Central Deficiency (Cleft Hand)

15

Toshihiko Ogino

Definition

Central deficiency of the hand is called cleft hand, split hand, lobster-claw, or central oligodactyly. Barsky defined cleft hand as a form of congenital absence of one or more digits in which the central rays of the hand are affected [1]. According to his definition, there are two types of cleft hand, typical and atypical. Typical cleft hand is characterized by a deep V-shaped or funnel-shaped defect in the central part of the hand; atypical cleft hand is a more severe anomaly in which the three central rays are missing and is associated with various degrees of hypoplasia of the thumb and little finger. In atypical cleft hand there are often rudiments of the missing fingers along the web between the thumb and little finger (Fig. 15.1). Atypical cleft hand has the common characteristic features of other types of symbrachydactyly: all cases were unilateral; various degrees of hypoplasia existed not only in the affected finger but also in the adjacent fingers and in the proximal part of the limbs, and are associated with pectoral muscle absence [2, 3]. Atypical cleft hand is considered to be a moderate grade of symbrachydactyly, and the Congenital Committee of the International Federation of Societies for Surgery of the Hand (IFSSH) has urged not to use the term atypical cleft hand in order to prevent confusion of terminology [4]. This chapter only deals with typical cleft hand.

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Incidence and Genetics

Birch-Jensen [5] estimated the ratio of occurrence of typical cleft hand as 1 in 90,000 births. The incidence of cleft hand among all anomalies of the upper extremity is 2.3 % of 1,476 patients in Flatt's Iowa series [6] and 2.6 % of 943 patients in Ogino's Sapporo series [7].

Regular autosomal dominant inheritance was evident in about 34 % of reported pedigrees. In other pedigrees, there are some different types of inheritance, such as lack of penetrance of autosomal dominant inheritance and markedly irregular dominant inheritance [8]. Vogel classified cleft hand into two types from the genetic aspect [9]. In type 1 pedigree affected members showed constant involvement of feet and had a consistent autosomal dominant inheritance. In type 2 pedigrees affected members showed variable involvement of feet and irregular inheritance.

Split hand/foot malformation (SHFM) is a congenital absence of the central rays of the hands and feet. Some authors use ectrodactyly to denote any absence deformity of the distal limbs and reserve SHFM for the typical malformation; others use ectrodactyly synonymously with SHFM [10]. The Human Genome Organization Nomenclature Committee determined in 1994 that split hand/foot malformation should be denoted SHFM. SHFM may present with syndactyly, median clefts of the hands and feet, as well as aplasia or hypoplasia of the phalanges, metacarpals, and metatarsals. In severe cases, the hands and feet have a lobster claw-like appearance [11]. However, the severity of SHFM is highly variable. In mildly affected patients, SHFM may be limited to syndactyly and several instances of non-penetrance have been documented. Clinical variability not only exists between patients, but also between limbs of a single individual [12]. The most common mode of inheritance is autosomal dominant with variable penetrance. Autosomal-recessive and X-linked forms occur more rarely and other cases of SHFM and may be caused by chromosomal deletions and duplications. Abnormality of six SHFM loci has been found [13, 14].

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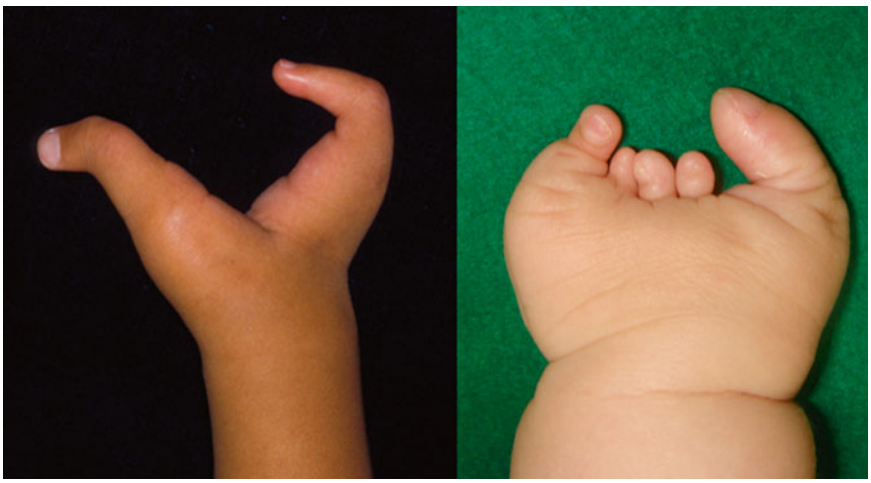

Fig. 15.1 Typical cleft hand (*left*) and atypical cleft hand (*right*)

Difference Between Cleft Hand and Other Types of Longitudinal Deficiency

The Swanson classification, which has been adopted as the IFSSH classification, has two major categories of congenital absence of digits [15, 16]: transverse deficiency (symbrachydactyly) and longitudinal deficiency. Atypical cleft hand is best classified as transverse deficiency or symbrachydactyly. In longitudinal deficiency, the congenital absence of digits is confined to the long axis of the upper limb and is classed as ulnar deficiency, radial deficiency, or central deficiency (cleft hand). In ulnar deficiency, there are various degrees of defects of ulnar fingers, such as the hypoplastic little finger, absence of the little finger, absence of the little and ring fingers, absence of the ulnar three digital rays, and absence of the ulnar four digital rays [17]. In radial deficiency, the mildest form of the hand deformity is hypoplasia of the thenar muscles and the most severe form is total absence of the thumb [18]. Non-opposable triphalangeal thumb, which is called five-fingered hand, is also one of the types of hypoplastic thumb. In some cases, the thumb and index finger are absent. In both radial and ulna deficiencies, there may be hypoplasia or aplasia of the forearm bones, and syndactyly and central polydactyly are not often seen.

Cleft hand is central deficiency, and the severe form is the absence of central three fingers, but in some cases thumb or little finger is also absent. The forearm bones are never involved, although defect or fusion of the carpal bones in distal carpal row may be seen in severe cleft hand. There are many cases in which central polydactyly, syndactyly, and cleft hand are associated in various combinations in an affected hand or in both hands of a patient [19, 20]. These anomalies also may occur in the members of the same family in various combinations. Manske [21] reported three cleft hands and one central polydactyly in four hands of identical twins. Satake et al. [22] reported a family of a mother with bilateral cleft hands, an elder daughter with the right cleft hand and the left central polydactyly, and young daughter with the left osseous syndactyly of the middle and ring fingers associated with cross bone between the middle and index fingers. There are some cases in which the middle finger is apparently missing but on X-ray, the middle and ring fingers are fused [23]. On the other hand, Müller (1936) [24] reported cases, which seemed to be cleft hands apparently, but skeletal changes were more consistent with a polydactyly of the middle finger. This issue has not been discussed in the literature for many years. Some authors reported that there were some cases in which the middle finger appears to be missing, but the metacarpus of the middle finger is duplicated [19, 25]. It is difficult or impossible to classify these cases into central polydactyly, syndactyly, or cleft hand. By these observations, cleft hand is seen to be an anomaly closely related to central polydactyly and syndactyly [19, 20, 26–29] (Fig. 15.2). When one looks at the radiographs of patients with osseous syndactyly between the middle and ring fingers, or polydactyly of the middle finger, if the defect occurs sufficiently proximal, then an appearance of cleft hand is seen (Figs. 15.3 and 15.4a, b) [28]. These observations support the concept that a common etiological mechanism is involved in the development of central polydactyly, cleft hand, and syndactyly. They also support that a common teratogenic mechanism might be the abnormal induction of finger rays in the process of formation of the fingers in the hand plate [20, 29]. From this point of view, cleft hand is one phenotype of abnormal induction of digital rays of the hand plate in which the central fingers are missing [29].

Teratogenic mechanisms of formation of cleft hand and other types of longitudinal deficiency:

In order to have a better understanding of the classification, it is necessary to clarify the development of longitudinal deficiency and cleft hand. The authors developed animal

