Emergency cesarean section in a patient with familial hypertrophic cardiomyopathy

Andrew C Leatherbarrow, Laura A Beard

Abstract

Introduction: Familial hypertrophic cardiomyopathy (HCM) is a genetic disorder affecting approximately 1 in 500 of the population. It is also the leading cause of sudden cardiac death in young adults. It is uncommonly encountered in the obstetric patient cohort and poses a number of challenges for the obstetric anesthetist.

Case Report: We report on a rare case where a 31-year-old Caucasian patient with known HCM presented for emergency cesarean section. The patient had a significant unprovoked gradient of 144mmHg across the left ventricular outflow tract. We discuss the pathophysiology of HCM. This includes how the disease process can produce a pathologically elevated left ventricular ejection fraction which may be falsely reassuring to clinicians. We describe how we managed general anesthesia for this case and discussed in detail the principles of managing anesthesia for emergency cesarean section in this high-risk patient cohort.

Conclusion: The key to successful management of anesthesia for cesarean section in the presence of HCM is meticulous attention to the maintenance of hemodynamic stability. Avoidance of hypovolemia, tachycardia, and excessive vasodilatation is crucial to safely manage this patient cohort.

Keywords: Cardiomyopathy, hypertrophic, cesarean, anesthesia.

Introduction

Hypertrophic cardiomyopathy (HCM) was initially described by Teare in 1958 [1]. Although the hereditary nature of the disease became apparent in the 1960s, it was not until 1990 when the first causative genetic defect was elucidated [2, 3]. It affects approximately 1 in 500 of the population and is the leading cause of sudden cardiac death in young adults [4]. An average sized obstetric unit should therefore expect to deliver patients with HCM each year. Worryingly, these patients are likely to be undisgnosed carriers of the disease. Although HCM may be diagnosed following presentation with classical symptoms including syncopal episodes and palpitations, the majority of cases are diagnosed following screening of relatives of affected individuals.

Pregnancy poses a significant physiological challenge to this cohort of patients. Systemic vascular resistance falls by around 30–70% in later pregnancy which exacerbates the gradient across the left ventricular outflow tract. Blood volume increases by 40–50% which may overload a poorly functioning hypertrophic ventricle. Cardiac output increases by around 50% which may lead to myocardial ischemia and arrhythmias in the presence of significant hypertrophy. It is, therefore, unsurprising that a recent case series described 74% of patients developing worsening
symptoms and 13% developing congestive cardiac failure in later pregnancy [5].

We report on a rare case where a patient with known HCM and a significant left ventricular outflow tract gradient of 144mmHg presented for emergency cesarean section. We discuss how the pathophysiology of HCM influences the management of anesthesia for cesarean section and describe how we successfully managed anesthesia for this patient.

**Case Report**

A 31-year-old multiparous patient with a background of familial hypertrophic obstructive cardiomyopathy (HCM) was admitted to the delivery suite at 35 weeks gestation in her sixth pregnancy.

The patient's first four pregnancies had ended with uneventful normal vaginal deliveries at term. Late during her fifth pregnancy, HCM was diagnosed through echocardiographic screening following a diagnosis in the patient's mother. She delivered vaginally, but labor was complicated by the development of cardiac failure. In view of this, an elective cesarean section was planned for her sixth pregnancy.

On the day of her delivery the patient attended maternity triage due to reduced fetal movements. Doppler ultrasound assessment of the umbilical artery demonstrated reversal in end-diastolic flow which carried a significant risk of perinatal mortality. The obstetricians decided to perform a category three cesarean section. The patient did not complain of any chest pain, palpitations, or syncopal episodes although she had noticed a gradual decline in exercise tolerance throughout the third trimester. Clinical examination and vital signs were normal. A 12-lead electrocardiograph (ECG) showed a left ventricular strain pattern in precordial leads V4-V6, and met voltage criteria for the left ventricular hypertrophy. Serial transthoracic echocardiography during pregnancy had shown asymmetrical septal hypertrophy with a ventricular wall thickness of 2cm. Systolic anterior motion (SAM) of the mitral valve was also present along with mild mitral regurgitation. The left ventricular ejection fraction was 74%. The gradient across the left ventricular outflow tract had been gradually worsening as pregnancy progressed. At 35 weeks gestation, the gradient was measured 144mmHg (normal gradient <30mmHg). Antenatally, the patient had been reviewed by the cardiology team and started on bisoprolol 2.5mg and aspirin 75mg daily. The patient had never had a general anesthetic before and reported no family history of problems with general anesthesia. Airway assessment revealed no concerns.

