and hematological systems. By contrast, Cobras and kraits release neurotoxic venom that can lead to rapid respiratory paralysis and death. The management of any snake bite should focus on airway, breathing and circulation, and if available, timely administration of antivenom. Surgical debridement and fasciotomy for compartment syndrome are not commonly required.

In conclusion, adverse fetal effects of a venomous snake bite can occur before maternal signs of systemic poisoning. Following a snake bite, the mother and fetus should be observed for at least 12 h, particularly because the snake species and the amount of venom injected can be unclear. More robust reporting of envenomation during pregnancy should be encouraged to improve information on management and maternal and fetal outcomes.

References

Anesthetic management for resection of cor triatriatum during the second trimester of pregnancy

W. Bai, S. Kaushal, S. Malviya, K. Griffith, R.G. Ohye

Department of Anesthesiology; Department of Surgery; Perfusion Services, University of Michigan Health System, Ann Arbor, Michigan, USA

ABSTRACT

Hemodynamic changes during pregnancy can result in cardiovascular decompensation in women with pre-existing cardiac diseases. Despite optimized medical treatment, some patients with severe structural cardiac abnormalities may need surgical intervention during pregnancy. We describe a woman who presented at 20 weeks of gestation with acute heart failure due to cor triatriatum, a rare form of congenital heart disease. This condition is characterized by a perforated fibromuscular membrane dividing the left atrium into two chambers. The clinical presentation varies from asymptomatic to acute heart failure depending on the size of the fenestrations in the membrane and the presence of associated cardiac malformations. In our patient, two severely restricted orifices in a membrane within the left atrium, moderate to severe pulmonary hypertension and good biventricular function were demonstrated by transthoracic echocardiography. Without surgical resection, the increased blood volume and cardiac output associated with pregnancy could have resulted in cardiovascular decompensation. She underwent urgent corrective open heart surgery with cardiopulmonary bypass. Perioperative anesthetic management included prevention of tachycardia, atrial dysrhythmias and pulmonary hypertension, close monitoring for and prompt treatment of maternal hypotension, maintaining euvoolemia and good cardiac contractility and avoiding hemodilution and hypothermia. These approaches, together with minimizing bypass time, resulted in successful maternal and fetal outcome.

Keywords: Anesthesia; Cor triatriatum; Pregnancy; Cardiopulmonary bypass

Accepted April 2009
Correspondence to: Wenyu Bai, MD, Department of Anesthesiology, Section of Pediatric Anesthesiology, F3900 C.S. Mott Children’s Hospital, 1500 E. Medical Center Dr, SPC 5211, Ann Arbor, MI 48109-5211, USA. Tel.: +1 734 763 2455; fax: +1 734 763 6651.
E-mail address: wenyubai@med.umich.edu

Introduction

Cor triatriatum is a rare form of congenital heart disease that is usually diagnosed in infancy or childhood. It is
characterized by a perforated fibromuscular membrane dividing the left atrium into two chambers. Occasionally imperforate, this membrane usually contains fenestrations ranging from small restrictive openings to large wide-open defects. Typically, the proximal or superior chamber receives the pulmonary venous blood while the distal or inferior chamber contains the atrial appendage and connects with the mitral valve. In children, cor triatriatum is frequently associated with a variety of major cardiac anomalies, such as anomalous pulmonary venous drainage, atrioventricular septal defect or tetralogy of Fallot. When cor triatriatum presents for the first time during adulthood, it is usually an isolated anomaly or associated with minor cardiac defects such as an atrial septal defect or persistent left superior vena cava. The clinical presentation depends on the size of the fenestrations in the membrane and the presence of associated cardiac malformations.

Patients with restrictive membrane openings present earlier, with a clinical picture similar to patients with mitral stenosis. Children with less restrictive membrane openings or mild malformations may survive into adulthood without any symptoms until substantial hemodynamic changes occur. During pregnancy significant increases in blood volume and cardiac output can result in cardiovascular decompensation in women with pre-existing cardiac diseases. Despite optimized medical treatment, some patients with significant cardiac structural abnormalities may need surgical intervention during pregnancy. We describe a woman at 20 weeks of gestation with no known cardiac history who presented with acute heart failure secondary to cor triatriatum and required urgent corrective open heart surgery with cardiopulmonary bypass (CPB).