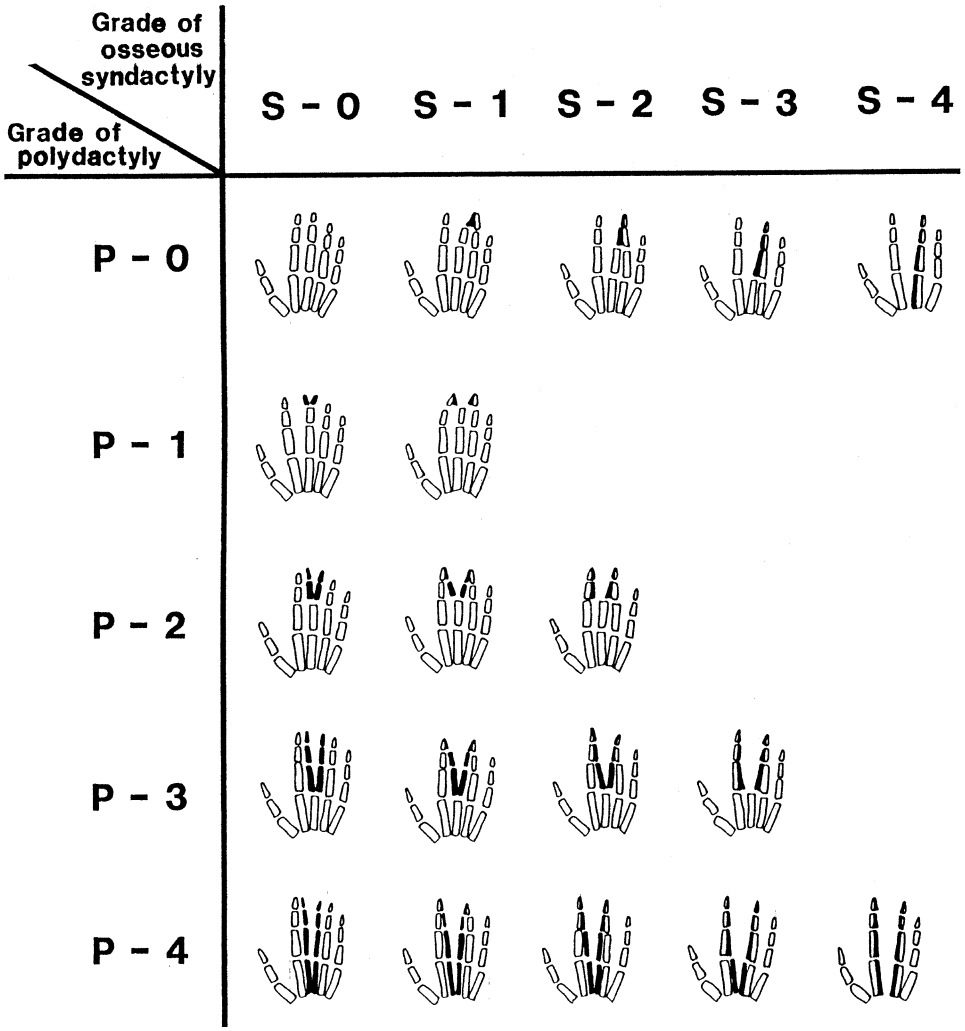

Fig. 15.2 Cleft hand formation processes from central polydactyly and/or osseous syndactyly. Reprinted with permission from Ogino T. A clinical and experimental study on teratogenic mechanism of the cleft hand, polydactyly and syndactyly. J Jpn Orthop Assoc.1979;53: 535–43

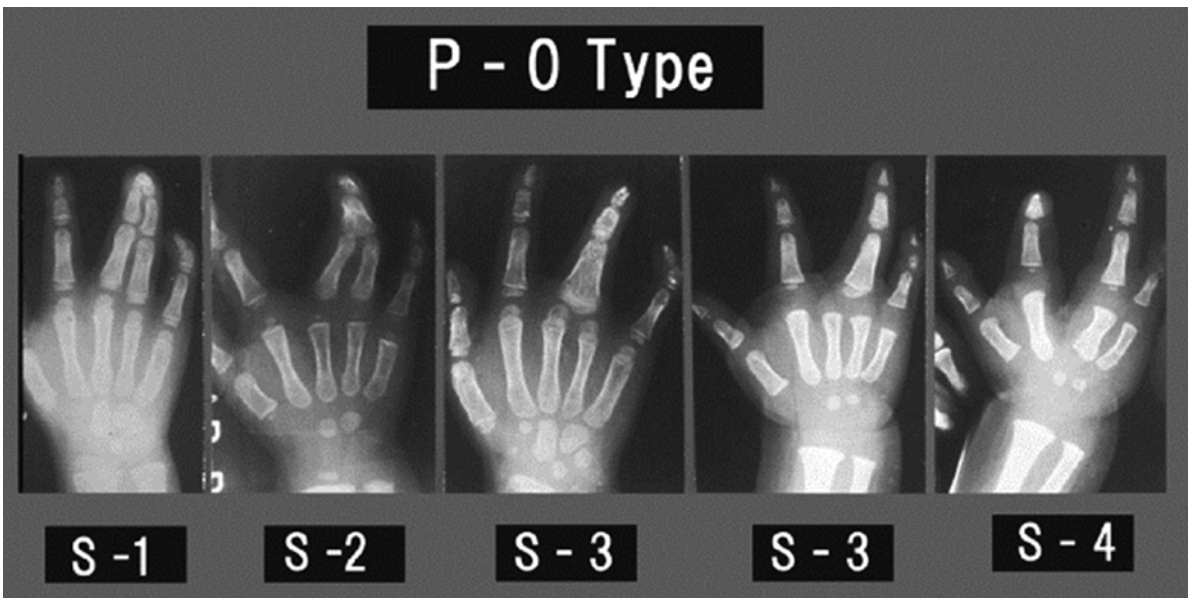

Fig. 15.3 The skeletal changes of P-0 type of anomalies in clinical cases. They seem to show that cleft hand formation proceeds from osseous syndactyly. Reprinted with permission from [28]

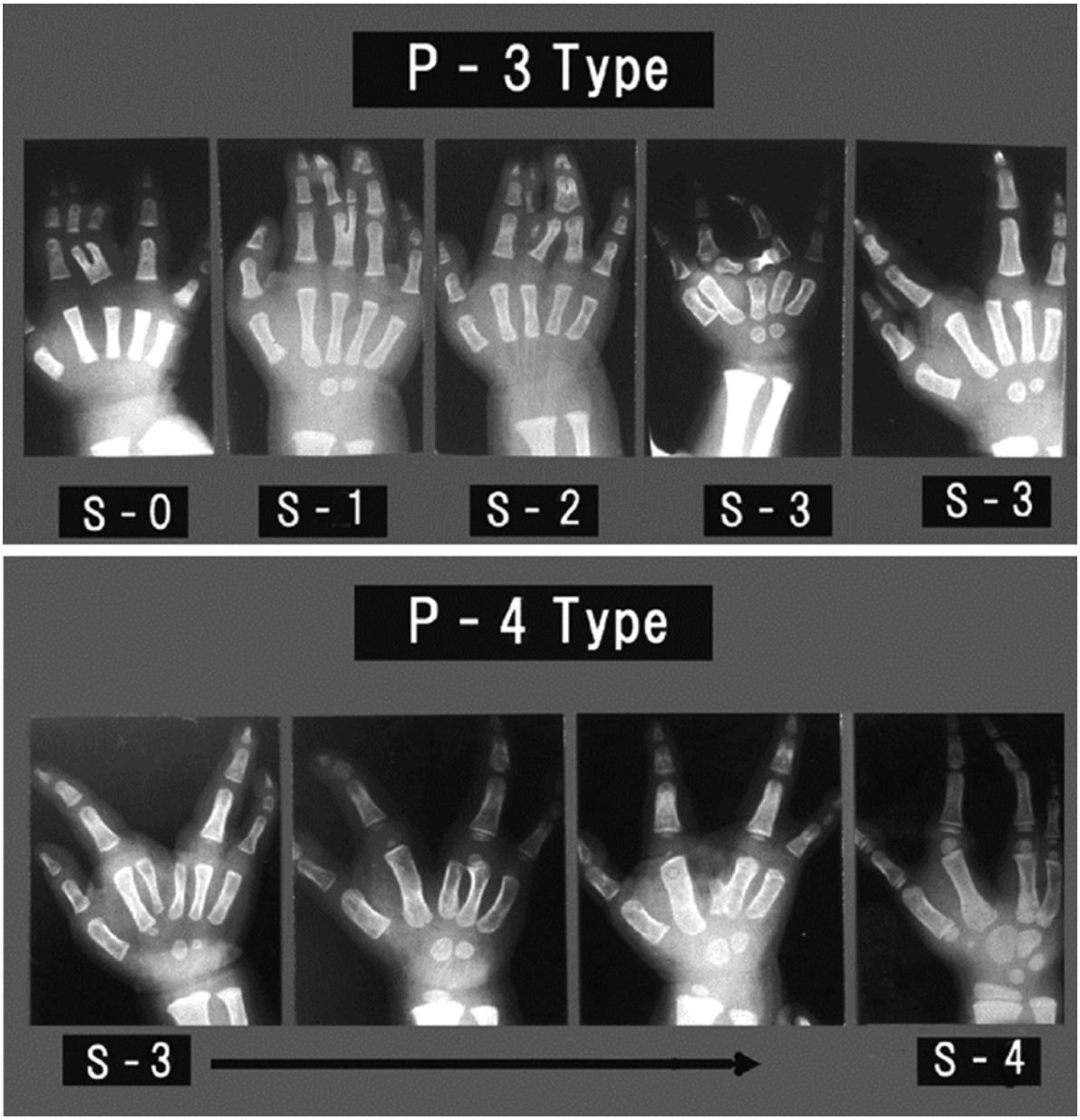

Fig. 15.4 (**a**, **b**) The skeletal changes of P-3 and P-4 types of polysyndactyly in clinical cases. They seem to show the cleft hand formation proceeds from central polydactylies. Reprinted with permission from [28]

models of these deformities using cleft foot as a model of cleft hand [17, 30–33]. The antineoplastic drug busulfan is given to pregnant rats, and radial deficiency and ulnar deficiency have been induced [19, 30]. The skeletal changes in busulfan-induced ulnar and radial deficiencies were similar to those of clinical cases. The critical period of ulnar deficiency in rats is about 1 day earlier than that of radial deficiency, and the critical period of radial deficiency in rats is just before limb buds appear [17, 30]. It was found that the dead mesenchymal cells were distributed evenly, and there was no localized cell deficiency inside the limb bud [33]. It was clear that the absence of digits in longitudinal deficiency was not caused by the localized deficiency of the limb bud.

A single cause affecting the limb bud in a certain receptive period of the development of the limb bud can induce central polydactyly, cleft hand, and syndactyly. When busulfan was given to rat fetuses at a critical period of these anomalies, later than that of longitudinal deficiency, cleft hand, central polydactyly, and osseous syndactyly were induced. The deformities were seen in varying stages of severity of osseous fusion. It was postulated that cleft hand was induced by the same etiology as osseous syndactyly and central polydactyly [29].

