We are IntechOpen, the world's leading publisher of Open Access books
Built by scientists, for scientists

6,900
Open access books available

185,000
International authors and editors

200M
Downloads

154
Countries delivered to

Our authors are among the

TOP 1%
most cited scientists

12.2%
Contributors from top 500 universities

BOOK CITATION INDEX
CLARIVATE ANALYTICS
INDEXED

WEB OF SCIENCE™

Selection of our books indexed in the Book Citation Index
in Web of Science™ Core Collection (BKCI)

Interested in publishing with us?
Contact book.department@intechopen.com

Numbers displayed above are based on latest data collected.
For more information visit www.intechopen.com

Congenitally Corrected Transposition of the Great Arteries

English C. Flack and Thomas P. Graham
Vanderbilt University
USA

1. Introduction

Congenitally corrected transposition of the great arteries (ccTGA) is a rare defect combining atrioventricular discordance with ventriculoarterial discordance. The atria are connected to the opposite ventricle (left atrium to right ventricle via a tricuspid valve) and the ventricles are connected to the incorrect great artery (right ventricle to aorta). Thus oxygenated blood is circulated systemically by the morphologic right ventricle (RV) and deoxygenated blood returns to the right atrium to be pumped out the left ventricle (LV) to the lungs (Figure 1). The defect is therefore "corrected" because of the physiologic flow of blood through the body. For the purposes of this review, univentricular hearts, those with common atrioventricular (AV) valves and those with aortic atresia will not be discussed.

2. Anatomy

The most common anatomy of ccTGA is that of {S,L,L}, representing atrial and visceral situs solitus (right-sided inferior and superior vena cavae returning deoxygenated blood to a right sided atrium), L-looped ventricles (the morphologic LV with mitral valve positioned on the right), and L-transposed great arteries (aorta arising off the left-sided morphologic RV and therefore situated anterior and leftward of the pulmonary artery). The RV serves as the systemic ventricle and, in the absence of other defects, oxygen saturation is normal. The most common positions of the heart in the chest are levocardia (apex to the left) or mesocardia (midline). Patients with levo- or mesocardia and visceral situs inversus have a high likelihood of ccTGA and therefore must carefully by assessed for atrial, ventricular, and arterial concordance. Dextrocardia, in which the apex of the heart is to the right, occurs in approximately 20% of patients (Graham & Markham, 2010). In cases of dextrocardia with mirror-image anatomy the anatomic designation is {I,D,D}.

2.1 Associated defects

The most common associated defects in ccTGA are ventricular septal defects (VSDs), which occur in 60-80% of cases, pulmonary stenosis (PS) in 30-50%, and tricuspid valve (TV) anomalies in 14-56%. The VSDs are usually large, perimembranous, and subpulmonary in location. Muscular inlet defects as well as multiple VSDs may also be seen. Pulmonary stenosis, more appropriately referred to as left ventricular outflow tract obstruction

(LVOTO), may be caused by fibromuscular tissue, valvar stenosis, or aneurysmal tissue of the membranous ventricular septum. The associated combination of LVOTO and VSD represents the largest group of ccTGA patients. TV anomalies occur along a spectrum of which an Ebstein-like anomaly is often the most clinically severe. Furthermore, as the TV is subjected to systemic pressures, even normally formed valves display progressive regurgitation with age. Less common defects occurring in association with ccTGA include atrial septal defect, patent ductus arteriosus, pulmonary atresia, double-outlet RV, aortic regurgitation, mitral valve abnormalities, and subaortic stenosis (Graham & Markham, 2010; Hornung & Calder, 2010; Van Praagh et al., 1998).
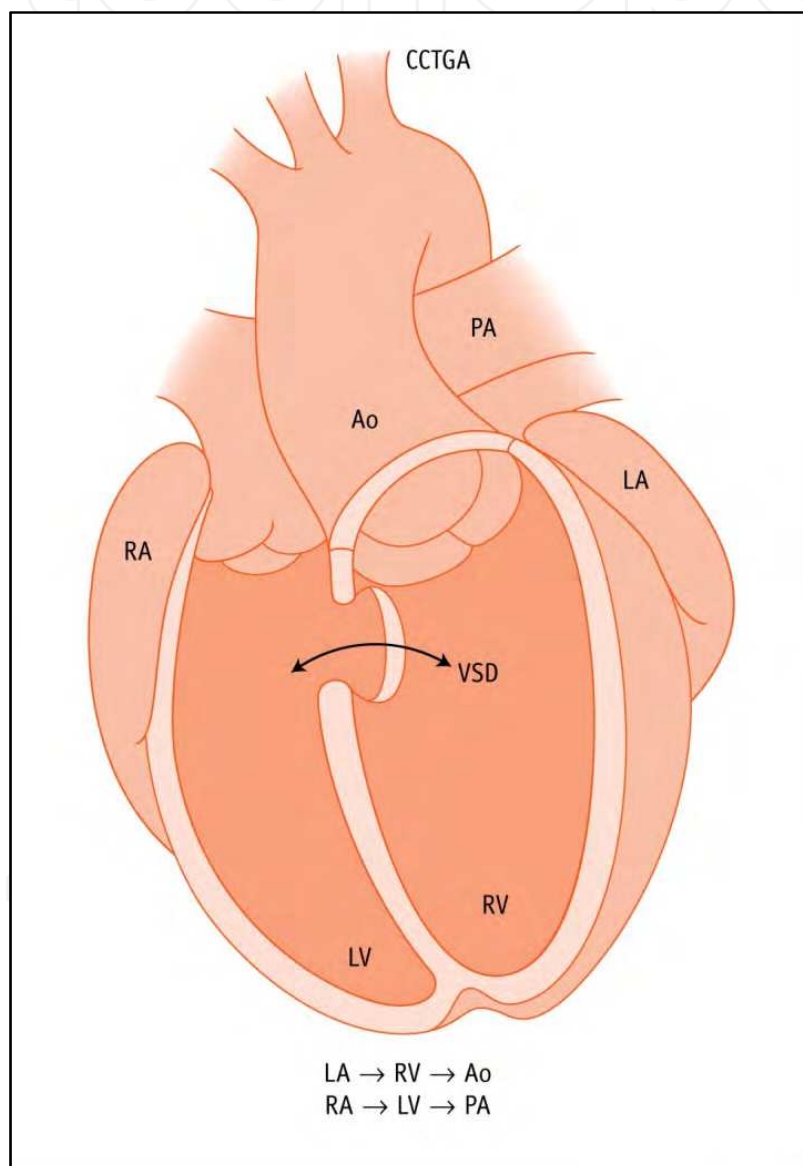

Fig. 1. Congenitally corrected transposition of the great arteries (ccTGA) with ventricular septal defect (VSD). (With permission from Springer Science + Business Media: *Current Treatment Options in Cardiovascular Medicine*, Congenitally Corrected Transposition of the Great Arteries: An Update, Vol. 9, 2007, pp. 405-413, Graham, T.P., Markham, L., Parra, D.P., & Bichell, D., Figure 1).

