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The surgical treatment of congenital hand deficiency

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Abstract
This review describes the two types of congenital cleft hand deformity, stressing the important ways in which they differ and summarises the surgical techniques that may be used in their treatment.

Introduction

The two conditions are frequently grouped together under such titles as lobster-claw hand, ectrodactyly, oligodactyly and split hand. These terms are confusing, sometimes hurtful to parents and best discarded. Lange (1936) and Barsky (1964) have differentiated the typical from the atypical cleft hand and the two conditions are distinct, imposing a different surgical approach while sharing similar functional problems.

Central longitudinal failure of development: the typical cleft hand

Barsky (1964) credits Hartsinck (1770) with the first description of a case and Birch-Jensen (1949) claims that the first use of the term cleft hand was by Kummell (1895). The deformity (Fig. 1) has been described by Flatt (1977) as being "a functional triumph and a social disaster". It is rare and has been estimated at invovling 0.4 (Rogala, *et al.*, 1974) to 0.14 (Birch-Jensen, 1949) per 10,000 live births. Cases occur sporadically but the condition may be familial and is inherited as an autosomal dominant with mixed penetration (Graham and Badgley, 1954; David, 1974). Recessive inheritance may occur (Flatt, 1977). Both hands and feet are frequently involved and the cleft is 'V' shaped. Other anomalies sometimes affect the limb and the condition may be associated with abnormalities in various organs. Genetic counselling is particularly important for these patients (David, 1974). In the common form the middle finger is missing, with or without the metacarpal, and suppression of more digits in a radial direction occurs in other cases. Ring finger absence alone is rare and when the digits are so severely suppressed as to produce a single digital variety this is the little finger. Syndactyly frequently affects the digits bordering the cleft and where the thumb is involved this can severely affect function. Several metacarpal variations are described. This bone may be totally missing in the cleft or two metacarpals may seem to support one finger which then resembles severe compound syndactyly on radiographs. Alternatively a bifid metacarpal may support two digits. Several subclassifications have been proposed (Watari and Tsuge, 1979; Nutt and Flatt, 1981; Tada *et al.*, 1981). The cause of the defect is probably due to an abnormality in the apical ectoderm of the limb bud which produces a wedge shaped gap in the hand (Muller, 1937). The much quoted centripetal theory (Maisels, 1970) attempts to explain a progression of clefting from a simple central soft tissue gap to complete suppression of all digits.

