

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

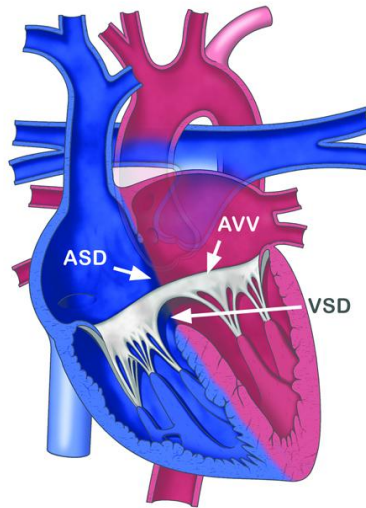
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I. Embryology

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

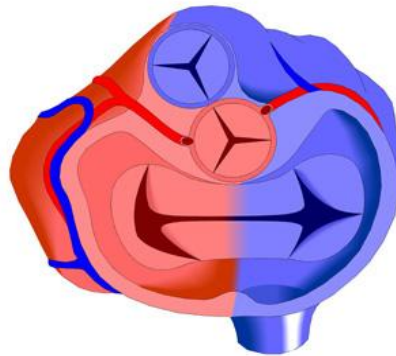
II. Anatomy

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



Complete Atrioventricular Septal Defect
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2. Abnormal development of the AV valves (Marx, 2006)
 - a. Involve septal portions of the mitral and tricuspid valve
 - b. Lower attachment on AV septum creates large Primum ASD
 - c. Higher attachment on AV septum results in larger VSD
 - d. Common AV valve
 - (1) No or abnormal septal attachments the AV valves
 - (2) 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.

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- B. Variations in terms based upon anatomic deformities (Warnes, 2008)
 1. May be called: complete, common, partial defects
 - a. Complete AVSD most common in Trisomy 21 (Down syndrome) patients
 - b. Partial AVSD most common in non-Down syndrome patients
 2. AV valves may be referred to as “right” or “left” sided AV valve instead of tricuspid or mitral valves
 3. “Atrioventricular septal defect” (AVSD) best descriptor of anomaly
- C. Posterior displacement of atrioventricular node
 1. Results in changes in electrocardiogram (In approximately 50% of patients)
 - a. Prolonged PR interval
 - b. “Superior” QRS axis (left axis deviation)
 2. Increases risk of surgically induced heart block
- D. Associated defects (Feldt, 2001)
 1. Tetralogy of Fallot
 2. Double outlet right ventricle
 3. Unbalanced ventricles
 4. Additional VSD
 5. Patent ductus arteriosus
 6. Subaortic stenosis

III. Physiology (Marx, 2006)

A. Left-to-right shunt

1. Similar to that seen with ASD and VSD
2. Increased shunt leads to pulmonary vascular disease (See Problem Section on Pulmonary Hypertension)
3. Increased incidence of pulmonary vascular disease may be related to genetic abnormality (Trisomy 21)

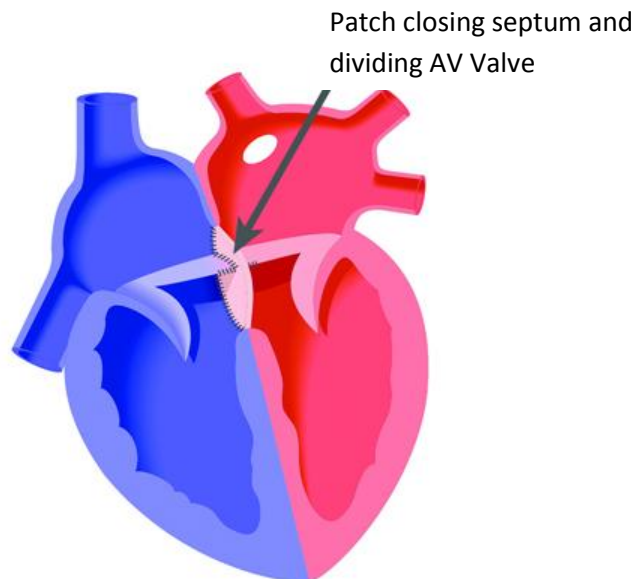
B. Abnormal atrioventricular valve(s)

