Emergent Appendectomy in a Pediatric Cardiac Patient

Stuart Hall, MD
Houston, TX

Scott Markowitz, MD
Philadelphia, PA

You’ve finished your scheduled cases for the day, and you learn that an emergency appendectomy with placement of PICC line has been put into your room. The patient has been brought to your preoperative holding area, and on arrival, you read the emergency room history and physical.

The patient is a 19-year-old male with trisomy-21 and a history of a complete AV-canal defect which was repaired at age 3 months. He is moderately developmentally delayed but is relatively cooperative. He presents to the emergency department with a one-day history of worsening abdominal pain which began as periumbilical pain but which now localizes to the right lower quadrant. He has had neither emesis nor fever, but has had loose stools for the past 24 hours. His review of systems reveals chronic nasal congestion, thought to be due to seasonal allergies.

He was admitted to the hospital 5 months ago for atrial flutter for which he was heparinized and treated with synchronized DC cardioversion. He was discharged from that admission on atenolol and coumadin. His mother mentions that he has “bad” mitral regurgitation and is scheduled to have cardiac surgery in 2 days to repair or replace his mitral valve, and in preparation he has not taken coumadin for 5 days. She also mentions that her son had an orchiectomy two years ago for a testicular mass. He has no history of anesthetic complications.

Allergies: none. Rx: digoxin 0.25mg qD, atenolol 50mg BID, furosemide 20mg BID, enalapril 5mg BID, coumadin (last dose 5 days ago), montelukast 10mg qHS, and nasal steroids. In the ER he received IV ampicillin, gentamicin, and clindamycin, along with a single 15mg dose of ketorolac.

Vital signs: wt 65.6 kg, BP 99/49, P 79, RR 20, T 99°F (PO), SpO₂ 99% on RA. Physical exam reveals a well-developed, obese, acyanotic male with typical trisomy-21 facies, good oral opening with a large tongue, neck with full range-of-motion. Cardiovascular exam reveals regular rate and rhythm, a III/VI systolic murmur at the LUSB and good peripheral pulses; he has no cyanosis, clubbing, nor edema. His abdominal exam shows diffuse pain maximal over the RLQ with rebound tenderness. He has a peripheral IV in the left antecubital fossa through which 0.9% NS is infusing at 100 cc/hr.
Labs: Na 134, K 4.8, Cl 98, CO₂ 25, BUN 13, Cr 1.3; H/H 16.9/50, WBC 15.6, Plt 165; PT 18.3, INR 1.5.
EKG: NSR, RBBB, LAD CXR: mild-moderate cardiomegaly with LA dilation and mildly increased PVM Ultrasound examination of the abdomen is consistent with acute appendicitis.

Questions:
1. What other information do you need before you anesthetize this patient?
2. What are the anesthetic implications of Down syndrome?
3. Does this patient require resuscitation prior to induction?

As you look over the chart and make your way to evaluate the patient and talk with the family, a cardiology fellow rushes in and apologizes for not having put her consultation on the chart sooner.
The consultation gives further detail about the admission for atrial flutter. A transesophageal echocardiogram performed at that time showed severe AV-valve regurgitation and found no evidence of thrombi. His last full echocardiogram shows severe left atrial dilation, a thickened mitral valve with a cleft anterior leaflet which has mild stenosis (5-6 mmHg gradient) and severe mitral regurgitation. LVEDD is normal and LV function is normal to slightly hyperdynamic (SF 45%). There is trivial tricuspid regurgitation.

Recommendations from the cardiology service include holding digoxin while the patient is NPO as well as keeping the patient off coumadin for the present time. They recommend SBE prophylaxis prior to surgery and suggest that the patient be admitted to a telemetry unit after surgery. They also write that “cardiac anesthesia is required for surgery.” The cardiology fellow tells you that they are quite concerned that the patient not become volume-overloaded, since the cardiologists worry that any volume load might stretch the left atrium and precipitate an atrial tachydysrhythmia that might not be well-tolerated hemodynamically. She says that they ordered a dose of furosemide to be given in the ER, but she is unable to determine if he received the diuretic.

Questions:
4. How does the consult affect your thinking about the patient?
5. Describe the anesthetic goals for patients with mitral valve disease.
6. What is “cardiac anesthesia?”
7. Can the mother give informed consent for this procedure?

In the OR, you apply ASA standard monitors, including pulse oximeter, noninvasive BP cuff, EKG, and an axillary temperature probe. You also place transcutaneous pacing/defibrillation pads. Preinduction vital signs are HR 90, BP 118/70, RR 20, SpO\textsubscript{2} 100%. You preoxygenate with FiO\textsubscript{2} 1.0 and then induce using a modified-rapid-sequence technique with cricoid pressure, giving the patient fentanyl 50mcg, etomidate 12mg, and succinylcholine 100mg IV. Laryngoscopy is easy with a Miller 3 blade and you intubate the trachea. The first BP after induction is 78/50; the next reading 52/18. The end-tidal CO\textsubscript{2} reads 12 mmHg. The pulse oximeter continues to read 100%, but the waveform is quite blunt. Heart rate is now 88 bpm.

