Atrioventricular Septal Defects
(AV Canal Defect, Endocardial Cushion Defects)

What the Nurse Caring for a Patient with Congenital Heart Disease Needs to Know

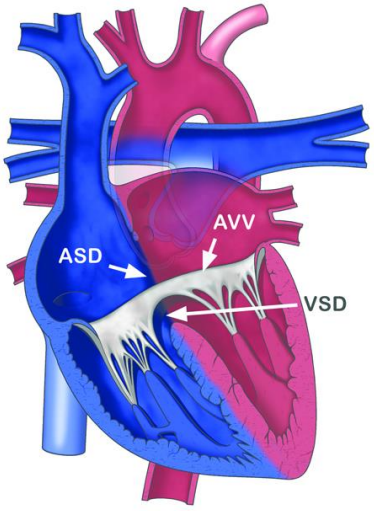
Mary Rummell, MN, RN, CPNP, CNS, FAHA,
Clinical Nurse Specialist, Pediatric Cardiology, Cardiac Services,
Oregon Health & Science University (Retired)

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)

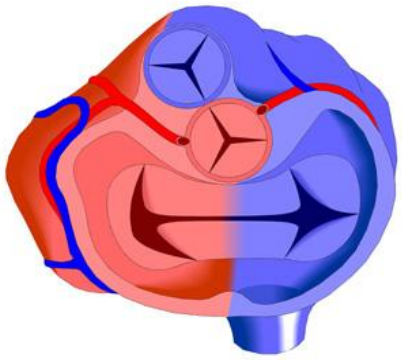
Anatomy

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

- 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

A cross section of the heart at the level of the valves illustrates the Common AV Valve in relation to the aortic (in red) and pulmonic (in blue) semi-lunar valves.

- 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

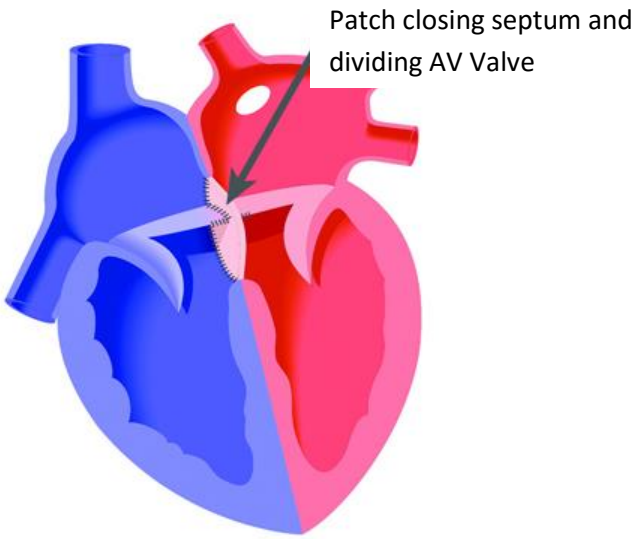
Physiology (Marx, 2006; Park, 2014)

- 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

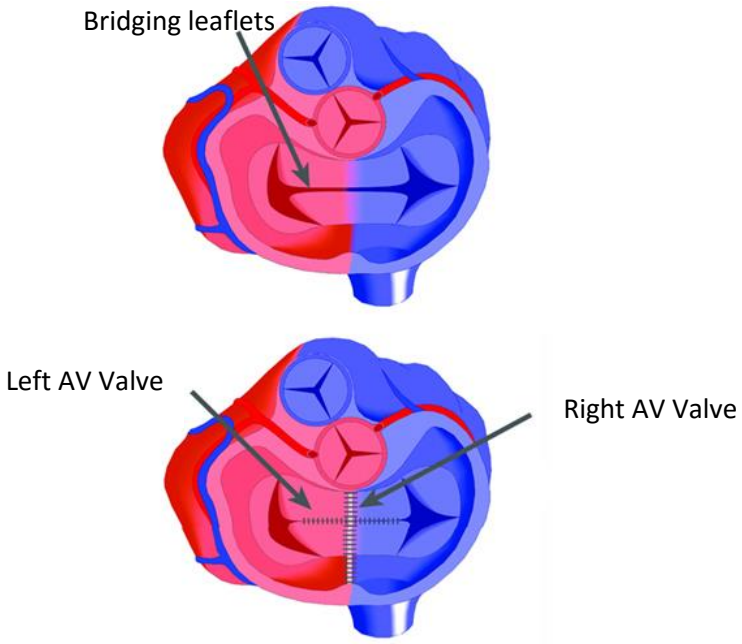
Type of Repair

- 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



Single Patch Repair of Complete AVSD
Illustrations reprinted from PedHeart Resource. www.HeartPassport.com.