2.2 Coronary arteries and cardiac veins

The coronary arteries are inverted in ccTGA, as described by Ismat et al. (2002). The most common coronary positions in {S,L,L} hearts are a right coronary artery off of the left posterior aortic cusp and a left common coronary artery off the right anterior cusp. Just as the morphologic LV is situated on the right side of the heart, the morphologic left coronary artery arises off the right aortic sinus. It is this right-sided coronary that bifurcates into the anterior descending artery, which lies in the interventricular groove, and the circumflex branch that runs posterior to the heart through its course in the right AV sulcus. Additional rare anomalies have been described in which both main coronaries arise from a single ostia or one main coronary gives rise to the other (i.e., anterior descending off the right coronary artery) (Hornung & Calder, 2010; Ismat et al., 2002). The cardiac veins seem to correspond to ventricular and coronary anatomy as described in a pathological series by Bottega et al (2009). Although the coronary sinus emptied as normal into the right atrium, dilated Thebesian veins and large collaterals were commonly noted on ccTGA specimens. Venous collateralization was noted between the two ventricles, allowing the morphologic LV to drain via Thebesian veins or collaterals to the coronary sinus. These venous anomalies are thought to be of benefit in providing access to both ventricles in some percutaneous procedures (Bottega et al, 2009).

2.3 Conduction system

The conduction system often consists of dual AV nodes and inversion of AV bundles. An increasing incidence of AV block, at a rate of approximately 2% per year, occurs even in the absence of surgical repair and is more likely in the presence of an intact ventricular septum (Daliento et al., 1986; Huhta et al., 1983). Anderson et al. (1974) consistently demonstrated the finding of an anterior and right-sided AV node that was situated anterolateral to the mitral-pulmonary valve junction. This node connects to the morphologic (right-sided) LV by a descending bundle of conducting tissue that travels anterior and lateral to the pulmonary outflow tract. The bundle branches are inverted, each typical of the morphologic ventricle they serve. In the presence of a subpulmonary VSD the descending AV bundle is located on the anterosuperior and anteroinferior borders of the defect. This is in contrast to concordant hearts {S,D,S} in which the conduction bundle travels along the posteroinferior margin of the VSD. Many ccTGA patients also have a posteriorly-situated AV node, which is often hypoplastic, in addition to a functional anterior node. Depending on the alignment of the interatrial and interventricular septae this posterior node may or may not have connections to the ventricles. Patients with appropriate alignment of the atrial and ventricular septae may be more likely to have two AV nodes with corresponding conduction bundles present. Invading fibrosis of the proximal AV node bundle as well as distal conduction bundles has been described on pathological specimens from older patients with correlating electrocardiogram (ECG) findings of complete heart block, suggesting fibrotic invasion is involved in the development of AV block (Anderson et al., 1974; Daliento et al., 1986).

3. Incidence and genetics

The incidence of ccTGA in patients with congenital heart disease (CHD) is approximately 0.5% with a slight male predominance (Graham & Markham, 2010; Piacentini et al., 2005). Although a specific genetic defect is yet to be defined for ccTGA, the recurrence risk of d-

TGA for siblings of ccTGA patients is 2.6% with an overall recurrence risk of 5.2% for ccTGA siblings to have some type of congenital heart defect (Piacentini et al., 2005). A recurrence risk of >5% is higher than expected, as the risk is typically thought to be 1-3% for unaffected parents to have an additional child with congenital heart disease (Van der Bom et al., 2011).

4. Natural history and outcome

The natural history of ccTGA depends largely on the presence of associated defects. Patients under 5 years old who also have VSD, LVOTO, and/or TV abnormalities represent the highest frequency of non-surgical deaths. However patients with isolated ccTGA (no associated lesions) may survive into their 4th and 5th decades (Hoffman, 2009; Presbitero et al., 1995). Many patients will demonstrate one or more complications including heart block, tricuspid regurgitation (TR), and congestive heart failure (CHF). Approximately 2-4% of ccTGA patients have ventricular pre-excitation (Wolff-Parkinson-White syndrome) and should undergo radiofrequency ablation of accessory pathways in cases of symptomatic reentrant tachycardia. Atrial tachycardia such as atrial fibrillation and flutter often occur with increasing age, atrial enlargement, and after surgical repair where suture lines and scars may support focal reentrant circuits. By 45 years of age 67% of ccTGA patients with associated defects will have developed CHF, as shown in Figure 2, whereas only 25% of ccTGA patients without associated lesions will have progressed to CHF by this age (Graham et al., 2000). Prieto et al suggests that outcome is dependent on morphology of the TV (the systemic AV valve), as this was the only predictor of severe regurgitation and RV dysfunction in a cohort of ccTGA patients described after mean follow-up of 20 years.
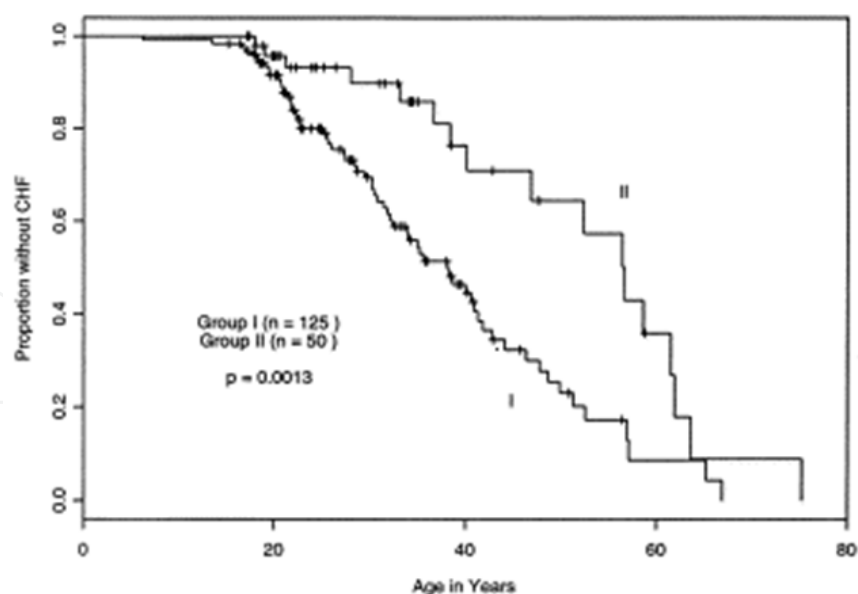

Fig. 2. Freedom from CHF in group I (associated lesions, n=125) and group II (no significant associated lesions, n=50) as a function of increasing age. (Reprinted from *Journal of the American College of Cardiology*, Vol. 36, No. 1, Long-term outcome in congenitally corrected transposition of the great arteries: A multi-institutional study, pp. 255-261, Copyright 2000 with permission from Elsevier).

