SPA PBLD, Spring 2015

Title: Anything's Possible in an Adult with Failing Fontan Physiology (…especially at your local Children's Hospital!)

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Goals/Objectives:

1. Recognize the causes, manifestations, and anesthetic risks of a failing Fontan circulation.
2. Describe the different types of Fontan circulations that may be encountered with their associated complications.
3. Assess the pros and cons of performing an anesthetic in an adult at a children's hospital, and what, if any, unique considerations should be anticipated.
4. Reflect on the impact of performing a complicated anesthetic in a remote location as opposed to an operating room.

Case history:

A 20 y/o male with a h/o tricuspid atresia s/p single ventricle palliation presents at your children's hospital with a perforated appendicitis. He has received all of his previous care at this hospital, including his fenestrated Fontan. He is scheduled now for an IR-guided intraperitoneal abscess drainage.

Questions:

1. What is a Fontan circulation?
    a. Are there different types? Are these differences significant?
    b. What is the significance of a fenestration?

The IR charge nurse with whom you've been working informs you that this case needs to be done urgently. The surgical team has called several times to expedite the process. You're asked if it's OK to send for the patient.

Before answering, you take a moment to review the patient's EMR and note that patient's last cardiology visit was 4 months ago. In the history, you note that he's complained recently of increasing shortness of breath, particularly with exertion, and increasing cyanosis (his friends have commented that his lips look blue after walking up the stairs at school). Recently, his socks and shoes also seem to feel a bit tighter, and he's noted that his abdomen is noticeably larger than in the past. While he's still eating, his appetite isn't what is used to be.

Questions:

2. What is a failing Fontan circulation?
    a. Does he meet criteria?
        i. What would you want to know to help decide this?

An echocardiogram at the last clinic visit demonstrated moderately depressed left ventricular function. His last cardiac catheterization, approximately 1 year ago, showed a Fontan pressure of 18mmHg. Multiple veno-venous collaterals were noted and subsequently coiled without an increase in Fontan pressure.

Questions:

3. What do you think about the Fontan pressure of 18mmHg?
    a. What are the possible causes?
    b. What else would you want to know from the cardiac catheterization?
4. What else would you look for from the echocardiogram aside from ventricular function?
5. Is it possible to have a failing Fontan with preserved systolic function?

The IR charge gently taps you on the shoulder and asks if you've made up your mind.

Questions:

6. Do you need any additional information to help with this decision?
7. Would you delay the procedure and request further preoperative workup prior to anesthetizing the patient?
    a. If so, what tests would you request or who would you consult?

An echo was performed on admission demonstrating essentially unchanged function and no change in mitral regurgitation (mild-moderate). The Fontan baffle appears unobstructed.

Current medications include ASA, budesonide, enalapril, and furosemide

Cardiology states that the patient is "as good as he's going to be," and the pediatric cardiac anesthesiologist who would have been available to help is now involved in an emergent TAPVR repair.

Questions:

8. Who should do this case?
    a. General anesthesiologist, pediatric anesthesiologist, pediatric cardiac anesthesiologist?
9. Where should you do it?
    a. Remotely located IR suite, general operating room, or cardiac operating room?
10. Is it appropriate to do this case at a children's hospital?
    a. Would your opinion change if it were a 40 year old?

You decide to proceed with the case in the IR suite, and the IR charge nurse calls for the patient. When the patient arrives to the holding area, you note that she has an NG tube to suction which is draining copious gastric secretions.

The patient's vital signs are T: 38.7, P: 115, BP: 78/50, RR: 24, SpO2: 92% on RA. On exam, she appears uncomfortable, has bilateral basilar crackles, abdominal tenderness, and edematous lower-extremities. One peripheral IV is present in the right arm but is not attached to any fluids.

Questions:

11. Do you have any concerns about the patient's vitals/physical exam?
    a. What are causes of desaturation in a Fontan?

12. How would you manage this patient?
    a. Would you consider MAC?
    b. Would you intubate and do a GA?
    c. Would you have any additional set up (ie, invasive lines, nitric, etc)?
    d. Does being in IR have any influence on your plan?

13. If you decide to do a GA, what drugs would you use to induce/muscle relax?

You decide to proceed with a general endotracheal anesthetic with a pre-induction arterial line. The induction is thankfully uneventful with no hemodynamic instability, but shortly afterwards you note the following vitals: P: 125, BP: 68/38, volume control: TV: 500, RR: 12, PEEP: 5, PIP 24, SpO2 90% on an FiO2 of 0.5.

Question:

14. What could explain the decrease in blood pressure and saturation?
    a. How would you manage this?

You decide to extubate at the end of the hour-long procedure with the vital signs having improved following your resuscitation to: P: 105, BP: 102/58, RR: 12, TV: 350 on SV, SpO2: 100% on an FiO2 of 0.6. The surgical service wishes to admit the patient to the surgical floor postoperatively.

Question:

15. Do you agree with this post-op location?

References:

1. Beyond a broken heart: circulatory dysfunction in the failing Fontan. Mori M, Aguirre AJ, Elder RW, Kashkouli A, Farris AB, Ford RM, Book WM. *Pediatr Cardiol*. 2014 Apr; 35(4):569-79.
2. Medical management of the failing Fontan. Ghanayem NS, Berger S, Tweddell JS.
   Ghanayem NS, Berger S, Tweddell JS. *Pediatr Cardiol*. 2007 Nov-Dec; 28(6):465-71.
3. Salvaging the failing Fontan: lateral tunnel versus extracardiac conduit. Morales DL, Dibardino DJ, Braud BE, Fenrich AL, Heinle JS, Vaughn WK, McKenzie ED, Fraser CD Jr. *Ann Thorac Surg*. 2005 Oct; 80(4):1445-51.
4. Adult congenital heart disease. Seal R. *Paediatr Anaesth*. 2011 May; 21(5):615-22.
5. The Fontan patient. Bailey PD Jr, Jobes DR. *Anesthesiol Clin*. 2009 June; 27(2):285-300.

6. The Fontan Circulation. Gewillig M. Heart 2005;91:839–846.

7. Four Decades of Fontan Palliation: A Review. de Leval, M. R. & Deanfield, J. E. *Nat. Rev. Cardiol.* 7, 520–527 (2010)