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CASE REPORT
Hand/Peripheral Nerve

Surgical Management of Hand Deformity in Epidermolysis Bullosa: Initial Experience and Technique

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Summary: Epidermolysis bullosa describes a rare group of genetic mucocutaneous disorders characterized by excessive epithelial fragility resulting in mechanically induced blistering and abnormal wound healing.[1,2] Its prevalence and incidence are 8.2 and 19.6 per 1,000,000 live births, respectively.[2] Gene therapy, protein replacement, and cell therapy strategies have been investigated, but there is currently no cure.[2] We describe a surgical technique to address the metacarpophalangeal joint flexion contracture and mitten deformity associated with EB. *(Plast Reconstr Surg Glob Open 2020;8:e2666; doi: 10.1097/GOX.0000000000002666; Published online 11 March 2020.)*

Clinically, epidermolysis bullosa (EB) is marked by bullae and secondary lesions including ulcerations and contractile scars that form in response to minor trauma.[1] Exposed areas (hands, feet and the oral cavity) are commonly affected, although generalized blistering can occur. Affected hands display flexion contractures at the metacarpophalangeal (MCP) and interphalangeal (IP) joints, as well as pseudosyndactly of the digits. Functionally, grasp is limited to weak approximation of a shortened thumb to the radial aspect of a shortened, flexed index finger. This results in significant physical disability and reduced quality of life. Management requires a multidisciplinary approach with surgery playing a critical role (Fig. 1).

We present the use of metacarpophalangeal flexion crease release and reconstruction with full-thickness skin graft (FTSG) for flexion contracture and mitten deformity associated with EB. Surgical goals include improved grasp and subjective increased range of motion at the MCP joints.

TECHNIQUE

Under general anesthesia, patients were draped in standard fashion with precautions to avoid skin and mucosal trauma. No tourniquets were used. A transverse volar incision at the MCP flexion crease is made through the skin. Dissection of the subdermal soft tissues is completed sharply with a scalpel employing a pushing motion rather than cutting to release scar bands. The MCP joints are then manipulated into full extension (to neutral, 0-degree position) and the incision further opened with scissor dissection. The proximal IP joints are only manipulated as needed to facilitate exposure of the palm. Using a paper template of the palmar defect, an FTSG is harvested from unblistered skin of the patient's groin and secured into place with interrupted absorbable sutures (Fig. 2).

With the digits in extension, the grafted site is dressed with a bolster of xeroform, gauze, and Coban Self-Adherent Wrap (3M). The donor site is closed primarily with absorbable suture and covered with xeroform and dry gauze, secured with minimal Mepitac Soft Silicone Tape (Mölnlycke, Health Care, UK). Dressings remain in place for 1 week and are removed in clinic. Patients are allowed to begin hand therapy at 3 weeks post surgery (Fig. 3).

RESULTS

This procedure has been performed on 4 hands on 2 patients. The average graft size in this series was $10.8 \times 4\,cm^2$. Both patients demonstrated excellent graft take and no evidence of neurovascular deficit or joint subluxation. At 1 month postoperatively, patients could extend at the MCP joints and form a fist with noticeable improvement. At 6 months, all 4 grafts remained soft, and patients could extend at the MCP joint (approaching neutral from 80-degree flexion, Fig. 4) and perform pincer grip to hold objects. No hypertrophic scarring occurred in graft or donor sites. Both patients were satisfied with outcomes (Fig. 4).

DISCUSSION

Morbidity in EB results from compromised structural and functional integrity of the skin on a cellular level. Deleterious mutations in at least 18 genes

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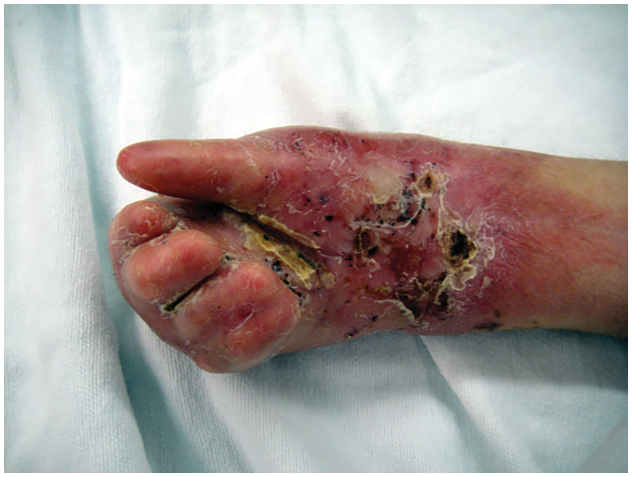

Fig. 1. This figure demonstrates the typical appearance of a hand with changes associated with EB, that is "mitten deformity."

