“Just a quick add-on”: A Young Woman with Failing Fontan Physiology and Respiratory Distress for Paracentesis

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Goals:
- Discuss key and unique aspects of Fontan anatomy and physiology
- Summarize the long-term implications of single ventricle palliation for congenital heart disease
- Describe complications and management issues related to large-volume paracentesis alone and in the context of Fontan physiology
- Describe the advantages and disadvantages of different approaches to airway and ventilatory management in this setting
- Discuss major facets of perioperative pacemaker management and Fontan-associated rhythm disturbances

Case history:
The patient is a 15-year-old, 55kg female with hypoplastic left heart syndrome, palliated more than 10 years ago by complete cavo-pulmonary anastamosis (Fontan), who presents for large-volume paracentesis.

Questions:
What is hypoplastic left heart syndrome (HLHS)?
How often does it occur?
What would be the natural progress of HLHS?
What are the three stages of today’s operative management of HLHS?
What is the Fontan operation?
What is the difference between intracardiac and extracardiac Fontan?
What is a fenestrated Fontan and what purpose does the fenestration serve?
Discuss aspects of “normal” Fontan physiology: Expected oxygen saturation, sensitivity to intravascular volume, sensitivity to spontaneous and controlled ventilation.
Describe how these factors influence your anesthetic plan.

Case history and physical examination (continued):
Her degree of ascites has increased over the past several months resulting in abdominal discomfort, nausea, and anorexia, now further complicated by increasing respiratory difficulty. The etiology of her ascites is uncertain, but has been generally ascribed to “failing Fontan” physiology. She has not been considered as a cardiac transplantation candidate due to noncompliance with medications and inability to quit smoking.
Questions:
Describe the main issues of “failing” Fontan physiology.
What factors are thought to be related to protein losing enteropathy?
Describe complications and management issues related to large-volume paracentesis in the context of Fontan physiology.
What would be your strategy for volume replacement?
Which fluids would you use?

Case history and physical examination (continued):
She also has an epicardial pacemaker for sick sinus syndrome. Aside from the ascites she is thin with a normal airway exam. She is anxious and crying. Her respiratory rate is 25 with room air oxygen saturation 85%. Noninvasive blood pressure is 90/60, heart rate is atrially paced at 70 bpm. She demands to be “totally out” for this procedure.

Questions:
So what is your anesthetic plan? Local vs. sedation (MAC) vs. general anesthesia?
What is your plan for IV access?
Do we need any special monitoring and if so, what would you use?
What airway management would you choose and how would you obtain it?
What are your anesthetic hemodynamic goals for this patient?
What would you use for induction of anesthesia/sedation?
What would you use for analgesia?
Would you order any preoperative studies?

Preoperative studies:
A recent echocardiogram showed mildly depressed RV function, moderate tricuspid regurgitation, and an open Fontan fenestration with flow into the right atrium.

Questions:
Is an ECHO typically sufficient cardiac evaluation in Fontan patients?
How that relates to this minor procedure we are planning on doing?
What rhythm disturbances can be seen in these patients?
Do you need to interrogate the pacemaker?
Where might the pacemaker generator be located?
What type of leads might this patient have?
Is there anything else needed to optimize cardiac status prior to surgery?
What risks and possible complication should be disclosed to the patient?
Would you mention death as a possible complication?

Intraoperative care:
The patient has an uneventful induction of anesthesia, but becomes hypotensive with a blood pressure of 60s/30s.

Questions:
How do you respond?
What vasoactive medications would you consider using for this patient?
Postoperative care:
The patient emerges uneventfully and wants to go home as soon as possible after the procedure. The cardiologist mentions that there are no beds available in the ICU. He could arrange admission to a monitored floor bed, but the patient would have to go to PACU first.

Questions:
What admission or discharge arrangements would you make for this patient?

Discussion:
The Fontan procedure has been performed since the 1970s. Typically the final, total cavopulmonary anastomosis physiologic arrangement is achieved after a series of surgeries. The exact treatment course varies somewhat, but most frequently patients are treated with a neonatal shunt procedure, a bidirectional superior cavopulmonary anastomosis (Glenn procedure), followed by the total cavopulmonary anastomosis (Fontan). Different methods of attaining total cavopulmonary anastomosis have been preferred at one time or another. A connection from the right atrium to pulmonary artery may be seen in patients who underwent their operation mostly before 1990. However the atrio-pulmonary approach is unlikely to be performed today. The prevailing approaches today include the intracardiac (lateral tunnel) Fontan and the extracardiac Fontan.

Intracardiac Fontan: The conduit directing inferior vena cava blood to the pulmonary artery is located within the right atrium. The intracardiac Fontan is commonly preceded by a hemi-Fontan procedure in which the connection between the superior vena cava and right atrium is preserved even though blood flow between the superior vena cava and right atrium is halted. A fenestration can be created in the Fontan circuit. These patients are at higher risk for arrhythmias in comparison to the extracardiac patients.

Extracardiac Fontan: The conduit directing blood from the inferior vena cava to the pulmonary artery travels outside the heart. Similar to the intracardiac Fontan a fenestration can be left in the Fontan circuit. Currently, this approach seems to be favored.

Patients with Fontan physiology are surviving to adulthood. These patients can be expected to present for noncardiac surgery, and some may decide to become pregnant. A one-size-fits-all approach to anesthetic management is inappropriate, but certain principles deserve routine consideration:
Details of cardiac function must be known. Recent echo and cardiac catheterization will review important details such as contractility, valve function, vascular stenoses, condition of the atrial septum (restrictive or unrestrictive), presence of collateral vessels and the status of a fenestration.
One must carefully review of surgical and medical history. Older patients may not have had the precursor surgeries you are accustomed to. (BT shunt versus modified BT shunt, hybrid stage 1 procedure). Patients may have had other relevant procedures such as treatment for subglottic stenosis, etc. These patients may have compromised vascular access secondary to numerous central lines placed, multiple cardiac
catheterizations, or ECMO. A history of arrhythmia and/or pacemaker is crucial. Single ventricle patients are prone to rapid hemodynamic decline with cardiac arrhythmias. Patient-specific data can then be placed in the context of single ventricle Fontan physiology. Pulmonary blood flow is passive, and driven by systemic venous pressure. Overall circulatory function will be best with low pulmonary vascular resistance. Vigilant, thoughtful, high-quality anesthesia results in low pulmonary vascular resistance, and is usually adequate for managing pulmonary vascular resistance. Hyperventilation is not useful. Prophylactic pulmonary vasodilators (Nitric oxide or Epoprostenol) are not routinely indicated. When feasible, spontaneous (negative pressure) ventilation is preferred over positive pressure ventilation. Furthermore, adequate venous blood return is necessary for pulmonary perfusion. Hypovolemia or relative intravascular depletion should be avoided. Situations of diminished cardiac output or contractility will be poorly tolerated. Regardless of the selected anesthetic technique, maintenance of cardiac output is a core consideration as the single functioning ventricle must provide pulmonary as well as systemic perfusion.

References:


