A Child with Glenn Cardiac Physiology and Hypoxic-ischemic Brain Injury Presenting for Major Hip Surgery

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Objectives:

- Review the stages of and indications for single-ventricle palliation of complex congenital heart disease
- Review pertinent features in pre-operative assessment of children with palliated congenital heart disease and its complications
- Plan intra-operative care of a complex heart disease patient presenting for major orthopedic surgery
- Discuss strategies for managing pain following major hip surgery
- Weigh risks of regional anesthesia techniques in children receiving anti-platelet therapy

Case History:

A three year-old, 15 kg female with palliated complex congenital heart disease presents for bilateral hip osteotomies and adductor release. She was born with a double-inlet left ventricle, which was palliated with pulmonary banding at 3 weeks of age. She had post-operative cardiac arrest necessitating the use of ECMO, resulting in hypoxic-ischemic brain injury, which led to cerebral palsy, epilepsy, and severe developmental delay. She underwent bidirectional Glenn shunt at 8 months of age.

Questions:

What is the anatomy of double-inlet left ventricle? How does this anatomy affect cardiac function? What are the implications of this condition? How is it managed surgically? What is a bidirectional Glenn shunt? How does the surgical palliation process proceed?

Case History and Physical Examination (continued):

Following the PA banding at 3 weeks of age, the patient had post-operative cardiac arrest necessitating the use of ECMO, resulting in hypoxic-ischemic brain injury, which ultimately led to cerebral palsy, epilepsy, and severe developmental delay. Her medications include levetiracetam, diazepam, lansoprazole, clonidine patch, gabapentin, baclofen, olanzapine, aspirin, enalapril, and oxycodone. She has no allergies.

On examination, the patient is resting comfortably without distress. Cardiac exam is unremarkable, with no murmurs. Lungs are clear to auscultation. G-tube is present
on abdominal exam. The patient is generally hypotonic but moves all extremities spontaneously with contractures evident in lower extremities. She does not make eye contact or verbalize. She has a history of very difficult IV access and comes from home today without IV access present. Vital signs include a room air saturation of 87%, heart rate of 114/minute, respiratory rate of 20/minute, and blood pressure 112/56 mmHg.

Questions:

Do the medications raise any concerns? What other questions would you like answered before taking this patient to the OR for anesthesia? What is your induction plan? What monitors do you require for this case? Are there any studies that you would like to see before proceeding?

Preoperative Studies:

Routine echocardiogram was performed 2 months prior to the orthopedic surgery. It showed a univentricular heart with double inlet connection (Holmes Heart) with patent Glenn shunt, multiple aortopulmonary collateral vessels, low normal to mildly depressed ventricular systolic function, mild left AV valve regurgitation, patent aortic arch, and ventricular hypertrophy.

Metabolic panel performed one week prior was all within normal limits.

CBC performed one week prior showed WBC 6.5, Hgb 15.7, Hct 45.1%, Plt 283K

Blood type is A, Rhesus D positive with negative antibody screen

Questions:

What is your plan for IV access? What if you are unable to obtain peripheral IV? Would you consider central line placement? How would you go about this in a patient with a bidirectional Glenn? How does the site of surgery impact your decision? Will you place an invasive arterial line? What about neuraxial anesthesia for post-operative pain control? Which aspects of the history are relevant with regard to this? What if the surgeon or CICU team plans to administer heparin (either unfractionated or low molecular weight) post-operatively?

Case Progression:

The patient undergoes inhalational induction using oxygen and sevoflurane in the operating room with pulse oximeter, non-invasive blood pressure, five-lead ECG, and end-tidal CO2 monitor. A 24 g PIV is placed in the left foot after one attempt. Vecuronium is administered and intubation proceeds uneventfully. A 22 g PIV is placed in left hand. A right radial arterial line is placed under ultrasound guidance due to difficulty in placement. As the nurse places a Foley catheter, the wrist is hyper-extended against the nurse’s hip, and the arterial line waveform is lost. It is no longer possible to aspirate blood from it. The surgeon is pacing around the
operating room. A left ulnar arterial line is then placed, again with ultrasound guidance.

Questions:

What are your concerns for intraoperative management of a palliated single-ventricle patient? What considerations must you make when setting ventilator parameters? How do PEEP and respiratory rate affect the Glenn shunt? What are your acid-base goals, and how can this affect flow through the shunt? How is management of the Glenn patient different from the Fontan patient? How will you administer intraoperative analgesia? Do you plan to extubate the patient at the end of the case? Do you think the patient should recover in the PACU and then the floor, or should she be directly transported to CICU for recovery and care?