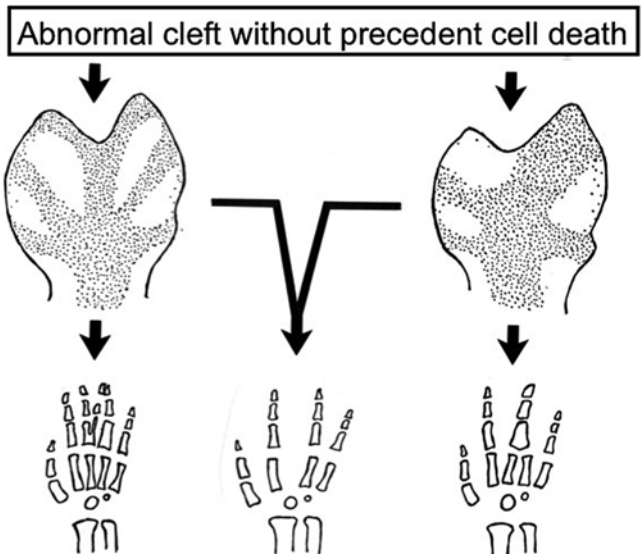

Fig. 15.5 Abnormal induction of digital rays. The early morphological changes leading to central polydactyly, syndactyly, and cleft hand were growth reduction and abnormal clefts in the central parts of the hand plates. The abnormal cleft was induced without precedent cell death and the cleft became deeper without cell death. If the abnormal cleft is induced on the edge of digital radiation, it might induce polydactyly or cleft hand. If the abnormal cleft is induced on the interdigital tissue, it might induce syndactyly or cleft hand. Reprinted with permission from Ogino T. Teratogenic mechanisms of congenital absence of digits. Locomotor System: Advances in Research, Diagnosis and Therapy 2011;18:173-93

In order to examine the underlying mechanism of busulfan-induced cleft hand, central polydactyly, and syndactyly, the authors evaluated localized apoptosis by Nile Blue staining and TdT-mediated dUTP nick end labeling (TUNEL) assays in treated rat embryos [34]. The authors further evaluated the potential disruption of major developmental pathways linked to digit number and syndactyly using Fgf8, Bmp4, and Shh as markers of these pathways. In busulfan-treated embryos, there was no difference of expression of Fgf8, BMp4, and Shh in the limb bud and footplate. The early morphological changes leading to central polydactyly, syndactyly, and cleft hand or foot were growth reduction and abnormal clefts in the central parts of the footplates. The abnormal cleft was induced without precedent cell death and the cleft became deeper also without cell death [34]. If the abnormal cleft were induced on the edge of digital radiation, it might induce polydactyly or cleft hand or foot. If the abnormal cleft were induced on the interdigital tissue, it might induce syndactyly or cleft hand or foot. The authors conclude that the abnormal cleft formation without precedent cell death was an early change leading to central polydactyly, syndactyly, and cleft hand or foot by a teratogen (Fig. 15.5 [35]). Abnormal cleft formation without precedent cell death might be caused by localized inactivation of the apical ectodermal ridge (AER) in the central part of the footplate [36].

Results of recent studies on split-hand/split-foot malformation (SHFM) using murine Dactylaplasia mutant (Dac) have shown that the central segment of the AER degenerates, leaving the anterior and posterior segments intact [37]. From this observation, it was suggested that localized failure of ridge maintenance activity was the fundamental developmental defect in Dac and it might also be suggested in SHFM [10]. Therefore, the teratogenic mechanism of formation of cleft hand/foot is the same both in drug-induced cleft hand in rats and in mutant mice with cleft hand.

Position of Cleft Hand in Japanese Modification of Swanson's Classification

Based on the clinical and experimental studies, the author modified the IFSSH classification in 1986 and added a fourth new category—abnormal induction of digital rays [38, 39]. In IFSSH classification, brachysyndactyly is classified into undergrowth and transverse deficiency into failure of formation of parts, and there is no item of cleft of the palm. However, analysis of clinical cases showed that brachysyndactyly, atypical cleft hand, or transverse deficiency seemed to be morphological variants of symbrachydactyly [2, 3]. Therefore, these deformities are included in a similar concept to transverse deficiency in failure of formation of parts in modified classification.

On the other hand, central polydactyly is classified as duplication, syndactyly as failure of differentiation of parts and typical cleft hand as failure of formation of parts in the IFSSH classification. However, these congenital deformities appear when the same teratogenic factor acts on embryo at the same developmental period. Because they have a similar causation, central deficiency, osseous syndactyly, central polydactyly, and cleft of the palm may be grouped together, and are included in the same category of abnormal induction of digital rays in modified classification [38, 39] (Table 15.1). Recent literature has reported that chromosome abnormality and also abnormalities of the positional gene may cause these anomalies [40–42].

The Japanese Society for Surgery of the Hand adopted our modification of the IFSSH classification in 1996 and it is now called the Japanese modification [43]. As a skin manifestation, there are syndactyly and cleft of the palm. As a skeletal manifestation, there are osseous syndactyly, central polydactyly, and absence of central finger rays (cleft hand), and triphalangeal thumb associated with cleft hand.

The author reviewed his own cases of abnormal induction of digital rays affecting 186 hands in 125 patients. Eighty-three cases were male and 42 female. The right side was affected in 47 cases, the left in 17 cases, and both in 61 cases. The deformities of the affected hand were expressed by the combination of cleft on the palm, cutaneous syndactyly,

Table 15.1 The Japanese Society for Surgery of the Hand Modification of the IFSSH classification revised by Ogino T. (2013)

- I. Failure of formation of parts (arrest of development)
  - A. Transverse deficiency
    - 1. Peripheral hypoplasia type
    - 2. Short webbed finger type
    - 3. Tetradactyly type
    - 4. Tridactyly type
    - 5. Didactyly type
    - 6. Monodactyly type
    - 7. Adactyly type
    - 8. Metacarpal type
    - 9. Carpal type
    - 10. Wrist type
    - 11. Forearm type
    - 12. Above elbow type
  - B. Longitudinal deficiencies
    - 1. Radial deficiencies:
      - (a) Dysplasia of the radius
      - (b) Deformities of the hand
      - (c) Dysplasia of the elbow
    - 2. Ulnar deficiencies:
      - (a) Dysplasia of the ulna
      - (b) Deformities of the hand
      - (c) Dysplasia of the elbow
  - C. Phocomelia
  - D. Tendon or muscle dysplasia
  - E. Nail dysplasia
    - 1. Aplapsia/hypoplasia of the nail
    - 2. Nail defect with brachytelephalangia
- II. Failure of differentiation of parts
  - A. Synostosis
  - B. Radial head dislocation
  - C. Symphalangism
  - D. Contracture
    - 1. Soft tissue
      - (a) Arthrogryposis multiplex
      - (b) Webbed elbow (pterygium cubitale)
      - (c) Clasped thumb
      - (d) Windblown hand
      - (e) Camptodactyly
      - (f) Aberrant muscles
      - (g) Nail deformities:
        - (i) Nail deformity with clinodactyly
        - (ii) Nail deformity with hypoplasia of the distal phalanx
        - (iii) Nail deformity or palmar nail with hypoplastic digit and symphalangism
    - 2. Bone
      - (a) Kirner deformity
      - (b) Delta bone (longitudinal epiphyseal brancket)
      - (c) Madelung deformity
    - 3. Others
  - E. Tumorous conditions
- III. Duplication
  - A. Thumb polydactyly
  - B. Central polydactyly (it should be category IV)
  - C. Polydactyly of little finger
  - D. Opposable triphalangeal thumb
  - E. Other hyperphalangism
  - F. Mirror hand
  - G. Dorsal and palmar duplication
    - 1. Double dorsal limited in the digit
    - 2. Double dorsal including the hand
    - 3. Double palmar limited in the digit
    - 4. Double palmar including the hand
- IV. Abnormal induction of digital rays
  - A. Soft tissue:
    - (1) Cutaneous syndactyly, (2) Cleft of the palm
  - B. Bone:
    - (1) Osseous syndactyly, (2) Central polydactyly, (3) Absence of central finger ray(s), (4) Triphalangeal thumb with cleft hand
  - C. Ulnar cleft hand:
    - (1) Cleft of the fourth web space: (a) with absent ring finger, (b) without absent ring finger
    - (2) Stiff finger: (a) symphalangism; PIP and/or DIP joint, (b) partial ankylosis; PIP and/or DIP joint
    - (3) Nail deformity: (a) clam nail, (b) claw nail, (c) circumferential nail
  - D. Abnormal induction of digital rays with hypoplastic hand:
    - (1) polydactyly, (2) syndactyly, (3) central finger absence
- V. Overgrowth
  - A. Macrodactyly
  - B. Hemihypertrophy
- VI. Undergrowth
  - A. Microcheiria (hypoplastic hand)
  - B. Brachydactyly
  - C. Clinodactyly
- VII. Constriction band syndrome
  - (1) Constriction ring, (2) Lymphedema, (3) Acrosyndactyly, (4) Amputation type
- VIII. Generalized skeletal abnormalities and a part of syndrome
- IX. Others (including unclassifiable cases)

osseous syndactyly, absence of central digit(s), which is absence of all phalanges of the digital ray(s), and central polydactyly. Of the 186 abnormal hands, a single deformity appeared in 86 hands: cutaneous syndactyly alone in 65 hands, osseous syndactyly alone in 17 hands, and cleft on the palm without absence of digit in 4 hands. In 100 hands, multiple deformities appeared in the same hand of a patient. Polydactyly and syndactyly were present in the same hand in 16 cases; a combination of cleft on the palm and syndactyly in six cases; a cleft, polydactyly, and syndactyly in one case; an absence of central digit and cleft on the palm in 37 cases; an absence of central digit, cleft on the palm, and syndactyly

in 34 cases; an absence of central digit, cleft on the palm, and polydactyly in one case; and an absence of central digit, a cleft on the palm, central polydactyly, and syndactyly in five cases. In these cases there were eight hands with triphalangeal thumb associated with absence of the index finger, and there was one hand with a floating little finger. In bilaterally affected cases, same type of expression was evident in both the right and left hands in 47 cases. Different abnormalities occurred in the right and left hands in 14 cases. Hand deformities were expressed by combinations of cutaneous syndactyly, cleft on the palm, osseous syndactyly, central polydactyly, absence of central digit, and triphalangeal thumb with cleft hand with absence of the index finger. This review suggested that abnormal induction of digital rays may explain simultaneous occurrence of differing abnormalities within the same hand. The concept of abnormal induction of digital rays seemed useful for classification of congenital hand differences.

