Management of Complications of Congenital Hand Disorders

Garet C. Comer, MD, Amy L. Ladd, MD*

KEYWORDS

• Syndactyly • Polydactyly • Thumb duplication • Radial longitudinal deficiency • Radial club hand • Thumb hypoplasia • Pollicization • Epidermolysis bullosa

KEY POINTS

- A thorough knowledge of patient presentation, natural history, and treatment options is essentially to effectively manage patients with congenital hand differences.
- Although some treatment complications are inevitable, judicious preoperative planning, meticulous technique, and knowledge of surgery-specific lessons help to decrease the risk of complications.
- Management of syndactyly, camptodactyly, ulnar polydactyly, radial polydactyly, thumb hypoplasia, radial longitudinal deficiency, and epidermolysis bullosa has evolved with refined understanding of disease processes and treatments.

INTRODUCTION

Advances in the care of patients with congenital hand differences have made dramatic progress in the modern era. These advances include refined surgical procedures, improved classifications, new technology, improved understanding of the embryology and pathogenesis of disease, and an increased understanding of the psychological implications of having a congenital hand difference This article focuses on 7 conditions frequently treated by hand surgeons: syndactyly, camptodactyly, ulnar polydactyly, radial polydactyly, thumb hypoplasia, radial longitudinal deficiency (RLD), and epidermolysis bullosa (EB). We review each topic and discuss methods to avoid complications in the treatment of patients with these conditions.

SYNDACTYLY

Syndactyly occurs in 1 in 2000 to 3000 live births, making it one of the most common congenital hand anomalies.[1] It most commonly appears in isolation, but may also be associated with congenital syndromes, such as Apert syndrome and Poland syndrome.[2] Syndactyly most commonly affects the third web space followed by the fourth, second, and first web spaces, respectively. It is frequently bilateral and is more commonly seen in males than females.[1,3–5] In addition to the hand, patients should be evaluated for foot involvement and associated syndromes.

The timing of surgical release of the digits is debatable, although surgery is most frequently undertaken at 12 to 18 months of age. Although good results have been reported with delaying

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Department of Orthopedic Surgery, Robert A. Chase Hand & Upper Limb Center, Stanford University, 450 Broadway Street, Pavilion C, Redwood City, CA 94063, USA
* Corresponding author.
E-mail address: alad@stanford.edu

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surgery until 4 or 5 years for simple syndactyly,[1] separation is typically not delayed past 12 months for digits of unequal length to avoid deformity caused by the shorter digit tethering the longer.

The surgical treatment of syndactyly has evolved dramatically over the years. In the 19th century, digits were separated with straight dorsal and palmar incisions with attempted primary closure.[6] However, this led to unacceptable results from wound closure under tension and contracture formation.[7] Currently, surgery is based on the following principles:

1. Division of the web with zigzag incisions,
2. Achieving tension-free closure by utilizing free skin graft when primary closure is not possible, and
3. Applying dorsal flap for commissure reconstruction.

Building on these principles, subsequent reports have focused on refining these techniques and improving outcomes. Specifically, recent literature has focused on split-thickness versus full-thickness skin grafts, commissure reconstruction techniques, and the use of tissue expanders to obviate the need for skin grafting. Several authors have advocated for the superiority of full-thickness skin grafts over split-thickness grafts by citing a lower rate of contracture and web creep.[1,5,8] However, others have noted comparable outcomes with both techniques.[9]

Many commissure reconstructions techniques have been developed. These include the Flatt dorsal hourglass-shaped rotational web flap,[10,11] V-Y advancement flaps,[12,13] trilobed dorsal flaps,[14] and triangular flap designs.[15,16] Barabás and Pickford[17] reported on 144 webs reconstructions by a modified Flatt technique at a mean of 5 years follow-up. The rate of complications was low with 7 (4.9%) graft failures, 6 patients (4.2%) with web creep, and only 4 patients (2.8%) requiring reoperation. The authors recommend avoidance of longitudinal wound lines extending midaxially from the commissural flap as seen when a volar T-shaped incision is used.

Common complications include (1) web creep—losing the deep commissure performed at surgery - with an incidence of 2% to 24% (**Fig. 1**).[8] (2) Skin graft slough is common and may contribute to web creep, especially with the most proximal grafts set distal to the dorsal flap. (3) Vascular compromise is a potential challenge in complex syndactyly and polysyndactyly, such as Apert syndrome and rotational deformities owing to anomalous transverse or even absent arterial flow (**Fig. 2**). The long-held dictum in syndactyly surgery is that separating only 1 digit at a time in multiple contiguous syndactyly prevents vascular compromise and loss of a digit. We find, however, this is seldom a problem in simple syndactyly when a full complement of neurovascular structures is the rule. (4) Nail deformity is often underappreciated and seen in complex syndactyly, especially Apert syndrome and fused distal phalanx presentations.[18] (5) Finally, rotational deformity may be unmasked in complex syndactyly when the digits are freed and allowed to develop unencumbered (**Fig. 3**). However, this is less a complication and more an unanticipated outcome because deformity development may be unpredictable; nonetheless, the possibility should be reviewed with the family at the initial surgery.

Minimizing complications in the surgical treatment of syndactyly requires meticulous operative technique, tension-free closure, and reduction of shear forces across skin graft sites. Using

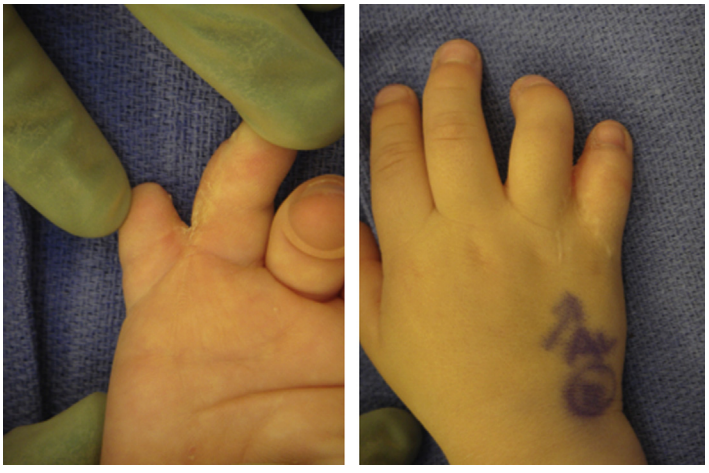

Fig. 1. Web creep after the release of a complete syndactyly.