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Elias H Kahan

Brian Temple

Philadelphia College of Osteopathic Medicine

Logan Carr

Elizabeth Zellner

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An Update and Report Failure of Surgical Syndactyly Repair in Harlequin Ichthyosis

Elias H. Kahan, BA*
 Brian Temple, DO†‡
 Logan Carr, MD‡
 Elizabeth Zellner, MD‡

Summary: Harlequin ichthyosis (HI) is a rare congenital skin disorder caused by irregular epidermal differentiation. Syndactyly in HI is associated with thick hyperkeratotic skin flexion and angulation deformity of the hand and fingers resulting in limited function of the upper extremity. Traditional syndactyly release is limited as full-thickness skin grafts typically used in reconstruction are composed of diseased skin and require donor sites in a patient predisposed for adverse wound healing. This case report is a follow-up to a previous viewpoint written about digital escharotomies in a newborn with HI and outlines a second and fourth webspace syndactyly release with a dermal substitute. Despite early evidence of adequate release and improved hand function, recurrence of syndactyly was observed within 4 months of surgical release. Our experience described within this case report may suggest the limitations and possible alternatives of surgical release of syndactyly in the HI population. (*Plast Reconstr Surg Glob Open* 2022;10:e4594; doi: 10.1097/GOX.0000000000004594; Published online 17 October 2022.)

We previously wrote regarding digital escharotomies to save finger viability in harlequin ichthyosis (HI).¹ Our patient, a newborn that had been undiagnosed prenatally, underwent surgical escharotomy release of hyperkeratinic constriction bands across wrist creases, along individual fingers, and on bilateral ankles on the first day of life. Although retinoids have had success relaxing the hyperkeratosis, we advocated for surgical management to minimize limb ischemia and the risk of autoamputation.² The patient had favorable results immediately postoperative as well as improved digital function, development of web spaces, and minimal scarring 6 months following the procedure.

Since that time, our patient has become a thriving four-year-old, and her parents have created an active social media presence and a foundation to help other patients and families experiencing HI (link: @harlequindiva).

Although her hands healed, the patient's proclivity for skin production resulted in bilateral, incomplete

syndactyly, mild flexion contractures at the metacarpophalangeal (MCP) joints, and slight ulnar deviation. To improve digit function, we performed a syndactyly release in December 2020 on the second and fourth web spaces of the right hand. Significant subcutaneous fat was removed from the MCP joint to two thirds up the digits of the neurovascular bundle. Given the patient's complex skin condition and the uncertainty regarding healing a graft, we used human acellular dermal matrix to close the remaining space (Fig. 1). Recent studies have shown that synthetic skin substitutes yield comparable outcomes to skin grafting among pediatric syndactyly repairs.³ Although other authors have reported successful use of skin grafts when conducting syndactyly release among patients with HI, available data are limited, and the authors were concerned about stimulating additional skin growth.⁴ A nonadherent dressing was fashioned to bolster the skin substitute and was removed within 5 days of surgery. The patient's incisions were nearly healed at 1 week postoperatively and she displayed improved hand motion at the 2-week postoperative visit. She began occupational therapy beginning on the second postoperative week. Reforming of syndactyly was apparent at 6 weeks postoperatively with full recurrence of syndactyly at the 4-month postoperative visit (Fig. 2).

Fourteen months following the procedure, the patient's digit function mirrors the preoperative state. This web creep is likely due to the nature of HI rather than the surgical technique. However, a potential revision to the operative technique may include use of skin graft, which has shown longer-term success without syndactyly recurrence, albeit among limited cases.^{4,5} Additional alterations to the procedure may

From the *School of Medicine, New York Medical College, Valhalla, N.Y.; †Philadelphia College of Osteopathic Medicine, Philadelphia, Pa.; and ‡Department of Surgery, Westchester Medical Center, Valhalla, N.Y.

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Fig. 1. Immediate postoperative photograph after syndactyly release and dermal substitute application.



Fig. 2. Four-month postoperative photograph.

include limiting the duration of postoperative surgical dressings to reduce inflammation as well as using the patient's predilection for skin growth to close the wound by secondary intention. Nonoperative alternatives consist of occupational therapy and continued use of retinoids to reduce the contractures at the joints; however, nonsurgical management will be unlikely to reduce the syndactyly.

We write this not to discourage colleagues from treating this patient population, but rather to share experiences and expectations for future patients.

SUMMARY

Syndactyly in HI presents a unique challenge to reconstructive surgeons due to the inherent nature of disordered epidermal differentiation. Limitations in donor site availability and likelihood of recurrence may preclude attempted release in this population. Our case report suggests a high likelihood of recurrence may be observed and thus should be readily discussed with caregivers before attempting surgical release of syndactyly in patients with HI.

Elizabeth Zellner, MD

Westchester Medical Center
Ambulatory Care Pavilion, 1st Floor
100 Woods Road
Valhalla, NY 10595

E-mail: elizabeth.zellner@wmchealth.org

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