SKIN

BRIEF ARTICLE

Accelerated Wound Closure in Neonatal Bart Syndrome with Negative Pressure Therapy

Simran K. Chandawarkar, BS[1], Helen A. Hannan[2], Ibrahim Amjad, MD[3]

[1] Northeast Ohio Medical University, Rootstown, Ohio, USA
[2] University of Michigan, Ann Arbor, Michigan, USA
[3] AmjadPlastics, Miami, Florida, USA

ABSTRACT

Bart Syndrome is a rare mechanobullous disorder and a subtype of epidermolysis bullosa (EB) that appears at birth with congenital localized absence of skin, absence or deformities of nails, and blistering of mucous membranes. Standard treatment reveals a variety of topical medical treatments with an average epithelization time of 3 months. Here, two newborn patients are presented with diagnosis of Bart Syndrome with significant EB and cutis aplasia on bilateral lower extremities. Both patients were successfully treated with Negative Pressure Wound Therapy (NPWT) and epithelization was achieved in only three weeks. This novel treatment underscores the importance of quick recovery time sin vulnerable neonatal populations and opens the door for further research to optimize NPWT for treatment of EB/Bart Syndrome.

INTRODUCTION

Bart Syndrome is a rare mechanobullous disorder and a subtype of epidermolysis bullosa (EB) that appears at birth.[1] It is an autosomal-recessive disorder affecting type VII collagen.[2] It is characterized by any combination of the following three features: congenital localized absence of skin, absence or deformities of nails, and blistering of mucous membranes.[3] Diagnosis is typically clinical, though biopsy or genetic testing may be used.[4,5,6] As Bart Syndrome remains a rare diagnosis, no standardized treatment has been developed. Various dressings, topical antibiotics, and hydro-surgical debridement have been used with limited success. A limited number of cases have revealed treatment with an assortment of topical dressings, antibiotics such as fusidic acid ointment and mupirocin ointment, and hydro-surgical debridement. Typical closure takes three months, risking infection and further breakdown. This gives rise to a multitude of complications from open wounds, further skin breakdown, and infection, especially in the neonatal population. Additionally, delayed healing times prolong NICU stays for many patients and increase the need for fluid replacement and IV therapy. Here we describe two neonatal cases of Bart Syndrome managed with a novel treatment: the use of negative pressure wound therapy (NPWT) to accelerate wound closure.

CASE PRESENTATIONS

Patient One

Patient One is a newborn white female who was noted to have skin changes consistent with epidermolysis bullosa and loss of skin of the bilateral lower extremities from the knees to the toes at birth. Significant dermal and epidermal loss noted by 12 hours (**Figure 1**). At this time, a NPWT dressing was placed at 75mmHg with a contact layer of Mepitel® that were changed three times a week. Healing began by day 2 with granulation tissue. Significant epithelization was noted at two weeks, and the contact layer was shifted to a 3M™ Adaptic™ dressing. By week three, full epithelialization was noted (**Figure 2**). She progressed with dressings of Mepitel®, Mepitel® transfer, and Kling.

Patient Two

Patient Two is a newborn black female who presented to a local community hospital with cutis aplasia of the bilateral lower extremities in a pattern very similar to Patient One (**Figure 3**). A pathology report and genetic testing confirmed the diagnosis of EB with a pathologic deletion in the ITGB4 gene on chromosome 17, encoding for beta-4 integrin, an integral part of the hemidesmosomes in epidermal basement membranes. On day-one of life, a NPWT

