

# Neuraxial Anesthesia for Delivery in a Parturient with Fontan Physiology

## OBJECTIVES

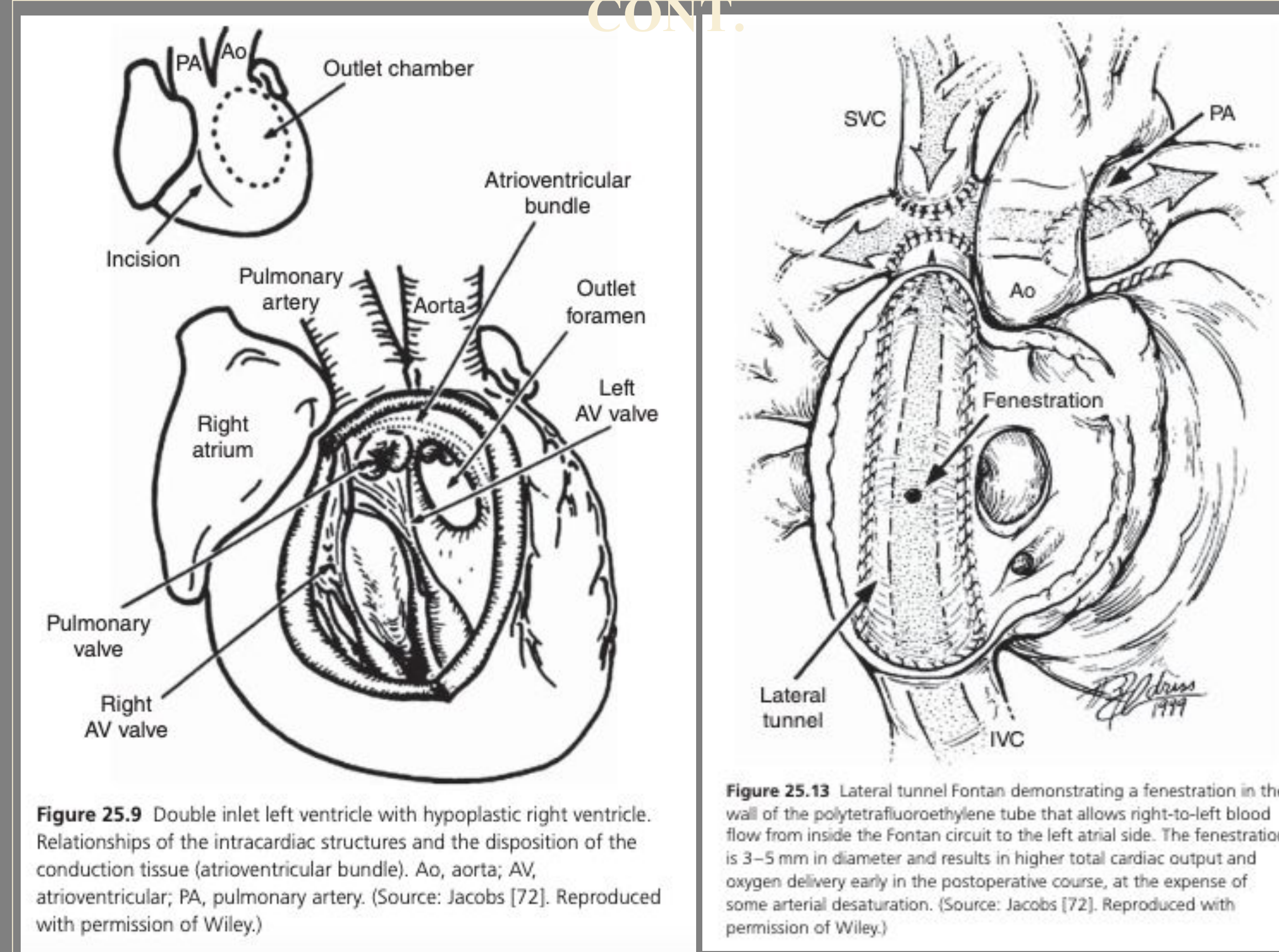
- Review characteristics of Fontan physiology
- Discuss neuraxial anesthetic considerations
- Optimize an anesthetic plan for a high-risk patient undergoing a concerning procedure

## CASE PRESENTATION

A 27-year-old parturient (G3P1011; ASA Class IV) was transported to the Riley Hospital OB floor after presentation with full-term, 5 days premature rupture of membranes and labor contractions at an outside hospital. She was 2 centimeters dilated and requesting an epidural upon arrival. All fetal monitoring results were within the normal range. Her past medical history included congenital heart disease, double inlet left ventricle, complete transposition of great vessels, pulmonary atresia, and questionable history of pulmonary emboli. She is currently taking ASA 81, pantoprazole, and is s/p lateral caval Fontan procedure with fenestration in the first year of life. Vitals all WNL. Lab results notable for platelet count of 93 k/cumm. An ECHO from two months prior revealed normal LV systolic function and a patent connection between the SVC and right pulmonary artery. A note from cardiology indicated a vaginal birth without pushing should be attempted but did not include an anesthesia consult. The anesthesia team admitted the patient to the OB ICU and prepared a plan for management.

## FONTAN PHYSIOLOGY

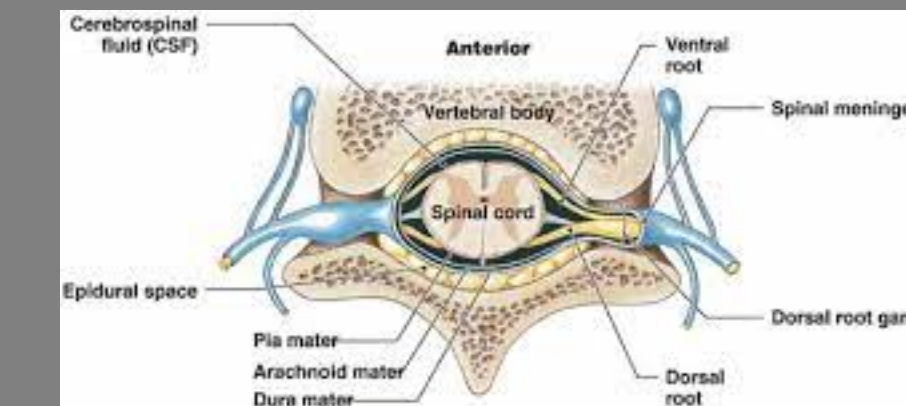
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## BASIC NEURAXIAL

CONSIDERATIONS

As stated by the ASA, "In the absence of a medical contraindication, maternal request is a sufficient medical indication for pain relief during labor." Neuraxial anesthesia, namely epidurals, are the most common and often safest anesthetic technique available for parturients. Due to concerns about anesthetic agents crossing the blood-placental barrier, avoidance of intravenous administration of medications as such as possible is ideal. Placement of a catheter within the epidural space allows for continuous or intermittent bolus injection of analgesics directly to the location of spinal nerve roots and allows for spread of the medication both above and below the puncture site. General disadvantages associated with epidural use mainly focus on the high incidence of sympathectomy, especially in hypovolemic or hypertensive patient populations. Signs of sympathectomy include profound hypotension, reflex tachycardia, hypothermia, and decreased response to stress. There is some debate as to what contraindications there are for neuraxial anesthesia; therefore, the only widely accepted contraindication is patient refusal.



## CASE CONTINUED

Prior to the initiation of neuraxial anesthesia, an awake arterial line was placed in the right radial artery in order to monitor blood pressure more closely. An appropriate waveform was noted and her preprocedural blood pressure was recorded to be 131/76 with a heart rate of 96 beats per minute. Placement of the epidural catheter went smoothly and was secured at the skin 5 cm past where loss of resistance was felt. A test dose of 3 mL 1.5% lidocaine with 1:200,000 epinephrine was given, and no response was observed. Common practice is to then give a loading dose through the epidural; however, due to this patient's high-risk cardiac history, no boluses were given throughout her labor, and she was not given the option for patient-controlled epidural analgesia (PCEA). Instead, the epidural infusion of Fentanyl-Bupivacaine (2 mcg/mL – 0.1% premix) began relatively low at 5.3 mL per hour and was gradually increased in 2 mL per hour increments until adequate analgesia was achieved. Over the 6-hour course of her labor, a total of 43.33 mL of the anesthetic was given. After dosing the epidural, her blood pressure was recorded to be 133/76, with a heart rate of 86 beats per minute. Due to the possible implications associated with this patient "bearing down", a vacuum-assisted delivery was performed by the OB team. Any potential elevations in intrathoracic pressure were avoided, and mom and baby made it through delivery without any serious complications. Mom was subsequently monitored in the ICU with the arterial line remaining in place for three days and was instructed to visit cardiology within two weeks. No issues with recovery were recorded.

