

Graded epidural anaesthesia for Caesarean section in a parturient with Shone's syndrome: a case study

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Pregnancy with underlying heart disease is a unique challenge both to the obstetrician and the anesthesiologist. Asymptomatic women with mild to moderate single lesions can successfully carry a pregnancy to term and undergo vaginal delivery. However, pregnancy can result in rapid clinical deterioration, which may lead to maternal and/or foetal mortality in symptomatic patients with complex heart diseases, like Shone's syndrome. A thorough understanding of the impact of pregnancy on the haemodynamic response to the patient's cardiac lesion is required for the management of labour and delivery. A meticulous approach is needed when planning anaesthesia for Caesarean section in such a case as the associated haemodynamic effects of both regional and general anaesthesia can have a serious deleterious effect on both the mother and infant.

We report on the successful management of a parturient known to have Shone's syndrome undergoing Caesarean section under graded epidural anaesthesia.

Keywords: Caesarean section, epidural anaesthesia, pregnancy, Shone's syndrome

Introduction

Shone's syndrome is a rare congenital cardiac condition comprising multiple obstructive left-sided cardiac lesions involving the supralvalvular mitral ring, parachute deformity of the mitral valve, subaortic stenosis and coarctation of the aorta.¹ It is an extremely rare syndrome, for which there is a guarded prognosis.²⁻⁴ The continuation of pregnancy, and the management of labour and delivery, are very challenging owing to the associated haemodynamic effects.

We successfully managed a parturient known to have Shone's syndrome undergoing Caesarean section under epidural anaesthesia.

Case study

A 26-year-old primigravida, weighing 65 kg with a height of 155 cm and known to have Shone's syndrome, was referred to our hospital at 36 weeks' gestation, for management of the delivery of her infant. The patient had been diagnosed with Shone's syndrome at the age of five, when she first developed symptoms in the form of fatigue and dyspnoea on exertion. An echocardiography was performed at that time and revealed mitral stenosis, aortic stenosis with subvalvular aortic membrane and coarctation of the aorta. Aged 10, the patient underwent corrective surgery, which involved the placement of a Dacron patch for the coarctation, and excision of the subvalvular aortic membrane. She was found to be free of any overt manifestations during regular medical reviews for several years. Her growing years were normal, and she was symptom free from conception until the first trimester of her pregnancy. However, with the onset of the second trimester, she started to experience palpitations, fatigue and breathlessness, even on mild exertion. She consulted a private physician, and was prescribed 20 mg furosemide tablets once daily, which improved her condition.

She was then referred to our facility for the management of her delivery.

On first presentation, she was comfortable at rest, but dyspnoeic on mild exertion. Her heart rate was 82 beats per minute (bpm) at rest, blood pressure (BP) in the supine position was 136/80 mmHg in the right upper limb, 126/70 mmHg in the left upper limb, and 120/70 mmHg in both lower limbs. On auscultation in the mitral area, a systolic and a diastolic murmur was heard, as well as a systolic murmur in the aortic area, which was transmitted to the right carotid artery. The electrocardiogram (ECG) revealed a sinus rhythm and left ventricular hypertrophy. The ECG (Table 1) showed mitral regurgitation (Figure 1), a bicuspid aortic valve with subaortic membrane (Figure 2), severe aortic stenosis (Figure 3) and coarctation of the descending aorta (Figure 4). Based on these findings, a diagnosis of Shone's syndrome and rheumatic heart disease with recoarctation of the aorta was made. An obstetric examination revealed a single viable foetus of 36 weeks, with a foetal heart rate of 140 bpm.

Foetal cardiography was performed twice a week from the day of admission. During one of these cardiography episodes, beat-to-beat variability of < 5, with a baseline heart rate of 130 bpm, and two decelerations in half an hour, were observed by the obstetrician. As the pregnancy was close to term (37 weeks), the managing team, comprising an obstetrician, anaesthesiologist in charge and cardiologist, opted for delivery by Caesarean section. A graded epidural technique with invasive haemodynamic monitoring was planned because a subarachnoid block and general anaesthesia for Caesarean section are both more likely to induce cardiovascular instability.