Following discussion with both cardiologist and cardiac anesthetist, a decision was made to proceed under general anesthesia due to the extreme gradient present. Premedication consisted of ranitidine 150mg PO on delivery suite followed by 30ml of sodium citrate 0.3M when the patient arrived in theatre. Venous access consisted of two 16G peripheral venous cannulae along with a radial arterial line. A central venous catheter was not inserted as we were concerned about the risk of inadvertently inducing a cardiac arrhythmia during guidewire insertion. Cardiac arrhythmias are poorly tolerated in patients with HCM and can lead to rapid cardiovascular collapse. Other measures to minimize the risk of cardiac arrhythmias included the initiation of beta-blocker therapy antenatally and IV potassium replacement pretheater as her level was at the lower end of normal at 3.5mmol/l. We also had a defibrillator on hand in theater in the event of an arrhythmia. Once in theater, a modified rapid sequence induction was performed using cardiodegradable induction agents. A remifentanil target-controlled infusion (Minto model/C targeting) was slowly titrated up to an effect site concentration of 3ng/ml while the patient was preoxygenated. We utilized remifentanil to smooth induction and obtund the sympathetic response to intubation, especially to avoid a tachycardia, which is poorly tolerated in patients with HCM. This was followed by etomidate 0.3mg/kg and suxamethonium 100mg. Intubation was uneventful. Maintenance of anesthesia was with 50% nitrous oxide/50% oxygen and sevoflurane at 1.2% end tidal concentration. 15mg of atracurium was administered to maintain neuromuscular blockade.

The patient required a total of 2mg phenylephrine which was administered as 100µg boluses to maintain systolic blood pressure. The oxytocin bolus was omitted due to the risk of rapidly decreasing systemic vascular resistance. The 1unit/ml postnatal infusion was run at an accelerated rate of 60ml/h until 5 units were delivered, and then, the infusion rate was reduced to 10ml/h. Estimated blood loss was 300ml. A total of 1.5l of Hartmann’s solution were infused intraoperatively. During emergency and extubation, we again needed to minimize sympathetic stimulation and tachycardia. We utilized the method of continuing the remifentanil target-controlled infusion at a reduced effect site concentration of 2ng/ml while the sevoflurane washed out. Once verbal communication was made with the patient, she was extubated and the remifentanil infusion was then stopped. This permitted a smooth extubation with avoidance of tachycardia and coughing. Post-operative analgesia was supplemented with ultrasound-guided transversus abdominis plane blocks. The newborn was delivered in excellent condition with Apgar scores of 8 at 1 minute and 10 at 5 minutes. The umbilical vein pH was 7.31 at Birth. Mum was transferred to the high dependency unit (HDU) post-surgery.

**Discussion**

Approximately one-third of patients with a diagnosis of familial HCM will clinically deteriorate in pregnancy [6]. This patient cohort, therefore, requires regular antenatal follow-up in joint obstetric and cardiology clinics with early anesthetic assessment.

Regarding pre-operative assessment, there are several key aspects to assess in pregnant patients with HCM in addition to the standard anesthetic history and examination. First, it is important to determine whether the patient has symptoms of HCM and their severity. Patients may complain of syncopal episodes, palpitations, dyspnea, or angina. Symptoms may have developed in pregnancy or if preexisting could have worsened. This may indicate the hypertrophic myocardium is obstructing the left ventricular outflow or the patient is experiencing dangerous arrhythmias. The presence of these features places the patient in a high-risk category. Patients with prior episodes of arrhythmias may have an
implantable cardioverter-defibrillator fitted which poses its own management issues. The patients exercise tolerance should also be noted and medication should also be reviewed. Beta-blocker therapy will usually have been initiated by the cardiology team as the reduction in contractility improves cardiac output by reducing the gradient and in addition reduces myocardial oxygen requirements in the hypertrophied myocardium. The patient may also be on other antiarrhythmic agents.

Serial echocardiogram studies should also be reviewed. The degree of hypertrophy should be noted. A left ventricular wall thickness of 1.2 cm is normal. If > 1.5 cm, this is abnormal and one of the diagnostic features of HCM (providing there is no other explanation for the hypertrophy, e.g., severe hypertension). Over 3 cm represents severe hypertrophy and is a major risk factor for sudden cardiac death [7]. The gradient across the left ventricular outflow tract should also be noted. This is normally < 30 mmHg. A gradient above 30 mmHg is significant and above 50 mmHg is severe. Echocardiography may also demonstrate the phenomena of SAM of the mitral valve and associated mitral regurgitation. SAM is thought to occur due to a venturi effect induced by the narrowed left ventricular outflow tract [8]. Essentially, the ejected blood velocity increases leading to a pressure drop in the outflow tract. The anterior mitral valve leaflet is pulled by the pressure drop toward the left ventricular outflow tract. This has two consequences. First, this will cause a varying degree of the left ventricular outflow tract obstruction and reduce cardiac output. Second, the mitral valve leaflets will be pulled apart in systole leading to mitral regurgitation. It is important to note that contractility is pathologically elevated due to the septal hypertrophy. This combined with reduced diastolic filling leads to an abnormally high and falsely reassuring ejection fraction (typically > 75%). In our case, the ventricular wall thickness was 2 cm, gradient 144 mmHg, ejection fraction 74%, and SAM was demonstrated which overall placed our patient in a high-risk category.

