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Review Article

Congenital Malformations of the Tricuspid Valve: Diagnosis and Management - Part II

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Abstract

Congenital malformations of the tricuspid valve, namely, Ebstein's malformation of the tricuspid valve, and Ebstein's type of anomaly of the morphologic tricuspid valve on the left-side in subjects with congenital corrected transposition of the great arteries are reviewed. The pathologic and pathophysiologic features of these lesions are enumerated followed by a discussion of techniques of diagnosis and management methods. It was concluded that these congenital abnormalities of the tricuspid valve can be effectively diagnosed with the currently available non-invasive and invasive investigative techniques and these defects can successfully be managed with the existing therapeutic medical and surgical methods.

INTRODUCTION

A number of congenital malformations of the tricuspid valve have been described and these include, tricuspid atresia, tricuspid stenosis, tricuspid insufficiency, Ebstein's anomaly of the tricuspid valve, and Ebstein's type of abnormality of the morphologic tricuspid valve on the left-side in patients with congenital corrected transposition of the great arteries (CCTGA) [1,2]. In these reviews, tricuspid atresia was discussed in Part I of this series. In this Part II, Ebstein's anomaly of the tricuspid valve and Ebstein's type of abnormality of left atrio-ventricular (AV) valve in CCTGA will be discussed. Similar to Part I, pathologic anatomy and pathophysiology of each defect will be presented first followed by the diagnostic features by multiple laboratory techniques and methods of management.Ebstein's Anomaly of the Tricuspid Valve.

Ebstein's anomaly of the tricuspid valve is a cyanotic, congenital heart defect (CHD) characterized by a downward (apical) displacement of the insertion of the septal and posterior leaflets of the tricuspid valve with the resultant tricuspid regurgitation and right atrial (RA) enlargement of varying degrees [2]. This anomaly, now called Ebstein's, was first described by Wilhelm Ebstein in 1866 in an autopsy of a 19-year-old laborer who had had cyanosis and dyspnea since early childhood [3]. Ebstein's anomaly of the tricuspid valve accounts for 0.3 to 0.6% of all CHDs [4,5].

PATHOLOGIC ANATOMY

The chief pathology of Ebstein's is a downward (or apical) displacement of insertion of the septal and posterior leaflets of the tricuspid valve from the original annulus level. This displacement makes a portion of the right ventricle (RV) become part of the RA and is described as an "atrialized" RV (aRV). Consequently, the RV distal to the displaced insertion of the tricuspid valve is smaller and lacks some of the inlet portion. The third (anterior) leaflet of the tricuspid valve in this lesion is usually large, sail-like, and redundant. But, the coaptation of these leaflets is inadequate, leading to varying degrees of tricuspid regurgitation. There are also varying grades of dysplasia of the tricuspid valve tissue contributing to the tricuspid regurgitation. Persistent tricuspid regurgitation leads to RA enlargement. In some patients, the RV outflow tract may be obstructed secondary to valvar pulmonary stenosis or atresia.

The presence of an accessory conduction pathway between the atrium and ventricle is seen in 30% of Ebstein's anomaly cases, manifesting as Wolf-Parkinson-White (WPW) syndrome. Some patients may develop supra-ventricular tachycardia (SVT) from a concealed pathway [6,7]. Associated lesions include patent foramen ovale (PFO), atrial septal defect (ASD), ventricular septal defect (VSD), patent ductus arteriosus (PDA), tetralogy of Fallot, double-outlet right ventricle, mitral valve prolapse or stenosis, transposition of the great arteries, non-compaction of the myocardium, and absent pulmonary valve syndrome [8].

PATHOPHYSIOLOGY

In patients with mild Ebstein's, the tricuspid valve function is close to normal and the defect may not be detected until later in life. In moderate to severe forms, with each atrial contraction, the blood is propelled into the aRV. With the ventricular contraction that follows, the blood is forced back into the RA. With the succeeding atrial contraction, this blood is forced back into the aRV. This back-and-forth blood flow, the so-called ping-pong effect [2], causes RA dilatation and increases the RA pressure; the latter results in a right-to-left shunt across the ASD/PFO. This shunt results in arterial desaturation and pulmonary oligemia. These pathophysiologic abnormalities are further accentuated

by the usual high pulmonary vascular resistance (PVR) in the newborn babies.

In severe cases of Ebstein's anomaly, the functional portion of the RV may be too small to generate enough systolic pressure to open the pulmonary valve and permit forward flow via the pulmonary valve. This is compounded by high PVR and a PDA leading to the inability of the pulmonary valve leaflets to open. This leads to what is referred to as "functional" pulmonary atresia. In such patients, the pulmonary valve may open after the PVR decreases, either naturally or by medical treatment. Therefore, there may be associated functional pulmonary valve atresia, especially in the immediate neonatal period when there is transient pulmonary hypertension. Right-to-left shunting at the atrial septal level, leading to cyanosis in the newborn, may resolve as the PVR decreases along with the establishment of forward flow via the pulmonary valve. The degree to which such a resolution occurs depends on the severity of the defect. Cyanosis may return in later childhood or adolescence when the tricuspid valve function deteriorates (causing regurgitation).

DIAGNOSTIC METHODS

Chest X-ray

In moderate and severe cases, severe cardiomegaly is the striking feature, secondary to right atrial enlargement (Figures 1 and 2A). In milder cases, the cardiac silhouette may be closer to normal (Figure 2B). The lung fields are either normal or oligemic, depending upon the magnitude of the pulmonary blood flow. Severe cardiomegaly due to right atrial enlargement along with pulmonary oligemia in a cyanotic newborn is unique to Ebstein's anomaly. However, a similar x-ray findings may be seen with critical pulmonary stenosis or pulmonary atresia with intact ventricular septum, and rarely tricuspid atresia, when there is associated obstruction at the PFO level causing severe right atrial enlargement. Echocardiography will differentiate these defects from Ebstein's anomaly.

Electrocardiogram (ECG)

Tall and peaked P waves indicating RA enlargement (Figure 3) are typically seen. Prolongation of PR interval is noted in 2/3rd of the neonates [9-11]. Relatively-low QRS voltages and right bundle branch block pattern are usually seen in Ebstein's anomaly. The features of WPW syndrome, with a short PR interval and delta wave (Figure 4) are reported in approximately 30% in Ebstein's anomaly patients, while a few (12%) have evidence of pre-excitation with a normal PR interval [12].

