associated anomalies	Total
Solitary	36	17	53
Multiple	7	4	11
Total	43	21	64

Table 3: Different syndromes encountered in the study

Syndrome	Number
Constriction band syndrome	3
Split hands split feet syndrome	2
Apert's syndrome	1
Pfeifer's syndrome	1
Poland syndrome	1
Total	8

Table 4: Types and frequency hand anomalies in the study

Type	Number	Percentage
Duplication	25	38
Ulnar polydactyly	19	27
Radial polydactyly	6	11
Syndactyly	24	36
Constriction band syndrome	5	8
Clinodactyly	5	8
Cleft hand	5	8
Camptodactyly	4	6
Vascular malformation	3	4
Symbrachydactyly	3	4
Brachydactyly	3	4
Congenital trigger finger	1	2

Table 5: Polydactyly cases of the study

Polydactyly cases	Unilateral		Bilateral	Total
	Right	Left		
Ulnar polydactyly	3	4	12	19
Radial polydactyly	2	4	0	6
Total	5	8	12	25

The 3rd web was the most commonly affected [Figure 3]; it was involved in 17 cases (71%) followed by the 2nd web, involved in 10 cases (42%). Eighteen patients (75%) were operated of whom dorsal triangular flap technique was applied for 13 patients, dorsal rectangular flap technique for 4 patients and one patient needed butterfly flap technique for web deepening [Figures 4 and 5]. Fifteen patients needed grafting (83%) of which full thickness grafting was used in 6 cases (40%). Eight out of operated cases (44%) had complications. Web creeping was the commonest and was detected in 6 cases (33%). Other complications, including flexion contracture, hypertrophic scarring and wound infection, were noticed [Figure 6].

Four patients had camptodactyly; two males and two females. All patients were presented after the age of 15 years old. The little finger was the most frequently affected digit; was involved in three patients [Figure 7]. All patients had unilateral deformity and were operated using multiple Z-plasties of skin, release of flexor



Figure 1: Bilateral ulnar polydactyly and brachydactyly Type D. (a) Both hands, dorsal aspect. (b) X-ray of both hands



Figure 2: Right hand radial polydactyly Type II with dominant radial digit. The patient had positive family history of similar condition (father); (a) Right hand, lateral view. (b) Right hand after excision of the ulnar accessory digit. (c) Right hand of patient's father

