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Case Report of an Awake Craniotomy in a Patient With Eisenmenger Syndrome

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Abstract

We present a detailed report of an awake craniotomy for recurrent third ventricular colloid cyst in a patient with severe pulmonary arterial hypertension in the setting of Eisenmenger syndrome, performed 6 weeks after we managed the same patient for a more conservative procedure. This patient has a high risk of perioperative mortality and may be particularly susceptible to perioperative hemodynamic changes or fluid shifts. The risks of general anesthesia induction and emergence must be balanced against the risks inherent in an awake craniotomy on a per case basis.

Adult patients with congenital heart disease (CHD) increasingly present for noncardiac surgery (NCS).^{1,2} Among adult CHD patients, those with Eisenmenger syndrome (ES) have a particularly high peri-operative risk. ES develops as a complication of cyanotic CHD and is characterized by a bidirectional or right-to-left intracardiac shunt, pulmonary hypertension, and systemic hypoxemia. Case series of NCS in adult ES patients over the past 80 years report mortality rates between 4% and 27%,^{3–8} with lower estimates in more recent series and higher estimates associated with obstetric patients. Even with minor procedures,⁵ mortality, usually from cardiac arrest, is associated with factors that worsen right heart function: oxygen desaturation, severity of right-to-left shunt fraction, right ventricular dysfunction,⁷ and fluid shifts related to hemorrhage and resuscitation.^{3,4,6,8} Available case

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DISCLOSURES

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data do not clearly favor monitored anesthesia care (MAC), regional anesthesia (RA), or general endotracheal anesthesia (GETA) for NCS in ES patients.^{5–8}

We recently reported on the intraoperative management with RA and MAC of a 32-year-old man with ES undergoing fenestration of a third ventricle colloid cyst and ventriculoperitoneal shunt (VPS) placement for obstructive hydro-cephalus.⁹ The cyst and associated symptoms recurred in a period of weeks. Symptomatic colloid cysts of the third ventricle may result in neurological deterioration or sudden death, risks that cannot be stratified on the basis of cyst size, symptom duration, or the presence of hydrocephalus.^{10,11}

We present the case of this patient now undergoing awake craniotomy for total resection of the same lesion. In light of our recent prior experience with this patient and the unique intraoperative risks facing this patient, we again proceeded with RA and MAC, but with a modified hemodynamic management in this second procedure, allowing a unique comparison across cases. A written Health Insurance Portability and Accountability Act authorization to use or disclose existing protected health information was obtained.

CASE DESCRIPTION

A 33-year-old man (53.8 kg, 170 cm) with severe pulmonary arterial hypertension (PAH; World Health Organization group 1), New York Health Association class II, and ES due to residual congenital ventricular septal defects (VSDs) presented for craniotomy to remove a 1.5-cm colloid cyst of the third ventricle.

His medical history was significant for attempted closure of a congenital VSD at 3 years of age, with bidirectional shunting via residual small membranous VSD and large complex muscular VSD. Cardiac catheterization 3 years before his VPS surgery revealed these pressures (mm Hg): right ventricle (RV): 114/16/80; right pulmonary artery (PA): 120/53; right pulmonary capillary wedge: 10; and mean arterial: 89. Pulmonary vascular resistance was 13.8 Wood units (normal range, 1–2 Wood units), and cardiac output was 4.15 L/min. Inhaled nitric oxide (iNO) trial showed no change. Transthoracic echocardiography 1 week before craniotomy showed moderate RV hypertrophy with moderately reduced systolic function, preserved left ventricular systolic function, and severe tricuspid regurgitation with estimated RV systolic pressure of 112 mm Hg (systemic blood pressure, 105/61 mm Hg); this was not significantly different from his pre-VPS study. The patient was stable on diltiazem, furosemide, and triple-PAH therapy (ambrisentan, sildenafil, and inhaled treprostinil). Regarding his functional status, he was able to play table tennis 1.5 hours per day and used supplemental O₂ 3 times per week for 1 to 2 hours. Four days before surgery, he was admitted to the hospital for transition from inhaled to subcutaneous treprostinil infusion. Room air O₂ saturation (SpO₂) was 88% to 92% at rest. Laboratory studies were unremarkable; N-terminal prohormone of brain natriuretic peptide was 330 pg/mL, down from 948 pg/mL after VPS placement.

