Congenital Heart Disease – Part 1 and Part 2

Ilene Claudius and Ashley Stroebel

- The **most common** surgery to repair a congenital heart defect is **ventricular septal defect** (VSD) repair.
- The **most complex** surgery to repair a congenital heart defect is **transposition of the great arteries**.
  - The modern approach to this condition is an arterial switch procedure where the aorta and pulmonary artery are switched.
  - This surgery is less commonly associated with atrial dysrhythmias than past procedures.
- Children with one functional ventricle (eg, hypoplastic left heart syndrome)
  - Either ventricle may be involved.

- Procedures aim to task the one ventricle with the important part of the circulation (ie, systemic) and provide passive flow back to the pulmonary circulation.
- Patients typically have staged surgeries and more than 70% of these children now survive to adulthood.

- Staged surgeries for children with a single ventricle
  - **Stage 1**: Norwood Procedure (happens at birth or shortly thereafter)
    - The main goal is to allow mixing to occur and to still use the single ventricle to do systemic and pulmonary circulation.
    - The first part of the surgery is the creation of the Blalock-Taussig or Sano shunt. These replace the patent ductus arteriosus.
      - The Blalock-Taussig shunt connects the subclavian artery and the pulmonary artery. The sano shunt connects the RV to the pulmonary artery. The surgeon will pick one of these based on their preference.
      - They will also have an atrial septostomy to create an atrial septal defect (ASD), if they don't already have one, as well as a neo-aorta when you want that systemic flow to be more robust.
  - **"Interstage period"**: After the Norwood, the patients will be discharged home because they need to grow before the next procedure. This period between surgeries is a scary time because the babies have a very tenuous physiology. They have about a 10% risk of death during this period.
    - Often, these children live far away from the center where they had their surgery and so half of the deaths happen at the local community hospital where they live.
    - Most commonly, patients present with something like a URI which may lead to hypoxemia, bronchiolitis with mucus plugging and V-Q mismatch, or acute gastroenteritis with dehydration. Their physiology makes them particularly vulnerable.
      - **These patients are completely preload dependent so the intrathoracic pressure is critical for blood flow.**
    - Elevated intrathoracic pressure prevents blood return to the heart and their ability to pump systemically. In addition, their lungs aren't getting blood flow so they aren't getting oxygenated blood. Babies lack preload and get hypoxic, which worsens the scenario.
    - These patients should be admitted for at least 24 hours of observation, even with a viral URI.
    - **Normal O_2 saturation for these patients in the interstage period will be 75-80%.**
      - If they are below that, you have to ask why? Is it hypoxemia from the viral URI or could they have shunting?

        - Ask parents what the child's baseline O_2 sat is at home and then give them a trial of nasal cannula oxygen with the baseline O_2 saturation as your target.
        - Examine perfusion including blood pressure.
        - Order CXR, labs +/- BNP. Do a POCUS evaluating IVC, B-lines, pericardial effusion, and cardiac function.
    - In terms of fluid, if they seem dehydrated or hypovolemic, you can give IV fluid in small aliquots (5-10 mL/kg) and check serial capillary refills and ultrasound reassessments.
    - The first line pressor, if needed, would be norepinephrine or epinephrine.
- After initial assessment, if their perfusion is still low:
    - There is a formula called the QP index (pulmonary blood flow) and the systemic blood flow is called the QS. There should be a 1:1 ratio between these.
    - To calculate this quickly, use 25 for pulmonary blood flow and use 100 minus the O_2 saturation for your systemic blood flow.
        - If you can target 25:25 as the 1:1, that's a good place to consider targeting..
        - If the systemic blood flow is less than 25, you need some SVR support
        - If you feel you've gotten to a maximum on fluids, a vasopressor would be indicated.
- Vasopressor options
    - Norepinephrine, epinephrine, or phenylephrine can be used.
    - Milrinone may also be requested by a consultant but is more of an inotrope and can vasodilate.
- These shunts can thrombose and this can occur in the setting of dehydration.
    - The Blalock-Taussig shunt and Sano shunt have machinery-like murmurs and you want to listen for that murmur. If you don't hear the murmur, consider thrombosis. Patients can rapidly decompensate if this occurs.
    - Start heparin if thrombosis is a strong possibility.
- **Stage 2**: Bidirectional Glenn procedure (occurs after 4-6 months)
    - This is a partial caval-pulmonary anastomosis where the SVC is connected to the pulmonary artery. This is the first time that the single ventricle is unloaded and part of the blood flow is allowed to go passively to the lungs.
    - The Blalock-Taussig or Sano shunt is also removed at this stage.
    - The O_2 sat rises to the low 90's.
    - This stage is associated with fewer complications and is a "holding period".
    - Hemodynamic status is much less tenuous.

- **Stage 3**: the Fontan procedure (occurs around 18-36 months).
  - The IVC is connected to the pulmonary artery. Now the SVC and the IVC are both connected to the pulmonary artery with a space in between that almost acts like a right atrium.
  - There may be a fenestration created here to allow for some regulation of pulmonary pressures so that the pulmonary system does not get overloaded.
  - At this point, these patients are relatively stable but they are at risk of dysrhythmias because of the structural changes that have been made.
    - **The most common dysrhythmias are atrial: atrial flutter, atrial fibrillation, and SVT.**
    - Treatment is basically the same as in other patients. Adenosine is the first line and procainamide would be a good next choice. Cardioversion is indicated for unstable patients.
  - Another complication is thrombosis so most patients are on aspirin or an antiplatelet/anticoagulant agent.
    - It is important to ask because this is an age common for falls and trauma.
- Adults who have survived these surgeries into adulthood are also at risk for dysrhythmia.
  - Treatment is the same as for any other adult.

Related Segments:
EM:RAP HD Congenital Heart Disease:
https://www.emrap.org/episode/pedsem-enduring/congenitalheart

CorePendium chapter - Pediatric Cardiac Disorders:
https://www.emrap.org/corependium/chapter/rec9l5sGLCse76gzR/Pediatric-Cardiac-Disorders