Adult Congenital Heart Disease in General Echocardiography Practice

An Introduction

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ADULT CONGENITAL HEART DISEASE IN GENERAL ECHOCARDIOGRAPHY PRACTICE

The spectrum of adult congenital heart defects seen in echocardiography practice varies according to institutional practice and expertise. Half a century ago, survival with severe congenital heart disease was less common. Today, nearly 80% of such patients in industrialized societies now survive into adulthood. Most are followed up in centers that specialize in adult congenital heart diseases (CHDs), but it is not uncommon for such adults to be seen in general echocardiography practice.

Most CHDs are compatible with survival to adulthood without need for surgical intervention (Table 1). Indeed, some lesions, e.g., mitral valve prolapse, bicuspid aortic valves (BAVs), patent foramen ovale, atrial septal defects, and Marfan syndrome may be completely asymptomatic. More severe or complex defects require interventions early in life (Table 2).

This chapter introduces the fundamentals of echocardiographic assessment in CHD followed by a concise summary of the more common CHD seen in general adult echocardiography practice. All images shown are from adults who were seen in general echocardiography practice.
Table 1
Congenital Heart Disease in Adults Compatible With Survival to Adulthood With No Prior Surgery or Intervention

<table>
<thead>
<tr>
<th>Condition</th>
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<tbody>
<tr>
<td>Mitral valve prolapse</td>
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<tr>
<td>Bicuspid aortic valve</td>
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<td>ASD</td>
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<tr>
<td>Marfan syndrome</td>
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<tr>
<td>Isolated restrictive or moderately restrictive ventricular septal defects</td>
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<tr>
<td>Mild aortic stenosis (valvular, supravalvular, subvalvular)</td>
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<tr>
<td>Mild pulmonary valve stenosis</td>
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<tr>
<td>Small patent ductus arteriosus</td>
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<tr>
<td>Ostium primum ASD (AV septal defect)</td>
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<tr>
<td>Ebstein’s malformation</td>
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<td>Corrected transposition (transposition of the great arteries)</td>
</tr>
<tr>
<td>(AV-to-ventriculoatrial discordance)</td>
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</tbody>
</table>

ASD, atrial septal defect; AV, atrioventricular.

Echocardiography in Adult CHD

Echocardiography, both transthoracic (TTE) and transesophageal (TEE), play central roles in the diagnosis and management of CHD (see Chapter 4, Table 15 for Class I indications for echocardiography in adult CHD). Optimal echocardiographic examination and interpretation requires knowledge and experience in the following:

- Normal cardiac anatomy and physiology.
- The spectrum of adult CHDs.
- The segmental approach to echocardiography examination.
- Palliative and corrective surgical and transcatheter interventions and techniques (past and present).
- Postoperative and postintervention residua and complications.
- Superimposed acquired age-related heart disease, e.g., hypertension, coronary artery disease.

Table 2
Spectrum of Congenital Heart Disease

I. Chambers and valves in normal sequence and position (S,D,S)
   A Shunting predominant
      1. Atrial septal defect (Chapter 21)
      2. Atrioventricular septal defects (Chapter 21)
      3. Isolated ventricular septal defect
      4. Patent ductus arteriosus
   B Stenosis or obstruction predominant
      1. Absent AV connections (tricuspid and mitral atresia)
      2. Absent or obstructed ventriculoarterial connections (pulmonary atresia, aortic atresia, subaortic obstruction, aortic stenosis)
      3. Obstructed great arteries (coarctation of the aorta, aortic atresia)
      4. Obstructed venous flow (total anomalous pulmonary venous return)
   C Anomalous valve position
      1. Ebstein’s malformation (anomaly)

II. Chambers and valves not in normal sequence or relationship
   A Anomalies of relationships between atria and ventricles
      1. Double-inlet left or right ventricle (with univentricular heart)
      2. AV discordance (congenitally corrected transposition of great arteries [L-TGA])
   B Anomalies of relationships between ventricles and great arteries
      1. Tetralogy of Fallot
      2. Double-outlet right and left ventricles
      3. Truncus arteriosus
      4. Ventriculoarterial discordance (transposition of the great vessels D-TGA)

*Discussed in this chapter.
AV, atrioventricular.