Laparoscopic Ladd’s Procedure in a Pediatric Patient Post Bi-Directional Glenn Shunt

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Objectives:
1. Discuss the safety of laparoscopic surgery in pediatric patients with single ventricle physiology.
2. Review single ventricle physiology and the impact of pneumoperitoneum.
3. Discuss the management of pulmonary hypertension in a pediatric patient with single ventricle physiology.
4. Discuss the anesthetic management options for pediatric patients with single ventricle physiology undergoing laparoscopic surgery.

Case History:
A 23-month-old male child, 8.6 kg, diagnosed with malrotation of the intestine, heterotaxy syndrome and asplenia is being moved into your operating room for a Ladd’s procedure. You are notified by your resident that the surgeon plans on performing this surgery laparoscopically. He also tells you that this child had a bi-directional Glenn shunt at 6 months of age and repair of his common atrioventricular valve one month ago.

Questions:
What is heterotaxy syndrome? Are there associated anomalies with heterotaxy syndrome? What is a bi-directional Glenn shunt? Any anesthetic concerns with this patient and his planned Ladd’s procedure?
**Case History and Physical Examination:**

This child was born via vaginal delivery at 35 weeks estimated gestational age. He has complex congenital heart disease involving an unbalanced atrioventricular canal defect, double-outlet right ventricle, pulmonary stenosis and pulmonary artery hypoplasia. Shortly after birth he underwent bilateral Blalock-Taussig shunt placement. He underwent surgery for a bi-directional Glenn shunt at 6 months of age and repair of his common atrioventricular valve for moderate-severe regurgitation one month ago. His last cardiac catheterization a week ago reports pulmonary hypertension. He is currently on 1 L of oxygen at home with his oxygen saturation ranging from 63 – 84%. He also has a history of seizures (last one was 4 months ago) and developmental delay. He was admitted this morning, and is currently sitting up in the crib, but starts to cry when you attempt to listen to his heart and lungs. His mother tells you he hasn’t had any problems with his prior surgeries. His current medications include: Ampicillin, Pulmicort, Vasotoc, Pepcid, Lasix, Poly-vi-Sol, Allegra, Viagra, Xopenex and Phenobarbital.

**Questions:**

Is there any further information you would be interested in? Does a cardiac anesthesia team need to be taking care of this child? Your resident is asking if there is anything special you want in setting up the room? Would you like to give any preoperative medications to alleviate separation anxiety?

**Preoperative Studies:**

The child had an echocardiogram done the same day as his cardiac catheterization a week ago which reports mild right ventricular dysfunction, mild left ventricular global dysfunction, and a cleft in the common atrioventricular valve with mild regurgitation. His electrocardiogram shows normal sinus rhythm, right atrial enlargement, and right ventricular hypertrophy. Recent laboratory values are as follows:

- WBC 16.3  HGB 15.5  HCT 46.7  PLTS 464
- Na 141  K 4.3  CL 107  CO2 20  BUN 10  Cr 0.3  Gluc 97
**Questions:**

The surgeon wants to know what is taking so long. He was apparently told that the echo looked okay. The surgeon says it is no big deal since it is just going to be laparoscopic and there won’t be much blood loss. The child will be admitted to the regular ward post-operatively because it is just a laparoscopic procedure.

Do you agree with the postoperative care plan? What are the advantages to laparoscopic surgery for this patient? Do you think this patient is optimized from a cardiac standpoint? Will you proceed?

The nurse in the holding area tells you that neither she nor the nurse from the IV team could start an IV. What are your anesthetic plans? Do you need an IV prior to induction? Do you need a specimen sent for a type and screen? Do you need any invasive monitors for this laparoscopic procedure? Do you think intra-operative TEE monitoring might be useful for this patient? Do you plan to extubate this patient at the conclusion of surgery?

**Intraoperative Course:**

The child is now back in the operating room and crying uncontrollably. It is very difficult to hold him still to attempt an IV start. You proceed with an inhalational induction and the oxygen saturation begins to fall to the 50’s as you attempt to start an IV. What do you think is happening? What will you do next?

With improved mask ventilation the oxygen saturation rises back to 88%. You are able to only place a working 24 gauge IV. Is this adequate for the planned surgery? If you decide to attempt a central venous line where will you place it or does it matter? What is a transpulmonary pressure gradient? What should it be? Do you think you will need an arterial line? If so does it matter where you place the arterial line? The surgeon asks why do you need an arterial line for a laparoscopic surgery—what do you say in response? Does this child need endocarditis prophylaxis?

