Congenital Heart Defects

Wendi S Redfern MSN, RN, ACNS-BC
Unit Based APN PICU
Objectives

- To have an introductory understanding of congenital heart defects
- To verbalize the clinical indices of inadequate cardiac output in pediatrics.
What is the heart’s job?

- The circulatory system’s job is to deliver oxygen and nutrients to all the body’s cells and to remove waste products of cellular metabolism!
Anatomy of the Heart

Made up of three layers

❤ Pericardium- 3 functions
  ➢ Prevents displacement of the heart
  ➢ Physical barrier that protects
  ➢ Contains pain and mechanoreceptors

❤ Myocardium- cardiac muscle

❤ Endocardium- Internal lining
The Normal Heart
Cardiac Performance

What is Cardiac Performance?

Cardiac Output!!!
What is Cardiac Output?

HR \times SV = CO
Heart Rate

In young children HR is fast and their SV small.

Infants are HR dependent for their CO.
When is HR too fast?

Tachycardia helps to maintain CO during times of distress but there is a marginal limit to HR improving CO.

Infants HR > 220
Children HR > 180

Result in compromise in ventricular diastolic filling time & coronary artery perfusion time.
Heart Rate too Slow

Bradycardia, transient may be normal but significant or sustained can be life threatening.
Stroke Volume

Averages 1.5 ml/kg

Stroke Volume is the product of

- Preload
- Afterload
- Contractility
Ventricular Preload

What is it?

It is the amount of myocardial stretch before contraction.
The greater the stretch on of the myocardial fibers the greater the contraction.

- With more volume in the ventricle there is more stretch on the muscle fibers which causes more tension which results in increase force for contraction
Factors Increasing Preload

- Hypervolemia
- Decreased intrathoracic pressure
- Systemic vasoconstriction
- Trendelenberg positioning
Factors Decreasing Preload

- Hypovolemia
- Systemic vasodilatation
- Increase intrathoracic pressure
- Increase intraperitoneal pressure
- Asynchronous atrial contractions
Atrial Septal Defect (ASD)

- [http://www.pted.org/?id=atrialseptal1](http://www.pted.org/?id=atrialseptal1)
- A hole in the atrial septum, or muscle wall
  - that separates the right and left atria
  - allows oxygenated blood from the lungs shunt left to right
  - This blood proceeds into the right ventricle, which pumps it back to the lungs rather than to the body.

- vary in size and in the severity of symptoms
- 5 and 10 per cent
- 2x more prevalent among girls than boys
- [http://www.pted.org/?id=atrialseptal3](http://www.pted.org/?id=atrialseptal3)
Ventricular Septal Defect (VSD)

- Hole in the ventricular septum
  - the muscular wall that separates the right and left ventricles

- Allows "shunting," of blood between the ventricles
  - Most commonly, "left to right shunt."
    - oxygenated blood from the left ventricle enters the right ventricle because there is greater pressure in the left ventricle and the resistance in the lungs is significantly lower than in the body tissues.
Most common form of congenital heart disease
- 21% of all cases
- single or multiple
- may occur in different parts of the ventricular septum

Small holes usually close spontaneously in the first year or two of life.
Large holes almost always require surgical closure in the first year of life.
VSDs may be present with other heart defects.
Affects boys and girls with equal frequency
VSD

- Left to right shunt
  - Blood that just returned from the lungs crosses the VSD and goes back to the lungs again. This causes increased pulmonary (lung) blood flow.
  - A heart murmur occurs because there is a pressure difference between the two ventricles and there is turbulent blood flow crossing the hole. The smaller the hole, the louder the murmur.

- Children are affected differently depending on the size, location, and number of holes in the ventricular septum. Small holes generally cause little or no difficulty and often close naturally as the child grows.
VSD

- Larger holes
  - child's feeding and growth
  - rapid breathing
  - Irritability
  - excessive sweating
  - poor weight gain

- The vessels which carry blood from the heart to the lungs and back again may become congested, or overloaded, with blood, resulting in congestive heart failure (CHF). This usually occurs when the child is 6 to 8 weeks old.

- With large VSDs, the lungs are receiving increased blood under higher than normal pressure. This can result in pulmonary hypertension (high blood pressure in the pulmonary artery). If this pressure becomes too high, the heart may be unable to function properly.

http://www.pted.org/?id=ventricularseptal2
Coarctation of the Aorta

- Blockage in the aorta
  - This may consist of a narrowing of the vessel or a shelf-like obstruction

- Located on the descending aorta near the heart
  - Usually immediately past the point (further from the heart) where the subclavian artery (SA in the diagram) exits the aorta on its way to the upper body.

