Congenital Heath Disease and the Adult

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The number of adults with congenital heart disease (ACHD) has increased due to significant advances in diagnosis, medical and surgical care over the past five decades; at the moment in the United States the population of patients with ACHD is approaching one million. (1). The incidence of in the United States is estimated to be 6.2 per 1000 live births. Approximately 85% of these infants are expected to survive to adulthood (2). Survival of patients with ACHD rates depend on multiple factors; year of birth, age at diagnosis, complexity of the lesion and whether or not a lesion has been surgically corrected or palliated (Table 1) (2). Definitive surgical repair at a very early age and the improved care of patients with complex CHD will result in improved survival in the future.

Congenital heart disease can be classified in several ways:

A) Simple lesions include isolated lesions such as intracardiac communications; complex pathology includes severe malformations or malpositions of the cardiac structures.

B) Acyanotic versus cyanotic lesions: Cyanotic conditions are usually associated with restrictive pulmonary blood flow in the presence of intracardiac shunting or complete arterial and venous admixtures.

C) Shunt, obstructive, regurgitant and mixed lesions: This classification is based on the physiologic spectrum of CHD. Shunt lesions may occur at the intracardiac or extracardiac levels. The direction and magnitude of the shunt depends on the size of the communication and the relative resistances of the pulmonary and systemic circulation. Shunts that are primarily left to right result in pulmonary overcirculation, ventricular volume overload and signs and symptoms of congestion. Right to left shunts due to pulmonary outflow tract obstruction or elevated pulmonary vascular resistance are seen in with hypoxemia. Obstructive lesions, of varying severity, may affect inflow or outflow regions of the heart. Regurgitant lesions rarely occur in isolation. Mixed lesions have complete mixing of the pulmonary and systemic venous return and circulation that exist in a parallel rather than in series arrangement.

Atrial Septal Defects

Atrial septal defects (ASD) account for one quarter to one third of all congenital lesions, occurring more commonly in women (3). There are four types of atrial septal defects:

1. Ostium secundum defect (75%) which occurs in the mid septum region of the fossa ovalis. Varying degrees of mitral valve prolapse and mitral regurgitation can occur.

2. Ostium primum defect (15%), which is also a form of a partial atrioventricular, canal defect and consists of a defect in the lower aspect of the interatrial septum. Abnormalities of the atrioventricular valve can occur with a “cleft” in the anterior mitral valve and septal tricuspid valve leaflet with variable degrees of regurgitation.

3. Sinus venosus defect (10%) is usually superior and posterior in relation to the superior vena cava (more frequently) and/or inferior vena cava. These defects are commonly associated with an anomalous drainage of one or more pulmonary veins into the right atrium or superior vena cava (4).

4. Coronary sinus defects (extremely rare) occur between the left atrium and coronary sinus and can be associated with a persistent left superior vena cava.

A defect in the interatrial septum allows pulmonary venous blood to pass from the left to the right atrium. A patient with a large defect can develop atrial enlargement, dilation of the right ventricle and pulmonary arteries and develop pulmonary hypertension. The potential detrimental effects of chronic...
right ventricular volume overload, paradoxical embolization and atrial dysrhythmias are considered indications for intervention.

Primary or patch closure of an atrial septal defect in childhood provides excellent operative results and nearly normal long-term survival in adults (5,6). Atrial arrhythmias may be seen especially after the third decade of life. Operative patch closure is generally recommended if the shunt is large with a pulmonary blood flow-to-systemic blood flow ratio of 1.8:1 or greater and right ventricular enlargement. In some cases with favorable anatomy, closure of the secundum defects may be accomplished by the transcatheter route (7).

Intraoperative TEE is performed to confirm the presence, size and location of the defect, degree of atroioventricular valve regurgitation, ventricular function and to evaluate for associated anomalies such as anomalous pulmonary veins, systemic venous obstruction or erroneous diversion of systemic venous drainage to the left atrium. Color flow Doppler echocardiography can demonstrate flow across the atrial septum and detect mitral or tricuspid regurgitation. To evaluate for small shunts agitated saline (contrast) can be used. Postoperatively, TEE is used to detect residual shunts by color Doppler and contrast, assess ventricular function and assess for pulmonary venous obstruction and valvular regurgitation.

**Ventricular Septal Defects**

Ventricular septal defects (VSDs) are the commonest cardiac abnormality in infants and children (8). Adults with unoperated VSDs are encountered less frequently than those with ASDs because large defects are usually closed surgically in childhood when there is evidence of congestive heart failure or pulmonary hypertension. Also, in infancy and childhood, defects have a greater rate of spontaneous closure (90% of those that close do so by the time the child is 10 years of age) (9). Small perimembranous and trabecular VSDs may completely close spontaneously even in adulthood (9).

Ventricular septal defects can be classified by anatomic location into four types

1. Perimembranous defects (70%) are found in the membranous region of the septum and can extend into the muscular, inlet or outlet regions.

2. Muscular defects (20%) are located within the trabecular portion of the septum, and can be in the central or apical areas. Multiple defects can occur, either two or three or multiple small ones known as “swiss cheese septum”.

3. Doubly committed or subarterial ventricular septal defects — so called “supracristal” defects (5%) are found just below the aortic and pulmonary valves and may have associated aortic cusp herniation and aortic regurgitation.

4. Inlet ventricular septal defects (5%) occur close to the atroioventricular valves in the posterior portion of the septum.

These septal defects permit left-to-right shunting at the ventricular level and the physiologic consequences are determined by the size of the defect and the relative resistances of the systemic and pulmonary vascular beds. If the defect is restrictive, as described above, there is a large pressure gradient between the ventricles in systole and the flow across the ventricular septal defect is limited. If the defect is large (nonrestrictive), there is minimal to no pressure difference between the left and the right ventricles and the magnitude of the shunt is dependent upon the ratio between the pulmonary and systemic vascular resistance.

In adults with VSD, the overall 10-year survival after initial presentation is approximately 75%. Patients who survive to adulthood comprise two groups, those whose VSD’s have closed spontaneously or those with nonrestrictive defects that can develop elevated pulmonary vascular resistance. About 10% of patients with nonrestrictive VSD’s develop Eisenmengers (9), syndrome or reaction, which is defined as nonrestrictive VSD, pulmonary vascular disease and reversed shunt.
TEE is particularly helpful in evaluation of these defects. TEE should be used to assess the location and size of the defect, to determine the chamber sizes and PA dimensions, to detect associated anomalies, to identify ventricular septal aneurysms if present, to evaluate the aortic valve for herniation, prolapse or insufficiency, and to determine if there is pulmonary hypertension. Color flow Doppler can be used to identify the direction and magnitude of shunt flow, and the presence of any associated valvular regurgitation. Pulsed and continuous wave Doppler can be used to determine the peak velocity across the VSD. When restrictive, the velocity is high whereas when nonrestrictive the velocity would be low. The peak velocity is then used to calculate the right ventricular systolic pressure (RVSP) which is equal to the pulmonary artery systolic pressure (PASP) in the absence of any outflow obstruction.

The formula used is \( \text{RVSP} = \text{SBP} - (\text{peak velocity of VSD jet})^2 \) (10). Postoperatively, TEE is used to detect residual shunts by color Doppler and contrast, assess ventricular function and residual aortic or pulmonic insufficiency and right ventricular outflow tract obstruction.

**Complete AV Septal Defect**

In complete AV septal defect (AVSD), or atrioventricular canal defects, there is failure of the endocardial cushions to close the atrial and ventricular septum, affecting the complete formation of the mitral and tricuspid valves. As a result, patients have a ventricular septal defect, an atrial septal defect, and varying degrees of AV valve regurgitation. AVSD are classified into several forms:

**Partial:** where the ventricular septum is intact, almost always a primum ASD and cleft in the mitral valve

**Intermediate:** the rarest form with a restrictive VSD, primum ASD and cleft mitral valve

**Complete:** Nonrestrictive VSD, primum ASD and a common orifice for the atrioventricular valves.

With large left-to-right shunts at the atrial and ventricular levels and systemic pressures in the right ventricle, pulmonary vascular disease occurs very early. Most adults with uncorrected complete atrioventricular septal defects (AVSD) are cyanotic with Eisenmenger's syndrome and are not candidates for correction. The lesions in these patients are usually corrected in infancy. The surgery consists of patch closure of the atrial and ventricular septal defects and repair of the AV valves. The unoperated adult with a complete AV septal defect is rarely a candidate for a complete repair because of the development of pulmonary vascular disease, but they may be a candidate for heart-lung transplantation or lung transplantation with intracardiac repair.