The authors concluded that severe TV insufficiency leading to RV dysfunction has the greatest impact on long-term survival in both operated and unoperated patients. In patients who underwent surgical intervention for ccTGA, 20-year survival rate was 90% for patients with competent TVs, whereas survival was only 35% for patients with severe TV insufficiency. Furthermore, patients who were diagnosed with severe TV insufficiency demonstrated a rapid deterioration in clinical status with RV failure occurring on average 5 years after onset of insufficiency (Prieto et al., 1998). Overall natural history in the ccTGA patient without associated defects is promising, as patients may remain relatively asymptomatic through early and mid-adulthood. However the frequent development of complications in the 4th and 5th decades often culminates in the progressive development of RV (systemic) dysfunction and heart failure, requiring aggressive medical management and possible surgical intervention (Presbitero et al., 1995).

5. Diagnosis

Just as the natural history is largely dependent on defects associated with ccTGA, so is timing of presentation and diagnoses.

5.1 Prenatal diagnosis

Fetal diagnosis of many forms of CHD continues to improve. However the fetus with ccTGA and mild or no additional intracardiac anomalies may be overlooked by routine ultrasound screening. Distinct features notable on prenatal ultrasound that may improve detection of ccTGA are parallel course of the great arteries in combination with dextrocardia, abnormal insertion of the papillary muscles, and/or an abnormal TV (McEwing & Chaoui, 2004; Paladini et al., 2006; Shima et al., 2009). A retrospective review by Wan et al. found no difference in the number of cardiac interventions, timing of surgery, or survival between a cohort of ccTGA patients diagnosed prenatally (n = 14) and postnatally (n = 26). However, because 70% of this cohort required cardiac intervention prior to 3 years of age, the authors suggest prenatal diagnosis is important for preparation and counseling of the family (2009). A recent review of 11 cases of fetal ccTGA diagnoses describes the use of four-dimensional echocardiography and spatiotemporal image correlation (STIC), in which the relationship of the great arteries can be assessed in several different orthogonal planes by placement of a reference dot on images reconstructed from acquired volume data sets (Zhang et al., 2011).

5.2 Early presentation and diagnosis

Diagnoses of infants and children may occur after murmur evaluation, as VSDs are commonly associated lesions. In cases of large VSDs or severe TV regurgitation, some infants may present in CHF with diaphoresis, pallor, tachypnea, inability to gain weight, hepatomegaly, and a gallop on exam. Auscultation of the ccTGA patient may also reveal a loud, single second heart sound (S2) at the left 2nd intercostal space, with absence of S2 over the right 2nd intercostal space (Friedberg & Nadas, 1970). The presence of VSD combined with LVOTO may lead to a cyanotic presentation from decreased pulmonary blood flow. However, some degree of LVOTO may be protective of the lung bed in patients with large VSDs, and may delay a CHF presentation despite the normal decrease in pulmonary vascular resistance.

5.3 Late presentation and diagnosis

Interestingly, if there are no additional associated defects ccTGA may go unnoticed until adolescence or adulthood. Case reports have even cited incidental findings and late diagnoses of ccTGA in adults in the fifth to eighth decades of life (Chang et al., 2009; Jennings et al., 1984; Orchard et al., 2010; Scardi et al., 1999). A cohort of patients with ccTGA over 18 years of age who presented to an adult CHD clinic over a 15 year period is described by Beauchesne et al (2002). Sixty-six percent of these patients were over 18 years of age when diagnosed, and 17% of the cohort was over 60 years old at the time of diagnosis. Common reasons for referral in such patients range from abnormal ECGs and cardiomegaly on chest radiographs to complete heart block and murmurs (Presberito et al., 1995).

6. Evaluation

6.1 Chest radiograph

The CXR in ccTGA patients with mesocardia or levocardia typically demonstrates a straightened upper-left cardiac border from the leftward-positioned ascending aorta. Dextrocardia usually occurs with normal situs and, as stated previously, occurs in 20% of ccTGA patients (Figure 3). The presence of abdominal situs solitus and dextrocardia should raise suspicion of ccTGA. In the patient without any associated defects, an atypical cardiac position in an otherwise normal CXR may be the only indication of ccTGA.
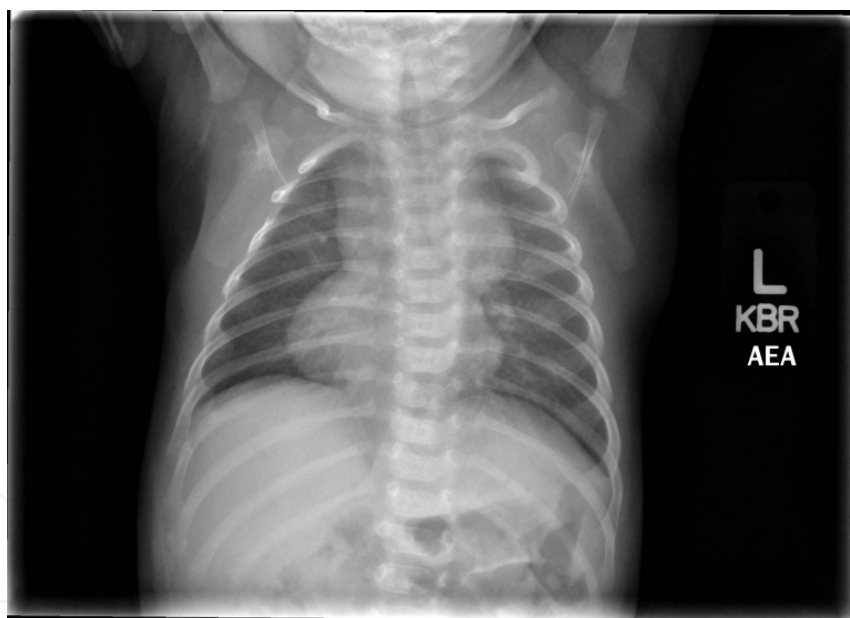

Fig. 3. CXR of infant with dextrocardia, abdominal situs solitus, and ccTGA. Note the position of the cardiac apex pointed to the right. The left heart border demonstrates the prominent left-sided ascending aorta. The thymic shadow is seen over the right mediastinum.

