Ventricular Septal Defect

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

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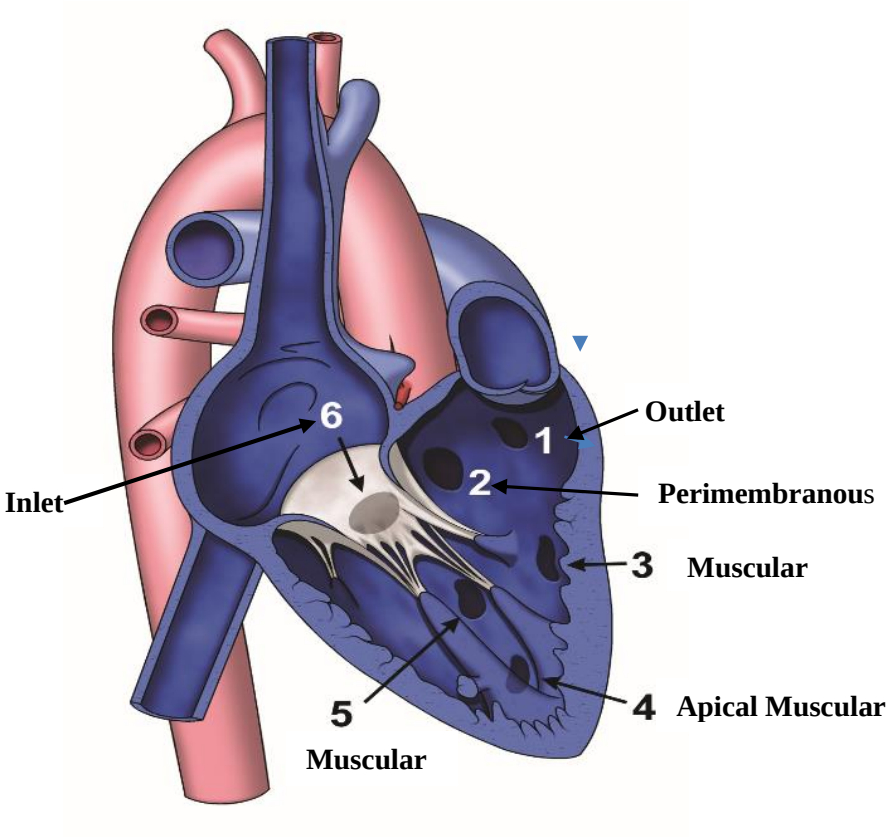
Embryology

- 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

- May occur in any part of the septum
- May occur in more than one location



Illustrations reprinted from PedHeart Resource. www.HeartPassport.com.

- 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 $\geq$ systemic pressures
            - Rapidly fall with first few breaths after birth
            - Reaches normal adult pressures within first 2 months of age (Normal mean pressures $\leq$ 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 ½ 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

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