

REVIEW

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Echocardiographic assessment of transposition of the great arteries and congenitally corrected transposition of the great arteries

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Abstract

Echocardiographic assessment of patients with transposition of the great arteries and congenitally corrected transposition requires awareness of the morphology and commonly associated lesions. The pre-operative echocardiography should include a full segmental and sequential analysis. Post-operative assessment is not possible without awareness of the type of surgical procedure performed and consists of assessing surgical connections and residual lesions.

Key Words

- ▶ congenital heart defects
- ▶ congenitally corrected transposition of great arteries
- ▶ transposition of the great arteries

Introduction

Echocardiographic assessment of patients with transposition of the great arteries (TGA) and congenitally corrected transposition of the great arteries (CCTGA) requires a good understanding of the morphology of both defects with their commonly associated lesions. Insight into the physiology and the impact of different surgical treatment options are crucial when evaluating these patients pre-operatively and postoperatively.

Transposition of the great arteries

Transposition of the great arteries (TGA) is a conotruncal anomaly where the ventriculo-arterial connections are discordant; the aorta aligns with the right ventricle (RV) and the pulmonary artery aligns with the left

ventricle (LV) (Figs 1 and 2). When there are concordant atrioventricular connections, TGA results in severe cyanosis because the circulation is in parallel rather than in series; the deoxygenated blood that returns from the systemic veins goes directly out the aorta and the oxygenated blood that returns from the pulmonary veins goes directly out of the pulmonary artery. Approximately 40% of children with TGA have an associated ventricular septal defect (VSD) that can be variable in size (1). The overwhelming majority of children with TGA present with cyanosis immediately after birth, particularly if the ventricular septum is intact. Those children with TGA in association with a large VSD may present somewhat later in infancy. TGA is the second most common cyanotic lesion after tetralogy of Fallot and affects males more than females.

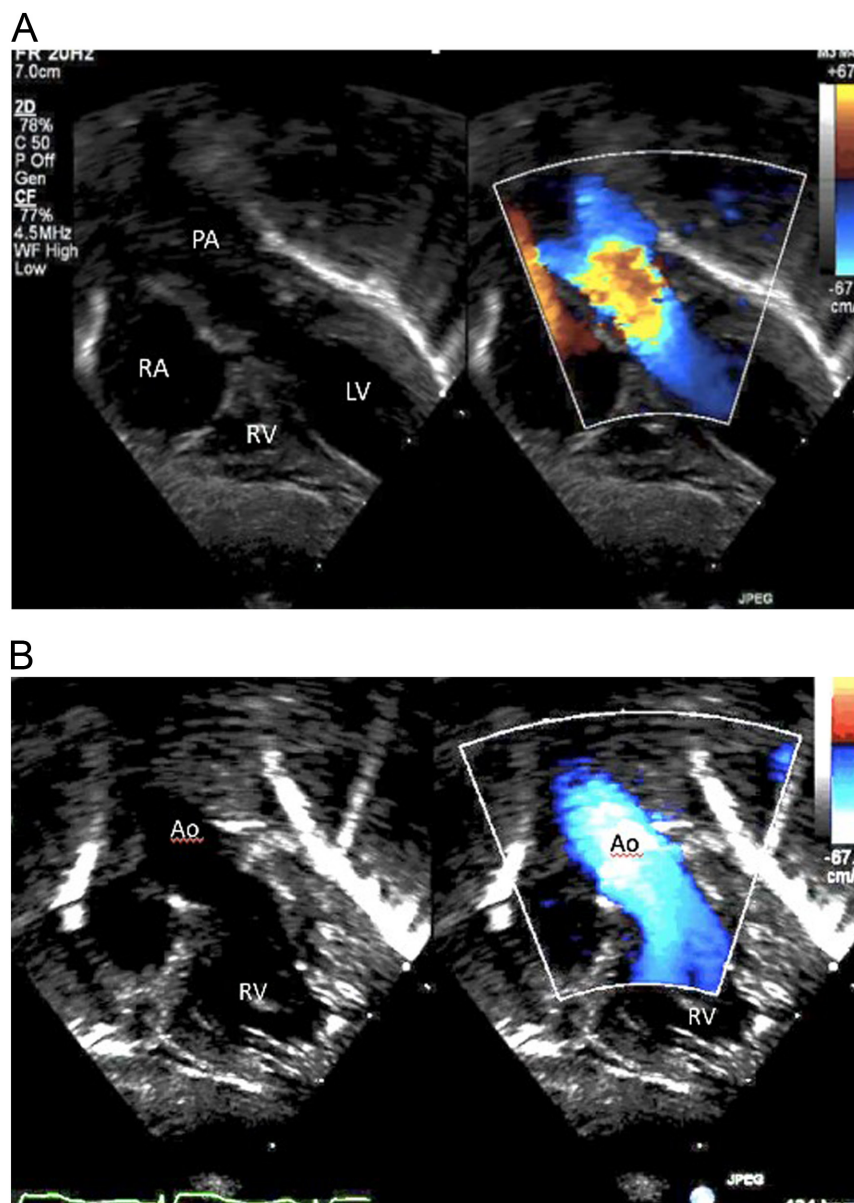


Figure 1
TGA: Discordant ventriculo-arterial connections. Subxiphoid coronal views in patient with TGA. In (A) the great vessel aligned with the left ventricle (LV) is the pulmonary artery (PA) and is seen bifurcating. In (B) the aorta (Ao) is seen aligned with the right ventricle (RV). This shows the discordant ventriculo-arterial connections.

