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Cesarean Delivery in a Patient With Hypertrophic Obstructive Cardiomyopathy

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Stem Case and Key Questions Content

A 35 year-old G1P0 with a twin pregnancy presents at 37 weeks to the Labor and Delivery suite for management of oligohydramnios. Her past medical history is significant for hypertrophic obstructive cardiomyopathy (HOCM) diagnosed as an infant, atrial and ventricular tachyarrhythmias, and gastroesophageal reflux disease (GERD). Twelve years prior to presentation, the patient had an automatic internal cardiac defibrillator (AICD) implanted for symptomatic, non-sustained ventricular tachycardia (NSVT) and primary prevention. Nine years prior to presentation, she experienced ventricular fibrillation with syncope, which caused her AICD to discharge appropriately. The patient also has occasional chest pain and shortness of breath due to paroxysmal atrial fibrillation with a rapid ventricular response. She tells you that the pregnancy has been complicated by one episode of near syncope around 28 weeks.

1. What else do you want to know about her cardiac condition?
2. Is there any other information you would need before making an anesthetic plan?

You find that the patient's AICD was interrogated at the time of her near syncopal episode and shows 13 episodes of NSVT at 170 - 230 bpm and 1 episode of ventricular tachycardia. She had an echocardiogram at 34 weeks, which showed a left ventricular ejection fraction (LVEF) of 65 - 70%, asymmetric septal hypertrophy, a peak left ventricular outflow tract (LVOT) gradient of 34 mmHg, normal right ventricular size and function, and no valvular abnormalities. The patient denies any other medical history. Her medications are metoprolol 150 mg daily and propranolol 10 mg as needed for palpitations. She has no known allergies. She has a normal body habitus, and her airway is a Mallampati class 2. She is anxious about having anesthesia and wants to know what your anesthetic plan is going to be.

1. What do you tell her?
2. Is her cardiac disease a contraindication for a neuraxial technique?
3. What are your concerns? What is she at risk for?
4. Do you need to talk to anyone or do any other preoperative testing?

You discuss this patient with a multidisciplinary team, including the patient's obstetrician, cardiologist, and electrophysiologist. Because of the patient's cardiac disease and her history of poorly tolerated arrhythmias, the decision is made for cesarean delivery.

1. Do you agree with the team's plan for cesarean delivery? Why do you think it was chosen?
2. Do you think there are advantages to vaginal delivery in this patient?
3. What type of anesthesia do you think is best for this patient?

You review the patient's laboratory studies and find that her hemoglobin is 12.6, her hematocrit is 35.4, and her platelet count is 104,000. She has normal prothrombin (PT) and partial

thromboplastin (PTT) times, as well as normal serum chemistries. You call electrophysiology to interrogate the patient's AICD and deactivate tachytherapy prior to going to the operating room (OR). You place defibrillating electrodes on her chest and back then bring her to the OR.
1. What is your anesthetic plan?
2. What monitors are you going to use?
3. What medications or special equipment are you going to have available for her?
4. What are you going to use for gastric acid prophylaxis?
5. What about deep venous thrombosis prophylaxis?

In the OR, you place an arterial line and two 18 gauge peripheral IVs. With the patient in sitting position, you place an epidural at the L3-L4 interspace. Prior to dosing the epidural, you initiate both phenylephrine and esmolol infusions. You load the epidural with a total of 20 cc of 2% lidocaine in slow, divided doses. After twenty minutes, she only has a T10 level bilaterally.
1. How are you going to improve the anesthetic level?
2. What medications and dosages are you going to use for the anesthetic?
3. How long should you wait for your initial dose to take effect?

You remove the epidural catheter, but you decide to place another one, this time at the L2-3 interspace. You use a 27G Whitacre needle to confirm correct placement of the Tuohy needle, but do not give a spinal dose. You give a total of 10 cc of 2% lidocaine in divided doses. After waiting an additional twenty minutes, the level of anesthesia is only T6, and the patient has pain with application of an Allis clamp above her umbilicus.
1. How would you position the patient for the second epidural?
2. Why do you think two epidurals failed to achieve adequate surgical anesthesia?
3. What are you options for anesthesia now?

After consulting with the patient, you decide to convert to general anesthesia. You continue the phenylephrine and esmolol infusions, pre-oxygenate the patient, and perform a rapid sequence induction with etomidate 10 mg and succinylcholine 100 mg. The airway is secured on the first attempt, and the neonate is delivered 3 minutes after skin incision. You start the oxytocin infusion immediately, and within 5 minutes, you need significantly more phenylephrine to maintain her blood pressure. You alert the obstetricians, and they tell you the uterus is not contracting well and she is bleeding more than usual.
1. What are you going to do to maintain hemodynamic stability?
2. How are you going to treat her uterine atony?

Despite your interventions, the bleeding continues. The obstetricians decide that the only way to control the bleeding is to perform a hysterectomy. The entire operation lasts 4 hours. You are able to extubate the patient at the end of the case. In the OR, she complains of pain. Her visual analog score for pain is 8/10.
1. What are you going to offer the patient for postoperative pain control?
2. Where should this patient be transferred post-operatively?
3. Do you think the general anesthetic contributed to her uterine atony?
4. Would you attempt epidural anesthesia on this patient in the future if she needed a different type of surgery?