Are there any recommendations for laparoscopic surgery in a single ventricle patient? If so what are they? Is there any utility to placing a caudal block in this child undergoing a laparoscopic procedure?

The surgery proceeds based on your recommendations. It has now been two hours and you notice the patient is becoming hypotensive despite the fact there has been minimal blood loss. What do you think is the cause of the hypotension? Do you think that he needs volume or is something else going on? If you choose to volume load what will you give—crystalloid, colloid or blood? Should you start an inotropic agent at this time? If so, what inotropic agent will you choose and why?
The patient’s blood pressure rises in response to a bolus of albumin. It has now been two-and-a-half hours, and the surgeon announces that he has to open. He cannot complete the surgery because of visualization problems due to the low intra-abdominal insufflation pressure. Just then the patient’s oxygen saturation falls to 60% and the blood pressure is falling again.

What could be causing this? Is it time to start an inotropic agent now?

You send a blood gas and find out the PaCO2 is 69 despite your ETCO2 reading 36. What causes this discrepancy? You further increase your respiratory rate, yet the oxygen saturation is still in the 60’s and dipping down further along with the patient’s blood pressure. You start to notice the child is now becoming bradycardic.

What else can be done to improve this child’s oxygen saturation? Is it time to start an inotropic agent now?

CPR is started but no significant response. You call for help. A colleague places a TEE probe and notes minimal if any ventricular motion. The patient is emergently taken to the cardiac cath lab. Despite attempted intervention, the child cannot be resuscitated. What happened?

Discussion:

With the improvement in survival now being seen in children with complex congenital heart disease, children with single ventricle physiology are coming to the operating room on a regular basis for non-cardiac surgery. These children will be at various stages in their palliative repair. A large percentage of children with single ventricle physiology also have other associated anomalies and medical diseases, hence it is imperative to have a solid understanding of their unique physiology to be able to formulate a safe anesthetic plan for each individual patient.

The single ventricle pathway consists of three stages: a Blalock-Taussig shunt, bi-directional Glenn shunt and total cavopulmonary shunt (Fontan). The Blalock-Taussig shunt is performed in the first few days of life to serve as the main source of pulmonary blood flow. It is classically described as a systemic-to-pulmonary shunt from the subclavian artery to the corresponding pulmonary artery on that side. Currently, its modification utilizes a Gortex shunt bridging the two vessels. The shunt is either 3.5 mm or 4.0 mm in size based on the surgeon’s choice. Modification of the classical technique may cause less distortion of the pulmonary artery architecture and is technically easier to take-down at the next stage of palliation. The expected oxygen saturation following completion of the BT shunt is 75-85%. A higher oxygen saturation signifies excessive pulmonary blood flow which can result in pulmonary edema and congestive heart failure.
The second stage of palliation is the bi-directional Glenn shunt performed at approximately six months of age. The superior vena cava is anastomosed directly to the right pulmonary artery allowing venous return to flow into both the right and left pulmonary arteries. If the pulmonary arteries are hypoplastic, an augmentation is performed at this time. The BT shunt is taken down as well. The expected oxygen saturation following is 75-85% due to the continued venous admixture from blood returning via the inferior vena cava.

The third and final stage of palliation is the Fontan or total cavopulmonary connection performed at approximately two years of age. The venous return from the lower half of the body is directed to the right pulmonary artery by either an intra-atrial baffle or extracardiac conduit. The use of an intra-atrial baffle and its extensive atrial suture lines is felt to put patients at higher risk for developing arrhythmias later in life. The use of an extracardiac conduit however may place the patient at a higher risk for developing thrombosis or compression of the conduit. The use of fenestrations allows for a “pop-off valve” should the pulmonary vascular resistance become too high to enable adequate flow to fill the systemic ventricle. The expected oxygen saturation would be lower as there is still some venous and arterial admixture with the fenestration.

Upon completion of the third stage, pulmonary blood flow is completely passive and sensitive to changes in pulmonary vascular resistance. The transpulmonary pressure gradient (central venous pressure – atrial pressure) provides an estimate of pulmonary vascular resistance. The model gradient is 7 – 8 mm Hg to insure adequate preload to the systemic ventricle. Factors that increase pulmonary vascular resistance include acidosis, hypoxia, hypercarbia, hypothermia and elevated airway pressures.

