Congenital Heart Disease

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Objectives

The learner will:

- Identify 3 areas in the developing heart that result in congenital heart defects.
- Discuss 3 important components of a cardiovascular assessment.
- List the 3 cardinal signs of congenital heart disease.
- Describe the clinical symptoms in each category.
- Identify 3 defects that are included in each category.
A “congenital” anomaly originating in the developing fetus is often considerably modified, at least physiologically, by the dramatic circulatory adjustments at birth. Weeks, months, or even years may then elapse before the anomaly evolves into the “typical” clinical picture. Both physiologic and structural changes subsequently continue, or conversely, the malformation may “vanish.” Joseph Perloff (1978)

**Definition:**
- Congenital (Latin) Con = together, Genital = Born
- Actually occurs seven months before birth
- Most defects are compatible with intrauterine life until a term birth
Incidence of Congenital Heart Disease

- 8-10 per 1000 live births (approx. 1 %)
  - (Does not include preterm infants with PDAs or people with slight abnormalities of aortic valve)
- Increases to approx 3% with subsequent siblings
- Risk increases with left sided lesions – some quote 15%.
- Also increases in offspring of parents with CHD
- Remains one of the greatest causes on neonatal mortality & morbidity
- Leading expense for neonatal health care
Etiology of Congenital Heart Disease: Multifactorial

- **Maternal:**
  - Disease: Diabetes, Collagen disease (Lupus), Seizure disorders
  - Medications: anticonvulsants, diazepam, progesterone/estrogen, alcohol, street drugs (cocaine), Retin-A, lithium, thalidamide, lithium, warfarin, aspirin, ACE inhibitors

- **Genetic:**
  - Chromosome abnormalities: 13, 18, 21;
  - Turner’s Syndrome,
  - DiGeorge Syndrome (22q11 deletion),
  - Williams Syndrome

- **Environmental:**
  - Viral infections (Rubella, CMV), ???
  - Toxins
Embryologic Timeline

- **Week 3**: Cardiogenic plate, endocardial tubes
- **Week 4**: Fusion of endocardial tubes to single median tube, first contraction at day 22, cardiogenic looping
- **Week 5**: Beginning of true circulation, septation of primum atrium, AV valves to 3 chamber heart
- **Week 6**: Septum secundundum, septation of bulbus and ventricle, divided truncus arteriosus
- **Week 7**: Four chambered heart, absorption of pulmonary veins
Cardiac Looping
Blood Flow Through Partitioning Heart

A: Truncus arteriosus
   Sinoatrial valve
   Sinus venosus
   Primordial atrium
   Atrioventricular canal
   Bulbus cordis
   Primordial ventricle

B: Primordial atrium
   Ventral
   Dorsal
   Endocardial cushions
   Primordial right ventricle

C: Plane of section D
   Right atrium
   Sinoatrial valve guarding orifice of sinus venosus
   Foramen primum
   Left atrioventricular canal
   Fused AV endocardial cushions
   Arrow passing through right atrioventricular canal

D: Septum primum
   Sinoatrial valve guarding orifice of sinus venosus
   Foramen primum
   Left atrioventricular canal
   Fused endocardial cushions
   Primordial interventricular septum

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Partitioning of Truncus Arteriosus
Development of the Ventricular Septum
Development of Aortic Arch
Fetal Circulation
Newborn Physiology
Closure of fetal shunts

- **Ductus arteriosus**
  - Constrictures by 10 to 14 days. Anatomic closure and formation of ligamentum arteriosus by 12th week
  - May remain open much longer with prematurity or hypoxia.

- **Foramen ovalae**
  - Functional closure at birth, anatomic closure has been seen by 3 months, 50% by 5 years, 25% of adults still have PFO.

