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Case Reports

FREE JEJUNAL GRAFT FOR TREATMENT OF ESOPHAGEAL STRICTURE DUE TO FAMILIAL EPIDERMOLYSIS BULLOSA

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ABSTRACT

In this paper, two patients with severe dysphagia and esophageal stricture secondary to epidermolysis bullosa are presented and discussed. They are siblings of an affected family. Primary resection and anastomosis had been performed previously in both patients, but dysphagia had recurred. We treated these patients with free jejunal graft to the esophagus with excellent results. Both patients are well and free of dysphagia after five years.

Keywords: Stricture, esophagus; epidermolysis bullosa; graft, jejunal, free.

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INTRODUCTION

Epidermolysis bullosa (EB) comprises a group of genetically-determined disorders characterized by blistering of the skin and mucosae, especially the mouth and esophagus. The blisters may either result from minor mechanical trauma or apparently arise spontaneously.[1]

Classification of this complex and heterogeneous group of syndromes is difficult, and is not helped by the large variety of names and eponyms applied to each syndrome. Early classification was largely based on clinical and genetic observations. While these observations are clearly important, the electron microscope has now demonstrated that the level of cleavage in the skin is different in the major groups of syndromes, namely simple, junctional and dystrophic epidermolysis bullosa. Aside from this, there are ultrastructural and biochemical abnormalities, especially in skin collagenase, which have helped to distinguish between the two main types of dystrophic EB. It seems logical, therefore, to modify the classification in the light of these data.[1]

In the current classification, the genetic forms of EB are:

1) Simple, in which there is intraepidermal splitting, and autosomal dominant or autosomal recessive inheritance,

2) Junctional, in which splitting occurs in the lamina lucida and is accompanied by skin atrophy, and inherited

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by an autosomal recessive gene,

3) Dystrophic, in which separation occurs in the sublamina densa, and is accompanied by scarring. It may be inherited by an autosomal dominant gene,[1] or by an autosomal recessive gene which has four subtypes:

a) generalized (Hallopeu-Siemens),

b) generalized non-mutilating,

c) localized, and

d) inverse.

Esophageal strictures due to epidermolysis bullosa can be a life-threatening problem and may be extremely difficult to treat effectively. In this report we present our experience with treatment of recurrent esophageal stricture and dysphagia in two siblings of a family.

CASE REPORTS

Case one

The patient is a 57 year old female, the third sibling of an eight-member family. The parents' marriage was weakly consanguinous. Two of her brothers had died in the first few months of life due to this disease. Her 47 year old brother is discussed in Case two.

Bullae of the skin and mucous membranes had been frequently present since birth. Dysphagia developed when she was six years old and had progressed insidiously to the extent that she was not able to consume solid foods, only liquids. She had surgery for the first time at 37 years of age, and the skin incision scar was present at the anterior border of the sternocleidomastoid muscle (Fig. 1). We do not have details of the operative procedure performed, but esophageal stricture and dysphagia persisted and the left recurrent laryngeal nerve had been injured. At 40 years of age, the stricture at the cricopharyngeal level was operated by Belsey in the U.K., and resection and primary anastomosis was performed. The patient was well for six years after this operation, but again dysphagia recurred and progressed. Upon presentation to us, she had severe weight loss and could barely consume clear fluids. Barium swallow revealed recurrence of severe stricture at the anastomotic line (Fig. 2).

Case two

This 47 year old male had symptoms and signs similar to his sister (Case one), only more severe and aggressive in nature. Severe dystrophy and scarring of fingernails and toenails was present (Figs. 4,5), and aside from the buccal mucosa, esophagus and skin, bullae were present in anoderm and urethral mucosa. Dysphagia had developed at six years

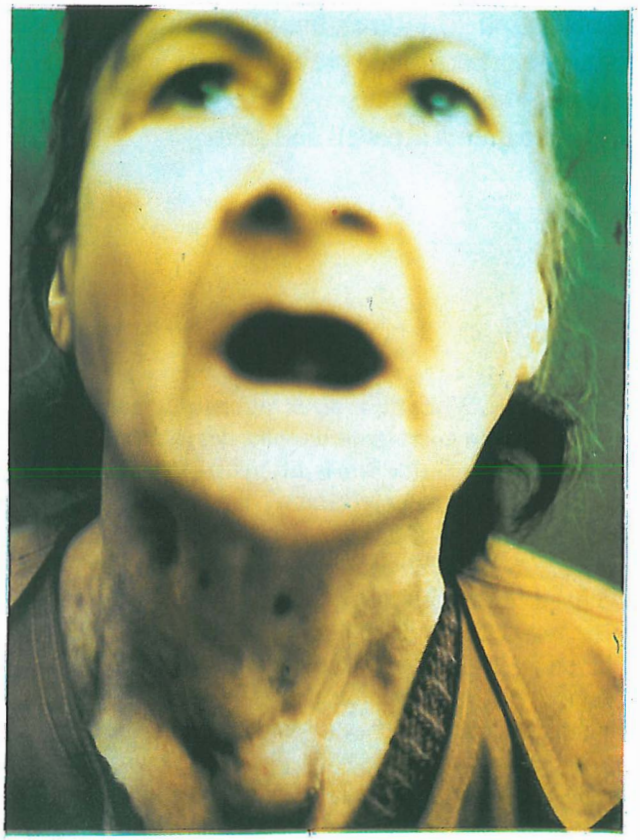

Fig. 1. Patient one after jejunostomy closure. Note limitation in degree of mouth opening.

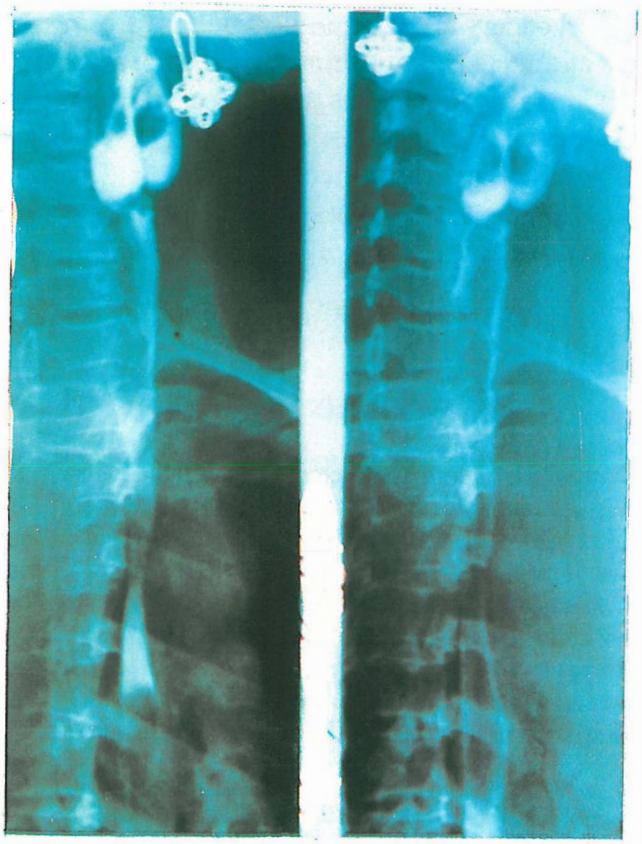

Fig. 2. Barium esophagram demonstrating stricture at cricopharyngeal level.

of age, and had become complete at 23 years of age at which time he could not swallow his saliva.

At 25 years of age, he was hospitalized in Israel and dilatation was attempted, but even the smallest size bougie could not be passed and tube gastrostomy was performed. Barium swallow disclosed a stricture at the cricopharyngeal level which moved with swallowing (Figs. 6,7). At that time, the stricture was resected and primary anastomosis performed by Belsey in the U.K. He also was well for six years after operation, but dysphagia recurred and progressed. Upon presentation to us, he could consume only clear fluids and was cachectic. He was treated by free jejunal graft to the esophagus and dysphagia was alleviated.