Management

The first question is: do these patients need

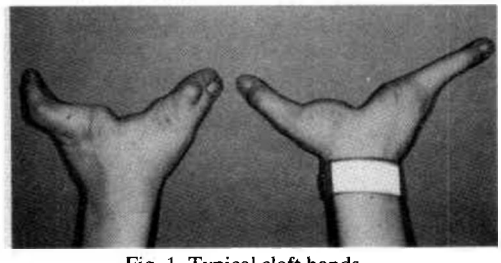

Fig. 1. Typical cleft hands

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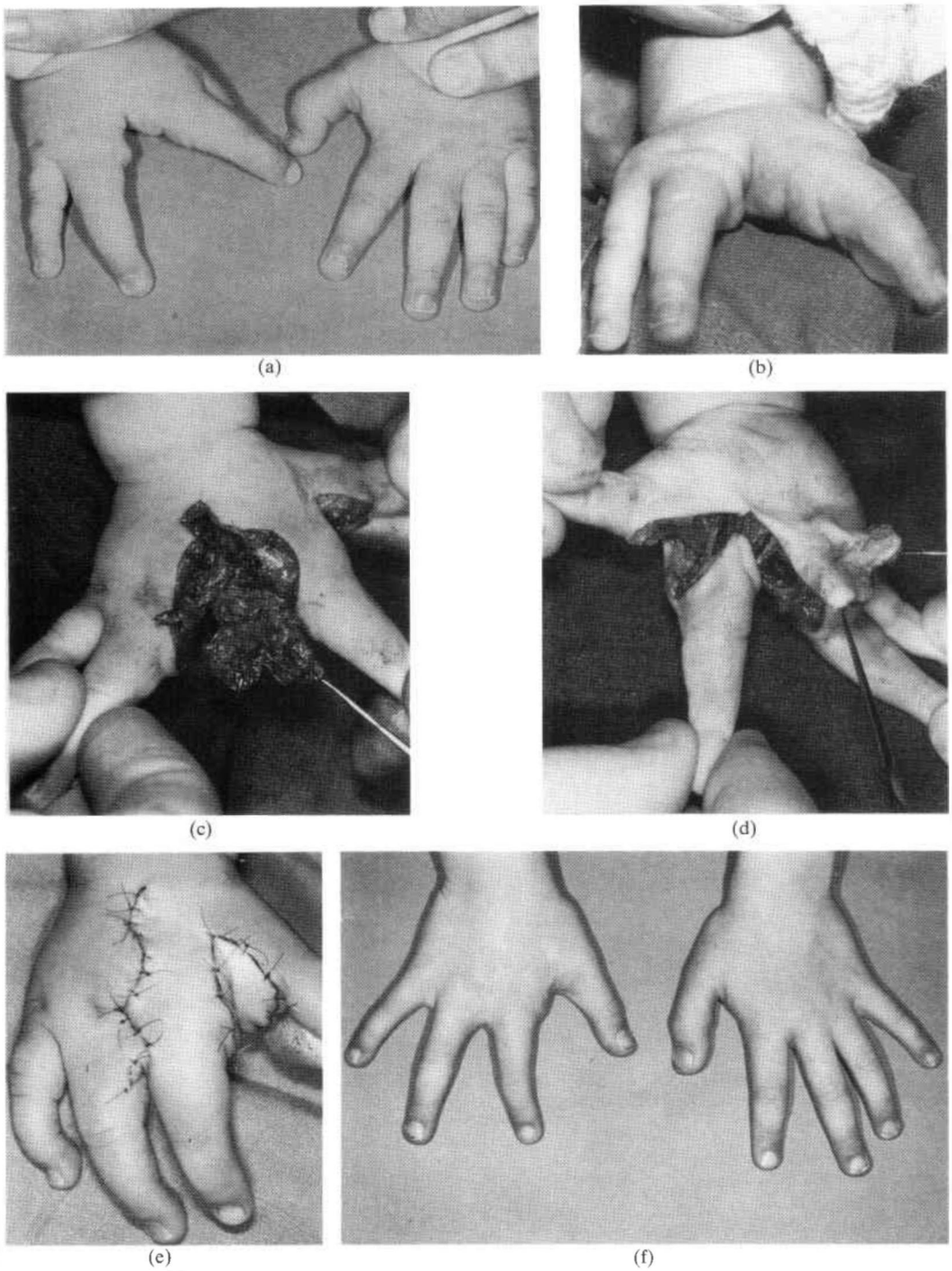


Fig. 2. Reconstruction of typical cleft hand by the Snow-Littler technique
(a) and (b) Pre-operative views
(c) and (d) Flap elevation
(e) Flap inset into thumb web and cleft closure
(f) Post-operative view

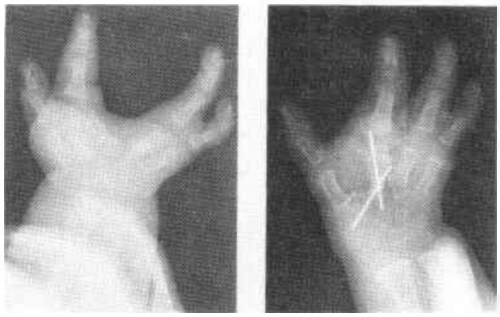


Fig. 3. Snow-Littler technique. Radiographs to show pre- and post-operative bony anatomy. The third metacarpal has been partially excised. The index metacarpal has been transected proximally, shifted ulnarwards and fixed to the base of the third metacarpal.

