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Evaluation of Non-Coverage Cost Effective Technique for Management of Epidermolysis Bullosa Mitten Hands in Developing Countries

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ABSTRACT

Background: Epidermolysis bullosa (EB) is a group of rare genetic disorders known by susceptibility of the skin epidermis and mucosa that separate from underlying tissues and cause increase in skin fragility, blistering, and scarring in response to minor traumatic injuries. Many methods were described in the literatures for soft tissue coverage after the release of mitten hands which include cultured keratinocytes; using epithelium raised from cocoon, to split skin grafts or full-thickness skin grafts; and cellular allograft dermal matrix.

In this study, we described a simple and efficient non-coverage technique, as it eliminates the donor site complications when using STSG or FTSG, and also minimizes cost management by avoid using cellular allograft dermal matrix, specially in developing countries, where cost effectiveness due to financial issues should be considered.

Aim of this Study: Is to describe and evaluate the non-coverage technique based on the Birmingham Epidermolysis Bullosa Severity Score Sheet (BEBSS) in patients suffering from epidermolysis bullosa mitten hands.

Patients and Methods: We operated upon nine patients suffering from epidermolysis bullosa mitten hands by using the non-coverage technique to obtain full release of all fingers and web spaces. Evaluation and scoring of preoperative and postoperative hand scars were based on the 'Birmingham Epidermolysis Bullosa Severity Score Sheet.

Results: Showed major improvement in the mean of post-operative BEBSS with lower rate of recurrence and postoperative complications. Hand function was remarkably improved within 2 months which was demonstrated subjectively by the capability of grasping and pinching objects such as cups, spoons and pens using unilateral hand.

All patients and care givers were satisfied with the improvement in function and the ability to perform more tasks than preoperatively.

Conclusion: Managing soft tissue defects after release of EB mitten hands can be left with no coverage and with appropriate dressing for self-re-epithelialization can be ideal method especially in developing countries where allografts are relatively expensive and not available.

Key Words: *Non-Coverage – Cost – Epidermolysis Bullosa – Developing Countries.*

INTRODUCTION

Epidermolysis bullosa (EB) is a group of rare genetic disorders; the overall incidence and prevalence of this disease are approximately 1:53,000 live births and 1:125,000 respectively. It is known by susceptibility of the skin epidermis and mucosa that separate from underlying tissues and cause increase in skin fragility, blistering, and scarring in response to minor traumatic injuries **[1]**. Clinical presentations of these patients and severity of the disease show variability depending on multiple factors. Based on the location of the fragility and the level of skin cleavage, EB is classified into 4 major types: EB simplex, junctional EB, dystrophic EB and Kindler syndrome. Epidermolysis bullosa is categorized into two main subtypes: Autosomal dominant EB (DEB) and recessive EB (REB) **[2]**.

REB individuals present with clinical picture at birth, including generalized blisters, ulceration of the mucous membrane, atrophic scars yet the exact mechanism is still unknown. Symptoms gradually deteriorate with disease progresses and it causes chronic blood loss, recurrent infections and malnutrition, which can result in anemia, late puberty, and osteoporosis. REB patients who survive past childhood frequently develop metastatic squamous cell carcinoma (SCC), which often leads to death **[3,4]**.

Hands of REB patients are susceptible to repeated cycles of blistering, ulceration, and healing, which may cause chronic scarring. This can also provoke series of inflammation which promote the development of chronic scars and tissue fibrosis. Severe fibrosis can induce finger contractures, progressive digital fusion (pseudo-syndactyly),

flexion contractures and adduction contractures in the thumbs. In some cases, sever mitten like deformities can cause severe dysfunction that leads to loss of the ability to perform even the basic daily tasks. These hand deformities are known as "cocoon hands" **[5,6]**.

Many methods were described in the literatures for soft tissue coverage after the release of mitten hands which include cultured keratinocytes; using epithelium raised from cocoon, to split skin grafts or full-thickness skin grafts; and cellular allograft dermal matrix **[7]**.

In this study, we described a simple and efficient non-coverage technique, as it eliminates the donor site complications when using STSG or FTSG, and also minimizes cost management by avoid using cellular allograft dermal matrix, specially in developing countries, where cost effectiveness due to financial issues should be considered.

Aim of this study is to describe and evaluate the non-coverage technique based on the 'Birmingham Epidermolysis Bullosa Severity Score Sheet (BEBSS) in patients suffering from epidermolysis bullosa mitten hands.

