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Surgical management of windblown hand: results and literature review

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Abstract
Purpose Windblown hand is a congenital anomaly characterized by multiple hand deformities. The condition is extremely rare as shown by the paucity of cases reported in the literature. The deformities, which can result in a cosmetically unsatisfactory appearance and, if left untreated, maladaptive behavior, are progressive in nature. Consequently early treatment is necessary.
Methods We operated upon 23 hands in 18 patients (age range at surgery 6 months to 16 years) at a tertiary care center over a period of 7 years. The patients were followed for an average period of 7 years. The surgical approach was chosen based on the severity of the condition according to Zancolli and Zancolli's classification [Hand Clin 1:443–456 (1985)].
Results According to the criteria of Wood and Biondi [J Hand Surg 15A:431–438 (1990)], of the 23 hands operated upon, 17 had excellent cosmetic results, and 15 had excellent functional results. The results were better in patients undergoing early surgery—before the age of 2 years. Relapse of the deformity to a lesser extent than the original condition was seen in two hands at the last follow up.
Conclusion Definitive conclusions on this condition cannot be drawn due to limited experience in the surgical management of this rare condition. We believe that early surgical management is probably the best option available for the patient based on the results obtained. Early surgery and good post-operative compliance from patients can facilitate successful management of this rare condition with predictable results.

Keywords Congenital · Deformity · Flexion contracture · Ulnar drift · Windblown hand

Introduction

Windblown hand, also known as the congenital ulnar drift of fingers, is a rare congenital anomaly with characteristic hand deformities. It was first described by Boix in 1897 [1], and since then various etiologies and syndromic associations have been proposed. Zancolli and Zancolli [2] described this condition as a type of segmental arthrogryposis and divided it into three types based on increasing severity. Deformities include ulnar drift of the fingers at the metacarpophalangeal joint, flexion and adduction contracture of the thumb and first web space contracture; they are usually bilateral and symmetrical. The association of windblown hand with syndromes like Freeman–Sheldon syndrome is well known. Associated craniofacial and foot anomalies may also be present [3–5]. Published studies advise early surgical treatment for these patients as a severely deformed hand may lead to functional limitations and an unacceptable cosmetic appearance [6]. However, to date, there is no standard management protocol due to the

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rarity of the condition; consequently, the basic principles in managing congenital deformities of the hand are followed. Nonsurgical management in the form of splinting and surgical techniques, including soft tissue procedures and bony correction, have been reported. A number of surgical techniques, ranging from contracture release, excision of abnormal bands of tissues, tendon transfers and osteotomies, have been described in the literature based on a few reported cases [7–9]. The efficacy of splinting, the effectiveness of surgery, the incidence of relapse following nonsurgical and surgical management and the appropriate timing of surgery are still a matter of debate. In most instances, management is based on individual expertise and experience.

Materials and methods

We operated upon 18 patients (12 males and six females; age range 6 months to 16 years) with windblown hand presenting at the hand surgery unit in our institution from 1996 to 2002. Fifteen patients presented before 2 years of age, one patient at 5 years and two were adolescents older than 12 years. All cases were bilateral, and 23 hands in these 18 patients were operated on. The right hand alone was operated in ten patients, the left hand alone in three patients, and five patients underwent bilateral staged surgical correction. Associated craniofacial anomalies were present in ten patients, and foot anomalies were present in nine patients (congenital talipes equinovarus, five patients; congenital vertical talus, two patients; syndactyly of the toes, two patients). All patients presented to us were accompanied by their parents. Complaints included an inability or difficulty in grasping and holding onto objects; older children had complaints of difficulty with writing and participating in sports activities. All patients complained of worsening of the deformity, and two adolescent patients had difficulty in perineal care with their left hand. Despite these disabilities, up to presentation at our hospital, the patients had sought treatment mainly for the unpleasant cosmetic appearance. All deformities in a single hand were addressed at the same sitting. Bilateral deformities were staged with a minimum interval of 6 months.