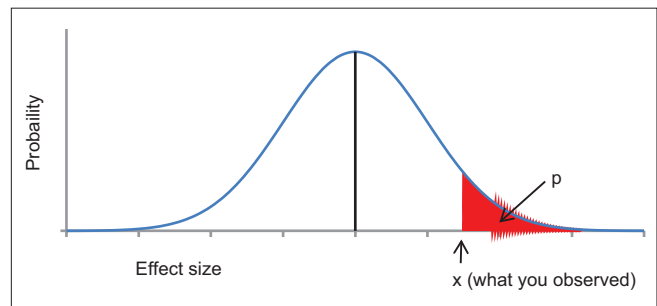


Figure 3: Frequency of web affection in syndactyly cases

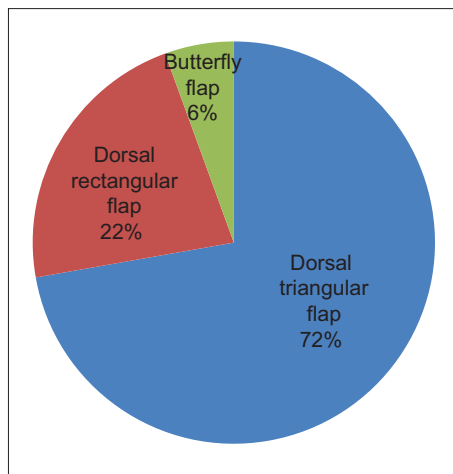


Figure 4: Different techniques used for web reconstruction in syndactyly

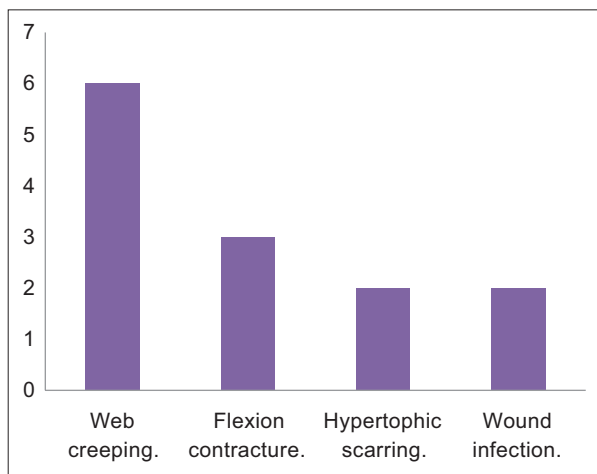


Figure 6: Types and frequency of complications after syndactyly separation

digitorum superficialis tendon and checkrein ligament with K-wire application for at least 3 weeks. One patient needed anterior capsulotomy of proximal interphalangeal joint. With follow-up, joint stiffness and residual flexion deformity of varying degrees were detected in all operated cases.

Five patients had clinodactyly of whom two patients had familial clinodactyly [Figure 8]. Patients with familial clinodactyly had bilateral hand affection. The little finger was involved in four patients [Table 7]. One patient was operated using multiple Z-plasties of skin with K-wire application for 3 weeks. By follow-up residual, angular deformity was noticed.

Five patients had constriction band syndrome; three males and two females [Table 8]. Type IV was the most common type and was encountered in all patients [Figure 9]. Three patients were operated with multiple Z-plasties of

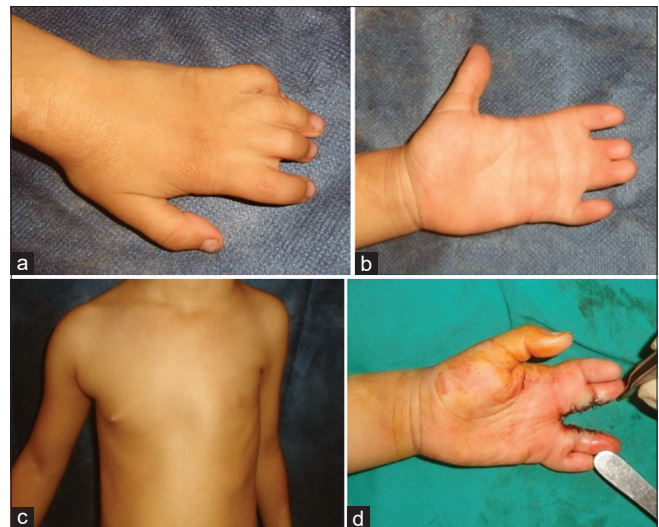


Figure 5: Simple incomplete syndactyly of 2nd and 3rd webs of the left hand associated with hypoplastic LT pectoralis major muscle (Poland's syndrome). (a) Left hand, dorsal aspect. (b) Left hand, palmar aspect. (c) Hypoplastic left pectoralis major muscle. (d) Left hand after separation of 3rd web

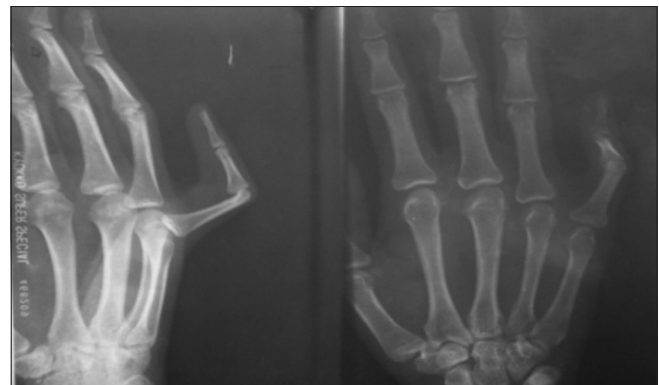


Figure 7: X-ray of the right hand, anteroposterior and lateral views revealed flexion contracture of the right little finger, narrow proximal interphalangeal joint space and hyperextension of metacarpophalangeal joint

Table 6: Syndactyly cases of the study

Syndactyly type	Simple incomplete	Simple complete	Complex complete	Total
Solitary anomaly	9	5	3	17
Associated with other anomalies	2	1	2	5
Part of a syndrome	1	0	1	2
Unilateral	5	5	3	13
Right	3	3	2	8
Left	2	2	1	5
Bilateral	7	1	3	11

the constricting rings were done for Type I and Type II deformities, digits separation with web reconstruction using dorsal triangular flap and grafting was done for Type III deformity (acro-syndactyly). For Type IV deformity with shallow 1st web, web deepening using volar rectangular flap was done.