In this chapter, the author has revised the Japanese modification of the IFSSH (see Table 15.1). The abnormality of the nail has not been clearly classified, and the abnormalities of the dorso-ventral plane of the hand also have not been described in the previous Japanese modification. First, "E. Nail dysplasia" in I. Failure of formation of parts was subdivided into:

1. Aplapsia/hypoplasia of the nail.
2. Nail defect with brachytelephalangia.
3. Others.

Secondly, II. Failure of differentiation of parts had (g) Nail deformities added as a sub-category, and it is subclassified into:

1. Nail deformity with clinodactyly
2. Nail deformity with hypoplasia of the distal phalanx
3. Nail deformity or palmar nail with hypoplastic digit and symphalangism (This is the same deformity observed in "ulnar cleft hand: VI. C")
4. Others

Thirdly, III. Duplication had "G. dorsal and palmar duplication" added and is sub-classified into:

1. Double dorsal limited in the digit
2. Double dorsal including the hand
3. Double palmar limited in the digit
4. Double palmar including the hand
5. Others

Fourthly, the categories IV. Abnormal induction of digital rays added the categories:

C. Abnormal induction of digital rays, ulnar cleft hand
D. Abnormal induction of digital rays with hypoplastic hand

After these changes, it becomes easier to differentiate true typical cleft hand described by Barsky and other cleft hand-like deformities.

Clinical Characteristics

Blauth and Falliner's reported incidence of cleft hand: they found bilateral involvement in 50 %, and in unilateral involvement, the ratio of right hand to left hand is 60 to 40 %. The ratio of male to female is 60 to 40 % [44]. In approximately one out of three cases of cleft hand there was associated cleft foot.

Defect of the central finger rays varies [45]: there are hands with deep cleft formation on the palm without absence of the finger rays. This is a type of central deficiency, and therefore a form of cleft hand (Type 0) [46]. There are cleft hands with one finger absent, two fingers absent, three fingers absent, or four digits absent. In one finger absence type (Type 1), the middle finger is most commonly absent. In that case, the ring finger often will have camptodactyly. This deformity is not actually true camptodactyly, but a claw finger deformity due to abnormal lumbrical or interosseus muscles. Most often, the affected ring finger has no joint contracture when the child is young. If the metacarpophalangeal joint of the ring finger is passively flexed to neutral, the patient can actively extend the PIP and DIP joints of the affected finger.

Some cases with mild excessive cleft and absence of the middle finger will have normal looking third metacarpal bone radiologically. However, in some cases the third metacarpal bone deviates ulnarly and has a common MP joint with the fourth metacarpal bone and proximal phalanx of the ring finger. Alternately, third metacarpal bone will deviate radially and have a common MP joint with the second metacarpal bone and proximal phalanx of the index finger. In the former case, the ring finger is wider than normal and in the later case the index finger is wider than normal. When middle finger ray including the third metacarpal is absent, the cleft is deeper than usual. The deeper the cleft is, the more often syndactyly of the first web space and the fourth web space occur. In some rare cases of deep cleft, hypoplasia of the little finger, or the fusion of the fourth and fifth metacarpals, is seen. In Type 1 cleft hand, the index finger or ring finger also may be absent [47, 48]. When the index finger is absent, the thumb is often triphalangeal and will deviate radially, in contrast to most triphalangeal thumbs, which deviate ulnarly. A "Y" shaped second metacarpal bone between the thumb and middle finger, or two thumb metacarpal bones may be seen on X-ray. When the ring finger is absent, the little finger is small and stiff, and this may be called "ulnar cleft hand." When the finger is not absent and the cleft is in the forth web apace, this is often associated with stiffness of the IP joints, palmar nail, and the dorsal skin on the palmar side of the little finger. This may be called "double dorsal deformity of the finger." The etiology of this deformity is considered to be different from other types of cleft hand (see below).

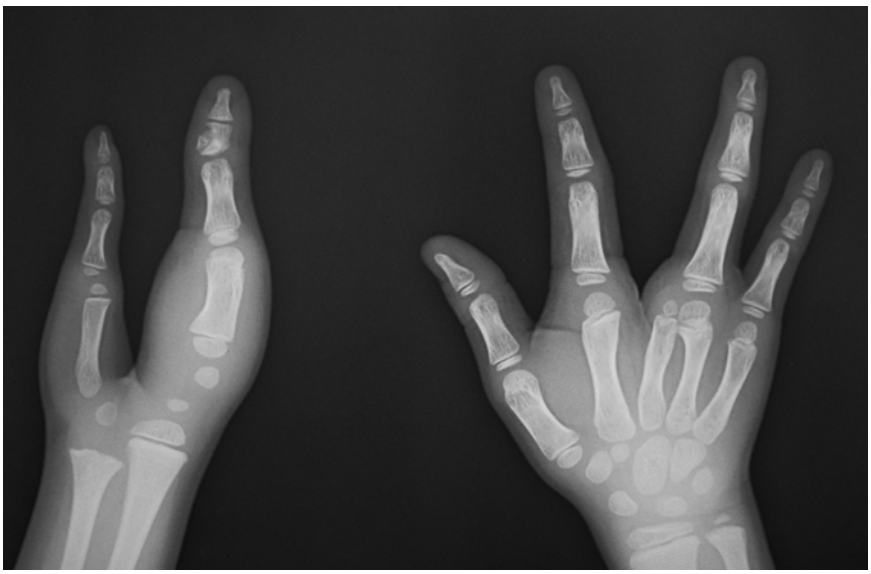

Fig. 15.6 Different degree of absence of the fingers associated in both hand of a patient. *Left*: type 3 cleft hand with central three-finger absence and triphalangeal thumb with radial deviation of the IP joint. *Right*: type 1 cleft hand with absence of the middle finger

In two-finger absence type (Type 2), the index and middle fingers are absent more commonly than the middle and ring fingers. When the index and middle fingers are absent, the thumb is usually triphalangeal. In three-finger absence type (Type 3), if the thumb is opposable then pinch is possible between the thumb and the little finger. If the thumb is in the plane of the hand however, pinch between the thumb and little finger is impossible [49]. In four-digit absence type (Type 4), the radial four digits are absent in most commonly, but very rarely the ulnar four digits will be absent and only the thumb remains [46]. In radial-four-digits absence type, the metacarpals of the thumb and affected fingers are usually only partially absent or not involved.

A cross bone is a transverse or oblique bone lying in the base of the cleft. It is regarded as the displaced remnant of the metacarpal or proximal phalanx of the missing digit and it bridges between the end of the metacarpal bones of the missing digit and the proximal phalanx, MP joint, or the metacarpal bone of the adjacent finger. There may be two cross bones which might be duplicated proximal phalanges of the missing digit and are located between missing digit and adjacent fingers between the end of the metacarpal bones of the missing digit and the proximal phalanx, MP joint, or the metacarpal bone of the adjacent finger. There may be two cross bones, which might be duplicated proximal phalanges of the missing digit, and are located between missing digit and adjacent fingers. There may be solid bone union or cartilaginous continuity between the cross bone and the proximal phalanx of the adjacent finger in the skeletally immature patient. In some cases, the proximal phalanx of the adjacent finger will have a deltaphalanx (longitudinal epiphyseal bracket). X-ray films may show, two metacarpals supporting one digit, side-to-side fusion of the neighboring metacarpals, broad metacarpals, or duplicated metacarpals.

Surgical Classification of the Cleft Hand

Saito et al. [45] classified typical cleft hand into four types on the basis of the number of defective finger rays:

Type 1: deep cleft formation on the palm without missing finger.
Type 2: defect of a single finger ray.
Type 3: defect of two finger rays.
Type 4: defect of three finger rays.

Watari and Tsuge [27] classified typical cleft hand according to the same idea. In their classification there is no type without absence of the finger, but a type in which four finger rays are absent. They divided single ray defect type of cleft hand into proximal and distal types. In the proximal type all phalanges and the metacarpus are missing and in the distal type only phalanges are missing. The author modified these classifications as follows (Fig. 15.6):

Type 0: cleft hand without missing finger
Type 1: defect of a single finger ray
Type 2: defect of two finger rays
Type 3: defect of three finger rays
Type 4: defect of four digital rays

In every type, when the index finger is absent, the thumb is mostly triphalangeal and deviated radially.

Manske et al. [50] proposed surgical classification for cleft hand based on the characteristics of the thumb web space, because he thought the thumb web was more important to the function of the hand than the central deficiency. According to his report, cleft hand is classified into five types:

Type 1: normal first web
Type 2A: mildly narrowed first web
Type 2B: severely narrowed first web

Type 3: syndactylized first web
Type 4: merged web in which index ray suppressed, thumb web space is merged with the cleft
Type 5: absent web, in which thumb elements suppressed, ulnar rays remain and thumb web space no longer present

One benefit of sub-classification of the congenital hand deformities is that one can better picture the deformity of the hand, from description with the classification. For example, when told or read: "Type 4 thumb polydactyly in Wassel's classification [51]" or "Type 3 hypoplastic thumb in Blauth's classification [18]," one can clearly image the deformities of the hand and the possible associated deformities. Both hypoplastic thumb and thumb polydactyly may have the narrowing of the first web. This is an important factor not only when treating cleft hand, but also hypoplastic thumb and thumb polydactyly. Manske reported that the progressive narrowing of the thumb web correlated with progressive severity of the central defect. The author also observed the same findings and published it in 1977 [19]. Therefore, one can imagine the possible condition of the first web associated with cleft hand, using Saitou's classification based on the number of defective finger rays. Surgical treatment is not only directed to the first web, but also the deep or wide cleft. I feel that sub-classification should be valuable not only for the patient but also for communication of the people who are treating these deformities. Based on this viewpoint, Saitou's classification based on the number of defective finger rays seems more valuable for sub-classification of the cleft hand. If one uses Manske's classification [50], it would be more useful if combined with Satou's classification [45]. Falliner [52] classified cleft hand into three types as follows:

Radial cleft hand
- Hand deformities including osseous syndactyly of the thumb and index finger
- Absence of the index finger
- Absence of the thumb and index finger
- Absence of the radial three or four digits

Central cleft hand
- Central defect with absence of the middle finger
- Central defect with absence of the central two fingers
- Central defect with absence of the central three fingers
- Cleft hand with only the thumb and little finger present

Ulnar cleft hand
- Absence of the ring finger
- Absence of the ring and middle fingers, with or without of hypoplasia of the little finger

This classification seems to be too simple for clinical use. Moreover, ulnar cleft hand has characteristic clinical features such as deep cleft in the fourth web space, absence of the ring finger and/or hypoplastic little finger associated with stiffness of the PIP joint and palmar nail, and it is considered to differentiate it from other types of cleft hand. When one uses Japanese modification of the IFSSH classification [43], in abnormal induction of finger rays, the hand deformities are expressed with combination of the cleft, syndactyly, and other phenotypes. Therefore, if one describes deformities combined with the degree and the location, one can expresses the deformity of the cleft hand and other combined deformities precisely.