We classified the hand deformities as per Zancolli and Zancolli's classification [10] (Table 1) into three types, and the type of surgery was decided according to the severity of the deformity. In our series, 11 hands belonged to type I, eight hands, type II and four hands, type III. Patients with type I deformity underwent surgical procedures ranging from correction of finger contracture by release of abnormal subcutaneous bands and full thickness skin graft [11] to cover the defect, correction of thumb web contracture and local flap cover by the way of dorsal rotation or index transposition flap. Patients with types II and III deformity depending on severity needed in addition relocation of the subluxed extensor tendons, metacarpophalangeal arthrodesis of the thumb, metacarpal corrective osteotomy and crossed intrinsic transfers [12]. The surgical procedures carried out in our series are tabulated in Table 2.

Surgical techniques

Finger contracture

The flexion contracture of the fingers was addressed through a transverse relaxing incision just distal to the metacarpophalangeal joint on the volar aspect. The tight subcutaneous tissue and the abnormal bands were released in all hands. The long flexors to the fingers were not disturbed. The resultant defect after contracture release was covered with a full thickness skin graft. The corrected position was maintained with Kirshner's (K) wires.

Patients with types II and III deformity also underwent relocation of the extensor tendons through a transverse dorsal incision in the region of the metacarpophalangeal joint. Centralization of the extensor tendons was achieved by plicating the radial sagittal bands. The ulnar drift was corrected by means of crossed intrinsic transfers. The ulnar intrinsics were released at the level of the

Table 1 Zancolli and Zancolli's [10] classification of hand deformities in windblown hand

Type	Deformity	Our series
I	Fasciocutaneous deformity, with short skin and abnormal subcutaneous bands	11 hands
II	Type I + short tendons of the wrist, thumb, and fingers	8 hands
III	Type I and II + short ligaments and capsules with bony deformity	4 hands

Table 2 Surgical procedures

Reconstruction procedures done	Number of hands
Fingers	
Contracture release and skin graft	19
Extensor tendon relocation	12
Metacarpal corrective osteotomy	4
Crossed intrinsic transfers	9
Thumb	
Release and dorsal rotation flap	8
Release and index transposition flap	13
Metacarpophalangeal arthrodesis	2
Skin graft	2

metacarpophalangeal joint and transferred radially. In the case of a bony abnormality, a radially based closing wedge corrective osteotomy of the metacarpals in the subcapital region that avoided the physes was performed to correct the ulnar deviation of the metacarpal head through the same incision and subsequently fixed with K wires.

Thumb and first web contracture

Reconstruction of the defect following the first web contracture release was performed by means of local transposition or rotation flaps. No distant flaps were used, even in type III deformities. Local flaps, either an index transposition flap or a dorsal rotation flap, were used in the majority of cases. In two dominant hands belonging to the adolescent patients, a metacarpophalangeal joint arthrodesis of the thumb was carried out because of severe deformity and to provide greater stability in writing and other skill-requiring activities. In two hands of adolescent patients, the wide defect following release of first web space and thumb adduction contracture was reconstructed using a full thickness skin graft from the groin. The surgical procedures are explained in Figs. 1 and 2.