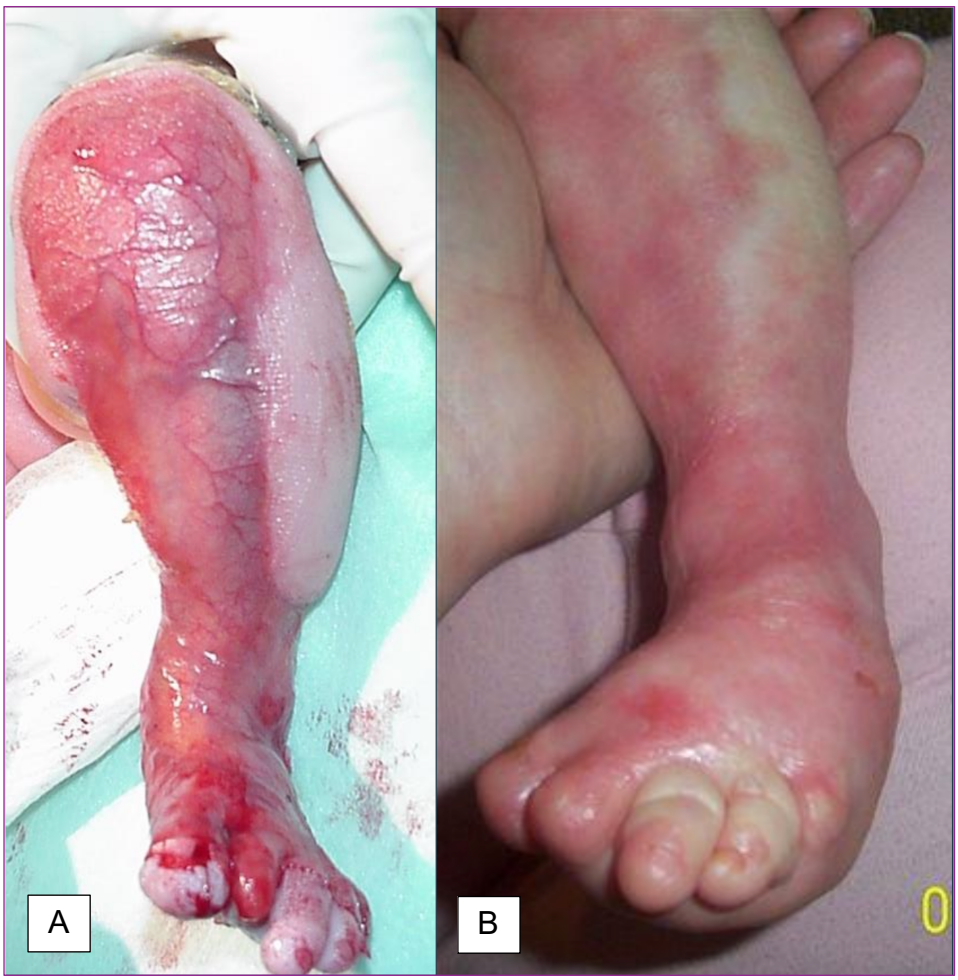

Figure 1. (A) A photo of Patient One's lower extremity presentation at birth with cutis aplasia. (B) A photo of Patient One after 3 weeks of treatment using NPWT with full epithelization of the lower extremity.

dressing was placed with Mepitel® as the contact layer and black foam at 125 mmHg, after which, granulation tissue developed. Significant epithelization was noted by 2.5 weeks, after which the dressing was changed to 3M™ Adaptic™, and NPWT was continued. After an additional week, full closure was achieved.