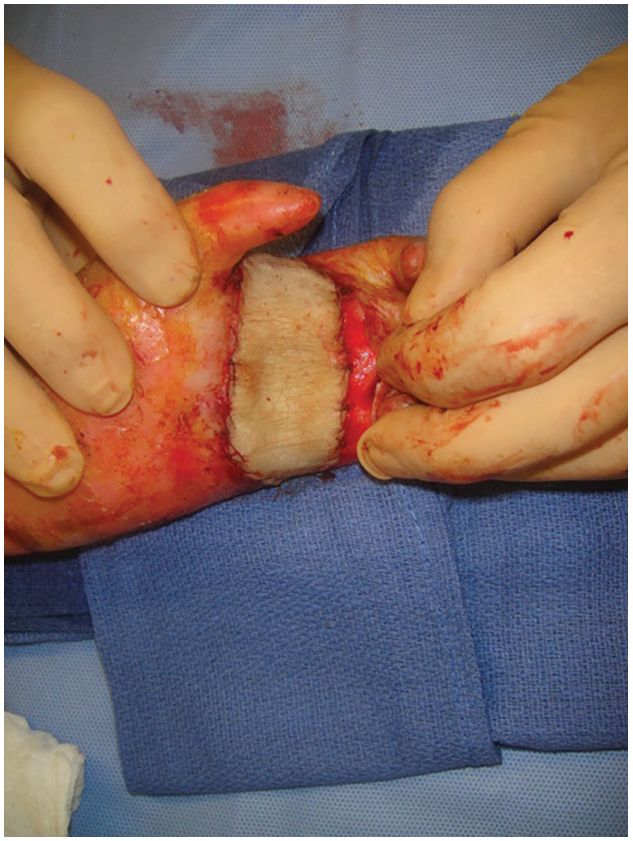

Fig. 2. This figure demonstrates a full thickness skin graft in place over MP flexion crease after scar release.

encoding keratin filaments in the skin and mucous membranes have been implicated.[1,2] These alterations impair epidermal anchoring, predisposing patients to tissue dehiscence.

Although 30 subtypes are described, there are 4 main clinical types of EB: simplex, junctional, dystrophic, and Kindler syndrome.[1] Diagnosis is confirmed by histology, with immunofluorescence mapping and mutational analyses.[2] Affected genes are expressed in epithelialized urogenital and mesenchymal organs, resulting in extracutaneous involvement. Thus, EB comprises a multisystem disorder with significant morbidity.

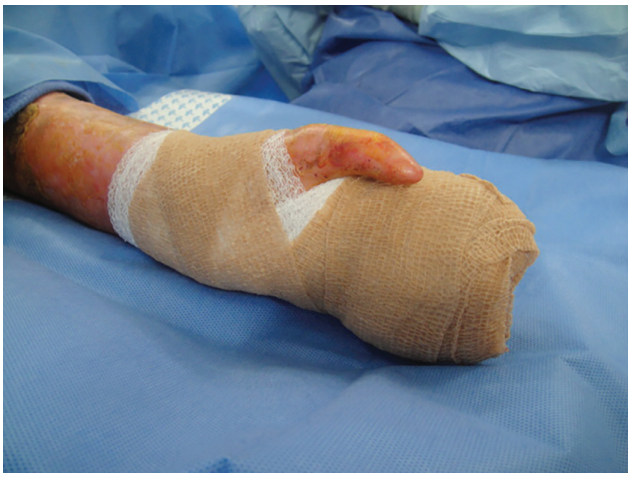

Fig. 3. This figure demonstrates a Coban dressing in place at the completion of the operation.

Approximately 61% of recessive dystrophic EB patients require hand surgery, typically needing 5 or more operations.[3] Challenges include risk of iatrogenic trauma, impaired wound healing, and frequent contracture recurrence. Recurrence of contractures and deformity is common within 2 years of intervention.[4] However, surgery to correct severe deformity facilitates patients' independence and can improve quality of life. In 1995, Ciccarelli et al[5] described precise indications for surgery: palmar contracture, contracture of the proximal IP joint >30 degrees, significant SF deformity, pseudosyndactyly extending to the proximal IP joint, or impairment of activities of daily living. Most surgeons operate when deformity prohibits independence and is intolerable to the patient.

Various methods for wound management and coverage following contracture release are described, yet no consensus exists. Rapid wound healing with long-lasting results that allow early hand therapy is ideal. While healing by secondary intention is described with acceptable recovery of limb function,[6] larger wounds require coverage to avoid infection, prolonged healing, and undue fibrosis and contracture. In these cases, use of skin grafts and allogeneic dermal substitutes, such as Apligraf[7] or Biobrane,[4] has been described. Split-thickness skin grafts, however, can create difficult-to-heal donor site wounds secondary to the lack of primary closure, and can be associated with iatrogenic trauma to surrounding skin or within the graft itself from the dermatome. In a case series of 5 patients treated with Biobrane gloves following contracture release, wound healing was longer than we experienced (at least 4 weeks to completion), and the first dressing changes uniformly required general anesthesia.