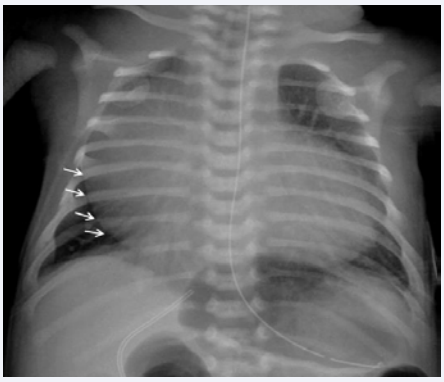

Figure 1 A chest X-ray of a neonate with Ebstein's anomaly. The X-ray shows severe cardiomegaly secondary to right atrial enlargement (arrows). The pulmonary vascular markings appear normal in this example although pulmonary oligemia is more typical in severe Ebstein's anomaly. Reproduced from Reference [9].

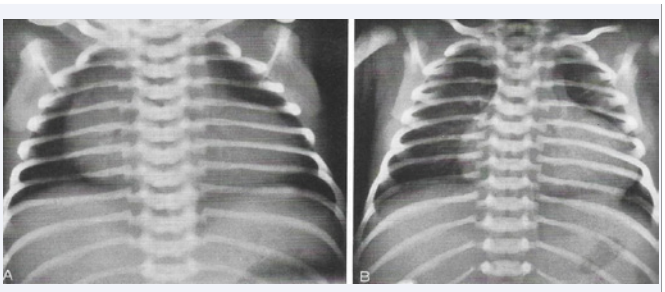

Figure 2 Chest X-rays (postero-anterior views) of two neonates with Ebstein's anomaly. A shows moderate to severe cardiomegaly secondary to right atrial enlargement. B shows mild cardiomegaly. The pulmonary vascular markings appear markedly decreased in both A and B. Reproduced from Reference [2].

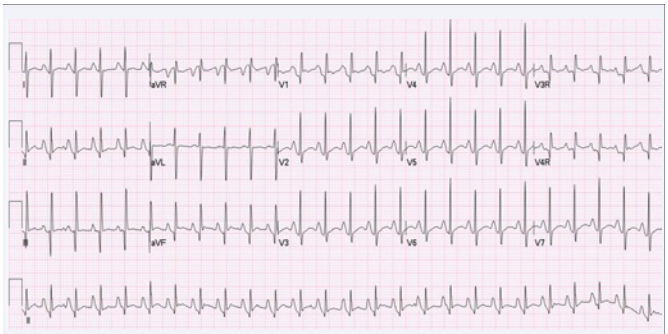

Figure 3 An electrocardiogram of a newborn with Ebstein's anomaly. Note the tall P waves representing right atrial enlargement. Reproduced from Reference [12].

Echocardiography

Echocardiography provides almost all of the necessary information for a complete diagnosis along with an assessment of the severity of the lesion [2,13-15]. M-mode echo may demonstrate delayed closure of the tricuspid valve relative to that of mitral closure [2]. Two-dimensional (2D) echocardiogram shows RA enlargement and downward dislodgment of the tricuspid valve leaflets (Figures 5A & 6A). The anatomy of the leaflets, including valve dysplasia and abnormalities of the chordal attachments should also be evaluated. Doppler studies show tricuspid insufficiency (Figure 6B) and shunting from RA to left atrium (LA) via the PFO (Figure 6B). Peak tricuspid regurgitation jet velocity may be used in estimating the RV/pulmonary artery (PA) systolic pressure by the use of a simplified Bernoulli equation. Assessment of the shunt at the atrial level across the PFO or ASD (Figure 6) is important and will correlate with the degree of cyanosis noted clinically. The patency of the ductus should also be evaluated. Even though the atrialized part of the RV bulges into the left ventricular (LV) outflow tract

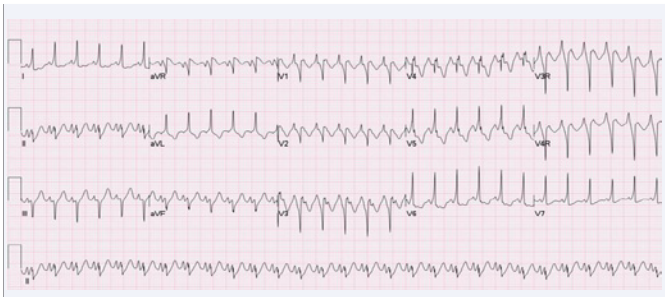

Figure 4 An electrocardiogram of a neonate with Ebstein's anomaly and Wolf-Parkinson-White (WPW) syndrome. This example shows tall, peaked P waves (lead II) consistent with right atrial enlargement. A short PR interval and Delta wave are consistent with WPW syndrome. Reproduced from Reference [9].

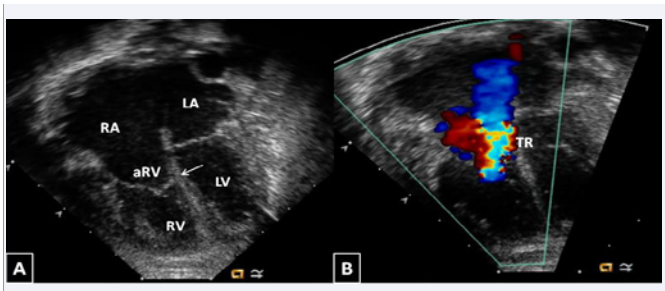

Figure 5 Apical four-chamber views of an echocardiogram of a newborn with a mild to moderate form of Ebstein's anomaly. Panel A shows a two-dimensional image demonstrating the insertion of the septal leaflet of the tricuspid valve (arrow) which is displaced apically in comparison to the insertion of the mitral valve. The atrialized right ventricle (aRV) is shown. Panel B shows a color Doppler image demonstrating moderate tricuspid valve regurgitation (TR). LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle. Reproduced from Reference [12].

(Figure 7), clinically significant LV outflow tract obstruction rarely occurs.

While the septal and posterior leaflets of the tricuspid valve are displaced downwards, the anterior leaflet is large and sail-like and has abnormal chordal attachments which may lead to RV outflow tract obstruction. This may be detected during echo studies. The pulmonary valve may fail to open adequately; this is either due to anatomic valve leaflet fusion (true pulmonary valve atresia) or due to functional atresia of the pulmonary valve due to the inability of the RV to generate systolic pressure high enough to overcome the pressure in the PA. The PA pressure may be elevated in the early neonatal period due to high PVR or a large PDA transmitting aortic pressure to the PA. Sometimes it is not easy to distinguish functional type from true atresia of the pulmonary valve, but if a pulmonary insufficiency jet on color Doppler imaging is seen, that would indicate functional type of the pulmonary valve atresia.

Prediction of Outcome based on Echocardiography

Several methods to predict outcomes have evolved over time; these are largely based on echocardiographic parameters in the fetus or newborn, and these include the Celermajer index [16] and Simpson-Andrews-Sharland (SAS) score [17].

Celemajer Index

Apical four-chamber views of the echocardiogram (Figure 8) may be used to derive Celermajer index [16]. The index is calculated as follows: ratio of the area of the RA + the atrialized component of the RV to the pooled area of the noninvolved RV, LV and LA (Figure 8). The higher the grade, greater is the mortality: 1. Grade 1 (a ratio of < 0.5) - mortality of 0%, 2. Grade 2 (a ratio of 0.5 to 0.99) - mortality of 10%, 3. Grade 3 (a ratio of 1.0 to 1.49) - mortality of 44%, and 4. Grade 4 (a ratio of ≥1.5) - 100% mortality (Figure 9) [16].

Simpson-Andrews-Sharland Score

The SAS score is based on observations from the first prenatal echocardiogram and includes

1. Cardiothoracic ratio in fetal echocardiogram.
2. Celermajer index.
3. RV-LV ratio.
4. Reduced/absent pulmonary valve flow, and
5. Retrograde ductus arteriosus flow [17].

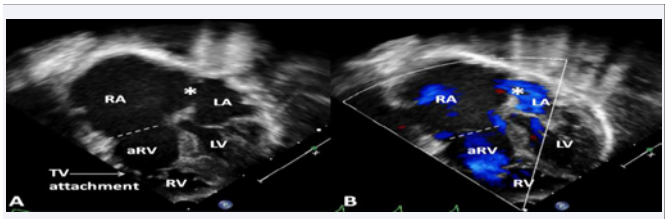

Figure 6 An echocardiogram of a neonate with Ebstein's anomaly. Apical four-chamber views are shown. A shows a two-dimensional image. B shows the same image with color Doppler flow mapping. Note the significant downward displacement of the tricuspid valve (TV) attachment (solid arrow labeled "TV attachment") from the true annulus of the tricuspid valve (marked by a dotted line). The portion of the right ventricle (RV) between the tricuspid valve true annulus and the tricuspid valve attachment is the atrialized portion of the right ventricle (aRV). * represents the location of the patent foramen ovale (PFO). The color Doppler image in B shows right-to-left shunting (blue color). Note the markedly enlarged right atrium (RA) in both A and B. LA, left atrium; LV, left ventricle. Reproduced from Reference [9].

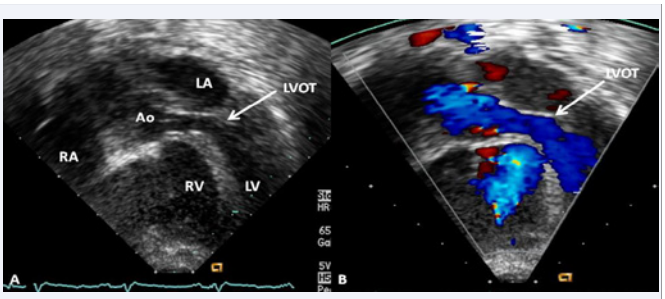

Figure 7 An apical five-chamber view of an echocardiogram showing a two-dimensional frame (A) and with color Doppler (B) showing the bulging of the right ventricle (RV) into the left ventricular outflow tract (LVOT). In this example, there is no significant obstruction; note the laminar flow in the LVOT and aorta (Ao). LA, left atrium; LV, left ventricle; RA, right atrium. Reproduced from Reference [12].

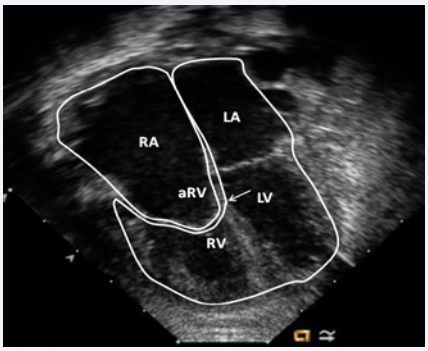

Figure 8 An apical four-chamber view showing all chambers of the heart in end diastole. A tracing on the image shows the two areas used to obtain the ratio (Celermajer index) for prediction of outcome in fetal and neonatal echocardiograms (see text for further details). The abbreviations are the same as those used in Figure 5. Reproduced from Reference [12].

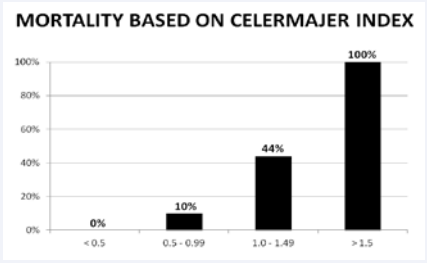

Figure 9 Mortality rate based on the Celermajer index. Data were derived from the original study [16]. Reproduced from Reference [12].

As one can see many items used in this score are derived from echo parameters. This score predicts survivors vs. non-survivors and is useful for counseling parents during pregnancy. The possible scores range from 0 to 10. Similar to the Celemajer index, higher scores correlate with higher mortality. When the scores were ≤ 3, survival was 91%. There were no survivors when the score was ≥ 5 in their original study [17].

Magnetic Resonance Imaging (MRI) and Computed tomography (CT)

MRI and CT studies are not necessary for evaluation and diagnosis in infants and children. However, in adolescents and adults with poor acoustic windows, MRI and CT may help establish the diagnosis and evaluate severity of the lesion [18].

Cardiaccatheterizationandselective Cineangiography

As alluded to in the preceding section, the diagnosis of Ebstein's can be suspected by clinical, chest X-ray, and ECG findings and confirmed by echocardiographic studies, and therefore, cardiac catheterization and selective cineangiography are not necessary to establish the diagnosis. In the past, simultaneous recording of pressure and intracardiac electrocardiograms as the catheter is withdrawn from the RV to aRV and RA (Figure 10) to demonstrate aRV [2], but this is no longer routinely performed because of the diagnosis can easily be made by echocardiographic studies. Similarly, angiograms are not necessary in all cases. Examples to demonstrate angiographic anatomy are shown in Figures 11 and 12.

Ebstein's Malformation of the Tricuspid Valve with Atresia

A rare combination of Ebstein's with atresia of the tricuspid valve has been reported [19]. Angiographic and pathologic figures of this anomaly are shown in Figures 13 to 15.

