

Short Communication

Considerations for delivery in pregnancies complicated by maternal hypertrophic obstructive cardiomyopathy

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Abstract

We present the management of a pregnancy complicated by maternal hypertrophic obstructive cardiomyopathy (HOCM), an uncommon but significant cardiac lesion. While it is accepted practice to manage such cases in a tertiary setting with multidisciplinary involvement, we reflect that the ultimate management of the birth should centre on obstetric principles. A discussion and literature review of HOCM, pregnancy and anaesthetic management is provided.

Key words: cardiac disease, complications, hypertrophic obstructive cardiomyopathy, pregnancy.

Introduction

There have been several reports of pregnancies in women with hypertrophic obstructive cardiomyopathy (HOCM).^{1–3} The potential for cardiac deterioration and overt cardiac failure remain the cause for concern within the antenatal, intrapartum and postnatal periods.² Concerns regarding maternal decompensation during pregnancy and birth have led to an increase in operative delivery rates in women with HOCM. These interventions have occurred primarily to avoid labour stresses and catecholamine release, with the potential compromise of cardiac output from increased heart rate and impaired myocardial contractility.⁴

A case is presented of a pregnancy in a woman with known HOCM whose management centred on a combined medical approach with obstetric physician, cardiology and obstetric and anaesthetic involvement. On cardiological advice, her delivery was managed by Caesarean section that unexpectedly resulted in increased morbidity from surgical complications. It has been suggested that the frequency of Caesarean birth in these cases reflects excessive caution by medical practitioners rather than evidence-based management.²

In a cohort of 40 pregnant women with hypertrophic obstructive cardiomyopathy, Autore reported 68% of patients diagnosed with this condition pre-pregnancy were managed with Caesarean delivery compared with 10% of women in whom the diagnosis was made during or after pregnancy.² Clinical deterioration or the development of adverse cardiac symptoms was uncommon, occurring in 15% of women who were asymptomatic prior to pregnancy. In a study of 127 pregnant women symptom deterioration was observed in fewer than 10% of cases.¹ Similarly, a British study of 54

pregnancies in 23 women with HOCM reported good maternal outcomes in hypertrophic cardiomyopathy.³

Hypertrophic obstructive cardiomyopathy is a myocardial disorder characterised by left ventricular hypertrophy. It is now diagnosed more frequently, secondary to increased clinical awareness and familial screening.¹ It is increasingly being found to occur in an autosomal dominant disorder with variable degrees of penetrance.³ The disorder has been estimated to occur in 0.05–0.2% of the general population, yet forms around 0.5% of the outpatient population referred for echocardiography.

Because of the rigidity of the hypertrophic myocardium, ventricular filling is critically dependant on adequate preload and the maintenance of sinus rhythm.^{2,4} Disease severity is in direct proportion to the degree of left ventricular outflow obstruction. Additionally, there may be functional outflow obstruction arising from the hypertrophied ventricular tissue. Tachycardia or a reduction in afterload may compromise perfusion of the hypertrophic myocardium.

Symptomatology in those affected with hypertrophic obstructive cardiomyopathy is extremely variable. Some patients with hypertrophic obstructive cardiomyopathy may be asymptomatic. Those with symptoms can develop dyspnoea, orthopnoea, and paroxysmal nocturnal dyspnoea as a result

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of congestive heart failure. Arrhythmia may occur causing syncope, palpitations and dizziness or even sudden cardiac death.

