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Urgent cesarean delivery and prolonged ventilatory support in a parturient with Fontan circulation and undiagnosed pseudocholinesterase deficiency

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ABSTRACT
A parturient with Fontan circulation required general anesthesia for urgent cesarean delivery and subsequent prolonged postoperative ventilation for newly-diagnosed pseudocholinesterase deficiency. Anesthetic management necessitated a thorough understanding of the hemodynamic principles of the Fontan circulation and physiologic adaptations during surgical delivery and recovery in the intensive care unit.

Keywords: Fontan: circulation, procedure; Pseudocholinesterase deficiency; Pregnancy; Positive pressure mechanical ventilation; Negative pressure mechanical ventilation; Cesarean delivery; Tricuspid atresia

Introduction

Parturients with congenital heart disease pose significant anesthetic challenges in the peripartum period. Pseudocholinesterase deficiency, which occurs in 1:3200 parturients, also complicates anesthetic management, especially if the condition is undiagnosed before induction of general anesthesia. This combination of conditions in a parturient presents unique challenges to the anesthesiologist and critical care physician in both ventilatory management and hemodynamic control. We describe a parturient with tricuspid atresia and Fontan circulation who underwent general anesthesia for urgent cesarean delivery and required prolonged postoperative mechanical ventilation for undiagnosed pseudocholinesterase deficiency. This case has been published by our obstetric colleagues,[1] but in this report we discuss the implications of anesthesia and mechanical ventilation.

Case report

A 26-year-old 63 kg G3P0 parturient at 33 + 6 weeks of gestation presented for urgent cesarean delivery. Her cardiac history was significant for congenital tricuspid atresia with hypoplastic right heart, for which she underwent surgical palliation with Fontan circulation at age seven - years. Her functional cardiac status before pregnancy was New York Heart Association (NYHA) class I, and she was able to participate in moderate regular exercise. Her past medical history was otherwise significant for scoliosis and an elective blepharoplasty, at an outside institution, for which she was unable to recall the type of anesthesia. She denied any family history of anesthetic complications.

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Her pregnancy remained relatively uneventful until 23 weeks, at which time she developed mild peripheral edema and dyspnea. Blood pressure and heart rate parameters were within her normal range: 104/65 mmHg and 81 beats/min, respectively. Her symptoms progressed to significant dyspnea with mild exertion and venous congestion headaches by 27 weeks, and she was admitted to the antepartum unit for bed rest, diuresis, and betamethasone therapy in preparation for potential preterm delivery. An echocardiogram showed the Fontan connection and bidirectional Glenn anastomosis to be widely patent, with an ejection fraction of 58%. The left ventricle demonstrated borderline enlargement, the right atrium was severely enlarged and a restrictive ventricular septal defect with bidirectional flow into the hypoplastic right ventricle was present. Oral hydrochlorothiazide 25 mg daily and furosemide 20 mg daily (with concomitant potassium supplementation) improved symptoms of dyspnea, headaches, and edema without adversely affecting blood pressure. Subcutaneous unfractionated heparin (10 000 IU twice daily) thromboprophylaxis was administered due to concern for venous thromboembolism from the combination of activity limitation and the passive flow of the Fontan circulation.

Because of collective cardiology and obstetric concerns for anticipated maternal intolerance of the hemodynamic stress of labor, elective cesarean delivery under neuraxial anesthesia was scheduled for $34 + 2$ weeks. However, spontaneous labor ensued at $33 + 6$ weeks, necessitating urgent cesarean delivery. Her most recent dose of heparin had been administered 8 h previously, and an activated partial thromboplastin time of 41 s (normal 21–33 s) was obtained immediately preoperative. Because of parturient preference, her scoliosis, and her anticoagulated status, a general anesthetic was selected for the procedure.

On arrival to the operating room, she was placed in the supine position with left uterine displacement, and two peripheral intravenous catheters and a left radial artery catheter were placed. A rapid sequence intravenous induction was performed with etomidate 0.3 mg/kg and succinylcholine 1.5 mg/kg. Blood pressure transiently increased to 160/100 mmHg during intubation and spontaneously returned to baseline. Anesthesia was maintained with sevoflurane 1.7% and an air:oxygen mixture with FiO_2 of 0.54. Three minutes after induction, a vigorous neonate was delivered. Following placental delivery, intravenous oxytocin 30 IU/L was initiated to treat mild uterine atony. Long-acting neuromuscular blocking agents were not administered. She remained hemodynamically stable and well oxygenated throughout the remainder of the procedure. Sevoflurane was discontinued after incision closure. Twenty minutes after discontinuation of sevoflurane, the patient remained apneic. Stimulation of the ulnar nerve demonstrated absence of a twitch response. Stimulation of the contralateral ulnar nerve also demonstrated absence of a twitch response, confirming prolonged neuromuscular relaxation. Pseudocholinesterase deficiency was suspected; a propofol infusion was initiated, and she was transferred to the intensive care unit (ICU) for continued sedation and mechanical ventilation.

Sedation was maintained using a titrated propofol infusion of 15–45 μg/kg/min with intermittent bolus doses of midazolam 1–2 mg. Mechanical ventilation mode was initially synchronized intermittent mandatory ventilation (SIMV) with pressure support (PS), with settings of tidal volume 450 mL, rate 14 breaths/min, FiO_2 0.5, Positive end expiratory pressure (PEEP) 5 cmH_2O, and PS 10 cmH_2O. Airway pressures were maintained at a mean of 10 cmH_2O, and peak inspiratory pressure ranged from 15 to 20 mmHg. She gradually regained muscular strength with improved hand grasp 7.5 h after the succinylcholine dose, but continued to require ventilatory support due to inadequate tidal volumes. After approximately 7.5 h on SIMV, she was weaned to spontaneous ventilation using continuous positive airway pressure (CPAP) 5 cmH_2O with PS 10 cmH_2O. Ninety minutes later, and 9 h after the dose of succinylcholine, she was extubated and oxygen was administered by nasal cannulae. Blood gas analysis during weaning from the ventilator demonstrated normal oxygenation, ventilation and acid–base status. Despite her Fontan circulation, there were no significant hemodynamic derangements during mechanical ventilation. Laboratory analysis demonstrated a serum pseudocholinesterase activity $<$100 U/l (normal 1800–6600 U/l). A dibucaine number could not be determined. She denied any recall of events during the period between anesthetic induction and just before extubation in the ICU. The remainder of her postoperative hospital course was unremarkable, and she was discharged to home on postoperative day five.

Discussion

The Fontan circulation is a single ventricle cardiac physiology in which blood flows passively into the pulmonary arteries (Fig. 1). Preload is therefore the most important determinant of cardiac output.[2] The systemic ventricle may be either the morphologic right or left ventricle, depending on the type of congenital heart lesion. Common cardiovascular complications resulting from Fontan circulation include poor ventricular filling from restricted pulmonary or conduit flow, systolic or diastolic failure of the systemic ventricle (especially when the ventricle is morphologically right-sided), and venous thromboembolism. Obstetric complication rates are substantial, including a 48% risk of both delivery before 34 weeks of gestation and small for gestational age infants.[2] Infants born to mothers with congenital heart disease are also more likely to have congenital heart disease. Despite these potential risks, individual reports and