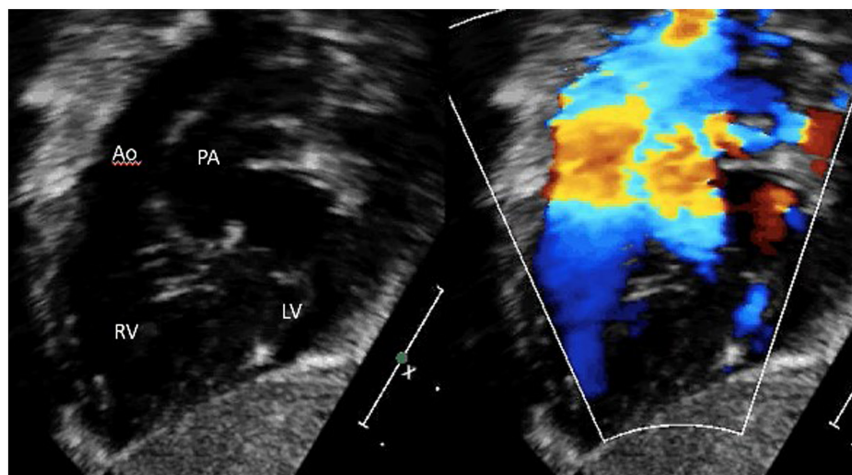
The etiology of TGA remains unknown and in isolation it is rarely associated with a syndrome or a known genetic disorder. Maternal diabetes has been reported as a risk factor for fetal TGA (2). There is clearly a developmental abnormality such that the conotruncus does not go through its normal rotation and results in the great arteries aligned abnormally.

Important morphometric features of TGA include the relationship of the aorta and the pulmonary artery. In contrast to the normal heart, the great arteries in TGA arise in parallel rather than spiral around each other (Fig. 2). Most typically, the aorta sits anterior and rightward of the pulmonary artery; however, the great arteries may be side by side or directly anterior/posterior to each other (Fig. 3).

Very rarely, the aorta may sit posterior to the pulmonary artery. The conal anatomy can also vary in TGA. Conus is the circumferential muscle under a semilunar valve that tends to be associated with the anterior great artery. Conus can be present under either great artery, both or neither. In the normal heart, there is subpulmonary conus only. The majority of children with TGA have subaortic conus only (thus the aorta tends to be anterior), however, any conal anatomy is possible including bilateral conus (under both great arteries) (3).

Another important feature is LV outflow tract obstruction which can occur in 10% of patients with TGA (4). There are multiple possible causes including ventricular septal thickening, fibrous membranes, aneurysm of the

A



B

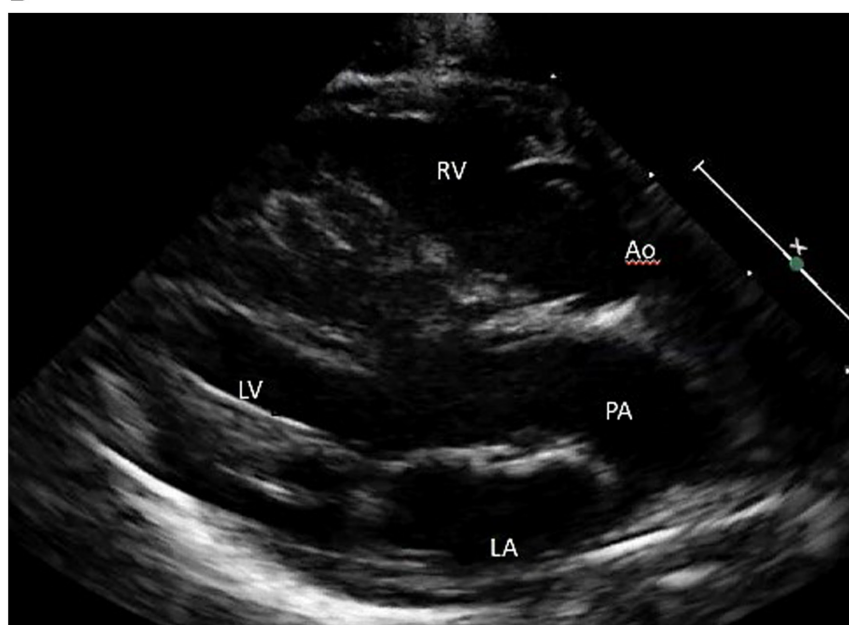


Figure 2

Parallel great arteries. (A) Subxiphoid short-axis view demonstrates the parallel orientation of the great arteries with the aorta (Ao) anterior to the pulmonary artery (PA). (B) Parasternal long axis view demonstrating the parallel great arteries.

membranous septum, conal septum malalignment and valvar pulmonary stenosis. RV outflow tract obstruction is more unusual except in those patients with an anterior malalignment VSD (discussed below). In those cases, more distal obstruction such as coarctation of the aorta or interruption of the aortic arch are also possible.

The coronary artery origins by definition are in an abnormal position in TGA because the aorta is anterior to the pulmonary artery. There is significant variability in coronary artery anatomy which is important to assess in the pre-operative echocardiogram (Fig. 6). Coronary artery variants such as circumflex artery originating from the right coronary artery are more common when the great arteries are more side by side. One of the most

concerning anatomic variations is an intramural course of the proximal coronary artery. This occurs when a coronary artery arises from the opposite sinus and courses between the great arteries typically in the wall of the aorta. In these cases, translocation of the coronary arteries (during the arterial switch operation) is more complicated and may result in kinking or narrowing of the vessel. In some cases, patients have only a single coronary artery or a coronary artery that has an abnormal course posterior to the pulmonary artery or anterior to the aorta. Studies report that coronary artery variants predict poor outcome after arterial switch operation (5, 6).