We argue that FTSGs are viable alternatives and report functional improvement with this technique. While FTSGs

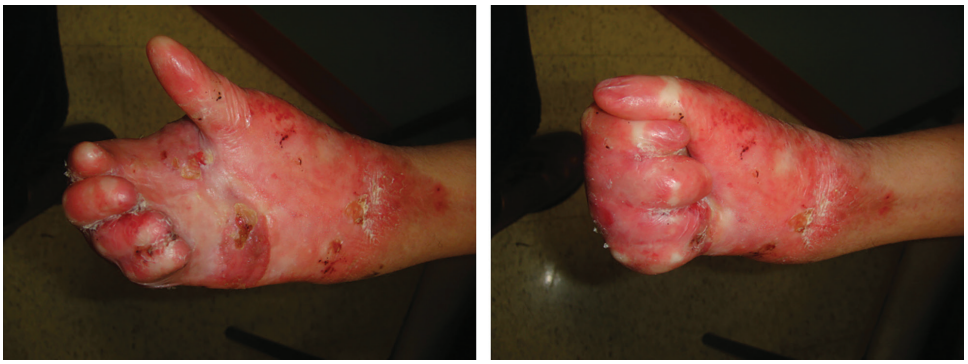

Fig. 4. This figure demonstrates the typical postoperative appearance after skin graft has healed, shown in full flexion and extension. This patient was seen 6 weeks postsurgery.

necessitate a donor site, definitive closure can be accomplished, reducing healing time. FTSGs also display less secondary contracture than split-thickness skin graft, theoretically reducing local soft tissue contracture.[8] Our technique yielded no donor site complications and enabled rapid enrollment in hand therapy by 3 weeks postsurgery. All dressing changes were performed without the need for sedation or the OR, in contrast to previously reported techniques.[9]

This study is limited by a small population size and a short follow-up period of 23 months. Further study is needed to evaluate long-term outcomes in more patients. Indeed, plastic surgeons should recognize the significant role they can play in addressing EB-related hand morbidity and seek to better understand available surgical techniques.

SUMMARY

EB comprises a rare group of genetic mucocutaneous disorders characterized by blister formation and mitten hand deformity. We describe a surgical technique for MCP flexion crease release and reconstruction with FTSG to address the flexion contracture and mitten deformity associated with EB. Early results for 4 hands show improved grasp and increased range of motion at the MCP joints. Plastic surgeons should recognize the significant role they can play in addressing EB-related hand morbidity and seek to better understand available surgical techniques.

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REFERENCES

1. Hsu CK, Wang SP, Lee JYY, et al. Treatment of hereditary epidermolysis bullosa: Updates and future prospects. *Am J Clin Dermatol.* 2014;15:1–6.
2. Laimer M, Prodinger C, Bauer JW. Hereditary epidermolysis bullosa. *J Dtsch Dermatol Ges.* 2015;13:1125–1133.
3. Fine JD, Johnson L, Weiner M, et al. Pseudosyndactyly and musculoskeletal contractures in inherited epidermolysis bullosa: experience of the National Epidermolysis Bullosa Registry, 1986–2002. *J Hand Surg Br.*2005;30:14–22.
4. Jutkiewicz J, Noszczyk BH, Wrobel M. The use of biobrane for hand surgery in epidermolysis bullosa. *J Plast Reconstr Aesthet Surg.* 2010;63:1305–1311.
5. Ciccarelli AO, Rothaus KO, Carter DM, et al. Plastic and reconstructive surgery in epidermolysis bullosa: clinical experience with 110 procedures in 25 patients. *Ann Plast Surg.* 1995;35:254–261.
6. Tian F, Li B, Tian LJ. Treatment of severe hand deformities caused by epidermolysis bullosa. *Orthopedics.* 2011;34:e780–783.
7. Fivenson DP, Scherschun L, Cohen LV. Apligraf in the treatment of severe mitten deformity associated with recessive dystrophic epidermolysis bullosa. *Plast Reconstr Surg.* 2003;112:584–588.
8. Berezovsky AB, Pagkalos VA, Silberstein E, et al. Primary contraction of skin grafts: a porcine preliminary study. *Plast Aesthet Res.* 2015;2:22–26.
9. Ladd AL, Kibele A, Gibbons S. Surgical treatment and postoperative splinting of recessive dystrophic epidermolysis bullosa. *J Hand Surg Am.* 1996;21:888–897.