

Case Report

Unusual case of cleft hand

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ABSTRACT

We present a case of a six-year-old male child with cleft hand deformity involving the dominant right hand. It was a rare case of atypical cleft hand with no missing tissue but cleft extending to metacarpal level and associated hypoplasia of thumb and index finger. As per Manske's classification of cleft hand our patient belongs to the Class III variety. There was associated malposition of the index finger with absence of first web space and syndactyly of thumb and index finger at the metacarpal level. A modified Snow-Littler procedure was planned. The surgical plan involved closure of cleft, release of thumb and index finger syndactyly and reconstruction of the first web space. The functional outcome was good considering hypoplasia of the index finger and thumb. Depending upon the function of the thumb tendon transfers can be planned to augment thumb function at a later date along with correction of rotational deformities of the index and middle finger.

KEY WORDS

Central hand deficit, cleft hand, split hand

Deficiencies of the central portion of the hand have been named as cleft hand, split hand, lobster claw, ectrodactyly by many authors.^[1] The incidence is 1 to 4 in 100000 live births.^[2] It usually appears bilaterally and is associated with cleft feet. Autosomal dominant inheritance is known. Associated anomalies^[3,4] include cleft feet, congenital heart, cataracts, defect of tibia, radioulnar synostosis and fusion of kidneys. There is a 60% male preponderance. Severe cases are associated with syndactyly of thumb and index finger and of ring and little finger with absence of third metacarpal ray.^[5] In more severe cases there may be absence of all digits of the hand. We present a mild variety of cleft hand wherein all digits of the hand were present with hypoplasia of the thumb and index finger. Flatt^[6] has classically described this anomaly as functional triumph and social disaster.

Cleft hand deformity is secondary to wedge-shaped

defect in primary hand plate. The apical ectodermal bridge does not develop normally and there is a defect in its differentiation. According to Maisel's^[7] theory of centripetal suppression of the primary hand plate, in the mild variety there is no missing tissue but cleft is present deep down to the metacarpal level. In the moderate variety, along with the cleft, digits on the radial side of the hand are missing and in the most severe form even digits on the ulnar side of the hand are missing. Our patient belongs to the second group with hypoplasia of radial digits of the hand rather than absence of the same.

CASE REPORT

We present a case of a six-year-old male child with moderate variety of cleft hand involving the right hand (dominant) with no missing tissue but cleft extending to the metacarpal level with associated hypoplasia of the thumb and index finger [Figures 1 and 2]. There was no positive



Figure 1: Pre-op - clinical photo – Volar side cleft hand



Figure 2: Pre-op –clinical photo – dorsal side cleft hand



Figure 3: Pre-op X-ray cleft hand



Figure 4: Pre-op markings. Flap A – for reconstruction of first web, Incision B – for release of first web, Incision C – for skeletanisation of index finger

family history. The child did not have other associated congenital anomalies involving the heart, long bones of the leg, feet or kidney. The other hand was normal. As per the widely accepted Manske's^[8] classification of cleft hand (surgical classification of central hand deficiencies according to thumb web) our patient belongs to the Class III variety which is in the midway range from Grade One to Five in the order of severity. There was associated malposition of the index finger with absence of the first web space and syndactyly of the thumb and index finger at the metacarpal level. There was associated rotational deformity at the metacarpo-phalangeal level both in the index and middle finger. The X-ray of the hand was done to confirm the same [Figure 3]. No other imaging was done due to cost constraints.

A modified Snow-Littler^[9] procedure was planned after obtaining fitness for anaesthesia. The surgical plan

involved closure of cleft, release of thumb and index finger syndactyly, release of adduction contracture thumb and reconstruction of the first web space. This procedure was chosen over others because it releases the thumb and index finger syndactyly down to the metacarpal level and broadens the first web with transfer of palmar skin flap from the cleft. Corrective osteotomies for the index and middle ray would be done at a later stage in view of the possibility of associated vascular anomalies. The indications for surgery in this case were adduction contracture of the first web and closure of the cleft for cosmetic, social and psychological reasons.