PATIENTS AND METHODS

We operated upon nine patients during the period of March 2017 till October 2019. Patients underwent full laboratory investigations as well as radiological evaluation. Most patients had low hemoglobin level which necessitated admission to hospital two days prior to surgery for blood transfusion. The surgical plan and follow-up were thoroughly explained to the patients and care givers. Only one hand was operated at a time. Evaluation and scoring of preoperative and postoperative hand scars were based on the 'Birmingham Epidermolysis Bullosa Severity Score Sheet' According to BEBSS, six grades indicating different degree of hand malfunction are included as follows: Birmingham Epidermolysis Bullosa Severity Score (BEBSS): 0=No scarring, 1=Milia and/or atrophic scars, 2=Just detectable contractures or webbing, 3=Obvious contractures or proximal webbing, 4=Between 3 and 5, and 5=Mitten formation with fingers all fused **[8]**.

Also, subjective evaluation was done postoperatively for patients and families for satisfaction with the results of treatment.

Anesthesia:

General anesthesia was used in all patients using laryngeal mask which was fixed by a soft ribbon tied around the back of the skull with cotton padding between it and the skin. Care was taken while transferring the patient to the operating table by lifting the patient gently to avoid any friction to the skin.

Surgical details:

Surgery was performed under pneumatic tourniquet which was padded by layers of cotton more than normally used. Skin was prepared using betadine solution applied gently to the skin with no frictional scrubbing. A suture was placed at the tip of each finger for retraction. A trial of preserving the cocoon surrounding the fingers and hand was always done, although not always successful. An incision was made to separate the fingers, which was followed by using dissecting scissors to open the plane which usually existed between fingers. This was followed by transverse incision to release any flexion contracture and severing any fibrous tissue. During dissection, soft tissue covering over tendons should be preserved to avoid tendons exposure. Gentle extension of the fingers was usually enough to reach full extension with no vital structures exposed. A K-wire was used to maintain the extension reached by the release.

The first web was released through an incision from the dorsum of the web to the thenar muscles. The fascia of the first dorsal interosseous and adductor muscles was released and sometimes release of the attachment of the first dorsal interosseous muscle was needed. In two patients a transverse wire was placed between the first and second metacarpals.

Tourniquet was released to ensure that full extension was not affecting the vascularity of fingers.

Dressing:

Vaseline gauze and antibiotic ointment were used. Proper separation of fingers and a bulky dressing was placed in the first web space. This was followed by bulky overall dressing with no splint used.

Postoperative:

At the beginning dressing was done every third day under general anesthesia (GA) using a face mask gently held to the face. As more patients were operated upon dressing was done every week under GA for three to four weeks depending on the condition of the wounds. The K-wires were removed after three to four weeks and replaced by a well-padded fabricated hand splint. At this time, the physical therapy was started under the supervision of a specialized hand therapist.

Fig. (1): Showing intraoperative full release with non-coverage technique.

Fig. (2): Postoperative bulky dressing without use of splint.

RESULTS

This study included 9 patients with mitten hands, 5 males and 4 females with mean age 16.4±10.2 years old (range 4-33 years).

Mean of preoperative BEBSS score was 4.67± 0.5 (range 4-5). Patients were operated with the previously described technique.

Mean of postoperative BEBSS score improved to be 0.78±0.97 (range 0-3) with average follow-up period 7.4±3.7 months (range 3-12 months).

Mean of postoperative healing time was 25.67± 3.7 days (range 21-33 days) with no complications seen as there were no infections, no signs of de-vascularization following fingers release and fixation by K-wires. Mean of postoperative follow-up period 7.4±3.7 months (range 3-12 months). The higher recurrence of contractures was mainly noticed in the first web space.

Table (1): Showed patients' characteristics, results, pre- and post-operative BEBSS scores.

Pt. No.	Age	Gender	Hand	Preop. BEBSS score	F.U	Postop. BEBSS score	Healing
1	29	F	Dominant	5	12	1	23
2	33	M	Dominant	5	12	1	25
3	15	F	Dominant	4	6	0	24
4	24	F	Dominant	5	7	0	33
5	4	M	Dominant	4	9	0	28
6	16	M	Dominant	5	3	1	21
7	5	F	Dominant	4	11	0	22
8	12	M	Non-Dominant	5	4	3	28
9	10	M	Dominant	5	3	1	27

Hand function was remarkably improved within 2 months which was demonstrated subjectively by the capability of grasping and pinching objects such as cups, spoons and pens using unilateral hand.