MANAGEMENT

Medical and Transcatheter management

The medical management is largely dependent upon the severity of the symptoms and age at presentation [5,9,12,20,21]. Cyanotic neonates who are otherwise asymptomatic do not need any active treatment unless the cyanosis is severe. Cyanosis usually resolves to a variable extent as and when the PVR decreases with time. The treatment for severe cyanosis at birth consists of temporarily keeping the ductus open by administering PGE_1 infusion (0.05-0.1mcg/kg/min) [22-25] until the pulmonary vascular resistance drops. The use of inhaled nitric oxide (iNO) in the first few days of life to reduce the PVR may help to improve the pulmonary blood flow and hence the systemic oxygenation. Such therapy is usually needed for a few days only, after which the infant can be safely weaned off of iNO. Intubation and positive

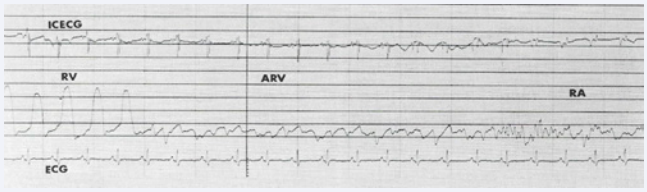

Figure 10 Simultaneous intracavitary electrocardiogram (ICECG) at the top and pressure recordings (in the middle) as the catheter is withdrawn from the right ventricular (RV) body to the atrialized RV (ARV) and then to the right atrium (RA) in a patient suspected of having Ebstein's anomaly of the tricuspid valve. Surface electrocardiogram (ECG) is shown at the bottom. Note ventricular ICECG and ventricular pressure in the body of the RV (left panel), ventricular ICECG and atrial pressure (middle panel) and atrial ICECG and atrial pressure (right panel). This pattern is characteristic for Ebstein's anomaly of the tricuspid valve. Reproduced from Reference [2].

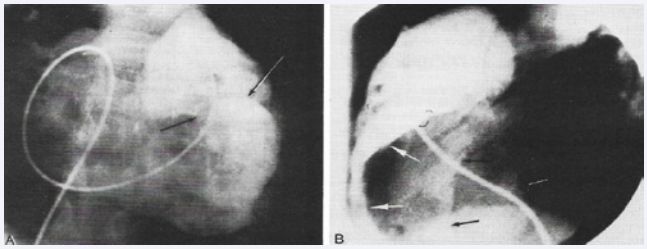

Figure 11 Selected frames from a right ventricular cine-angiogram in anteroposterior (A) and lateral (B) views in a patient with Ebstein's anomaly of the tricuspid valve. In A, black arrows point to displaced tricuspid valve attachment. In B white arrows point to displaced tricuspid valve attachment while black arrows point to the true annulus of the tricuspid valve. Reproduced from Reference [2].

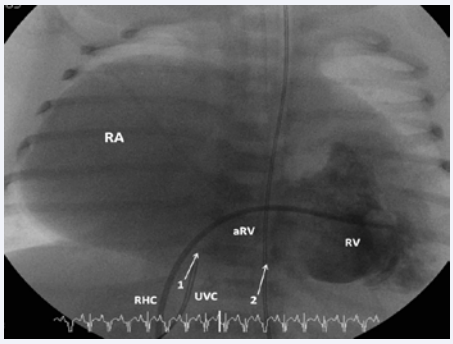

Figure 12 A right ventricular (RV) angiogram in postero-anterior view from a patient with Ebstein's anomaly. Severe tricuspid valve regurgitation has led to the opacification of a markedly enlarged right atrium (RA). In this example, the pulmonary arteries are not opacified, indicating the presence of anatomic or functional pulmonary atresia. There is a trilobed appearance with two notches; the first (arrow 1) separates the RA and the atrialized portion of right ventricle (aRV) and represents the location of the true tricuspid valve annulus. The second notch (arrow 2) represents the origin of the displaced tricuspid valve leaflets. RHC, right heart catheter; UVC, umbilical venous catheter. Reproduced from Reference [12].

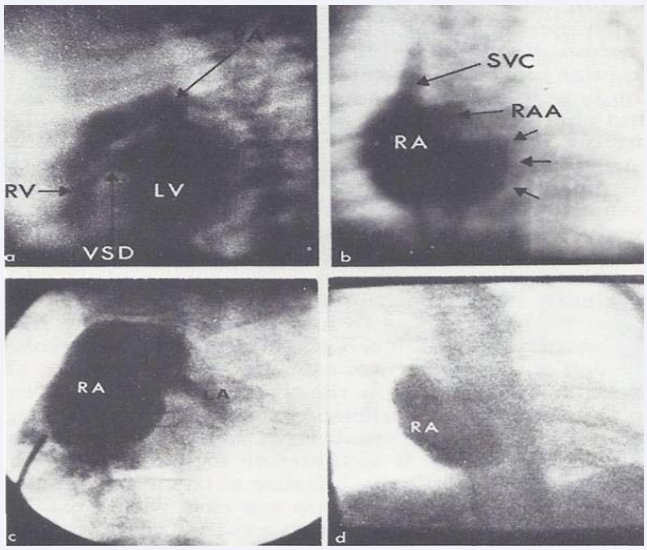

Figure 13 a. Selected left ventricular (LV) cineangiographic frame in left anterior oblique view demonstrating a ventricular septal defect (VSD). The right ventricle (RV) is opacified via the VSD and looks close to normal in size. b. Selected right atrial (RA) cineangiographic frame in postero-anterior view prior to opacification of the left atrium (LA). The contrast material delineates the atretic tricuspid valve and is shown by small unlabeled arrows. This is rounded and extends more to the left than in other tricuspid atresia cases. c and d. Right atrial angiographic frames from two patients with classic tricuspid atresia are shown for comparison with figure b. These frames are frozen prior to full opacification of the LA. PA, pulmonary artery; RAA, right atrial appendage; SVC, superior vena cava. Reproduced from Rao PS, et al., Amer J Cardiol 1973; 32:1004-9 [18].

pressure ventilation may help to manage pulmonary hypertension more effectively. Deep sedation and muscle relaxants may be necessary for few days to manage pulmonary hypertension. The correction of metabolic acidosis with bicarbonate infusion and inotropic (dopamine/dobutamine) infusion for low cardiac output may be needed. Some neonates may require a surgical systemic-pulmonary shunt [26,27] or stenting of the ductus [28-30] to maintain adequate pulmonary blood flow, and thus maintain adequate systemic oxygen saturation.

Symptoms of heart failure secondary to severe tricuspid regurgitation may be treated with anti-failure medications such as furosemide and digoxin. SVT related to WPW or other accessory pathways or atrial flutter because of an enlarged RA should be controlled using appropriate anti-arrhythmic medications; adenosine for acute control of SVT and suitable anti-arrhythmic medication such as propranolol, flecainide and amiodarone may become necessary for long-term control. Ablation of the accessory pathway causing SVT is reserved for older children and adults. Restrictive PFOs are rare with Ebstein's and do not usually require balloon atrial septostomy. While balloon atrial septostomy may relieve systemic venous congestion, one should be cognizant of the increase in cyanosis that may occur due to increased right-to-left shunt at the atrial level after the septostomy. RV outflow tract obstruction, secondary to anterior leaflet attachments can't be addressed with medical or transcatheter therapy. However,

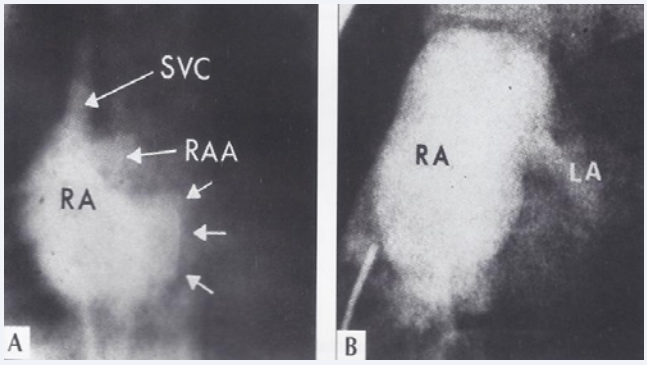

Figure 14 A & B: Figures 13 b and c are put together to more clearly illustrate right atrial angiographic differences between Ebstein's and classic types of tricuspid atresia. Abbreviations are same as those used in Figure 13. Reproduced from Reference [19].

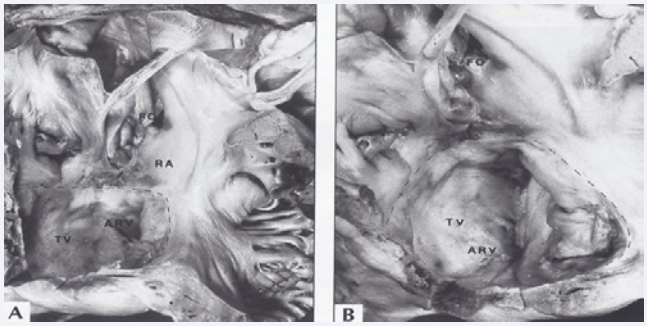

Figure 15 A. The interior of the right atrium (RA) and the atrialized right ventricle (ARV) are shown. B. This is close-up of A. The interrupted line demarcates the true tricuspid valve annulus. The tricuspid valve (TV) leaflets are displaced downward (Ebstein's malformation) and fused, incorporating a part of the right ventricle into the RA. FO, fossa ovalis. Reproduced from Rao PS, et al., Amer J Cardiol 1973; 32:1004-9 [18].

balloon pulmonary valvuloplasty is likely to help in patients with associated valvar pulmonary stenosis [31].

Surgical management

Every attempt should be made to avoid surgery in the neonatal period. However, in severe cases, survival may be difficult without surgical intervention [32]. Specific surgery that will help the patient may vary depending upon the pathophysiologic and clinical features. A number of surgical procedures are available for use [32-36]. Simple closure of an ASD (surgical or transcatheter) to reduce cyanosis from right-to-left shunting at the atrial level, though it may be used in older children, is not a viable option in the neonate. An aortopulmonary shunt [26,27] may be needed to overcome diminished pulmonary blood flow secondary to severe RV outflow tract obstruction. Stenting the ductus [28-30] has also been used as an alternative to surgical shunts. Repair of the tricuspid valve should be avoided in the neonatal period due to extremely poor results. The Starnes procedure may be performed in severe cases of Ebstein's anomaly in which the patients are severely symptomatic as neonates. The Starnes procedure [36] is an RV-exclusion procedure consisting of patch-closure of the tricuspid valve and a Blalock-Taussig shunt to provide pulmonary blood flow (Figure 16). Such patients are destined for a single-ventricle route, eventually undergoing two more surgeries including a bidirectional Glenn operation around 6 months of age and a Fontan operation at 2-4 years of age [32], as described in Part I of this series.

Multiple approaches to the repair of Ebstein's anomaly have been described [12,34,37-39] and all such procedures are reserved for older children, adolescents and adults. The essential components of surgical repair in Ebstein's anomaly include repair or replacement of the tricuspid valve, plication of the tricuspid valve annulus, plication of the aRV, attempting to bring the leaflets to the anatomic tricuspid valve annulus, and plication or reduction-plasty of the right atrium [40]. The most modern of the repair procedures is the cone repair described by de Silva and colleagues [39]. Some patients with moderate to severe forms of Ebstein's anomaly who do not require neonatal surgical repair may be candidates for the so-called one-and-one-half ventricle repair [41]. This procedure consists of performing a bidirectional Glenn operation [42] and leaving the intracardiac lesions without any repair. In one-and-one-half ventricle repair, the SVC flow is directed into both the branch pulmonary arteries, thus reducing the amount of blood that the RV has to handle. The atrial defect may be closed either surgically or via transcatheter methods. For patients with arrhythmia, transcatheter or surgical ablation therapy may be performed in older children and adults. These treatment options have been recently reviewed elsewhere [12] for the interested reader.

EBSTEIN'S ANOMALY OF THE MORPHOLOGIC TRICUSPID VALVE WITH CONGENITAL CORRECTED TRANSPOSITION OF THE GREAT ARTERIES

Congenital corrected transposition of the great arteries is a rare complex CHD in which the ventricles are reversed, right to left. CCTGA was initially described by von Rokitansky in 1875

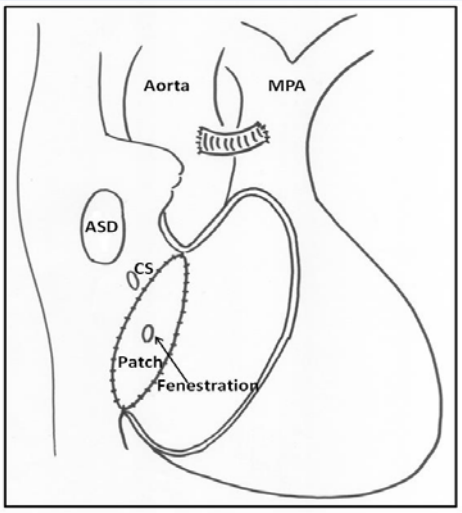

Figure 16 Schematic diagram showing the principal features of the Starnes Procedure. Patch-patch closure of the tricuspid valve orifice. Fenestration-this is created to allow for the escape of blood that returns through the Thebesian veins to the right ventricle. ASD-atrial septal defect. A systemic-to-pulmonary arterial shunt ("central shunt) is shown in the figure. Alternatively, a Blalock-Taussig shunt may be performed. CS, coronary sinus. Reproduced from Reference [12].

