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Anesthesia for a Hypoplastic Left Heart Syndrome Parturient

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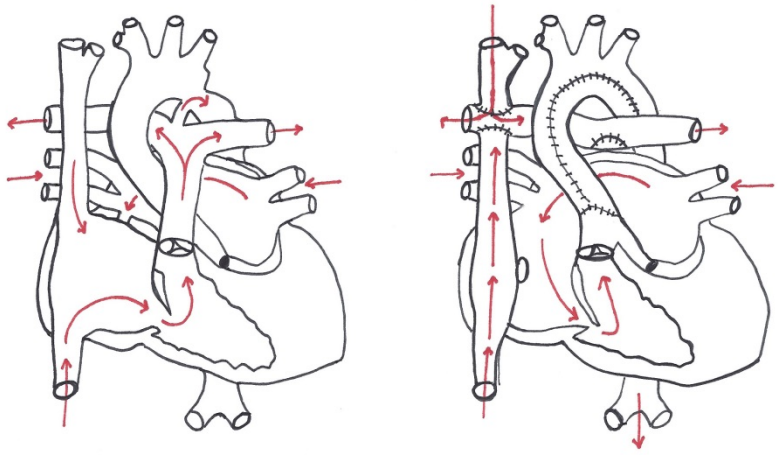
Stem Case and Key Questions Content

You are consulted on a 19 year old G_3P_{0020} singlet intra uterine pregnancy at $24^{1/7}$ weeks gestation. The patient is reportedly completely healthy with no medical issues except those related to the "complete correction" of her Hypoplastic Left Heart Syndrome (HLHS). A quick literature search finds case reports of successful pregnancies involving single ventricle physiology, but none specifically of an HLHS Fontan parturient. A mildly anxious parturient hands you an impressively large medical file. She tells you that her "complete correction" is an extra-cardiac Fontan.

Chart Review

This HLHS patient's ductus arteriosis was maintained using a PGE_2 infusion for the first 5 days of her life before undergoing a Norwood Stage 1 operation with a right modified Blalock-Tausig shunt (mBTS). Six months later, she was converted to a bi-directional Glenn (BDG) and the mBTS was removed. Sixteen months later, she underwent conversion to an extra-cardiac fenestrated Fontan. Recent echocardiogram reports indicate that the fenestration has spontaneously closed.

Trace the complete circulation of a single red blood cell (RBC) through the Fontan schematic. What pathway supplies pulmonary blood flow (PBF)? Given the complete passive pulmonary blood flow of a Fontan, what physiologic parameter(s) would you expect to influence the cardiac output?



Your literature search identified possible complications following Fontan operation; (1) low cardiac output syndrome, (2) hypoxemia, (3) complications associated with systemic venous hypertension, (4) dysrhythmias, and (5) thrombosis. With these in mind, consider the implications of different anesthetic techniques in a non-parturient.
Consider how these issues could evolve during pregnancy, labor and delivery, and post-partum given the cardiovascular physiologic changes accompanying pregnancy.

Phase	Estimated Blood Volume	Cardiac Output	Albumin (g%)	P_{onc} (mmHg)
/1\	↑ ~ 10%	↑ ~ 30%	3.9	25
/2\	↑ ~ 30%	↑ ~ 40%	3.6	23
/3\	↑ ~ 45%	↑ ~ 50%	3.3	22
Latent phase Ctx		↑ ~ 65%		
Active phase Ctx		↑ ~ 80%		
Expulsive phase		↑ ~ 95%		
Post-partum		↑ ~ 130%		

Developing an Anesthetic Plan in this Parturient
The obstetrician (OB) plans an elective delivery at 36 weeks gestation. Given a favorable cervix, labor would be induced, followed by an instrument-assisted delivery to prevent having the patient perform a valsalva maneuver; otherwise, an elective cesarean delivery would be performed. Until this time, she will be medically managed by both her OB and cardiologist.
She has a Mallampati Class II airway, a 5 cm oral opening with good dentition, a 6 cm thyroid-to-mandible distance, the ability to subluxate her mandible, and a full range of motion in her neck.

