PHACE(S) Syndrome: Unmasking the Anesthetic Implications
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Introduction: PHACE (Posterior fossa brain malformations, facial Hemangiomas, Arterial anomalies, Cardiac anomalies and/or aortic coarctation, and Eye abnormalities) - or PHACES (when a ventral defect including Sternal clefting is involved), is a neurocutaneous syndrome (1) with significant anesthetic implications. A rare entity, the majority of articles on this topic is comprised of isolated case reports. Furthermore, PHACE(S) syndrome has not been extensively addressed in the anesthesia literature.

Case Report: F.S. was a 4.1kg female with a delivery complicated by apnea requiring intubation for 5 hours. On preoperative evaluation, physical exam revealed a facial hemangioma, micrognathia, and a sternal V-shaped cleft. Diagnostic testing revealed a normal posterior fossa and bilateral optic nerve hypoplasia. She had numerous cardiovascular abnormalities including an absent left carotid artery, a small patent ductus arteriosus and a patent foramen ovale. There was an unusual branching pattern of the aortic arch with abnormal rightward origin of the left subclavian artery and prominent ductal diverticulum originating from the left side of the aorta opposite to the origin of the left subclavian artery. She was brought to the operating room on day of life 15 for elective repair of the sternal cleft. The anesthetic sequence involved induction with sevoflurane, placement of peripheral intravenous and arterial lines, intubation with vecuronium and ductal diverticulum originating from the left side of the aorta opposite to the origin of the left subclavian artery. She was maintained with isoflurane and remifentanil. The surgical component of the procedure involved a midline vertical incision, partial sternotomy and sternectomy, placement of sutures under direct vision through intercostal spaces and then approximation of the two sternal bars via traction of the crossed sutures. During the repair alternating displacement of each hemithorax was required to place sutures under direct vision through intercostal spaces and then placement of the pulse-oximeter (on the right foot) was confirmed and a second pulse-oximeter probe was placed on the right hand to measure preductal saturation. A 10% difference in pulse-oximeter readings was noted during the episodes of desaturation, indicating the possibility of a shunt. Oxygenation improved after pressure was relieved from the hemithorax. The patient’s blood pressure remained stable throughout the operative course with volume maintenance and deficit replacement. Blood loss was negligible. The closure of the sternum was tested, with no change in peak inspiratory pressures or hemodynamic parameters. After chest closure, the FiO2 was gradually decreased to 30% with oxygen saturations in the right hand and right foot equalizing and remaining stable at 99 – 100%. Correct placement of the pulse-oximeter (on the right foot) was confirmed and a second pulse-oximeter probe was placed on the right hand to measure preductal saturation. A 10% difference in pulse-oximeter readings was noted during the episodes of desaturation, indicating the possibility of a shunt. Oxygenation improved after pressure was relieved from the hemithorax. The patient’s blood pressure remained stable throughout the operative course with volume maintenance and deficit replacement. Blood loss was negligible. The closure of the sternum was tested, with no change in peak inspiratory pressures or hemodynamic parameters. After chest closure, the FiO2 was gradually decreased to 30% with oxygen saturations in the right hand and right foot equalizing and remaining stable at 99 – 100%. Neurovascular blockade was reversed. End-tidal carbon dioxide was stable at 45 torr during spontaneous respiration. The patient’s trachea was extubated at the end of the procedure and she was discharged from the hospital two days later in good condition.

Discussion: We present the anesthetic management of a 15-day-old female with PHACES syndrome for repair of a sternal cleft. The intraoperative course was complicated by episodes of desaturation that, based on the differential pulse-oximeter readings and the clinical scenario in which they occurred, were likely due to a ductal shunt. Although the anesthetic management for this patient was straightforward, it is important to be cognizant of the possibility for potential complications when caring for patients with PHACES syndrome - a diagnosis that should be considered in any infant with a large, plaque-like facial hemangioma. Extracutaneous hemangiomas are reported in 22% of cases, with the subglottic airway being the most common location (2). Arterial pathology in the head and neck including stenosis or absence of an internal carotid artery, or occlusion of the proximal anterior, middle or posterior cerebral arteries has been reported. These abnormalities can lead to a Moya-Moya like condition with extensive collateralization. The majority of patients with cerebral and arterial anomalies develop secondary neurological sequelae such as seizures, developmental delay, and/or contra-lateral hemiparesis (3). There are at least two reports of death from cerebrovascular infarction, one of which occurred postoperatively (4, 5). Various associated cardiovascular anomalies have been reported including PDA, VSD, ASD, pulmonary stenosis and coarctation of the aorta. If present, sternal clefting should be repaired neonatally while the chest wall is highly compliant and the risk of significant compression of thoracic structures is minimal (6). In summary, the anesthesiologist should be aware of the potential for multi-organ pathology when presented with a PHACE(S) syndrome patient.

References: