Ventricular Septal Defect
What the Nurse Caring for a Patient with CHD Needs to Know

Courtney Petro, BSN, RN, CCRN
Registered Nurse, Cardiovascular ICU,
Lucile Packard Children’s Hospital at Stanford

Melanie Sojka, MSN, RN, CPNP-AC/PC
Pediatric Nurse Practitioner, Cardiac & Thoracic Surgery,
University of Chicago Medicine, Comer Children’s Hospital

Grace Macek, MSN, RN, PNP-BC
Pediatric Nurse Practitioner, Cardiac & Thoracic Surgery,
University of Chicago Medicine, Comer Children’s Hospital

Jennifer Newcombe, MSN, CNS, CPNP-AC/PC
Nurse Practitioner, Pediatric Cardiothoracic Surgery,
Loma Linda University Children’s Hospital

Dorothy M Beke, RN, MS, CPNP-PC/AC
CICU Clinical Nurse Specialist and Cardiology Clinic NP,
Boston Children’s Hospital

Embryology
- One of most common congenital heart defects (CHD)
- Intraventricular septum divides right (RV) & left (LV) ventricles
  - Consists of 3 separate septa
  - Beginning in 5th week embryonic development
  - Completely formed and closed by 7th-8th week embryonic development
- Septum results from:
  - Growth of muscular portion upward from ventricular floor towards endocardial cushions
  - Growth of subendocardial tissue from right side of endocardial cushion
    - Fuses with aorticopulmonary septum
    - Fuses with muscular portion
- Causes of ventricular septal defect (VSD)
  - Unclear
    - Multifactorial
    - Genetic/chromosomal syndromes (trisomy 13, 18, 21/ Holt-Orem, Cornelia de Lang)
  - Majority not associated with other defects or syndromes
  - More common in premature or low-birthweight infants

Anatomy
- Results when interventricular septum fails to close (See illustration below for locations and types of VSDs)
- Abnormal communication along septum between right & left ventricles
  - Disruption in fusion of 3 separate septa
  - Size and anatomical location
    - May be single or multiple defects
    - Varies with 4 major locations
      - Perimembranous (Membranous, infracristal, conoventricular malalignment including tetralogy of Fallot and double outlet defects) (Number 2 in above illustration)
        - Located in upper portion of septum
        - Most common - 70-80%
        - Frequently close in first year of life – 30-50%
        - Conoventricular defects do not spontaneously close
      - Outlet (Supracristal, conal, subpulmonary, subarterial) (Number 1 in above illustration)
        - Incomplete fusion along aortopulmonary septum with endocardial cushions & muscular portion
        - Located just beneath pulmonary valve
May involve prolapse of aortic valve leaflet
  - Results to damage in aortic valve
  - May result in aortic valve insufficiency
- Spontaneous closure uncommon

**Inlet or Canal VSDs (Number 6 in above illustration) (Number 6 in above illustration)**
- Lie beneath septal leaflet of TV
- May be referred to as an atrioventricular septal defect, but does not involve either atrioventricular valve
- Will not spontaneously close

**Muscular or trabecular VSDs (Numbers 3, 4, & 5 in above illustration)**
- Less common – 5-20%
- Completely surrounded by muscular tissue
- May appear as single defect on LV side and multiple defects on RV side due to trabeculations (criss-crossing fibrous and muscular tissue strands)
- May close spontaneously
- “Swiss cheese” septum
  - Multiple defects
  - Involve all septal regions

- Associated with other cardiac defects: Atrial Septal Defect (ASD), Patent Ductus Arteriosus (PDA), Coarctation of the Aorta (CoAo), subvalvar Aortic Stenosis (AS), or subpulmonic stenosis (PS)
- Multiple VSDs often present with: Tetralogy of Fallot (TOF), Double-Outlet Right Ventricle (DORV)
- May be acquired in older patients from post-surgical leak, trauma, or myocardial infarction

