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Case Report Open Access

Anesthetic Challenges in a Child with Pulmonary Agenesis: A Case Report and Review of Literature

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Abstract

Pulmonary agenesis is a rare congenital anomaly, which results in a complete absence in the development of lung tissue, bronchi, and pulmonary vessels. Pulmonary agenesis can occur on its own or it can often be found in conjunction with other congenital defects as part of VACTERL association. Anesthesia for patients with pulmonary agenesis can be challenging particularly when other defects or comorbidities exist. Here, we describe the successful anesthetic and ventilation management of a pediatric patient with right-sided unilateral pulmonary agenesis with VACTERL association.

Keywords: Pulmonary agenesis; VACTERAL association; One lung ventilation

Introduction

Pulmonary agenesis is a rare congenital anomaly occurring during lung development involving lung tissue, bronchi, and pulmonary vessels which results in complete absence or severe hypoplasia of either unilateral or bilateral lungs. Bilateral pulmonary agenesis is not compatible with life. Persons afflicted with unilateral pulmonary agenesis can generally live a normal life provided that their associated malformations are properly managed [1]. Pulmonary agenesis can occur on the right or left side. Although left sided agenesis is more common, agenesis of the right side presents with higher mortality due to the severity of associated anomalies [1]. It can occur on its own, or it can be found in conjunction with other congenital defects such as VACTERL association (vertebral defects, anal atresia, cardiac defects, tracheo-esophageal fistula, renal anomalies, and limb abnormalities).

Anesthesia for these patients often poses a challenge especially when other comorbid conditions exist. Pulmonary agenesis essentially acts as a single lung with delicate tissue; as a result the tissue is more fragile when compared to a normal patient. Therefore, positive pressure ventilation poses a higher risk for barotrauma, pneumothorax, pneumomediastinum, and interstitial emphysema on the side with lung tissue. In addition, other co-malformations can result in further anesthetic challenges such as drugs with renal excretion require careful titration due to renal agenesis or anomalies and cardiac defects effecting drug circulation.

Case Report

A 6-year-old girl (18 kg) presented to the hospital with food impaction after ingestion of a hotdog the night before and multiple episodes of vomiting. Birth history was significant for twin gestation delivered at 35 weeks *via* cesarean section (birth weight of 2400 kg). At four months, she was diagnosed with VACTERL association after she was found to have right pulmonary agenesis, right renal agenesis, congenital scoliosis and asymmetric facies with right facial hypoplasia, mild left ventricular hypertrophy, and tracheomalacia.

Airway exam revealed Mallampati Class 2 and missing upper incisors. A prior computed tomography (CT) of her chest showed atresia of the right mainstem bronchus, aplasia of the right pulmonary artery, and complete right lung agenesis with compensatory hyperinflation of the left lung and herniation across the anterior junction line (Figure 1). There was a complete shift of the heart and mediastinal structures into the right hemithorax, which was also smaller than the left. At baseline, the patient required no additional oxygen therapy and was able to participate in moderate to strenuous activities at school. Her oxygen saturations were 99% on room air.

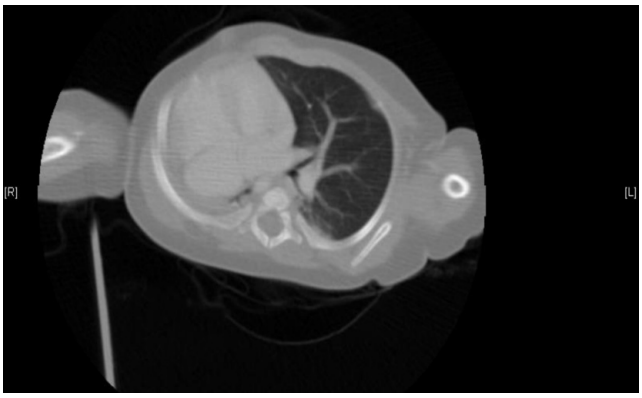

Figure 1: CT chest shows absent right lung and mediastinal structures including the heart shifted to the right.

An esophagogastroduodenoscopy (EGD) for removal of the foreign body was decided. Given the recent history of vomiting, a rapid sequence intubation with cricoid pressure, 45 mg propofol and 30 mg succinylcholine intravenously was done. Direct laryngoscopy with a MAC 2 blade revealed a grade 1 view and a 4.5 ETT was placed

atraumatically, secured at 14 cm. Breath sounds were heard on the left and transmitted across the right hemithorax. End-tidal carbon dioxide ($EtCO_2$) was confirmed. Pressure control mechanical ventilation was initiated with the following settings: inspiratory pressure 15 cmH_2O, respiratory rate of 16, PEEP 3, and 0.5 FiO_2. She was placed in the left lateral decubitus position for the procedure. Tidal volumes remained 110 mL and $EtCO_2$ 32 mmHg.

The patient was maintained on 2.8% sevoflurane. 15 min into the procedure she experienced frequent desaturations to 80%-82% SpO_2 and bradycardia (heart rate (HR) low 60 s). Immediately, FiO_2 was increased to 100%, 0.2 mg IV Glycopyrrolate was given intravenously and the HR responded appropriately, however the desaturations continued to mid-80s. She was taken off the ventilator and recruitment maneuvers were tried, however, her O_2 saturation remained in the mid-80s. The patient was then repositioned from left lateral decubitus to supine and her SpO_2 increased to 90% to 95%. She was continuously hand-ventilated, until she was maintaining spontaneous respirations on the ventilator. A decision was made to discontinue the sevoflurane and switch to IV anesthetics. Small boluses of propofol 5 mg IV were titrated for the remainder of the case. The procedure concluded without any complications and she was extubated uneventfully. The patient was discharged later that evening.