Intra-operative Course:

Given that the patient is on aspirin at moderate dose (and must remain so for patency of the Glenn shunt), the patient is unable to communicate, and has poor lower extremity function at baseline, the risks of epidural analgesia were deemed to outweigh possible benefits. The patient remains hemodynamically stable throughout the case. She is maintained on desflurane after induction. Analgesia is accomplished with 100 mcg fentanyl (6 mcg/kg), 4 mg morphine (0.26 mg/kg), and 230 mg IV acetaminophen (15 mg/kg). Total fluid balance is: crystalloid: 750 ml, 5% Albumin: 160 ml, blood loss: 250 ml and urine output: 135 ml. Her final ABG reports hematocrit of 29%. She is kept paralyzed throughout the case, and at case end is reversed with neostigmine and glycopyrrolate. She is extubated awake and transported uneventfully to the CICU where a safe-handoff is performed.

Questions:

What is the requirement for anticoagulation in the patient on a single-ventricle palliation pathway? What is the practice in your institution? What is the evidence base?

Post-operative Care:

The CICU manages the patient given her complex cardiac history. She later transfers to the inpatient cardiac floor, before eventual discharge. While in hospital the acute pain service manages her pain with nurse/parent-controlled analgesia morphine, diazepam as needed for muscle spasm, and scheduled IV acetaminophen. Methocarbamol is added when muscle spasms are identified as a source of pain. When gastrostomy-tube feedings resume, oxycodone is given per G-tube, and the PCA discontinued. Her home baclofen dose is also resumed.
Discussion:

This case raises issues of providing anesthesia for patients with palliated congenital heart disease for non-cardiac surgery, pain management for children with cerebral palsy, and regional anesthesia with concurrent anti-thrombotic medication.

This patient was born with a double-inlet left ventricle. As a biventricular circulation cannot be fashioned for children with DILV, they have to be placed on a staged pathway to a univentricular circulation with passive perfusion of the lungs. For this child, as a first step, a pulmonary band was placed to prevent overcirculation to the lungs. This procedure, or alternately, a modified BT shunt or Norwood procedure, is performed in the neonatal period. After allowing the child and her vasculature to grow, at about 4-8 months of age, a bidirectional Glenn shunt is performed. The Glenn shunt procedure, also known as superior cavopulmonary anastomosis, separates the SVC from the right atrium and connects it to the right pulmonary artery. The main pulmonary artery is usually disconnected from the single ventricle. Venous blood is thus rerouted from the upper body to the lungs, bypassing the heart. This reduces the volume load on the single ventricle and the degree of cyanosis due to mixing. The final step of the palliation is to complete the Fontan circulation by delivering venous blood from the lower body to the lungs by connecting the IVC to the PA. In this final stage, all systemic deoxygenated blood returns directly to the lungs and oxygenated blood in the single ventricle is pumped out to the body.

Neonatal cardiac surgery carries a substantial risk of neurological insult. This patient suffered a devastating brain injury, which led to severe hypotonia and cerebral palsy. Contractures resulting from such insults often require orthopedic intervention as the child ages. Taking a child with complex cardiac disease to the operating room for non-cardiac surgery presents many challenges to the anesthesiologist. These include access, monitoring and pain management. Neuraxial anesthesia may be precluded by the concomitant use of anti-platelet agents, which is common in children undergoing single ventricle palliation. Our patient was on an aspirin regimen. The current ASRA guidelines state: "NSAIDs seem to represent no added significant risk for the development of spinal hematoma in patients having epidural or spinal anesthesia. NSAIDs (including aspirin) do not create a level of risk that will interfere with the performance of neuraxial blocks." The patient in this case presented additional concerns, particularly her already compromised lower extremity function and the inability to report sensory or motor changes indicative of epidural hematoma. Thus, the anesthesia, cardiac, and orthopedic teams agreed to forego epidural analgesia.

To optimize the delivery of care to this complex patient, many teams communicated prior to all decisions and interventions. The anesthesia team, acute pain service, cardiologist, and orthopedic surgeon worked together to ensure that all her needs were met and no considerations were overlooked. She was successfully managed pre-, intra-, and post-operatively, and was ultimately discharged home without incident.
References:


