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An unusual case of intraoperative hypoxemia in a four year old boy

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Key points

During an intraoperative decrease of oxygen saturation, potential respiratory causes need to be ruled out including a low inspired oxygen concentration, ventilation-perfusion mismatch, hypoventilation or diffusion problems. Rare congenital cardiac abnormality have to be always considered even if in older patients.

Abstract

Various factors may be responsible for intraoperative hypoxemia including either primary pulmonary or cardiac issues. Primary cardiac issues resulting in a right-to-left shunt are generally identified prior to arrival in the operating room. During an intraoperative decrease of oxygen saturation, potential respiratory causes need to be ruled out including a low inspired oxygen concentration, ventilation-perfusion mismatch, hypoventilation or diffusion problems. Of these, ventilation-perfusion mismatch related to bronchospasm, preexisting lung disease, acid aspiration, or inadvertent main stem intubation remain the primary causes of intraoperative hypoxemia. We present a 4-year-old boy who presented for surgical repair of an atrial septal defect and a ventricular septal defect during a surgical trip to a developing country. Following endotracheal intubation, despite the absence of respiratory issues, the pulse oximeter value varied from 82-92% with a PaO2 of 60-70 mmHg. Although the patient was initially presumed to have pulmonary hypertension resulting in right-to-left shunting at the ventricular level, surgical exploration revealed the presence of total anomalous systemic venous return. The anatomy of this rare congenital cardiac abnormality is discussed, previous reports from the literature presented, and its inclusion into the differential diagnosis for hypoxemia is discussed.

Keywords: hypoxemia; intraoperative, pediatric

Introduction

Various factors may be responsible for intraoperative hypoxemia including either primary pulmonary or cardiac issues. Primary cardiac issues with a right-to-left shunt are generally identified prior to arrival in the operating room. The primary focus during intraoperative hypoxemia is identification of potential respiratory causes including a low inspired oxygen concentration, ventilation-perfusion mismatch, hypoventilation or diffusion problems. When patient-related problems are ruled out, monitor-related problems should be investigated. With pulse oximetry, movement artifact,

low perfusion states, or poor signal quality may interfere with accurate readings.

Connection of the systemic venous return to the anatomic left atrium also known as total anomalous systemic venous connection (TASVC) is an extremely rare form of congenital heart disease (CHD). Typically, a right superior vena cava, the inferior vena cava, and the coronary sinus empty into the left atrium. As this results in the return of all systemic blood to the left side of the heart, an atrial septal defect (ASD) must exist for survival to allow for pulmonary blood flow. The return of unoxygenated venous blood to the left side results in cyanosis and arterial desaturation thereby making TASVC a form of cyanotic CHD. Key signs and symptoms of this unusual form of CHD include early fatigue, embolic events, cerebral abscesses, and cyanosis.[1] Early in infancy, the only obvious sign may be cyanosis, which is present in the absence of respiratory distress.

TASVC can generally be diagnosed with echocardiography although given the variable anatomy, a definitive diagnosis may be difficult. As such, a high index of suspicion is necessary and bubble contrast, angiography or cardiac magnetic resonance imaging may be needed for a definitive diagnosis. These diagnostic modalities may be in scare supply in developing countries making the diagnosis of this pathology difficult. The authors present an unusual case of perioperative hypoxemia in a 4-year-old boy who presented for surgery for repair of an ASD and VSD. Although the patient was initially presumed to have pulmonary hypertension resulting in right-to-left shunting at the ventricular level, surgical exploration revealed TASVC. The anatomy of this rare congenital cardiac abnormality is discussed, previous reports from the literature presented, and its inclusion into the differential diagnosis for hypoxemia is discussed.

Case Report

Institutional Review Board approval is not required for single case reports at Nationwide Children's Hospital (Columbus, Ohio). Presentation of this case and review of the hospital course was approved by Heart Care International (Greenwich, Connecticut). The patient was cared for during a cardiac surgical trip to Lima, Peru. The patient was a 4-year-old, 13 kilogram boy, who presented initially with fatigue. He had been experiencing increasing fatigue over the past six months. On physical examination a murmur was heard and he was evaluated with a transthoracic echocardiogram. His echocardiogram showed both atrial and ventricular septal defects without evidence of tricuspid. His pathology was interpreted as showing isolated atrial and ventricular septal defects. His cardiovascular exam revealed mild distal clubbing, and a grade III/VI systolic murmur that was loudest along the lower left sternal border. He had no neurological abnormalities on physical exam. His initial hemoglobin level was 14.1g/dL. The preoperative chest radiograph showed an enlarged cardiac silhouette and dextrocardia. His preoperative workup did not contain a spot-check pulse oximetry reading as it is not routinely available in this developing country. After an appropriate *nil per os* period of 6 hours the patient was transported to the operating room where standard American Society of Anesthesiologists' monitors were placed. The initial oxygen saturation upon the patient's arrival to the operating room was 82 percent while breathing room air. The patient was upset and moving around and the pulse oximeter value was attributed to motion artifact. The patient had not been reported to be cyanotic by the local cardiology team. Anesthetic induction included the inhalation of increasing concentration of sevoflurane in 100% oxygen. Once peripheral venous access was obtained, fentanyl (5 µg/kg) and vecuronium (0.2 mg/kg) were administered. Throughout the induction process the patient's oxygen saturation was never greater 92% despite the administration of an inspired oxygen concentration (F_iO_2) of 100%. Following tracheal intubation, a second peripheral intravenous cannula, an arterial cannula, and a central venous

cannula were placed. Maintenance anesthesia included fentanyl (total dose of 15 µg/kg) and isoflurane (exhaled concentration 1-2%). An arterial blood gas analysis confirmed the low pulse oximeter value with a PaO_2 value of 62 mmHg with no evidence of a base deficit. No change in the pulse oximeter value was noted when varying the F_iO_2 from 40 to 100%. The persistently low pulse oximeter value was discussed with the cardiac surgeon and given the reported cardiac pathology of a large ASD and VSD, sildenafil (1 mg/kg) was administered via an orogatric tube to treat presumed pulmonary hypertension. Other than hypoxemia, the patient was otherwise hemodynamically stable during the entire period prior to cardiopulmonary bypass (CPB). Following sternotomy and placement of arterial and venous cannulas, CPB was initiated without difficulty.

