

Case Report

Congenital Syndactyly of the Hand: A Case Report and Literature Review

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Abstract: The most common congenital defect of the hand is syndactyly. This is caused by incomplete separation of the fingers during limb development. In this article, we discuss the embryological basis, risk variables, and classification methods that guide surgical decision-making. The aim of reconstructive surgery is to produce a functional, aesthetic, and independent web space without any sequelae such as scar contracture or digital necrosis. We discuss indications for intervention, including timing of surgery according to the type of syndactyly and associated disorders. Current surgical techniques involve full-thickness skin grafts, dorsal advancement flaps, and a modified dorsal pentagonal flap.

Keywords: Congenital Hand Anomaly, Syndactyly, Reconstructive Surgery, Pediatric Hand Surgery, Web Space Reconstruction.

INTRODUCTION

Syndactyly is the most common congenital anomaly of the upper limb and accounts for 10%–28% of all congenital hand deformities, with an estimated incidence of 1 in 2,000–1 in 2,500 live births [1]. This condition is characterized by incomplete or failed differentiation of the digital rays during limb morphogenesis, resulting in abnormal interdigital fusion that may involve only the skin and soft tissues (simple syndactyly) or extend to osseous structures (complex syndactyly) and, in some cases, include accessory phalanges or anomalous nails (complicated syndactyly) [2]. There is a significant male preponderance with a male-to-female ratio of approximately 2:1. Bilateral disease occurs in about half of the patients and is often asymmetric [3].

CASE REPORT

An 18-month-old child presented to the pediatric hand clinic with a fused ring and middle finger on his left hand. The infant was born at term, and there was no family history of congenital malformations. On examination there was webbing of the ring and middle fingers extending beyond the distal interphalangeal joint, without synonychia. No shortening of the affected digits was noted, and range of motion at metacarpophalangeal, proximal, and distal interphalangeal joints was intact. General examination did not reveal any syndromic symptoms. Examination of feet and contralateral hand was unremarkable. The kid had surgical release for simple incomplete syndactyly. A modified Flatt approach was employed with the reconstruction of the webspace with interdigitating zigzag flaps and a dorsal hourglass flap.

EMBRYOLOGY

The upper limb buds appear 26-27 days after fertilization, and the lower limb buds appear 28-30 days after fertilization. Fingers on the upper limb separate at 41-43 days and are completely separate by 52-53 days. The mesenchymal core of the limb bud is surrounded by the ectoderm. The genetic control of limb bud development and differentiation is

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complicated. Three directions of development are specialized signal centers. The proximal-distal outgrowth needs the apical ectodermal ridge (AER). Experimental removal of the AER decreases growth [4]. The progress zone is the mesodermal cells beneath the apical ectodermal ridge that receive signals to produce certain cell types for certain tissues in certain locations in the limb. The mesodermal cells of the postaxial zone of polarizing activity in the developing limb control the anteroposterior or radioulnar axis. This region suppresses the growth of the thumb and encourages the growth of the ulnar digit. Cell translocation from the polarizing zone to the preaxial or radial limb diminishes thumb development. The third signaling center regulates the dorsal ectoderm cells that determine the dorsal-ventral properties of the limb. Transplanting dorsal ectoderm to the ventral side of the limb bud results in the formation of dorsal limb structures on both sides [5]. The three signaling centers talk to each other by releasing specific morphogens. These morphogens influence the growth of the limb via chemical pathways. Engrailed 1 and Lmx-1b cooperate with the sonic hedgehog protein (SHH), fibroblast growth factors (FGFs), and Wnt-7a to pattern the dorsal and ventral and to modify the apical ectodermal ridge. The myelin cells die through a process known as apoptosis. By 8 weeks, digital separation is almost complete, and ossification centers for the phalanges are beginning to form. 23 Members of the BMP family may be involved in apoptosis. To kill cells, the AER has to secrete proteins, fibroblast growth factors, and members of the homeobox (Hox) gene family [6]. And when the AER is cut out, the process is complete. BMP-4 is released in the interdigital mesenchyme and joint primordium when the distal digits separate. The digits in the embryo are completely separated from one another, and BMP-4 expression is found only in the areas where joints are forming. Syndactyly is induced by low doses of BMP-4, which block apoptosis in the mesodermal tissue between the digits. It is correlated with the width of the limb bud, which is determined by the length of the basal ectodermal ridge. Changes in BMP-4 reduction and SHH expression can trigger polydactyly [7].