The advantages cited for laparoscopic procedures include decreased postoperative pain, shorter postoperative recovery time, decreased postoperative pulmonary dysfunction and a decreased incidence of wound infection. However, despite these advantages, laparoscopic techniques can have detrimental effects on single ventricle physiology. Laparoscopy involves the use of carbon dioxide (CO2) insufflation to create a pneumoperitoneum. Studies have demonstrated an increased gradient between arterial CO2 and end-tidal CO2 in children with cyanotic congenital heart disease, as well as the unreliability of peripheral pulse oximetry with an oxygen saturation < 80%. The absorption of CO2 leading to hypercarbia and acidosis can raise pulmonary vascular resistance impeding forward flow, as well as increase the risk for arrhythmias. Due to the lack of an active pumping chamber on the venous side, impediment of forward flow can cause stagnation on the venous side leading to possible shunt thrombosis. The increased abdominal pressure decreases venous return which leads to diminished systemic preload and cardiac output. The increased abdominal pressure also limits ventilation leading to decreased tidal volumes, atelectasis and V/Q mismatch which can worsen existing cyanosis. Increasing minute ventilation to counteract changes related to CO2 absorption can further worsen the already compromised ventilation. Pneumoperitoneum can also increase systemic vascular resistance which causes increased workload on the functioning single ventricle leading to
myocardial dysfunction. All of these physiologic changes can provide for a significant negative impact on an already compromised or tenuous single ventricle circulation.

Treatment for pulmonary hypertension includes the use of parenteral or inhaled pulmonary vasodilators. Oxygen is a strong pulmonary vasodilator which is readily available in combination with hyperventilation to lower elevated CO2 levels. The use of nitrates such as nitroglycerin or nitroprusside tends to have strong systemic vasodilating effects in addition to their pulmonary vasodilatory effects. Milrinone, a phosphodiesterase inhibitor, allows right ventricular support and pulmonary vasodilation, but again has been associated with systemic vasodilation requiring norepinephrine use to counteract it. An increase in the systemic vascular resistance causes greater work for the existing single ventricle to pump against. Other pulmonary vasodilators include intravenous prostacyclin or its synthetic analog iloprost. Inhaled nitric oxide administered at 5 – 40 parts per million is probably the most selective pulmonary vasodilator available, yet carries the risk of methemoglobinemia. Sildenafil and Bosentan use is becoming more prominent in pediatric congenital heart disease for maintenance therapy, but there is very limited clinical research data.

From the limited number of case reports and small series of patients with single ventricle physiology undergoing laparoscopy for gastrostomy or Nissen fundoplication, a few recommendations can be made. In hopes of minimizing increased intra-abdominal pressure affecting venous return and ventilation, CO2 insufflation should be limited to 8 – 12 mm Hg as a decrease in systemic cardiac output was noted with higher insufflation pressures of 15 – 20 mm Hg. All patients in these case reports received an arterial line regardless of age or stage of palliation. An arterial line provides a rapid way to measure CO2 or PaO2 should any question arise, as well as providing a means for closer monitoring of the patient’s volume status and systemic cardiac output, especially if any arrhythmias occur. A good working intravenous (IV) catheter to maintain adequate volume status is vital. It goes without saying that surgical time should be minimized if possible.

In regards to the choice of anesthetic management, there is no “best anesthetic” for this group. The main thing to keep in mind is that children with cyanotic congenital heart disease have an increased risk for morbidity and mortality for both major and minor non-cardiac surgical procedures. An IV line prior to induction is considered to be a luxury depending on the age of the patient. These patients have usually undergone a number of procedures, not necessarily surgical, and are small for their age. This can make IV access quite difficult especially for those less than 1 year of age. An inhalational induction with sevoflurane can be safely accomplished, but remember to avoid stimulation when the child is still “light” because this can trigger an increase in pulmonary vascular resistance and subsequent drop in oxygen saturation. Maintenance anesthetic choice truly depends on whether you think that you will be able to extubate the child at the end of the procedure. This decision is based heavily on their pre-operative functional status. One would be more inclined to opt for a high-dose narcotic technique for a child who has marginal functional status evidenced by an increasing need for
oxygen at home and demonstrating little weight gain, as opposed to a child who has a room air
oxygen saturation of 83% and eating well at home with appropriate weight gain. However, with
a high-dose narcotic technique this will require post-operative ventilation placing the child at risk
for subsequent respiratory infection. The risks and benefits of each choice must be carefully
weighed. All children with single ventricle physiology, even if extubated at the conclusion of
surgery should be monitored at least overnight in an intensive care setting.

Anesthetic management requires individualization because there are typically many
conflicting factors involved. There is no simple answer.

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