DISCUSSION

Epidermolysis bullosa involves the squamous epithelium of the entire body. The bullae may developed spontaneously or by even slight trauma (Nikolsky's sign). Corneal erosion, lacrimal duct stricture, ectropion, and laryngeal edema and bullae have been reported. Common problems encountered in patients include malnutrition, aspiration pneumonitis, and unconsciousness.

In one report, 16 patients were studied radiologically. Esophageal abnormalities occurred at sites of relative stasis (aortic knob, carina, gastroesophageal junction) and in areas subject to peptic esophagitis. Esophageal strictures have been reported in the upper third of the esophagus in 50% of patients, in the lower third in 25% and at multiple sites in 25% of cases. Eight (50%) of the 16 esophageal strictures occurred in the proximal third and five (31%) had multiple strictures. The strictures varied in length from 2 mm to 15 cm. Long strictures had tapered margins. Short segmental stenoses or webs were also seen. Less common radiologic findings included pseudodiverticulum formation, prestenotic dilatation, ulceration and spasm. Strictures sometimes resembled caustic or peptic strictures.[3]

In the report of Kern et al., seven patients from 4 to 14 years of age received medical treatment and dilatation.[4] Bullae and ulcers were treated effectively by corticosteroids, however they can also be treated by dilatation.[4] Verapamil is effective for relieving dysphagia due to esophageal spasm. Acute and chronic strictures in children can be relieved by dilatation.[4] Oral phenytoin is effective for prevention of bullae and ulcers. Its effect on skin lesions at a serum level of 10 μg/mL has been approved and is effective at the onset of esophageal symptoms, but it is not effective during dilatation. Repetitive dilatation may be necessary, however some of these patients have been followed for ten years without recurrence. Dilatation was done under general anesthesia and intubation, and the risk of perforation is generally low. Because it is difficult to

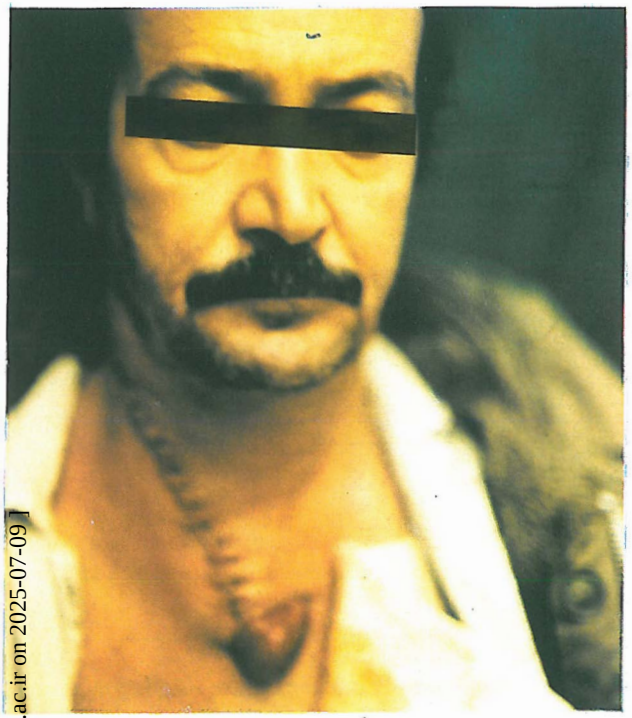

Fig. 3. Patient two after operation. Note jejunostomy at sternal notch.

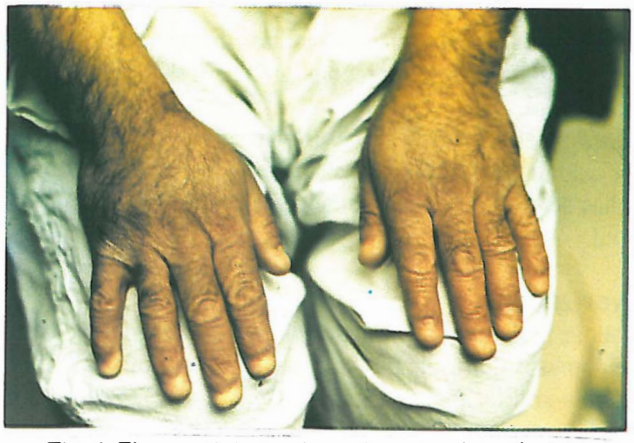

Fig. 4. Fingernail dystrophy and scarring in patient two.