- **Ductus venosus**
  - Constriction of vessel within hours of birth as portal pressure falls with removal of placenta
Normal Heart Pressures and Saturations
Cardiac Output:
Heart Rate X Stroke Volume

Normal Stroke Volume
- Newborn = 400 mL/kg/min
- Infant = 200 mL/kg/min
- Child = 100 mL/kg/min

Increased oxygen demand (consumption) increase cardiac output: Heart rate and/or stroke volume
Heart Rate:
The number of contractions/minute

Principles:

- Increased heart rate improves Cardiac Output until diastolic filling time and coronary perfusion are compromised

- Decreased heart rate necessitates an increase in stroke volume to maintain Cardiac Output

- Neonates are heart rate dependent for Cardiac Output
Stroke Volume:
The volume of blood ejected by the ventricle with each contraction

Factors affecting Stroke Volume:
- Preload
- Afterload
- Contractility
Cardiac Output:
Amount of blood pumped from the ventricle/minute

- MUST meet the metabolic demands of the body.
- Normalized to BSA (Body Surface Area)
- The cardiovascular assessment is your measurement of how well your patient is meeting the metabolic demands.
Assessment

- Systematic
- Consistent
- Head to toe
- Assess other systems with CV system
- Always start with inspection and note general appearance

SICK or WELL
Cardiovascular Assessment

- End-organ perfusion
- Skin color and temperature
- Pulses (peripheral & central)
- Capillary refill
- Heart rate/rhythm
- Heart sounds
- Blood pressure
Inspection

- Head/neck
  - Color
    - Cyanosis - central vs peripheral
    - Pallor - hemoglobin
  - Work of breathing
    - Nasal flaring, retractions
- Neck veins
Palpation

Peripheral pulses
Rate/rhythm
Quality
Equality
Location
Carotid
Central – Brachial, femoral
Peripheral – Radial, dorsalis pedis, posterior tibial
Palpation

- **Anterior chest**
  - Thrills
  - Precordium – Point of Maximum Impulse

- **Skin**
  - Peripheral perfusion
  - Temperature – one of best ways to assess CO
  - Moisture

- **Abdomen**
  - Liver
Conduction System

- Superior vena cava
- Aorta
- Sinoatrial node (S.A. node or pacemaker)
- Right atrium
- Atrioventricular node (A.V. node)
- Right ventricle
- Left atrium
- Left ventricle
- Inferior vena cava
- Purkinje fibers
- Right and left branches of atrioventricular bundle (bundle of His)
Rhythm Interpretation

❤Sinus Arrhythmia
Auscultation

- Key is to listen to one sound at a time!
- Listen in 2 positions.
- Listen in systematic pattern.
Heart Sounds

- $S_1$ – beginning of systole, AV valves close
- $S_2$ – end of systole, closure of semilunar valves
- $S_3$ – normal in children, related to rapid filling of ventricle
- $S_4$ – abnormal, heard late in diastole or early systole, heard in CHF, decreased ventricular compliance
Murmurs

Definition: “Whooshing” sounds that occur in various stages of diastole and/or systole due to turbulent blood flow.

Causes:

- Normal heart murmur
- Increased rate of blood flow
- Blood flow forced forward through an incompetent/constricted/irregular valve
- Blood flows back (regurgitant) through an incompetent valve, a septal defect, or a patent ductus arteriosus
Characteristics of Murmurs

- Timing
- Frequency or pitch
- Location
- Intensity (Graded I – VI)
- Radiation
- Quality
- Effect of respiration
- Effect of position
Significance of Murmurs

- A systolic murmur may be normal. A diastolic murmur is never normal.
- The intensity of the murmur does NOT indicate the severity of heart disease.
- You may not hear a murmur in serious CHD.
- You may hear a Grade 4-5 murmur in insignificant CHD.
Normal Heart Murmurs

- Newborn – Peripheral pulmonary stenosis
- Infant/child – Pulmonary ejection murmur
- Child – Still’s murmur, vibratory murmur
- Teenager – Venous hum
Blood Pressure

- Cuff size – EXTREMELY important
- Extremity – use right arm
- Normal Range
- LAST vital sign that will indicate that the child is in trouble
### Normal Dinamap Blood Pressure Ranges