**Case report**

A 20-year-old pregnant patient (G1P0) at 20 weeks of gestation presented to the pediatric cardiology clinic with progressive exercise intolerance, orthopnea, paroxysmal nocturnal dyspnea, and occasional chest pain that started at the beginning of the second trimester. The patient was previously healthy with no known history of cardiovascular disease. Cardiovascular examination revealed normal S1, narrowly split S2, and a grade II/VI systolic murmur at the left sternal border. Transthoracic echocardiography demonstrated a fibromuscular membrane with two severely restrictive orifices within the left atrium, consistent with cor triatriatum (Fig. 1). The mean gradient across her cor triatriatum membrane was 17 mmHg. There were no other cardiac anomalies, although the patient was noted to have moderate to severe pulmonary hypertension with an estimated pressure of 65 mmHg, mild right ventricular enlargement, and good biventricular function. Her electrocardiogram showed normal sinus rhythm with possible right ventricular hypertrophy. Fetal ultrasound examination revealed an apparently healthy fetus with a heart rate consistent with gestational age. Although counseled by her cardiologist, obstetrician and cardiothoracic surgeon that her current cardiac status required urgent repair and placed her and her fetus at significant risk for adverse events, the patient desired to continue her pregnancy. Medical therapy such as supplemental oxygen, furosemide and digoxin were reserved for hemodynamic instability before surgery, but not instituted.

On the day of surgery at 20+6 weeks of gestation, her temperature was 35.5°C, heart rate 75 beats/min, respiratory rate 16 breaths/min, blood pressure 94/64 mmHg and oxygen saturation (SpO₂) 99% on room air. Her weight was 54 kg with her height 169 cm. There were no signs of heart failure, including no jugular venous distension, hepatomegaly, or peripheral edema. Her hematocrit was 33%. Fetal heart rate (FHR) by an external fetal monitor was 150 beats/min.

In the operating room with the patient supine with left uterine displacement, SpO₂, cardiac rhythm and blood pressure were monitored. After rapid-sequence induction and intubation with fentanyl, lidocaine, etomidate, and succinylcholine catheters were placed for central venous and arterial pressures. Anesthesia was maintained with isoflurane, equal mixtures of oxygen and air, continuous morphine infusion and vecuronium for muscle relaxation. Volume expansion with Ringer’s lactate 5-10 mL/kg and ephedrine 5-10 mg were administered before CPB to maintain the mean arterial pressure above 65 mmHg. Cardiographic monitoring was not used intraoperatively because of potential technical difficulties due to frequent fetal movements and CPB.

Intravenous heparin 22 000 units was administered and bicaval CPB was initiated with mean flows of 3.5 L min⁻¹ m⁻²; mean arterial pressure was maintained at 65-70 mmHg and the hematocrit between 30-34%.

![Fig. 1](image-url)
Shortly after initiation of CPB, maternal mean arterial pressure decreased to 50 mmHg; it was treated with phenylephrine 100 μg and increased pump flow. After resection of the membrane through the left atrium, maternal cardiac activity resumed spontaneously and the patient was weaned from CPB with dopamine 5 μg·kg⁻¹·min⁻¹. Transesophageal echocardiography confirmed total resection of the abnormal structure with unobstructed blood flow in the left atrium. Aortic cross-clamp time was 9 min and CPB time was 18 min; normothermic CPB was maintained throughout.

After surgery, the patient was transferred to the cardiac surgery intensive care unit in stable hemodynamic condition; the FHR was 150 beats/min. She was weaned off the dopamine infusion and extubated 2 h postoperatively. The FHR was monitored for the first postoperative day because of the potential fetal risks from maternal hemodynamic instability, abnormal coagulation and fetal bradycardia from maternal opioid analgesia. The patient had no uterine contractions and normal FHR values. She was discharged home on postoperative day 4 with oral pain medications. She continued her pregnancy uneventfully. A healthy infant was delivered at term with labor pain medications. She continued her pregnancy uneventfully.

Discussion

Cor triatriatum is a rare cardiac lesion accounting for only 0.1% of all congenital heart defects; it is usually diagnosed in early childhood. It is characterized by an incomplete fibromuscular membrane that persists between the common pulmonary veins and the body of the left atrium, creating an accessory chamber or essentially a third atrium. Pulmonary venous flow is obstructed by the restrictive opening(s) in the membrane and elevated pressure in the left atrial accessory chamber. Right ventricular dysfunction and pulmonary arterial hypertension may follow. It may be an isolated defect in 30% cases or associated with other cardiac defects. Depending on the severity of obstruction to the pulmonary venous return, or orifice size through the membrane or presence of other cardiac anomalies, cor triatriatum usually presents in early childhood as recurrent upper respiratory infections, dyspnea, congestive heart failure and failure to thrive. Echocardiography is the primary diagnostic modality for diagnosing cor triatriatum and identifying associated anomalies.