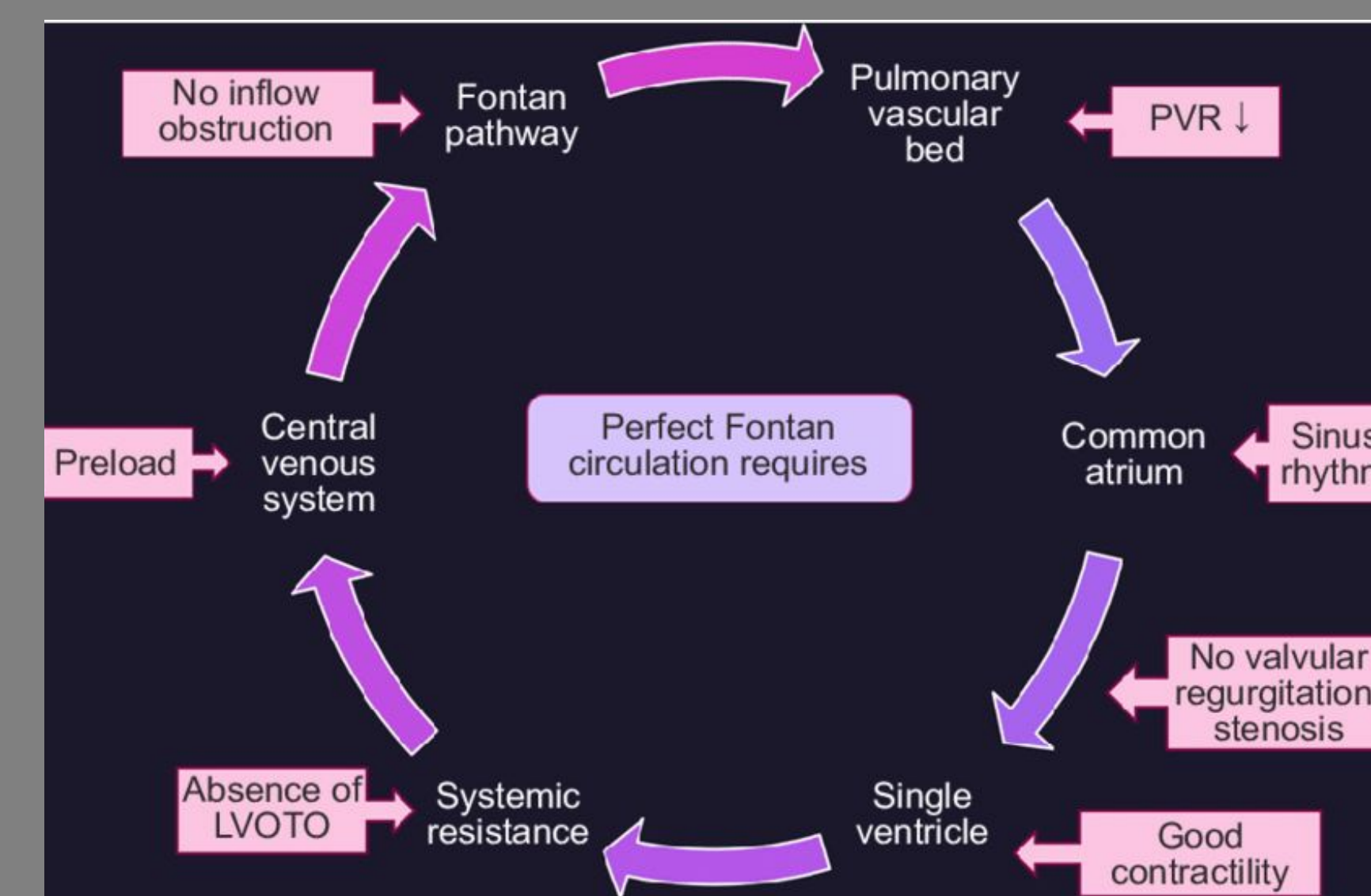
## FONTAN PHYSIOLOGY

Many forms of congenital heart disease exist, and all necessitate special attention from the anesthesia care team, even when surgically managed decades prior. As shown in Figure 25.9, a double inlet left ventricle forms when both atria are connected to the left ventricle, with a hypoplastic right ventricle. In addition, complete transposition of the great vessels indicates that the anatomic locations of the pulmonary artery and aorta are swapped. Both types of lesions ultimately disrupt systemic and pulmonary circulation by causing blood to flow through the system in parallel, rather than in series. Patients with serial circulation present with arterial desaturation and cyanosis due to deoxygenated blood bypassing the pulmonary system and mixing with oxygenated blood. Prior to surgical intervention, the balance between pulmonary and systemic vascular resistances must be optimized to prevent preferential blood flow throughout only one system. Factors that can increase pulmonary vascular resistance and should be avoided include hypercapnia, acidemia, hypothermia, and increased concentrations of circulating catecholamines. One surgical technique performed to manage these issues is the Fontan procedure, usually completed in sequential steps throughout the first year of life. In the final procedure, the patient undergoes cardiopulmonary bypass as the right atrium is opened, and a lateral tunnel is fed through to connect the IVC and SVC to the pulmonary artery. Sometimes, a hole, or fenestration, is made in the wall of the tunnel present within the atrium to act as a pop-off valve for the system in the instance of pressure overload. After the procedure, the single ventricle pumps blood only into the systemic vasculature, with

## FONTAN PHYSIOLOGY

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pulmonary blood flow becoming completely passive and dependent upon venous return from the periphery. In these patients, it is important to maintain their baseline blood pressure to prevent hypoxemia. Additionally, avoidance of positive pressure ventilation (PPV) and high positive end-expiratory pressure (PEEP) is beneficial to prevent elevated intrathoracic pressures and subsequent decline in venous return. The same can be said of patient positioning in steep Trendelenburg, or positions that directly decrease venous return, such as extreme reverse Trendelenburg and "beach-chair". Long-term issues that may arise after the Fontan procedure include atrial arrhythmias, reduced cardiac output, heart failure, and frequent air or thrombus embolization.