In the operating room, all intravenous (IV) lines were meticulously debubbled. Oxygen was delivered via nasal cannula capable of capnography. Respiratory rate was monitored by impedance plethysmography via both electrocardiogram and end-tidal CO₂ (EtCO₂).

Respiratory rate and Etco₂ remained 14 to 18 breaths/min and 33 to 36 mm Hg, respectively, for the duration of the case. The utility of trending Etco₂ with nasal capnography was confirmed by Paco₂ from an arterial blood gas (pH/Pco₂/Po₂/HCO₃ of 7.36/37.6 mm Hg/52.3 mm Hg/20.5 mEq/L, respectively).

To limit PAH exacerbation by hypoventilation-related hypercarbia and pain or stress, sedation and anxiolysis were maintained with midazolam (8 mg given in 1 mg increments over 6 hours) and dexmedetomidine (1 µg/kg bolus over 30 minutes + 0.4–0.8 µg/kg/h infusion). Depth of sedation was monitored during the case by respiratory rate and intermittent verbal communication with the patient. Local injection of 2% buffered lidocaine preceded placement of all lines and invasive monitors, including 16- and 18-g peripheral IV catheters, 20-g left radial arterial line, and left subclavian triple-lumen central venous pressure line. A scalp block was performed with 0.5% bupivacaine with epinephrine, and Mayfield pin sites were anesthetized with 2% buffered lidocaine. Acetaminophen 1000 mg IV was given during closure.

Before any medication, Spo₂ was 85%. We trended Spo₂ as an index of intracardiac shunt fraction, and this value ranged from 79% to 90% during the case. Vasopressin infusion (0.005–0.04 U/min) was initiated with sedation and slowly titrated during invasive monitor placement to target mean arterial pressure >60 mm Hg and Spo₂ > 80%. iNO therapy was available during the procedure, but was not initiated. ST segment size was continuously monitored for signs of right heart strain. Dobutamine infusion (1–2 µg/kg/min) was initiated after incision, anticipating potential acute right heart strain associated with venous air embolism (VAE) or rapid volume loss resuscitation. Both vasoactive infusions and sedation were discontinued at the conclusion of surgery. Our intraoperative management is schematized in the Figure.

We sought to minimize volume-related strain on his right heart with the competing goal of avoiding venous air entrainment. The patient was positioned in a semirecumbent position, his head approximately 5 cm above his heart. We confirmed ready availability of saline to flood the surgical field. A precordial Doppler monitor was placed, and its sensitivity was confirmed by the agitated saline test. Initial central venous pressure after central line insertion, leveled at the site of surgery, was –3 mm Hg at which time the patient was deemed euvolemic, and fluid therapy was targeted to maintain this value. In total, 500 mL Normosol (Hospira Inc, Lake Forest, IL) and 250 mL normal saline were given. Estimated blood loss was 150 mL.

The patient tolerated the 4.5-hour procedure without response to noxious stimuli, significant hemodynamic changes, respiratory depression, or dysrhythmias. He was transported to the intensive care unit for monitoring, where he required vasopressin infusion intermittently over the first 36 hours after surgery. He otherwise had an unremarkable postoperative course.

DISCUSSION

ES and severe PAH pose multiple elevated risks, which we considered in planning this patient's anesthetic: (1) high volume blood loss and resuscitation, a likely risk factor for

perioperative mortality in ES patients undergoing NCS^{6,8}; (2) exacerbation of PAH and RV systolic dysfunction due to hypercarbia, hypoxemia, acidemia, stress associated with stage 2 physiology, and positive pressure ventilation (PPV); and (3) VAE, which could manifest as a pulmonary embolism, causing RV strain, as a paradoxical air embolism (PAE) entering the most nondependent coronary vessel, the right coronary artery (RCA), or as a PAE-associated stroke.

The lesion's location above the third ventricle required surgical dissection in the midline near the sagittal sinus. In ES patients, accelerated blood loss can precipitate a series of adverse events that enhance each other's impact. Blood loss and the resulting systemic arterial hypotension increase right-to-left shunting, reduce blood's O₂ carrying capacity, and reduce perfusion pressure and cardiac output of increasingly hypoxemic blood to the RCA. These acute changes synergistically impair O₂ supply to an RV that is now faced with a higher demand from PAH, acutely worsened by hypoxemia and evolving acidemia due to systemic hypoperfusion.