The surgical treatment for TGA is an arterial switch operation. In this procedure, the great arteries become

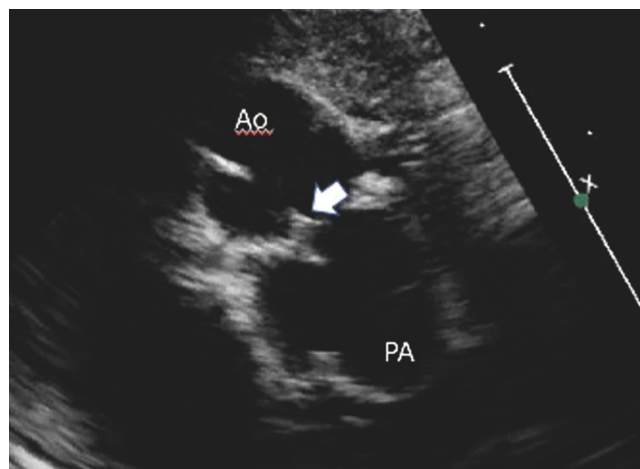


Figure 3
Relationship between great arteries. Parasternal short-axis view demonstrating the aorta (Ao) is positional anterior and rightward relative to the pulmonary artery (PA). The arrow demonstrates the alignment between the commissures of both semilunar valves.

surgically aligned with the appropriate ventricle (pulmonary artery to RV, aorta to LV) by way of transection and re-anastomosis of the great arteries. In addition, the coronary arteries are translocated to the new 'neo' aorta (7). During the arterial switch operation, the Lecompte maneuver is often performed; this procedure tucks the aorta behind both branch pulmonary arteries to prevent obstruction (8). Prior to the 1980s, attempts at the arterial switch operation had been unsuccessful most typically because of the challenges of moving the coronary arteries and properly aligning the branch pulmonary arteries.

Before the arterial switch operation was a feasible strategy, the atrial switch procedure was utilized starting as early as the late 1950s. Drs Senning and Mustard devised methods to 'switch' the atria rather than the great arteries such that systemic and pulmonary venous blood was baffled to the contralateral ventricle to allow for a circulation in series rather than parallel (9, 10). This procedure resulted in the RV and tricuspid valve functioning in the systemic position.

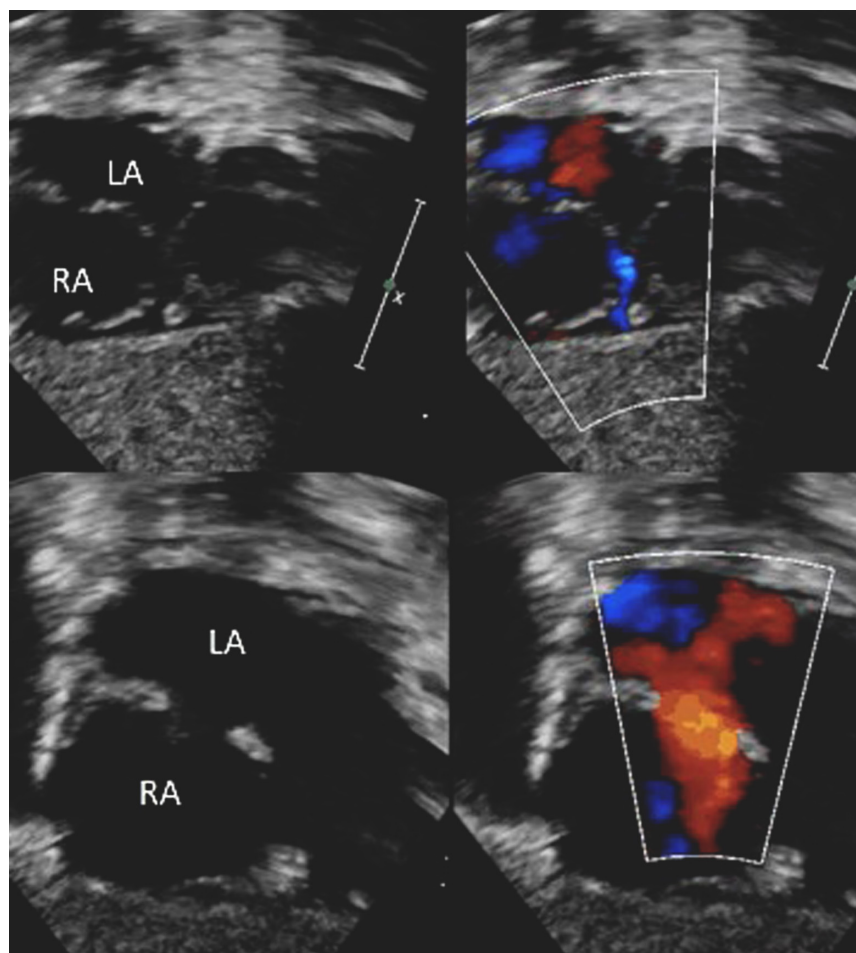


Figure 4
Role of balloon atrial septostomy. Newborn with TGA born with very restrictive ASD with very limited L-R shunting shown in subxiphoid long-axis view. Preductal saturation in this patient was 40%. Balloon atrial septostomy was urgently performed with creation of good sized unrestrictive atrial communication with preductal saturation improving to 85%. LA, left atrium; RA right atrium.

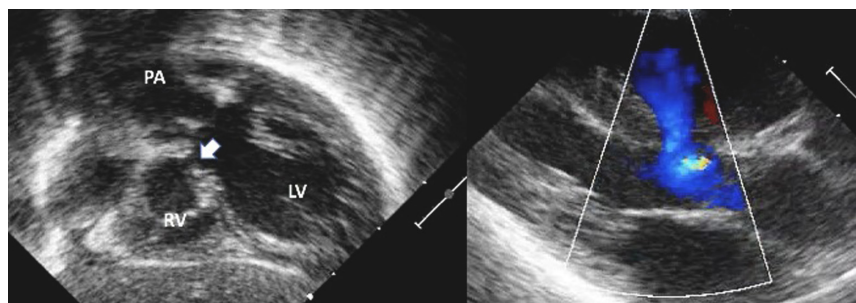


Figure 5

TGA with perimembranous VSD. In the subxiphoid anterior oblique view the arrow indicates the location of the VSD in the perimembranous part of the septum. There is fibrous continuity between the tricuspid valve and the pulmonary valve. The image on the right shows a tilted parasternal long-axis view with shunting from the RV to the LV through the VSD.

TGA with intact ventricular septum

In TGA without a VSD, cyanotic presentation at birth typically leads to immediate recognition in the delivery room or newborn nursery. Because the circulations are in parallel, mixing of deoxygenated and oxygenated blood must occur for survival. Generally, an arterial duct is inadequate as the only source of mixing, and thus an adequate atrial communication is essential. In those with an intact or nearly intact atrial septum, extreme cyanosis may occur (Fig. 4). In these cases, an urgent intervention is required, namely a balloon atrial septostomy. This procedure, first described by Dr William Rashkind in 1966, utilizes a balloon catheter to tear a hole in the septum primum portion of the atrial septum (11). This procedure can be life-saving for infants with TGA.

