Echo for Congenital Heart Disease
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Atrial Septal Defects (ASDs):

4 types:
1. Ostium primum
2. Ostium secundum
3. Sinus venosus
4. Coronary sinus

Ostium Primum:
- Associated with trisomy 21 (Down’s syndrome)
- Located near the AV valves and often associated with cleft anterior mitral valve leaflet or cleft septal tricuspid valve leaflet
- Primum ASD + Inlet VSD = Complete Atroventricular canal (CAVC)

Ostium Secundum
- Most common type (70-80%)
- Associated with mitral valve prolapse
- Defect in septum primum located in the Fossa Ovalis

Sinus Venosus
- Associated with anomalous pulmonary venous return

Coronary Sinus:
- Associated with a persistent left superior vena cava
Figure 1:

Figure 2:
Classification of VSDs is complicated, and confusing. Many articles and texts use multiple synonyms for the same lesion, and to make matters worse there are multiple ways of describing and classifying VSDs.

**VSDs can be classified based on:**

**Anatomic location within the right ventricle (RV):**
- Relative to Crista Supraventricularis:
  - Infracristal vs Supracristal
- Relative to RV Inflow & Outflow tracts:
  - Inlet vs Outlet
- Relationship to Semilunar Valves
  - Subaortic vs Subpulmonic
- Relationship to the Septum
  - Malalignment (anterior vs posterior)
  - Membranous vs Muscular

Types of VSDs:
- A. Type 1 = Outlet = Subpulmonic = Subvalvular = Supracristal = Conal = Intraconal = doubly committed
- B. Type 2 = Membranous ~ Peri, Paramembranous = Subaortic = Infracristal ~ Conoventricular
C. Type 3 = Inlet = AV Canal Type
D. Type 4 = Muscular = Trabecular

Outlet = Subpulmonic = Subvalvular = Supracristal = Conal = Intraconal = doubly committed VSDs are located just below the pulmonic valve and are associated with herniation of the right coronary cusp of the aortic valve creating aortic insufficiency (see figure 4, A). This is the only lesion located above the crista supraventricularis and is therefore termed supracristal. Differentiating this lesion from a membranous VSD is best done from a right ventricular inflow-outflow view, where this lesion is seen closer to the pulmonic valve than the tricuspid valve (see figure 2, letter A).

Membranous ∼ Peri, Paramembranous = Subaortic = Infracristal ∼ Conoventricular VSDs comprise the most commonly seen VSD. These VSDs occur just below the aortic valve inferior to the crista supraventricularis (infracristal) near the junction of the septal and anterior tricuspid valve leaflets. Membranous VSDs are sometimes associated with intraventricular aneurysms that form when tricuspid valve tissue acts as a windsock and plugs the hole in the septum. These lesions are sometimes associated with herniation of the aortic valve cusps, but herniation is more commonly seen with subpulmonic VSDs.
Figure 4:

- A = Outlet = Subpulmonic = Supracristal = Subarterial = Conal = Intraconal
- B = Membranous = Peri. Paramebranous = Subaortic = Infracristal = Conoventricular = Malalignment
- C = Inlet = Complete AV Canal Type
- D = Muscular = Trabecular

Crista Supraventricularis
A. Outlet = Subpulmonic = Subvalvular = Supracristal = Conal = Intraconal = doubly committed.

B. Membranous  ~ Peri, Paramembranous = Subaortic = Infracristal  ~ Conoventricular

Note the area between A & B (*) is occasionally referred to as a conoventricular VSD, most authors would include this in membranous category, but others (Boston Children’s hospital) are specific regarding the distinction.
Malalignment of the ventricular septum can result in membranous VSDs. Tetralogy of Fallot is associated with malalignment of the septum anteriorly and rightward which creates obstruction of the right ventricular outflow, along with a VSD and an overriding aorta. (fig 6).

**Figure 6: Anterior Malalignment of the ventricular septum.**

![Anterior Malalignment of the ventricular septum](image)

**Figure 7: Complete AV Canal Consists of Primum ASD & an Inlet VSD. Associated with cleft antroventricular valve leaflets.**

![Complete AV Canal](image)
Inlet VSDs (figure 4, C) are associated with trisomy 21 (Down’s syndrome) and cleft atrioventricular valve leaflets. Primum ASD + Inlet VSD = CAVC. These are located more posterior than membranous VSDs. Trabecular (muscular) VSDs (figure 4, D) are defects in the muscular ventricular septum.

References:
1. Sidebotham et al. *Practical Perioperative Transoesophageal echocardiography*
2. Mullins atlas of congenital heart disease
3. *A practical Approach to Transesophageal Echocardiography* by Perrino AC, and Reeves, Scott
   *Ventricular Septal Defects in Adults*