Atrial and Ventricular Septal Defects

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Objectives

- To understand the embryology of atrial and ventricular septal development
- To understand the anatomy of the atrial and ventricular septums
- To understand the common structural anomalies seen in the atrial and ventricular septums
ASD/VSD Incidence UNC 2016

- Total 8060 echocardiograms
- 237 PFO (3%)
- 368 ASD (4.6%)
- 845 VSD (10.4%)
ASD - Embryology

29 days

33 days

37 days

55 days

Birth
Foramen Ovale

- Directs highly oxygenated blood from placenta to left heart in utero
- LA pressure increase pushes flap left to right and causes functional closure of PFO
- Up to 30 % of adults have a probe patent foramen
ASD Location
Secundum ASD

- 7-10% of all CHD
- Female:Male ratio - 2:1
- Mostly sporadic, but associated with NKX2.5 (5q34) and TBX5 (12q31) gene mutations
- Can be seen with syndromes
  - Holt-Oram, Down syndrome, Ellis van-Creveld syndrome, Opitz syndrome, Costello syndrome
- Associated anomalies
  - Pulmonary veins
  - Cleft MV
Clinical features

- Mostly asymptomatic
- Fatigue and dyspnea with large shunt usually in older kids
- Rare subset of symptomatic infants with FTT, CHF, and LRI’s
- If untreated, adults > 40y may develop pulmonary HTN and Eisenmenger’s Syndrome
Anomalous Pulmonary Venous Connections
Intracardiac Echocardiography
Primum ASD

- Accounts for 15-20% of all ASD’s
- Failure of the endocardial cushions to fuse and send tissue superiorly to close the primum septum (inferior limbic band)
- Associated with cleft MV (partial AVSD)
- Will not spontaneously close
Sinus Venosus ASD

- 5-10% of all ASD’s
- Abnormal insertion of the SVC or IVC overriding the atrial septum
- Sinus venous superior defect
  - At SVC insertion (superior limbic band)
  - Almost always with anomalous right pulmonary veins
- Sinus venous inferior defect
  - At IVC insertion
  - Can be seen with PAPVR
ASD Repair

Hanslik, A, Pediatrics 2006:118(4)
Device Closure

The Amplatzer Device

Device
- 0.004-0.0075” Nitinol
- Two Flat Disks
- 4mm Waist
- Dacron Mesh
- 4-34 mm Sizes
- Delivery Cable
- 7-12F
Ventricular Septal Defects
Prevalence

- Delay in the closure of the interventricular septum after 7wks gestation
- Most common type of CHD seen 0.1-0.2% of all livebirths
- 15% of these will need intervention
- May be isolated or with other defects eg. tetralogy of Fallot
- Exact incidence is underestimated due to spontaneous closure
- Increased incidence in premature infants
Embryology

- Closure of the interventricular foramen
  - Growth from the endocardial cushions
  - Growth of tissue superiorly at crest of muscular septum
  - Downward growth of infundibulum at conotruncus
- All meet at membranous septum
Anatomy
## Incidence of Spontaneous VSD Closure