Discussion

Pulmonary agenesis is a congenital malformation that results in the complete absence of a lung. The exact cause of pulmonary agenesis is still unknown, and neither is its true incidence as 50% of cases are stillborn and over 20% die at birth or within their first few months [2]. However, it has been thought to have an autosomal recessive inheritance pattern with other associated factors such as consanguineous marriage, vitamin A deficiency, salicylate use, intrauterine infections, and environmental factors [1,3,4] Males are affected more often affected than females [4]. Those who present with right sided pulmonary agenesis have a worse prognosis due to higher incidence of associated cardiac anomalies and greater mediastinal shift which results in tracheal compression [3,5].

A majority of patients with pulmonary agenesis also present with other associated anomalies, which include tracheal stenosis, patent ductus arteriosus, pulmonary artery atresia, cardiac malformation, horseshoe kidney, and VACTERL association [4]. In a review of 114 published reports by Schechter et al., pulmonary agenesis was found to be associated with anomalies of the skeleton (26%), great vessels (24%), cardiac anatomy (23%), urogenital system (13%), upper respiratory tract (11%), face and cranial nerves (11%), lower intestinal tract (7%), and the esophagus (6%) [6]. While congenital anomalies are associated with both left and right sided agenesis, there is a significant increase in associated anomalies in patients with right sided agenesis [4].

Due to the absence of lung on the affected side, there is a mediastinal shift to the ipsilateral side, which can lead to extrinsic tracheal compression, airway stretching, vascular compression and kinking [7]. There can also be distension and emphysematous changes in the contralateral, normally developed lung [7]. Severe mediastinal shift can cause cardiovascular collapse due to compression of the great vessels and heart. To prevent life-threatening consequences of mediastinal shift, tissue expanders have been inserted into the empty space [2].

Symptoms of pulmonary agenesis can be present at birth as respiratory distress syndrome. Severe recurrent respiratory infection is a common finding during infancy. The infections result from either imperfect drainage of lung secretions or spillover of pooled secretions from a blind bronchial stump into normally developed lung parenchyma on the contralateral side [4]. Often times this causes patients to present with cough, tachypnea, wheezing, respiratory distress, or chronic non-productive cough due to inflammation and bacterial colonization resulting in pneumonia and death before the age of five [3]. Patients may remain asymptomatic until adulthood when the anomaly is identified as an incidental finding on routine examination or during workup for an alternative problem [3]. CT of the chest is considered to be the best diagnostic test for pulmonary agenesis as it allows for visualization of the bronchial tree, parenchyma, and vasculature [4].

Anesthetic management for these patients often poses a challenge, especially if other comorbidities exist. Pulmonary agenesis essentially acts as a single lung with delicate tissue as tissue is underdeveloped and friable due to abnormal blood flow during early embryonic development. Therefore, positive pressure ventilation poses a higher risk of barotrauma, pneumothorax, pneumomediastinum, and interstitial emphysema on the unaffected side. As a result, a lung protective strategy with low tidal volume and increased respiratory rate are recommended. Additionally, the single lung leads to increased airway resistance, requiring higher peak pressures to overcome resistance and prevent shunting and hypoxemia. Airway management secondary to an underdeveloped or stenotic trachea necessitates use of a smaller endotracheal tube, further increasing airway resistance.

Positioning of a patient with pulmonary agenesis is also an important factor to consider intraoperatively. The lateral decubitus position alters pulmonary function in patients without pulmonary agenesis as a combination of the weight of the mediastinum and increased cephalad pressure of abdominal contents on the dependent diaphragm decreases the compliance of the dependent lung. Underventilation of the dependent lung occurs while favoring ventilation of the nondependent lung, and results in a ventilation perfusion (V/Q) mismatch as perfusion is increased to the dependent lung due to gravity [8]. In patients with pulmonary agenesis there is only one functioning lung, therefore while in the lateral decubitus position they are more likely to experience V/Q mismatch than patients with normal lung anatomy and function. In our case, placing a patient with right lung agenesis in a left lateral decubitus position resulted in decreased left lung compliance, changes in the V/Q ratios secondary to the left lung receiving 100% of the cardiac output, and no compensatory ventilation due to agenesis of the right lung. The patient desaturated to mid-80% SpO_2 while in the left lateral decubitus position, which failed to improve with recruitment maneuvers or increase of FiO_2, and saturations promptly improved to mid-90% SpO_2 after changing the patient to supine. Although the explanation of physiology of the positional change is unclear, we believe that the overall lung volume would be reduced, and pulmonary blood flow would favor the dependent periphery rather than the central portions of the lung; hence with a change in position there is an improvement in the oxygenation. The displacement of mediastinal contents in the lateral position with compression of the heart, great vessels, and lung parenchyma could also be a factor in the bradycardia and oxygen desaturations.

While the desaturation in our case arose from position, it is still important to consider other potential etiologies including light anesthesia, atelectasis, return from neuromuscular blockade, and pneumothorax.

Jet ventilation and neuraxial anesthesia have been explored as alternatives to overcome the challenges faced with ventilating a patient with pulmonary agenesis. Babita et al. reported the successful use of combined spinal epidural anesthesia for a cesarean section in a patient with left lung agenesis [9]. In our case, we changed the position of our patient to overcome the additional V/Q mismatch produced by lateral decubitus position which clinically presented as desaturations and bradycardia. Given the events of this case, we would recommend preoperative positioning in the lateral decubitus position in patients with pulmonary agenesis to assess impact on oxygenation prior to anesthetic interventions.

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