Cleft of the Fourth Web Space of the Hand

Cleft of the fourth web space of the hand is associated with or without absence of the ring finger. These deformities are called ulnar cleft hand [48]. However, its clinical features are different from various types of abnormal induction of digital rays including cleft hand. Moreover, characteristic clinical features of cleft of the fourth web space with absence of the ring finger are different from those without absence of the ring finger, although the little finger is hypoplastic in both conditions [47, 53]. There is no appropriate terminology and precise classification for the sequence of these congenital hand deformities. Ulnar cleft hand without absence of the ring finger is characterized with combination of various degrees of cleft of the ring and little finger, hypoplasia of the little finger, hypoplasia of hypothenar muscles, extension contracture or symphalangism of the little finger and clam nail, claw nail or circumferential nail deformity, and dorsal skin of the palmar little finger [53] (Fig. 15.7). In the opposite hand, the same deformity, polydactyly of the little finger, ulnar deficiency, or partial duplicated distal phalanx of the ring finger may be seen. In this anomaly, there are various associated deformities of the hand. Hand and nail deformities in this anomaly are similar to those of ulnar-mammary syndrome or Schinzel syndrome [54]. The teratologic sequence of the variety of hand deformities with ulnar cleft of the fourth web without absence of digits is most likely a different entity from abnormal induction of finger rays.

Abnormal Induction of Digital Rays (Including Cleft Hand) Associated with Hypoplastic Hand

Abnormal induction of digital rays in the hand plate means induction of abnormal number of the digital rays in the hand plate. Therefore, excessive or decreased number of induction of digital ray occurs, but nearly all of the hands with deformities of abnormal induction of digital rays do not seem to have hypoplasia of the hand. However, there are hand deformities, with combinations of syndactyly, cleft of the palm, central polydactyly, osseous syndactyly, or absence of the central fingers associated with hypoplasia of the affected hand. This deformity is most often unilateral.

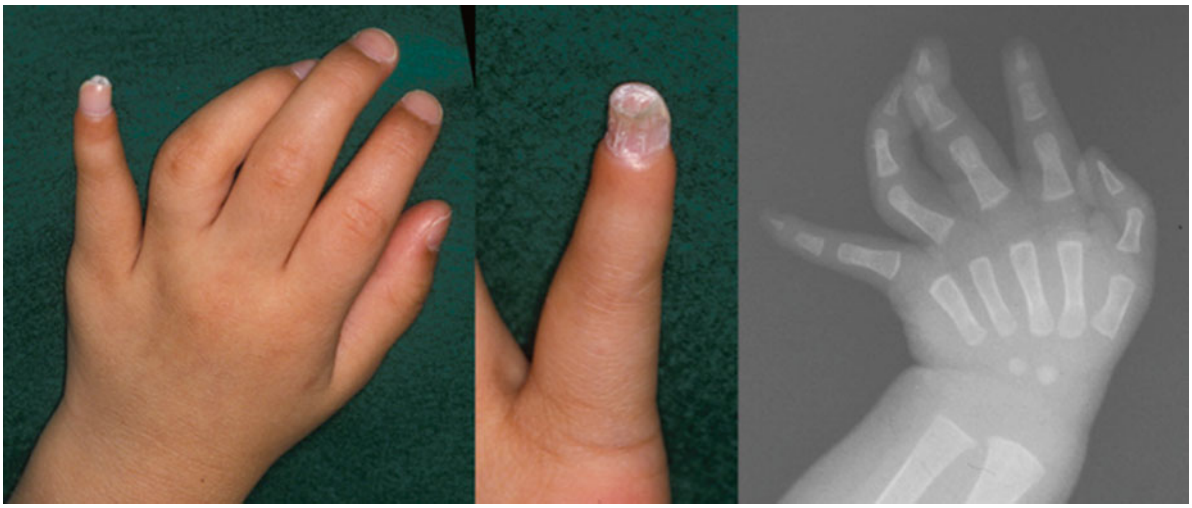

Fig. 15.7 Cleft of the fourth web space without absence of the finger. *Left*: cleft of fourth web space associated with stiff PIP joint (symphalangism) and hypoplastic little finger. *Center*: circumferential nail and dorsal skin on the palmar side of the little finger. *Right*: synchondrosis of the PIP joint of the little finger

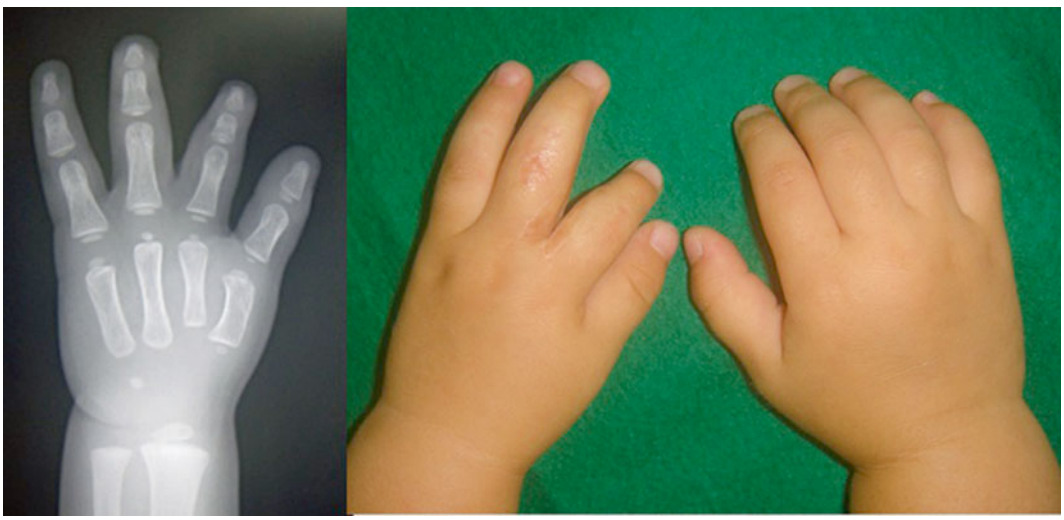

Fig. 15.8 Abnormal induction of the finger rays associated with hypoplasia of the affected hand. The characteristic features of this condition seem to be those of transverse deficiency, which are unilateral involvement and hypoplasia of the whole affected hand compared to the opposite hand, and those of abnormal induction of digital rays, which are that the hand has cleft of the palm, syndactyly, central polydactyly, osseous syndactyly, and/or absence of the central fingers. It is difficult to say which finger rays are missing in X-ray film

This condition seems to have both the characteristic features of transverse deficiency (hypoplasia of the affected hand and unilateral involvement) and those of abnormal induction of digital rays (central polydactyly, syndactyly, and cleft hand). This condition is not associated with polydactyly, syndactyly, and/or central deficiency of the opposite hand and the feet. In the affected hand, thenar and hypothenar muscles are relatively well formed and it is sometimes difficult to say which finger rays are missing in the central finger rays, although the thumb and the little finger are never absent [55, 56] (Fig. 15.8).

Associated Anomalies

As regional association, syndactyly of the thumb and index finger or that of the ring and little fingers, and brachydactyly of the little finger are most common. Triphalangeal thumb often occurs in cleft hand associated when the index finger is absent. Polydactyly of the thumb and side-to-side synostosis of the fourth and fifth metacarpals are rarely seen. Occasionally, some patients will be affected bilaterally in which there is a cleft hand on one side and on the other, another type of anomaly, such as cutaneous syndactyly, osseous syndactyly, or cen-

tral polydactyly. The central deficiency in SHFM patients may also be accompanied by other distal limb anomalies including central polydactyly and/or syndactyly [57].

Foot deformities, such as cleft foot, syndactyly, central polydactyly, and tibial ray deficiency, are also associated with cleft hand.

Cleft hand appeared as a part of syndrome, such as, ectrodactyly–ectodermal dysplasia–clefting (EEC) syndrome, de Lange syndrome, split hand/split foot with mandibulo-facial dysostosis, split hand with perceptive deafness, split hand with congenital nystagmus, fundal changes and cataract, anonychia with ectrodactyly, and the acrorenal syndrome.

As mentioned above, split hand/foot malformation (SHFM), or central ray deficiency, can occur as an isolated malformation or as a part of syndrome, such as in the EEC syndrome. Rüdiger et al. in 1970 [58] named an anomaly complicated three malformations the EEC syndrome, based on their initials, namely, ectrodactyly, ectodermal dysplasia and clefting syndrome. The main clinical signs, in order of frequency observed in Rodini and Richieri-Costa [59] reported group, were ectodermal dysplasia (100 %), ectrodactyly (78 %), tear duct anomaly (71 %), cleft lip/plate (58 %), genito-urinary anomalies (15 %), deafness (9 %), and mental retardation (2 %). The clinical expression of the EEC syndrome is quite variable; any one of the above signs may be absent except ectodermal signs. Ectrodactyly may occur only in hands (25 %) or in both hands and feet (65 %). Ten percent of the patients had no limb involvement. Cleft hands and feet are characteristic anomalies of this syndrome, but syndactyly, polydactyly of the central digital ray may be associated with this syndrome [57, 60, 61]. Majewski and Küster [62] stated that ectrodactyly is not an obligatory symptom. Skin anomalies related to ectodermal dysplasia are fine, thine smooth skin, hyperkeratosis, and dermatoglyphic alterations. Trichodysplasia, dental defects, onychodysplasia, and dyshidrosis may be associated. In EEC syndrome, most cases have *p63* gene mutations. In contrast, *p63* mutations were detected in only a small proportion of patients with isolated SHFM [63].