Transposition with associated ventricular septal defect

The anatomy of TGA in association with a VSD is quite variable. As with patients that have normally related great arteries, any anatomic type of VSD may be present in association with TGA. The nomenclature of VSDs was recently reported by the International Society for Nomenclature of Paediatric and Congenital Heart Disease (12). Those with a simple type of VSD, typically have a perimembranous central or muscular defect (Fig. 5).

If the VSD is small, it may be left alone during the arterial switch operation. If large, it is typically repaired at the time of the procedure.

There are several more complex types of VSDs that may be present. An outlet (doubly committed) VSD is an unusual defect that occurs when there is absence of the conal (outlet) septum such that the aortic and pulmonary valves are in fibrous continuity (12). In these cases, closure of the VSD may result in distortion of the semilunar valve leaflets and subsequent semilunar valve regurgitation. Malalignment type VSDs are outlet defects where the conal (outlet) septum is out of its normal alignment with the muscular ventricular septum (12). These type of VSDs are more common than the outlet defects without malalignment and result in outflow tract obstruction. Posterior malalignment of the conal septum with resultant VSD is typically associated with subpulmonary outflow tract obstruction. In TGA with significant posterior malalignment type VSD, an arterial switch procedure cannot be performed because the aortic outflow would be obstructed after surgery. In these cases, the Rastelli operation is typically performed, whereby the VSD is baffled through the VSD to the aorta and a pathway (typically a conduit) is made between the RV and the pulmonary artery. Another option for this anatomic substrate is the Nikaidoh operation whereby the aorta is translocated closer to the LV (through the pulmonary annulus) in order to avoid LV

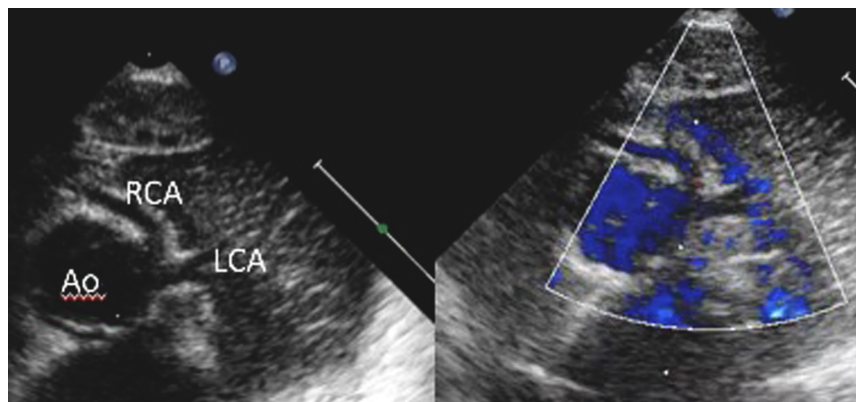


Figure 6

Patient with TGA and single coronary artery. In this high parasternal view, a single coronary artery that comes from the left sinus (sinus 1) is seen trifurcating into the anteriorly looped right coronary artery (RCA) and left coronary artery (LCA) which immediately bifurcates into the left anterior descending and circumflex arteries.

outflow tract obstruction postoperatively. Anterior and/or rightward malalignment of the conal septum typically results in RV outflow tract obstruction. Since the aorta arises from the RV, the annulus of the aortic valve may be small and distal obstruction (coarctation of the aorta or interruption of the aortic arch) may be present. In these cases, a more complex arterial switch operation is required that often includes VSD closure, aortic arch repair and in some cases, enlargement of the RV outflow tract (similar to what is performed in tetralogy of Fallot repair).

Inlet (canal) type VSDs are unusual in TGA but can be seen most typically in patients with isomerism (heterotaxy syndrome). They are typically large defects that require repair at the time of arterial switch operation. They can also be seen in association with straddling tricuspid valve which may preclude a two ventricle repair (13).

Echocardiographic assessment of preoperative TGA

- Assess the position of the great arteries from subxiphoid frontal and sagittal views or from parasternal long-axis view (Figs 1 and 2).
- If cyanosis is present, examine the presence and size of the atrial septal defect in subxiphoid views to determine if balloon atrial septostomy is required (Fig. 4).
- Identify presence, type, number and size of VSD using multiple views (Fig. 5).
- Identify presence and type of LV outflow tract obstruction using subxiphoid, apical five-chamber and parasternal long-axis views.
- If malalignment of the conal (infundibular) septum is present, assess severity of outflow tract obstruction and more distal obstruction in multiple views.
- Assess aortic arch in cases of anterior/rightward malalignment of the conal septum in suprasternal sagittal view.
- Assess in multiple views for coronary artery anatomy including:
 - Assessment for coronary arteries passing posterior to the pulmonary artery (circumflex artery from right coronary artery).
 - Assessment for coronary arteries passing anterior to the aorta (right coronary artery arising from left facing coronary cusp) (Fig. 6).
 - Assessment for coronary arteries passing between the great arteries (intramural course).

Arterial switch operation

The outcome for the arterial switch operation is generally excellent in the modern era (14). However, there are

some issues that must be assessed over the short term and long term after this procedure. Ideally, this procedure is performed in the first few days of life to obviate the cyanosis. The other reason to perform the operation early in the neonatal period is the high pulmonary blood flow that occurs (despite the cyanosis). Over a relatively short period of time, these patients can develop pulmonary vascular disease (15). Furthermore with decrease in pulmonary vascular resistance, the LV becomes deconditioned and may fail after the switch procedure when exposed to the much higher systemic vascular resistance. In patients with a large VSD the surgery can be postponed for a few weeks or months based on surgical preference. Recent data however suggested that postponing surgery beyond 2 weeks of life may affect neurodevelopmental outcomes (16).