Questions:
8. Comment on this induction, Is the monitoring adequate?
9. What is your differential for the decrease in BP? The decrease in ET CO\textsubscript{2}? Tx/Rx?
10. Where should this patient go after surgery?

During your survey of the patient and equipment, you notice that the sampling line has become dislodged (but not disconnected) from the gas analyzer. You begin a crystalloid bolus and place an arterial line while the surgeon places a PICC line in the right antecubital fossa. The arterial blood pressure reads 112/65, the HR 88, and SpO\textsubscript{2} is 100% on FiO\textsubscript{2} 0.4. An arterial blood gas measurement taken under these conditions shows pH 7.48, PaCO\textsubscript{2} 30, PaO\textsubscript{2} 371, BE +1, Hct 51%, Na 135, K 4.5, iCa++ 1.1, glucose 124, serum lactate 2.1.

Questions:
11. Is it appropriate to proceed with surgery?
12. In light of the patient’s response to induction, what does the blood gas tell you?
13. Do you plan to extubate?
Model Discussion

Endocardial cushion defects such as a complete atrioventricular canal defect occur with a frequency somewhere between 0.11-0.36/1000 live births, and such defects comprise approximately 4% of all congenital heart defects. Trisomy-21 or Down syndrome is the most common cause of canal-type defects. The specific genetics of this lesion are beyond this discussion, but it will come as no surprise that a large region of the chromosome 21 is implicated. In one institutional survey, 80% of infants under one year of age with canal-type defects had Down syndrome; conversely, 35-40% of patients with Down syndrome have canal-type defects.

The most common indication for reoperation after repair of a complete AV-canal defect is left AV-valve regurgitation. The most common indication for exploratory laparotomy in children in North America is acute appendicitis. It has been encountered in all age groups, from newborns to adults, but its incidence peaks from ages 6 to 10 years. In all children, 30-50% have perforated by the time of surgery. Appedicitis is on the differential for any child with fever, malaise, loss of appetite, or other nonspecific constitutional symptoms. Perforated appendicitis can lead to peritonitis, intra-abdominal abscesses, empyema, and bowel obstruction.

Questions 1-4: Preoperative evaluation and management

There are several ways to approach the congenital heart patient who presents for noncardiac surgery. Often, it can be helpful to address the cardiac issues first. Is the patients cardiac disease of primary concern to the situation, or is it a secondary issue which may or may not impact management of the presenting complaints? Second, what is the nature of the planned procedure? In the case of emergency surgery, or surgery to be performed on a child who is either unrepaired or palliated, the patients are often more critically ill and may require very close attention to their preoperative management.

When available, old medical records can give details on previous surgeries and interventions, and can give an overall impression of the childs clinical course. Diagrams, often the product of information gathered at cardiac catheterization, can be quite helpful in understanding the physiology of complex lesions. First and foremost, though, particularly in the case of an emergency procedure, it is important to question the childs parents on his or her current and recent state of health. How is the childs exercise tolerance when compared to siblings or peers? Has he or she become markedly less active lately? Has cyanosis increased? Have there been any recent URI symptoms . . . even though outcome data have suggested that URIs may not increase mortality, or have a significant effect on perioperative morbidity, children with congenital heart disease may not tolerate the increase in pulmonary vascular resistance or airway resistance associated with intercurrent respiratory disease. Determine what medications the child is taking, and when the last doses were taken.

Physical examination, in addition to evaluating the childs presenting complaints, should pay particular attention to the thorax. Increased work of breathing, wheezing, or any change in the cardiac examination may warrant further investigation prior to induction. A 12-lead EKG can reveal dysrhythmias or, at the very least, document the childs rhythm. Echocardiography can help give an up-to-date picture of cardiac function, intracardiac shunting, valve function, etc. Laboratory evaluation can be directed toward the patients particular situation: it may be prudent to check electrolytes for a patient on diuretic therapy, to check the hematocrit of a patient with cyanotic disease, etc.
Emergency surgeries present their own problems. The child who has not fasted appropriately or who has an obstructive bowel process presents an increased risk for aspiration. There may not be time to get old records, let alone involve a cardiologist in consultation to assist with preoperative optimization of management. Above all, a child’s clinical status will dictate how long he or she can wait for surgery: the child with (ruptured) appendicitis will tolerate an hour or two for consults better than a trauma patient with a head injury, for example.

The American Heart Association has published guidelines for SBE prophylaxis, and many patients with heart disease carry a copy of these guidelines with them. Briefly, this patient, who has no allergies and who fits AHA guidelines as a moderate risk patient undergoing a procedure on the gastrointestinal system, would need 50mg/kg ampicillin IM or IV within 30 minutes before the procedure. The triple antibiotic therapy he has received preoperatively is sufficient prophylaxis.