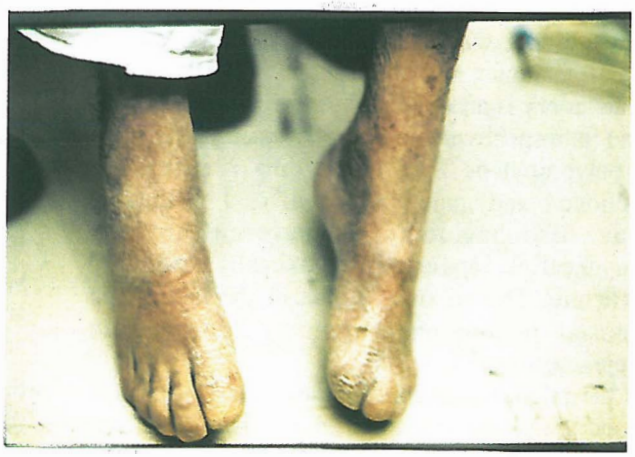

Fig. 5. Severe scarring and dystrophy of toenails in patient two.

distinguish between bullae, ulcers, spasm and organic stricture, management is therefore generally begun conservatively to reduce bullae formation, relieve symptoms and prevent aspiration.[4]

High dose corticosteroids (2.5 mg/kg prednisone or equivalent) relieve dysphagia within 48 hrs, and the dose is subsequently gradually tapered over the next two weeks. If this treatment fails, dilatation is begun and it is better to begin after remission of skin symptoms.[4]

Several surgical techniques have been proposed for the treatment of esophageal stricture due to epidermolysis bullosa. Primary resection and anastomosis may be applied for short strictures. Also, gastric pull-up, reversed gastric tube flap and pedicled flap of colon have been applied, but these are all major operations for these severely malnourished and immunosuppressed patients. Anastomotic leak and stricture recurrence have been reported. Further, if these procedures are used as a bypass rather than esophageal replacement, further esophageal study by barium swallow or esophagoscopy can not be performed.

In the new technique which we will describe, several advantages are noteworthy:

1) It is not as extensive a procedure as those described above,

2) It does not preclude esophageal studies after operation,

3) It is potentially useful for reconstruction of short and long strictures,

4) It does not eliminate the possibility for future gastric, jejunal or colonic flap or graft,

5) It has several technical advantages which will be pointed out.

Operative technique

An oblique incision is made at the anterior border of the sternocleidomastoid muscle on the right side (both of our patients had a similar incision scar on the left side). The middle thyroid vein and superior thyroid artery and vein are ligated and divided. The common carotid artery, internal jugular vein, trachea, esophagus and right recurrent laryngeal nerve are exposed. Laparotomy is then performed, and a 10 cm segment of jejunum with its artery and vein is harvested. The jejunal artery is anastomosed to the common carotid artery and its respective vein to the internal jugular vein with 7-0 polypropylene (Fig. 8). A feeding gastrostomy is then fashioned and maintained until oral feeding is underway. Esophagotomy is performed at the cricopharyngeal level and extended proximally and distally to the stricture. The proximal stoma of the jejunum is anastomosed to the esophagotomy (end-to-side esophagojejunostomy).