After surgical management, the active range of motion (AROM) of MP joint was between 0 and 60 degrees. AROM of proximal IP joint reached 80 degrees and distal IP joints was between 0 and 15 degrees. Each digit could perform adduction and outreach in coordination. The restored thumb achieved much increased flexibility and opposition.

All patients and care givers were satisfied with the improvement in function and the ability to perform more tasks than preoperatively.

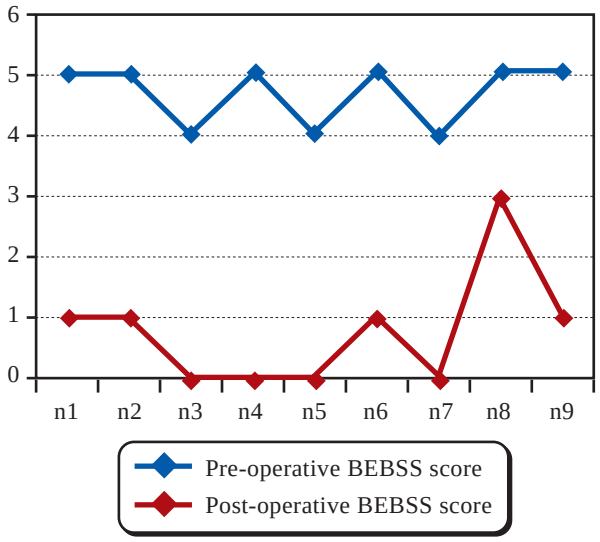

Fig. (3): Pre-operative and Post-operative BEBSS Scores.

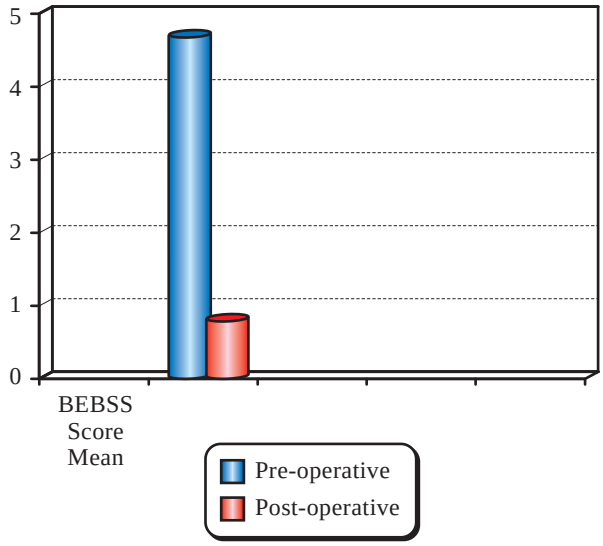

Fig. (4) Means of Pre-operative and Post-operative BEBSS Score.

Case (1)

Fig. (5): Preoperative EB Mitten Hand.

Fig. (6): Nine months postoperatively.

Fig. (7): Patient held a pen and wrote her name nine months postoperatively.

Case (2)

Fig. (8): Preoperative EB Mitten hand.

Fig. (9): Twelve months postoperatively.

Fig. (10): Thumb opposition twelve months postoperatively.

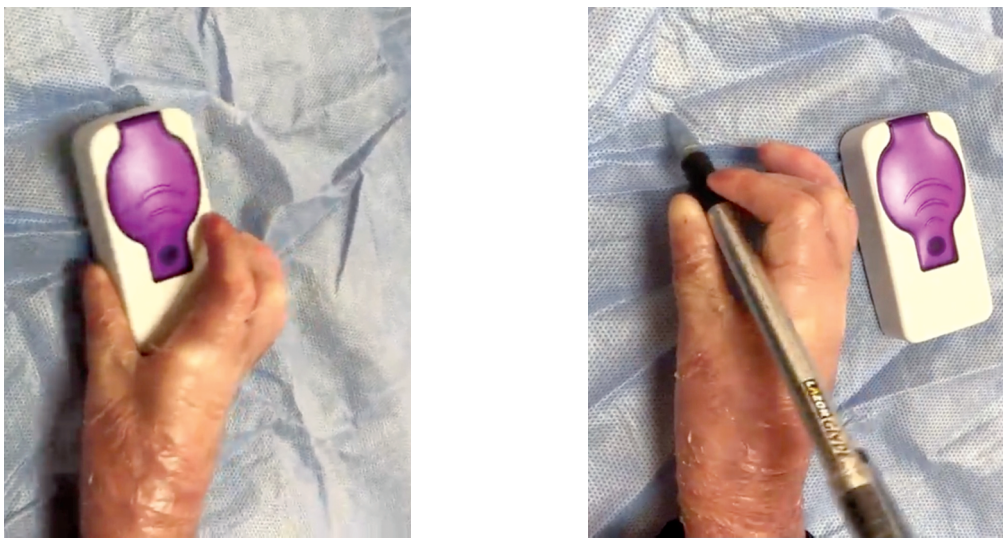

Fig. (11): Holding objects and writing twelve months postoperatively.