There are several short-term and long-term concerns after the arterial switch operation. The most concerning issue immediately after surgery is whether the translocation of the coronary arteries is successful (17). Coronary artery problems typically present as ventricular dysfunction, difficulty with separation from cardiopulmonary bypass and ventricular tachyarrhythmias. This can be assessed in the operating room by transesophageal echocardiography or by epicardial imaging (18). Other short-term concerns after the arterial switch include narrowing at the anastomotic sites above the neopulmonary valve and neo-aortic valves (14) (Fig. 7). Finally, the Lecompte maneuver is usually performed with the arterial switch operation to assure that the branch pulmonary arteries lie without obstruction (Fig. 8). The maneuver entails tucking the aorta behind both pulmonary arteries. Branch pulmonary artery narrowing is a potential complication of this procedure typically because of stretching or kinking of the proximal pulmonary artery branches (19).

In the long term, other concerns may emerge. The coronary arteries may become narrowed over time or in rare cases may become atretic (20, 21). Coronary artery problems are more common with more unusual pre-operative coronary artery patterns (6, 20, 21). Coronary artery imaging may be needed by CT, MRI or angiography. Coronary artery problems may present with regional wall motion abnormalities of the ventricle. The neo-aortic root often becomes enlarged over time after the procedure. Though many risk factors for root dilation have been reported, it is likely a consequence of the procedure and the suture line that is required for the anastomosis (22, 23). In rare cases, aortic root replacement has been reported (24). Neo-aortic valve regurgitation is also common but does not usually require aortic valve surgery (22, 23).

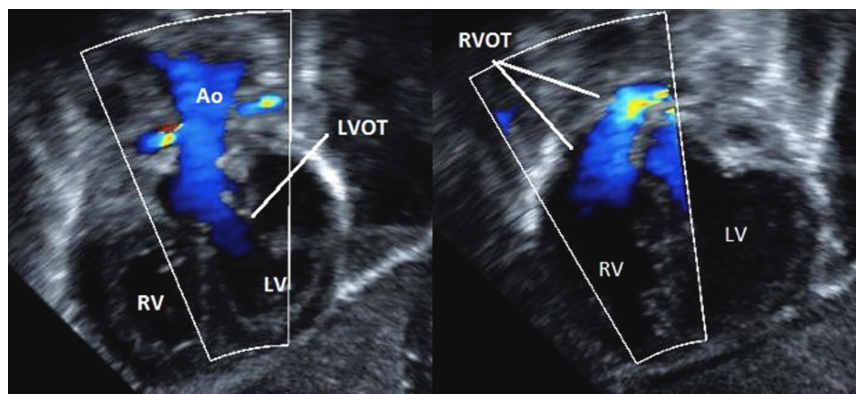


Figure 7

Position of the outflow tracts after the arterial switch operation. Subxiphoid left anterior oblique view demonstrates the parallel orientation of the outflow tracts after the arterial switch procedure with the aorta coming off the LV and the pulmonary artery from the RV. Note the acceleration of flow distal to the pulmonary valve from mild supra-valvar pulmonary narrowing. Also note the branch pulmonary arteries seen on either side of the aorta (Ao).

Echocardiographic assessment of arterial switch operation

- Assess the outflow tracts for supra-valvar narrowing and neo-valvar regurgitation from subxiphoid, apical and parasternal views (Fig. 7).
- Assess for regional wall motion abnormalities from apical and parasternal views.
- Try to identify color flow in the translocated coronary arteries in parasternal short-axis view with very low color scale.
- Assessment of the branch pulmonary arteries with Doppler interrogation using the high parasternal view (Fig. 8).
- In longer term assessment, measure the neo-aortic root in the parasternal long axis view and assess for neo-aortic and neopulmonary valve regurgitation in multiple views.

Especially in adults after the arterial switch operation, echocardiographic images may be limited and additional cross-sectional imaging by CT scan or MRI may be

required to image the pulmonary artery branches and coronary arteries.

Atrial switch operation

Most patients who have undergone the atrial switch operation are now adults as the arterial switch operation has replaced this procedure in almost all cases. In the current era, the atrial switch operation is reserved for patients with a late presentation of TGA in whom the LV pressure and mass are inadequate to function in the systemic circulation. The concerns for the atrial switch operation are completely different than the concerns regarding the arterial switch operation.

The most common complications of the atrial switch operation include systemic venous baffle obstruction, pulmonary venous baffle obstruction and baffle leak (25, 26, 27, 28). The superior vena cava portion of the systemic venous pathway is particularly vulnerable to narrowing because it is a smaller structure and because some of these patients have transvenous pacing wires (25).

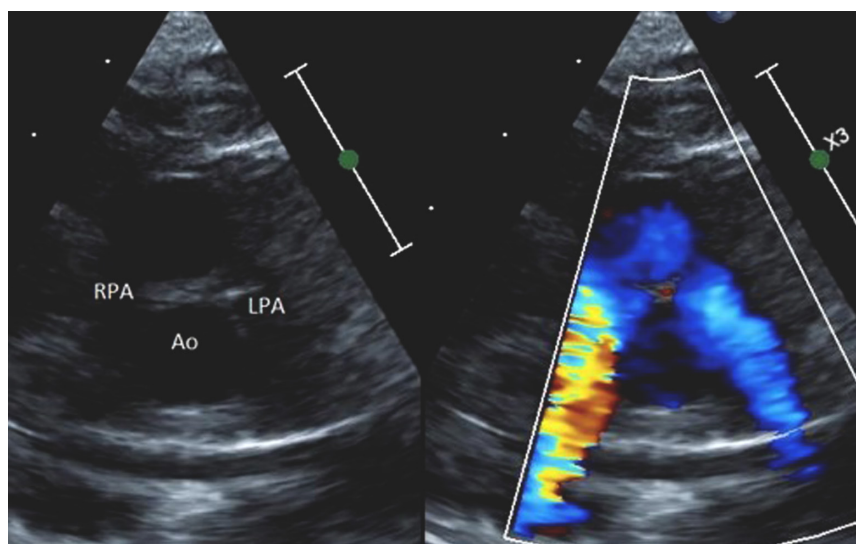


Figure 8

Position of the branch pulmonary arteries after the Lecompte maneuver. In this high parasternal view, the pulmonary artery bifurcation is positioned anterior to the aorta with both pulmonary arteries draped over the ascending aorta.