Since the stricture was limited to the cervical esophagus just beyond the pharynx and extending down to but not reaching the mediastinal esophagus, after opening the stricture longitudinally and extending the incision up into the normal pharynx and distally into the normal esophagus, the proximal end of the free jejunal loop was anastomosed to the side of the pharyngo-esophageal incision as depicted below. The width of the beveled jejunal loop end was long

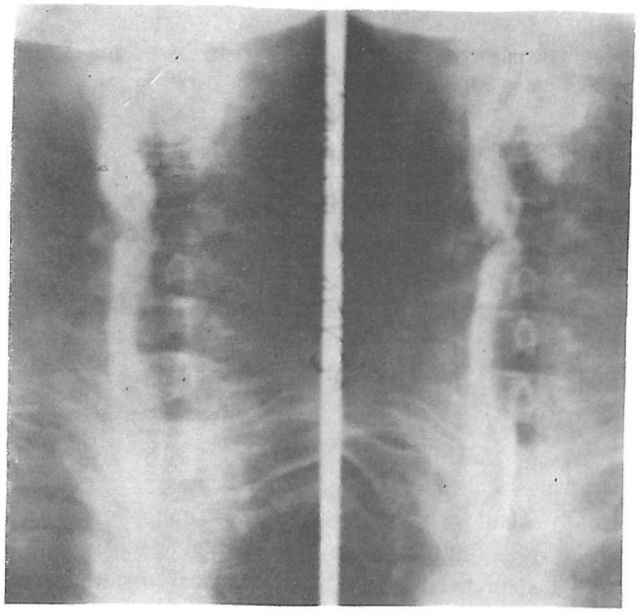

Fig. 6. Barium esophagram, anteroposterior view, demonstrating severe stricture in patient two. Note stricture at cricopharyngeal level (lateral view).

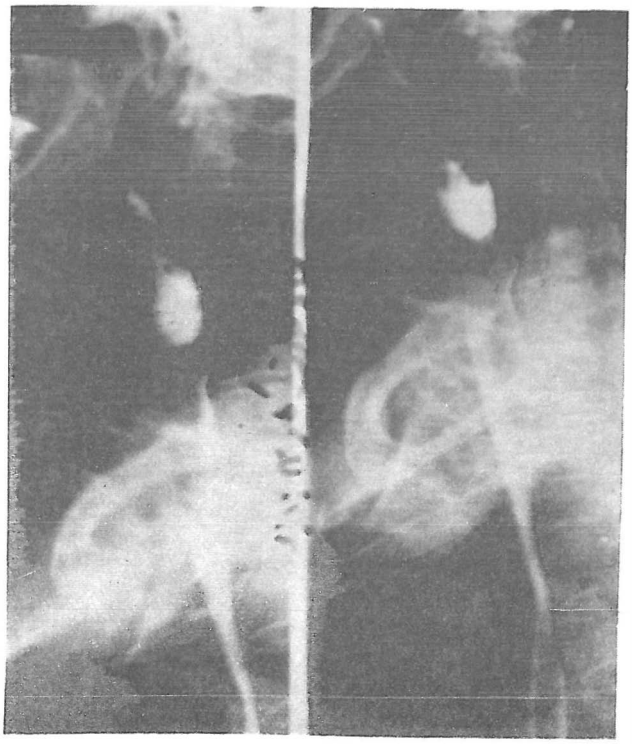

Fig. 7. Barium esophagram in patient two. Note stricture at cricopharyngeal level (lateral view),

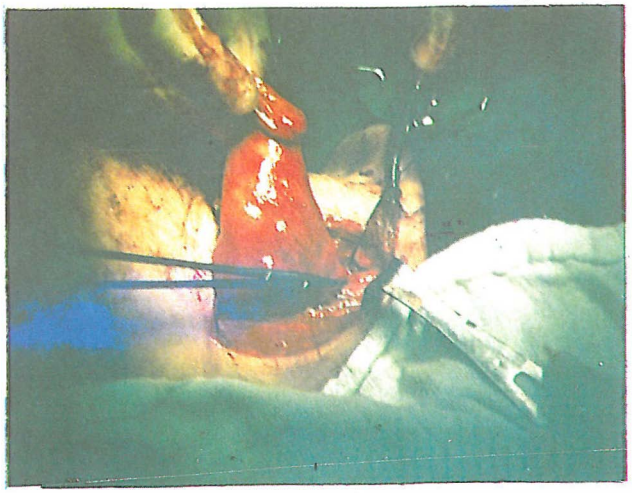

Fig. 8. Operative view demonstrating distal stoma (held) and vascular pedicle (tip of hemostat).