However marked cardiomegaly, left atrial enlargement, and an increase in pulmonary vasculature may be present in patients with a large VSD and significant left to right shunt. A CXR with impressive cardiomegaly and left atrial enlargement may also be indicative of an Ebstein-like malformation of the TV. The presence of pulmonary stenosis or atresia will demonstrate darkened lung fields from attenuated pulmonary blood flow. Overall, the degree of cardiomegaly and amount of visible pulmonary vascularity is dependent on the

presence and direction of shunting, as well as the severity of LVOTO (Carey & Ruttenberg, 1964).

6.2 Electrocardiogram

The ECG in patients with ccTGA is most significant for a superior QRS axis and atypical septal activation. As discussed previously, the conduction system in ccTGA consists of inverted AV bundles. Therefore the septum is activated from right to left, demonstrating presence of septal Q waves in the right precordial leads (QR pattern in leads V4R and V1) and absence of Q waves in the left precordial leads (rS pattern in lead V6). In fact, undiagnosed ccTGA patients with such a pattern on ECG have been diagnosed with remote inferior infarcts (Jennings et al., 1984; Warnes, 2006). Preexcitation may be observed in those patients with ccTGA and Wolff-Parkinson-White. Finally, varying degrees of AV block may be present, as well as patterns of right or left-sided chamber enlargement.

6.3 Echocardiography

Transthoracic echocardiography (TTE) as an imaging modality is relatively inexpensive, widely available, and noninvasive. As with many types of CHD, TTE is the first line and most useful modality in the diagnosis of ccTGA. The anatomical designation (most commonly {S,L,L} as discussed previously), is first assigned by demonstrating atrial position, ventricular looping, and arterial looping. Morphology of the RV is seen on TTE by the presence of coarse trabeculations and a moderator band, whereas the LV has a smooth-walled endocardium and a funnel-shaped appearance. The level of the TV is inferior to the MV, which may also give a clue to ventricular inversion. In evaluation of the outflow tracts, the aorta in ccTGA is usually anterior and to the left of the PA. Once the diagnosis of ccTGA is made through demonstration of discordance between atria and ventricles as well as ventricles and great arteries, several anatomic objectives should be defined in the TTE evaluation. Semilunar and AV valve morphology as well as presence and severity of regurgitation warrant full description. Coronary origins should be identified and their proximal courses described. The degree of LVOTO is important as well as any additional defects present, as these will impact whether and what type of surgical repair is necessary (Oechslin, 2009). Transesophageal echocardiography (TEE) has been shown to have greater accuracy over TTE in correctly defining atrial situs and chordal AV valve attachments in adult patients with ccTGA (Caso et al., 1998). TEE is also more useful for investigation of intracardiac vegetations in cases of suspected endocarditis and in evaluation of thrombus in the atrial appendages, which may be applicable to the ccTGA patient with sustained atrial arrhythmias.

6.4 Cardiac catheterization

Rather than a modality for diagnosis, cardiac catheterization (Figure 4) is typically reserved for the post-surgical patient who would benefit from an intervention such as LV to pulmonary artery (PA) conduit dilation or stent placement. For patients undergoing surgical palliation for single-ventricle ccTGA anatomy, catheterization is performed to assess pressure, function, and valve regurgitation prior to surgery. Most interesting, however, is the adult patient who presents with ischemic heart disease and is discovered on cardiac catheterization to have ccTGA after abnormal catheter passes or inversion of coronary arteries on angiography (Jennings et al., 1984).
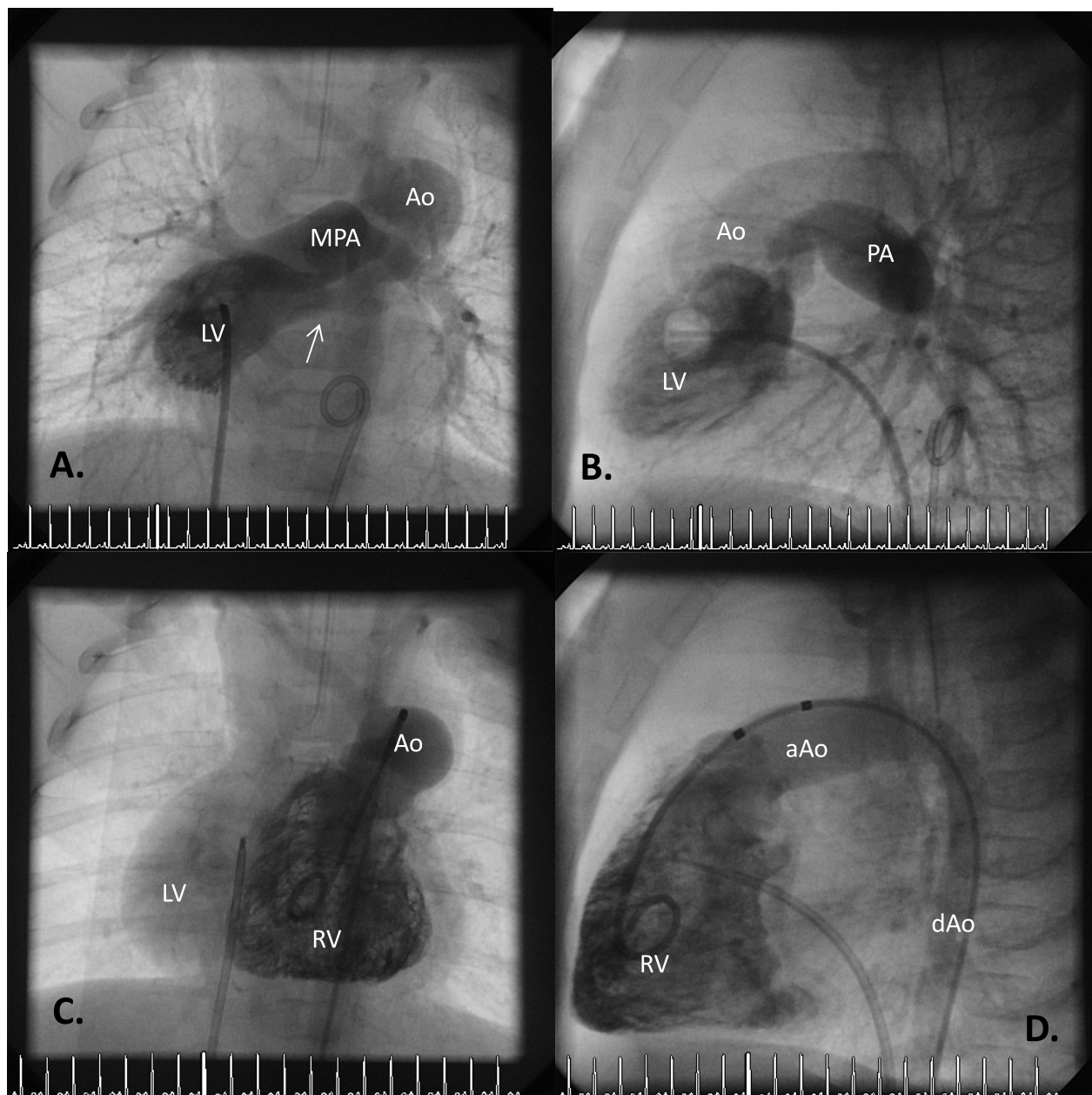
Fig. 4. Cardiac catheterization of ccTGA infant with dextrocardia, pulmonary stenosis, and VSD (same infant as in Fig. 3). (A.) Anterior-posterior projection. A catheter is positioned in the right-sided morphologic left ventricle (LV). Contrast fill the LV, pulmonary trunk, and pulmonary arteries. Contrast flows right to left across the VSD (arrow) and fills the aorta. (B.) Lateral projection. Contrast from the LV flows through the LV outflow tract, across the pulmonary valve, and fills the pulmonary arteries. The aorta fills by right to left shunting through the VSD. Note the aorta is anterior to the pulmonary artery. (C.) Anterior-posterior projection. A catheter is positioned retrograde into the left-sided morphologic right ventricle (RV). Contrast fills the trabeculated RV and the leftward aorta. (D.) Lateral projection. Contrast fills the large RV, ascending, and descending aorta. LV, left ventricle; RV right ventricle; Ao, Aorta; aAo, ascending aorta; dAo, descending aorta; MPA, Main pulmonary artery; PA, Pulmonary artery.