Therefore, our overriding perioperative hemodynamic goal was to maintain a minor left-to-right shunt with pulmonary and systemic pressures close to baseline. While PA pressures can be directly measured with a PA catheter, we decided against placing one due to the risk of arrhythmia and PA rupture. In lieu of a PA catheter, we used his SpO₂ to monitor his shunt fraction, titrating a combination of dobutamine and vasopressin infusions.

Notably, we modified our hemodynamic management compared to his prior procedure,⁹ for which epinephrine infusion and iNO were used. Anticipating highly stimulating portions of the patient's VPS placement procedure,⁹ which could exacerbate PAH, we favored prophylactic pulmonary vascular resistance reduction (iNO) and RV inotropy (epinephrine). Continuous dobutamine/vasopressin may have some advantage over epinephrine/iNO for a lengthy (4.5 hours) procedure with potential blood loss, namely, prioritizing RV inotropy (dobutamine) and RCA perfusion (vasopressin). In addition, epinephrine has been associated with transient lactic acidosis, tachycardia, and possibly arrhythmia when compared to a dobutamine–norepinephrine regimen.¹² Furthermore, given the patient's minimal SpO₂ improvement with iNO during his prior procedure (94% from 92%) and history of fixed PAH, we held iNO in reserve and did not initiate therapy.

We opted to begin the case with RA and MAC. Fortunately, surgical management of these lesions is generally well tolerated with minimal hemodynamic disturbances.^{10,13} Therefore, while prepared for potential volume resuscitation, we weighed the risks of PPV more heavily. PPV, by raising intrathoracic pressure, can place strain on the RV, exaggerate the fall in preload due to hypovolemia, and promote a right-to-left shunt. By this latter mechanism, PPV has been shown to promote PAE,^{14,15} a potentially devastating complication in this patient. PPV does not appear to affect the incidence of VAE in adults,¹⁶ in contrast to pediatric patients,¹⁷ and may in fact trigger VAE during the release of positive pressure.¹⁸ If large volume resuscitation required conversion to GETA intraoperatively, we anticipated that our patient could tolerate PPV. Due to RV remodeling and preconditioning, ES patients appear better able to compensate for RV strain compared with group 1 PAH patients with

noncongenital disease,¹⁹ a conclusion supported by the many available case reports of GETA with PPV in ES patients.^{3,5–8}

Having an awake patient may limit control of $Paco_2$ and brain swelling, and conditions such as obstructive sleep apnea may lead to airway compromise during periods of moderate sedation. To mitigate these concerns, we closely monitored our patient's respiratory state with nasal capnography (validated by $Paco_2$) and chose sedatives with low risk of respiratory depression, avoiding propofol. We required a low Fio_2 to minimize the risk of fire in the presence of surgical electrocautery. Our ability to control Fio_2 was limited by using the nasal cannula and resulted in an undesirably low Pao_2 . If faced with a similar situation, we would use the recently available high-flow humidified nasal cannula system. In the event that obstruction or hypercapnia became significant enough to affect hemodynamics or operating conditions, we were readily equipped for a controlled induction to GETA with ketamine and rocuronium, planning for an asleep fiberoptic intubation, without removing the patient's head from the Mayfield pin fixation.

Proceeding with surgery under these circumstances would typically be considered a last resort, without compelling evidence that such a patient could tolerate surgical stress of this nature. Our patient had an urgent need for definitive surgery, and he tolerated a cyst fenestration and VPS placement well only 6 weeks before. His previous procedure gave us useful data regarding his response to hemodynamic interventions and his tolerance for RA. Taken together with existing case data, our experience suggests that multiple anesthetic options can be safely used in this high-risk group.

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Figure.

Intraoperative timeline of surgical and anesthetic events in the described awake craniotomy, accompanied by potential adverse hemodynamic changes relevant to patients with PAH at each time point. *Choice of vasoactive drugs is described in the main text. DEX indicates dexmedetomidine; MID, midazolam; PAE, paradoxical air embolism; PAH, pulmonary arterial hypertension; PVR, pulmonary vascular resistance; SVR, systemic vascular resistance; VAE, venous air embolism.