**Physiology**
- Abnormal blood flow across defect in ventricular septum
- Affected by:
  - Size of defect
    - Primary variable
    - Impacts shunt and need for repair
      - VSDs <25% of the aortic annulus diameter
        - Small
        - Minimal, if any, left-to-right shunting
        - Potential for spontaneous closure based upon location
      - VSDs 25%-75% of the aortic annulus diameter
        - Little to moderate left to right shunting
        - No pulmonary artery hypertension
        - May be mild to moderate pulmonary overcirculation
        - May have symptoms of congestive heart failure (CHF)
          - Can be managed with medications
- May improve as the patient grows and the defect starts to close
- VSDs >75% of the aortic annulus diameter
  - No restriction to flow
  - Moderate to large volume shunt
  - Pulmonary overcirculation with CHF symptoms
    - Increased pulmonary venous return
      - LA and LV dilation
      - LV hypertrophy
    - Increased RV volume
      - Increased pulmonary blood flow
      - Increased pulmonary pressures
      - May result in pulmonary artery hypertension (PAH) (See Peds/Neo Problem Guideline on Pulmonary Hypertension)
    - Long standing PAH may result in Eisenmenger’s Syndrome (See Adult Problem Guideline on Eisenmenger’s Syndrome)
  - Most likely requires closure
- Resistance to flow
  - Pulmonary vascular resistance (PVR)
    - Newborn
      - Pulmonary pressure ≥ systemic pressures
      - Rapidly fall with first few breaths after birth
      - Reaches normal adult pressures within first 2 months of age (Normal mean pressures ≤ 25 mmHg after first few weeks of life)
      - Decrease accelerated by:
        - Supplemental oxygen
        - Pulmonary vasoactive medications (iNO, sildenafil)
    - Decreased resistance increases flow across defect (left-to-right shunt)
      - Normal maturation of pulmonary vascular bed
        - Usually occurs by 2 months of age
        - RV pressure usually drops to ~ 1/3rd to 1/2 of LV pressure by ~ 2 weeks; however in the presence of a VSD, RV pressure may take longer to decrease.
      - Allows for development of pulmonary overcirculation
  - Systemic vascular resistance (SVR)
    - Newborn
      - Systemic pressures = pulmonary pressures
      - Left sided lesions increase systemic resistance
        - Coarctation of the aorta (CoA), aortic valve (AV) stenosis
        - Vasoconstriction
• Increased left-sided systolic pressure increases flow to RV (left-to-right shunt)

**Procedures/Interventions**

• **Indications for Intervention**:
  - Infants with greater than 2:1 shunt
    - Large sized VSDs and significant CHF
    - Medical treatment of CHF
      - Diuretic therapy
      - Digoxin/ angiotensin converting enzyme (ACE) inhibitors
      - Increase caloric intake
      - Goals: control symptoms and allow infant to grow
    - Surgical closure
      - Early closure (~ less than 3 months of age)
      - Unable to manage CHF and provide for somatic growth
  - Infants with shunts 1.5:1
    - Moderate sized VSD
    - May usually be followed for up to 5 years of age to maximize chance of spontaneous closure
  - Shunts less than 1.5:1
    - Small VSD
    - Require close follow-up
  - Outlet VSDs
    - May see prolapse of leaflet of AV and develop progressive aortic regurgitation (AR)
    - Repair before significant aortic regurgitation (AR) develops

• **Surgical intervention - complete repair via open heart surgery (OHS)**
  - Patch closure
    - Requires cardiopulmonary bypass & sternotomy
    - Majority of membranous & inlet VSDs closed through the transatrial approach
    - Repair thru tricuspid valve (TV)
      - May have obstructed visualization from valve leaflet
      - Requires retraction and sometimes detachment of valve leaflet and subsequent repair of TV
    - Some defects require a right ventriculotomy or pulmonary artery approach
    - Closed with patch material (Dacron or Polytetrafluoroethylene [PTFE])
  - Direct suture closure for very small defects
  - **Risks**
    - Injury to AV or conduction system
    - Residual VSD
    - Diminished RV function with ventriculotomy
  - **Intraoperative transesophageal echocardiography (TEE)**
    - Assess repair
    - Rule out residual VSD
- Assess competence of TV and AV
- Assess ventricular function

- **Surgical intervention** – palliative
  - Pulmonary Artery Band (PAB)
    - Requires sternotomy; cardiopulmonary bypass not required
    - Controls symptoms related to CHF
    - Allows infants to grow and reach appropriate size for surgical repair
  - Rarely indicated
    - Exceptions: infants < 2.5 kg with multiple/complex defects and/or intractable CHF

- **Combined Surgical/cardiac catheterization intervention** (Hybrid intervention)
  - **Indications**
    - Patients too small for percutaneous catheter system
    - Part of repair of complex lesion
    - Visualization of VSD difficult
  - **Periventricular closure**
    - Requires sternotomy but avoids cardiopulmonary bypass
    - Placement of percutaneous device directly through the right ventricular (RV) free wall

- **Cardiac catheterization intervention**
  - **Percutaneous closure**
  - Avoids sternotomy & cardiopulmonary bypass
  - Device is placed during cardiac catheterization
  - Muscular VSDs may be amenable to device closure

