PBLD #31: Navigating the Anesthetic Challenges Associated with Evaluation and Treatment of a Child with Newly Discovered Suprasystemic Pulmonary Hypertension

Rachel G. Clopton, MD; Richard J. Ing, MB BCh FCA (SA)
Children's Hospital Colorado

Objectives

Children with newly suspected pulmonary hypertension (PH) must undergo an extensive and invasive evaluation in order to establish a definitive diagnosis of PH and to identify an underlying cause. In this problem based learning discussion, a previously healthy 14kg 4 year old girl with new-onset syncope, thrombocytopenia, and echocardiographic evidence of suprasystemic PH must undergo multiple anesthetics in a variety of locations during the initial work-up to characterize her newly-discovered disease. The learning objectives for this discussion are:

1. To identify the common signs, symptoms, and causes of PH in the pediatric population.
2. To review the considerations for general anesthesia and sedation in the patient with untreated suprasystemic PH undergoing investigative procedures.
3. To discuss the thoracic surgical risks in a patient with suprasystemic PH.
4. To catalog the preparations required for emergency surgery in a critically ill patient with suprasystemic PH.

Case History

A previously healthy 14kg 4 year old girl who lives on a farm presented to her PCP with a 6-week history of fatigue, cough, and dyspnea. Her mother also reported mild snoring. She had a syncopal episode during the appointment and was transferred to a local Emergency Department for further evaluation. Laboratory studies were remarkable for thrombocytopenia (platelet count 50,000/μL). CXR showed reticulonodular interstitial opacities and central vascular prominence (see Figure 1). She was diagnosed with community acquired pneumonia, prescribed a course of augmentin, and referred to a hematologist who auscultated a murmur. The patient was sent to a cardiologist who performed a trans-thoracic echocardiogram which estimated her right ventricular systolic pressure to be 140mmHg, coinciding with a systemic systolic blood pressure of 90mmHg. She was transferred to Children's Hospital Colorado for further evaluation and treatment of newly discovered suprasystemic PH.

Questions

1. What is the definition of PH? What are the most common presenting signs and symptoms?
2. How is the classic presentation different in children compared to adults?
3. What is the differential diagnosis for new PH (not associated with congenital cardiac disease) in a child? How do the thrombocytopenia and findings on CXR affect this list?

Case History Continued

The patient underwent a CT Scan that showed lymphadenopathy, numerous pulmonary nodules, enlarged pulmonary arteries, and engorged bronchial arteries. However, laboratory testing for infectious or rheumatologic causes were negative. Therefore, she was scheduled for bronchoscopy, PICC line insertion, cardiac catheterization with atrial septostomy, and thoracoscopic lung biopsy in the hybrid cardiac catheterization laboratory/operating room.

Data on DOS:

Temp 36.6 °C HR 83 BP 121/55 SpO2 96% on 1LPM O_2 via NC RR 23

TTE: moderate to severe tricuspid regurgitation, Vmax 6.33m/s estimating RVSP of 160mmHg (BP=127/60). Severe flattening of the ventricular septum, bowing into the left ventricle. Severe right ventricular enlargement with moderately decreased systolic function. Mild right ventricular hypertrophy. Moderate right atrial enlargement. Left ventricle was compressed by the right ventricle, subjectively normal left ventricular systolic function. No change in response to oxygen.

Hct 29.6%, Platelets 186,000/μL

Airway exam was normal.

Questions

1. Discuss the anesthetic management of a patient with suprasystemic PH in the cardiac catheterization laboratory undergoing multiple diagnostic procedures. Is ECMO stand-by necessary?
2. What is your plan for induction and emergence? Assuming no mis-adventures in surgery, will you plan to extubate in the OR or transfer to PICU intubated and sedated?

Intra-operative Management

Bronchoscopy findings were unremarkable. Cardiac catheterization confirmed suprasystemic PH, non-reactive to oxygen and inhaled nitric oxide (see figure 2). No pulmonary artery or pulmonary vein stenosis was detected. An atrial septostomy was performed.

Questions

1. What are the risks and benefits of atrial septostomy in this patient?
2. Discuss the increased risks associated with thoracoscopic surgery in this patient. Should lung biopsy proceed now or wait?
3. During left thoracoscopic wedge resection, bleeding was noted at the biopsy site. At what point should blood transfusion be considered?
4. The EKG shows ST changes. What is the cause? What is your treatment strategy?
5. The procedure is over and she is ventilating spontaneously with a RR of 20 and TV 8-10ml/kg. Would you extubate her? If not, what is your post-operative sedation plan?