<table>
<thead>
<tr>
<th>Reference (year)</th>
<th>Incidence</th>
<th>Age at closure and length of follow-up period</th>
<th>Number of patients*</th>
<th>Types of VSDs</th>
</tr>
</thead>
<tbody>
<tr>
<td>1969</td>
<td>25%</td>
<td>Infancy and childhood</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>1979</td>
<td>75%</td>
<td>Infants, by 10 years of age</td>
<td>–</td>
<td>All</td>
</tr>
<tr>
<td>1979</td>
<td>83%</td>
<td>Infants</td>
<td>–</td>
<td>Muscular</td>
</tr>
<tr>
<td>1985</td>
<td>5%</td>
<td>1.5, 6.5, 13, and 14.5 years of age</td>
<td>4/87</td>
<td>Perimembranous</td>
</tr>
<tr>
<td>1987</td>
<td>45%</td>
<td>Infants and children, follow-up of 12 months</td>
<td>20/44</td>
<td>Muscular and membranous</td>
</tr>
<tr>
<td>1987</td>
<td>37%</td>
<td>Infants and children</td>
<td>7/19</td>
<td>Membranous</td>
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<tr>
<td>1987</td>
<td>17%</td>
<td>Composite group consisting of a hospital and referred patients</td>
<td>11/66</td>
<td>Membranous</td>
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<tr>
<td>1987</td>
<td>50%</td>
<td>Infants and children</td>
<td>5/10</td>
<td>Muscular</td>
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<tr>
<td>1987</td>
<td>22%</td>
<td>Composite group consisting of a hospital and referred patients</td>
<td>7/32</td>
<td>Muscular</td>
</tr>
<tr>
<td>1992</td>
<td>76%</td>
<td>Neonates (6 months)</td>
<td>32/42</td>
<td>Muscular</td>
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<tr>
<td>1998</td>
<td>44%</td>
<td>Infants, follow-up of 12 months</td>
<td>490/692</td>
<td>All</td>
</tr>
<tr>
<td>1998</td>
<td>71%</td>
<td>Infants, follow-up of 19 years</td>
<td>490/692</td>
<td>All</td>
</tr>
<tr>
<td>1998</td>
<td>23%</td>
<td>Children</td>
<td>24/106</td>
<td>–</td>
</tr>
<tr>
<td>2000</td>
<td>8%</td>
<td>Infants (3–18 years after birth, median 9.5 years)</td>
<td>8/36</td>
<td>Membranous</td>
</tr>
<tr>
<td>2000</td>
<td>74%</td>
<td>Infants (3–18 years after birth, median 9.5 years)</td>
<td>119/161</td>
<td>Muscular</td>
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<tr>
<td>2004</td>
<td>47%</td>
<td>Average age at closure: 9 months</td>
<td>75/159</td>
<td>Perimembranous</td>
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<tr>
<td>2004</td>
<td>83%</td>
<td>Average age at closure: 9 months</td>
<td>29/35</td>
<td>Muscular</td>
</tr>
<tr>
<td>2008</td>
<td>23%</td>
<td>Adolescence (mean, 10.6 ± 5.3 years)</td>
<td>11/48</td>
<td>Perimembranous</td>
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<tr>
<td>2010</td>
<td>4%</td>
<td>&gt;16 years</td>
<td>8/200</td>
<td>Perimembranous</td>
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<tr>
<td>2011</td>
<td>84%</td>
<td>Infants (&lt;3 months)</td>
<td>126/150</td>
<td>Muscular</td>
</tr>
<tr>
<td>2014</td>
<td>75%</td>
<td>The first year of life</td>
<td>33/44</td>
<td>Muscular</td>
</tr>
</tbody>
</table>

*Numbers represent numerator/denominator.
Spontaneous closure of ventricular septal defects followed up from <3 months of age

(a) CHF -
(b) CHF +

(a) Perimembranous
(b) Muscular

Mikaya, Ped Internat 46(2) 2004
Mechanisms of VSD Closure
Perimembranous VSD

- Most common (70-80%)
- Extension into other regions
- Associated with minor TV anomalies
- Can have some malalignment
- LV-RA shunt (Gerbode defect)
Perimembranous VSD spontaneous closure
Muscular VSD

- 5-20% of all VSD’s
- May be multiple
- Central, midmuscular - may have multiple RV openings
- Marginal - along RV- septal junction
- Swiss cheese septum - large number of multiple defects
Apical muscular VSD

- 5% of all VSD’s
- Can be large
- Hard to see with RV trabeculae
Inlet VSD (AV canal type)

- 8% of all VSD’s
- Associated with cleft MV
- Only seen in apical 4 chamber view
- Commonly found in an atrioventricular septal defect
- Will not close spontaneously
Supracristal VSD (outlet, subpulmonary)

- 5-7% of all VSD’s
- More common in Asians
- At risk for AI due to right coronary cusp prolapse
Treatment

- Small vs. large defects
- Location
- Surgery vs. device
  - Patch closure
  - Primary closure
  - PA Banding
Conclusion

- ASD’s and VSD’s are very common and a majority will spontaneously resolve within the first few years of life.
- Echocardiography allows precise diagnosis of location, size, and associated anomalies.
- The goal of management is to prevent fixed pulmonary vascular resistance through medical treatment of CHF and closure of the defect.