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A Parturient Presenting in Premature Labor With Single-Ventricle Physiology Complicated by Breech Presentation and Paroxysmal Supraventricular Tachycardia: What's Your Labor Management

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Stem Case and Key Questions Content

A 27-year-old G3P0, 163 cm, 70 kg female at 24.6 weeks gestation was admitted to the labor and delivery suite secondary to preterm premature rupture of membranes and worsening dyspnea on exertion. She has a history of single ventricle physiology and paroxysmal supraventricular tachycardia (PSVT). The fetus was noted on ultrasound to be in breech presentation.

Key Questions:
1. What further information is required at this time?
2. Is medical consultation warranted? From whom?
3. What is the significance of her worsening dyspnea on exertion?
4. How does a history of paroxysmal supraventricular tachycardia affect anesthetic management?

Upon review of the medical records, her congenital heart disease consisted of left atrial isomerism, a common single ventricle, a common atrio-ventricular valve, transposed great arteries, moderate mitral regurgitation and a hemiazygous continuation of the inferior vena cava. Her previous heart surgeries included a pulmonary artery banding at 3 weeks of age, a subsequent bilateral bidirectional Glenn procedure at 7 months of age, followed by a Fontan procedure at 3 years of age.

Key Questions:
5. Why is pulmonary artery banding performed in the neonate and how does this improve cardiac blood flow oxygenation?
6. How does a bilateral bidirectional Glenn procedure improve oxygenation in a patient with single ventricle physiology?
7. What is a Fontan procedure? How is this different from a Glenn procedure?
8. What are the hemodynamic goals for labor and delivery in this patient?

Two previous pregnancies resulted in spontaneous first trimester abortions. Her medications included digoxin, enalapril, and aspirin.

Key Questions:

9. What is the cause for her two previous spontaneous first trimester abortions?
10. Which medications would you continue? Which if any would you discontinue?

On physical exam, she was noted to be cyanotic appearing with significant clubbing. She complained of mild shortness of breath during ordinary physical activity. Cardiac exam revealed a grade 3/6 high pitched pansystolic murmur on her left sternal border and lung auscultation revealed mild bibasilar rales. Airway examination was Mallampati Class II with full cervical range of motion. Vitals signs were a heart rate of 92 bpm, a blood pressure of 116/70 mmHg, and an oxygen saturation of 80-88% on room air. An 18G peripheral intravenous catheter was in place.

Key Questions:
11. What is your assessment of her maximum oxygen saturation of 88% on room air?
12. What is the significance of her cyanosis and clubbing? How would this affect anesthetic management?
13. What New York Heart Association (NYHA) classification is she? Why is this classification important in clinical management?
14. What laboratory examinations would you order?

Electrocardiogram revealed an ectopic atrial pacemaker, bilateral atrial enlargement, right bundle branch block, and left ventricle hypertrophy. An echocardiogram showed unobstructed blood flow into both pulmonary arteries. Arterial blood gas revealed a pH = 7.47, CO2 = 31, HCO3 = 22 and O2 = 52 on room air. Hemoglobin and hematocrit levels were noted to be 18.1 and 53.4, respectively with an INR of 1.1, PT of 12 and PTT of 36.4. A thromboelastogram (TEG) was within normal limits.

Key Questions:
15. What does the arterial blood gas reveal?
16. Why are her hemoglobin and hematocrit levels elevated?
17. What is your assessment of her coagulation studies?

The patient was admitted for antenatal surveillance and betamethasone was administered for fetal lung maturity. A PICC (peripherally inserted central catheter) line was inserted secondary to poor intravenous access and long term antenatal surveillance. She was placed on low flow oxygen (3 liters-per-min via nasal cannula), and continued on her aspirin and digoxin medications.

Key Questions:
18. Why was she kept on low flow oxygen?
19. Why was digoxin continued?
20. Does aspirin therapy affect your decision for performing regional anesthesia?

At 30 weeks gestation, she experienced abrupt vaginal bleeding and went into preterm labor. Breech presentation was confirmed by ultrasound. The multidisciplinary team consisting of obstetric anesthesia, obstetrics, maternal fetal medicine, pediatric cardiology, critical care medicine, and pediatrics met and recommended an immediate cesarean section.

Key Questions:
21. Why is it important to have a multidisciplinary approach for this patient?

22. What additional monitors are required for this case? Does this patient need a central line? Does this patient need an arterial line?
23. What are the advantages and disadvantages of regional anesthesia for this patient?
24. What are the advantages and disadvantages of general anesthesia for this patient?
25. What anesthetic do you chose for her cesarean delivery? Why?
26. How would you induce and maintain anesthesia for her cesarean delivery?
27. If this was a STAT cesarean section, would this change your management? In what way?