treatment? Cases which have not undergone surgery for a variety of reasons adapt and function extremely well often possessing excellent pinch and grasp despite deformaties which are very disfiguring. Experience has shown, however, that function can be improved by early surgery and this is usually performed between 1 to 2 years. Direct closure of the soft tissue cleft can be performed in minor cases and attempts should be made by using a local flap (Barsky, 1964) to reconstruct a web commissure. Recurrent finger separation by drifting of the border metacarpals following surgery should be prevented by the reconstruction of a transverse metacarpal ligament protected by temporary transmetacarpal Kirschner wiring. Free tendon grafts are frequently used for this technique and may be available locally as both the extensors and flexors of the missing finger are usually present in the ray and are fused over the end of the metacarpal remnant. If not, a palmaris longus graft can be used to hold the adjacent metacarpals together (Ueba, 1981). Frequently after excision of a redundant metacarpal and before closure of the cleft the collateral ligaments of the metacarpo-phalangeal joint need reconstructing using local tissue flaps including periosteum to prevent instability and deviation. In more severe cases where the residual digits are widely separated by a deep gap, spare skin from either side of the cleft should not be discarded. Local flaps fashioned from this tissue provide excellent skin for separation of the syndactyly which frequently affects the adjoining digits. This is a particularly valuable technique when the first web is involved with the thumb adducted and is the treatment of choice for the common situation where the middle finger is missing and the thumb partially fused to the side of the index. It can be used in combination with a shift of the index finger metacarpal in a ulnar direction with an osteosynthesis at the base of the middle finger metacarpal. The Snow and Littler procedure (1967) (Figs. 2 and 3) is an excellent technique which employs these two manoeuvres but other methods combining skin flaps from the cleft and a metacarpal shift have been described more recently (Miura and Komada, 1979; Ueba, 1981). For the less severe cases of central longitudinal deficiency the surgery is challenging, rewarding and gives excellent functional and aesthetic results (Fig. 4). Where suppression has been more severe the aims are less ambitious. When there are only two digits a combination of flap surgery, rotation osteotomy and tendon readjustment may restore a crude pincer grip. Where only 1 digit remains, treatment is more difficult and the results less satisfactory. Some of the methods discussed in the next section then become applicable to restore a simple prehensile function to the hand. These include microsurgical free toe transfer (May *et al.*, 1981).

Transverse failure of development distal to the carpus: the atypical cleft.

This condition is a different entity (Fig. 5). Treatment is generally more difficult and functional results less rewarding. The digital rays are missing at variable levels from the metacarpal distally. In less severe forms the thumb and little finger may be totally or partially preserved so that the child has a crude grasp and a useful pinch. The cleft of the hand is then 'U' shaped and the gap between the

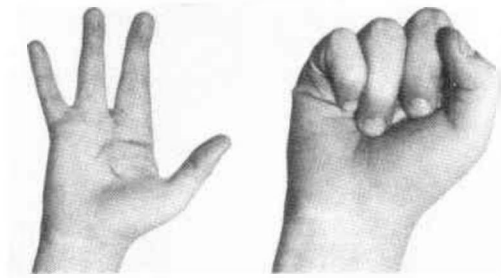


Fig. 4. Post operative function following Snow-Littler procedure for repair of typical cleft hand.

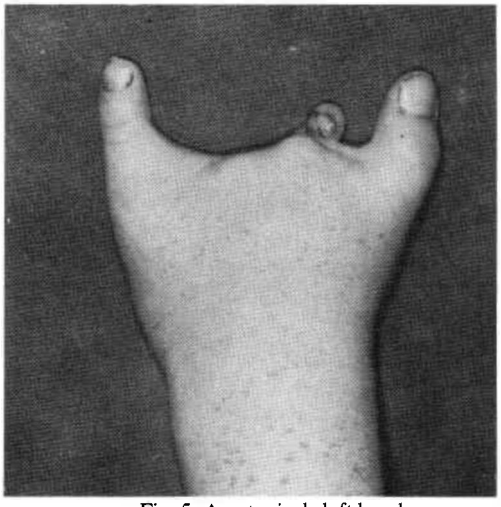
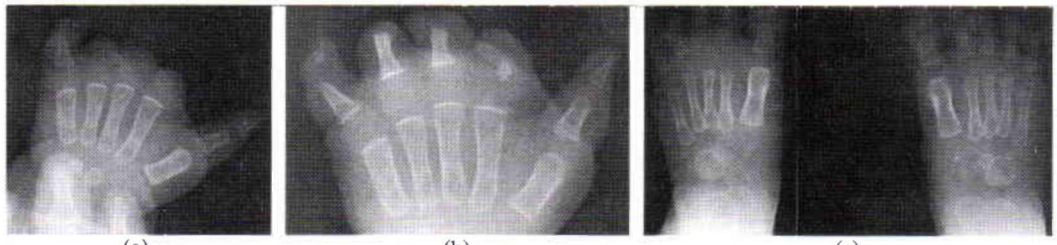


Fig. 5. An atypical cleft hand.

border digits is occupied by soft tissue nubbins with rudimentary nails representing finger remnants. This has led Buck-Gramcko (1971) to classify the deformity as a form of symbrachydactyly. The condition is sporadic and one hand is usually affected. Associated anomalies are less frequent. Unlike the typical cleft hand the monodactylous type of this deformity spares the thumb. The aetiology of the condition is probably a primary transverse failure of bone formation with subsequent disturbance of the soft tissue.