The patient underwent routine sternotomy and after opening the pericardium the apex of the heart was located in the right chest, the pulmonary artery (PA) originated anterior and rightward of the posterior aorta. There was bilateral superior vena cava (SVC), with the inferior vena cava (IVC) in the midline. The patient was placed on cardiopulmonary bypass with cannulae in the bilateral SVC and IVC. He was cooled to 32 degrees Celsius. The right atrium was opened and the left SVC, right SVC and IVC drained into the posterior left atrium. The atrial septum was intact with a small superior patent foramen ovale. The pulmonary veins drained into the LA and were posterior to the systemic venous drainage.The atrial septum was excised. Intra-atrial baffles were created connecting the right SVC and IVC to the anterior atrium using CorMatrix ECM®, CorMatrix, Roswell, GA. The VSD was closed using a GORE-TEX® patch, Gore Medical, Flagstaff, AZ, through the tricuspid valve. The atrial septum was reconstructed between the IVC and right SVC baffle, and along the annulus of the tricuspid valve posterior and to the posterior opening of the L SVC. Care was taken to avoid obstruction to the pulmonary veins on the right side. After an aortic cross-clamp time of 118 minutes and a total CPB time of 161 minutes, the patient was successfully weaned from the CPB without inotropic support. His post-CPB PaO2 was greater than 500 mmHg with an inspired oxygen concentration of 100%. The patient's chest was closed, residual neuromuscular blockade reversed, and his trachea was extubated without incident in the operating room. The patient was transferred to the cardiac ICU where he recovered overnight. The remainder of his postoperative course was uneventful. Transthoracic echo revealed unobstructed systemic venous flow to the right atrium, no residual shunts, and unobstructed pulmonary venous return to the left atrium. He had excellent biventricular function.

Discussion

Total anomalous systemic venous connection can exist in multiple different forms. It may be accompanied by dextrocardia with or without situs inversus and occur in association with heterotaxy syndromes. The diagnosis of TASVC has been reported in only a small number of patients in the literature. Although it remains an uncommon form of cyanotic CHD, it should be considered in the differential diagnosis of hypoxemia without accompanying lung disease and in the absence of respiratory distress. Several of the patients reported in the literature presented with the isolated finding of a low oxygen saturation that was incidentally noted on pulse oximetry. Such was the case in our patient, who did not appear clinically cyanotic, and yet had an oxygen saturation in the 80% range. As noted in our patient, the oxygen saturation in patients with true shunt related to CHD is not generally affected by changing the inspired oxygen concentration. This clinical feature may give a clue to the differential diagnosis when trying to differentiate cardiac causes of hypoxemia from respiratory causes as the latter generally responds to increasing the F_iO_2.

TASVC has been reported in all age ranges from neonates to adults.[2,3] Our review of the literature

revealed a total of approximately twenty reported cases with diagnosis attained from a combination of contrast echocardiography and angiography. The presenting signs and symptoms have included congestive heart failure, brain abscesses, other embolic phenomenon, hypoxemia, cyanosis, and decreased exercise tolerance.[4-6]

Specific perioperative issues included associated congestive heart failure related to volume overload, arrhythmias due to atrial dilatation, polycythemia due to prolonged hypoxemia, and the potential for paradoxical emboli related to the right-to-left shunt.[7] The latter mandates meticulous technique to avoid the administration of air or particulate matter. The right-to-left shunt results in a more rapid onset of intravenously administered anesthetic agents and a delayed onset of inhalational anesthetic agents. Although our patient's preoperative echocardiogram demonstrated adequate ventricular function, the choice of anesthetic agents may need to be modified in patients with associated myocardial dysfunction or congestive heart failure as well as the avoidance of agents that may precipitate tachycardia and arrhythmias (pancuronium and desflurane).[8] The presence of chronic volume overload and exaggerated pulmonary blood flow also presents the potential for perioperative pulmonary hypertension especially when the diagnosis and repair are performed outside of infancy. Additional concerns may be related to the presence of associated heterotaxy syndromes as well as anomalies of the systemic venous drainage which impact on the techniques of cannulation for CPB.[9,10]

Several factors led to the delay in diagnosis in our patient including limited previous access to primary healthcare, issues related to the quality of the echocardiogram due to the equipment available, and lack of the use of preoperative evaluation with the route use of pulse oximetry. Given that the echocardiogram provided a presumptive diagnosis of an ASD and VSD, cardiac catheterization was not deemed necessary. Despite this, the possible diagnosis of TAPVC should be included as a cause of cyanotic CHD and should be considered when patients present with hypoxemia in the absence of respiratory disease, regardless of the age of the patient. Associated signs and symptoms may include exercise intolerance as well as comorbid conditions related to the chronic right-to-left shunt including cerebrovascular accidents and cerebral abscesses. The diagnosis and associated cardiac malformations can generally be identified using contrast echocardiography although occasionally cardiac catheterization or MRI may be required.

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