[43]. The incidence of this defect is close to 0.5% of all CHDs [44,45].

Pathologic anatomy

In the most common form, the atria are normally positioned (atrial situs solitus), atrio-ventricular discordance is present, the morphologic left ventricle (MLV) is on the right and the morphologic right ventricle (MRV) is on the left, and ventriculo-arterial discordance exists. In essence, the ventricles are reversed. The RA passes the systemic venous return into a right-sided, MLV and then into the PA, and the LA empties the pulmonary venous blood into a left-sided, MRV and then into the aorta; thus, the blood circulation is normal [2,46,47]. Because the aorta arises from the right ventricle and the pulmonary artery comes off the LV, transposition of great arteries is deemed to be present. However, a normal blood flow pattern is preserved, hence the term CCTGA, or corrected transposition for short. Normally the aorta is positioned to the right of the PA; however, in this defect, the aorta is located to the left of the PA; hence the term l-transposition of the great arteries.

Although patients without any other cardiac defects have been reported [48,49], most CCTGA patients have significant associated defects such a VSD, pulmonary stenosis (PS), atrioventricular (AV) block (spontaneous, or following catheter or surgical intervention), and/or an Ebstein's-like malformation of the left-sided, morphologic tricuspid valve [2,46,50].

Pathophysiology

As mentioned in the preceding section, the blood flow pattern is normal in CCTGA and therefore, there is no physiological blood flow abnormality in patients without any associated cardiac defects. In patients with moderate to large VSD, the pulmonary

blood flow is increased. In subjects with VSD and severe PA, the pulmonary blood flow is decreased. In patients with moderate to severe left AV valve regurgitation, there is MLV volume overloading.

DIAGNOSTIC METHODS

Chest X-ray

Straightening of the left upper border of the heart related to an l-transposed ascending aorta may be seen. Moderate to severe cardiomegaly may be seen in patients with moderate to severe left AV valve insufficiency.

Electrocardiogram

ECG usually demonstrates characteristic features including the absence of a Q wave in the left precordial leads and the presence of a Q wave in V1, indicating the reversal of septal depolarization (Figure 17). Atrio-ventricular conduction block (first-, second- or third-degree heart block) may be present in some patients.

Echocardiogram

Echocardiogram is diagnostic, although it may be a challenge for the beginner to perform and interpret. An absence of continuity between the left AV valve and the aortic valve due to the presence of a conus on the left side (MRV) is one of the echocardiographic features. Scrutiny of the atrioventricular valve attachment may help diagnose ventricular inversion; a higher level of attachment of the mitral valve on the right compared to the tricuspid valve attachment on the left indicates that the ventricles are inverted (Figure 18). The degree of downward displacement of the left-sided tricuspid valve may be assessed in a four-chamber view (Figure 19A). The left AV valve insufficiency can also be demonstrated by color Doppler (Figure 19B). Enlargement of the LA and MRV can also be shown by m-mode and 2D imaging and the degree of their enlargement is proportional to the magnitude left AV valve insufficiency. The size, location and shunting across the VSD and the degree of PS may be shown by echo-Doppler studies, but will not be illustrated in this paper dealing left AV valve abnormality.

MRI and CT

MRI and CT studies are not necessary for evaluation and diagnosis in infants and children. However, in adolescents and adults with poor acoustic windows, MRI and CT may help establish the diagnosis and evaluate severity of the lesion [12].

Cardiaccatheterizationandselective Cineangiography

Catheterization and angiography are not ordinarily necessary, although, on occasion, when the diagnosis is not clear, catheterization and angiography may be helpful. The diagnosis of Ebstein's anomaly of the tricuspid valve by simultaneous recording of intracavitary electrocardiograms and pressures across the tricuspid valve is a well-established technique [2]; such a method to diagnose Ebstein's anomaly of the left AV valve in patients with CCTGA has also been utilized [50]. Simultaneous intracavitary electrocardiographic and pressure recordings across the left atrioventricular valve in a 13-month-old infant with angiographically confirmed CCTGA and left AV valve insufficiency is illustrated in Figure 20 [50]. These recordings were similar to those obtained in classic Ebstein's anomaly of the tricuspid valve in a normally positioned (non-reversed) RV

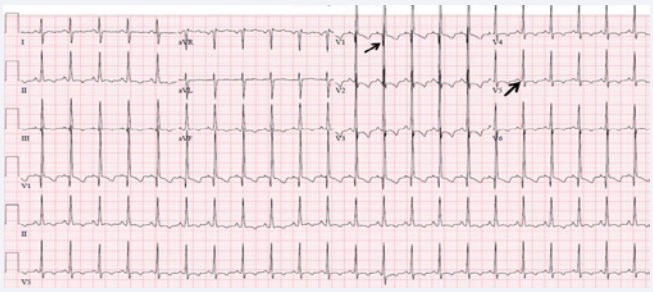

Figure 17 An ECG of a patient, demonstrating Q waves in the right chest leads and no Q waves in the left chest leads. This is highly suggestive of ventricular inversion, although such a pattern may also be seen with severe right ventricular hypertrophy. The P wave vector is normal, suggestive of situs solitus. Reproduced from Reference [51].