Post-Operative Course

The patient was transferred to the PICU for post operative monitoring. Approximately 3 hours after her arrival to the PICU, she became tachypneic, pale, hypotensive (BP 70s-80s/50s. baseline SBP 100s-120s), tachycardic with ST-segment depression, and hypoxic with SpO2 in 70s on increased oxygen and 40ppm iNO. The PICU attending calls anesthesia for help with further management.

Questions

1. What is the differential diagnosis for her current hemodynamic changes and desaturation?
2. Could this be a pulmonary hypertensive crisis? How would this patient have looked prior to her procedures in the setting of a pulmonary hypertensive crisis?
3. What tests would you ask the PICU attending to order?
4. Would you call anybody else?
5. ST-Changes are profound with systolic blood pressures <120mmHg. Why? Discuss an induction plan for this patient.

Further Testing

Lung biopsy failed to identify a cause of PH, showing plexiform arteriopathy characteristic of severe PH. Extensive work up continued to be unrevealing. Clinically the patient's symptoms of right heart failure and PH began to improve after initiation of therapy including: treprostenil infusion, prednisolone, lasix, and sildenadil. However, she continued to suffer from intermittent fevers and thrombocytopenia. At this point (3 weeks after her first anesthetic), the primary team requested anesthesia for a PET-CT Scan, scheduled for 1 hour. She was required to lie completely still but breath holds were not necessary.

The most recent TTE showed:

Atrial septal defect (s/p balloon septostomy) with left to right shunting. Moderate right atrial enlargement. Moderate tricuspid regurgitation Vmax of 5.5 m/sec, corresponding to RVSp 121 mmHg (SBP 155mmHg). Moderate flattening of the ventricular septum. Moderate right ventricular enlargement with low normal systolic function. Hyperdynamic, underfilled left ventricle. Moderate hypertrophy of the right ventricle. Compared to the previous study, the right ventricular function has improved with left to right atrial level shunt and moderate septal flattening.

Questions

1. What is your anesthesia plan for this radiologic study?
2. What is your plan for airway management?
3. Discuss medication considerations for this procedure.

Ground Hog Day-Another lung biopsy

Unfortunately, an identifiable cause of the patient's PH remained elusive. In a final attempt to completely rule out a neoplastic condition that would exclude the patient from candidacy for lung transplantation, she was scheduled for bone marrow biopsy, repeat bronchoscopy, port placement (for chronic intravenous pulmonary vasodilator therapy), CT localization of a prominent pulmonary nodule, and open biopsy of the pulmonary nodule.

Questions

1. Based on her surgical history, how would you plan anesthetic approach this time?

A Happy Ending

No underlying etiology could be identified, so the patient was diagnosed with idiopathic primary PH. She was discharged home after a 46 day hospitalization on the following medication regimen: amlodipine, treprostenil continuous infusion, prednisolone, spironolactone, warfarin, furosemide, lansoprazole, cellcept, and oral sildenafil.

Her most recent TTE (~3 months after discharge) showed RVSp 56mmHg, corresponding to a SBP 118/73. She is an active 4 year old going to preschool.

Discussion

For a child with newly discovered PH, the path from initial discovery to final diagnosis can be treacherous. The disease is often diagnosed late because early symptoms are vague (fatigue and cough) and the disease is rare (incidence 1/100,000)[1]. At least one anesthetic to facilitate cardiac catheterization is required to obtain definitive diagnosis. Children with PH are at increased risk for perioperative complications, with patients with suprasystemic pulmonary arterial pressures being the most vulnerable. According to Carmosino and colleagues, major perioperative complications, including cardiac arrest and pulmonary hypertensive crisis, occurred in 4.5% of children with PH undergoing procedures under anesthesia. Additionally, among children with PH, the presence of suprasystemic PH further increases the risk of major complications eight fold.[2]

This PBLD demonstrates that multiple techniques for anesthesia can be used safely in a patient with suprasystemic PH. Regardless of technique chosen, great care must be taken to ensure adequate myocardial perfusion and to avoid acute increases in PVR and right ventricular dysfunction, which could precipitate a pulmonary hypertensive crisis under anesthesia.