Pre-Anesthetic Workup
Would you want to examine her again before delivery day? When, or at what frequency? What exam changes would you expect?
Would you order chest X-rays and/or pulmonary function studies? When should they be performed? How would the results influence your anesthetic plan?
Would you order a cardiac catheterization? If so, why and when should it be done? How would the results influence your anesthetic plan?
CBC and electrolyte panel are normal for a 24 week gestation. Would you want other tests? If so, which one(s), why and when should they be done?

Anticipated Anesthetic Monitoring for the Delivery
In addition to standard ASA monitors, would you want invasive monitoring (A-line, CVP, PAC)? If so, which one(s) and why? Placed at what anatomic location?

Additional Preparations for the Delivery
Assuming no suspicion of placenta previa or accreta, what blood products would you want to have available? Are type-specific products adequate or is cross-matching required? Special concerns? What hemoglobin level would prompt you to transfuse red blood cells in this patient? She has never experienced cardiac arrhythmias or heart failure. Would you apply DC cardioversion pads for the delivery? What vasoactive drug(s) would you have available? How do they affect inotrope, pulmonary vascular resistance (PVR), and systemic vacular resistance (SVR)?

The Plan
Assuming the pregnancy progresses without incident, you plan a scheduled OR delivery, with left uterine displacement, a large-bore IV and left internal jugular CVP, an indwelling A-line, all fluids warmed, a 5 lead ECG, DC cardioversion pads applied, the vasoactive drugs on infusion pumps, and prophylactic antibiotics against bacterial endocarditis.
The plan is for a neuraxial anesthetic (NA) with an instrument-assisted vaginal delivery following spontaneous or induced labor. What are the anesthetic goals? How could these be achieved using a NA technique? Assume the plan is for a combined spinal-epidural (CSE) anesthetic infusing 0.2% ropivicaine with 2 µg/mL of fentanyl. What would you choose as the intrathecal drug(s)? Special concerns?

Would you alter this plan if she has been taking an aspirin daily, with no history of thrombus? What if she takes prophylactic or therapeutic doses of low molecular weight heparin?
The plan is for a general anesthetic if the OB wants to perform a “stat” cesarean section for obstetric indications without an established NA block or if the patient condition contra-indicates placing it.

On the Day of Delivery
BP is 115/75 sitting, HR 99, RR 24, T 36.5C, and the SpO_2 is 97% on room air. The physical exam is unchanged and she is visibly anxious. The OB believes the cervix is favorable for induction.

Premedication
Would you give her an anxiolytic? Special concerns?
Would you order gastroesophageal reflux disease (GERD) prophylaxis for a vaginal delivery? If so, which drug(s) and when would they be given? With no known drug allergies (NKDA), when would you administer antibiotics against bacterial endocarditis? With a penicillin allergy, is there a concern with vancomycin?

Induction of Anesthesia
Assume the pregnancy progressed without complications, the monitors are in place, the intravenous and intra-arterial access has been established, and prophylactic drugs have been given. The patient’s hemoglobin level is 9.5 g/dL. Would you give a fluid bolus prior to initiating the NA block? If yes, what fluid would you administer and why? Would you place the CSE with the patient in a sitting or lateral position? Would you identify the epidural space using loss of

resistance to air or saline? Would you give a “test dose”? If so, what drug(s) would you use? The OB induces labor using oxytocin. What effects would oxytocin have on the Fontan circulation?
Assume an uneventful labor, followed by a vacuum-assisted vaginal delivery of a baby boy; APGAR scores are 7 and 9.
Although oxytocin is infusing, the OB says the uterus is “boggy” and asks you to treat her. What would you give and how would you give it? How would it affect the Fontan circulation? Would you choose something else if the delivery were by cesarean section?