Pulmonary venous baffle obstruction is less common and typically occurs immediately after surgery (26). Elevated left ventricular pressure estimate (the pulmonary ventricle) is often a sign of progressive pulmonary venous baffle obstruction. Baffle leaks can occur anywhere between the two atrial chambers but most typically occurs near the inferior vena cava. From a physiologic standpoint, these leaks act as atrial septal defects with a left-to-right shunt and can result in LV dilation (28). They are usually amenable to device closure in the cardiac catheterization laboratory. Transesophageal echocardiography may be required to assess for complications of the atrial switch operation because of poor acoustic windows in adulthood.

Though not a complication of the surgery itself, many patients who undergo the atrial switch operation develop RV dysfunction and tricuspid valve regurgitation because these structures are in the systemic position (29). Atrial arrhythmias are also common and often require pacemakers. Assessment of RV systolic function is challenging but methods like measurement of fractional area change, tissue Doppler velocities, and strain imaging can be used in longitudinal follow-up. Progressive tricuspid regurgitation occurs in the context of RV failure. Finally more recently late occurrence of pulmonary arterial hypertension has been recognized as an important long-term problem in patients after the atrial switch (30). Monitoring LV systolic pressures is essential part of the follow-up.

Echocardiographic assessment of atrial switch operation

- Assessment of the systemic and pulmonary venous pathways for obstruction using subxiphoid, apical and parasternal views (Fig. 9).
- Color assessment or contrast injection to assess for baffle leaks.
- Spectral Doppler assessment of mitral regurgitation to estimate pulmonary artery pressure using modified Bernoulli equation.
- Assessment of mechanism and severity of tricuspid regurgitation using apical and parasternal views as well as 3D imaging.
- Assessment of RV performance using various techniques including tissue Doppler velocities, fractional area change, speckle tracking imaging to measure RV longitudinal strain.

In case of poor imaging windows, cardiac MRI or CT may be required to image the pathways and monitoring right ventricular function.

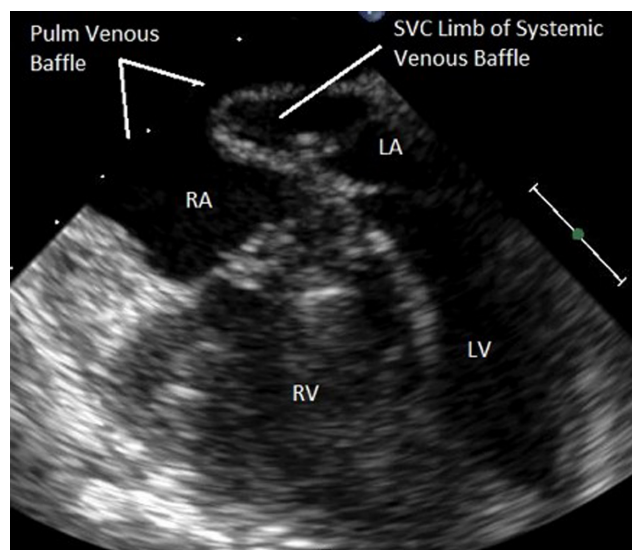


Figure 9
Venous pathways after the atrial switch procedure. Apical four-chamber view from transesophageal echocardiogram demonstrates the pulmonary venous baffle connecting the pulmonary venous return to the RV and the superior vena cava (SVC) limb of the systemic venous return baffle connecting to the LV.

Congenitally corrected transposition of the great arteries

CCTGA is characterized by the presence of discordant connections at the atrioventricular and ventriculo-arterial junctions (31). In the presence of usual atrial arrangement (situs solitus), this implies that the right-sided morphologically right atrium is connected to the right-sided morphologically LV, while the left-sided morphologically left atrium aligns with the left-sided morphologically RV (Fig. 10). The LV gives rise to the pulmonary trunk and the RV empties in the aorta. Typically the aorta is positioned anterior and to the left of the pulmonary trunk. In about 10% of patients with CCTGA there is mirror-image atrial arrangement (situs inversus) resulting in a right-sided left atrium connecting to a right-sided RV that empties in the aorta, while a left-sided right atrium connects to a left-sided LV aligned with the pulmonary trunk. In this configuration the aorta is rightward and anterior to the pulmonary trunk. In CCTGA and similar to TGA, the outflow tracts are parallel to each other. Often there is an abnormality of the position of the interventricular septum which results in the ventricles aligning side-by-side with an added superior-inferior rotation. The coronary arteries typically follow the ventricular morphology. Because of malalignment between the atrial septum and the inlet ventricular septum related to the wedged position of the

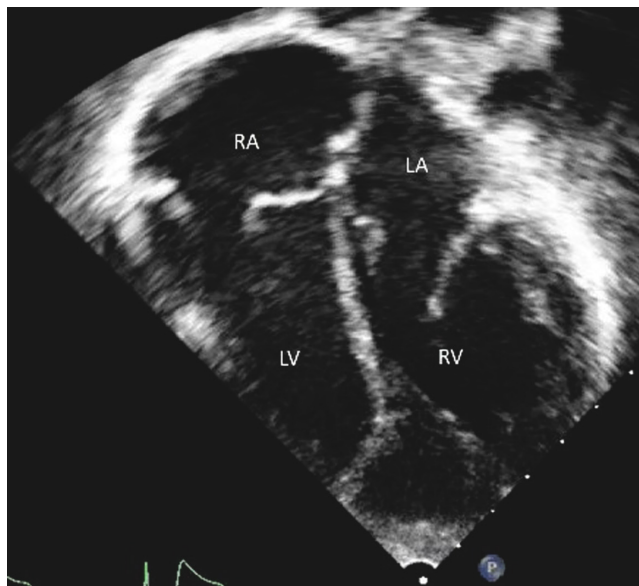


Figure 10

CCTGA. In this apical four-chamber view, the atrioventricular connections are demonstrated with the right atrium (RA) connected to the LV and the left atrium (LA) to the RV. The mildly lower insertion of the septal leaflet of the tricuspid valve can be appreciated.

pulmonary outflow tract, the conduction tissue typically runs abnormally which is important when performing surgery or interventions as there is increased risk for complete heart block, which can also occur spontaneously in the follow-up of patients with CCTGA.