Hypertrophic obstructive cardiomyopathy has been classified into different functional classes according to the New York Heart Association (NYHA).⁵ Autore reported the deterioration of symptomatology correlated with the degree of functional impairment as accorded by the NYHA functional class.² However, no single clinical or echocardiographic feature correlated with adverse events in pregnancy.¹

Case report

A 27-year-old woman diagnosed with HOCM at age 19, was referred to our institution for prenatal care in her first pregnancy. Her father and sister were also affected, with her sister dying at age 26 from peripartum complications of HOCM and her father at age 32 from a cardiac arrhythmia. Echocardiography at 16-weeks gestation demonstrated HOCM with a severe resting gradient of 146 mmHg, moderately severe mitral regurgitation and moderate pulmonary hypertension with an estimated systolic pulmonary artery pressure of 53 mmHg. There was normal left ventricular size with asymmetrical septal hypertrophy involving the inferoseptal, anteroseptal and anterior walls. The septal thickness was 22 mm. As the extent of myocardial hypertrophy was widespread and not restricted to the interventricular septum, therapies such as surgical septal ablation were inappropriate. She had an implantable cardioverter defibrillator for primary prevention against malignant ventricular arrhythmia inserted 4 years ago. The medical therapy of atenolol and verapamil were maintained throughout the pregnancy and cardiology functional class grading was class II according to the NYHA functional criteria.

Regular physician and cardiology review were provided throughout the antepartum period in addition to obstetric care. During the course of the pregnancy there was increasing fatigue, particularly in the third trimester. There was no dyspnoea or clinical evidence of cardiac failure. Fetal growth was appropriate to clinical and ultrasound assessment. Her cardiology team advised delivery by elective Caesarean birth in order to avoid potential cardiovascular complications during labour.

At 38 weeks gestation, Caesarean delivery occurred at a general tertiary hospital with consultant anaesthetist and cardiologists present. Prior to delivery the defibrillator was inactivated to avoid discharge during diathermy. Pre-operatively arterial and central venous catheters were inserted and cardiac output was monitored intraoperatively with *trans*-thoracic ultrasound. Anaesthesia for the delivery was provided with a combined spinal epidural technique. With the patient sitting erect, 7.5 mg hyperbaric 0.5% bupivacaine and 15 µg fentanyl were administered intrathecally. This was followed 15 min later by the administration of 5 mL plain bupivacaine 0.5% via the epidural catheter on three occasions at 5 min intervals. A phenylephrine infusion (100 µg/mL at 50 µg/min) was commenced at the time of the spinal anaesthetic as prophylaxis against hypotension. This infusion was increased to 75 µg/min

just prior to surgery. There was meticulous attention to afterload maintenance with the assistance of *trans*-thoracic echocardiography, central venous and arterial pressure measurements, permitting titration of crystalloid to maintain the left ventricular volume. This regimen was consistent with other documented cases of managed delivery of patients with HOCM.⁶

The Caesarean section proceeded without incident, with a live male infant of birth weight 3200 g delivered. Extreme sensitivity to intravenous oxytocin occurred. A marked decline in systolic blood pressure (from 130 to 75 mmHg) occurred following the administration of 1 IU of oxytocin via a slow intravenous bolus. Echocardiography demonstrated an increase in left ventricular contractility. The phenylephrine infusion was transiently increased correcting the hypotension. Subsequently a further four 1-unit boluses of oxytocin were given for a total of 5 IU, without further haemodynamic compromise. An oxytocin infusion was then commenced at 3.6 IU/h. Prophylactic antibiotics were given to protect against endocarditis and surgical infectious morbidity.

At post-procedure she experienced chest pain. The ECG, serum troponin and arterial blood gases were normal. The day following delivery, her haemoglobin dropped from 115 to 50 g/L, necessitating transfusion of 5 units of packed cells. Ultrasound imaging revealed a large haematoma in the left broad ligament requiring radiological embolisation of the internal iliac artery to achieve haemostasis. Subsequent postpartum progress was satisfactory and an intrauterine progesterone device for contraception was recommended. There were several discussions antepartum regarding future pregnancies and tubal ligation offered at the time of Caesarean delivery; however, the patient declined this intervention. Paediatric cardiology follow up was recommended in view of the risk for inheritance of this autosomal dominant condition and the mother has ongoing medical supervision by her cardiologist.

Two months post-delivery she developed recurrent atrial fibrillation and secondary hypotension necessitating the commencement of amiodarone therapy.