Management

Where bone and soft tissue are preserved distal to the carpus surgical reconstruction with the provision of sensate prehension has advantages over the use of an opposition device. Various options are available and each case must be considered separately after careful assessment of what is available and what is required. Simple excision of small nubbins of skin and deepening of the cleft between an adequate thumb and little finger may enhance function by a simple operation with low morbidity but when border digits are very short some lengthening operation must be undertaken. Free phalangeal transfer from the toes has had mixed success over the years (Carroll and Green, 1975; Goldberg and Watson, 1982) but the technique described by Buck-Gramcko and Pereira in 1990 in which a proximal toe phalanx is transplanted with its periosteum has been consistently successful (Fig. 6). These authors have shown that the phalanx survives and grows for a longer period if transplanted early and have been able to reconstruct a joint. The bone requires an adequate skin pocket for survival and when nubbins of tissue are large these should not be discarded too readily before considering this technique. Unstable digits with missing phalanges, in particular the thumb and little finger when a small terminal phalanx is unsupported on the metacarpal, can be lengthened and stabilised by a phalangeal transfer between the two bones of the digit. Alternatively a bone graft, which is less readily absorbed in this situation may be used. Other lengthening procedures are available but technically difficult. Rudimentary parts of digits including bone and soft tissue may be transposed on vascular pedicles to lengthen short adjacent fingers, and distraction lengthening of metacarpals has been reported (Smith and Gumley, 1985) but not gained widespread popularity. Complex reconstruction using a free bone graft covered by a skin flap to create an insensate opposition post for a



(a) (b) (c)

Fig. 6. An atypical cleft hand treated by phalangeal transfers from the toes.
(a) Pre-operative view
(b) Post-operative view
(c) Post-operative view of feet — proximal phalanges removed from left 2nd and 3rd toes. Middle phalanx removed from right 2nd toe.

solitary mobile digit is another option. This multi-staged procedure which is technically demanding does carry risks and has limited indications. A better alternative made possible by advances in microsurgery is the reconstruction of a digit using single stage toe transfer. The technique was first used following traumatic loss of the thumb. The great toe or the second toe is used but the latter has advantages in that it leaves a better donor site in the foot (O'Brien, *et al.*, 1978). The use of this method in reconstruction for congenital deformities is still in its infancy, but with improved microvascular technique survival is no longer the main problem (Gilbert, 1982; Lister *et al.*, 1983: Lister and Scheker, 1985). It seems likely that in carefully selected cases this technique will offer considerable benefit in the future. The operation is indicated for congenital amputations, especially of the thumb, when these occur in the "ring constriction syndrome". In these cases, nerves, vessels, tendons and bones have suffered an intrauterine amputation at the same level and are predictably available for reconstruction. The technique also has a place in the treatment of severe forms of transverse failure of development so that a useful prehensile pattern can be restored to the hand.

Conclusion

Typical and atypical cleft hand deformities occur with varying degrees of severity. Children learn to adapt from an early age and frequently develop remarkable function. If surgical treatment is used it should be carefully planned for each patient and performed early. Many techniques are described for improving hand function and, when the deformity is severe, these involve complex bone and soft tissue reconstruction.

REFERENCES

BARSKY A. J. (1964). Cleft hand: classification, incidence and treatment. *J. Bone Joint Surg.*, **46A**, 1707–1720.

BIRCH-JENSEN, A. (1949). Split-hand and atypical split-hand in congenital deformities of the upper extremities. Odense, Denmark, Andelsbog-trykkerriet. p 46–67.

BUCK-GRAMCKO, D. (1981). Allgemeines Wahloperationem. Handchirurgie. Band I. — Stuttgart: Thieme.

BUCK-GRAMCKO, D., PEREIRA, J. A. R. (1990). Proximal toe phalanx transplantation for bony stabilisation and lengthening of aplastic digits. *Ann. Chir. Main. Memb. Super.*, **9 (2)**, 107–118.