Post-Partum
Blood loss is estimated at 1000 mL and adequate hemostasis is achieved. Should the parturient be admitted to an ICU post-partum? What unusual complication(s) could occur in a post-partum Fontan patient?
Would you remove the epidural catheter prior to ICU transfer? How would you provide post-partum analgesia for the vaginal delivery? What about for a cesarean delivery? What if low molecular weight heparin is to be administered for prophylaxis against deep vein thrombosis? Would epidural morphine be appropriate in either of these settings? Would scheduled Ketorolac be a reasonable adjunct?

Model Discussion Content

The Fontan parturient is uncommon[1-3], but is expected to occur more frequently. Caring for these patients requires a review of the available literature[4] on Fontan parturients, analysis of the different patient management alternatives[5], and finally extrapolation of this information to the HLHS Fontan parturient.

Chart Review
We will base the discussion of physiological implications on this patient’s cardiovascular anatomy[6,7].

Developing an Anesthetic Plan
First, elucidate the goals of an anesthetic for the Fontan parturient. With these goals in mind, consider the rationale for/against performing extensive pre-delivery workup and testing. Emphasize those tests which influence the anesthetic management and peripartum course. Second, consider different types of hemodynamic monitoring with their indications and risks. And third, consider contingencies for addressing evolution of the peripartum course. A particular plan will be suggested, as well as some associated benefits and risks for evolution of the peripartum course.

Pre-Anesthetic Workup
A primary objective is to indentify the current cardiopulmonary anatomy and assess its functional status. Current anatomy should be delineated from the patient chart or determined using color Doppler trans-thoracic echocardiography (TTE) technology. A cardiac catheterization would be required to delineate specifics like coronary anatomy and to assess pulmonary pressures. Assessing the functional status would include a TTE interpreted by a

pediatric cardiologist combined with the patient's exercise tolerance. This may change with stresses placed on the cardiopulmonary system associated with pregnancy. Examining the patient on the day of delivery may be sufficient, provided that she has been closely followed by both the cardiologist and obstetrician, and that provision is made to re-examine her if a complication is identified.

The predictive value of a complication history related to her Fontan correction is unavailable; however, issues occurring and resolved in the early post-operative period including low cardiac output syndrome, hypoxemia, increased systemic venous pressures, dysrhythmias or thrombosis would be less concerning than if the same were to occur later. A recent history of supra-ventricular tachycardia (SVT) may indicate a pre-disposition to SVT in the parturient. The current ECG will identify a p-wave originating from the SA Node. Presence of a re-entrant pathway would prompt an in-depth discussion with the cardiologist regarding the best anti-arrhythmic drug and possible alternatives.

In the absence of co-morbid lung disease, pulmonary function testing is unlikely to provide additional information impacting the anesthetic plan. Serial chest X-rays would demonstrate pleural/pericardial effusions as well as interval changes. As the gravid uterus continues to elevate the diaphragm, the magnitude of cardiopulmonary compromise resulting from an effusion will increase, perhaps prompting an intervention.

Baseline laboratory tests are ordered to identify anemia, thrombocytopenia, hypoalbuminemia, coagulation dyscrasias, and electrolyte disorders. Collins et al[8] found a 29% prevalence of anemia overall in adults with single ventricle physiology. Determining the maternal blood type and screening for antibodies is prudent given the history of multiple exposures to blood products. Hypoalbuminemia and/or coagulation dyscrasias may indicate decreased hepatic synthetic function and initiate further investigation to differentiate between a low cardiac output syndrome and systemic venous hypertension as the cause. An elevated bicarbonate level may indicate respiratory compromise given the incompletely compensated respiratory alkalosis expected at term.

Anticipated Anesthetic Monitoring

An A-line for both vaginal and cesarean deliveries is planned to provide beat-to-beat hemodynamic monitoring and to facilitate blood sampling to guide therapy. Although 2 large bore IV's are adequate for volume resuscitation, the true indication for central venous access is the need to infuse vasoactive or inotropic drugs.

Monitoring CVP as a guide for fluid management is tempting; however, to date no data are available to suggest optimal values in the Fontan circulation. Internal jugular over subclavian vein access minimizes the risk of pneumothorax during placement. Femoral vein access may be compromised by the lithotomy position and CVP values would only reflect systemic venous pressure if the uterus does not impede IVC blood flow. A pulmonary artery catheter (PAC) would be technically challenging to place, as it would not follow the expected anatomical

pathway.

Additional Preparations
Administering fractionated blood products targets therapy without adding unnecessary volume. Cross-matched products may minimize the likelihood of transfusion reactions; even a transient increase in pulmonary pressures from a transfusion reaction could be devastating to the Fontan circulation. At a minimum, type-specific and cross-matched packed red blood cells should be available. No data are available to suggest optimal hemoglobin values in the Fontan patient; however, maximizing oxygen delivery and avoiding the increase in PVR that results from hypoxia is the goal.

Tachy-arrhythmias, particularly those where atrioventricular synchrony is lost, are poorly tolerated in Fontan patients. Some will have an automated implantable cardioverter-defibrillator (AICD) to maintain a sinus rhythm. Amiodarone is particularly effective at treating atrial flutter and junctional ectopic tachycardia in children, and presumably is effective in these patients; although it's generally avoided in pregnancy due to fetal thyroid toxicity. However, DC cardioversion to restore a sinus rhythm may be necessary, so placing external cardioversion pads seems prudent. Vasoactive drugs are chosen to support cardiac output and uterine blood flow.

Milrinone, a potent phosphodiesterase-3 inhibitor, is an effective inotrope-vasodilator which significantly reduces PVR and SVR and increases cardiac output primarily due to increasing stroke volume. The presumption is that uterine blood flow would be supported by the increased cardiac output, in spite of the decrease in SVR. Epinephrine would have a similar effect, although it may cause some uterine relaxation from β-adrenergic stimulation. Phenylephrine increases SVR to support maternal blood pressure and uterine blood flow, but also increases PVR, reducing preload; a lower preload combined with an increased afterload could decrease cardiac output. However, phenylephrine has been used successfully in a number of case reports.

The Plan
The anesthetic goal specific to a Fontan parturient is to maintain preload to the systemic pumping chamber which is derived from completely passive pulmonary blood flow in order to provide cardiac output adequate to support uterine blood flow. Planned interventions are directed at avoiding conditions which 1) decrease venous return to the heart (hypotension, hypovolemia, sympathectomy, vasodilation, positive pressure ventilation), 2) cause myocardial depression (cardiodepressant drugs, anesthetics), or 3) increase pulmonary vascular resistance (hypoxia, hypercarbia, acidosis) in order to avoid decreasing pulmonary blood flow, atrial filling pressures, and cardiac output.

A neuraxial (NA) anesthetic, with adequate intravascular volume, initiated gradually to minimize an acute sympathectomy precludes positive pressure ventilation and its possible deleterious impact on systemic venous return. A NA technique, using drugs with minimal myocardial

depressant effects, eliminates myocardial depression from volatile anesthetic agents. Dose dependent respiratory depression from systemic opioid use leading to hypoxia, hypercarbia, and acidosis could be mitigated by using neuraxial opioids. This respiratory depression is eliminated by using only local anesthetic (LA) drugs in the NA.

A combined spinal-epidural (CSE) using an intrathecal opiate offers some theoretical advantages during an induction of labor. Choosing low dose intrathecal fentanyl provides analgesia for the visceral pain component of latent phase labor with minimal risk of fetal bradycardia. It would not cause a sudden sympathectomy associated with intrathecal LA drugs. It permits interpretation of an epidural "test dose". It provides analgesia during infusion of the LA drug into the epidural space to gradually initiate a block for the somatic pain component of active phase labor. It permits demonstration of a bilateral LA block following its initiation. Finally, it avoids delayed respiratory depression experienced with the use of less lipophillic opiates.
In the absence of thrombocytopenia, neither prophylactic aspirin nor low molecular weight heparin (LMWH) changes the plan for a NA anesthetic technique, provided that prophylactic LMWH administration is discontinued 12 hours prior to placing a NA block[9].