- In some cases, the aortic valve (AV), through which blood enters the aorta from the left ventricle, is abnormally formed in this defect, with only two valve leaflets rather than the usual three (Bicuspid Aortic Valve).

- [http://www.pted.org/?id=coarctation1](http://www.pted.org/?id=coarctation1)
Coarctation of the Aorta

- The obstruction causes high blood pressure in the left ventricle
- Blood pressure in the upper body becomes high
- Blood pressure in the lower body is low because of the reduced blood flow through the aorta.
- If the obstruction in the aorta is severe, infants will develop severe heart failure after the patent ductus arteriosus (PDA) closes in the first several days after birth. If the problem is not diagnosed promptly, the infant may die.
Tetralogy of Fallot

- 10% of the cases of congenital heart disease.
- most common cyanotic heart defect
- involves four (Greek tetra = four) anomalies of the structure of the heart:
  - 1) A large ventricular septal defect (VSD)
  - 2) A narrowing (stenosis) of the outflow tract (infundibular stenosis) from the right ventricle into the pulmonary artery and/or pulmonary valve narrowing
  - 3) The aorta is enlarged and "overrides," or sits directly above, the ventricular septal defect (VSD).
  - 4) A thickening of the muscle wall of the right ventricle resulting in a right ventricular hypertrophy (thickening).
- A right sided aortic arch is present in 1/4 to 1/3 of patients.

http://www.pted.org/?id=tetralogyfallot1
Tetralogy of Fallot

- affects boys and girls with equal regularity
- In some infants and children, it can act more like a simple VSD. In these patients the main problem is too much blood flow.
- In some infants and children, there can be profound narrowing of the right ventricular outflow tract. Because of the severe narrowing, it is easier for the blood to cross the VSD right-to-left and go out the aorta instead of going to the lungs. If this is the case, the infant or child can become quite blue (cyanotic).
- In fact, the most severe form of Tetralogy of Fallot is with pulmonary atresia where no blood can cross from the right ventricle to the pulmonary arteries and lungs. In these infants it is necessary to begin a medication (Prostaglandin E1) to help keep open the Ductus Arteriosus (a vessel connecting the aorta to the pulmonary artery that usually closes soon after birth - see PDA) to maintain some bloodflow to the lungs.
- Babies with Tetralogy of Fallot may experience intermittent spells of extreme cyanosis, termed "tetralogy spells," or hypercyanotic spells. These can be serious and even life-threatening.
- [http://www.pted.org/?id=tetralogyfallot2](http://www.pted.org/?id=tetralogyfallot2)
HLHS

- Term used to describe spectrum of malformations with varying degrees of underdevelopment of left-sided heart structures
  - AA/MA, AA/MS, AA/unbal AVC, AS/MA, AS/MS, DILV w/ L-TGA, TA w/ D-TGA
HLHS

- Fourth most common cardiac defect presenting in infancy (7.5% of newborns with CHD)
  - 25% of cardiac deaths during first week
  - 15% of cardiac deaths during first month
HLHS - Pathophysiology

- Left ventricle is nonfunctional
- Pulmonary venous through RA via ASD, PFO or TAPVC
- Right ventricle supplies both pulmonary and systemic circulations in parallel
- Systemic flow through PDA
- Ductal closure leads to progressive acidosis, ischemia and death
HLHS - Pathophysiology

- Qp:Qs ratio represents balance of pulmonary and systemic circulation
- As PVR drops in newborn period, increase in Qp, thereby inc. volume load on RV to maintain adequate Qs
  - most common cause of CHF in 1st month
  - leads to high output failure, ischemia, death
Stage 1 Palliation of HLHS – Surgical Goals

- Complete relief of systemic outflow obstruction
  - Surgical arch reconstruction
  - Stenting of patent ductus arteriosus
- Maintain coronary blood flow without obstruction
- Prevent pulmonary venous hypertension through creation of nonrestrictive ASD
- Restrict excessive pulmonary blood flow with creation of systemic to pulmonary artery shunt
  - Modified Blalock-Taussig (BT) shunt
  - Right ventricle to pulmonary (RV-PA) conduit
  - Branch pulmonary artery banding
Different strategies – common issues

- Inefficient parallel circulation, and the need for precise partitioning
- Dependence on a functionally inferior systemic right ventricle
- Optimal energy utilization
Tissue oxygen delivery in critical illness