Cleft hand was detected in five patients. All patients were females with two patients were identical twins and had cleft hands cleft feet syndrome [Figure 10]. Four patients (80%) had other hand anomalies; syndactyly. Type IIa was the most common type (40%). One patient was operated in whom excision of the V-shaped defect with web space reconstruction using transpositional flaps from the adjacent fingers were done.

Symbrachydactyly was detected in three patients. Short finger type was the most common type; was detected

Table 7: Clinodactyly cases of the study

Finger affected	Unilateral		Bilateral	Total
	Right	Left		
Little finger	0	1	3	4
Ring finger	1	0	0	1
Total	2	0	3	5

Table 8: Constriction band syndrome cases of the study

Type of deformity	Unilateral	Bilateral
Type I	0	1
Type II	0	1
Type III	0	1
Type IV	2	3



Figure 8: Right little finger clinodactyly; (a) right hand, dorsal aspect. (b) Right hand, palmar aspect. (c) X-ray of right hand, anteroposterior view

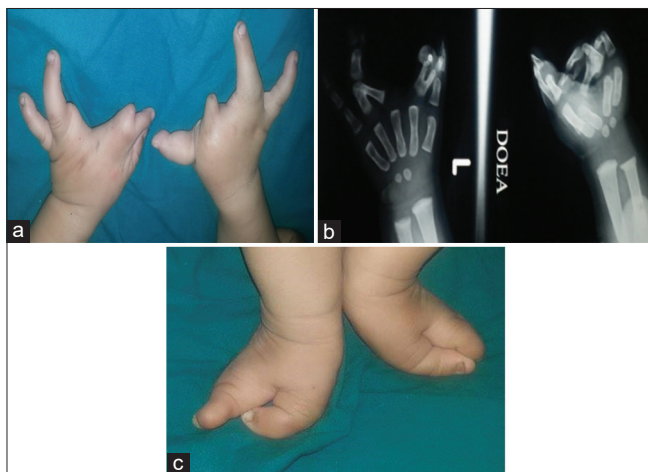


Figure 10: Split hands split feet syndrome, bilateral cleft hand Type 3 with complex complete syndactyly of the right 1st web, simple incomplete syndactyly of the left 1st web, left thumb duplication Type II and duplicated proximal phalanx of the left index and distal phalanx of the left ring finger. (a) Both hands, dorsal aspect. (b) X-ray of both hands, anteroposterior view. (c) Both feet

in all cases [Figure 11]. All patients had associated hand anomalies.

Three patients had brachydactyly; two females and one male. Types of brachydactyly encountered in the study were Type E₁ (short metacarpal bone), Type D (short distal phalanges of both thumbs) and Type A₁ (short middle phalanges). Type E₁ was unilateral; involving the left 4th metacarpal bone whereas patients with Type D and Type A₁ had bilateral hand deformity. The patient with Type E₁ brachydactyly was operated using distraction osteogenesis for 6 weeks [Figure 12]. About 4 mm distraction was achieved, but the case was complicated with earlier bony consolidation.

Vascular malformations were detected in three patients; two females and one male. All of them were venous malformations as demonstrated by soft tissue ultrasonography.

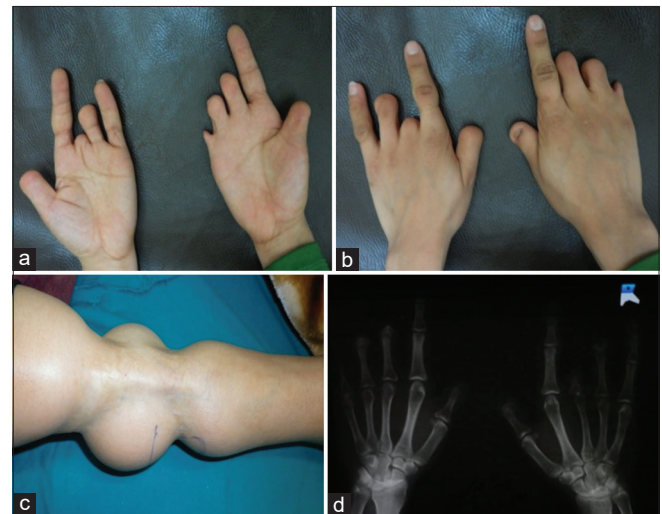


Figure 9: Constriction band syndrome of both hands Type IV associated with right leg constriction rings. (a) Both hands, palmar aspect. (b) Both hands, dorsal aspect. (c) Right leg showing constriction rings. (d) X-ray of both hands, anteroposterior view