CCTGA can occur as isolated lesion but most patients (>90%) have associated congenital defects. VSDs occur in more than half of the patients and are usually perimembranous central. This type of VSD is characterized by fibrous continuity between the pulmonary and the tricuspid valves. All other types of VSD have been described in patients with CCTGA (muscular, inlet, doubly committed, malalignment). LV outflow tract obstruction also occurs in about 50% of patients with usual atrial arrangement and can be subvalvar or valvar, most commonly combined. Subvalvar stenosis can be muscular or fibrous, or can be caused by the subvalvar mitral valve apparatus. Tricuspid valve dysplasia is a very common feature of CCTGA with apical displacement of the septal and inferior leaflets resembling Ebstein malformation seen in normally connected hearts. In contrast to Ebstein, however, the leaflet displacement does not result in a rotational abnormality of the inflow and is not associated with myocardial RV thinning. The tricuspid valve leaflets can be dysplastic adding to variable degree of tricuspid regurgitation. Often this is progressive as RV dilatation caused by

tricuspid regurgitation will cause further tethering of the septal leaflet and due to septal displacement this results in more severe regurgitation of the valve. Severe tricuspid regurgitation adds volume loading to a pressure loaded RV and is associated with progressive RV dilation and dysfunction. Mitral valve anomalies like the presence of an isolated mitral valve cleft or parachute mitral valve can also be associated with CCTGA.

Echocardiographic assessment of pre-operative CCTGA

The initial echocardiographic evaluation of a patient with CCTGA requires a full segmental echocardiographic evaluation. Dynamic 'sweeps' from different views, especially from subxiphoid windows, are extremely helpful for describing the connections and the associated lesions.

Important components of the echocardiographic assessment are:

- Describe the atrial arrangement by determining the veno-atrial connections. This is most commonly done from subxiphoid or apical imaging (Fig. 10).
- Determine the cardiac orientation from subxiphoid imaging. This describes the position of the heart within the chest (levocardia, mesocardia, dextrocardia). Abnormal cardiac orientation is common in CCTGA.
- Determine the ventricular morphology based on typical ventricular features like offset of the tricuspid valve, moderator band and coarse trabeculations for the RV. Once atrial arrangement and ventricular morphology are determined, the atrioventricular connection can be identified as discordant. Typically different imaging planes are combined including subxiphoid and apical views.
- Determine the morphology of the great arteries. This can be based on different views, most commonly from subxiphoid and parasternal views. The great arteries are parallel. Once the great arteries are described, the ventriculo-arterial connections are established as discordant.
- Determine the relationship of the aorta and the pulmonary artery. This is most often accomplished in subxiphoid cross-sectional imaging and high parasternal views (Fig. 11).
- Assess for the presence of associated lesions including:
 - Tricuspid valve anomalies and degree of tricuspid regurgitation. Different views are combined. The apical views are the most commonly used (Fig. 12).

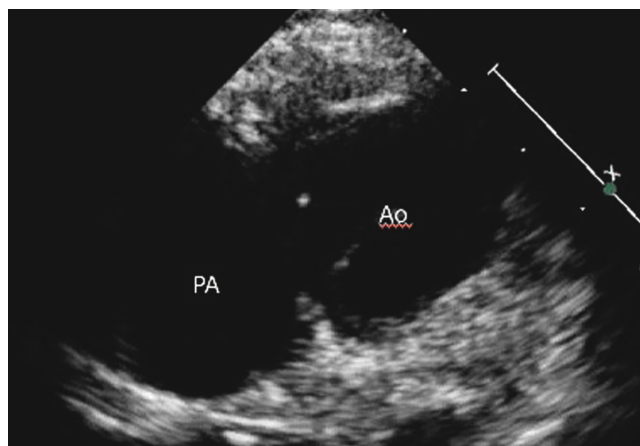


Figure 11
CCTGA relative position of great arteries. High parasternal short-axis view demonstrates the leftward and anterior position of the aorta (Ao) relative to the pulmonary artery (PA).

- The presence of a VSD: determine location, size, shunting. If inlet VSD, look for straddling of the atrioventricular valves. This combines different views typically subxiphoid and parasternal (Fig. 13).
- LV outflow tract obstruction: describe mechanism (subvalvar and valvar pulmonary stenosis) and severity (gradient). All views can be used Alignment with the outflow tract is typically determined from subxiphoid or apical views.
- Look for any other lesion like atrial septal defect, patent ductus arteriosus, arch anomalies, or left superior vena cava.
- Determine the coronary artery origins typically from parasternal short-axis view of the great arteries. This is particularly important if a double-switch operation is to be considered.

Once the morphology has been described in detail, follow-up echocardiograms will mainly focus on the assessment of tricuspid regurgitation, RV function and hemodynamic impact of associated lesions like LV outflow tract obstruction, VSD or any other shunts. Tricuspid regurgitation assessment is largely qualitative as there can be multiple jets. RV function assessment is challenging related to the complex morphology but also related to the associated lesions, particularly the presence of tricuspid regurgitation. Quantitative techniques like fractional area change, tricuspid annular excursion (TAPSE), tissue Doppler and strain imaging can be useful in serial follow-up but all these measurements are influenced by changes in volume and pressure loading. In the follow-up comparison between current and previous studies is crucial and longitudinal changes over time should be monitored. In case of poor imaging windows cardiac MRI or gated CT scan can be considered for assessment of ventricular function. Surgical management of CCTGA is highly variable and is influenced by the associated defects. Two approaches are used (32). The first approach is based on correcting physiologic abnormalities while leaving the anatomic connections unchanged. This can involve VSD closure, tricuspid valve repair or replacement, surgery on the LV outflow tract, and so forth. In recent years pulmonary artery banding has been used in patients with moderate to severe tricuspid regurgitation as higher LV pressure shifts the ventricular septum and can reduce septal leaflet tethering. In younger patients, pulmonary artery banding can ‘retrain’ the LV if functioning at lower pressures and prepare the patient for an anatomic correction (double-switch procedure). When a physiologic repair is used, the RV remains the systemic ventricle which is a concern long-term because progressive RV failure can develop.