- Goal (regardless of mechanism of shock)
  - Delivery of oxygen adequate to meet the metabolic demands of tissues
  - Delivery of oxygen in excess of requirements - Reserve

- Optimize oxygen delivery to tissues
  - Cardiac output
  - Reduced sympathetic output
Shock is best defined as:

- 1. Uncontrolled blood or fluid loss
- 2. Blood pressure less than the 5th percentile
- 3. Altered mental status and low urine output
- 4. Reading about the salaries of professional athletes
- 5. None of the above
Definition of Shock

- Inadequate tissue perfusion to meet tissue demands
- Usually the result of inadequate blood flow and/or oxygen delivery
- **Shock is NOT a blood pressure diagnosis**
- Ultimately, if uncorrected, this state progresses to irreversible cell damage, collapse of homeostatic mechanisms, and death
Characteristics of Shock

- End organ dysfunction:
  - reduced urine output
  - altered mental status
  - poor peripheral perfusion
- Metabolic dysfunction:
  - acidosis
  - altered metabolic demands
Cardiogenic Shock

- Hypoplastic Left Heart Syndrome
- After surgical repair of CHD
- Myocardial impairment
- Cardiac dysrhythmias
## Septic Shock

### warm vs. cold

<table>
<thead>
<tr>
<th><strong>WARM</strong></th>
<th><strong>COLD</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>- fever</td>
<td>- fever/hypothermia</td>
</tr>
<tr>
<td>- tachycardia</td>
<td>- tachycardia</td>
</tr>
<tr>
<td>- bounding pulses</td>
<td>- poor pulses</td>
</tr>
<tr>
<td>- altered mental status</td>
<td>- altered mental status</td>
</tr>
<tr>
<td>- wide PP</td>
<td>- narrowed PP</td>
</tr>
<tr>
<td>- decreased perfusion</td>
<td>- decreased perfusion</td>
</tr>
<tr>
<td>- decreased UO</td>
<td>- decreased UO</td>
</tr>
<tr>
<td>- =capillary refill</td>
<td>- decreased capillary refill</td>
</tr>
<tr>
<td>- +/- mottled extremities</td>
<td>- mottled extremities</td>
</tr>
</tbody>
</table>
Common perfusion indices

- Blood pressure
  - $\text{BP} \sim \text{CO} \times \text{SVR}$

- Clinical exam (pulse amplitude, CRT, ext temperature)
  - Influenced by autoregulation/temperature control
  - Limited in its ability to accurately assess cardiac output

- Organ function (urine output, mental status)
  - Multifactorial, non-quantitative and late

- Metabolic acidosis (lactate, base deficit)
  - Lactate: Oxygen delivery dependent oxygen consumption already in effect. Late
    - $\uparrow$ Lactate predict need for ECMO – Charpie, JTCVS 2000
  - Base Deficit: Other causes
Shock
Perfusion Indices

- Mental Status: Level of Consciousness
  - is the child...
    - restless
    - anxious
    - agitated
    - lethargic
    - listless
    - comatose
    - able to make eye contact & recognize parent
Shock
Perfusion Indices

- Skin
  - rash
  - turgor
  - color
  - temperature
  - mottling
  - moistness
Shock
Perfusion Indices

- Nail beds: color and cap refill
- Mucous membranes: color and moistness
- Chills
- Fever
- Fontanel
Shock
Perfusion Indices

- Work of Breathing
  - rate and depth
  - chest wall dysfunction
  - inspect airway
  - restrictive problems
Shock
Perfusion Indices

- Cardiovascular
  - pulse: rate, quality, rhythm
  - peripheral veins: collapsed, distended
  - blood pressure
  - pulse pressure
Shock

Perfusion Indices

- Urine output
- weight loss
- dysuria
- metabolic acidosis
- CVA tenderness
Shock
Perfusion Indices

- Abdominal
  - distention
  - rebound
  - tenderness
  - bleeding
Shock Treatment

- Restore & Maintain Hemodynamic Stability:
  - volume resuscitation
  - vasopressors &/or inotopes

- Support oxygenation & oxygen delivery:
  - supplemental oxygen
  - mechanical ventilation

- Identify Source/Etiology
  - antimicrobials
Shock Treatment

- CORRECT METABOLIC ABNORMALITIES/DIC:
  - acidosis, hypoglycemia, hypocalcemia, anemia

- ANTICIPATE & PREVENT COMPLICATIONS:
  - stress ulcers, barotrauma, MOSF

- SURGICAL INTERVENTION:
  - congenital heart disease