Prophylactic antibiotics to guard against endocarditis were given one hour before surgery. Lactated Ringer's solution and

Table 1: The patient's echocardiography report

Mitral valve	Thickened, with diastolic doming present A mitral valve area of 2.17 cm ² Grade III regurgitation
Aortic valve	Thickened and bicuspid valve with severe stenosis A peak gradient of 80.4 mmHg, maximum velocity across the valve of 4.5 m/second A cusp opening of 15 mm Subaortic membrane present
Left atrium	Dilated, with a left atrial diameter of 52 mm
Left ventricle	Concentric left ventricular hypertrophy Left ventricular internal diameter during diastole of 50.3 mm
Aorta	Coarctation of the descending aorta A peak gradient of 37 mmHg

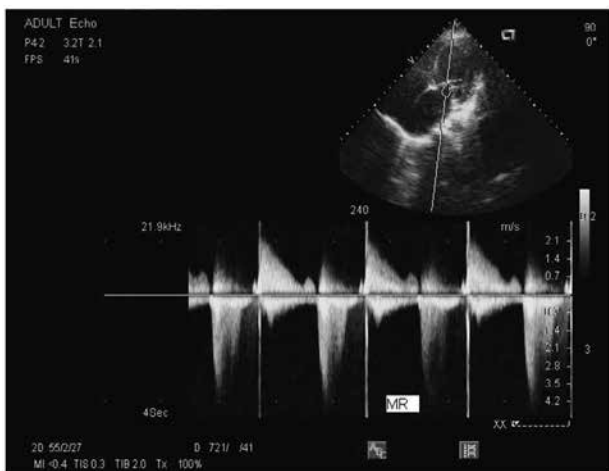


Figure 1: A preoperative transthoracic echocardiographic image, showing the thickened mitral valve with mitral regurgitation.

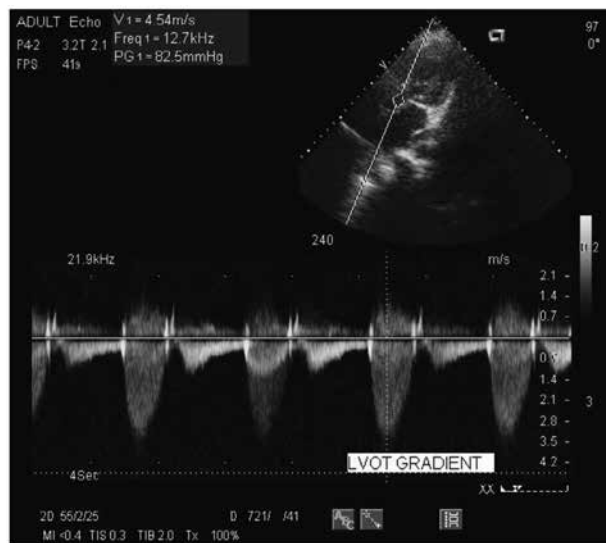


Figure 3: A preoperative transthoracic echocardiographic image, showing severe aortic stenosis with a peak gradient of 82.5 mmHg.

continuous monitoring of the ECG, non-invasive BP and oxygen saturation was started on the operating table. A baseline heart rate of 86 bpm and BP of 140/96 mmHg in the right upper limb were observed. An 18-G epidural catheter was inserted at the L2–L3 interspace in the sitting position, and a test dose of 3 ml lignocaine administered. Epinephrine was avoided as it can lead

to tachycardia. The patient was then made supine with left uterine displacement.



Figure 2: A preoperative transthoracic echocardiographic image, showing the subaortic membrane with bicuspid aortic valve.



Figure 4: A preoperative transthoracic echocardiographic image, showing coarctation of the aorta.

A right radial arterial line, a left femoral arterial line and a central venous cannula in the right internal jugular vein were inserted under local anaesthesia. Twelve millilitres of plain bupivacaine (0.5%) and 50 µg of fentanyl were injected through the epidural catheter in a graded manner until a level of T6 was achieved. Simultaneously, a phenylephrine infusion was started at a very low dose (40–60 µg/hour), titrated to avoid the fall in BP due to vasodilatation. A Caesarean section was performed and a female infant with an Apgar score of nine at one minute was delivered within five minutes of incision. An oxytocin infusion at 10 U/hour was started post delivery. Good uterine muscle tone was achieved. The phenylephrine infusion was stopped gradually. The procedure lasted 80 minutes, during which a total of 1 500 ml of fluid was infused, including 500 ml of colloid and 1 000 ml of crystalloid. Urine output of 300 ml was obtained. The patient was shifted to the post-anaesthesia care unit (PACU), where 0.0625% bupivacaine infusion was continued through an epidural catheter for 24 hours for the purpose of providing postoperative pain relief. The patient was shifted from the PACU on the fourth day, and discharged in a satisfactory condition on the tenth day.

Discussion

Shone's syndrome is a rare entity of multiple obstructive defects affecting the left side of the heart. It can occur in its complete form with the entire four lesions, viz. the supra-ventricular mitral ring, parachute deformity of the mitral valve, subaortic stenosis and coarctation of the aorta. The incomplete variety is observed more frequently when only two or three of the lesions are present.¹ A few cases of this rare anomaly have been diagnosed in adulthood. Cases of diagnosed prenatally have also been reported.^{2,5–7} Our patient was known to have Shone's syndrome. The associated mitral regurgitation was attributed to rheumatic heart disease. After corrective surgery was performed for her childhood episode of cardiac decompensation, she was free of overt symptoms until she fell pregnant. The physiological haemodynamic changes associated with pregnancy, such as the increase in plasma volume, heart rate, stroke volume and cardiac output, and the decrease in systemic vascular resistance, are known to have profound implications for patients with heart lesions, especially stenotic ones.⁸

Although the patient had tolerated her pregnancy fairly well, the chances of cardiac decompensation occurring during the peripartum period were considered. The fear of foetal distress, suggested by the cardiocography finding, prompted the decision to proceed with a Caesarean section.

Our patient experienced obstruction at three levels, i.e. mitral stenosis, aortic stenosis and coarctation of the aorta. The optimal choice of anaesthetic technique for delivery by Caesarean section in patients with stenotic valvular lesions is controversial.⁹ Neuraxial anaesthesia and analgesia may lead to afterload reduction, hypotension and tachycardia; unacceptable in a case of fixed cardiac output.¹⁰ General anaesthesia is associated with sympathetic stimulation in response to endotracheal intubation, which can be detrimental to the patients with existing cardiac disease, in addition to the unwanted impact of a general anaesthetic and narcotics on the infant.¹¹

We determined that foetal decompensation can be avoided and a better foetal outcome could be achieved with regional anaesthesia, providing an acute decrease in afterload, tachycardia and volume overload could be avoided, and maintaining the BP within the preoperative limits. Arterial lines were established prior to the epidural anaesthesia with these goals in mind. Anaesthetic drugs

were given in incremental doses until the desired level of T6 was achieved. Phenylephrine infusion was started simultaneously and was titrated to maintain the BP strictly within $\pm 10\%$ of the preoperative value. Fluid administration was guided by central venous pressure, which was kept between 10 cmH₂O and 12 cmH₂O. Strict control of the preload was vital as a decrease therein would lead to compensatory tachycardia, which could prove dangerous with such a tight aortic stenosis. On the other hand, an increase in the preload could lead to pulmonary oedema.

There was a difference in BP between the right upper limb and the other three limbs. Vasodilatation due to regional anaesthesia was anticipated in the lower part of the body causing an exaggerated fall in BP beyond the coarctation of the aorta; and, thereby, compromising the uteroplacental blood supply. Thus, arterial lines were established in the right radial and left femoral artery. However, such a fall did not occur because the titrated infusion of phenylephrine was strictly controlled.

In general, cardiac output increases during pregnancy, leading to an increase in left ventricular work. This demands an augmentation of coronary blood flow, which, in turn, is associated by the diastolic BP. Regional anaesthesia has been associated with ischaemic ECG changes in healthy parturients.¹² Our patient did not experience ischaemia; neither symptomatically nor as ECG changes. Compared to mephentermine and ephedrine (both α and β agonists) bolus doses, a phenylephrine (pure α agonist) infusion maintained the BP to the desired level without inducing tachycardia, thus maintaining adequate coronary blood flow in our patient.¹³

Koelble et al. reported on a case involving a parturient with Shone's syndrome associated with an aortic aneurysm. The patient delivered her infant successfully by Caesarean section performed under general anaesthesia.¹⁴ The only other report in which an epidural anaesthesia for Caesarean section was performed in a parturient with Shone's syndrome is that by Sachse and Hannallah.¹⁵ However, the case was complicated by maternal hypotension and foetal bradycardia. Emergent conversion to general anaesthesia was also required.¹⁵ Our patient was managed successfully by the establishment of arterial pressure monitoring prior to the incremental dosing of the epidural anaesthesia and by maintaining the BP meticulously with a phenylephrine infusion.

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