Questions 4-5: Anesthetic Plan

The goals for the management of mitral stenosis are, at their simplest, to avoid increases in heart rate while preserving systemic vascular resistance. Fluids should be tightly controlled to avoid volume overload, acute LV failure, and pulmonary edema. Mitral stenosis impedes diastolic filling of the left ventricle; the gradient across the valve can increase pressure in the left atrium, which can translate into increased pulmonary vascular resistance when the LA pressure is chronically greater than 25 mmHg. The left atrium often dilates in response to the increase in pressure, which leads to atrial fibrillation and an increased risk for intra-atrial thrombus formation.

Mitral regurgitation leads to a decrease in effective left ventricular cardiac output because of the volume lost through the mitral valve during systole. Acquired mitral regurgitation is often found with mitral stenosis as a result of rheumatic heart disease; acute mitral regurgitation can be the result of papillary muscle dysfunction following myocardial infarction, or chordal rupture with bacterial endocarditis. The goals of management of the patient with mitral regurgitation include maintenance of normal sinus rhythm with a low-normal rate and maintenance of systemic vascular resistance. Intravascular volume should be maintained to ensure adequate venous return and optimal forward LV output.

Down syndrome is associated with a large tongue, small mouth, and a short neck. These patients can suffer from atlantoaxial instability as well. They are not usually difficult to intubate, but it is perhaps safest to minimize manipulation of the head and neck during laryngoscopy. Some advocate using antisylologues in these patients, but a history of atrial tachyarrhythmias might be a relative contraindication.

The most important concerns for the perioperative management of appendicitis are fluid management and attention to the septic process. Sepsis can sap the reserves of an otherwise healthy child: appropriate fluid resuscitation and administration of antibiotics (to cover both skin and gut pathogens) are crucial. Rapid sequence induction is often indicated because of concomitant vomiting and concern for decreased gut motility.
Questions 6-7: Medicolegal and social issues

Consultants should tread with great caution when suggesting a course of action which lies within the purview of another specialty. Within pediatric anesthesia, cardiologists sometimes raise the specter of cardiac anesthesiawithout explaining what is meant by that title. Someone who practices solely pediatric cardiac anesthesia? Someone with fellowship training in cardiac anesthesia? Do pancuronium and high-dose narcotics constitute a cardiac anesthetic? It is unclear that making such a notation in the chart has medicolegal ramifications, but certainly, a practitioner with no specific training in cardiac anesthesia may feel uncomfortable caring for such a patient when this line is found in the medical record.

In most jurisdictions in the United States, parents cannot give consent for their offspring once the child has reached the age of majority (18 years). Such surrogate consent may serve to make medical practitioners feel better in the case of an emergency procedure, but its legal standing is doubtful. Where applicable, parents who are the caretakers of their adult offspring who are, for whatever reason, unable to give consent should be counseled to become guardians of these patients, and should have medical power of attorney for their children.

Questions 8-14: Recognizing and treating a post-induction arrest

Once the anesthesia provider recognizes the apparent arrest, he or she should call for help and for the crash cart/defibrillator. Tube placement should be verified: in this case, it was noted quickly that the ETCO₂ sampling line had become dislodged from the capnometer. Auscultation of bilateral breath sounds, detection of end-tidal CO₂, and direct visualization of the endotracheal tube passing through the cords all serve to confirm tube placement. This patient has intravenous access in place.

Hypotension after induction has myriad causes, including in this case possible measurement error, hypovolemia, and possibly anesthetic overdose. It may be difficult to obtain an adequate non-invasive blood pressure from an obese patient: an appropriately-sized cuff must be used, and even pediatric institutions should have cuffs for large adults. Hypovolemia in this patient is probably a combination of his abdominal disease process and zealous diuresis by the cardiology service. Despite the most careful of inductions, the combination of a decrease in SVR and the decreased venous return under positive-pressure ventilation can unmask hypovolemia in the patient with poor reserves.

The PALS algorithm for a pulseless arrest incorporates the treatment of asystole, pulseless electrical activity, ventricular fibrillation, and unstable ventricular tachycardia. VF and unstable VT are treated first with defibrillation, 2 J/kg then 4 J/kg; ACLS recommends 200J/300J/360J for adults. After defibrillating and initiating chest compressions, epinephrine 10 mcg/kg given, followed by another maximal (4 J/kg to 360J) shock. Lidocaine 1 mg/kg or amiodarone 5 mg/kg can also be given. PALS allows for escalation to 100 mcg/kg of epinephrine for subsequent doses, while ACLS no longer recommends single doses of greater than 1 mg. PALS has not incorporated vasopressin into its algorithm. Asystole and PEA are treated with chest compressions and doses of epinephrine 10 mcg/kg at 3-5 minute intervals.

This apparent arrest after induction allows for a rather extensive differential to be developed: is the problem related to the patients cardiac disease, to sepsis from his appendicitis, to his fluid status, to his medications . . . to the method of induction . . . to equipment failure or malfunction. The actual exact etiology of the arrest is probably a combination of factors, the foremost of which are perhaps hypovolemia (with several manifestations) and the confounding factor of a gas sample line dislodged during the intubation.
REFERENCES

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SELECTIVE REFERENCES

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