Pulmonary agenesis: anesthetic experience with four neonates presenting for major surgery

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INTRODUCTION

Pulmonary agenesis (PA) is defined as the total absence of pulmonary parenchyma, blood vessels, and the bronchial tree beyond the tracheal bifurcation. Unilateral congenital PA is rare and its etiology is unknown. Four neonates with PA presenting for major surgery are described.

CASE 1

A two-day-old 32 week premature 2.3 kg male with Trisomy 21 and left renal hydronephrosis presented for emergent exploratory laparotomy for intraperitoneal free air. Preoperative chest radiograph (CXR) demonstrated an opacified left hemithorax and the report attributed this to atelectasis (figure 1). At intubation attempts to pass a 3.0 and 2.5 endotracheal tubes (ETT) failed. The airway was eventually secured with a 2.0 ETT. There were no other untoward intraoperative events. Bowel perforation on the basis of Hirschsprung disease explained the free air. Postoperative CT showed a left PA with critical long segment tracheal stenosis with circumferential rings (figures 2 and 3).

CASE 2

12-day-old 2.3 kg female born at 36 weeks gestation presented for tracheoesophageal fistula (TEF) repair. Delayed referral resulted in severe dehydration with 25% weight loss. During her resuscitation over 3 days, a right PA, ventral septal defect (VSD), Pierre Robin sequence, and pelvic kidney were noted. A gastrostomy was performed on day 15. She died of sepsis prior to definitive repair of her TEF.

CASE 3

An 8-day-old 3.2 kg male product of a consanguineous marriage was born with multiple anomalies. These included Tetralogy of Fallot, tracheoesophageal fistula, right PA, thymic cysts, cerebral cysts, congenital dislocated hips and hemivertebrae. In view of the multiple anomalies gastrostomy and esophagostomy was initially performed. This child subsequently had definitive cardiac surgery and esophageal repair without incident.

CASE 4

A 2-day-old 3 kg female 38 weeks gestation presented for repair of TEF. Left PA and VSD were noted. Apart from an episode of supraventricular tachycardia there were no intraoperative issues.

DISCUSSION

Unilateral congenital pulmonary agenesis is rare and thus may be missed if not suspected. Patients most commonly present in the neonatal period with respiratory distress (1) and there is frequent association with other anomalies (2). Breath sounds may or may not be auscultated and if present are typically diminished. Pulmonary agenesis may be detected antenatally with ultrasound or postnatally with CT or MRI. CXR typically features complete opacity of the affected side with mediastinal shift toward the opacity. Abnormalities of the trachea are a common association. TEF was present in three of our series and tracheal stenosis, when present, may involve a long segment of trachea or even the entire trachea (3). Right-sided PA has been associated with higher mortality than left-sided agenesis. One neonate with right PA in our series died of sepsis and renal failure.

CONCLUSION

While rare, unilateral congenital pulmonary agenesis may coexist with conditions requiring surgery early in life. In view of the associated anomalies careful anesthetic planning and management is required.

REFERENCES