Regarding intraoperative management, it is important to understand that patients have both systolic and diastolic dysfunctions. Systolic function is impaired due to the left ventricular outflow tract obstruction. This is due to a combination of fixed obstruction from septal hypertrophy and dynamic obstruction if SAM is present. Diastolic dysfunction occurs as the hypertrophied ventricle is stiff and relaxes poorly. The left ventricle is, therefore, dependent on high filling pressures to maintain stroke volume meaning hypovolemia must be avoided. Tachycardia must also be avoided, this will reduce diastolic filling time and further reduce cardiac output. In view of these factors, the use of invasive blood pressure monitoring is crucial.

As mentioned previously, we managed this patient with general anesthesia view of the extreme pressure gradient across the left ventricular outflow tract. We administered a 500 ml crystalloid preload before induction to ensure the patient was well filled. We used remifentanil for this case as it is a cardiostable agent which also has the advantage of inducing a protective bradycardia. The rapid offset of remifentanil also minimizes the risk of neonatal sedation following delivery. Falls in systemic vascular resistance must be avoided as this will worsen the gradient and reduce cardiac output. Due to this, it is important to use cardiostable induction agents and minimize the dose of inhalational anesthetic. Furthermore, for these reasons, spinal anesthesia is particularly hazardous due to the rapid fall in systemic vascular resistance and is not recommended in the presence of HCM. As an alternative to general anesthesia, patients with HCM can be safely managed with a slow incremental epidural top-up. This provides the advantages of avoiding complications of general anesthesia and improved pain control in the post-operative period. The advantages gained by improved pain control and reduced sympathetic stimulation need to be carefully balanced against the risks of hypotension and exacerbating the gradient across the left ventricular outflow tract. Falls in systemic vascular resistance must be minimized through up-titration of a vasopressor infusion as the epidural block develops. A major disadvantage of incremental epidural top-up is the inability to reverse the physiological effects of the epidural should the patient deteriorate.

In our case, we elected not to insert a central venous catheter due to the risk of causing an arrhythmia during insertion. Any arrhythmia will compromise atrial ejection and lead to rapid decompensation into cardiac failure. Tachyarrhythmias are particularly dangerous in patients with HCM and can lead to irreversible cardiovascular collapse.

Careful consideration must be given to the use of vasoactive drugs to maintain systemic vascular resistance. Agents with positive inotropic and chronotropic properties must be avoided as they will worsen the outflow tract gradient and reduce diastolic filling time. Phenylephrine with its pure alpha agonist effect is the optimal vasopressor, whereas ephedrine with its sympathomimetic properties should be avoided. Care must also be taken when administering utoeretic drugs post-delivery. Oxytocin causes a rapid fall in systemic vascular resistance and tachycardia when given as a bolus [9]. In the presence of HCM, both side effects will severely compromise cardiac output. Therefore, the oxytocin should be administered as an accelerated infusion. Ergometrine is also best avoided as it induces coronary artery vasospasm which would exacerbate an already ischemic hypertrophied ventricle [10]. Uterotonic prostaglandins including carboprost and misoprostol are generally contraindicated in the presence of cardiac disease as they can exacerbate hypertension and can rarely cause pulmonary edema. Although in the presence HCM and ongoing hemorrhage due to uterine atony, the potential risks may be outweighed by that of the need to control ongoing hemorrhage. Strategies may include administering reduced doses of ergometrine and prostaglandins or utilizing non-pharmacological methods of control. Balloon tamponade is the first-line intervention in our institution following failure of pharmacological control as it is least invasive and can be deployed rapidly. Second-line methods include insertion of uterine sutures and interventional radiology techniques such as internal iliac balloons.

Postoperatively, patients with HCM must be observed in a high dependency environment with continuous ECG monitoring. Analgesia must be titrated to avoid pain-mediated tachycardia. Our patient was monitored in HDU for 36 h before discharge to the postnatal ward.
Conclusions

HCM poses a number of challenges to the obstetric anesthetist. A ventricular wall thickness >3 cm, outflow tract gradient >50 mmHg, and the presence of SAM of the mitral valve indicates severe disease and high risk of sudden death. The key to successful management of anesthesia for cesarean section in the presence of HCM is meticulous attention to the maintenance of hemodynamic stability. Avoidance of hypovolemia, tachycardia, and excessive vasodilatation is crucial to safely manage this patient cohort.

References


Conflict of Interest: Nil. Source of Support: None

How to Cite this Article