Times of airway manipulation are fraught with danger for patients with suprasystemic PH. Adequate depth of anesthesia must be achieved prior to any planned airway instrumentation, as tracheal stimulation has been associated with significant increases in PVR in children with PH.[3]

The third anesthetic highlighted options for deep sedation in patients with PH. With the use of supplemental oxygen and monitoring of end-tidal carbon dioxide, no signs of increased pulmonary arterial pressures were observed and this is consistent with newer literature on the topic.[4,5]

Finally, there is a paucity of literature regarding thoracic surgical complications associated with PH.

References

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Further Reading

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Figures

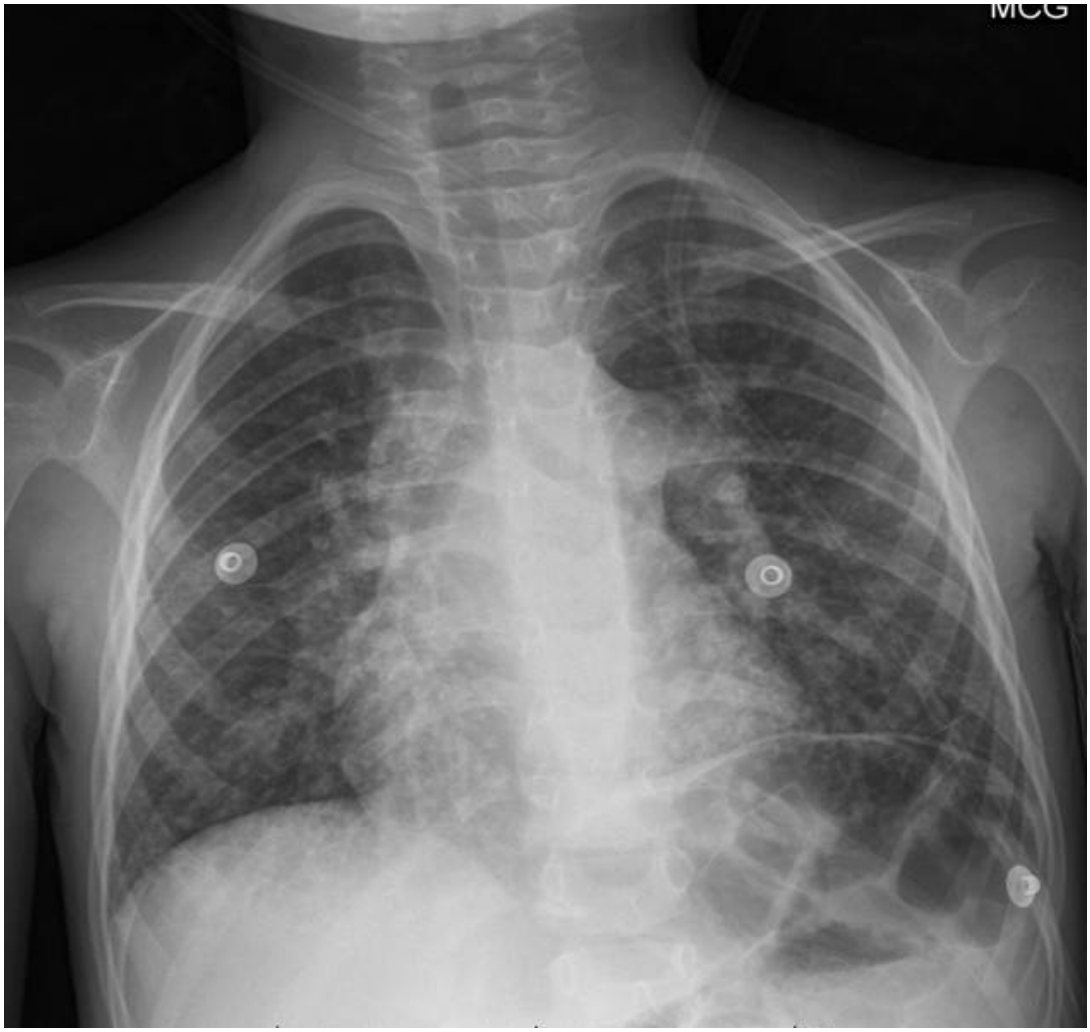

Figure 1: initial CXR showing reticulonodular interstitial opacities and central vascular prominence.

Figure 2: Pulmonary arterial pressure waveform (yellow) superimposed on systemic pressure waveforms (red) during administration of 33% oxygen confirming the presence of suprasystemic PH.