Figure 11: Female patient, 5 years old with right hand symbrachydactyly (short finger type) and simple incomplete syndactyly and right hypoplastic thumb (Type II). (a) Right hand, dorsal aspect. (b) X-ray of right hand, anteroposterior view

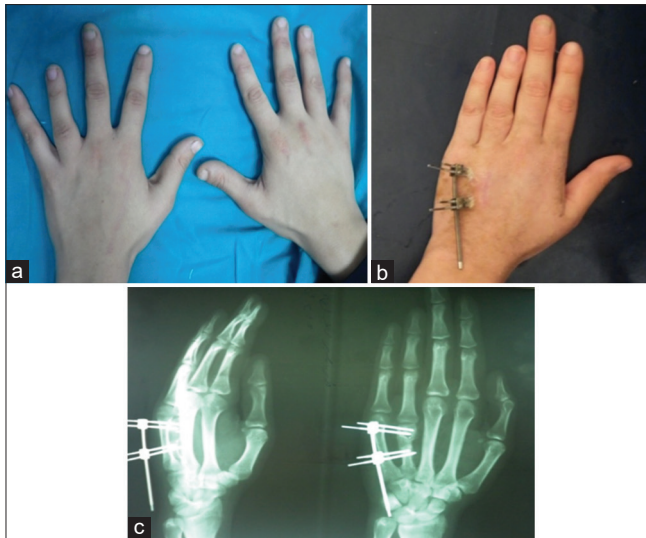


Figure 12: Left ring finger brachydactyly Type E₁. (a) Both hands, dorsal aspect. (b) Left hand with distractor applied. (c) X-ray of left hand, anteroposterior and lateral views

Congenital trigger finger was detected in one patient who was presented at the age of 4 months with middle fingers of both hands were affected. Repeated extension splintage was applied for 2 months with mild improvement of the deformity was noticed.

DISCUSSION

Congenital hand anomalies are more common among males with male to female ratio is 3:2.^[4] In this study, 59% of patients were females. This can be explained by social concepts as the presence of such anomalies in a female child threatens her social life and marriage opportunity.

Hand anomalies are either isolated, associated with other anomalies or may be a component of a syndrome. Some anomalies have genetic background with positive family history whereas others occur sporadically.^[8] This study revealed that 67% of patients had isolated hand anomaly/anomalies whereas 33% of patients had multiple anomalies. Family history was positive for 26 patients.

Although the aetiology of most hand anomalies in our community is still unknown, the common positive history of consanguinity in Upper Egypt may be a significant etiological factor for the occurrence of certain hand anomalies.

The mean age of cases presentation is significantly late. The delay of seeking medical advice can be attributed to lack of awareness and education with low socioeconomic

status and wrong traditional concepts in Upper Egypt community.

Failure of differentiation and duplication are the most frequent anomalies with polydactyly is the most common individual diagnosis with an incidence of about 1 per 1000 live births.^[2,8] This study also revealed that failure of differentiation and duplication anomalies were most frequent deformities (90.6%) with polydactyly was the most common anomaly encountered.

In this study, ulnar polydactyly was more frequently found which is similar to literature.^[3] The majority of patients were females (72%), but this can be explained by relatively higher percentage of females included in the study (59%).

According to Wassel classification, radial polydactyly was classified into seven types based on the level of duplication and number of bony components.^[9,10] Wassel Type IV is the most common type.^[8] In this study, Wassel Type II was the most frequent type which can be attributed to the limited number of radial polydactyly cases in the study.

Syndactyly has an incidence of 1 per 2000–2500 live births. Male to female ratio is 2:1. It is more commonly bilateral.^[11] In this study, syndactyly was relatively common (36% of patients) with females were more commonly affected which can be explained by relative higher percentage of females in the study and small number of cases.

The 3rd web is the most commonly involved, followed by 4th web and the 2nd web.^[6] In this study, the 3rd web was the most common web affected followed by the 2nd web (42%), however, firm conclusions regarding percentages cannot be obtained due to small number of cases.

Syndactyly treatment is mainly surgical with the optimum age is 2 years old. However, in certain situations, earlier intervention may be essential. Several techniques were described including dorsal triangular flap or dorsal rectangular flap with zigzag incision and full thickness grafting if needed.^[5,12] Mean age for surgical treatment of syndactyly in this study was 2.5 years. Various techniques were utilized with zigzag incision, and full thickness graft was the most common surgical modality applied.

Complications after syndactyly separation are related to scar contracture and include web creep (2–24%), flexion

contracture, deviation of digits, rotational deformity and nail deformity.^[12] In this study, these complications especially web creep were encountered.

Camptodactyly affects 1% of live births, and it is bilateral in 2/3 of cases.^[13] However, in this study, the deformity was unilateral in all cases, but this cannot be conclusive finding due to the limited number of camptodactyly cases (four patients).

Conservative treatment (extension splintage) is indicated for mild flexion deformities whereas surgical treatment is indicated for cases with severe deformities. Several procedures were described and the choice of the proper one depends on the degree of flexion deformity, status of periarticular structures and the balance of forces around the joint.^[13] In this study, all patients needed surgical intervention because of severe flexion deformity and late presentation. The frequent occurrence of post-operative complications can be attributed to late presentation, lack of effective physiotherapy and delayed or irregular follow-up visits.

The incidence of clinodactyly is underestimated as many cases are asymptomatic. Familial clinodactyly is the commonest type. It is usually bilateral and involves the little finger.^[14] Clinodactyly was detected in five patients of whom familial clinodactyly was encountered in two patients (40%), but this can be related to small number of cases. Residual angular deformity was noticed in the operated case which can be explained by the fact that no corrective bony osteotomy was done.

Constriction band syndrome affects 1 per 12,000–15,000 live births. Four types were identified based on the severity of the deformity.^[15,16] The management is dependent on the severity of the deformity and procedure choice depends on the type of the deformity present.^[17] In this study, five patients had constriction band syndrome. Treatment plan was individualised according to type of deformity.

Cleft hand may be associated with other anomalies such as syndactyly, polydactyly or triphalangeal thumb.^[18] Five types of cleft hand were recognised based on the status of the 1st web space.^[19] It is usually bilateral and familial.^[6] In this study, cleft hand was more commonly bilateral (60%) with Type IIa was the most common type and syndactyly was the most frequent associated anomaly; however, these results may be biased by the limited number of cases.

Symbrachydactyly has low worldwide incidence and usually sporadic.^[6] Four types were described based on severity of the deformity.^[5] In this study, three patients had symbrachydactyly with all cases were sporadic.

Brachydactyly may be sporadic or heritable. It may be present as an isolated anomaly, associated with other anomalies or may be a component of a syndrome. The surgical treatment of aims at lengthening of the involved bony element which can be achieved through two methods; distraction osteogenesis or single stage lengthening using bone graft with soft tissues release.^[20] In this study, all brachydactyly cases were sporadic; however, this may be attributed to limited number of cases. The earlier bony consolidation that complicated the operated case can be attributed to lack of follow-up and lack of patient's awareness regarding daily distraction.

CONCLUSION

The incidence of congenital hand anomalies in Upper Egypt community is underestimated. Early and effective management is important to provide adequate hand function and to ameliorate the serious social and psychological impacts on the patient and the parents. It is better to be achieved by multidisciplinary team and should be individualised for every patient.

According to the Central Agency for Public Mobilization and Statistics, the official governmental organisation responsible for population census, the estimate population of Sohag Governorate by the end of 2014 was about 4.6 millions.^[21] Our University Hospital is a tertiary referral center; it receives patients from all the areas of Sohag Governorate and it also receives some cases from the governorates south to it, namely, Qena, Luxor and Aswan. However, we are not the only hospital that receives these anomalies and even in our hospital; there are two departments dealing with this scope of anomalies, namely, plastic surgery and orthopaedic surgery departments. Therefore, we cannot conclude that this is the real population-based prevalence of hand anomalies, but indeed it represents a great part of real prevalence. These estimate prevalence is underestimated due to several cultural and social reasons. Of these reasons, some parents do not seek medical advice for their children because they believe that it is not good to try to change what was created by God's hand.

The post-operative complications can be minimised by adequate pre-operative assessment of patients, proper choice of the operative procedure and adequate post-operative physiotherapy.

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Conflicts of interest

There are no conflicts of interest.

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