Right Heart Lesions

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IMAGING OF THE RIGHT HEART

The algorithms used for left ventricular assessment cannot always be applied to the right ventricle (RV), which differs in morphology, shape, and function. The RV has a complex geometrical shape with a thinner and more trabeculated myocardium than the left ventricle (LV). The chest x-ray is a preliminary imaging tool, with the lateral view best suited to detect RV enlargement. Echocardiography is the mainstay for assessment of the RV, with published adult and pediatric guidelines. Cardiac magnetic resonance imaging (CMR) is the current gold standard for RV assessment and tissue characterization. Gated cardiac computed tomography angiography (CTA) is an alternative when CMR is contraindicated.

The retrosternal location of the right ventricular outflow tract (RVOT) and pulmonary valve (PV) makes echocardiography difficult, especially in adolescents and adults. CMR and CTA allow detailed assessment of anatomy, valve mobility, presence of poststenotic dilation, and associated condition of the branch pulmonary arteries (PAs). With current CT scanners, image acquisition can be completed within seconds or less and with a short breath hold. CTA is excellent for assessment of proximal and distal PAs. CMR is excellent for sequential assessment of the PV. Anatomy and function of the RVOT and PV can be assessed by balanced steady state free precession (SSFP) CMR imaging. Turbo spin-echo with blood suppression technique is very useful for imaging RVOT and branch PAs especially in patients with metallic implants causing artifacts. Through-plane and in-plane phase contrast quantification of flow across the PV and branch PAs provides comprehensive functional assessment including valve stenosis or regurgitation, and differential pulmonary blood flow. Magnetic resonance angiography (MRA) is an alternative to CTA for assessment of extracardiac structures.

EBSTEIN ANOMALY

Overview. Ebstein anomaly accounts for fewer than 1% of all cases of congenital heart disease. Associated malformations include atrial septal defect (ASD), ventricular septal defect (VSD), pulmonary stenosis and atresia, tetralogy of Fallot, congenitally corrected transposition of the great arteries, and patent ductus arteriosus (PDA). Conduction system abnormalities and arrhythmias, including Wolff–Parkinson–White syndrome, have been seen in 22% to 42% of patients.

Etiology, Pathophysiology, and Clinical Presentation. Wilhelm Ebstein provided the first description of this abnormality in 1866. It is characterized by abnormal development and positioning of the tricuspid valve (TV) leaflets, with apical displacement of the septal and posterior leaflets. This phenomenon leads to partitioning of the RV into an atrialized basal inlet segment and an apical outflow chamber. The displaced valve leaflets may be adherent to the RV wall. The anterior valve leaflet tends to be normally positioned, however, it is frequently large and redundant, and sometimes adherent to the RV free wall with a “sail-like” appearance.

The pathophysiology of Ebstein anomaly varies, depending on the degree of dysplasia and displacement of the TV leaflets and the presence and severity of associated defects. With greater degrees of dysplasia and displacement, the severity of tricuspid insufficiency increases, resulting in elevated right atrial pressures and thus right-to-left shunting at the atrial level. Patients with mild forms of Ebstein anomaly may be asymptomatic in childhood and present with symptoms of cyanosis, right heart failure, or arrhythmia in adulthood. Patients with severe forms present in early neonatal life with severe combined cyanosis and acidosis, and a ductal-dependent circulation.

Imaging. Classically, the chest radiographic appearance includes a globular or box-shaped heart as a result of right atrial enlargement associated with normal to diminished pulmonary vascularity (Fig. 74.1). Findings can vary by clinical presentation. Patients with significant valvar dysplasia and severe tricuspid insufficiency have marked right heart enlargement and decreased antegrade pulmonary blood flow. With less marked valve displacement and tricuspid insufficiency, only mild right heart enlargement may be present (e-Fig. 74.2).

Ebstein anomaly can be readily diagnosed by both prenatal and postnatal echocardiography. Echocardiography and CMR allow for detailed anatomic and functional evaluation. The characteristic features include displacement and tethering of the septal and posterior leaflets of the TV, dilation of the right atrium, and atrialization of the RV (Fig. 74.3). Imaging goals include evaluation of the TV and severity of tricuspid regurgitation; evaluation of right-to-left shunting through the ASD or patent foramen ovale, evaluating volume and function of the RV including the atrialized portion; delayed enhancement in the thin atrialized ventricular wall and septum; PV and branch PA stenoses; left ventricular size and systolic function; and aortic arch anomalies.