Surgical excision of the intra-atrial membrane is the preferred treatment for cor triatriatum. CPB, hypothermia, aortic cross-clamping and cardioplegia are used to facilitate surgical repair. Long-term results usually are excellent in patients without residual obstruction or other cardiac defects. However, children with mild malformations may survive to adulthood without any symptoms until substantial hemodynamic changes occur, such as during pregnancy, labor and post partum. Physiologic changes of pregnancy including increased plasma volume, heart rate, and cardiac output are of particular concern in parturients with cor triatriatum. Labor and delivery are stressful events causing tachycardia and acute right heart failure in these patients. Acute pulmonary edema and atrial fibrillation have been reported during pregnancy and the immediate postpartum period with previously undiagnosed cor triatriatum. Medical treatments such as diuretics, β-adrenergic blocking agents and digitalis glycosides have been successfully used to relieve symptoms temporarily. However, patients who progress to congestive heart failure, pulmonary hypertension or atrial fibrillation despite medical management are likely to experience worsening symptoms and hemodynamic decompensation toward the end of gestation, during labor and immediately post partum. In such patients and in those with severe mechanical obstruction, surgical intervention during pregnancy is mandatory.

Although no large series of open heart surgery involving CPB during pregnancy has been published, cumulative case reports and reviews have shown a consistently low maternal but high fetal mortality of 0-3% and 20-24%, respectively. In patients with cor triatriatum and pulmonary hypertension, anesthetic management should minimize tachycardia, atrial dysrhythmias and pulmonary hypertension, and maintain euvoolemia and good cardiac contractility. Mean arterial pressure should be maintained between 65 and 70 mmHg.

Fetal distress with resultant bradycardia is prone to develop at initiation of CPB, and is presumably induced by hypotension, hypothermia, placental hypoperfusion, and non pulsatile uterine blood flow. Clinically, various strategies have been described and used as protective measures to maintain fetal viability during CPB including maintenance of high perfusion pressure in the range of 65-70 mmHg, high flow rate, and avoidance of hypothermia. These strategies were used successfully in our case. Crystalloid hemodilution combined with mild hypothermia have also been previously used with success, despite concerns related to fetal distress with low maternal hematocrit. Sutton et al. suggest that maintenance of higher hematocrits of 33-35% with normothermic CPB may be ideal to optimize oxygen transportation to the fetus. A target hematocrit above 30% was chosen for our patient.

Cardiotocographic monitoring throughout these cases could be beneficial for early detection of fetal distress and uterine contractions, and may facilitate appropriate and timely CPB interventions. However, such monitoring may be difficult due to frequent fetal movements, particularly during the second trimester. Since a short CPB time, normothermia and high pump flows were anticipated in our case, fetal monitoring was not used during the surgical procedure. Nonetheless, maternal hypotension after initiation of CPB was treated promptly with phenylephrine and an increase in pump flow. Fetal heart tones were closely monitored in the postoperative period.
In summary, successful cardiac operations requiring CPB have been reported during pregnancy, but experience is still limited. In this parturient undergoing repair of cor triatriatum, successful perioperative management included prevention of tachycardia, atrial dysrhythmias, and pulmonary hypertension, close monitoring for and prompt treatment of maternal hypotension, high CPB index, and avoidance of hemodilution and hypothermia. These strategies, as well as minimizing aortic cross clamp and CPB time, were used with successful maternal and fetal outcomes.

Acknowledgement

We thank Dr. Gregory Ensing for his echocardiographic assistance.

References


Anesthetic management of a patient with cleidocranial dysplasia undergoing various obstetric procedures

A. Ioscovich, A. Barth, A. Samueloff, S. Grisaru-Granovsky, S. Halpern

ABSTRACT

Patients with cleidocranial dysplasia, a rare autosomal dominant genetic syndrome, possess abnormal anatomical features of the head, mouth, neck and spinal column. These features may result in perioperative problems such as difficult airway and complicated regional anesthesia. We report the anesthetic management of a young woman with cleidocranial dysplasia undergoing four caesarean sections, one vaginal delivery and a dilatation and curettage, employing different modes of anesthesia. Anesthetic management in this disorder presents challenges for both general and neuraxial anesthesia.

Keywords: Anesthesia; Pregnancy; Cleidocranial dysplasia; Marie-Sainton syndrome, Uncommon diseases

Introduction

Cleidocranial dysplasia (CCD) is a rare autosomal dominant disease that has also been called cleidocranial dysostosis, mutational dysostosis, osteodental dysplasia or Marie-Sainton syndrome. CCD is characterized by abnormal clavicles, patent cranial sutures and fontanels, supernumerary teeth, a high-arched palate, delayed