CARROLL, R. E., GREEN, D. P. (1975). Reconstruction of the hypoplastic digits using toe phalanges. *J. Bone Joint Surg.*, **57A**, 727.

DAVID, T. J. (1972). Dominant ectrodactyly and possible germinal mosaicism. *J. Med. Genet.*, **9**, 316–320.

DAVID, T. J. (1974). The differential diagnosis of the cleft hand and cleft foot malformations. *Hand*, **6**, 58–61.

FLATT, A. E. (1977). Cleft hand and central defects. In: The Care of Congenital Hand Anomalies. — St. Louis: C. V. Mosby. p265-285.

GILBERT, A. (1982). Toe transfers for congenital hand defects. *J. Hand Surg.*, **7**, 118–124.

GOLDBERG, N. H., WATSON H. K. (1982). Composite toe (phalanx and epiphysis) transfers in the reconstruction of the aphalangic hand. *J. Hand Surg.*, **7**, 454–459.

GRAHAM, J. B., BADGLEY, C. E. (1954). Split-hand with unusual complications. *Am. J. Hum. Genet.*, **7**, 44–50.

HARTSINCK, J. J. (1770). Beschryving van Guiana, of de wilde Kust in Zuid-America. — Amsterdam: Gerrit Tielenburg. Vol. 2, p811-812.

KUMMEL, W. (1895). Die Missbildungen der Extremitaeten durch Defekt, Verwachsung und Ueberzahl. In: Biblioteca Medica. Section E, Vol. 3. — Cassel, T. G.: Fischer and Co.

LANGE, M. (1936). Grundsatzliches uber die Beurteilung der Enstehung und Bewertung atypischer Hand-und Fussmissbildungen. *Verhandl. Deutschen Orthop. Gesellschaft*, **31**, 80–87.

LISTER, G. D., KALISMAN, M., TSAI, T. M. (1983). Reconstruction of the hand with free microneurovascular toe-to-hand transfer: experience with 54 toe transfers. *Plast. Reconstr. Surg.*, **71**, 372–386.

LISTER, G. D., SCHEKER, L. (1985). The role of microsurgery in the reconstruction of congenital deformities of the hand. *Hand Clin.*, **1**, 431–442.

MAISELS, D. O. (1970). Lobster-claw deformities of the hands and feet. *Br. J. Plast. Surg.*, **23**, 269–282.

MAY, J. W., SMITH, R. J., PEIMER, C. A. (1981). Toe-to-hand free tissue transfer for thumb construction with multiple digit aplasia. *Plast. Reconstr. Surg.*, **67**, 205–213.

MIURA, T., KOMADA, T. (1979). Simple method for reconstruction of the cleft hand with an adducted thumb. *Plast. Reconstr. Surg.*, **64**, 65–67.

MULLER, W. (1937). Die angeborenen Fehlbildugen der menschlichen Hand. — Leipzig: Thieme.

NUTT, J. N., FLATT, A. E. (1981). Congenital central hand defect. *J. Hand Surg.*, **6**, 48–60.

O'BRIEN, B. M., MACLEOD, A. M., SYKES, P. J., BROWNING, F. S. C., THRELFALL, G. N. (1978). Microvascular second toe transfer for digital reconstruction. *J. Hand Surg.*, **3**, 123–133.

ROGALA, E. J., WYNNE-DAVIES, R., LITTLEJOHN, A., GORMLEY, J. (1974). Congenital limb anomalies: frequency and aetiological factors. *J. Med. Genet.*, **11**, 221–233.

SMITH, R. J., GUMLEY, G. J. (1985). Metacarpal distraction lengthening. *Hand Clin.*, **1**, 417–429.

SNOW, J. W., LITTLER, J. W. (1967). Surgical treatment of cleft hand. In: Transactions of the Society of Plastic and Reconstructive Surgery: 4th Congress in Rome, Amsterdam, Exerpta Medica Foundation. p888–893.

TADA, K., YONENOBU, K., SWANSON, A. B. (1981). Congenital central ray deficiency in the hand — a survey of 59 cases and subclassification. *J. Hand Surg*, **6**, 434–441.

UEBA,Y. (1981). Plastic surgery of the cleft hand. *J. Hand Surg.*, **6**, 557–560.

WATARI, S., TSUGE, K. (1979). A classification of cleft hands, based on clinical findings: theory of developmental mechanisms. *Plast. Reconstr. Surg.*, **64**, 381–389.