

Pyloric Atresia Associated with Epidermolysis Bullosa

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ABSTRACT

We present 5 cases of pyloric atresia associated with junctional epidermolysis bullosa, from 2003 to 2005. Patients underwent laparotomy after stabilization. Four neonates had gastroduodenostomy, and the other had excision of membrane and pyloroplasty. Four survived and one died from fulminant septicemia. Although the association of pyloric atresia with epidermolysis bullosa has been reported to be fatal, our study showed good survival rate.

Key words: *Epidermolysis bullosa, Pyloric atresia, Surgery, Survival.*

INTRODUCTION

Pyloric atresia (PA) is a rare malformation which is estimated to be responsible for less than 1% of gastrointestinal atresia with an incidence of 1 in 100,000 live births(1). Epidermal bullosa (EB) is a group of hereditary disorders of skin where the primary cause is the formation of blisters following minor trauma. It is categorized as EB simplex (EBS), junctional EB (JEB), and dystrophic EB (DEB). Evidence suggests that PA-EB association is a distinct entity, referred to as the PA-EB syndrome(2). Hitherto, the association of PA-EB was thought to be fatal because of septicemia, electrolyte imbalance and protein loss seen in EB. It was even recommended in the past that surgical treatment be withheld in these patients. We report good results with surgery in these children and better survival rate.

METHODS

This study was conducted in the pediatric surgical units of Boo-Ali hospital in Sari, Iran from 2003 to 2005. We had 5 patients with PA associated with JEB. Biopsy of bullae was done in all patients for

confirmation of diagnosis. All children were subjected to surgery. The details are shown in **Table I**.

RESULTS

There were 4 boys and one girl. Mean age of presentation was 8.4 days (3 to 15 days). There were multiple bullous skin lesions over the extremities in all of the patients (**Fig.1**). An abdominal X-ray in first patient showed distended stomach with no distal gas (**Fig.2**). All underwent laparotomy after stabilization. Four neonates with type 2 PA underwent gastroduodenostomy, and the one patient with type 1 PA had excision of membrane and pyloroplasty. By 6 to 36 months follow-up (mean 21.6 months), all 4 surviving infants are doing well. One child died on 12th postoperative day.

DISCUSSION

The earliest description of PA is credited to Caleder in 1749(3), and Swinbure and Kohler first described the association of PA and EB in 1968(4,5). The typical presentation of PA is non bilious vomiting soon after birth without abdominal distention. The diagnosis is made easily on plain abdominal X-ray,

TABLE I DETAILS OF CASES OF PYLORIC ATRESIA - EPIDERMOLYSIS BULLOSA

	Case 1	Case 2	Case 3	Case 4	Case 5
Sex	girl	boy	boy	boy	boy
Type of delivery	cesarean	vaginal	cesarean	vaginal	vaginal
Prenatal ultrasonography	normal	normal	polyhydramnios	NA*	polyhydramnios
Referring time (day)	8	8	3	11	5
Physical examination	multiple bullous eruptions over the extremities	bullous lesions all over the body especially ankles and hands	bullous eruptions on the palm and pressures sites	bullae on hand feet and trunk	multiple bullous eruptions over the extremities
Surgery	gastro duodenostomy	Heineki-Mikulicz pyloroplasty	gastroduodenostomy	gastroduodenostomy	gastroduodenostomy
Outcome (time for follow up)	survived (3 yr)	survived (3 mo)	survived (6 mo)	survived (6 mo)	died on 12th postoperative day

* Not available



FIG. 1 Ruptured bullae over the hand.

which shows a single gas field bubble representing the distended stomach with no distal gas. Antenatal ultrasound may show polyhydramnios and dilated stomach(6). Prematurity and polyhydramnios are found in most of these patients as in our first and third patients. The surgical treatment of pyloric atresia depends on the type of atresia. In type 1 PA, membrane is excised and Heieke-Mikulicz pyloroplasty is performed(7,8). For type 2 atresia the surgical treatment is excision of atretic segment and gasterodeodenostomy(7,9). For type 3 also the surgical treatment is gastroduodenostomy(10). Gastrojejunostomy is better avoided because of 59% failure and 55% mortality(11).



FIG. 2 Plain abdominal radiograph revealing single gas bubble with no gas distally.

Epidermolysis bullosa presents with blisters, crusted erosions and scar formation. Of the 3 types of EB, JEB is mostly associated with PA(12,13).

Whereas the surgical management for PA is straightforward, septicemia, electrolyte imbalance,

WHAT THIS STUDY ADDS?

- Stable neonates having an association of pyloric atresia and epidermolysis bullosa have good survival; surgery should not be denied.

protein loss and failure to thrive complicate the severe exudative skin lesions, often leading to death. This has led to recommendation that surgical treatment be withheld in patients with PA-EB. Four of our cases were stable before operation, and tolerated it well. They are all well on follow-up visits. Follow up was from 6 to 36 months. One case was not in good condition and died of sepsis after operation. Postoperatively, all 4 survived patients tolerated feeding well and went home with no problem. Another series by Hayashi, *et al.*(11) also recommends that stable infants with PA-EB should be treated surgically. Samad, *et al.*(2) reported 3 cases of PA-EB; only one survived.

They concluded that although the prognosis is poor in this condition, surgery should not be withheld on assumption of high mortality(2). We also suggest surgical treatment for neonates with PA-EB who are stable.

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