## FONTAN + PREGNANCY + NEURAXIAL

Cardiovascular disease has been named as the leading cause of death in the parturient, with pre-existing heart disease resulting in the highest risk. Pregnancy induces many cardiovascular changes that can endanger even the healthiest of patients, such as an increased blood volume and stroke volume, increased heart rate, and increased cardiac output demands. For the parturients at an elevated risk, the World Health Organization (WHO) established a classification of cardiovascular disease in pregnancy; those listed as Class III or higher must have their pregnancy closely monitored by a team including a cardiothoracic anesthesiologist. Prior to labor and delivery, this team creates a detailed birthing plan in which labor is induced in a controlled environment. Class III and higher cardiac lesions include mechanical valves, cyanotic heart disease, Fontan circulation, pulmonary hypertension, severe mitral stenosis, and a left ventricular ejection fraction less than 30%. Moms that fall in the classes above may even be advised to terminate their pregnancy due to the significant risk for maternal mortality and morbidity. WHO suggests that the ideal management for these parturients is vaginal delivery aided by early neuraxial anesthesia, with ICU monitoring for 24-to-72 hours post-partum. The hope for early use of an epidural is to minimize cardiac swings in response to pain and contractions, decreasing risk for arrhythmia and thromboembolic events.

Despite the suggestions made by WHO, our patient presented without a detailed birthing plan or consultation by an anesthesiologist. Additionally, premature presentation to a pediatric institution meant that there was no access to cardiology or ECMO should the need arise.

## CONCLUSIONS

Although rare congenital defects and their perioperative management may seem most relevant to the pediatric anesthesia care team, as surgical interventions improve and patients are able to live longer, healthier lives, these patients will continue to present to our operating rooms and still deserve the meticulous care we give to their younger counterparts. Additionally, the parturient patient presents unique challenges for the anesthesia team, when what is thought to be best for the patient does not align with patient goals and expectations.

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