

LETTER TO EDITOR (VIEWERS CHOICE)

EPIDERMOLYSIS BULLOSA WITH ESOPHAGOBRONCHIAL FISTULA*Jenisha Jain*, Mayank Jain, Ajay Jain, Shohini Sircar, C K Waghmare, Sagar Adkar*

A three years five month old boy born of a third degree consanguineous marriage, presented with blisters and erosions following minimal trauma since birth and history of blood in vomitus one week prior to admission. He was given blood transfusion prior to referral to our centre. He had history of severe constipation which responded partially to stool softeners. He was had been taking only milk and water since birth. His parents reported that the mother had suffered a first trimester abortion in the first pregnancy and another male child born after the patient had similar lesions and had died on eighth day of life following severe skin infection. On examination, the child weighed 9.7 kgs (below third centile) and was 86 cms tall (below third centile). He has multiple blisters all over the body. Polydactyly, nail dystrophy, enamel hypoplasia, dental caries, oral and palatal ulcers were noted. The abdomen was distended and bowel loops were palpable. He was diagnosed to have epidermolysis bullosa based on the classical history and examination. Skin biopsy was not done. Sigmoidoscopy was normal and oesophagogastroduodenoscopy showed narrowing in mid esophagus and multiple esophageal ulcers and webs. Barium swallow revealed a distal esophageal stricture. The child was planned for esophageal dilatation. During the second endoscopy, two openings were noted in the esophageal lumen at 15 cms. By passing a guidewire under fluoroscopic guidance, it was demonstrated that one opening continued into the narrowed esophagus while the other communicated with right sided bronchus. A 15 F Freka's tube was inserted across the esophageal stricture for feeding under fluoroscopic guidance. Feeding was started within four hours of the procedure and the child was discharged on tube feeding. This was done to allow healing of the esophagobronchial fistula and improve nutrition. Constipation improved with adequate fluids and proper nutrition.

Epidermolysis bullosa is a prototype of mechanobullous disease of stratified squamous epithelium that causes blistering and erosions secondary to minor trauma. (1) It predominantly affects the skin and mucus membranes. The gastrointestinal tract, trachea, bladder and urethra are often involved. (2) The common gastrointestinal manifestations include dysphagia, esophageal stricture, pyloric stenosis, anal stricture, chronic constipation and fecal impaction. (3) The reported prevalence of the disease is 1 in 3,00,000 live births. A genetically abnormal collagenase, the structural gene of which is located on chromosome 11, is thought to induce collagen dissociation. (4) Esophageal strictures are seen in children with epidermolysis bullosa. These occur secondary to minor trauma like swallowing solid food. Several studies have recommended total replacement of thoracic esophagus as the treatment of choice and avoidance of procedures like bougienage and endoscopy. (5-7) Techniques of five second inflation using rigiflex balloon have been reported of relieving esophageal stricture. (2) High dose corticosteroids and phenytoin too have been tried. (8,9) Colonic replacement is advised in recurrent obstruction, recurrent dysphagia and esophageal perforation. Our patient was also planned for dilatation following the barium swallow and first endoscopy. On second endoscopy prior to dilatation, an abnormal second opening was noted. Various options like placement of retrievable esophageal stent, surgery and Ryle's

tube placement were considered. Due to underlying skin disease, the chances of esophageal injury, ulcer formation and poor healing would have been higher with these treatment options. The placement of Freka's tube and feeding to improve nutrition and healing was considered to be the most effective treatment option in this case.

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E-published: 1st July 2012. **Art#45**

DOI:10.7199/ped.oncall.2012.45

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