The patient received a combined spinal epidural with 0.75% bupivacaine (5 mg) plus fentanyl (25 mcg) in dextrose 8.5% intrathecally. The patient was placed in the supine position with left uterine displacement and oxygen was administered via face mask at 4 liters per minute. The patient was then bolused 10mls of 2% lidocaine with epinephrine in 2ml increments to achieve a T4 bilateral sensory level.

Key Questions:
28. What are the goals in anesthetic management of this patient for cesarean delivery?
29. What is the advantage of giving a low dose spinal followed by incremental dosing via the epidural catheter?

Seven minutes after surgical incision, a male infant weighing 1213gm was delivered through a low transverse uterine incision with Apgar scores of 4 and 7 at 1 and 5 minutes. The procedure was completed uneventfully.

Key Questions:
30. Where should the patient recover? Why?
31. Would you obtain any additional laboratory examinations at this time?

Model Discussion Content
Medical advances in the treatment of congenital heart disease (CHD) have led to an increase in parturients with heart disease. It is important to have a fundamental knowledge of the cardiopulmonary physiology of each of these lesions and the impact that pregnancy and labor will have on this physiology.

Single Ventricle Physiology
Multiple congenital heart defects may lead to single ventricle physiology. To survive at birth the single ventricle must supply cardiac output to both the systemic and pulmonary circulations. In order to achieve this requirement there must be a communication at the atrial level to allow mixing of oxygenated and deoxygenated blood and a systemic to pulmonary artery connection, such as a patent ductus arteriosus (PDA), to allow flow to the pulmonary and systemic circulations.

The palliation of single ventricle defects is usually done in three stages. The first stage and the least likely to be seen in a parturient, is the patient with a single ventricle and a systemic to pulmonary artery shunt, such as a stented PDA, or a surgically created central shunt, such as the Blalock Taussig shunt (Figure 1). This is a palliative step usually performed in young infants when it is necessary to provide are liable blood supply to the pulmonary system during the period of higher pulmonary vascular resistance (PVR) seen in the neonatal stage. Blood is

driven into the pulmonary bed by the systolic blood pressure, resulting in a systemic arterial oxygen saturation that is dependent on systemic blood pressure. Therefore, the higher the systolic blood pressure, the greater the amount of pulmonary blood flow and the higher the arterial oxygen saturation.

The second stage is usually surgically created around 4 to 6 months of age when the PVR has dropped to normal levels, allowing blood to flow passively to the lungs. The central shunt is surgically removed and a systemic venous to pulmonary artery (PA) connection is created, which is called a Glenn procedure (Figure 2). The Bi-directional Glenn connection involves connecting the SVC (superior vena cava) to the side of the right PA, which maintains continuity of the left and right PA's facilitating the flow of blood to both lungs. An alternative to the Bidirectional Glenn is called the Hemi-Fontan, which involves connecting the junction of the right atrium and SVC directly to the right PA with patch augmentation of the central PA. One benefit of the Hemi-Fontan procedure is it is thought to prevent central PA hypoplasia. At this stage of surgical palliation, blood is now directed passively to the lungs via the SVC, and the mean pulmonary artery pressure is equal to the systemic venous pressure. This decreases the workload on the single ventricle by diverting the blood from the upper half of the body directly to the lungs. The IVC at this point remains connected to the right atrium, creating a right to left shunt and resulting in a systemic oxygen saturation of 75-80% (Figure 2). The timing of this second stage of surgery is important and should be undertaken prior to the potential development of pulmonary hypertension secondary to the high pressure, high volume state of the central shunt.

The final and third operation to separate the two circulations and stop the right to left shunting is completed around 18 months of age and is called the Fontan procedure (Figure 3). During this procedure IVC blood is diverted through a lateral tunnel conduit to the pulmonary artery. The pulmonary and systemic circulations are now ***in-series*** with little mixing of oxygenated and deoxygenated blood and the patient's systemic arterial oxygen saturation should be above 95% (Figure 3). In both the Bidirectional Glenn and Fontan circulations, the pulmonary circulation is now driven by the remaining kinetic energy from the single ventricle output, the transpulmonary gradient (the difference between the mean central venous pressure and the systemic ventricle end diastolic pressure), and the negative intra-thoracic pressure created by inspiration.[1].

Pregnancy in Single Ventricle Patients

Approximately 85% of children born with CHD are now surviving into adulthood due to advancements in surgical and medical management of such patients.[1,2] This increase in survivability translates into more parturients presenting with CHD, and therefore it is necessary that practitioners in the obstetrical setting have an in-depth understanding of the physiology of such diseases. Cardiac disease is a major non-obstetric cause of death during pregnancy, labor, and delivery, and an estimated 25% of maternal cardiac deaths now involve CHD.[3] Furthermore, the risk to the fetus is well correlated with the degree of hypoxia and cardiac disease.

Siu et al.[4] published a risk assessment analysis of parturients with congenital and acquired heart disease and concluded that there are four predictors for a primary cardiac event during pregnancy: 1) a prior cardiac event including arrhythmias, heart failure ,transient ischemic attacks or a stroke before pregnancy; 2) a baseline New York Heart Association (NYHA) class > II or cyanosis; 3) a diagnosed left heart obstruction defined by a mitral valve area < 2 cm^2, aortic

valve area < 1.5 cm^2, or a peak left ventricular outflow tract gradient > 30 mm Hg; 4) a reduced systemic ventricular systolic function with an ejection fraction < 40%. Using their index, the risk of having a cardiac event in pregnancy in a patient who has heart disease was 5% if none of the events as described above were present, 27% if one, and 75% if greater than one.[4] This index is applicable to women with single ventricle physiology given that they are at risk for arrhythmias, may have cyanosis depending on the stage of surgical palliation achieved, may have deterioration in NYHA class during pregnancy, and often have ventricular dysfunction due to persistent hypertrophy and ventricular dilation.[1,2,5,6] Despite the fact that many single ventricle patients present with good cardiac function, they remain at high risk for a significant cardiac event during pregnancy and delivery .Parturients with poor functional status, an ejection fraction less than 40%, pulmonary edema, and symptomatic arrhythmias are at risk for cardiac events.[4] Ideally, patients with a single ventricle should have preconception counseling. This is done to apprise the patient of the risks and possible complications of pregnancy, and for an evaluation that may reveal an anatomic or functional problem that should be assessed prior to pregnancy which may result in a safer and less complicated pregnancy. A multidisciplinary approach is optimal when discussing the risk of pregnancy and whether or not pregnancy is advisable.[1] Complications are common in pregnancies of women with single ventricle physiology. Most single ventricle patients today have been palliated with the Fontan operation and their systemic arterial oxygen saturation is usually over 95% and hemoglobin is normal. If a patient is cyanotic, she has a greater chance of complications. Drethen et al.[7] performed a literature review of 2,491 pregnancies in women with CHD. Cyanotic patients (the single ventricle with cyanosis was not a separate category) had a 40% spontaneous abortion rate, 3% developed arrhythmias,14% developed heart failure and 3% developed endocarditis. In contrast, Fontan patients had rates of spontaneous abortion and dysrhythmias equal to 40% and 16%, respectively, but they had no episodes of heart failure or endocarditis. Perinatal mortality was 3% in the cyanotic group and less than 1% in the Fontan group. Such findings demonstrate a benefit to being acyanotic.[7] Arrhythmias are common complications during pregnancy in women who have had a Fontan procedure.[1,3,5,6] Studies have shown up to 26% of these patients develop supraventricular arrhythmias at some point during their pregnancy. The mechanism behind this complication involves scar tissue formation in the right atrium, damage to the sinoatrial node during prior atriotomy, and exposure of right atrial tissue to elevated pressures. Arrhythmias tend to become more frequent as the pregnancy progresses, correlating with increased plasma volume and elevated levels of estrogen and progesterone causing augmentation of adrenergic receptor activity.[5,6]

Furthermore, these patients are at increased risk for having first trimester spontaneous abortions, intrauterine growth retardation, small-for-gestational age offspring and fetal demise.[1,2,5,7] This is due to both placental insufficiency and the increased likelihood of premature rupture of membranes and preterm labor in Fontan patients.[1,5] In cyanotic parturients, the risk to fetal well being is correlated with the degree of maternal hypoxia and the incidence of fetal demise is 1-4%.[8] Patients following Fontan surgery have been shown to be at an increased risk for intracardiac thrombus formation.[1,2,9] This risk is due to a combination of factors including the existence of prosthetic material, increased risk of atrial arrhythmias, and potential for diminished cardiac output resulting in a low flow state.[10] Additionally, there is evidence that single ventricle physiology is associated with a baseline deficiency in anticoagulant factors, such as antithrombin III and proteins C and S.[9] Overall, there remains controversy regarding the need for anticoagulation in patients following Fontan surgery. Some groups, such as patients with a history of thrombosis, low cardiac output, arrhythmias, and atrial

dilation, are more routinely anticoagulated.[6] Furthermore, pregnancy further increases the risk of thromboembolism in such patients. Despite full anticoagulation, thromboembolic events can still occur in these patients.[11] Aspirin is continued through the day of delivery and resumed as soon as there is no longer a significant risk for postpartum bleeding. In practice, patients who have had a significant thrombosis are placed on heparin or enoxaparin, given the teratogenic effects of coumadin. In such patients, prophylactic doses of enoxaparin should be stopped 12 hours prior to any regional technique and can be restarted 2 hours after a single shot procedure or after catheter removal in the case of a continuous catheter technique. According to the American Society of Regional Anesthesia and Pain Medicine Evidence-Based Guidelines, therapeutic heparin infusions should be stopped 4 hours prior to regional anesthesia and can be restarted 2 to 4 hours after discontinuation of the needle or catheter.[12]

Patients with single ventricle physiology are often on multiple cardiac drugs. Most cardiac medications should be continued throughout pregnancy, but the lowest possible effective dose should be used. Single ventricle patients are particularly at risk for arrhythmias and sinus rhythm should be maintained with medications, such as beta-blockers, calcium channel blockers, and digoxin, and with cardioversion, if necessary, in a hemodynamically unstable parturient.[3,5,9] Angiotensin converting enzyme (ACE) inhibitors should be stopped during pregnancy as they are linked to renal tubular dysgenesis and neonatal renal failure.[2] Importantly, dosing of medications will change during pregnancy given the increased volume of distribution, increased plasma volume, increased hepatic clearance, and altered protein binding of drugs. For example, a patient taking digoxin 0.25 mg per day may need to increase her dose to 0.5 to 0.75 mg per day by 30 weeks gestation. Medications should be continued throughout labor with doses returning to pre-pregnancy levels after delivery. The cardiovascular changes that occur in pregnancy can be divided into three phases, each with its own risks. The first phase, *gestation period,* is characterized by an increase in total blood volume (35%), plasma volume (45%) and red blood cell volume(20%). There is a decrease in systemic vascular resistance (SVR) and PVR and an increase in cardiac output, stroke volume and heart rate.[3,5] These normal cardiovascular changes of pregnancy can have substantial consequences in parturients with single ventricle physiology, and thus frequent cardiac assessment should occur throughout the pregnancy.[5] Plasma volume and cardiac output peak near the end of the second trimester or approximately 20 weeks gestation.[2]. At this time, a follow-up echocardiogram can be beneficial in assessing overall ventricular size and function and valve gradients, and in determining if any change has occurred since early pregnancy that may impact the delivery plan.[3]

Labor and delivery is the second phase. During labor the cardiovascular system is affected by pain, uterine contractions, medications, maternal position, and the type of delivery. Pain and anxiety should be controlled to minimize any further increases incardiac output. This is achieved using regional techniques, intravenous pain medications, and other non-pharmacological techniques. During a contraction the cardiac output increases another 30-40% as blood is displaced from the uterus to the venous system. Aortocaval compression is particularly clinically relevant in patients with passive blood flow to their lungs and should be avoided.[2] The immediate *post partum period* is the third phase and is characterized by another 30% increase in cardiac output. Blood volumes are variable at this point. At delivery an estimated 500-1000 milliliters (mL) of blood is lost; however, blood volume does expand after delivery due to the loss of the placental circulatory bed and the contracting uterus. Given this increase in cardiac output and expanded blood volume, patients with poor cardiac function or pulmonary hypertension may

be at risk for sudden death up to one week postpartum.[1,2] The use of oxytocin immediately postpartum may be associated with vasodilation and hypotension, which can result in devastating cardiovascular consequences in single ventricle patients. Oxytocin should be infused at the slowest effective dose.[2]

Anesthetic Management
It is fundamentally important to understand that Glenn and Fontan circulations are *volume responsive states* that require an adequate preload and normal to low afterload to maintain cardiac output. The anesthetic goals are to preserve cardiac systolic function by minimizing the cardiac depressant effects of any agents, keeping the PVR low and maintain sinus rhythm, a crucial aspect of maintaining good cardiac output. A normal to low PVR is beneficial in maintaining systemic arterial oxygen saturation by encouraging passive pulmonary blood flow into the lungs. A decrease in PVR is facilitated by providing supplemental oxygen, keeping the PCO2 normal to low normal, and preventing and treating respiratory acidosis. Spontaneous ventilation is beneficial in that the negative intra-thoracic pressure created during inspiration augments blood flow to the lungs and improves oxygenation.[1] Positive pressure ventilation may result in decreased preload and thus a decreased cardiac output.