TEE is useful for defining the type and extension of the septal defects (Table 2) and the morphology of the “bridging leaflets”, which span the common orifice. Proposed by Rastelli et al. (13), the attachment of the anterosuperior leaflet defines the various types of AVSD’s (a, b and c). TEE of an AVSD demonstrates complete absence of the crux of the heart, with both low atrial and high ventricular septal defects. In the adult, color-flow imaging and Doppler studies usually show regurgitation of both AV valves and evidence of pulmonary artery hypertension.

**Tetralogy of Fallot**

Tetralogy of Fallot, the most common cyanotic defect after infancy refers to a combination of lesions consisting of a) an interventricular septal defect, b) right ventricular outflow tract obstruction c) an aorta overriding the ventricular septal defect d) right ventricular hypertrophy, which is a compensatory response to the pressure load. The right ventricular obstruction and unrestricted, large ventricular septal defect result in an elevated right ventricular pressure that is similar to systemic pressure.

Most patients with tetralogy of Fallot have had palliative operations or corrective surgery by the time they are teenagers. Occasionally, a patient reaches the third decade of life without surgery. Sometimes patients present with only palliative systemic to pulmonary arterial shunts such as Blalock-Taussig shunt (subclavian artery to pulmonary artery), Potts shunt (descending aorta to left pulmonary artery), or Waterston shunt (ascending aorta to right pulmonary artery), central interposition tube graft, infundibular...
reseption (Block procedure) or pulmonary valvotomy, right ventricular to pulmonary artery conduit without VSD closure or with fenestrated closure. Prior to surgical correction, most patients died in the second decade of life.

Although it is an extremely successful operation, complete repair of tetralogy of Fallot has several potential significant postoperative residua, including residual right ventricular outflow tract obstruction, pulmonary regurgitation, peripheral pulmonary artery stenosis of one or both pulmonary arteries and ventricular septal patch leaks. Survival in postoperative tetralogy of Fallot is about 90% at about 30 years after surgery. Thus, most adults with tetralogy of Fallot come to surgery for correction of significant hemodynamic sequelae, especially pulmonary outflow pathology and less frequently for residual VSD.

TEE is useful for definition of the pathology, which is best seen in the mid-esophageal long axis view. In patients with unrepaired Tetralogy of Fallot, TEE demonstrates various degrees of right ventricular outflow obstruction at the infundibular, valvar, supravalvar and branched levels. There is a large ventricular septal defect in the vicinity of the membranous septum with evidence of right-to-left shunting and a dilated overriding aorta. The gradient across the right ventricular outflow tract can be measured with spectral Doppler by interrogating the main pulmonary artery in a mid esophageal ascending short axis view of the aorta.

**Transposition of the Great Arteries**

These infants are born with great arteries arising from the inappropriate ventricle. The aortic valve arises anteriorly from the right ventricle and the pulmonic valve posteriorly from the left ventricle. Without mixing at the atrial or ventricular levels or patent ductus arteriosus, this lesion is incompatible with life.

Infants with this condition rarely survive without intervention. With only rare exceptions, adults have had a palliative atrial switch procedure (such as the Mustard or Senning operation), after which the right ventricle usually continues to serve the systemic circulation. One current preferred surgical approach is the arterial switch operation (Jatene procedure) designed to reconnect the great arteries to their proper ventricle, translocate the coronary arteries to the new aortic location, and restore a normal physiology.

TEE evaluation with multiple planes including contrast injection should include baffle assessment after the Senning and Mustard procedure to evaluate potential baffle leaks or venous obstruction. Evaluation of ventricular function is important as patients with an atrial switch procedure are at risk of right ventricular (systemic pump) failure. Patients with the arterial switch operation (Jatene procedure) are at risk for coronary artery obstruction, atrial or ventricular patch leaks, aortic regurgitation, and supravalvular obstruction at the site of the aortic and pulmonary artery anastomoses.

**Conclusions:**

Adults with CHD may come to the attention of anesthesiologists during:

a) cardiac surgery for the first time, for either definitive repair or palliation
b) cardiac surgery for definitive correction following palliative surgery
c) cardiac surgery for management of complications of prior intervention, or conversion of a repair to a more modern technique
d) non-cardiac surgery in the presence of untreated, palliated, or corrected lesions.

In many medical centers, TEE is now used for intraoperative assessment of patients with congenital heart disease undergoing cardiac surgery. In non cardiac surgery, where additional monitoring may be required, TEE can be used to evaluate ventricular volume, function, underlying pathology and assess any residual lesions.

**References:**