

Retrograde Coarctation in a Hybrid Stage 1 Hypoplastic Left Heart, Should I Be Concerned?

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Goals:

1. Describe the anatomy in hypoplastic left heart syndrome
2. Describe the different palliation procedures, including hybrid approach
3. Formulate an anesthetic plan for a patient with stage 1 palliation
4. Familiarize with complications of stage 1 palliation

A 3 month old boy with Hypoplastic Left Heart Syndrome s/p Hybrid Stage 1 palliation is scheduled for a laparoscopic Nissen Fundoplication.

1. What is Hypoplastic Left Heart Syndrome?
2. What are the palliation options? What is a hybrid palliation?
3. What preop data would you like before need to proceed?
4. What are the hemodynamic challenges with a laparoscopic procedure? Would you advocate for an open fundoplication?

TTE 2 weeks ago shows: RV functions is normal, minimal tricuspid regurgitation, ASD is widely open, and a retrograde aortic coarctation with a gradient of 10mmHG. EKG shows sinus rhythm. The baby is on aspirin, furosemide, ranitidine, enalapril, and enoxaparin. The cardiologist has deemed him as optimized for the procedure. VS: BP upper body 84/36, lower body 68/24, Sat 85% on the right arm. Wt 4.5kg Ht 55cm RR 40 Temp 36.5 C. Labs: Htc 44% Plts 241 BUN 18 Creat 0.6

5. What is your anesthetic plan?
 1. Induction: inhaled? IV? IM? Maintenance?

2. Monitors, arterial line? NIRS? Does the location of SpO₂ and A line matter?

3. Access, central line?

4. Blood products available? Irradiated? Leukoreduced? Washed?

5. PACU or ICU post op?

6. Extubate or leave intubated?

6. What is your target saturation? How high or low FiO₂ would you tolerate?

7. What is your target hematocrit? What would be your transfusion trigger?

You proceed to start the case. The induction and intubation were uneventful. You managed to place two 22g IVs and a femoral arterial line. Pre and post ductal pulse oximeters read 81% and 85%, respectively, on room air. Arterial line reads 64/28. ETCO₂ is 38. NIRS are reading low 40's not far from baseline. Blood gas is pH 7.35 pCO₂ 32 pO₂ 47 Hct 42% BE 3.4 Lac 1.1

8. How does the NIRS monitor work?

9. How would you use NIRS to guide your anesthetic? Which sites can be monitored?

The procedure starts and a pneumoperitoneum is created. Insufflation pressures are ~10mmHg. BP 59/26 NIRS 35/37 Sat 74%. Blood gas pH 7.25 pCO₂ 48 pO₂ 36 Hct 39% Lact 3.1

10. Would you treat the blood pressure? The NIRS?

11. Would you increase the FiO₂?

The case continues. The blood pressure continues drifting down. You notice PVCs and ST segment changes on ekg. BP 55/24 NIRS 31/35 Sat 70% Blood gas pH 7.20 pCO₂ 45 pO₂ 34 Hct 35% Lact 6.2

12. What would be your response to worsening hemodynamics?

13. What is your differential diagnosis?

14. Would you call pediatric cardiology to the OR?

15. Do you abort the surgery?

Pediatric cardiology arrives and performs a TTE. They report that heart function is moderately depressed and that the retrograde coarctation gradient is ~18mmHg.

16. What is the next step in management?

17. Do you need more lines? Would another a line help?

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