Model Discussion Content
Hypertrophic obstructive cardiomyopathy (HOCM) is an autosomal dominant disorder with a 0.1-0.2% incidence in pregnant women (1). It is characterized by the presence of a gradient across the left ventricular outflow tract (LVOT), which subjects the patient to cardiovascular compromise any time there is a reduction in ventricular size (2). For this reason, anesthetic management of parturients with HOCM is complex. Presentations and symptoms vary widely and patients may be at risk for sudden cardiac death, pre-syncope, syncope, or arrhythmias (3). When providing anesthesia care, it is important to avoid worsening obstruction of the left ventricular outflow tract; goals should include maintaining sinus rhythm, and avoiding tachycardia, reduced preload, and reduced afterload (4).

Despite significant demands on the cardiovascular system, pregnancy is generally well-tolerated by patients with HOCM (5). According to the limited data that exists for this subset of patients, pre-pregnancy clinical status is predictive of the risk during pregnancy (6). For some patients, however, the physiologic stress of delivery can lead to decompensation, even if the pregnancy has been otherwise uneventful. This may be especially true for those patients who undergo cesarean delivery. During operative delivery there tends to be more blood loss and fluid shifts, both of which may be less well-tolerated by patients with HOCM.

The severity of the patient's cardiovascular disease will ultimately determine the need for invasive monitoring at cesarean delivery. Since most of the women with HOCM who carry a pregnancy to term, or close to it, do well, it is difficult to argue in favor of routinely obtaining central access. An arterial line, on the other hand, is a relatively low-risk procedure that provides valuable information about changes in hemodynamic status. Certainly parturients with an automatic internal cardiac defibrillator (AICD) in place should have an arterial line.

The presence of an AICD poses an additional challenge to the care of parturients with HOCM. Ideally, these patients should be monitored closely in the antepartum period, and as part of this management, AICD interrogation should take place (preferably well before delivery) in order to avoid intraoperative complications. It is important for AICD interrogation to occur because even if the plan is for vaginal delivery, cesarean delivery may be necessary. Application of a magnet to the device is not always sufficient to avoid the unwanted effects of electromagnetic interference. Most devices have a pacemaker function, but not all pacemakers have a magnet response, and some AICDs have no magnet response, while others can be permanently disabled (7). Whenever the antitachyarrhythmia function is disabled, a defibrillator must be immediately available.

Neuraxial anesthesia is preferred to general anesthesia in all parturients for cesarean delivery for the usual reasons, unless otherwise contraindicated. For healthy patients, without cardiovascular disease, spinal anesthesia is usually the technique of choice for several reasons, including reliability and rapid onset of dense sensorimotor blockade. It is technically simpler than epidural anesthesia and requires a lower dose of drug. For patients with HOCM, however, the rapid onset of sympathetic blockade that occurs with spinal anesthesia is especially undesirable. The decrease in systemic vascular resistance (SVR) that occurs with sympathectomy can have negative consequences for both the mother and fetus. A significant decrease in SVR will worsen left ventricular outflow obstruction, which in the most severe cases, can lead to cardiac arrest. A significant decrease in SVR can also cause inadequate perfusion

of the utero-placental circulation and poor oxygen delivery to the fetus. Although by no means as catastrophic, but still important, significant hypotension (as a result of low SVR) often results in nausea and vomiting, which is unpleasant for the mother and less than ideal for the obstetrician.

For all of these reasons, epidural anesthesia is probably the better choice for patients with HOCM (or other cardiac disease) who require cesarean delivery. It allows for titration of the degree of sympathetic blockade, while providing adequate surgical anesthesia. Still epidural anesthesia is not without its own complications; it is less reliable than spinal anesthesia, and it takes longer to establish (5 minutes vs. 15 - 20 minutes). There is evidence to suggest, however, that dural puncture with a 25-G immediately prior to epidural initiation in labor improves sacral spread, onset and analgesia (8). It seems reasonable then to consider this technique for cesarean delivery in order to improve the likelihood for satisfactory surgical anesthesia. It is also important to remember that the epidural space is heterogeneous; anatomical variation can prevent the spread of local anesthetic. Lastly, even when given in slow, divided doses, epidural anesthesia can produce sympathectomy and hypotension. An infusion of phenylephrine, the vasopressor of choice in both pregnant patients and patients with HOCM, is usually sufficient to counteract these changes.

In summary, HOCM is characterized by left ventricular hypertrophy (in the absence of other cardiac or systemic causes) as well as LVOT obstruction and presents with a variable clinical course (9, 10). The major implications of HOCM in pregnancy are related to the potential for clinically significant decompensation, which may manifest as arrhythmia, heart failure, or even sudden cardiac death. The increase in intravascular volume that occurs during pregnancy likely reduces obstruction of the LVOT and may partially explain why most women with HOCM do not deteriorate during pregnancy (11). However, the stress of delivery, vaginal or cesarean, can result in deterioration despite an uncomplicated intrapartum course. Anesthetic management of parturients with HOCM, therefore, requires attention to heart rate, rhythm, preload, and afterload. Both epidural analgesia for labor and epidural anesthesia for cesarean delivery are safe and effective when slowly titrated to effect (12).

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