Atrioventricular Septal Defects
(AV Canal Defect, Endocardial Cushion Defects)
What the Nurse Caring for a Patient with Congenital Heart Disease Needs to Know

Mary Rummell, MN, RN, CPNP, CNS, FAHA,
Clinical Nurse Specialist, Pediatric Cardiology, Cardiac Services,
Oregon Health & Science University (Retired)

Embryology
- Occurrence: 4 -- 5 % of all congenital heart defects
- Development of the atrioventricular (AV) canal starts in the fourth week of gestation
- Endocardial cushions (Moore, 2008)
  - Develop from specialized extracellular matrix (cardiac jelly)
  - Form on walls of AV canal and fuse together
  - Fuse with septum primum to form lower portion of atrial septum
  - Fuse with the bulbar ridges to form the membranous (upper) part of the ventricular septum
  - Along with tissue from walls of AV canal form atrioventricular valves (tricuspid and mitral valves)

Anatomy
- Abnormal development of the structures that are derived from the endocardial cushions.
  - Abnormal development of the atrioventricular septum (Marx, 2006)
    - Involves the primum atrial septum and the inlet ventricular septum
    - Septal defects vary in size
      - Worst case involves the entire atrial and ventricular septa
      - Most common is large atrial septal defect (ASD) with small ventricular septal defect (VSD)
      - Rarely see small ASD with large VSD
    - Size of defects depend on position of atrioventricular (AV) valves

Complete Atrioventricular Septal Defect
© Scientific Software Solutions, 2016. All rights reserved.
Abnormal development of the AV valves (Marx, 2006)
- Involve septal portions of the mitral and tricuspid valve
- Lower attachment on AV septum creates large primum ASD
- Higher attachment on AV septum results in larger VSD
- Common AV valve
  - No or abnormal septal attachments of the AV valves
  - Single anterior and single posterior leaflets bridging the septal orifice
    (Illustration below shows Anterior and Posterior Leaflets of the Common AV Valve)

Common Atrioventricular Valve

© Scientific Software Solutions, 2016. All rights reserved.

Variations in terms based upon anatomic deformities (Warnes, 2008; Park, 2014)
- May be called: complete, common, partial, and unbalanced defects
  - Complete AVSD
    - Most common in Trisomy 21 (Down syndrome) patients
      - Two thirds of patients with uncomplicated AVSD have Down syndrome
      - Fifty percent of patients with Down syndrome have AVSD
    - Single AV Valve orifice
  - Partial AVSD
    - Most common in non-Down syndrome patients
    - Two separate AV Valve orifices
    - Usually asymptomatic
  - Unbalanced AVSD
    - Single AV valve committed either to right or left of midline
    - Creates differential flow into the ventricles
    - Usually results in one ventricle being smaller than the other
    - May make a two ventricular repair impossible
AV valves may be referred to as “right” or “left” sided AV valve instead of tricuspid or mitral valves

“Atrioventricular septal defect” (AVSD) best descriptor of anomaly

- Posterior displacement of atrioventricular node
  - Results in changes in electrocardiogram (In approximately 50% of patients)
    - Prolonged PR interval
    - “Superior” QRS axis (left axis deviation)
  - Increases risk of surgically induced heart block

- Associated defects (Park, 2014)
  - Tetralogy of Fallot
  - Double outlet right ventricle (DORV)
  - Unbalanced ventricles (Single ventricle)
  - Additional VSD
  - Patent ductus arteriosus
  - Subaortic stenosis

**Physiology** (Marx, 2006; Park, 2014)

- **Left-to-right shunt**
  - Not usually present in neonatal period [unless early decrease in pulmonary vascular resistance (PVR), i.e. with administration of supplemental oxygen for oxygen saturation <90% but >76%]
  - Occurs with decrease in pulmonary vascular resistance (PVR)
    - Usually around 2-4 weeks of age
    - May be accelerated with administration of supplemental oxygen
    - Rapid decrease in PVR may cause pulmonary edema
  - Congestive heart failure
    - Similar to that seen with ASD and VSD
    - Tachycardia
    - Tachypnea
    - Failure to thrive

- **Hypoxemia**
  - Mixing of systemic venous and pulmonary venous blood
  - Can occur with shunts at both ventricular and atrial level

- **Pulmonary vascular disease**
  - Elevated pulmonary artery pressure from excessive pulmonary blood flow (pulmonary overcirculation)
  - Continually elevated pulmonary pressure leads to pulmonary vascular disease (See Problem Section on Pulmonary Hypertension)
  - Chronic pulmonary overcirculation
    - Leads to Eisenmengers syndrome (See Problem Section on Eisenmengers syndrome)
    - Seen in older, unrepaired patients
  - Increased incidence of pulmonary vascular disease may be related to genetic anomaly (Down syndrome/Trisomy 21)