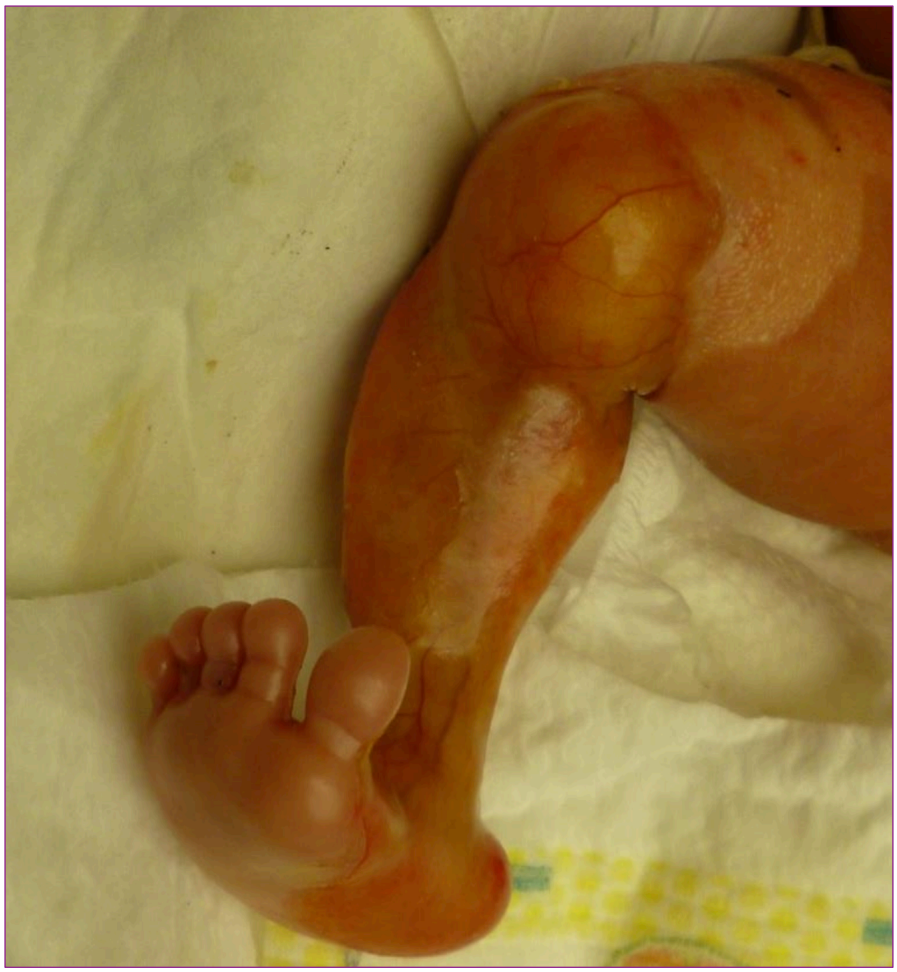

Figure 3. A photo of Patient Two's lower extremity presentation at birth with cutis aplasia.

DISCUSSION

Ideal treatment of Bart Syndrome focuses on the closure of wounds, prevention of infection, and reduction of complications. Conservative care requires 5–7 weeks, increasing infection risk.[7,8] Additionally, grafts like Apligraf® carry significant rejection risks. NPWT applies vacuum pressure to remove fluid and promote healing. Further, by promoting neo-angiogenesis, it initiates the formation of granulation tissue. The contact layer of Mepitel® was imperative for the protection of this granulation tissue. Rapid healing is especially valuable in neonates; Given their immune vulnerability, a speedy recovery is key to their ability to thrive. NPWT decreases the complications not only through reduced infection risk but also reduces the chance of further skin breakdown in the wound. Further, the decrease in dressing changes reduces the need for excessive sedation and pain management. Using NPWT only three times weekly achieved very promising results, which opens the door to

further research on optimizing this treatment to further promote healing.

CONCLUSION

EB/cutis aplasia associated with Bart Syndrome can be treated more effectively and efficiently with NPWT than with alternative current treatment regimens. Closure of these large open wounds of the bilateral lower extremities in a rapid fashion decreases the need for hospitalization and improves morbidity. NPWT is a viable, cost-effective treatment for EB in Bart Syndrome.

Conflict of Interest Disclosures: None

Funding: None

Corresponding Author:
Simran Chandawarkar
4209 State Route 44
Rootstown, OH, 44272, USA
Email: schandawarkar@neomed.edu

References:

1. Zelickson BH, Kist DJ, Matsumura K, Epstein EH, Bart BJ. Bart syndrome: ultrastructure and genetic linkage. *Arch Dermatol*. 1995;131(6):663-668. doi:10.1001/archderm.1995.01690180037006
2. Christiano AM, Bart BJ, Epstein EH Jr, Uitto J. Genetic basis of Bart syndrome: a glycine substitution mutation in type VII collagen gene. *J Invest Dermatol*. 1996;106(4):778-780. doi:10.1111/1523-1747.ep12346304. Corrected and republished in: *J Invest Dermatol*. 1996;106(6):1340-1342. doi:10.1111/1523-1747.ep12349293
3. Bart BJ, Gorlin RJ, Anderson VE, Lynch FW. Congenital localized absence of skin and associated abnormalities resembling epidermolysis bullosa: a new syndrome. *Arch Dermatol*. 1966;93(3):296-304. doi:10.1001/archderm.1966.01600210032005
4. Sharma H, Varma K. Case report of dominant dystrophic epidermolysis bullosa in siblings: a rare genetic disorder with a therapeutic challenge. *J Evol Med Dent Sci*. 2014;3(32):8856-8859. doi:10.14260/jemds/2014/3134
5. Falabella AF, Schachner LA, Valencia IC, Eaglstein WH. The use of tissue-engineered skin (Apligraf®) to treat a newborn with epidermolysis bullosa. *Arch Dermatol*. 1999;135(10):1219-1222. doi:10.1001/archderm.135.10.1219
6. Ozkaya O, Oreroğlu AR, Akan M. The use of acellular dermal matrix in treatment of mitten hand in epidermolysis bullosa patients. *J Hand Microsurg*. 2013;5(1):46-47. doi:10.1007/s12593-013-0090-6
7. Duran-McKinster C, Rivera-Franco A, Tamayo L, de la Luz Orozco-Covarrubias M, Ruiz-Maldonado R. Bart syndrome: the congenital localized absence of skin may follow the lines of Blaschko. Report of six cases. *Pediatr Dermatol*. 2000;17(3):179-182. doi:10.1046/j.1525-1470.2000.01747.x
8. Kim DY, Lim HS, Lim SY. Bart syndrome. *Arch Plast Surg*. 2015;42(2):243-245. doi:10.5999/aps.2015.42.2.243