**Normal Dinamap BP Values**  
[Systolic/Diastolic (Mean Arterial Pressure)]

<table>
<thead>
<tr>
<th>AGE-GROUP</th>
<th>MEAN</th>
<th>RANGE</th>
<th>90&lt;sup&gt;TH&lt;/sup&gt; PERCENTILE</th>
<th>95&lt;sup&gt;TH&lt;/sup&gt; PERCENTILE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Newly born</td>
<td>65/41</td>
<td>50-70/25-45</td>
<td>75/49 (59)</td>
<td>78/52 (62)</td>
</tr>
<tr>
<td>(Term Birth)</td>
<td>(50)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Neonate</td>
<td>84/</td>
<td>60-90/20-60</td>
<td>100/</td>
<td>104/</td>
</tr>
<tr>
<td>(To 28 days)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Infant</td>
<td>94/52</td>
<td>87-105/53-66</td>
<td>105/68 (83)</td>
<td>110/70 (86)</td>
</tr>
<tr>
<td>(1 – 12 mos)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Toddler</td>
<td>94/56</td>
<td>90-105/53-66</td>
<td>108/68 (82)</td>
<td>114/70 (85)</td>
</tr>
<tr>
<td>(2 – 5 yrs)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>School Age</td>
<td>98/62</td>
<td>97-112/57-71</td>
<td>114/74</td>
<td>118/78</td>
</tr>
<tr>
<td>(6 – 10 yrs)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Adolescent</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>(11 – 13 yrs)</td>
<td>106/66</td>
<td>112-128/66-80</td>
<td>120/78 (B)130/82</td>
<td>124/82 (B)136/86</td>
</tr>
<tr>
<td>(14 – 18 yrs)</td>
<td>(B)116/68 (G)108/66</td>
<td>66-80 (G)122/80</td>
<td></td>
<td>(B)126/82 (G)126/82</td>
</tr>
</tbody>
</table>
Cardiac Reserve

- Adaptive mechanisms activated to increase cardiac output.

  Mechanical factors
  - Hypertrophy
  - Dilation

  Biochemical factors
  - Increase energy production

  Activation of the adrenergic nervous system
Cardinal Signs of Heart Disease

- Cyanosis
- Decreased Systemic Perfusion
- Tachypnea
Cyanosis

### Clinical Presentation
- Central rather than peripheral.
- May not be immediately present.
- Not evident until a significant amount of reduced hemoglobin is present.
- Source of systemic arterial blood flow.
- Tachypnea without distress

#### Congenital Defects
- Transposition of the Great Arteries
- Tetralogy of Fallot
- Tricuspid Atresia
- Pulmonary Atresia
Consequences of Cyanosis

- Polycythemia
- Clubbing
- Hypoxic spells and squatting
- Central nervous system complications
- Bleeding disorders
Key Questions

- How does blood get to the lungs?
- How does blood get to the body?
- Is the pulmonary/systemic blood flow dependent on the ductus arteriosus?
- If the answer to any of these questions is yes, immediate intervention is mandatory.
  - Prostaglandin E1 (PGE)
  - Atrial septostomy
  - Arterial to pulmonary shunt
  - Palliative/reconstructive surgery
Prostaglandin E1 (PGE)

Indications for use:
- Right heart outflow obstruction
- Transposition of the great arteries
- Left heart outflow obstruction

Contraindications for use:
- Total anomalous pulmonary vascular return (TAPVR)
- Respiratory distress
- Left-to-right shunts with increase pulmonary blood flow
Prostaglandin E1

- Maintains patency of the ductus arteriosus in ductal dependent congenital heart defects to provide either pulmonary or systemic circulation.

- Side effects:
  - Cardiovascular: Vasodilation, arrhythmias, hypotension, edema
  - Neurological: Seizures, hyperthermia, irritability, lethargy
  - Respiratory: Apnea, hypoventilation, bronchospasm, tachypnea
  - Renal: anuria
  - Hematologic: Thrombocytopenia, hemorrhage, DIC
Modified Blalock-Taussig Shunt

- Bidirectional Glenn shunt
- Right pulmonary artery
- Super vena cava
- Waterston shunt
- Main pulmonary artery
- Subclavian artery
- Modified Blalock-Taussig shunt
- Potts shunt
Nursing Considerations:

- Intracardiac Mixing of Systemic and Pulmonary Circulation
- Assessment of Cardiac Output:
  - Shunt
    - Murmur present?
    - Hydration status?
    - Oxygen saturation?
Transposition of the Great Arteries

- 5% of all CHD
- Males > female by 3:1
- Must have communication between left and right sides of the heart
- Surgical repair before left ventricular muscle strength decreases
- Oxygen saturation may be greater in lower extremities
Arterial Switch Procedure
Coronary Artery Repair for TGA
Tetralogy of Fallot

- 10 % of all CHD
- Was most common cyanotic defect beyond infancy
- Cyanosis varies by degree of pulmonary stenosis
- 4 components:
  - Large VSD
  - Right ventricular outflow tract obstruction
  - Overriding aorta
  - Right ventricular hypertrophy
"Tet" Spells

Most frequently seen in unrepaired patients with Tetralogy of Fallot.

Symptoms:
- Rapid and deep respirations (hyperpnea)
- Increasing cyanosis
- Irritability and prolonged cry
- Decreased intensity or absence of heart murmur

Treatment:
- Pick up and hold infant
- Knee chest position
- Morphine sulfate: 0.1-0.2 mg/kg IM
- 100% oxygen
- Phenylephrine 0.02 mg/kg IV bolus followed by infusion
- Sodium bicarbonate: 1 mEq/kg IV
Tetralogy of Fallot with BT Shunt
Repair of Tetralogy of Fallot
Pulmonary Atresia

- With Intact Ventricular Septum  1% of all CHD
- With VSD 2% of all CHD
- Is ductal dependent
- Intervention necessary within first few hours after birth.
Tricuspid Atresia

- 1-2% of all CHD
- Male slightly more than female
- An associated defect is necessary for survival.
- Systemic saturation is directly related to the amount of pulmonary blood flow.
Tricuspid Atresia with BT Shunt
Tricuspid Atresia with Glenn Shunt
Tricuspid Atresia
Lateral Tunnel Fontan
Tricuspid Atresia
External Conduit Fontan
Decreased Systemic Perfusion

Clinical Presentation
- Poor feeding
- Pallor/grey
- Diaphoresis
- Tachypnea with respiratory distress
- Irritability

Congenital Defects
- Aortic Valve Stenosis
- Coarctation of the Aorta
- Hypoplastic Left Heart Syndrome
Nursing Considerations:

- Systemic Blood Flow?
- Intracardiac Mixing of Systemic and Pulmonary Circulations?
- Assessment of cardiac output
  - Color
  - Temperature
  - Perfusion
  - Blood Pressure
  - Heart Rate/rhythm
Aortic Valve Stenosis

- 5% of all CHD
- Males > female by 4:1
- Wide spectrum of disease.
- Babies with severe stenosis may be ductal dependent.
- Half of patients with Coarctation also have abnormality of aortic valve.
- Risk of reoccurrence increases by 15% or greater.
Coarctation of the Aorta

- 8 % of all CHD
- Male > female by 2:1
- 30 % of children with Turners Syndrome
- May not be evident until ductus arteriosus closes
- Can present with severe cardiogenic shock
Hypoplastic Left Heart Syndrome

- 2% of all CHD
- 10% of all CHD presenting within first month of life
- May not be identified until ductus arteriosus closes.
- Presents in shock.
- Treatment with oxygen will increase severity by decreasing pulmonary vascular resistance resulting in increase pulmonary blood flow and decreased systemic circulation.
Norwood Procedure for HLHS
Norwood Procedure/Sano
Glenn Shunt
HLHS with External Conduit Fontan
Tachypnea

Clinical Manifestations
- Tachypnea with mild respiratory distress
- Hepatomegaly
- Tachycardia
- Diaphoresis (secondary to sympathetic response to increased stress)
- Failure to thrive
  - Increased oxygen consumption
  - Increased work of breathing

Congenital Heart Defects
- Patent Ductus Arteriosus
- Atrial Septal Defect
- Ventricular Septal Defect
- Atrioventricular Septal Defect
- Total Anomalous Pulmonary Venous Return (TAPVR)
- Truncus Arteriosus
**Tachypnea:**

- Excessive Pulmonary Blood Flow

- Increased pulmonary blood flow results in pulmonary edema and congestive heart failure.