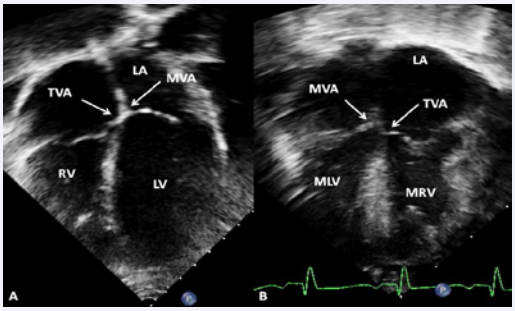

Figure 18 Selected video frames from apical four-chamber echocardiographic views of two infants. A. A normal ventricular relationship: note the higher level of attachment of the mitral valve (MVA) compared to the tricuspid valve attachment (TVA). B. Ventricular inversion: note the higher level of attachment of the mitral valve (MVA) on the right compared to the tricuspid valve attachment (TVA) on the left, indicating that the ventricles are inverted. LA, left atrium; LV, left ventricle; MLV, morphologic left ventricle; MRV, morphologic right ventricle; RA, right atrium; RV, right ventricle. Reproduced from reference [51].

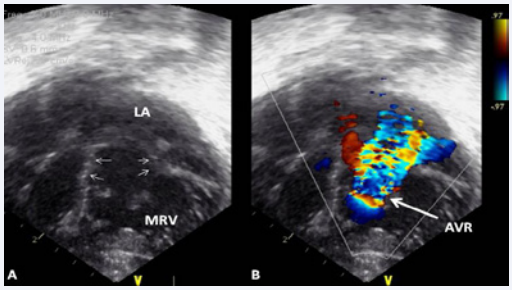

Figure 19 A. A selected modified apical view demonstrating the downward displacement of a left-sided morphologic tricuspid valve attachment (small arrows) in a patient with l-TGA. B. Color flow image of the same patient, showing severe atrioventricular valve regurgitation (AVR). LA, left atrium; MRV, morphologic right ventricle. Reproduced from reference [51].

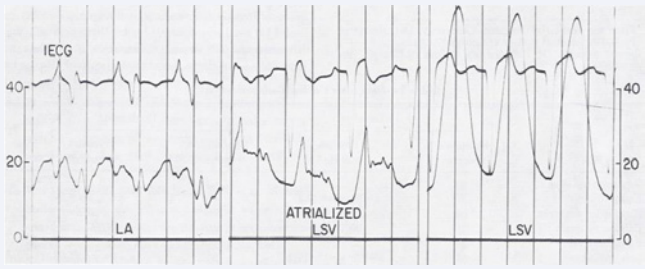

Figure 20 Simultaneous recording of intra-cardiac electrocardiogram (IECG) and pressures as the electrode/pressure recording catheter is slowly withdrawn from the left atrium (LA) to the left sided, systemic (LSV), morphologic right ventricle. Left panel shows atrial pressure curve with atrial electrogram. Middle panel shows atrial pressure curve with ventricular electrogram when the tip of the catheter is in the atrialized ventricular chamber. Right panel shows ventricular pressure curve with ventricular electrogram when the tip of the catheter is in the ventricular chamber. Pressure is marked in mmHg. The pressure in LSV is damped because of small size (lumen) of the catheter. Reproduced from Rogers JH, Jr, Rao PS. Chest 1977; 72:253-6 [50].

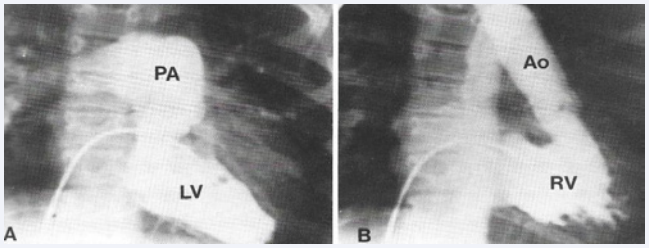

Figure 21 Selected ventricular cineangiographic frames in a patient with congenital corrected transposition of the great arteries. A. A selected frame from a right-sided ventricular cine-angiogram, demonstrating a smooth-walled morphologic left ventricle (LV) on the right side with opacification of the pulmonary artery (PA). Note that the pulmonary valve is located posterior (in the lateral view- not shown) and inferior to and rightwards from the normal position. B. A selected frame from a left-sided ventricular cine-angiogram via a catheter advanced from the right atrium to the left atrium via a patent foramen ovale and then into ventricle (not labeled), demonstrating a coarsely trabeculated morphologic right ventricle (RV) on the left side, with opacification of the aorta (Ao). Note that the aortic valve is located anterior (in the lateral view – not shown) and superior to and leftwards from the normal position. Reproduced from reference [2].

(Figure 10). Some typical angiographic pictures demonstrating the smooth-walled morphologic left ventricle on the right side (Figures 21A, 22a, 23) [46,50] and trabeculated morphologic right ventricle on the left side (Figures 21B, 22b, 24) [46,50] are shown. The degree of left AV valve insufficiency may be semi-quantified by the degree left atrial opacification (Figure 22b). Angiographic definition of ventricular morphology may also be demonstrated in dextrocardia patients (Figure 25) [51,52].

MANAGEMENT

Issues related to the management of associated cardiac defects, namely, VSD, VSD with PS and complete heart block will not be addressed in this paper dealing with left AV valve insufficiency in CCTGA patients. In patients with significant insufficiency of the morphologic tricuspid (an Ebstein's-like valve), afterload-reducing agents (Captopril/Enalapril) may initially be used to palliate the disease. If that is not able to control the CHF, surgical repair or valve replacement may become necessary [2,50,51]. Because of relatively poor long-term results, particularly related to leaving the morphologic RV to pump against the systemic circuit, a double switch operation was developed [53]. Discussion of the issues related to leaving the morphologic RV to pump against the systemic circuit and the relative advantages of simpler corrective procedures vs. the double switch operation [53,54] are beyond the scope of this paper.

SUMMARY

In this review, two uncommonly encountered congenital

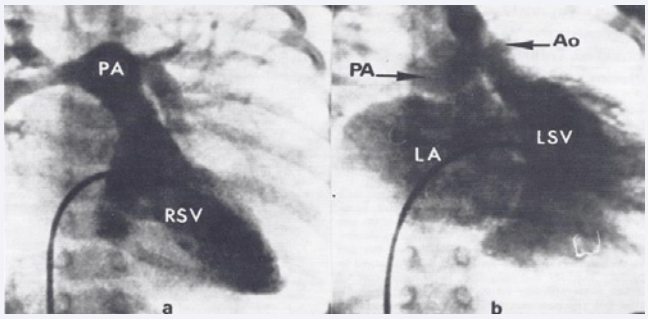

Figure 22 a. A selected frame from a right-sided ventricular (RSV) cine-angiogram, demonstrating a smooth-walled morphologic left ventricle on the right side with opacification of the pulmonary artery (PA). b. A selected frame from a left-sided ventricular (LSV) cine-angiogram, demonstrating a coarsely trabeculated morphologic right ventricle on the left side, with opacification of the aorta (Ao). Note the significant left-sided atrioventricular valve insufficiency, resulting in the opacification of the left atrium (LA). The PA is also opacified because of a left-to-right shunt via a ventricular septal defect (not marked). Reproduced from Rogers JH, Jr, Rao PS, Chest 1977; 72:253-6 [50].