Case (3)

Fig. (12): Preoperative EB hand (Contracture of MPs and 1st web space).

Fig. (13): EB hand 1 month post-operatively (MPs release and fixation by K wires).

DISCUSSION

Treatment of mitten hands of epidermolysis bullosa patients is a challenge in the practice of hand surgery. In developing countries, it is even more challenging with the lack of the appropriate equipment, supplies and finances; while in developed countries they can withstand the cost of cultured keratinocytes, cellular allograft dermal matrix and advanced types of dressings.

In this study, we showed our experience in managing mitten hands using the non-coverage technique with a simple low-cost appropriate dressing and early rehabilitation. The non-coverage technique was used by other authors with similar results **[9]**. Surgical outcome, results and quality of life described by authors who used more sophisticated techniques for coverage of the defects were not far than our results **[10,11,12]**.

This makes it more logical in developing countries to use simpler techniques, especially with the limitations in resources.

The recurrence rate of this study whether in the medial four fingers, the thumb, or first web space is in comparable and described by most authors **[13]**. Yet this degree still gave the patients the ability to perform much more tasks than preoperatively.

Our study lacks long-term follow-up for better assessment of rate of contracture recurrence and a more objective assessment tools and can be improved if more sample size was used and this was because of the rarity of the disease, although we believe our series is among the biggest mentioned in the literature.

Conclusion:

Managing soft tissue defects after release of EB mitten hands can be left with no coverage and with appropriate dressing for self re–epithelialization can be ideal method especially in developing countries where allografts are relatively expensive and not available.

REFERENCES

1- Tian F., Li B. and Tian L.J.: Treatment of severe hand deformities caused by epidermolysis bullosa. Orthopedics, 2011.

2- Fine J.D., Bruckner-Tuderman L., Eady R.A., et al.: Inherited epidermolysis bullosa: Updated recommendations on diagnosis and classification. J. Am. Acad. Dermatol., 2014.

3- Luria S., Radwan S., Zinger G., et al.: Hand surgery for dystrophic epidermolysis bullosa. J. Pediatr. Orthop., 2014.

4- Pfendner E.G. and Lucky A.W.: Dystrophic epidermolysis bullosa. NCBI bookshelf. A service of the National Library of Medicine [updated 2018 Sep 13]. National Institutes of Health; Seattle (WA), 2006.

5- Eisenberg Llewelyn M.D.: Surgical management of hands in children with recessive dystrophic epidermolysis bullosa: Use of allogeneic composite cultured skin grafts. Br. J. Plast. Surg., 1998.

6- Fivenson D.P., Scherschun L. and Cohen L.V.: Apligraf in the treatment of severe mitten deformity associated with recessive dystrophic epidermolysis bullosa. Plast. Reconstr. Surg., 2003.

7- Bernardis C. and Box R.: Surgery of the hand in recessive dystrophic epidermolysis bullosa. Dermatologic Clinics. Apr., 2010.

8- Wong A., Ward R., Hadley N., et al.: A scoring system for epidermolysis bullosa. Br. J. Dermatol., 2007.

9- Ladd A.L., Kibele A. and Gibbons S.: Surgical treatment and postoperative splinting of recessive dystrophic epidermolysis bullosa. J. Hand Surg., 1996.

10- Eisenberg Llewelyn M.D.: Surgical management of hands in children with recessive dystrophic epidermolysis bullosa: Use of allogeneic composite cultured skin grafts. Br. J. Plast. Surg., 1998.

11- Jutkiewicz J., Noszczyk B.H. and Wrobel M.: The use of Biobrane for hand surgery in Epidermolysis bullosa. Journal of Plastic, Reconstructive & Aesthetic Surgery. Aug., 2010.

12- Özkaya Ö., Öreroğlu A.R. and Akan M.: The use of acellular dermal matrix in treatment of mitten hand in epidermolysis bullosa patients. Journal of Hand and Microsurgery. Jun., 2013.

13- Zhou X., Zhang Y., Zhao M., Jian Y., Huang J., Luo X., Yang J. and Sun D.: Surgical management of hand deformities in patients with recessive dystrophic epidermolysis bullosa. Journal of Plastic Surgery and Hand Surgery. Jan., 2020.