

Simple Syndactyly

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Syndactyly is a common hereditary digit malformation where adjacent fingers are webbed due to a failure to separate during limb development. It may present as an isolated deformity or as part of a generalized syndrome.

The normal finger webspace slopes at 45 degrees, starting dorsally at the metacarpophalangeal [MCP] joint level and it slopes distally and palmarly to the mid-point of the proximal phalanx volarly. It is mostly comprised of dorsal non-glabrous skin. The natatory ligaments help form the web contour, while joining adjacent lateral digital sheets.

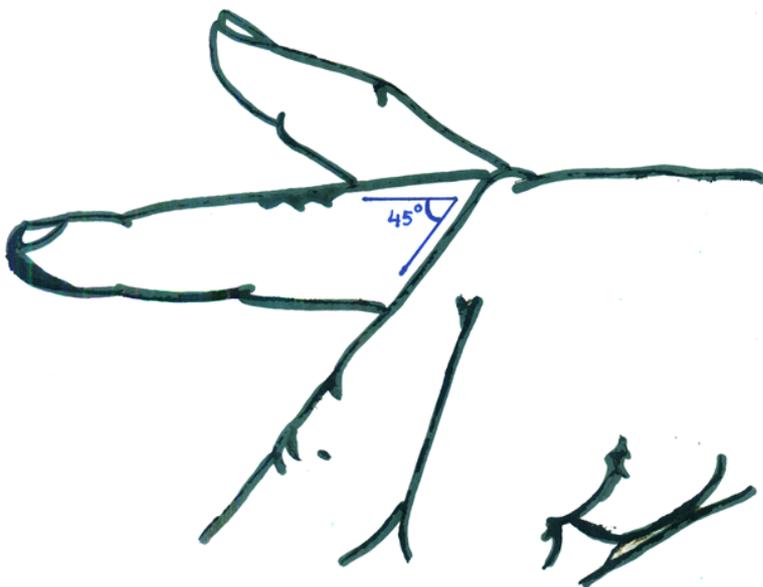


Fig. 1: The normal slope of the web space.

Embryology:

After the upper limb buds appear around day 27 of intra-uterine life, the distal ends of the buds flatten to form the hand plates by day 35. Digit rays begin to condense by day 41 mediated by the Apical Ectodermal Ridge [AER]. The rays are well defined by day 46. Between the rays are regions termed the Interdigital Necrotic Zones [INZs], which are fated to undergo apoptosis. This cell death is possibly mediated by Fibroblast Growth Factors [FGFs] and Bone Morphogenetic Proteins [BMPs]. By day 52, the rays are essentially separate.

Genetics:

Isolated syndactyly is almost always autosomal dominant. Candidate regions have been identified on the second chromosome (2q24-q36). Syndromic syndactyly is seen commonly with Poland, Apert, Crotzer and orofacioidigital syndromes.

Epidemiology:

Simple syndactyly is seen in about one in 3000 live births, and is twice as common in males as females. About half the cases are bilateral and the toes may be involved as well. Most common between the ring and long fingers (40% to 50%), followed by between ring and small fingers (25% to 28%). First web space syndactyly is the least common (7% to 9%).

Risk Factors:

Maternal smoking, poor nutritional status, lower socioeconomic status.

Classification:

Syndactyly is 'Simple' when digits are connected by only skin and soft tissue. This may extend to the fingertips ('Complete') or end proximal

to it ('Incomplete'). 'Complex' syndactyly contains bony fusion as well, while the presence of abnormal osseous elements is defined as 'Complicated' syndactyly.

Clinical Evaluation:

History / Examination

Physical examination must include entire affected upper limb, contralateral hand, chest wall, and feet to search for additional anatomical differences.

The presence of well-formed flexion and extension creases signify normal joint development and are commonly so in simple syndactyly. Clinically, osseous fusion can be ruled out by checking relative motion between the webbed fingers.

Before proceeding with the surgery, the parents should be explained about the relative lack of skin which will be evident following the release. It can be demonstrated by measuring the circumference of the parents' middle and ring fingers separately and comparing their combined measurement with the fingers held together. On average, there is a deficiency of 30% (*fig 2*).

