Epidermolysis bullosa is a genetically determined chronic disabling disease. The disease is classified in non-scarring or simple and scarring or dystrophic types. Various medical and surgical approaches are reported with no sustainable results. In this report, nine patients treated with split thickness skin grafting and flaps are reported. Due to recurrence of the contractures, a long term benefit is almost always impossible in these patients. However, early surgical interventions and physical therapy may be of help for the progression of the disease.

Keywords: Epidermolysis bullosa, graft, reconstruction

Case I
This 7 year old girl presented to the clinics with the complaints of hand deformity and recurring lesions on the legs. She was referred from a rural region where she had been treated with topical wound care and oral antibiotics on multiple occasions. On physical examination, she was found to have mitten hands and non-healing ulcers on the legs. She underwent contracture release and split thickness skin grafting under general anesthesia (Figure 1). Postoperatively, her hands were put on splint and physical therapy. However, she presented one year later with contracture in the flexion creases of the fingers.

Case II
This 14 year old boy presented with contractures in his hands. He underwent release of contractures and reconstruction with local flaps. Two years into his surgery, he presented with recurring contractures. He had a second surgery for release. He was put on physical therapy and splinting. He is currently complaining of contractures but refusing to undergo further surgery (Figure 2).

Case III
This 17 year old girl presented with severe contractures of his fingers (mitten hand) and toes with nonhealing wounds on arms and legs. She had release of contractures to some extent. However, it was not possible to obtain full extension due to contractures of the joints. She refused further treatment and lost to follow up (Figure 3).
Discussion

Scarring in dystrophic epidermolysis bullosa may result in disabling deformities particularly in the hands. Due to fragile skin and mucosa, these patients pose potential problems even during anesthesia. Treatment modalities of hand and foot deformities in epidermolysis bullosa are reported in various publications.8-10 Patients with epidermolysis bullosa particularly those with functional disabilities present with difficulties in skin care, surgical management and during anesthesia and postoperatively. It is reported that correction of advanced deformities by simply removing the epidermal cocoon is not sufficient for correction of deformities.1 The surgical treatment should be aimed for release of contractures over the web spaces and flexion creases with the use of split thickness skin grafts or skin flaps.

The relevant literature lacks a wide coverage of surgical treatment modalities in epidermolysis bullosa. It is noted that surgical interventions do not alter the course of disease. However, the corrections are not sustainable and the recurrences are inevitable. In our series, acute cases of epidermolysis bullosa have been treated as acute burn care. Particularly those with acute inflammation and epidermis loss, they have been managed with split thickness skin grafts or temporary skin dressings have been applied until epithelization occurred.

It seems apparent that acute cases of epidermolysis bullosa need to be approached like a burn wound to prevent bacterial infection and early occurrence of contractures. Those cases with contractures already present, conventional methods of reconstruction such as flaps and skin grafts need to be used. Postoperative splinting of the hands is also of utmost importance to prevent early recurrence of contractures. However,
long term outcomes are not always satisfactory. These patients should be aware of dismal outcomes of minute traumas. Therefore, early surgical treatment in childhood is mandatory to prevent advanced contractures of joints since there is no definitive curative surgical or medical treatment. These patients require close follow up and training for future problems. The role of reconstructive surgery in the management of epidermolysis bullosa should not be dismissed. Plastic Surgery must get involved in the early period of the disease progression.

Figure 2a, 2b: Severe contracture of the hand in a 17 year old boy.

Figure 3: Severe contractures of the web spaces in the feet of a 17 year old girl.

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