**Specific considerations**

- **Pre-operative considerations**
  - **Age**
    - Consider size and location of defect
    - Premature neonates
      - Consider size of defect relative to body size to determine timing of surgical correction
    - Neonates
      - Due to elevated PVR development of CHF rare in the first few weeks of life (See resistance to flow section)
      - Do not administer supplemental oxygen unless oxygen saturations persistently < 75%
    - Infants and children
      - May be managed medically
      - Require surgical correction
        - CHF unresponsive to medical management,
        - Growth failure
        - Outlet and inlet defects
  - **Health status**
    - Recent/frequent viral respiratory infections
      - Supportive management until cultures negative
• Surgery delayed until patients are symptom free to minimize post-bypass pulmonary complications
  – Failure to thrive
    – Consider nasogastric, high caloric feeding
    – Attempt to achieve positive caloric balance
• Postoperative (See Peds/Neo Problem Guidelines for Postoperative Management)
  – Most common open heart surgery for CHD
  – Ventilation
    – Patients beyond infancy may be extubated in OR
    – Neonates and infants may require ventilator support and aggressive diuresis before extubation
    – Patients with respiratory viral infections preoperatively may require extended intubation
      – Patients with pre-operative respiratory syncytial virus (RSV) should have negative cultures and be asymptomatic prior to surgery to minimize post-operative complications
    – Patients with pre-operative PAH will require pulmonary vasodilators (iNO, IV pulmonary vasodilators) (See Peds/Neo Problem Guidelines for Pulmonary Hypertension)
  – Inotropic support
    – Majority with minimal inotropic support
    – Repair of VSD with complex lesions or PAH
      – May also consider pulmonary vasodilators
  – Monitor for the following complications:
    – Arrhythmias (See Peds/Neo Problem Guidelines for Arrhythmia Management)
      – Most patients who have cardiopulmonary bypass surgery have temporary epicardial wires placed in OR
    – Complete Heart Block
      – Typically transient 24-48 hours
      – May be permanent and require placement of a permanent pacemaker, usually after 7-10 days
    – Supraventricular Tachycardia (SVT) or Junctional Ectopic Tachycardia (JET)
• Residual VSD
  – Common to have some residual leaks around patch which often eventually close with endothelialization
  – Assessment
    – Operating room
      – Analysis of RA and PA saturations
      – Echocardiography [Transesophageal (TEE) or Transthoracic (TTE)]
    – Intensive care unit
      – Desaturation
      – Increased pulmonary pressures
      – Decreased systemic pressures
VSD patch dehiscence with low cardiac output
- Pulmonary hypertensive crisis
- Patients with elevated PVR preoperatively or long-standing pulmonary over-circulation (See Ped/Neo Problem Guidelines for Pulmonary Hypertension)
  - Monitor PA pressures if PA line available
  - Follow pulmonary hypertension precautions
    - Avoid noxious stimulation
    - Fastidious pulmonary toilet (pre-medicate prior to suctioning)
    - Hyperventilation
    - Oxygenation
    - Strict acid / base control
    - Inhaled nitric oxide - potent pulmonary vasodilator
    - Sedation/paralysis

- Post-catheter device monitoring:
  - Bleeding at puncture site
  - Arrhythmias (Complete Heart Block)
    - Device may put pressure on the septum close to the left and right bundle branches
  - Valvar regurgitation
    - Risk of “trapping” the aortic, tricuspid, or mitral valve leaflets in the device
  - Device embolization

Long-Term Problems/Complications
- Structural Complications
  - Residual VSD
  - AI secondary to aortic cusp prolapse
  - Supravalvar pulmonic stenosis after prior placement of PAB
  - Subaortic membrane (rare)
  - Right ventricle muscle bundle hypertrophy (rare)
- Arrhythmias (See Peds/Neo Problem Guidelines for Arrhythmia Management)
  - Transient post-operative heart block
    - At risk of developing complete heart block
    - May require pacemaker placement
  - Ventricular arrhythmias
    - Monitor with periodic electrocardiograms (ECG), Holter monitor
    - Increased with ventriculotomy
- Heart Block requiring pacemaker placement (see Peds/Neo Problems Guidelines for Pacemakers)
  - Pacemaker interrogation every 3-6 months
  - Generator changes ~ every 6-10 years
  - Lead malfunction or fracture
  - Cardiovascular implantable electronic device (CIED) infection
Routine Cardiology Care for Surgical and Catheter Intervention

- Routine follow-up interval
  - Every 1-2 years by a cardiologist trained in Congenital Heart Disease (CHD) until 18 years of age
  - Adults
    - No residual VSD, no associated lesions and normal pulmonary pressure
      - Does not require continued follow-up at a regional ACHD center
    - Small residual VSD
      - Follow up visit every 3 to 5 years at an Adult CHD (ACHD) regional center
    - Device closure of a VSD
      - Follow up visit every 1 to 2 years at an ACHD center
      - Depends on the location of the VSD
- Cardiac studies as indicated by assessment/symptoms
  - Transient post-operative heart block, at least a yearly ECG
  - Echocardiogram
- Endocarditis prophylaxis recommendations (AHA, 2015)
  - Six months post-surgical repair/device placement
  - Longer if a residual defect is present

References:


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