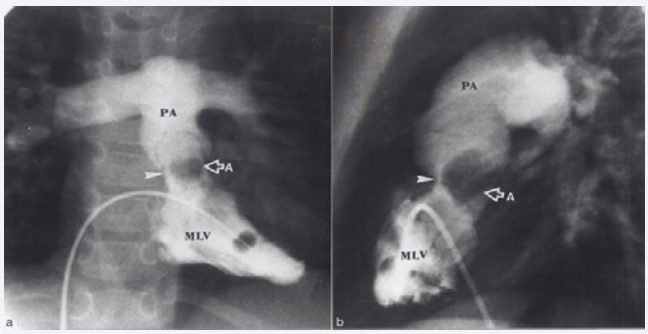

Figure 23 Selected frames from a morphologic left ventricular (MLV) angiogram in postero-anterior (a) and lateral (b) projections, demonstrating a finely trabeculated ventricle with opacification of the pulmonary artery (PA). Note that the pulmonary valve is located posterior and inferior to and rightwards from the normal position. The open arrow in 'a' points to a radiolucent aneurysm (A) in the sub-pulmonary region. The solid arrow in 'b' points to a narrow jet of contrast material going from the MLV to the PA, suggesting severe sub-pulmonary obstruction. Reproduced from Reddy SCB, Chopra PS, Rao PS, Am Heart J 1997; 133:112-119 [46].

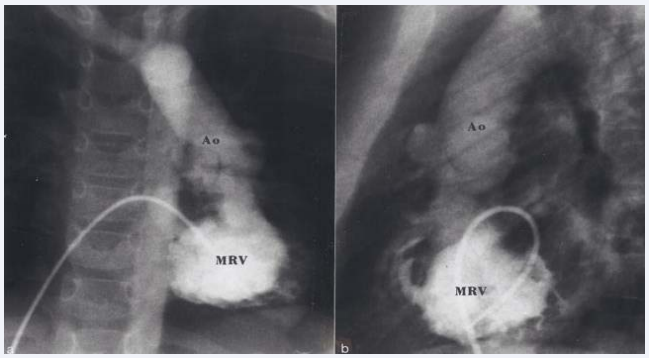

Figure 24 Selected frames from a morphologic right ventricular (MRV) angiogram in postero-anterior (a) and ateral (b) projections, demonstrating a coarsely trabeculated ventricle with opacification of the aorta (Ao). Note that the aortic valve is located anterior and superior to and leftwards from the normal position. Reproduced from Reddy SCB, Chopra PS, Rao PS, Am Heart J 1997; 133:112-119 [46].

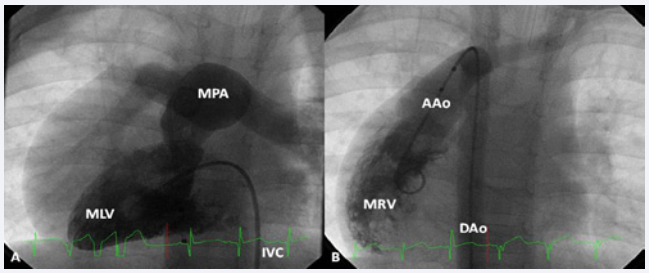

Figure 25 A. A selected frame from a morphologic left ventricular (MLV) cine-angiogram in a postero-anterior projection in a patient with dextrocardia, showing a finely trabeculated left-sided MLV which gives rise to a dilated main pulmonary artery (MPA). The inferior vena cava (IVC) which is connected to the left-sided right atrium (not shown) is on the left of the spine. B. A selected frame from a morphologic right ventricular (MRV) cine-angiogram from a postero-anterior projection in the same dextrocardia patient as is shown in A, demonstrating a coarsely trabeculated right-sided MRV which gives rise to the aorta (AAo). The descending aorta (DAo) is on the right of the spine. The aortic valve is anterior (in the lateral view; not shown) and superior to and rightwards from the pulmonary valve. This would indicate a d-loop of the ventricle and d-transposition of the great arteries in a patient with dextrocardia, a situation indicative of corrected transposition physiology. Reproduced from Yarrabolu TR, Thapar MK, Rao PS, Congenital Cardiology Today 2014; 12(7):1-8 [52].

malformation of the tricuspid valve, namely, Ebstein's anomaly of the tricuspid valve and Ebstein's type of abnormality of the morphologic tricuspid valve on the left-side in patients with CCTGA were discussed. Ebstein's anomaly of the tricuspid valve is a rare congenital heart defect accounting for 0.3 to 0.6% of all CHDs. This defect is characterized by a downward displacement of the insertion of the septal and posterior leaflets of the tricuspid valve with the resultant tricuspid regurgitation and right atrial enlargement of varying degrees. The diagnosis is relatively simple and can often be made on clinical features and simple laboratory studies such as chest roentgenogram and ECG and confirmed by echocardiography. Mild forms may be asymptomatic and may not need any treatment. Moderate forms may be managed with relative ease, and may improve as the PVR decreases with increasing age. Severe forms of the disease are a challenge to manage and may require surgical intervention. The prognosis depends on the severity of the lesion, the patient's age at presentation, and the type of surgical repair. Surgical outcomes have improved over time, but early presentation as a fetus and newborn is associated with a poor prognosis. CCTGA is a rare complex CHD and constitutes less than 0.5% of all CHDs. In this condition, there is atrial situs solitus, atrio-ventricular discordance, left to right reversal of the ventricles and ventriculo-arterial discordance with resultant normal blood circulatory pattern. While rare patients may not have other defects, most patients have associated defects and this paper addresses Ebstein's type of abnormality of the morphologic tricuspid valve on the left-side. This anomaly can easily be diagnosed by echo-Doppler studies. Afterload reducing agents and if those are not effective, surgical repair or replacement of the left AV valve may become necessary.

CONCLUSION

It was concluded that Ebstein's anomaly of the tricuspid valve and Ebstein's type of abnormality of the morphologic tricuspid valve on the left-side in patients with CCTGA can be effectively diagnosed with the currently available non-invasive and invasive investigative techniques and the defect can successfully be managed with the existing therapeutic medical and surgical methods.

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