6.5 Cardiac Magnetic Resonance Imaging (cMRI)

Cardiac MRI is now used in many types of CHD to further define anatomy and to quantify ventricular function and volume (Figure 5). For initial diagnosis, cMRI may be helpful in patients with restricted TTE windows, to define visceroatrial situs, and to delineate complex associated defects. In patients with interruption of the inferior vena cavae, systemic return from the lower body can be difficult to delineate by echocardiography, but is well defined by cMRI. Because echocardiographic evaluation of RV function in ccTGA patients is limited by geometric assumptions, cMRI has become the gold standard for RV function and volume assessment. TV morphology as well as degree of regurgitation can also be determined through cMRI. Prior to performing anatomic surgical repair in a ccTGA patient beyond infancy, cMRI may be useful in evaluation of LV mass, volume, and ejection fraction. Furthermore, if there are concerns about degree of LV dysfunction, perfusion studies with delayed enhancement MRI may be performed to directly investigate scarring of the LV myocardium prior to committing this ventricle to systemic workload. Cardiac MRI may therefore be a useful modality for evaluation of ccTGA patients not only as an adjunct to TTE for initial diagnosis, but also for assessment prior to surgical repair and serial follow-up of the systemic RV. If the presence of MRI-incompatible pacemaker or prosthetic valve precludes assessment by MRI, computed tomography (CT) scans can depict anatomy but cannot yield functional data as does MRI (Schmidt et al., 2000; Teo & Hia, 2011).
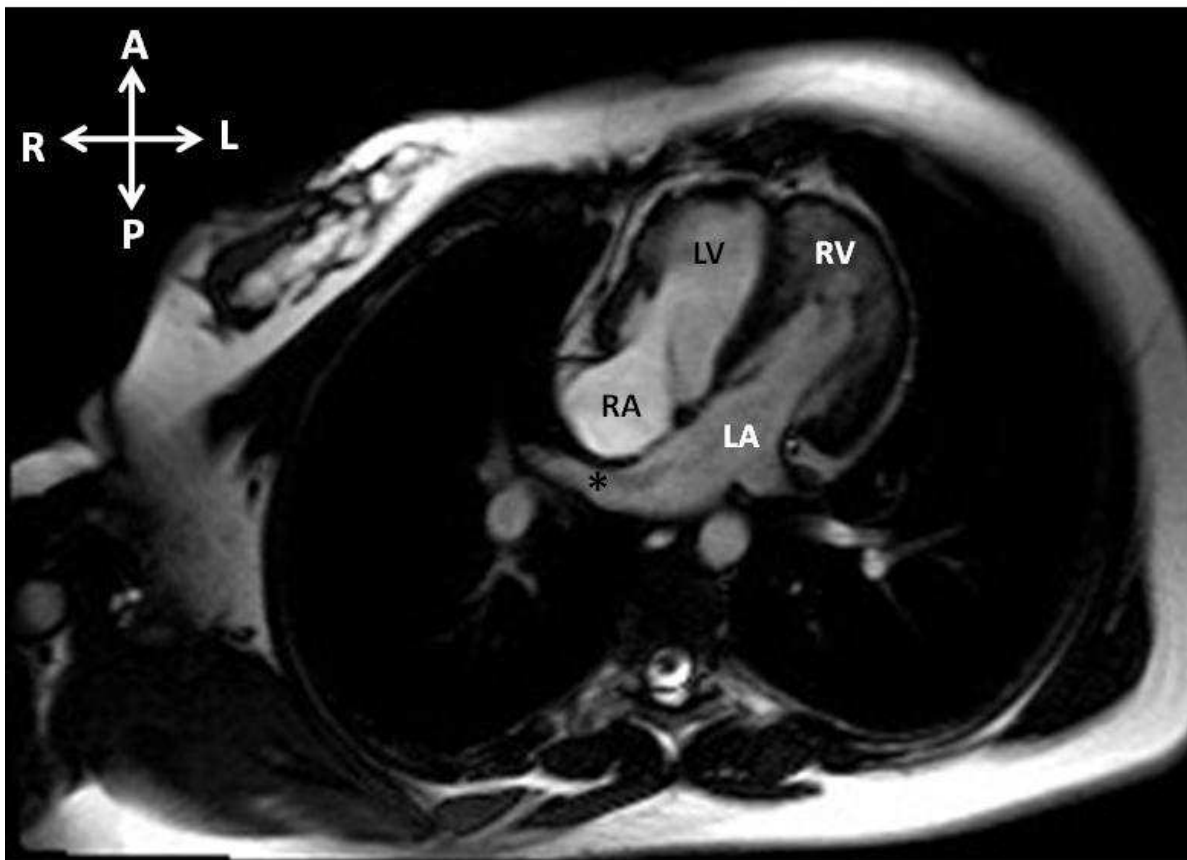

Fig. 5. Oblique cut T2-weighted MRI image of 4-chamber cardiac view of ccTGA patient with levocardia. The RA empties into a right-sided, smooth-walled, morphologic LV. A star (*) labels the entrance of a right pulmonary vein into the left atrium, which empties into a trabeculated, left-sided, morphologic RV. RA, right atrium; LV, left ventricle; LA, left atrium; RV, right ventricle.