Discussion

This case and the literature illustrate a number of salient points. Hypertrophic obstructive cardiomyopathy is being diagnosed more frequently, as a result of increasing awareness of the clinical syndrome and sophisticated imaging technology. With increased incidence occurring through better diagnosis, obstetricians are likely to encounter women with this diagnosis. An understanding of the interaction of pregnancy and the cardiac disorder is important to optimise outcome for the mother and her fetus.

Whilst it is reasonable to have a planned delivery procedure in this high risk circumstance, the Caesarean birth was principally selected on cardiology advice, seeking to minimise cardiovascular complications from labour. Whilst there is no evidence to indicate this woman was at a greater risk of broad ligament haematoma than any other, significant cardiovascular risk eventuated as a consequence of surgical complications from the procedure recommended to minimise

maternal morbidity – namely postpartum haemorrhage and anaemia. She tolerated the postoperative cardiovascular insult and combined with her uncomplicated antepartum course, in retrospect a vaginal delivery may have been a preferable option. Despite the clear need for specialist involvement in management of complex medical cases, it ultimately behoves the obstetrician to make decisions on a risk assessment basis regarding mode of delivery. On occasions this can prove difficult when non-obstetric practitioners provide a recommendation based on well-meaning, but incomplete knowledge of obstetric outcomes. Caesarean delivery, whilst in general a safe procedure, is associated with a variety of surgical complications which may occur despite all due surgical precautions and these adverse occurrences must be considered in the decision-making process for delivery.

Complexity of care in modern obstetrics is not uncommon. Obstetricians and physicians often work together using their respective skills in monitoring and managing pregnant patients with coexisting medical conditions such as autoimmune disease or thrombophilia.⁷ The management of pregnant women with pre-existing complex cardiac problems should ideally be undertaken by multidisciplinary teams in tertiary centres.

Much interest in HOCM has been generated in anaesthetic specialities where, in view of potential hypotensive effects of regional anaesthesia, this analgesic modality has been considered controversial.⁴ An additional concern for HOCM is the potential for fatal cardiac arrhythmia in the perinatal period. Minimising the cardiovascular stresses whilst maintaining adequate analgesia is critical issues for anaesthetists. The ventricular outflow obstruction must be minimised, whilst avoiding reduction in preload and afterload that may occur from sympathetic blockade caused by regional anaesthesia.⁴ The need for invasive monitoring is guided by individual case details however, is generally recommended.⁸ Prophylactic antibiotics both in labour and postnatal are recommended.²

Shortening of second stage by instrumental delivery is frequently employed to minimise cardiovascular compromise during labour in women with pre-existing cardiac disease. The maternal cardiac status should be optimised preoperatively whenever possible and planned elective delivery is preferable. Vaginal delivery is preferable when feasible, and, with careful incremental regional anaesthesia, is safe in most women with cardiac disease.⁹

A small dose of oxytocin resulted in haemodynamic instability in this case. Oxytocin is known to have negative inotropic and chronotropic effects mediated by cardiac oxytocin receptors.¹⁰ Intravenous oxytocin produces a biphasic dose-dependent change in mean arterial pressure. An initial pressor response accompanied by bradycardia and reduced cardiac output, is followed by a prolonged fall in mean arterial pressure accompanied by an increased cardiac output.¹¹ Oxytocic administration should be finely titrated and administered in small boluses as hypotension is typical in this situation.³ The cardiovascular effects of oxytocin are brief but if compensatory response cannot be effected may be disastrous. In retrospect it may have been preferable to have used an infusion alone and not mini-bolus oxytocin.

Pregnancy in a woman with hypertrophic cardiomyopathy is usually well-tolerated without an increase in sudden death.¹² This case, involving a woman with serious hypertrophic disease, discusses a combined approach involving obstetric, cardiology and anaesthetic teams and illustrates the need for obstetric guided management, with the literature and our experience suggesting that planned vaginal delivery may be a suitable mode of delivery. This case also demonstrates that severe left ventricular outflow tract obstruction and pulmonary hypertension complicating HOCM is not an absolute contraindication to a successful pregnancy outcome.

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