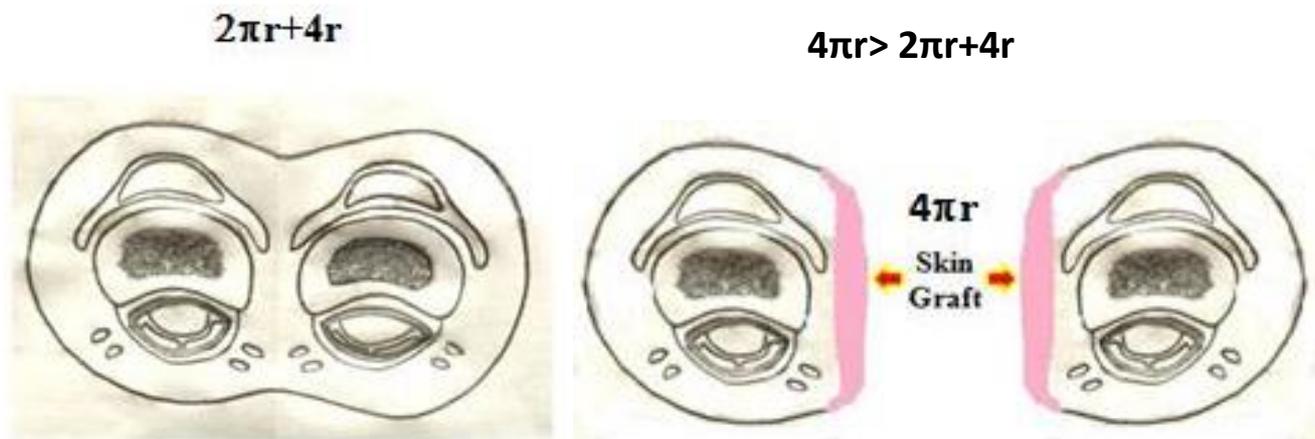


Fig 2: Greater skin requirement following syndactyly separation.

Imaging:

Plain radiographs of hand are taken to detect skeletal deformities, concealed extra digits, or articular deformities. Other investigations are needed only for syndromic cases.

Treatment:

“... ask not how soon the operation can be done but rather how late the functional demands will allow postponement of surgery” – Adrian Flatt.

Surgical release is indicated in almost every case except for a mild, incomplete syndactyly without functional impairment. However, the timing and technique are often matters of controversy.

Timing:

When two digits of unequal lengths are involved in a syndactyly, the longer digit suffers flexion and lateral angulation deformities as seen with the border digits. Hence, when the thumb or the small finger is part of a syndactyly, they require earlier release at around six months of age. For the 2nd and 3rd web, this can be postponed to around the 18th month without significant functional harm, when the hand becomes larger. In multiple web space syndactyly, more than one web can be released as long as one finger is not operated on both sides at once as the skin flaps can be jeopardized. Bilateral releases can be performed in the very young; however it can be very distressing for older children to have both upper limbs covered in dressings. Hence one side at a time is preferred.

Principles:

While many techniques exist, some principles which should be adhered to, while performing any technique, have been more or less accepted over the years.

- Only one side of a digit is released at a time.
- The web space is recreated from a local flap, usually dorsal.
- The sides of the fingers are closed with zigzag skin flaps.
- Flaps are defatted prior to closure.
- Bare areas are covered with full thickness skin grafts.

Surgical technique:

Reconstruction of the web commissure is the most critical part of syndactyly separation. It would determine the shape of the web space and chances of recurrence. Hence planning this flap is very crucial. A rectangular, proximally-based dorsal flap for the web space is the most common reconstructive design for the commissure. Numerous geometrical designs have been used for this flap, including rectangular, triangular, omega and multi-lobed shapes. The dorsal rectangular flap is planned extending to the two-third of length of a line joining the midpoint of metacarpal heads to the midpoint of proximal interphalangeal joints (*fig 3*). The reconstructed commissure should be created proximal than its normal position, to allow for the distal migration of the commissure or the web creep with growth of the child.