6.6 Exercise and stress testing

Cardiopulmonary exercise testing by treadmill is an important adjunct for ccTGA patient evaluation and management. In those patients able to perform treadmill tests, exercise capacity is determined through minute ventilation, carbon dioxide production, and oxygen consumption. Impaired exercise capacity in ccTGA patients has been shown to correlate with diastolic dysfunction in the form of increased RV filling pressures as measured by tissue Doppler imaging (Tay et al., 2011). Cardiopulmonary exercise testing in combination with gadolinium-enhanced MRI has been utilized to demonstrate RV myocardial fibrosis hypothesized to be responsible for RV dysfunction (Giardini et al., 2006). Systemic RV function can also be evaluated by dobutamine stress testing, in which MRI is performed at baseline and with dobutamine infusion. Objectively defining the capacity of the systemic RV to respond to stress may guide treatment on both initial and follow-up evaluations (Dodge-Khatami et al., 2002; Fratz et al., 2008). Sequential testing, performed either by exercise testing or by dobutamine stress test, is useful to assess overall cardiopulmonary function and response to medical or surgical therapy.

7. Management

7.1 Medical management

CHF medical management for the ccTGA patient with systemic RV has been extrapolated from CHF therapy for LV failure. This primarily includes β-adrenergic receptor blockade (β-blockers), diuretics and afterload-reducing agents with an angiotensin-converting enzyme (ACE) inhibitor (Winter et al., 2009). Digoxin may also be useful for its inotropic and antiarrhythmic effects. Angiotensin receptor blockade with losartan was evaluated in a multicenter, randomized, placebo-controlled clinical trial by Dore and colleagues (2005) but found to have no improvement on exercise capacity and no reduction in neurohormonal levels in patients with systemic right ventricles. Overall, evidence-based therapy for optimal CHF treatment in patients with systemic RV is lacking. Beyond medication, cardiac resynchronization has emerged as a therapy for patients with impaired systemic RV function and widened QRS morphology on ECG. Increased QRS duration as a result of bundle branch block or conventional pacemaker is typically greater than 120-140 ms with some patients having QRS duration >200 ms. Such electromechanical dyssynchrony creates inefficiency in ventricular ejection, whereas restoring synchrony has been shown to decrease QRS duration with improvement in RV filling time, ejection fraction, and overall CHF symptoms (Diller et al., 2006; Janousek et al., 2004; Kordybach et al., 2009). Takemoto et al. (2010) reports the use of transvenous permanent para-Hisian pacing in an 8 year old with ccTGA. Restoration of cardiac synchrony decreased the QRS duration from 198 ms to 94 ms, decreased interventricular conduction delay from 137 ms to 37 ms, and improved the patient's CHF symptoms from NYHA (New York Heart Association) class III to NYHA class II over a period of 6 months. Limitations in cardiac resynchronization therapy include difficulty in percutaneous lead delivery, although this has successfully been accomplished even in ccTGA cases of dextrocardia (Malecka et al., 2010).

7.2 Surgical management

Indications for surgical management in ccTGA patients of all ages continue to evolve and most often are determined on a case-by-case basis. Beauchanese et al. (2002) described a cohort of 44 unrepaired adult ccTGA patients. Of these, the 30 patients who required surgical intervention had significantly larger pre-operative cardiothoracic ratios on chest

radiographs, and had moderate to severe or severe systemic AV valve regurgitation. The ejection fraction of the systemic ventricle between the operated and unoperated groups was not statistically significant (Beauchesne et al., 2002). As discussed previously and depicted in Figure 2, nearly 2/3 of unrepaired ccTGA patients with associated defects will have developed CHF by the age of 45 years. Even asymptomatic adults with ccTGA have been shown by echocardiography to have RV dysfunction through the use of tissue Doppler quantification techniques (Bos et al. 2006). Thus the natural evolution of ccTGA for the majority of patients is eventual RV dysfunction and TV regurgitation. It is postulated that progression to failure in a systemic RV is unavoidable because the RV and TV are not anatomically suited to withstand the systemic pressure for which the LV and MV are intended. One mechanism thought to contribute to progressive RV decompensation is worsening TR from annular dilation and/or displacement of the septal leaflet of the TV as the RV remodels to accommodate systemic afterload.

ccTGA {S,L,L} Surgical Repair and Palliation			
Classic / Physiologic Repair		**Anatomic Repair**	
Associated Defect	**Repair**	**Associated Defect**	**Double Switch Repair**
VSD	VSD closure	VSD + normal PV	•VSD closure •Atrial switch (Senning / Mustard) •Arterial switch
VSD + PS	VSD closure + PS relief	PS / Atresia + VSD	•VSD closure + RV-PA conduit •Atrial switch + Arterial switch
VSD + PS/Atresia	Biventricular Repair: •VSD closure + LV-PA conduit	PS / Atresia + routable VSD	•RV-PA conduit • Atrial switch + Rastelli
	Univentricular Repair: • Systemic to PA shunt • Bidirectional Glenn • Fontan	Severe RV dysfunction Small RV Abnormal atrial anatomy	•Hemi-Mustard with bidirectional Glenn (modified atrial switch) •Arterial switch or Rastelli procedure
TR	Tricuspid valve repair or replacement	TR	PA banding to decrease TR or tricuspid valve repair / replacement

Table 1. ccTGA {S,L,L} Surgical Repair and Palliation. VSD, ventricular septal defect; PS, pulmonary stenosis; PA, pulmonary artery; PV, pulmonary valve; RV, right ventricle; TR, tricuspid regurgitation; BDG, Bidirectional Glenn

Depending on the age of presentation and extent of associated lesions, surgical repair may include one or more of several approaches (Table 1). In patients with a VSD and no LVOTO, "classic" or "physiologic" repair may include VSD closure only. Specific techniques must be employed in ccTGA patients to avoid damage to the conduction system during VSD closure. Because the AV conduction bundle descends along the anterior rim of the VSD and travels along the septal side of the right-sided morphologic LV, it is recommended to suture the VSD patch along the morphological right ventricular aspect of the septum. The surgical approach should be via right atriotomy and right-sided mitral valve. Ideally the VSD patch will lie partially on the morphologic LV septal aspect (to avoid damage to the TV superiorly) and partially on the morphologic RV aspect of the septum inferiorly (to avoid damage to the main conduction bundle) (Jonas, 2004). Physiologic repair may also include relief of