Treatment of Cleft Hand

Indication and Timing of Surgery

Cleft hand has several associated deformities. The goals of surgical treatment for cleft hand may need to address:

- Reduction of the excessive deep or wide interdigital space
- Separation of syndactyly of the first web or the interdigital space between the ring and little fingers
- Correction of claw finger deformity of the ring finger
- Correction of the deviation of the thumb due to triphalangeal thumb
- Correction of the deviation of the index finger due to trapezoidal shape of middle phalanx

Many authors stated that surgery of cleft hand is mainly performed for esthetic reasons. Reduction of the interdigital space is in fact performed mainly for the cosmetic reasons in cleft hand without missing finger (Type 0) and cleft hand with defect of a single finger ray (Type 1). However, correction of thumb deformities including the first web contracture and that of the deviation of the thumb due to triphalangeal thumb gives significant functional improvement. Such procedures may be performed simultaneously in order to prevent multiple surgeries. Reduction of the interdigital space, separation of syndactyly of the first web space, correction of the deviation of the thumb and that of claw finger deformity of the ring finger are usually performed at initial surgery. Claw finger deformity of the ring finger is corrected by reconstruction of the MP joint flexor with FDS tendon of the missing finger as in lasso procedure. It is easily done at initial surgery, as the flexor tendons are exposed in the palm and it is easy to select the transferred tendon before the cleft is closed. Alternatively, this procedure may be performed later as a second stage surgery, if the deformity is not corrected spontaneously after closing the cleft. The combination of the surgical procedures is different in each case according to the associated deformities. Reduction of the interdigital space should not be performed for some kind of cleft hand that is cleft hand with missing index finger, that with missing index and middle fingers, and cleft hand with defect of central three finger rays (Type 4). If reduction of the interdigital space is performed in these cases, the patient will have difficulty in grasping a large object. On the other hand, in cleft hand with trapezoidal proximal phalanx of the ring finger, the proximal phalanx is a delta phalanx and the ring finger has ulnar deviation. It may be corrected in some extent by physiolysis with free fat graft, if the surgery is performed in a young patient [64]. However, a secondary corrective osteotomy may be needed if satisfactory correction has not been achieved after physiolysis.

Closure of the excessive interdigital space (cleft) for cleft hand without missing finger (Type 0) or that with absence of a single finger (Type 1), separation of syndactyly between thumb and index finger and removal of the delta phalanx of the thumb, and correction of the claw finger deformity of the ring finger are performed at the age of 1 year. The author prefers to perform these surgeries simultaneously. Separation of syndactyly of the ring and little fingers is usually carried out at the second stage surgery, since the level of the interdigital web to be corrected can be more easily determined at that time. Separation of the side-to-side fusion of the metacarpals, and arthrodesis of the finger should be performed at a later stage as needed. Physiolysis with free fat graft should be performed around the age of 3–4 years. All necessary surgery should be completed by the time the child enters school at the age of 6 years.

Preoperative Care

Usually no preoperative care is needed. The author asks the parents to close the cleft of the hand manually by pushing the border digits at least one a day, when the simple closure of the cleft is indicated. If the cleft hand is associated with claw finger deformity of the ring finger, the author asks them also to prevent contracture of the finger with manual correction.

When the interdigital space is wide or deep and simple closure of the cleft is indicated, static splint may be applied in order to close the cleft until surgery. When we examine a patient, the first web contracture associated with cleft hand at the age of 1 or 2 years, sometimes the patient does not use the thumb for pinch but uses two fingers adjacent to the cleft for pinch. When the splint is applied to close the wide cleft soon after birth, the patient can learn normal pinch pattern. While the literature has generally not recommended splinting prior to surgery for cleft, the author believes that the splint may establish proper muscle balance in a corrected position and prevent secondary skeletal deformities in selected cases. Preoperative splinting facilitates correction of the deformity during surgery.

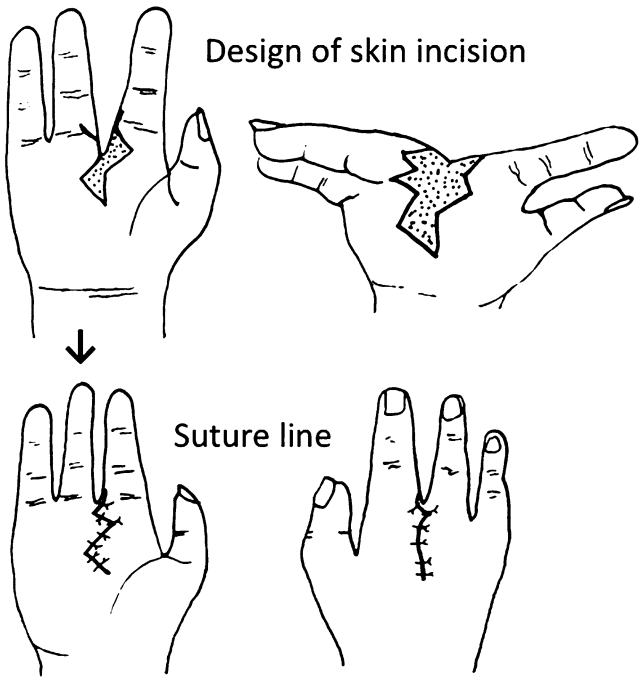

Fig. 15.9 Skin incision for reduction of the interdigital space using small triangular flap for the web. The *dotted area* of the skin will be excised

Closure of the Excessive Interdigital Space

In order to make natural slope of the interdigital web after closing the cleft, many procedures have been reported: Barsky [1] used a diamond-shaped flap based on one digit, Kelikian [65] used a rectangular flap from across the apex of the palmar cleft of the hand and Tsuge [66] used a triangular flap. The author prefers to use small triangular flap [46] (Fig. 15.9). Before skin incision, the second and fifth metacarpals are pushed toward each other and cleft is manually closed. The cleft can be closed easily, since the parents have been manually closing it for certain period before surgery. Then zigzag incisions may then be designed in expectation of an interdigitating closure. However, dorsal zigzag suture line may not give the best aesthetic result. In that case, the author uses dorsal straight incision instead of zigzag incision. An ulnar-based small triangular flap is raised by this incision. Excessive skin of a wide or deep interdigital space may then be removed. After necessary treatment of bone, tendon, and ligament, the skin incision is closed.

Treatment of Metacarpus and Cross Bone

There are different types of metacarpal bone deformities. In some cases, two metacarpals shift each other and support one digit. For example, in cleft hand with absent middle finger, the third metacarpal bone deviates ulnarwards and has common MP joint among fourth metacarpal bone and proximal phalanx of the ring finger or it deviates radial wards and has common MP joint among second metacarpal bone and proximal phalanx of the index finger. If the third metacarpal bone prevents to close the cleft manually in these types of deformities, the shortening or partial removal of the metacarpal shaft is indicated (Fig. 15.10a, b), but it was not necessary in most cases. There are also, side-to-side fusion of the neighboring metacarpals, broad metacarpus, and duplicated metacarpals. In the cleft hand with absent index finger, the Y-shaped second metacarpal bone is located between the thumb and middle finger, or two metacarpal bones exist in the thumb. These metacarpal deformities usually do not disturb hand function nor induce secondary deformities. Therefore, it is not necessary to treat them surgically.

On the other hand, many authors advocate removal of the cross bones and osteotomy of one or both of the adjacent metacarpals. Some authors thought that osteotomy is not essential. If the metacarpal remnants or cross bone prevent to draw the metacarpals together, these bone should be removed. If the second and fourth metacarpals could not be put into parallel after removing the third metacarpal, osteotomy of the second metacarpal or metacarpal transfer of the second metacarpal to the third one is recommended, but it is not essential. When these bones are removed, extensor hood and capsule of the metacarpophalangeal joint of the absent finger ray and/or adjacent finger ray must be incised. In such cases, repair of extensor hood

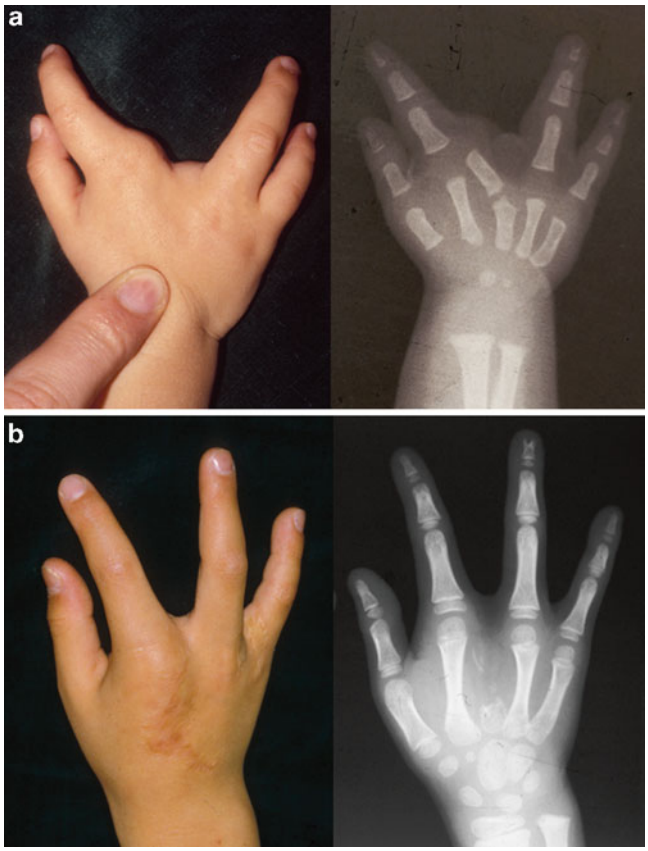

Fig. 15.10 Excision of the cross bone. (**a**) Preoperative appearance and roentgenogram. At the age of 1 year 2 months, the cross bone and the third metacarpal were resected. Osteotomy of the second metacarpal base was performed and the cleft was close. (**b**) Postoperative appearance and roentgenogram: after surgery, good alignment has been achieved

and joint capsule are necessary to prevent deformity after surgery. However, the tight soft tissue on the radial side does not allow the metacarpal to transfer ulnarward easily.