- Increasing inspired oxygen will decrease pulmonary resistance thereby increasing pulmonary blood flow. CHF symptoms will increase and may even have acute decompensation.

- Exception is TAPVR below diaphragm. This results in pulmonary venous congestion.
Nursing Considerations:

- Intracardiac Mixing of Systemic and Pulmonic Blood
- Supplemental Oxygen
- Assessment of Cardiac Output
- Failure to Thrive
  - Increased work of breathing
  - Increased heart rate
  - Decreased caloric intake
Patent Ductus Arteriosus

- 5-10 % of CHD except for preterm infant
- Female > male by 3:1
- Elective closure except in preterm infant
- Non-surgical closure with coils
Atrial Septal Defect

- 5-10% of all CHD
- 4th most common
- Females > males by 2:1
- Usually asymptomatic
- Primary types:
  - Secundum (site of fossa ovalis)
  - Sinus venosus (often associated with PAPVR)
  - Ostium primum (AVSD type defect)
Types of Atrial Septal Defects

Figure 12-1
Anatomic types of atrial septal defects (ASDs) viewed with the right atrial wall removed. IVC, inferior vena cava; SVC, superior vena cava.
Non surgical closure of ASD
Ventricular Septal Defect

- 20-25% of all CHD
- Most common defect
- Many will close spontaneously
- Intervention depends on location and symptoms
- Increasing use of cath closure devices
Figure 12-6 Anatomy of ventricular septum and ventricular septal defect (VSD). A, Ventricular septum viewed from the right ventricular (RV) side. The membranous septum is small. The large muscular septum has three components: the inlet septum (I), the trabecular septum (T), and the outlet (or infundibular) septum (O). B, Anatomic locations of various VSDs and landmarks, viewed with the RV free wall removed. a, outlet (infundibular) defect; b, papillary muscle of the conus; c, perimembranous defect; d, marginal muscular defect; e, central muscular defect; f, inlet defect; g, apical muscular defect. (From Graham TP Jr, Bender HW, Spach MS: Ventricular septal defect. In Adams FH, Emmanouilides GC, Riemenschneider TA (eds): Moss' Heart Disease in Infants, Children and Adolescents, 4th ed. Baltimore, Williams and Wilkins, 1989.)
Types of Ventricular Septal Defects

- **Membranous**
  - Also called perimembranous
  - Most common
  - Located below aortic valve
  - May require surgical repair

- **Muscular**
  - 70% close spontaneously

- **Outlet**
  - Located just below pulmonary valve
  - Risk of prolapse of aortic valve leaflet into defect
  - Requires surgical repair, important to protect integrity of aortic valve

- **Inlet**
  - Located below the leaflet of the tricuspid valve
  - Will need surgical repair
Atrioventricular Septal Defect (AVSD) (AV Canal Defect, Endocardial Cushion Defect)

- 2 - 5 % of all CHD
- 30-50% of children with Down’s Syndrome
- Usually surgically repaired by 4-6 months of age. Earlier if increasing CHF.
Total Anomalous Pulmonary Venous Return

- Less than 1% of all CHD
- No direct connection between PV and LA
- Must have an atrial communication for survival
- Types:
  - Supracardiac (50%)
    - Male = female
  - Infracardiac (20%)
    - Male > female by 4:1
  - Cardiac (20%)
    - Usually to coronary sinus
  - Mixed - Right veins involved twice as often as left
Truncus Arteriosus

- <1 % of all CHD
- Single trunk from both ventricles
- Must have a VSD
- Truncal valve may have 3 or 4 cusps
- Usually presents shortly after birth.
- Surgical repair when diagnosed.
Surgical Repair: Truncus Arteriosus
Ready for Discharge:

- Newborn Discharge Considerations
  - Hearing Test
  - Car Seat Angle Test
  - Newborn Metabolic Screen

- Medications

- Wound Healing

- Interstage monitoring
  - Oxygen Saturation
  - Weight Gain
  - Activity/fussiness
References


