Anesthetic management of a patient with PHACE syndrome undergoing a stereotaxic biopsy of a right brain ventricular mass

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### Introduction

#### Background

- PHACE syndrome is a neurocutaneous syndrome first described by Frieden et al in 1996 that consists of:
  - Posterior fossa malformations
  - Hemangiomas
  - Arterial anomalies
  - Coarctation of the aorta
  - Eye abnormalities
- Occurs more commonly in females at 8:9-1:1 (2)
- Results from a development defect occurring around 8 weeks of gestational age (1-2)
- These patients often need vascular, neurosurgical, ophthalmologic, or otolaryngology interventions (1)
- Due to multisystem involvement, anesthesia in patients with PHACE syndrome poses a major challenge

### Case History (continued)

#### History of Present Illness

- Past medical history was notable for:
  - Cerebellopontine angle mass
  - Partial anomalous pulmonary venous return (right upper pulmonary vein returns to the SVC)
  - Vascular ring- right aortic arch with aberrant left subclavian artery (Kommerell's diverticulum)
  - Dilated left common carotid artery with hypoplastic right common and internal carotid arteries
  - Hypoplastic left and right vertebral arteries
  - Atretic distal basilar artery
  - Multifocal arterial collaterals

#### Perioperative Course

- On the day of surgery, the patient was given oral midazolam as premedication
- An inhalational induction was performed with sevoflurane and oxygen
- A radial arterial line and 2 peripheral IVs were placed
- Direct laryngoscopy was performed and a size 4.0 microcuff endotracheal tube was placedatraumatically
- General anesthesia was maintained with sevoflurane/fentanyl/rocuronium
- The patient required frequent boluses of phenylephrine and eventually a dopamine infusion (3 mcg/kg/min) to maintain blood pressure
- Blood pressure ranged from 60-100 systolic and 20-60 diastolic while heart rate ranged from 90-18

### Discussion

- Perioperative complications arise mainly from:
  1. Cerebral hypoperfusion secondary to arterial malformations, which can result in seizure or stroke (1,3)
  2. Supra or subglottic hemangiomas that can result in airway obstruction and contribute to difficulty with intubation (1,3)
- Neuromonitoring with BIS or NIRS may be helpful to identify times of cerebral hypoperfusion to minimize chances of ischemia (1)

### References