On the Day of Delivery

Premedication

A short-acting benzodiazepine causes minimal respiratory depression and reduces anxiety-induced endogenous catecholamine release, thereby reducing PVR. Neonatal effects are easily managed. Aspiration would lead to increased PVR that could be devastating to the Fontan parturient; therefore, timely administration of metoclopramide and a long-lasting H_2-receptor inhibitor is beneficial for the duration of their action. Existing gastric pH is raised by administering a non-particulate antacid.

A conservative approach for prophylactic antibiotics against bacterial endocarditis is to administer them at the induction of labor and re-dose appropriately. A concern specific to vancomycin is the possible histamine release which increases PVR and decrease SVR, both of which are very undesirable in the Fontan parturient.

Induction of Anesthesia

Crystalloid fluid boluses given to single ventricle parturients prior to NA anesthesia have been reported; however, a colloid fluid bolus seem prudent given the Fontan parturient's sensitivity to fluid overload. The Fontan's propensity for developing effusions could be exacerbated during pregnancy through hemodilution and reduction in plasma oncotic pressures. Choosing a 5% albumin solution may counter this by contributing to an increased oncotic pressure. As this parturient is truly anemic, using PRBC's as the colloid solution would reduce maternal (and fetal) hypoxia and its resultant increase in PVR through increased oxygen-carrying capacity while increasing plasma oncotic pressure.

A CSE using intrathecal fentanyl only, then establishing a block by infusing the LA into the epidural space would not cause an acute sympathectomy. The sitting position facilitates access

to both the subarachnoid and epidural space. Identifying the epidural space using a saline loss-of-resistance technique is preferable to eliminate air introduction into epidural veins which are engorged with pregnancy and with the increased systemic venous pressures in a Fontan circulation.

A test dose using lidocaine without epinephrine administered through the epidural catheter would identify both intrathecal catheter placement by rapid block onset and intravascular catheter placement with sensorineural symptoms described by the parturient.
Oxytocin decreases both SVR and PVR. Both may be desirable unless the decrease in SVR causes hypotension, decreasing blood return to the pulmonary circuit, leading to decreased preload and cardiac output, further decreasing placental perfusion and leading to changes in fetal heart tones.

With a “boggy” uterus, infusing additional oxytocin would increase uterine tone and decrease PVR, promoting cardiac preload. Methylergonovine would undesirably increase PVR and should be avoided. As this patient is not an asthmatic, an intramuscular prostaglandin (15-methyl-$PGF_{2\alpha}$) injection would increase uterine tone with minimal risk of bronchospasm leading to an increase in PVR. During a cesarean delivery, intra-myometrial 15-methyl-$PGF_{2\alpha}$ injection may be the best alternative.

Post-Partum
The Fontan circulation continues to be stressed post-delivery by 1) increased blood volume and cardiac output from the autotransfusion associated with uterine contraction, and 2) peripheral fluid mobilization for many days post-partum, so ICU monitoring is required. The HLHS Fontan patient with the morphologic right ventricle acting as the systemic pumping chamber would be less adaptable to the acute volume increases than Fontan patients with a morphologic left systemic ventricle.

Requirements for post-partum analgesia differ by mode of delivery and concerns of respiratory depression from narcotic use are mitigated by ICU level monitoring post-partum. Analgesia needed after vaginal delivery is minimal, while additional analgesic needs following cesarean delivery would be adequately covered by infusing epidural morphine, supplemented by scheduled ketorolac. There are benefits to infusing a low dose epidural LA and narcotic solution. It would provide excellent analgesia, supplement peripheral vasodilation to mitigate the volume load from mobilization, contribute to a decreased risk of DVT formation, and could be used to provide anesthesia if returning to the OR becomes necessary.

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