

**PBLD Table #: 38**

**Pediatric Jehovah's Witness Patient with Shone's Complex and Mitral Stenosis and Pulmonary Hypertension Presenting for Posterior Spinal Fusion**

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**Objectives:**

1. Review the management and discuss the anesthetic implications of pulmonary hypertension in the setting of Shone's complex with mitral stenosis.
2. Discuss the ethical issues pertaining to the perioperative care of a pediatric Jehovah's Witness patient
3. Review techniques for blood conservation in scoliosis surgery
4. Review modalities for monitoring the cardiac patient for non-cardiac surgery

**Case history:**

A 15 year-old male Jehovah's Witness with an unspecified genetic syndrome and Shone's complex consisting of parachute mitral valve, bicuspid aortic valve, and aortic coarctation presented for staged T2-L4 posterior spinal fusion. He has severe thoracic scoliosis with resulting severe restrictive lung disease.

**Questions:**

What is Shone's complex and how is it managed? What are the anesthetic implications of left heart obstructive lesions and their long-term consequences? How does severe scoliosis affect the evaluation and work-up of this patient?

**Case history and physical examination (continued):**

The patient has dysmorphic facial features, bilateral sensorineural hearing loss and visual disturbance. He also has asthma, left diaphragm paresis and underwent tracheostomy secondary to subglottic stenosis, anterior tracheal vascular compression, epiglottic and tongue base collapse. He requires BiPAP at night. Additional surgeries include end-to-end repair of coarctation and mitral valve repair. He takes pulmicort and sparingly uses albuterol for his asthma. Physical exam reveals a slight 15 year old male with significant thoracic scoliosis and a capped trach in place. He has a blowing systolic murmur at the apex, ejection murmur at the LUSB, and diastolic murmur at the base. Lung exam reveals increased aeration on the left without wheezing. Vital signs are within normal limits including four-extremity BPs.

**Questions:**

Why does this patient have all of these murmurs? What further work-up would you require for his cardiac disease? Is an echocardiogram and ekg sufficient?

Are PFTs required? What other consultant services should evaluate this patient before clearing him for surgery?

What issues should you discuss regarding the patient and his parents religious beliefs? Is this patient old enough to have a private discussion regarding blood transfusion without his parents? Should you consult the hospital ethics committee? Do you have the option to not do this case?

**Preoperative studies:**

Echocardiogram showed normal biventricular function with no evidence of subaortic membrane or ventricular shunt. Cardiac catheterization showed moderate mitral stenosis (7 mmHg) and moderate pulmonary hypertension at rest (6 Woods units) that was responsive to inhaled nitric oxide and supplemental oxygen. He also has mild aortic stenosis (peak gradient 9 mmHg) and ½ systemic RVSP. A fluid challenge of 7.5 ml/kg resulted in a significant elevation of pulmonary pressures, mitral valve gradient, and LVEDP. Pulmonary function tests showed severe restrictive defect with FVC 28% and FEV1/FVC 50% predicted. Polysomnography showed severe OSA and hypoxia (92%) and hypercarbia (pCO<sub>2</sub> 55 mmHg) with trach capped.

**Questions:**

What is the significance of the fluid challenge? Should this patient be cleared for scoliosis repair or should he have a mitral valve replacement?

Why does he have pulmonary hypertension? Should the pulmonary hypertension be treated and optimized prior to surgery? How would you treat it?

Are you concerned about the sleep study? Does an airway exam need to be performed?

What preoperative labs would you require prior to surgery? Should any other medications be initiated?

**Case progression:**

The patient and his family were counseled about the severity of this scoliosis and the need to proceed with surgery prior to any additional cardiac surgeries. They understood the risk of the procedure including the potential for significant blood loss, however the patient and his family were firm in that no blood transfusion was allowed according to their religious beliefs.

**Questions:**

What is your anesthetic plan for this patient? How do you manage the tracheostomy in the prone position? What types of venous and arterial access are you planning? What other monitors do you want to use? Given the patient's extreme sensitivity to a fluid bolus, how do you plan on monitoring volume status? The surgeon is only planning to use SSEPs for neuromonitoring. What medications do you plan to use? What blood conservation techniques could be employed for this patient? The patient will allow the use of albumin and cell salvage (as long as there is continuous patient connection). Does patient positioning and type of table matter?