Axial imaging provides reliable information about the atrialized RV. A total right/left ventricular volume index obtained by CMR has been found to correlate with heart failure markers and disease severity.

Treatment. Patients with mild forms of Ebstein anomaly may not require surgical treatment, or the need for surgery may not arise until later in life until right heart dysfunction develops. Indications for surgical intervention/reintervention and/or medical therapy include limited exercise capacity (greater than New York Heart Association class II), increasing heart size (cardiothoracic ratio >65%), cyanosis (resting oxygen saturations <90%), severe tricuspid regurgitation with symptoms, progressive right ventricular dilation or dysfunction, transient ischemic attack, or stroke.

Surgery includes valvuloplasty with either the Carpentier or Da Silva’s cone procedures. The Carpentier procedure involves reimplantation of the anterior and posterior TV leaflets at the level of the neotricuspid annulus, resulting in a monocuspid or bicuspid valve configuration. More recently described “cone reconstruction” includes mobilization and re-anastomosis of the TV leaflets and subvalvar apparatus to form a cone-shaped valve with improved competence. Other possible treatments include TV replacement or, rarely, overseeing of the TV and creation of a central shunt in infancy with an ultimate single ventricle type palliation. Transcatheter closure of the ASD may be considered if severe cyanosis is present without tricuspid insufficiency necessitating valve repair. Rarely, cardiac transplantation may be indicated. Clinically significant arrhythmias develop in some patients and require transcatheter or surgical ablation procedures.
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TRICUSPID ATRESIA

**Overview.** Tricuspid atresia features an absence of the TV with a resultant lack of a direct connection between the right atrium and RV. It accounts for approximately 3% to 4% of congenital heart disease. Most cases are sporadic, but familial cases and association with 22q11 microdeletions have been reported.

**Pathophysiology and Clinical Presentation.** The TV is usually absent; instead, muscular tissue or, less commonly, fibrous tissue is present on the floor of the right atrium. An interatrial communication is present to allow blood to reach the left side of the heart from the right, and a VSD is usually present. The degree of RV development is related to the size of the VSD. In tricuspid atresia, there is complete mixing of systemic venous and pulmonary venous return and a variable degree of cyanosis. The severity of cyanosis is related to the degree of RV outflow obstruction, the size of the VSD, the origins of the great arteries (ventriculo-arterial connections are concordant in 70% of cases and discordant or transposed in 30% of the cases), the presence or absence of a PDA, pulmonary vascular resistance (associated variable degree of pulmonary or subpulmonary stenosis), and LV outflow obstruction. The most commonly used classification system was developed by Tandon and Edwards in 1974 describing tricuspid atresia as types I, II, and III (Table 74.1). An unrestrictive VSD with no pulmonary outflow obstruction leads to greater pulmonary than systemic blood flow, resulting in mild cyanosis with overcirculated, congested lungs. In contrast, restricted pulmonary blood flow results in more severe cyanosis.

**Imaging.** The heart usually appears normal or may be mildly enlarged on chest radiography. With pulmonary outflow obstruction, normal to decreased pulmonary vascularity is noted. With no obstruction, there is cardiomegaly and increased pulmonary vascularity (e-Fig. 74.4).

The characteristic imaging feature of tricuspid atresia is replacement of the TV with a ridge of muscular and fatty tissue positioned between the enlarged right atrium and the hypoplastic RV (e-Fig. 74.5). The size of the VSD and the relationship of the great arteries should be characterized. Echocardiography is the imaging modality of choice in infancy, but CMR and CTA can be used to determine anatomy in more complex cases. These modalities are playing an increasing role in preoperative evaluation in this disease, sometimes obviating the need for cardiac catheterization (see Treatment section). Important components of cross-sectional imaging after Glenn or Fontan procedures include assessment of anatomy and flow through the cavopulmonary and Fontan pathways, atrioventricular valve function, quantification of ventricular size and systolic function, and the presence of collaterals. New MR techniques like 4D flow can be used to evaluate the vascular geometry and complex blood flow patterns in Fontan patients.

**Treatment.** Ultimately, patients with tricuspid atresia require univentricular palliation because the hypoplastic RV is not capable of handling the entire cardiac output. The goal of univentricular repair is to have the single LV support the systemic circulation and for the systemic venous return to directly enter the PAs, thus bypassing the nonfunctional RV. The first stage of single ventricle palliation is performed in early infancy. Those with cyanosis may

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<td>I, normally positioned great arteries (70%)</td>
<td>Pulmonary atresia</td>
<td>Small ventricular septal defect, pulmonary artery hypoplasia</td>
<td>Large ventricular septal defect, normal pulmonary artery</td>
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<td>II, dextro-transposition of the great vessels (25%)</td>
<td>Pulmonary atresia/stenosis</td>
<td>Pulmonary or subpulmonary stenosis</td>
<td>Large pulmonary artery</td>
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<td>III, levo-transposition of the great vessels (5%)</td>
<td>Pulmonary or subpulmonary stenosis</td>
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require a prostaglandin infusion to keep the PDA open until a surgical aorto-pulmonary shunt (like the modified Blalock–Taussig shunt) is placed. In overcirculated infants with no pulmonary stenosis, diuretics and a surgical PA band are needed.

The second stage of the palliation is generally performed between 3 and 9 months of age in the form of a bidirectional Glenn anastomosis for palliation of tricuspid atresia.30,31 This procedure completes the separation of the systemic and pulmonary circulations and eliminates cyanosis. Other approaches to creating a cavopulmonary connection, including direct anastomosis of the right atrium to the PA, have been performed between 1 and 5 years of age, involves anastomosing the inferior vena cava to the PAs via an extracardiac conduit or a dilated right atrium; stenoses within the cavopulmonary pathway; thrombosis within the cavopulmonary pathway, especially in those cases.

**PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM**

**Overview.** Pulmonary atresia (PA) with intact ventricular septum (PA/IVS), a rare cardiac disorder that is highly variable in its appearance and presentation, represents approximately 1% to 3% of all cases of congenital heart disease.29 RV outflow tract obstruction is present because of atresia of the PV or the pulmonary infundibulum with an intact ventricular septum. Obstruction of the outflow from the right heart necessitates an interatrial communication or PDA to allow for pulmonary blood flow.10

**Etiology, Pathophysiology, and Clinical Presentation.** In contrast to patients with PA with VSD, patients with PA/IVS generally have well-developed, confluent central PAs.32 Morphology of the TV however is highly variable, with both severe stenosis and marked insufficiency seen. The spectrum of RV development is also variable, with varying degrees of hypoplasia (hypoplastic right heart syndrome). The RV is less hypoplastic in the setting of severe tricuspid regurgitation. Significant RV hypertrophy is common. Associated coronary artery anomalies may include RV to coronary connections. Some may exhibit an RV-dependent coronary circulation.11 In this condition, blood from the hypertensive RV perfuses the coronary arteries with possible associated coronary stenosis or even atresia. Significant drop in RV pressure (after pulmonary valvotomy) may result in myocardial ischemia and hemodynamic collapse in those cases. RV-dependent coronary circulation is typically seen in patients with small RVs and no significant tricuspid regurgitation.11 The major clinical manifestation of PA/IVS is cyanosis, which is typically seen shortly after birth when the PDA begins to close.

**Imaging.** In infancy, the size of the heart on a chest radiograph correlates with the degree of tricuspid insufficiency, with larger cardiac size correlating with increasing tricuspid regurgitation. Severe cardiomegaly producing the “wall-to-wall” heart may be present with severe tricuspid regurgitation (Fig. 74.9).11 Echocardiography yields diagnostic imaging in infancy, and CMR may be used in complex cases.1 Imaging features include atresia of the PV or infundibulum, hypoplastic RV and TV, an interatrial communication, and a PDA. Preoperative imaging goals include a comprehensive evaluation of the right heart structures. Attention to the anatomy and function of the hypoplastic TV
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and RV is necessary (Fig. 74.10). Coronary artery anatomy and ventriculo-coronary communications may not be well visualized on echocardiography, and thus additional evaluation with angiocardiology, CTA, or MRA may be necessary. Postoperatively, the type of imaging that is used will depend on the type of repair that has been performed. CMR may be important in these postoperative patients since RV evaluation is limited with echocardiography.

**Treatment.** Initial management of a neonate with PA/IVS includes the administration of prostaglandins to prevent PDA closure and maintenance of pulmonary blood flow. Interventions depend on the degree of RV and PA development and the coronary circulation pattern. A pulmonary valvotomy is contraindicated in the presence of an RV-dependent coronary circulation. A transcatheter pulmonary valvotomy and balloon dilation is performed if the morphology of the imperforate PV is favorable. When the pulmonary anatomy is not favorable for a transcatheter intervention, a surgical valvotomy may be performed. In the setting of significant RV hypoplasia, a one-and-one-half ventricular repair may be considered. This repair uses a bidirectional Glenn shunt to partially unload the right heart so that it receives venous return only from the inferior vena cava. Univentricular repair or cardiac transplantation may be considered with significant coronary anomalies.
RV hypertrophy with elevation of the cardiac apex and associated filling in of the upper retrosternal space can be seen. Echocardiography is the initial imaging method of choice. The role of CMR has increased and is particularly important in adult patients with limited echocardiographic windows. Imaging findings include valve thickening and systolic doming with poststenotic enlargement of the PAs (Fig. 74.12). With a dysplastic valve, the

Figure 74.10. Pulmonary atresia with intact ventricular septum after undergoing right classic Glenn and right atrium (RA) to pulmonary artery Fontan procedures in a 21-year-old. A four-chamber steady-state free-precession MR image shows the hypoplastic right ventricle (RV). The RA is mildly dilated. LA, Left atrium; LV, left ventricle.

Figure 74.11. Pulmonary valve stenosis. Frontal chest radiograph in a 10-year-old shows a normal heart size and pulmonary vascularity. Mild enlargement of the pulmonary trunk and left pulmonary artery is present (arrow).

Figure 74.12. Pulmonary valve stenosis. A steady-state free-precession MR image coronal to the right ventricular outflow tract (RV) shows a poststenotic jet of turbulent flow (arrow) across the pulmonary valve. Ao, Aorta; PA, pulmonary artery.

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pulmonary annulus is small. Imaging goals include evaluation of the level of the obstruction, valve anatomy, and determination of the peak systolic velocity and pressure gradient across the obstruction. The pressure gradient can be estimated from the flow velocity obtained by echocardiography or CMR phase contrast imaging using the modified Bernoulli equation, \( \Delta P = 4V^2 \) (where \( \Delta P \) is the pressure gradient and \( V \) is the peak velocity across the PV in m/sec). Lower peak velocities may be obtained on CMR relative to echocardiography. Quantitative assessment of the RV size and systolic function is necessary, as is assessment of the left heart. MRA or CTA allows determination of peripheral PA stenosis. After intervention, imaging is used to determine the degree of residual pulmonary stenosis and the severity of pulmonary regurgitation, and to evaluate RV size and systolic function.

**Treatment.** Infants with critical PVS are treated medically with prostaglandin E1 infusion to maintain ductal patency; inotropic support and intubation may be needed. Balloon valvuloplasty is the treatment of choice for isolated PVS in a nondysplastic valve, with excellent outcomes. A systemic-to-pulmonary artery shunt is rarely indicated for continued cyanosis. Balloon valvuloplasty often is used as the initial treatment for “dysplastic” PVs, but surgery is frequently needed because of the lower response rate. Peripheral pulmonary stenoses often require repeat angioplasties and/or stent placement.

Indications for catheter reintervention or surgery include residual RV outflow obstruction from severe infundibular hypertrophy, residual PVS, or peripheral arterial stenosis. PV replacement is indicated for symptomatic severe pulmonary insufficiency, RV dysfunction and dilation, or significant ventricular arrhythmia.

**SUGGESTED READINGS**

**REFERENCES**
Full references for this chapter can be found on www.expertconsult.com.