Esophageal Atresia and Tracheo-Esophageal Fistula with Right Pulmonary Agenesis

Y.K. Sarin

The association of pulmonary agenesis, with esophageal atresia with or without tracheo-esophageal fistula is exceedingly rare (1-7). This combination complicates anesthesia and usually mitigates against a successful outcome. Only 4 patients are there on record who have survived beyond neonatal period (5-7) and only 2 of them had long-term survivals (7). A similar case is being reported.

Case Report

A 1900 g full term male neonate born to a multi-gravida by normal vaginal delivery was admitted 19 hours after birth with mild respiratory distress and increased oral secretions. Attempts to pass nasogastric tube were unsuccessful. Examination showed absent right sided breath sounds and a cardiac apex beat in the right chest. Other obvious anomalies included right sided cleft lip with cleft palate. Chest radiograph revealed the coiled nasogastric tube in the blind proximal esophageal pouch. In addition, there was opacification of the right hemithorax with shift of the mediastinum to the right (Fig. 1). The ribs on the right side were crowded. Echocardiography showed normal intracardiac anatomy and left sided aortic arch.

From the Pediatric Surgery Unit, Department of Surgery, Ludhiana 141 008.
Reprint requests: Dr. Yogesh Kumar Sarin, Lecturer in Pediatric Surgery, Christian Medical College, Ludhiana 141 008.
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Definitive repair of the esophageal atresia with division of tracheo-esophageal fistula was done soon after the admission. No lung tissue was seen in the right chest. Three hours post-operatively, the child had sudden cardiorespiratory arrest from which he could be revived. On fifth postoperative day, he had another cardiac arrest while on intermittent positive pressure ventilation, and this time he could not be resuscitated. The parents did not agree for an autopsy.

Discussion

While hypoplasia of the lung is a common association of esophageal atresia with a quoted incidence of 2%(8), the more severe forms of pulmonary aplasia and agenesis are rare associations. Only 23 cases of severely underdeveloped lung in association with esophageal atresia have been recorded till 1989(1-7). There has been no predilection for sex or the side of agenesis/ apleias of lung. Bilateral pulmonary agenesis has been reported in association only twice(2,3). Other associated anomalies noted with this combination include those involving the heart, diaphragm, lip and palate, genitourinary tract, vertebrae and limbs.

The association of these two entities forms a lethal complex owing to the progressive respiratory embarrassment in neonate with already marked compromise of pulmonary reserve, and therefore, the protection of pulmonary system is the prime goal of the management. All such patients would require a prolonged course of intubation and intermittent ventilation. Mortality has been usually attributed to the associated cardiac anomalies(5,6).

REFERENCES