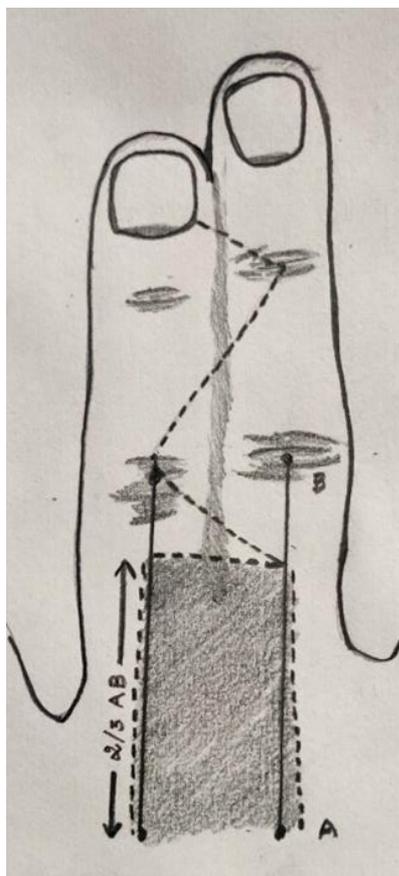


Fig. 3: Planning of the dorsal rectangular flap

In the fingers, multiple zig-zag incisions are used. Cronin's technique of matched zigzag incisions on the volar and dorsal surfaces is popular. Modified Cronin's technique with oblique zig zag incision can be used to avoid skin grafting on both sides of the commissure.

Withey and Sommerlad developed the "open-finger" technique, which modified the zigzag incisions into longer and narrower angled flaps that were left open to heal by secondary intention.

Most of the time, there will be residual raw areas on the sides of the released digits. Coverage of these soft tissue deficits is usually achieved with full-thickness skin grafts. Split-thickness skin grafts are at greater risk for contracture. Grafts should be avoided in the commissure, as this area requires supple skin. The common donor sites are mostly the groin, the medial malleolus region, volar wrist and elbow crease and instep of the sole.



Fig. 4: Steps of syndactyly separation

In children with simple, complete syndactyly, creation of the paronychia folds become challenging. In these cases, laterally based triangular flaps described by Buck- Gramcko can be used to create the paronychia folds and contour of the finger.

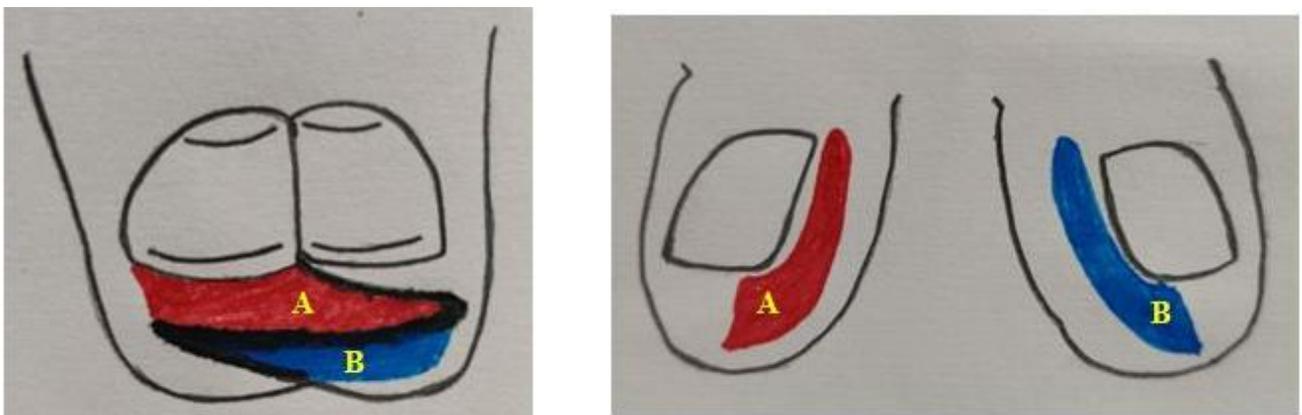


Fig. 5: Laterally based triangular flaps for paronychia folds in complete syndactyly.

Potential Complications:

Early complications include iatrogenic nerve or artery injury and graft or flap failure. Web creep is the most common long term complication. Joint contractures and nail deformities are also seen.

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