- 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



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:

Backer, C. L., Stewart, R. D., & Mavroudis, C. (2007). Overview: history, anatomy, timing, and results of complete atrioventricular canal. *Seminars in Thoracic and Cardiovascular Surgery*, 10, 3-10.

Backer, C. L., Stewart, R. D., & Mavroudis, C. (2007) What is the best technique for repair of complete atrioventricular canal? *Seminars in Thoracic and Cardiovascular Surgery*, 19, 249-257.

Cetta, F. (2009) Atrioventricular septal defects. In C. A. Warnes (Ed), *Adult Congenital Heart Disease*, Oxford, GB: Wiley-Blackwell.

Curley, M. A. Q., & Moloney-Harmon, P. A. (2001) *Critical Care Nursing of Infants and Children*, (2nd ed.). Philadelphia, PA: W.B. Saunders Company.

Everett, A. D., & Lim, D. S. (2010) *Illustrated Field Guide to Congenital Heart Disease and Repair*, (3rd ed.). Charlottesville, VA: Scientific Software Solutions, Inc.

Feldt, R. H., Edwards, W. D., Porter, C.J., et. al. (2001) Atrioventricular septal defects. In H. D. Allen, E. B. Clark, H. P. Gutgesell, & D. J. Driscoll (Eds), *Moss and Adams' Heart Disease in Infants, Children' and Adolescents*, (6th ed.). Philadelphia, PA: Lippincott Williams & Wilkins.

Marx, G. R., Fyler, D. C. (2006) Endocardial cushion defects. In J. F. Keane, D. C. Fyler, J. E. Lock (Eds), *Nadas' Pediatric Cardiology*, (2nd ed.). Philadelphia, PA: WB Saunders/Elsevier.

Mavroudis, C., & Backer, C. L. (Eds.). (2003) *Pediatric Cardiac Surgery*, (3rd ed.). St. Louis, MO: Mosby.

Moore, K. L., & Persaud, T. V. N. (2008). *The Developing Human. Clinically Oriented Embryology*, (8th ed.). Philadelphia, PA: WB Saunders/Elsevier.

Park, M. K. (2008). Atrioventricular Septal Defects. *Pediatric Cardiology for Practitioners*, (5th ed.). Philadelphia, PA: Elsevier.

Park, M. K. (2014). Left-to-right-shunt lesions. *Park's Pediatric Cardiology for Practitioners*, (6th ed.). Philadelphia, PA: Elsevier.

Pettersen, M. D. (2013). Pediatric complete Atrioventricular septal Defects. *Pediatrics: Cardiac Disease and critical Care Medicine*. http://www.emedicine.medscape.com/article/893914. Accessed online 9/2015.

Pillutla, P., Shetty, K. D., & Foster, E. (2009). Mortality associated with adult congenital heart disease: Trends in the US population from 1979 to 2005. *American Heart Journal,* 158(5), 874-879.

Rodriguez, F. H., Moodie, D.S., Parekh, D. P., et al. (2011.) Outcomes of hospitalization in adults in the United States with atrial septal defect, ventricular septal defect, and atrioventricular septal defect. *The American Journal of Cardiology,* 108, 290-203.

Rummell, M. (2013). Atrioventricular septal defect (AVSD)/AV canal defect, Chapter 8 Cardiovascular disorders, Specific diseases. In M. F. Hazinski (Ed.), *Nursing Care of the Critically Ill Child,* (3rd ed.) (320-326). Philadelphia, PA: Elsevier.

Slota, M. C. (Ed). (2006) *Core Curriculum for Pediatric Critical Care Nursing*. *American Association of Critical Care Nurses,* (2nd ed.). Philadelphia, PA: WB Saunders.

Warnes, A., & Williams, R. G. co-chair. et.al. (2008) Atrioventricular Septal defect in ACC/AHA 2008 Guidelines for adult Cs with CHD. *Journal of the American college of Cardiology* 52(23):e18, Available at: http://content.onlinejacc.org/cgi/content/full/52/23/e143 Accessed 7/15/2011

Reviewed/revised
10/2015
M. Rummell