An increase in SVR results in decreased oxygen delivery to the tissues and if hypotension is due to low systemic cardiac output raising the SVR may further decrease cardiac output.[6] Ephedrine and phenylephrine are the most commonly used vasopressors for obstetric patients. However, each has disadvantages for patients with single ventricle physiology. Ephedrine, secondary to its β-1 stimulation, may cause excessive tachycardia, which can cause decreased preload and cardiac output. Phenylephrine may cause increases in PVR and SVR, both of which may negatively affect cardiac output. Milrinone, an inotrope and vasodilator acting through increases in contractility and decreases in PVR, may be the drug of choice in patients with single ventricle physiology, significant baseline cyanosis, and a low cardiac out put state.[13]

The goals of any anesthetic should be to balance the pulmonary and systemic blood flow by preserving ventricular systolic function, minimizing the cardiac depressant effects of anesthetic agents, keeping the PVR low, and maintaining sinus rhythm, all of which are crucial in maintaining cardiac output. The second stage of delivery should be shortened if possible as Valsalva during this phase decreases venous return and increases afterload.[3] As mentioned previously, there is a fine balance in these patients and cardiac output can fall precipitously with decreases in preload.[1] Depending on the patient's ventricular systolic function and other comorbidities, small fluid boluses should be given as needed to maintain preload and cardiac output. With few exceptions, the method of delivery should be defined by obstetrical considerations. Vaginal delivery is generally preferable because it is associated with less blood loss, less risk of infection, and less thromboembolic events.[1,2] However, this should be undertaken at institutions with considerable experience in neuraxial blockade and forceps assisted vaginal deliveries. There is no consensus regarding absolute contraindications to vaginal delivery but cesarean delivery is reserved for obstetrical reasons and patients in severe heart failure.[2] Delivery by cesarean section provides the benefits of minimizing the deleterious cardiovascular effects caused by repeated Valsalva maneuvers and the increases in positive intrathoracic pressure required to deliver vaginally.[3] Delivery should be scheduled at a time when maximum clinical support is available.

Neuraxial anesthesia has a number of benefits in these patients. In addition to excellent analgesia, the lower body sympathectomy dilates the venous system and attenuates the large swings in blood volume seen at delivery.[3] This sympathectomy should be achieved gradually with an epidural or a combined spinal epidural anesthetic technique to avoid sudden changes in SVR and potential reversal of shunts leading to maternal hypoxemia.[1,6] Labor epidural analgesia may be induced via the epidural catheter with a dose of up to 10 mL of bupivacaine 0.0625% to 0.125% ± fentanyl (100μg) or an equivalent dose of ropivacaine given in small bolus dose increments. This should then be followed by a continuous infusion or patient controlled epidural infusion to provide adequate analgesia (T10-L1 dermatomes for the first stage of labor) while maintaining cardiovascular stability.[3] Given the more rapid changes in SVR seen with spinal anesthesia for labor analgesia, subarachnoid medications should be limited to opioids (fentanyl 20 μg) alone or opioids with a decreased dose of a local anesthetic (≤2.5 mg bupivacaine), as typical doses of subarachnoid local anesthetics may cause a significant sympathectomy.[3,14]

For cesarean delivery, an existing epidural can be carefully bolused in divided increments with 2% lidocaine (up to 20 mL) while maintaining cardiovascular stability or a combined spinal epidural (CSE) technique with hyperbaric intrathecal bupivacaine (5 to 7.5 mg), followed by bolus dosing the epidural catheter with 2% lidocaine titrated to a bilateral T4 dermatomal level before incision.[1, 14] In addition to the previously mentioned advantages of regional anesthesia, it provides the theoretical benefit of avoiding negative inotropic effects of most anesthetic agents. If general anesthesia is necessary, left uterine displacement is crucial in maintaining preload in patients with single ventricle physiology.[2] Hemodynamically stable induction agents, such as ketamine and etomidate, should be considered. Ventilator parameters should keep the peak airway pressures low to decrease PVR and maintain oxygenation. Additionally, the respiratory rate should be set to provide adequate minute ventilation with normal to low pCO_2 and the inspiratory time should be shortened to avoid decreases in preload. Spontaneous ventilation should be resumed as soon as possible to maximize cardiac flow. In patients with decreased cardiac function, large bore intravenous access, an arterial line for constant blood pressure evaluations, and reliable central line access for vasoactive medications infusion is advisable to optimize patient care. Pulmonary artery catheter monitoring is rarely indicated due to the risk of arrhythmias, thromboembolic complications, difficulty in interpretation, and distorted anatomy in these patients.[3] An echocardiogram assessing structure and ventricular function can help guide decisions to optimize the patient's cardiac function.[3,10] At the time of delivery, endocarditis prophylaxis is warranted due to their prior history of cardiac surgery.[1]

