PHACES Syndrome and anesthesia management for sternal cleft repair: A case report

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Introduction: PHACES Syndrome is an acronym used to describe a neurocutaneous syndrome encompassing the following features: posterior fossa brain malformations, hemangiomas, arterial anomalies, coarctation of the aorta, intra-cardiac defects, eye abnormalities, and sternal cleft defects. (1) PHACES syndrome is usually first diagnosed by the presence of an aggressive facial hemangioma, sometimes involving the airway; to date there have been no reported descriptions of anesthetic management. We wish to report our experience with the anesthetic management of a pediatric patient with PHACES syndrome during sternal cleft repair. Sternal cleft is a rare congenital malformation resulting from complete or partial failure of the two lateral sternal bars to fuse during the third month of embryologic development. (2)

Case Report: Our patient, a 2 month old 5.6 kg female, was born at term and noted to have a sternal cleft with tachypnea, mild retractions, and transient requirement for supplemental oxygen, prompting the presumptive diagnosis of PHACES syndrome. Evaluation with an MRI did not indicate any posterior fossa abnormalities, and echocardiogram demonstrated no malformations of the aortic arch or intracardiac anatomy. At 2 weeks of age she developed a right lower lip hemangioma. She presented for elective repair of the sternal defect at 2 months of age, and had no other problems noted on history, physical examination, or laboratory evaluation.

Anesthesia was induced with inhalational sevoflurane and spontaneous ventilation because of the possibility of a subglottic hemangioma, followed by intravenous line placement and easy mask ventilation. Upon direct laryngoscopy careful attention was taken to avoid trauma to the lower lip hemangioma and a grade 1 view was obtained. Her airway did not have any evidence of hemangiomas and a 3.0 cuffed endotracheal tube was easily inserted, which provided a leak at an inspiratory pressure of 10 cmH₂O. A 2nd intravenous line and right radial arterial line were placed. Pressure controlled ventilation was provided with tidal volumes of 10 cc/kg and peak inspiratory pressures of 18 mmHg. Anesthesia was maintained with sevoflurane and fentanyl. Surgery consisted of osteotomies of the 1st through 5th costal cartilages followed by wire closure of the defect. After closure of the sternal cleft, peak pressures of 20 mmHg were required to maintain same tidal volume. Postoperatively the patient remained intubated and was transferred to the intensive care unit. She was extubated the following morning and resumed breast feeding the same day. She received supplemental oxygen via nasal cannula at 0.25 L/min of oxygen for three days, and was discharged home in stable condition on post-operative day five. Pain throughout the post-operative period was managed with morphine infusion.

Discussion: PHACES syndrome was first identified as a constellation of abnormalities by Dr. Ilona Friedman in 1996. To date there have been no published reports of anesthetic management of these patients. The pathogenesis of the disorder is currently unknown. It does however have a female predominance and thus it has been speculated that the syndrome might represent an X-linked dominant condition (3). The most common initial presentation of the disorder is either a hemangioma or, as seen in our patient, a sternal cleft (1). The hemangioma is usually of a plaque morphology and located on the face. In one study where 14 patients were evaluated retrospectively the hemangioma usually followed the trigeminal division V1 distribution. It was also noted that if the lesion involved all three dermatomes then the patient had an increased risk of cerebrovascular anomaly (1). The instability of the sternum may result in ventilatory insufficiency, similar to that seen with a flail chest. Concerns for the anesthesiologist include detection and management of airway hemangiomas, and altered thoracic compliance with rib mobilization and closure of the underdeveloped sternum.
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