Reconstruction of the Deep Transverse Metacarpal Ligament

The deep transverse metacarpal ligament connects the anterior surfaces of the adjacent metacarpal heads. It normally blends with the volar plates of the metacarpophalangeal joints and prevents spreading of the fingers [67]. In cleft hand, the deep transverse metacarpal ligament is absent in the cleft where the finger is missing.

In order to obtain a satisfactory commissure and to prevent later spreading of the fingers, reconstruction of the deep transverse metacarpal ligament is necessary. Barsky makes two drill holes through both metacarpals adjacent to the cleft just proximal to the heads. Chromic catgut sutures are passed through these holes and tightened to approximate

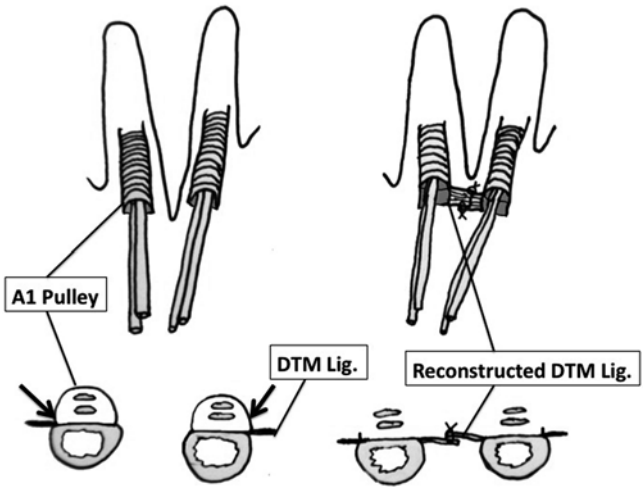

Fig. 15.11 Reconstruction of the deep transverse metacarpal ligament using flexor tendon sheath. Ligamentous flexor tendon sheaths are cut and ligamentous flaps are made. They are turned over and sutured each other

the diverging metacarpals on each side of the cleft. Flatt [6] used to fashion some sort of ligament out of the adjacent soft tissues, but he used also catgut or silk sutures in a technique similar to that reported by Barsky. Free tendon graft can be also used for tethering the adjacent metacarpals. However, one should know that excessive tethering of the metacarpals causes rotation of the metacarpals and cross over the fingers during grasping. Excessive force should be avoided to coapting the two metacarpal together. If excessive force is necessary to coapt the two metacarpal together, metacarpal osteotomy or metacarpal shift is recommended. In order to reconstruct the deep transverse metacarpal ligament, the author uses ligamentous flaps made out of the flexor tendon sheaths of the index and ring fingers (Fig. 15.11). The advantage of this method is that the reconstructed deep transverse metacarpal ligament is located in anatomical position and it is possible to avoid excessive tethering or rotation of the metacarpals. The index finger and ring finger are drawn together. If there is slackening of the extensor hood, it should be repaired by plication or tendon transfer. Then the deep transverse metacarpal ligament is reconstructed by flexor tendon sheath. If osteotomy or metacarpal shift is necessary, it should be carried out before reconstruction of the ligament.

Widening of the Thumb Web Space or Syndactyly Release of the Thumb and Index Finger

When the cleft of the hand is deep, the thumb web space is narrow. In this type of cleft hand, cleft closure and release of the adduction contracture of the thumb are necessary. Various

procedures have been reported to treat the cleft and syndactyly simultaneously. In every procedure, a rotation flap fashioned from the skin of the cleft is used to separate the web between the thumb and index finger and ulnar transposition of the index finger is performed to close the cleft. Snow and Littler used a palmar-based flap from the cleft, Takahashi and Yabe [68] used dorsal and palmar flaps from the cleft. Miura et al. [69], Ueba [70, 71], and Upton and Taghinia [72] solved this problem by transposition of the index finger ray to the ulnar side of the cleft by using skin incision around the base of the index finger. In all these procedures, an osteotomy is performed at the base of the second metacarpal or the index finger metacarpal is transferred to an ulnar finger ray. In these procedures, care must be taken to preserve adductor pollicis muscle and prevent injury of the ulnar nerve to the adductor pollicis.

On the other hand, Foucher et al. [73] stated that none of surgical techniques reported previously is easily applied to the treatment of very deep clefts accompanied by a significant divergence of the metacarpal bones. In such cases, the results of current techniques are disappointing. They proposed a new technique of "Translocation in the Radial Direction of the Ulnar Finger(s)" (TRUF) by intracarpal osteotomy. The reported cases were limited. The TRUF operation allowed closing of the cleft, alignment of the metacarpal bones, and preservation of carpometacarpal mobility. They transfer the little finger or the little and ring fingers with carpometacarpal joint(s) and hamate radially after intracarpal osteotomy. They put the hamate and ulnar fingers on the capitate. The best indication of this procedure is in the case of good alignment of the second metacarpal with the radius and no stump of the middle metacarpal but divergence of the ulnar finger(s). If the second metacarpal has severe radial inclination, a closing wedge osteotomy of the ulnar base of the index metacarpal should be performed.

Previously, the author used a dorsally based flap from the cleft to widen the first web. The skin incision outlines the sides of the cleft on the palmar surface of the index and ring fingers forming a zigzag incision with a proximal V-shaped apex. At the sides of the adjacent metacarpal heads, a ulnarly based small triangular flap is raised by this incision. As the incisions curve back onto the dorsal aspect, they run almost parallel the index finger to the cleft side of the midline of the two fingers. Additional incision starts on the palm of the thumb and index web at the same level as the V-shaped cleft incision. It runs distally parallel with the index finger, curves back onto the dorsal aspect of the thumb–index web and runs proximally and across in an ulnar direction to meet the dorsal index cleft incision. Fibrous bands between the thumb and index finger and fascia of the adductor pollicis and first dorsal interosseus muscle have to be released. Care must be taken to avoid the injuries to the neurovascular bundles. The index finger and ring finger are drawn together and deep transverse metacarpal ligament is reconstructed by a flap of the flexor tendon sheath. Osteotomy of the metacarpal may be performed, if it is necessary. The flap raised from the cleft is transposed to the thumb–index web and wound is closed in layers. However, the dorsal zigzag scar is not esthetically acceptable and in some cases necrosis of the distal tip of the flap due to poor circulation occurred. The author has used palmar rotation flap from the cleft to widen the first web (Snow–Littler procedure) for the past 25 years. The procedure is nearly the same as dorsal rotation flap [74, 75] (Figs. 15.12 and 15.13a, b). The digital artery is not included in the flap, but necrosis of the distal tip of the flap has never occurred.

When the cleft is very deep or there is complete syndactyly between the thumb and index finger, the Snow–Littler procedure is not indicated, as it is not easy to adapt the rotation flap from the cleft to the first web and the created deep V-shaped first web is not as esthetically acceptable. If there is complete or nearly complete syndactyly between the thumb and index finger, a palmar rotation flap from the cleft can be used but usually is not enough to cover the raw surface of the first web and a full thickness skin graft is necessary. In such cases, dorsal and palmar triangular flaps from the first web with free skin graft are better than Snow–Littler procedure in order to obtain functionally and esthetically good first web.

Syndactyly Release Between Ring and Little Finger

Separation of syndactyly between ring and little fingers is carried out by using dorsal rectangular flap combined with free skin graft. This surgery is usually performed when the patient is about 2 years old. Sometimes author performs cleft closure and separation of syndactyly between the ring and little fingers simultaneously. In such cases, there is a benefit to be able to use skin removed from the cleft for the free skin graft, if rotation skin from the cleft to the first web is not needed. If cleft hand is associated with the fourth and fifth metacarpal fusion, and partial cutaneous syndactyly between the ring and little fingers, deepening of the web space improves the appearance and the length of the fingers. In such cases, syndactyly release is indicated electively.