In conclusion, careful preparation and good multidisciplinary communication is key to a good outcome in patients with complex cyanotic heart physiology. Providers familiar with the treatment of adult congenital heart defects are necessary to guide treatment and any complications that may occur during the course of delivery.

Figure 1: Tricuspid atresia showing the hypoplastic right ventricle and the central shunt from the innominate artery to the right pulmonary artery. M = mixed oxygenated and deoxygenated blood, P = pink, oxygenated blood, B = blue, deoxygenated blood.

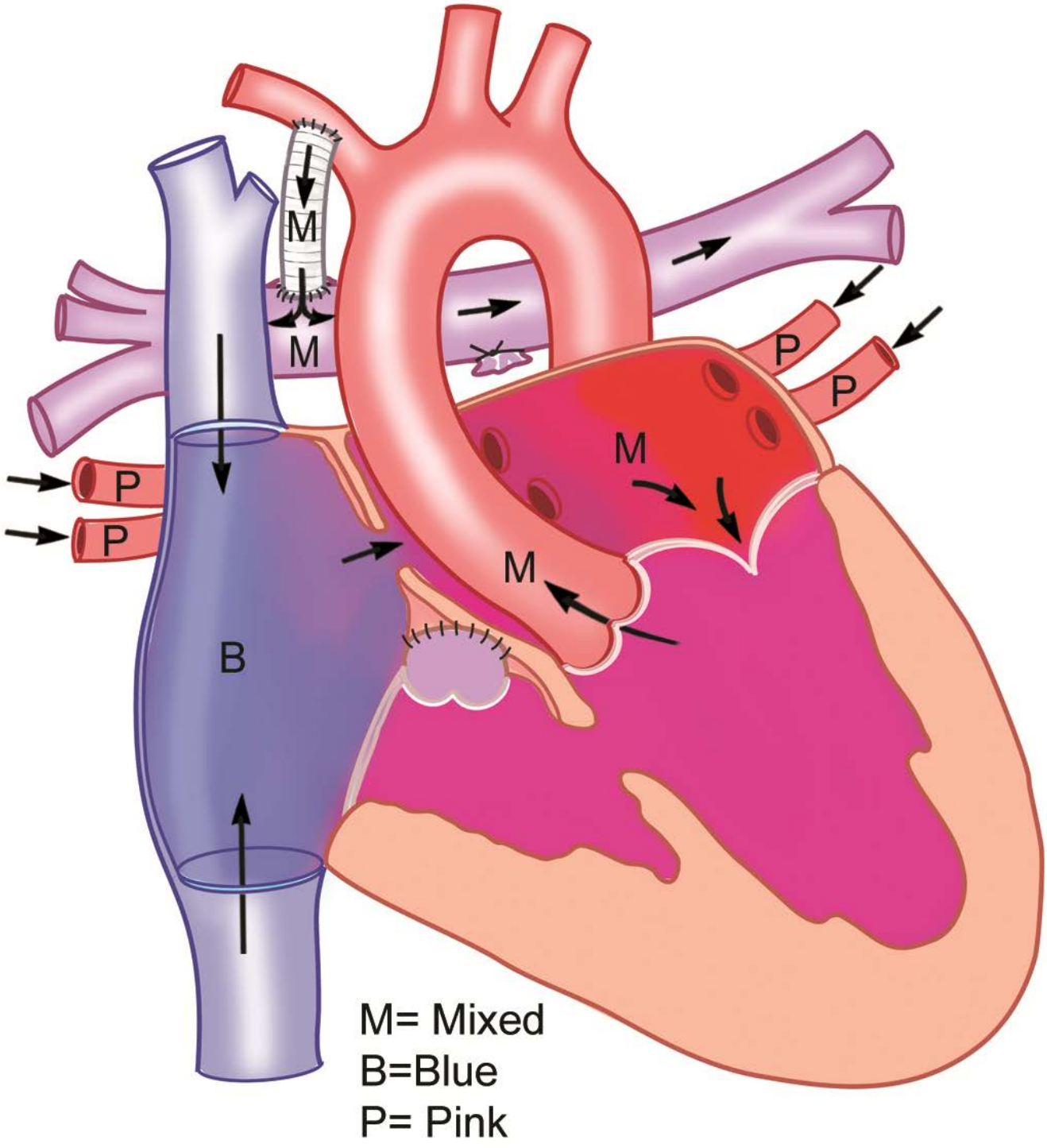


Figure 2: Bi-directional Glenn physiology. Tricuspid atresia showing the hypoplastic right ventricle, the central shunt has been removed and the superior vena cava connects to the right pulmonary artery. M = mixed oxygenated and deoxygenated blood, P = pink, oxygenated blood, B = blue, deoxygenated blood.