SINDACTYLY AND SWANSON CLASSIFICATION

The Swanson classification was developed by Dr. Adrian Swanson and was adopted by the International Federation of Societies for Surgery of the Hand (IFSSH) [8]. This classification system is the most accepted system for the classification of congenital upper limb anomalies. This classification is based on the concept of embryologic failure, classifying anomalies according to the developmental process that has been disturbed and not only on the anatomical appearance. Simple syndactyly is limited to soft tissue, while complex syndactyly involves fused bones (synostosis) between two or more fingers. Incomplete syndactyly refers to fusion at the distal phalanx, and complete syndactyly involves fusion to the proximal phalanx. Complicated syndactyly is the presence of bone growth between two digital rays (hidden polydactyly) and acrosyndactyly is when the distal phalanges have a proximal webspace [9].

SURGICAL TREATMENT

A clinical exam can tell you how much the fingers are involved. A clinical examination can distinguish between simple, complex, and complicated syndactyly. Active range of motion is present in the interphalangeal joints with well-defined flexion and extension creases suggestive of normal joint architecture and simple syndactyly. In the absence of flexion and extension creases and immobility of the joints, complex or complicated syndactyly should be suspected. The clinical diagnosis of synostosis is the immobility of the distal phalanges and the fusion of the nails, seen in Poland syndrome, Apert syndrome, amniotic band syndrome [10].

A plain X-ray examination of the affected hand is important in detecting synostoses, occult polydactylies, and other skeletal deformities, particularly in complex syndactylies. Treatment of syndactyly requires good preoperative planning and careful discussion of the risks and benefits with the family. In patients with irregular finger size, syndactyly release should be performed at 3 to 6 months of age only when there is deviation due to growth.

Alternatively, it may be postponed until the age of 1 to 2 years. Web spaces that are adjacent to each other in the same environment should not be disseminated to avoid vascular compromise. A simple longitudinal separation creates a lot of scar tissue that can interfere with growth and cause joint contractures and deformities. Zigzag closures have therefore been recommended as a means of minimizing longitudinal scarring. A commissure should be created with supple dorsal skin flaps to avoid grafts in this area to prevent web spread and scarring. Moreover, thumb-index syndactyly separation is more difficult than finger syndactyly separation. For moderate narrowing, a 2- or 4-flap z-plasty should be done, or a dorsal rotational advancement flap for isolated complete syndactyly [11].

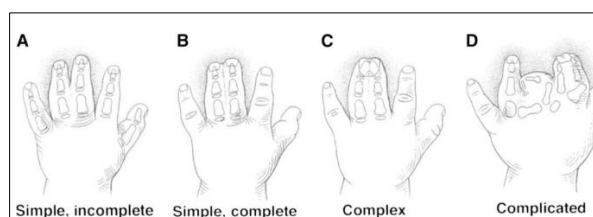


Figure 1: Syndactyly Classification



Figure 2: Ring and Middle Finger Sindactyly, Right Hand.



Figure 3: Simple and Complete Sindactyly



Figure 4: Preoperative marking



Figure 5: Volar View, Result after surgery



Figure 6: Palmar View, immediate outcome

DISCUSSION

Syndactyly is a common but relatively complex congenital hand defect. In the Swanson/IFSSH system, syndactyly is classified as Group II (failure of differentiation) [12]. This case supports some basic ideas presented in the literature. Timing of surgery is important; when the border digits (thumb-index or ring-little) are involved, the release must be performed between six and twelve months of age to prevent progressive rotational deformity and differences in growth. Intervention at 12-18 months is safe and effective in core web spaces. This reduces anesthetic risk and prevents functional adaptation. Choice of surgical technique directly influences results. The dorsal rectangular flap (Flatt) in combination with full-thickness skin grafts (FTSG) is still the gold standard [13]. The technique provides even, durable coverage of the web and reduces scar contracture. Comparative studies indicate that the results of full-thickness skin grafts (FTSG) from the groin or hypothernar eminence are better than split-thickness grafts, with less web creep and better cosmetic results [14]. Graftless procedures such as the dorsal pentagonal flap may be suitable for partial, simple syndactyly but require greater flap mobility and are not suitable for complex or complete syndactyly. Complications such as web creep (>2 mm distal migration), digital angulation, or neurovascular injury occur in 10 to 15% of cases [15]. Prevention involves a precise methodology: extensive defatting of the skin, zigzag incisions to avoid linear scarring, and tension-free application of the graft¹⁶. Postoperative splinting and scar massage are equally important.

CONCLUSION

Syndactyly is a congenital hand malformation caused by failure of interdigital apoptosis during embryogenesis. Accurate classification using the Swanson/IFSSH system is important for appropriate management and helps to guide the timing and technique of reconstructive surgery.

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