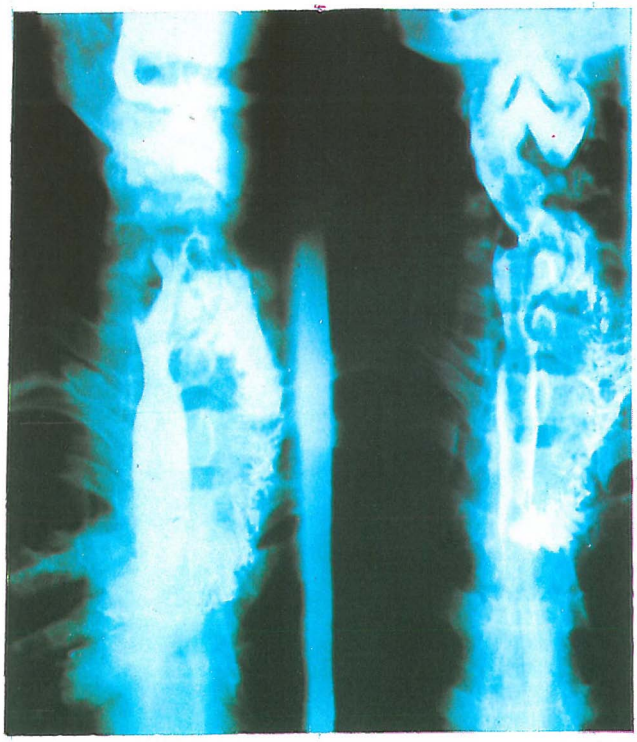

Fig. 9. Postoperative barium study in patient two, after jejunostomy closure. Note easy filling and emptying of jejunal loop.

enough to cross the stricture completely. Distal anastomosis of the loop was therefore unnecessary at this stage. We should mention that end-to-end anastomosis of free jejunal loop to the pharynx is unnecessary and extremely difficult as it involves more dissection and especially in these patients, minimal touch of mucosa by instruments would lead to a blister and separation of the mucosa from the underlying muscle layers (Nikolsky sign).

Since the remainder of the esophagus, albeit normal at this time, could develop a stricture in the future, we saved the rest of the jejunal loop and left it under the skin for future use if needed. The end of the loop was left open for drainage in the form of a jejunal stoma. After complete healing of the anastomosis and resuming oral feeding, this stoma was later closed and buried under the skin. Having part of the jejunal loop exposed in the form of a stoma would also help to evaluate its circulation and in case of a problem in blood supply, correction is possible before necrosis of the loop develops.

Jejuno-esophageal anastomosis was performed in one layer using modifed Gambee technique. We have made a modification in the technique of anastomosis which we call "parachute method". This technique is useful in low pelvic anastomosis and high jejunoesophageal anastomosis from the abdominal side also. All stitches are put in while keeping the ends apart from each other, and after all stitches are placed under direct vision with no guessing involved, by gently pulling on the sutures, the two ends come in proximity of each other and the sutures are then tied in order.

Nikolsky sign was evaluated by daily dressing of the stoma with sterile bandage, and was negative. The stoma was draining saliva freely, and thus the anastomotic site was free of tension or compression. After one week, barium study was performed, and no leak or abnormality was seen and regular oral diet was begun. Two weeks later, the jejunostomy was closed under local anesthesia, and after one week, another barium swallow study was done. There was no stricture at the anastomosic site, barium easily filled the jejunal graft, and the patch of jejunum readily emptied into the distal esophagus (Fig. 9).

At four years of follow up, there is no complaint and no signs of recurrence. We recommend this intermediate and flexible technique before resorting to more extensive procedures for these patients.

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