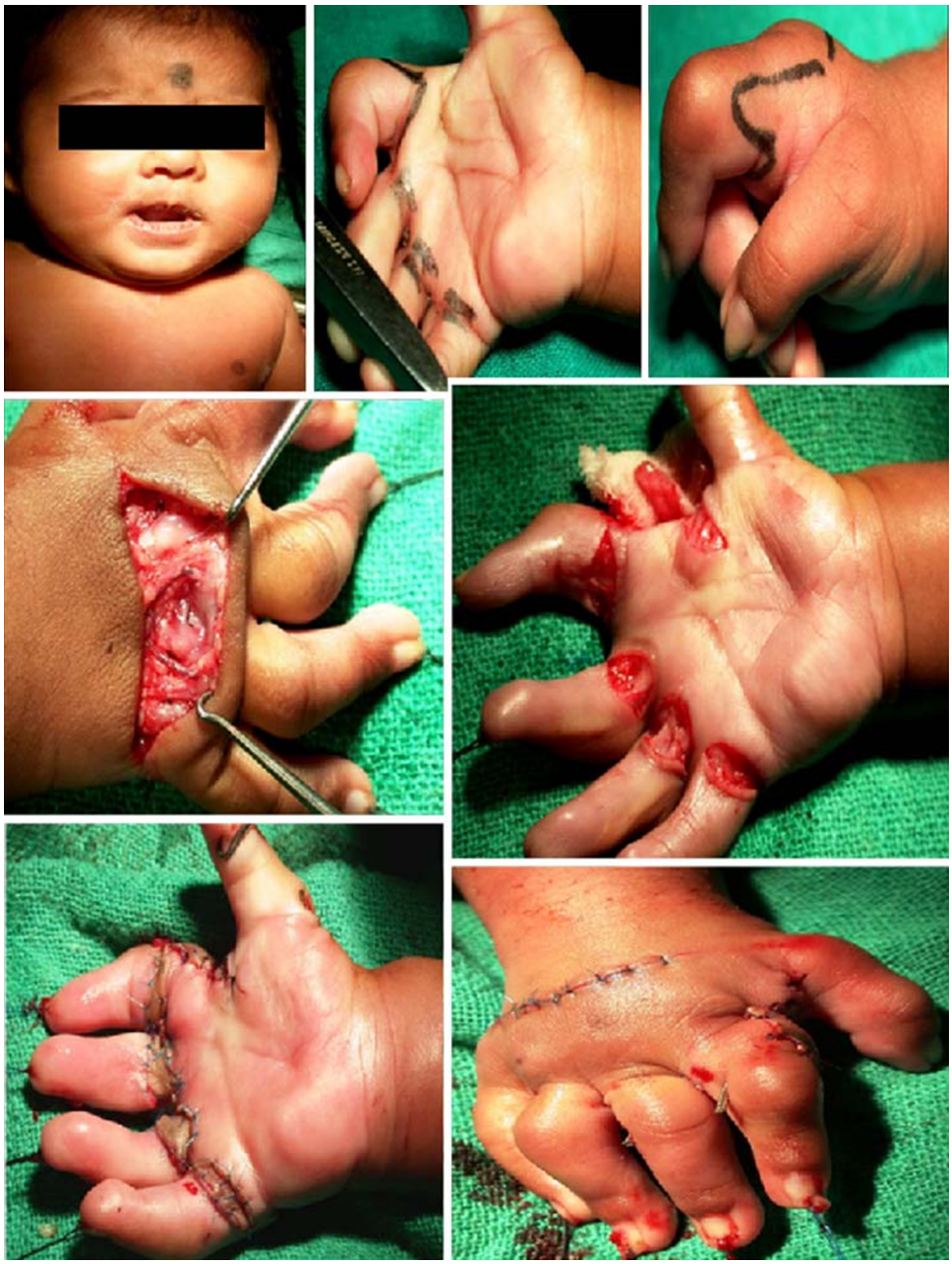

Fig. 1 A 6-month-old child with type II deformity underwent finger flexion contracture release with skin graft cover and K-wire stabilization. Relocation of the subluxed extensor tendons was performed through a dorsal transverse incision. The defect following release of the web contracture is covered with a local flap