1. Cleft in mitral valve
 - a. Common with ostium primum defect
 - b. Usually regurgitant leading to atrial enlargement
2. Common AV valve
 - a. May see cyanosis from mixing of pulmonary and systemic venous return
 - a. May be incompetent with regurgitant flow into atria
 - b. May 'override' ventricular septum
 - (1) Results in disproportionate blood flow into ventricles
 - (2) May result in hypoplasia of one ventricle

IV. Type of Repair

A. Repair of septal defects (Backer, 2007)

1. Patch repair with one or two patches
2. Repair usually completed in infancy
 - a. Timing based on:
 - (1) Symptoms
 - (2) Anatomy
 - (3) Associated defects/problems
 - b. Rarely requires reoperation for revision or repair of patch



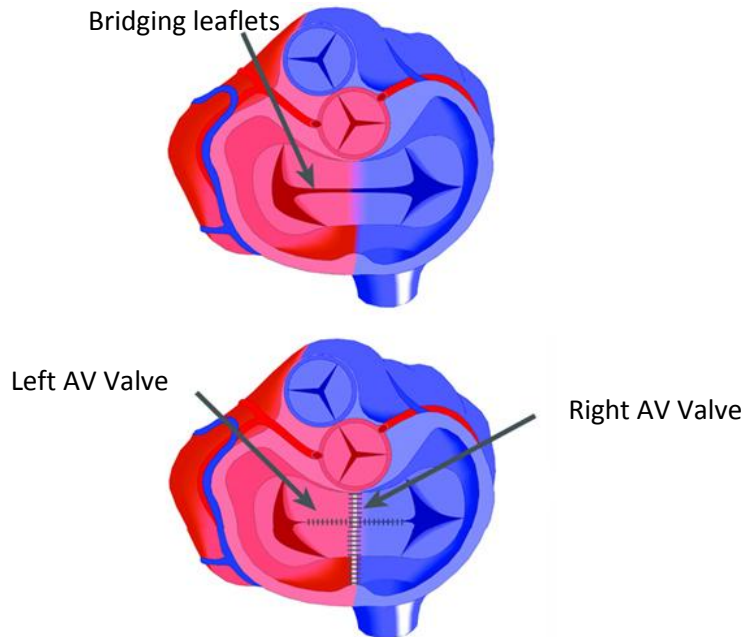
Patch Repair of Complete AVSD

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B. Repair of atrioventricular valves

1. Cleft mitral valve - least complicated to repair cleft
2. Common AV valve
 - a. Complexity depends on anatomy of valve
 - b. Goal to have a competent, non stenotic mitral valve
3. 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.

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C. Placement of band on pulmonary artery

1. Rarely done
2. Used for complex lesions

V. Long Term Complications/Interventions (Refer to Problem Section for specific complications) (Cetta, 2009; Rodrigues, 2011)

- A. Regurgitant/Stenotic AV valves
- B. Arrhythmias
- C. Sub aortic stenosis
- D. Pulmonary hypertension
- E. Greatest risk of mortality due to reoperations.

VI. Routine Cardiology Care (Warnes, 2008)

- A. Every 12-24 months by a cardiologist with experience in adult congenital heart disease

- B. Cardiac studies as indicated by assessment/symptoms
 - 1. Serial electrocardiograms (EKG)
 - 2. 24 hour ambulatory EKG monitor
 - 3. Imaging
 - a. AV valve functioning
 - b. Evaluation of left ventricular outflow tract
 - c. As indicated by assessment and clinical problems

VII. Consideration for pregnancy (Refer to Problem Section on Pregnancy for further discussion and management) (Warnes, 2008)

- A. Pre-conception cardiac evaluation
 - 1. Assess for residual hemodynamic lesions
 - 2. Counsel for pregnancy risk and preventive measures for women with Down syndrome
- B. Well tolerated in women who have been repaired and have no major residual defects
- C. Not advised in women with pulmonary hypertension

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7/2011