- **Abnormal atrioventricular valve(s)**
  - Cleft in mitral valve
    - Common with ostium primum defect
    - Usually regurgitant leading to left atrial enlargement
  - Common AV valve
    - May be incompetent with regurgitant flow into atria
Incompetent valves may result in,
- Pulmonary edema
- Increased pulmonary infections/pneumonia

Overrides (straddles) ventricular septum
- Usually in midline position
- Abnormal position committed to either right or left ventricle
  - Results in disproportionate blood flow into ventricles
  - May result in hypoplasia of one ventricle

### Type of Repair
- All types of AVSD require surgical repair, none will close spontaneously
- Repair of partial AVSD
  - Closure of primum ASD
  - Reconstruction/repair of AV valves
  - Timing
    - Symptomatic
      - Rare
      - If present, usually from regurgitant MV
    - Asymptomatic
      - Elective repair between 2-4 years
      - All babies with Down’s syndrome should be evaluated for a cardiac defect
- Repair of complete AVSD (Backer, 2007; Park, 2014) [See Illustrations from Scientific Software Solutions below]
  - Patch repair with one or two patches
  - Repair usually completed in infancy (between 2-6 months)
    - Timing based on:
      - Symptoms
        - Congestive heart failure
        - Pulmonary hypertension
        - Failure to thrive
      - Anatomy
      - Associated defects/problems
        - Genetic disorder – especially Down syndrome (Trisomy 21)
        - Presence of elevated pulmonary pressure may prevent symptoms
        - Additional factors related to babies with Down syndrome that influence hemodynamics and timing of repair
          - Chronic nasopharyngeal obstruction
          - Relative hypoventilation
          - Carbon dioxide retention
          - Sleep apnea
          - Decreased muscle tone
      - Rarely requires reoperation for revision or repair of patch
Repair of atrioventricular valves
- Cleft mitral valve - least complicated to repair cleft
- Common AV valve
  - Complexity depends on anatomy of valve
  - Goal to have a competent, non-stenotic mitral valve
- Most common cause for reoperation - left AV valve regurgitation

Single Patch Repair of Complete AVSD
© Scientific Software Solutions, 2016. All rights reserved

Repair of Atrioventricular Valve

Bridging leaflets

Left AV Valve

Right AV Valve

Repair of Atrioventricular Valve
Patch placement in anterior and posterior bridging leaflets form right and left atrioventricular valves.


- Placement of band on pulmonary artery
  - Rarely done
  - Possible Indications
    - Preterm or low birth weight
    - Severe CHF unable to manage with medications
      - FTT or weight loss
      - Unable to provide adequate caloric intake
    - Complex lesions
      - Unbalanced AVSD
      - DORV
      - Tet-canal

Post-operative risk factors/special considerations
- Pulmonary hypertensive crisis (See Pulmonary Hypertension Problem Guideline)
- Immediate, severe left AV valve regurgitation
- Arrhythmias – surgically induced heart block (See Arrhythmia Problem Guideline)

Long Term Complications/Interventions (Refer to Problem Section for specific complications) (Cetta, 2009; Rodrigues, 2011)
- Regurgitant/Stenotic AV valves
  - Residual AV valve insufficiency
  - Major determinant of long term outcome
- Arrhythmias
- LV outflow track obstruction
- Sub aortic stenosis
- Pulmonary hypertension
- Greatest risk of mortality due to reoperations.

Routine Cardiology Care (Warnes, 2008)
- Lifelong follow up
  - Infant/child - Every 6 months -1 year
    - Standard repair without residual defects
    - Increase frequency with development of incompetent MV or arrhythmias
    - Neurodevelopmental assessment/management of developmental delays
  - Adult - Every 12-24 months
    - Cardiologist with experience in adult congenital heart disease
- Cardiac studies as indicated by assessment/symptoms
  - Serial electrocardiograms (EKG)
  - 24 hour ambulatory EKG monitor
  - Imaging
    - AV valve functioning
    - Evaluation of left ventricular outflow tract
    - As indicated by assessment and clinical problems
Consideration for pregnancy (Refer to Adult CHD Problem Section on Pregnancy for further discussion and management) (Warnes, 2008)

- Pre-conception cardiac evaluation
  - Assess for residual hemodynamic lesions
  - Counsel for pregnancy risk and preventive measures for women with Down’s syndrome
- Well tolerated in women who have been repaired and have no major residual defects
- Not advised in women with pulmonary hypertension, residual VSD, poor ventricular function
- Risk of fetal CHD (range 10-15%)

References:


Reviewed/revised
10/2015
M. Rummell