The aims of reconstructive surgery were:

1. Restoration of the position of the thumb with preservation of its function
2. Transposition of the index finger ray to the ulnar side of the cleft



Figure 5: Intra-op picture. First web released, index ray skeletonised and flap A raised



Figure 7: Post-op X-ray cleft hand showing K wire holding second and third metacarpal



Figure 6: Intra-op picture. K wires *in situ* and first web reconstructed with flap A



Figure 8: Post-op clinical photo at six months showing thumb and index finger opposition

3. Adequate first web space
4. Reconstruction of transverse metacarpal ligament
5. Preservation of intact adductor pollicis and intrinsic muscles.

Patient was given general anaesthesia and surgery was performed under tourniquet control. Flap A was marked from the radial side of the middle finger. Incision B was planned for release of the adduction contracture of the first web. The index finger ray was skeletonised with a circumferential incision (Mark C) [Figure 4].

Figure 5 clearly shows skeletonised index finger with preservation of its musculotendinous and neurovascular units. Also seen is the released first web space with Flap A raised to reconstruct the same.

The index finger ray was transposed towards the middle



Figure 9: Post-op clinical photo at six months showing power grip with operated hand

finger ray and a transverse K-wire was passed into the second and third metacarpal to hold them together. Transverse metacarpal ligament was reconstructed using

local tissues. Kirschner wire was used to fix the thumb in functional position of abduction and Flap A was used to reconstruct the first web space [Figure 6]. The postoperative x ray shows k wire holding second and third metacarpals [Figure 7].

Closure of all wounds was achieved primarily after haemostasis and a padded dressing was given. Postoperative recovery was uneventful and sutures were removed on the tenth day. K wires were removed after a period of six weeks. Patient was advised physiotherapy thereafter. Figures 8 and 9 show final result after six months with good functional recovery and dramatic improvement in the cosmesis of the hand.

DISCUSSION

The ideal age for surgery of closure of cleft and first web reconstruction is around 18 months. If associated with syndactyly it should be separated earlier at the age of six months. Since our patient presented late at the age of six years we planned syndactyly release with closure of cleft and release of adduction contracture of the first web in a single-stage surgery. The syndactyly was of a complicated type as it was associated with hypoplasia of the thumb and index finger.

The traditional method of closing the cleft is to remove excess tissue, rudimentary bone if any and close the skin with interdigitating flaps to avoid scar contracture. Kelikian and Doumanian^[10] recommended fashioning a rectangular flap from the side of the cleft to bridge the gap.

Most cases present with marked First web contracture and hypoplasia of the thumb. Snow and Littler^[9] described transposition of second ray to third ray after removal of remnant of third metacarpal. They broaden the first web using skin from the palmar side. Miura and Comada^[11] have described a technique simpler than Snow-Littler's, wherein ulnar transposition of the index ray is done and first web is created by using the dorsal skin flap. In our patient all the metacarpals were well developed and the cleft was deep down hence it was not possible to use dorsal skin for first web reconstruction. We skeletonised the second ray and mobilised it towards the third and reconstructed transverse metacarpal ligament between two rays using local tissues from the flexor tendon sheath.

This will prevent it from springing apart. There was difficulty in mobilizing the second metacarpal ray towards the ulnar side as the second metacarpal could not be osteotomised in our patient.

The rotational deformity of the middle finger was not corrected in the same sitting in view of the possibility of associated vascular anomaly. This would require second surgery at a later date. The functional outcome was good considering the fact that the index finger and thumb are hypoplastic. Depending upon the function of the thumb tendon transfers can be planned to augment thumb function at a later date. We could reconstruct a wide first web space with the help of a flap from the volar skin of the middle finger as seen in Figure 6 which enable him to effectively abduct his thumb.

Both the child and parents are extremely happy and the child has started going to school. We are maintaining a close follow-up bimonthly and would offer the child further reconstructive procedures to improve function of his dominant hand.

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