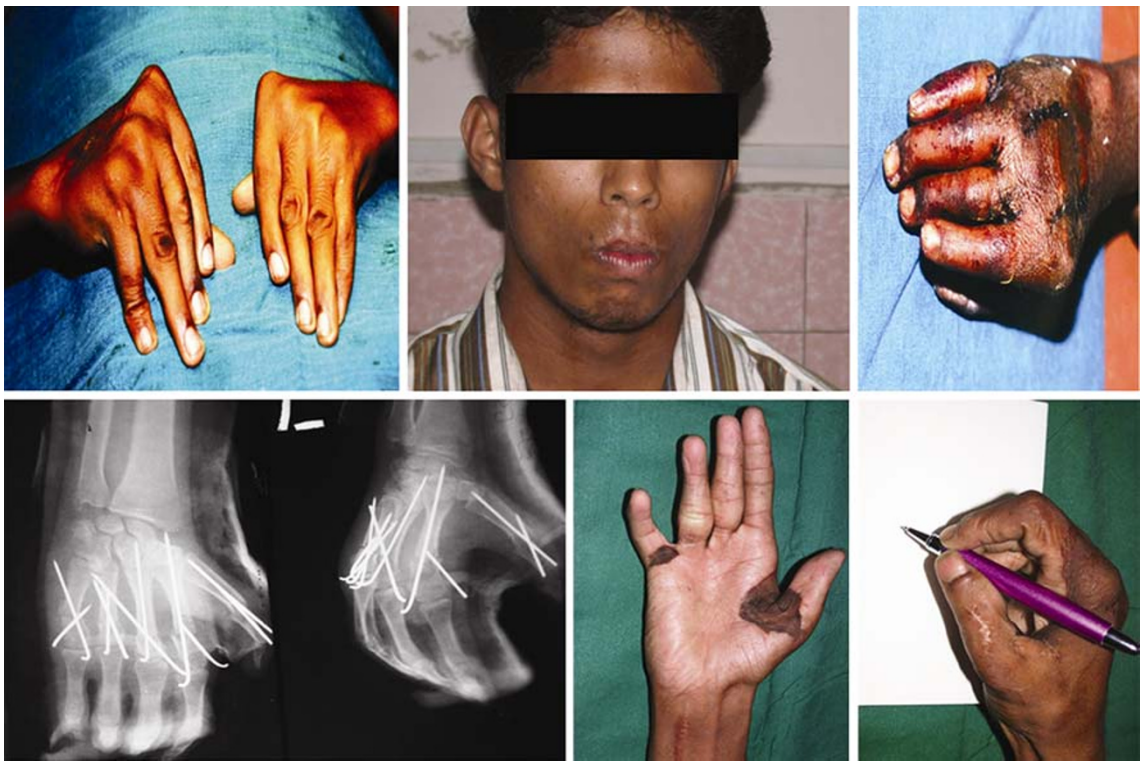

Fig. 2 An adolescent with type III deformity underwent sub-capital metacarpal osteotomies and metacarpophalangeal arthrodesis in the dominant hand in addition to the soft tissue procedures

Post-operative rehabilitation

The hands were immobilized in a plaster splint in the corrected position for a period of 2 weeks. The K wires were removed after 2 weeks once the grafts had settled, and active movements were encouraged using a dynamic corrective splint. The splint also helped in applying radial traction to maintain correction. The splint was used continuously for the first 3 months and then only at night time for the subsequent 2 months. In case of bilateral corrections, the other limb was operated upon after a minimum period of 6 months.

Results

The patients were followed up for an average period of 7 years (range 5–12 years). Our results were analysed using two parameters, namely, cosmetic appearance and functional outcome of each hand, as described by Wood and Biondi [13]. The cosmetic result was excellent in 17 hands, good in four hands and fair in two hands (Table 3). Of the 17 hands with excellent cosmetic results, 13 belonged to children who presented before 2 years of age. One of the patients whose cosmetic result was graded as fair had a poor functional outcome with limitations in activity.

Table 3 Results

Cosmetic outcome	
Excellent	17 hands (13 hands were operated before 2 years)
Good	4 hands
Fair	2 hands
Poor	Nil
Functional outcome	
Excellent	15 hands (14 hands were operated before 2 years)
Good	4 hands
Fair	3 hands
Poor	1 hand

The functional result was classified as excellent in 15 hands, good in four hands, fair in three hands and poor in one hand. Fourteen of the fifteen hands with excellent functional results were operated on before the patient was 2 years old.

Flap tip necrosis was seen in two patients, but the viability and survival of the flap was not compromised in any patient. At the last follow-up, relapse of the ulnar drift was seen in two hands, relapse of thumb web contracture was seen in one hand and relapse of finger contracture was seen in one hand. Although the cosmetic result was acceptable in these patients, one patient had a poor functional result according to the criteria of Wood and Biondi [13]. The

patient was advised surgical correction but was not willing to undergo another surgical procedure. Both patients (9%) who had a relapse were operated on before 2 years of age and had a type II deformity on presentation. There were no incidences of physeal injury and subsequent growth arrest in patients who underwent osteotomies and K-wire fixation.

Discussion

Windblown hand is a type of arthrogryposis with characteristic hand deformities, namely (1) flexion contracture of fingers, (2) ulnar drift of fingers at the metacarpophalangeal joint and (3) thumb adduction with first web contracture. Although the etiology is obscure, many hypotheses and syndromic associations have been described [3]. It is most commonly accepted to be a segmental arthrogryposis as described by Zancolli and Zancolli in 1984 [2], while abnormalities in palmar fascia were described as a causative factor by Jacquemain in 1967 [14]. . The Freeman–Sheldon syndrome (craniocarpotarsal dysplasia), described by these two researchers in 1938 [15], and whistling face syndrome, described by Burian in 1963 [4], are the most common syndromes associated with windblown hand, although the latter can occur in isolation. Associated deformities include clubfoot, congenital rocker bottom foot, small oral apertures, mask-like facies, long upper lip, small nose and skull abnormalities. The deformity is usually bilateral, but the unilateral condition has been described [16]. There has also been a report of familial associations and risks [17]. The ulnar drift of the fingers is the most common abnormality, but thumb and first web abnormalities may present a significant impairment for the patient.