Correction of the Deviation of the Thumb

Deviation of the thumb in cleft hand is often caused by triphalangeal thumb with a delta phalanx or rectangular extra phalanx [76]. Deviation of the thumb is corrected by remov-

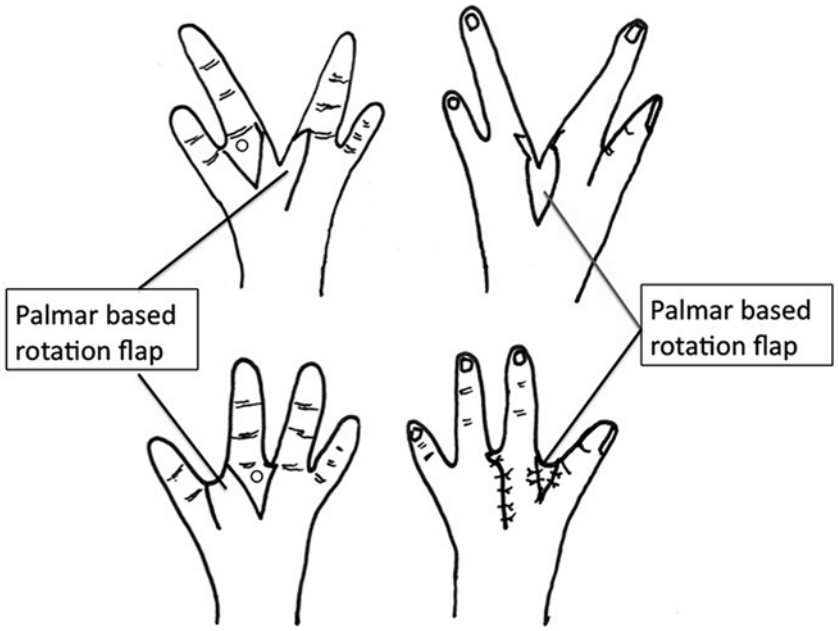

Fig. 15.12 Widening of the narrow first web by Snow–Littler procedure

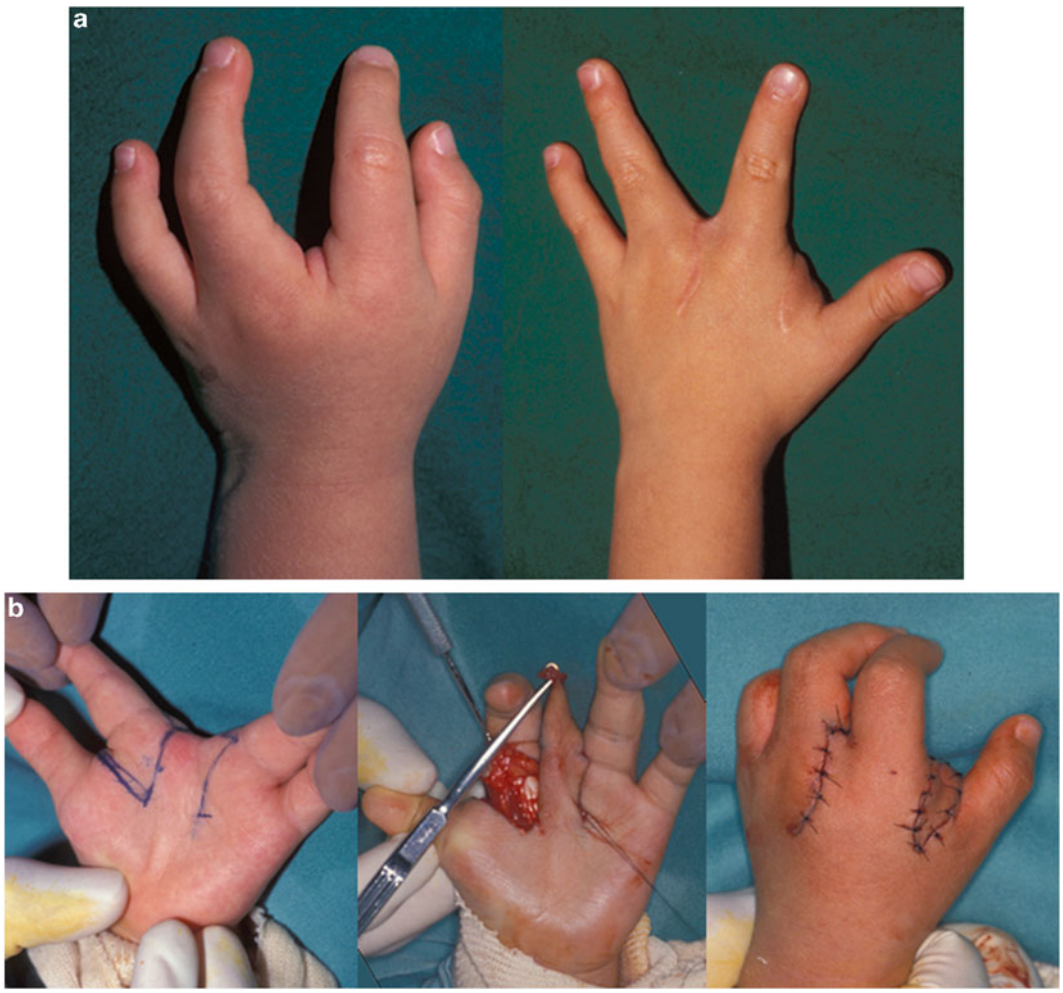

Fig. 15.13 (**a**) Snow–Littler procedure. *Left*: preoperative. *Right*: postoperative. (**b**) Snow–Littler procedure: during surgery. *Left*: design of skin incision. *Center*: palmar flap from the cleft. *Right*: transferred flap into the thumb web space and skin closure

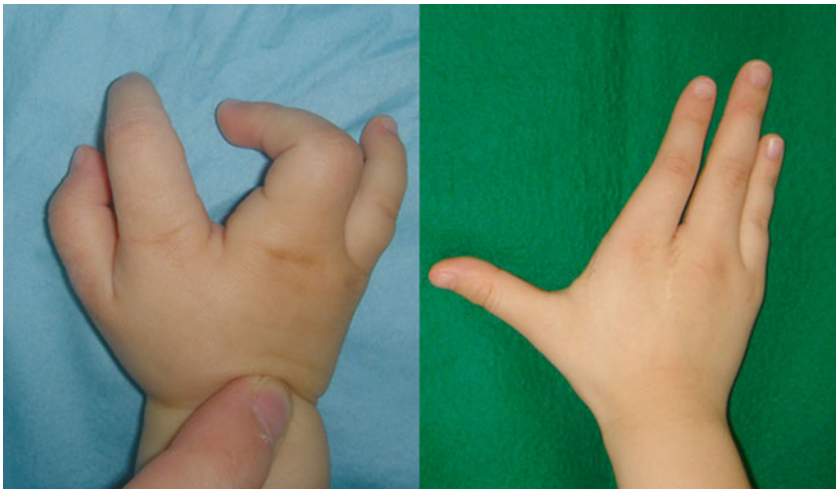

Fig. 15.14 Correction of the claw finger associated with cleft hand

ing the delta phalanx when the patient is less than 5 years. If the child is older than 5 years, the PIP joint or the DIP joint, where angulation occurs, is shortened and fused.

In removal of the delta phalanx, a short midlateral incision over the convex side of the thumb is used. The capsular structure including the collateral ligament is incised longitudinally and split. The delta phalanx is removed and the IP joint is fixed with a Kirschner wire for approximately 6 weeks. The collateral ligament is shortened and repaired, but the redundant skin is not excised, as it recovers spontaneously.

Correction of the Deviation of the Index Finger

Deviation of the index finger in cleft hand is caused by inclination of the DIP joint due to the rectangular middle phalanx. Most patients have no complaint due to this deformity. However, some patients will strongly desire correction of this deformity, when they reach adolescence. In such cases, corrective closing wedge osteotomy at the distal third of the middle phalanx is indicated.

Osteotomy of the phalanx: Longitudinal dorsal skin incision is carried out and middle of the extensor tendon is cut longitudinally. Closed wedge osteotomy is performed after subperiosteal exposure; fixation is carried out by crossed Kirschner wires or modified interosseous wiring.

Correction of Claw Finger Deformity of the Ring Finger

If the middle finger is absent, the ring finger may have a claw finger deformity. This deformity is described as camptodactyly associated with cleft hand in many papers. Flexion deformity of the PIP joint becomes rigid when the patients ages if passive correction of the PIP joint flexion deformity is not performed. This is not a true camptodactyly because the patient able to extend the PIP joint actively when the hyperextension of the MP joint of the affected finger is corrected to a neutral or slightly flexed position. Passive stretching and continuous splinting may correct this contracture. When cleft is closed around 1 year of age, spontaneous correction of the flexion deformity is sometimes observed, as hyperextension of the MP joint is usually corrected by the tension of the closed palmar skin. However, flexion deformity of the PIP joint or claw finger deformity should be corrected with tendon transfer at the initial surgery. In order to close a deep cleft, the structures including tendon and bone under the cleft are exposed. It is at this time that the surgeon has the best chance to select a tendon for transfer. In most cases of cleft hand, flexion of the PIP joint caused by the dysfunction of the intrinsic muscles. During surgery, we can often observe the extra flexor digitorum superficialis tendons of the ring finger or middle finger. One of them can be detached from the membranous insertion at the end of the stump of the missing finger. It is then transferred to the base of the proximal phalanx or the proximal end of the ligamentous flexor tendon sheath of the ring finger [77] (Fig. 15.14). If claw finger deformity is associated with divergence of the index and ring fingers, the detached flexor digitorum superficialis tendon may be divided into two slips. One slip is transferred to the radio-palmar periosteum of the base of the proximal phalanx of the ring finger and the other slip is transferred to the palmar periosteum of the ulnar base of the proximal phalanx of the index finger. The same type of procedure may be performed for extensor side. Extrinsic extensor tendon to the ring finger is transferred to the dorso-ulnar side of the expansion hood of the index finger and the extensor digitorum communis to the index finger is transferred to the dorso-radial side of the expansion hood of the ring finger. These procedures may prevent divergence deformity of the index and ring fingers when the fingers are extended.

Summary

Cleft hand is often associated with other deformities, which are phenotypes of abnormal induction of the finger ray numbers in the hand plate. When one classifies the congenital hand deformities, one has to face the problems how to classify the cases associated with cleft hand, central polydactyly, and syndactyly. It is easy to understand the association of these anomalies, once the concept of abnormal induction of the finger ray numbers in the hand plate has been accepted. The situation is the same as in congenital constriction band syndrome. One can easily understand the association among constriction band, acrosyndactyly, and amputation, if one has accepted the concept of congenital constriction band syndrome.

As I mentioned before, many authors think surgery of cleft hand is mainly performed for esthetic reasons, but some of the procedures have been performed for functional improvement. As in other congenital hand deformities, patient with cleft hand should use their hand skillfully when they become old, even if they have not surgically treated. Prof. P.C. Leung in Hong Kong, who is a respected hand surgeon and a person, delivered a lecture and told us as follows. Surgeons should be ambitious for treating the child with congenital hand problems. However, over ambitions may lead miserable surgeon and miserable patient. Surgery has its limitation and never forget "Don't make it worse!"

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