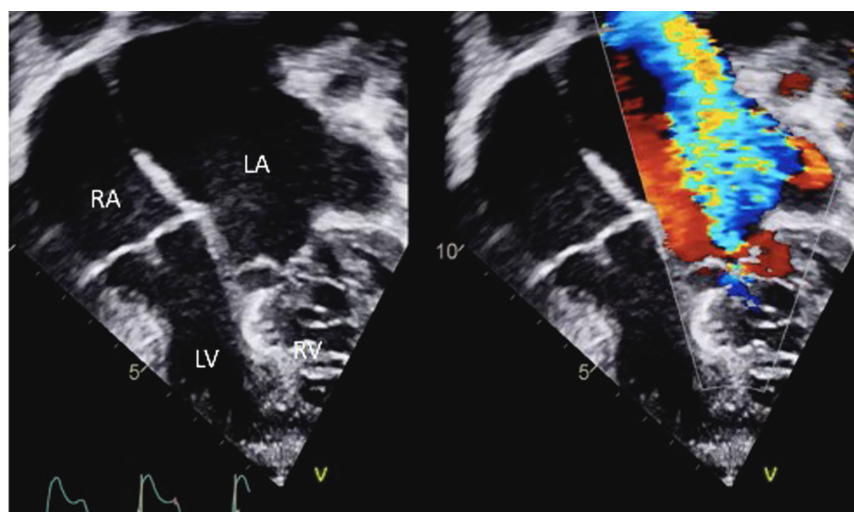


Figure 12
Ebstein-like malformation of the tricuspid valve with severe tricuspid regurgitation. In this apical four-chamber view, the severe tricuspid regurgitation can be appreciated by color Doppler. Note the more apically displaced septal leaflet of the tricuspid valve in contrast to Fig. 10.

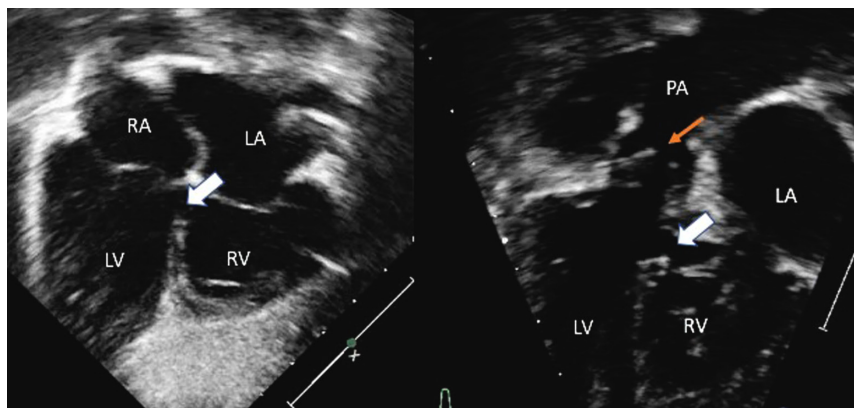


Figure 13

CCTGA with VSD and pulmonary stenosis. The subxiphoid image (left) shows that this patient has dextrocardia with the ventricular septum aligned perpendicular to the diaphragm. There is a VSD (white arrows) extending to the inlet and outlet portions of the ventricular septum. There is pulmonary valve stenosis with doming of the leaflets (orange arrow).

The second approach involves correcting the anatomic connections by rerouting the atrial connections by a Mustard or Senning procedure and reconnecting the ventriculo-arterial connections by doing an arterial switch procedure or a Rastelli procedure (connecting the LV to the aorta through the VSD and inserting RV to pulmonary artery conduit). This results in the LV becoming the systemic ventricle at the expense of extensive surgery. When performing a double switch procedure, the LV should be sufficiently trained to become the systemic ventricle after the procedure. This can be done by banding the pulmonary artery (33) but retraining the LV has been associated with long-term failure (34). In some cases the morphology precludes surgical repair and palliation is provided by performing the Fontan operation. Occasionally, heart transplantation is the most viable option.

Echocardiographic assessment of postoperative CCTGA

Echocardiographic assessment of postoperative CCTGA requires first knowing which type of surgical procedure has been performed. Before scanning, the surgical history must be reviewed.

Assessment of patients who underwent a physiologic type of repair or palliation always includes:

- Assessment of severity of tricuspid regurgitation.
- Assessment of RV function.
- Assessment of residual lesions: VSD, LV outflow obstruction; valvar stenosis and/or regurgitation.
- Assessment of pulmonary artery band position and gradient, if present and estimate of LV pressure based on mitral regurgitation jet.

For patients after anatomical repair, the assessment will include evaluating the surgical pathways:

- Atrial pathways looking for pathway obstruction or baffle leaks (similar to TGA after atrial switch operation).
- If Rastelli-type repair, assessment of the LV outflow tract to the aorta and the RV to pulmonary artery conduit function.
- If arterial switch, the assessment includes all the elements mentioned above for patients with TGA.
- Biventricular size and function needs to be monitored after anatomic repair as both LV and RV failure have been described over time, especially after late anatomic correction involving LV retraining.

In case of poor imaging windows, additional cross-sectional imaging using MRI or CT scan can be considered.

Conclusion

Echocardiographic assessment of patients with TGA and CCTGA requires a full segmental analysis with knowledge of the basic morphology and the associated lesions. Follow-up requires awareness of the type of surgery performed for assessment of the surgical repair and always includes a ventricular function evaluation and assessment of valve function. Additional imaging techniques including MRI, CT scan or angiography may be indicated for selected patients.

Declaration of interest

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