Early splintage for treatment of windblown hand had been shown to be effective [6, 18]. Although the results of surgical management in our series were better in younger children, we do not have similar controls to prove that early splinting is not beneficial. Splinting requires a great deal of compliance from the child and the parents. Even if correction is achieved by splinting, it takes a long time and correction may be suboptimal. Thumb adduction contracture had been shown to be difficult to correct with splinting alone [6]. Splinting may also result in a loss of valuable time in the child's formative years and interfere with schooling. The evidence currently available is inconclusive in terms of being able to determine whether splinting achieves consistent results over the long term.

All soft tissue structures have been shown to be abnormal in a windblown hand. Delay in surgical correction may lead to progressive contractures and bony abnormalities. In addition, early surgical correction is technically easier for the surgeon and also achieves a better correction, avoids bony procedures, allows early proper hand usage and minimizes maladaptive behaviors. The deformity has been shown to be best addressed before the patient is 3 years of age (Kallianen et al. [6]). The results in our series were also similar, but we experienced relapse of the deformities in two patients who were operated on before they were 2 years old. This once again raises the question of the appropriate time for surgery and the efficacy of early surgery in preventing relapse through skeletal maturity and the need for long term splinting. Anesthetic complications, such as an increased incidence of malignant hyperthermia, have been reported, and caution should be exercised [19].

The defect following the release of flexion contracture of the fingers was covered with a full thickness skin graft. We also used a K wire to maintain the fingers in extension and prevent graft contraction. Recently, local flap coverage of the defects has been described with good results. The use of a flap allows early mobilization without the need for a K wire and is also free from contraction. We achieved good results after using a skin graft and K wires and were able to maintain the correction and finger movements.

The ulnar drift of fingers represents a challenge to the surgeon, and this condition is prone for relapse. The flexor–extensor balance should be restored. Soft tissue procedures alone may not achieve a complete correction, and bony procedures have been shown to produce greater correction and achieve a better outcome. Metacarpal subcapital osteotomies (wedge or dome) [17] and osteotomy of the base of the proximal phalanx have been described [12]. We used a radially based closing wedge osteotomy in our patients. The distance between the osteotomy lines on the ulnar and radial side depend on the amount of flexion and ulnar deviation of the digits.

Following release of the thumb adduction and web space contracture, we used local flaps to cover the defects in majority of the hands. In 2 hands with severe first web contractures the defect after release could not be covered with a local flap. In these hands we used a full thickness skin graft from the groin to cover the raw area. Distant pedicled flaps have been described in these situations but [20] we did not prefer distant flaps because it needs to be staged, impairs mobilization, poor compliance in a child and associated with donor site morbidity.

We surgically fused the metacarpophalangeal joint of the thumb in two hands that belonged to adolescent patients with a severe deformity in the dominant hand. Arthrodesis offers a stable thumb in the corrected position, provides greater stability in the pinch and helps prevent relapse, which is common in hands with severe deformities.

Windblown anomaly is a rare condition and, therefore, experience in surgical as well as nonsurgical management is extremely limited. We personally believe that the hand

deformities are best managed surgically when the patient is very young. The functional results were definitely better in patients undergoing surgery at an early age, but early surgery also seems to be associated with an increased incidence of relapse. Consequently, long-term control-based studies are needed to provide data on which to draw definitive conclusions. However, due to the rarity of the condition, this may be practically difficult. Dynamic corrective forces facilitate correction of the deformities, and post-operative splinting is extremely valuable. Early surgery coupled with long-term dynamic splinting may provide the best option for these patients and minimize the chances of relapse. Parents should be warned of the relapsing nature of the condition before surgical management is contemplated. These children compensate well, such that these deformities rarely produce a major functional limitation. Maladaptive learned behaviors, even though they are functional, should be minimized, and a cosmetically normal hand in a growing child of the surgeons.

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