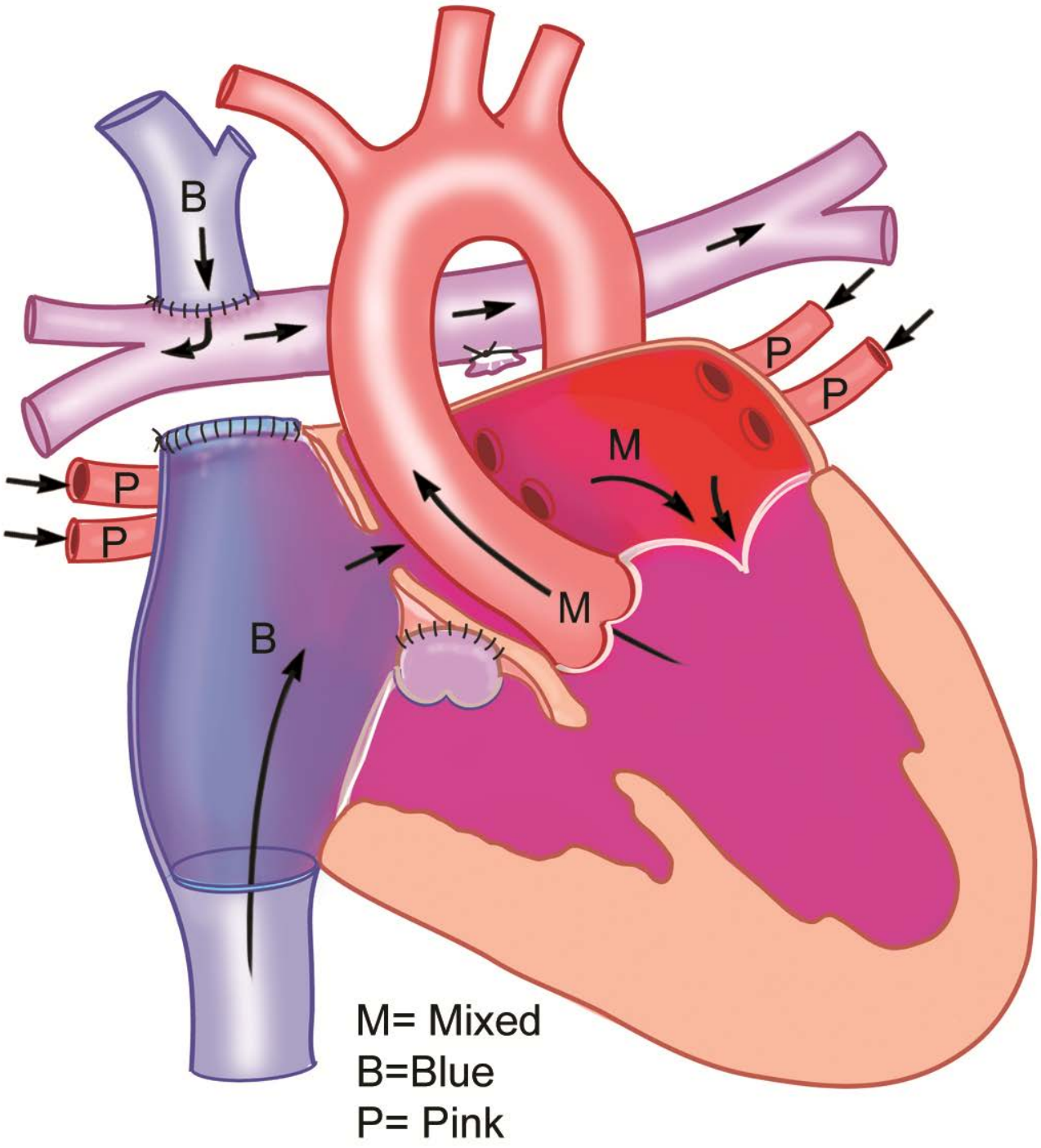
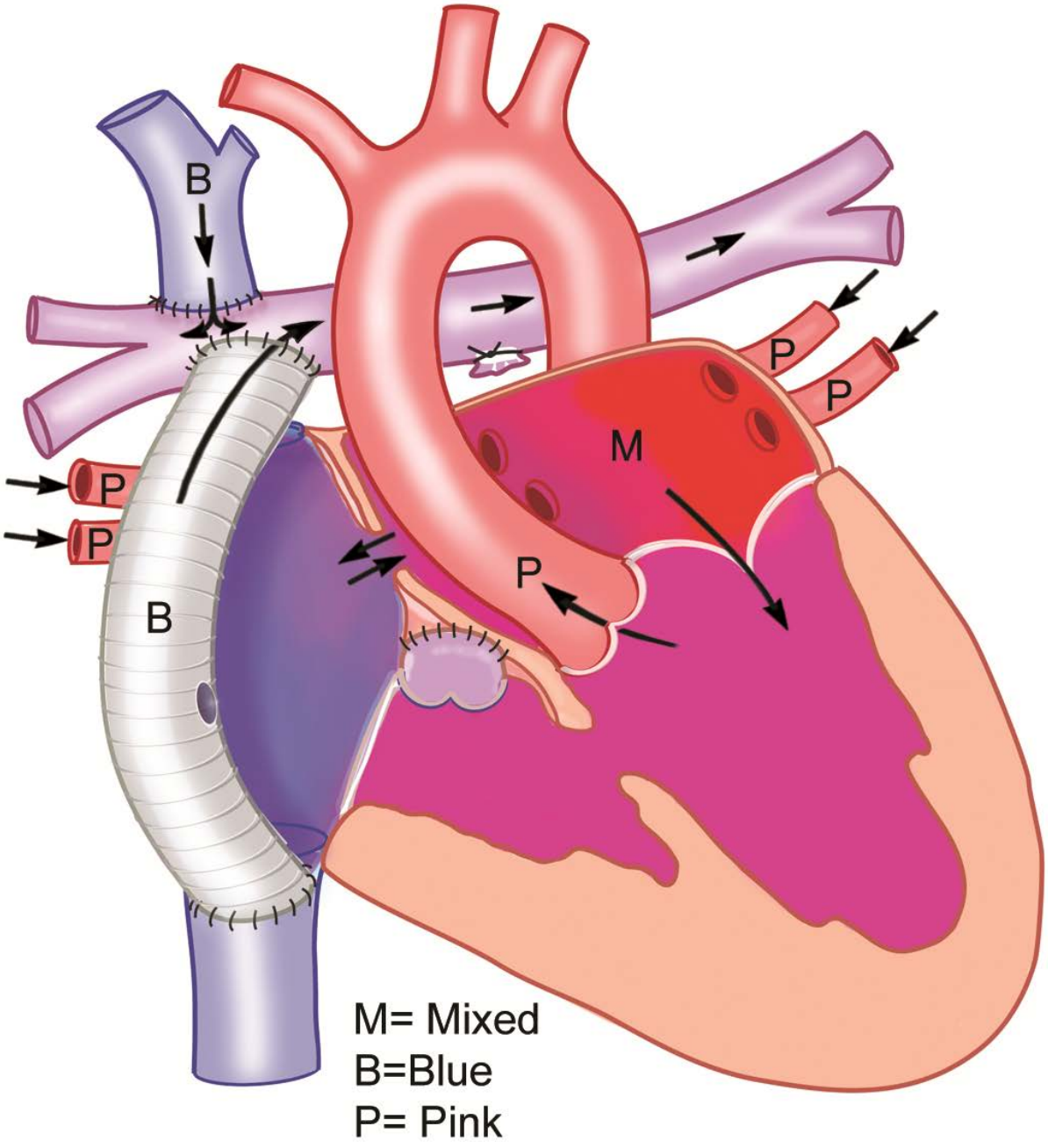


Figure 3: Fontan operation: Tricuspid atresia showing the hypoplastic right ventricle, the central shunt has been removed, the superior vena cava connects to the right pulmonary artery and now the inferior vena cava's blood is directed through an extra cardiac lateral tunnel to the right pulmonary artery. M = mixed oxygenated and deoxygenated blood, P = pink, oxygenated blood, B = blue, deoxygenated blood.



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