**Intraoperative care:**

Induction of anesthesia was uneventful via tracheostomy tube and large bore IV access was easily obtained. During placement of the arterial catheter the heart rate jumps from a base line 84 to 126 with an the NIBP of 59/36. How do you respond? What medications would you consider using for this patient?

You finish preparing the patient for surgery and successfully position the patient in the prone position without any hemodynamic instability. The surgery proceeds without complication although you notice ~250cc of blood as accumulated in the suction canister. What is your allowable blood loss for this patient? What is the lowest Hgb/Hct you would tolerate in this patient? Does the fact that the patient is a Jehovah's Witness influence that number?

As the surgeon is placing the last screw, he asks you whether it is wise to proceed with the anterior portion of the case? How do you respond? What factors influence your recommendations?

**Postoperative Care:**

The patient is awakened in the operating room and transported to the intensive care unit. The patient has a BP of 78/50 on arrival to the ICU, and the accepting fellow asks the nurse to initiate a 500 cc bolus of albumin. How do you respond? What are the challenges of fluid management in the setting of mitral stenosis? The attending arrives and asks why nitric oxide was not used during the case. What considerations would you have prior to initiating nitric oxide on this patient?

**Discussion:**

Shone's complex presents variable multilevel left heart obstruction consisting of a supralvalvar mitral ring, parachute deformity of the mitral valve, subvalvar aortic stenosis, and coarctation of the aorta. The severity of obstruction at each lesion may ultimately preclude a two-ventricle repair. Outcomes for patients following a two-ventricle pathway depend on the degree to which the mitral stenosis can be relieved.

Mitral stenosis involves mechanical obstruction to diastolic filling of the left ventricle. Anesthetic management of mitral stenosis involves sufficient preload to maintain left atrial pressure. While cardiac output is typically maintained with normal heart rates even in the setting of moderate disease, increases in heart rate can significantly compromise left ventricular filling and stroke volume. Pulmonary edema and right heart failure are significant risks in patients with severe mitral stenosis. Adequate pain control to prevent increases in heart rate and pulmonary vascular resistance is essential. Likewise careful titration of fluids is imperative to prevent sudden increases in left atrial pressure and the possible development of pulmonary edema.

Intraoperatively, standard ASA monitoring should be used in all cases, however additional monitors should be determined by the nature of the surgical procedure and the patient's clinical condition. Placement of arterial access is important in the case described above, though some providers may argue that perioperative monitoring with a Swan-Ganz catheter or echocardiography is also essential in the presence of pulmonary hypertension. While there is literature describing the use of Swan-Ganz catheters and echocardiography in the prone position, additional risk and potential for misinterpreted data can also occur with these monitors. A noninvasive monitor such as near infrared spectroscopy (NIRS) allows assessment of the adequacy of tissue perfusion and oxygen delivery. If the hemoglobin, SaO<sub>2</sub> and oxygen extraction remain relatively constant, NIRS can be used to follow changes in flow. A low reading can be an early sign of cardiovascular compromise.

A major risk of scoliosis surgery is blood loss and the need for blood product transfusion. The patient described above presenting for scoliosis repair was a Jehovah's Witness and did not want a transfusion according to religious beliefs. There are a number of blood conservation techniques available to limit transfusions for scoliosis surgery including hypotensive anesthesia, acute normovolemic hemodilution, cell salvage, antifibrinolytic therapy, and patient positioning. The risk of not transfusing blood as well as the use of albumin and cell salvage should be discussed in depth with a Jehovah's Witness patient prior to major surgery. The case of a minor Jehovah's Witness patient is particularly challenging since the decision-making capacity of a minor matures over time. It is relatively common practice in pediatrics to obtain court orders and transfuse children whose parents are Jehovah's Witnesses when blood is considered absolutely necessary to preserve life. Ultimately, the patient should be allowed to express their wishes free of coercion.

In summary, the case described above presents many challenges for the anesthesiologist. Scoliosis surgery presents the potential for major blood loss, which is significant for a patient with mitral stenosis sensitive to fluid boluses. Adding complexity to the case is the fact that the patient refuses blood product transfusion based on religious beliefs as a Jehovah's Witness. Careful perioperative planning and coordination by the anesthesiologist can create a relatively safe outcome.

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