pulmonary stenosis (PS) and/or LV to PA conduit placement. There is, however, the possibility that decreasing LV pressure by VSD closure and/or PS relief may allow the ventricular septum to realign towards the LV, resulting in displacement of the TV septal leaflet and increasing TR (Kral Kollars et al. 2010; Said et al. 2011). In a cohort of 123 patients with ccTGA presenting for classic biventricular repair over 33 years, the surgical group undergoing repair of VSD + PS demonstrated the greatest survival whereas patients requiring TV replacement at their initial operation exhibited the shortest survival. Risk factors for death in the VSD +/- PS relief groups included pre-operative RV end diastolic pressure greater than 17mmHg and complete heart block. Survival rates at 1-, 5-, 10-, and 15-years for patients who underwent classic repair were 84%, 75%, 68%, and 61%, respectively, although 17 of the 113 patients in this subgroup underwent Fontan and achieved 100% survival in short-term follow-up (Figure 6). The univentricular pathway with Fontan was assigned to ccTGA patients for which biventricular repair was contraindicated, as in patients with straddling AV valve tissue, inaccessible or multiple VSDs, or unbalanced complete AV canals (Hraska et al. 2005). More recently Bogers et al. (2010) confirmed that classic repair in which the RV remains the systemic ventricle results in significant incidence of reoperation and overall suboptimal survival.
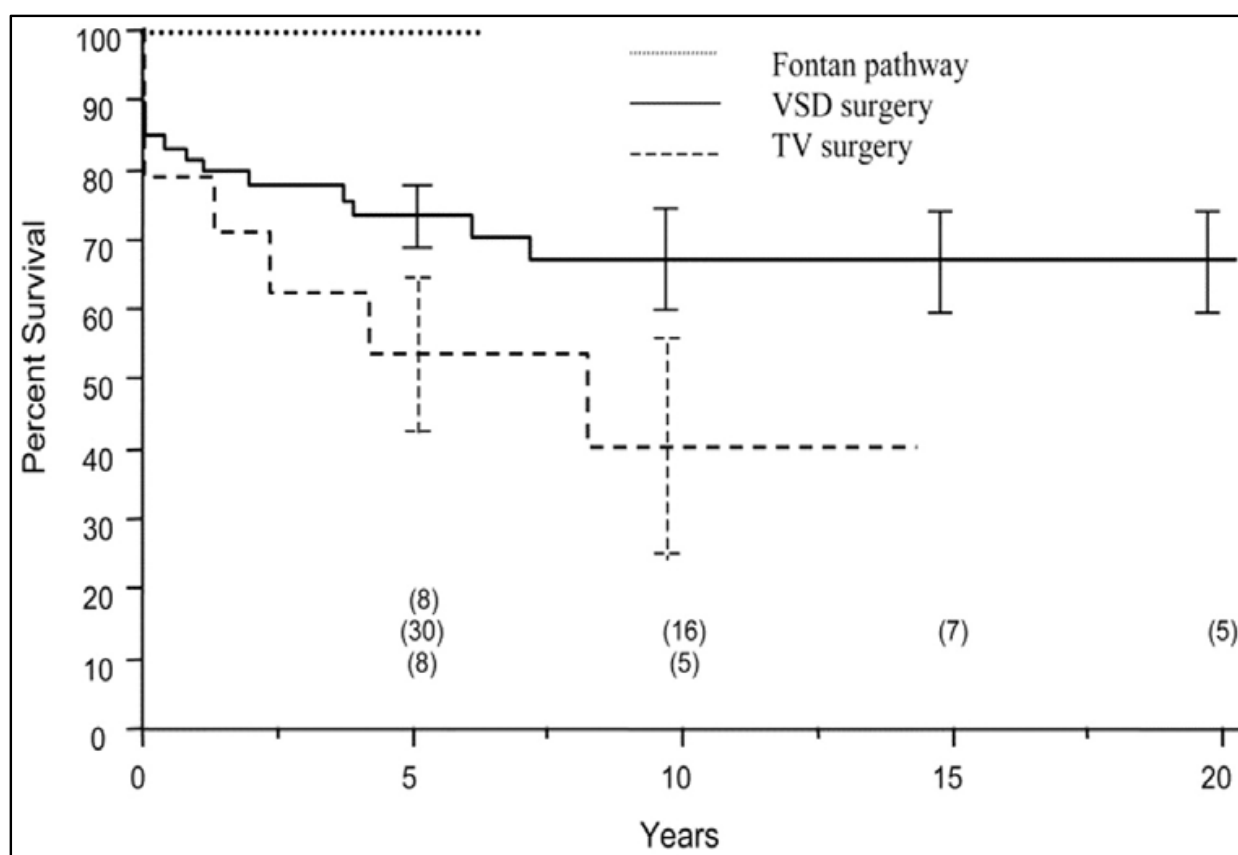

Fig. 6. Operative survival in ccTGA patients undergoing Fontan pathway (*dotted line*; n = 17), VSD surgery (*solid line*; n = 76), and TV surgery (*dashed line*; n = 14). Numbers of patients at risk are in parentheses. Error bars indicate 70% confidence limits. VSD, ventricular septal defect; TV, tricuspid valve. (Reprinted from *The Journal of Thoracic and Cardiovascular Surgery,* Vol. 129, No. 1, Long-term outcome of surgically treated patients with corrected transposition of the great arteries, pp. 182-191, Copyright 2005 with permission from Elsevier).

The "anatomic" or "Double Switch" (DS) operation was developed in response to unsatisfactory outcomes after the classic repair. Components of the DS (Figure 7A) include arterial switch with coronary artery transfer, VSD closure if necessary, and interatrial baffle by Senning or Mustard procedure. The Senning and Mustard operations, referred to as an "atrial switch," serve to direct systemic venous flow to the TV and RV and pulmonary venous flow to the mitral valve and LV. The purpose of the DS is to improve long term outcome by restoring the LV and MV to the systemic circulation. Requirements for this repair before committing the LV to the systemic workload include pre-operative LV pressure that is 80-100% systemic and normal LV wall thickness and function for a systemic LV (Duncan & Mee, 2005; Poirier et al., 2004). In the absence of LVOTO, pulmonary hypertension, or an unrestrictive VSD, the morphologic LV requires training prior to committing it to the systemic ventricle in the DS. LV training has been performed by placement of a pulmonary artery band (PAB) which is then serially tightened to introduce a greater pressure load nearing that of systemic pressure to the naïve LV. Median banding time for the purpose of LV retraining has been reported on average to be 13-14 months (Ly et al., 2009; Poirier et al., 2004; Winlaw et al., 2005). Morphologic LV reconditioning with PAB in patients with systemic RV after atrial switch for dextrotransposition of the great arteries (dTGA) has been described by Poirier et al (2004). PAB was performed in this population prior to anatomic correction or as bridge to transplant, and the success rate of completing adequate LV retraining was significantly less in patients beyond 12 years of age (20% of patients over 12 years completed the protocol, whereas 62% of patients less than 12 years were able to complete the PAB protocol, p = 0.02). Although a well defined standard for age of PAB placement in this setting is yet to be realized, it is apparent that candidacy for LV training with PAB beyond adolescence is questionable. Also concerning is report of late LV dysfunction in ccTGA patients who underwent DS operation after successful LV retraining by PAB placement (Quinn et al., 2008).

Rather than performing pulmonary artery banding in symptomatic ccTGA patients with intention of anatomic repair, Metton and associates (2010) advocate the use of PAB in asymptomatic ccTGA neonates and infants with intact ventricular septum to maintain rather than train the LV. In Metton's group the TV was not repaired at PAB placement, as it was thought that PAB placement may improve TR that was present prior to banding (Ly et al., 2009). This mechanism is described by Kral Kollars et al. (2010) in 14 patients who underwent PAB for LV retraining (median age 1.1 years, range 0 to 12 years). Eleven of the 14 patients had an increase in LV pressure of ≥2/3 systolic RV pressure with PAB and demonstrated significantly decreased TR as the LV geometry became more spherical and the interventricular septum shifted toward the morphologic RV. Patients who underwent classic ccTGA repair with procedures that reduced LV pressure below that of the RV, such as VSD closure with LV to PA conduit placement, demonstrated significantly increased TR postoperatively.

Although it is reasonable to medically manage mild TR with anticongestive therapy and afterload reduction, surgical intervention is indicated in cases of moderate or moderate to severe TR. TV repair for ccTGA patients is rarely successful, and most patients require valve replacement, which can be problematic in young children because of the relatively large prosthesis needed to allow for growth. Palliation with PAB may therefore be reasonable in infants and young children, since it has been shown that severe TV insufficiency leading to RV dysfunction has the greatest impact on long-term survival (Kral Kollars et al., 2010;

Prieto et al., 1998). Several groups have concluded that TV replacement should be considered at the earliest sign of RV dysfunction, with recommendations to consider operation before systemic ventricular ejection fraction (EF) decreases below 40 and 44% (Mongeon et al., 2011; Van Son et al., 1995).
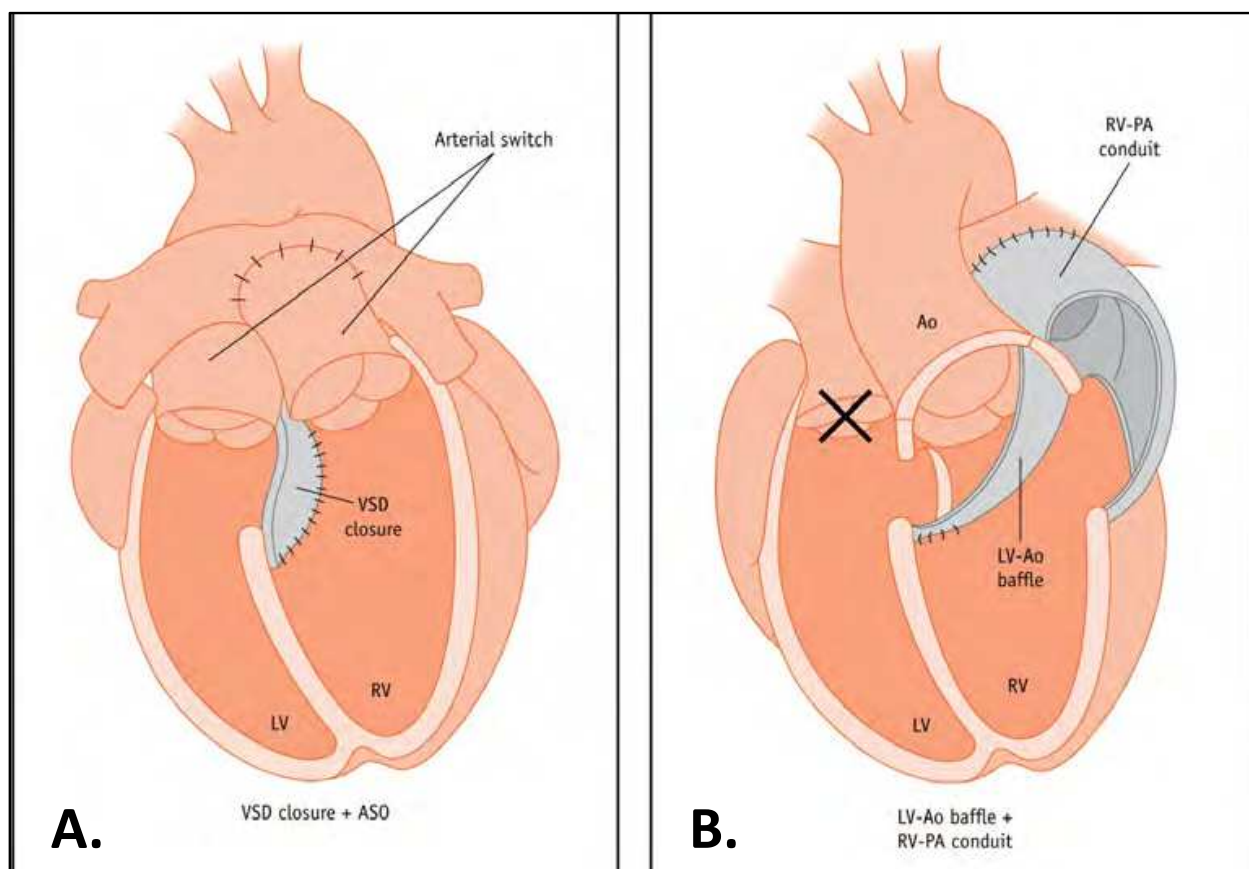

Fig. 7. The Double Switch operation for ccTGA. (A.) The double-switch anatomic surgical repair for ccTGA with VSD consists of an arterial switch, VSD closure, and atrial venous switch (not shown) by intra-atrial baffle operation (i.e., Senning or Mustard repair). (B.) The double-switch for ccTGA with left ventricle (LV) outflow tract obstruction includes an anatomical LV - aorta (Ao) baffle (i.e., Rastelli repair) and anatomical right ventricle (RV) - pulmonary artery (PA) conduit. Although not pictured, the baffle and conduit repair are also in combination with an atrial switch. (With permission from Springer Science + Business Media: *Current Treatment Options in Cardiovascular Medicine*, Congenitally Corrected Transposition of the Great Arteries: An Update, Vol. 9, 2007, pp. 405-413, Graham, T.P., Markham, L., Parra, D.P., & Bichell, D., Figure 1).

The combination of progressive systemic RV dysfunction and TR has lead to the consideration of a variation in DS operation for patients with LVOTO. Rather than combining the atrial and arterial switches, the Senning or Mustard atrial switch procedure is combined with a Rastelli operation, in which the LV outflow is channeled from the LV through a large VSD to the aorta and an RV to PA conduit is placed (Figure 7B). This operation is technically challenging and subject to the need for conduit replacements as well as possible reoperation for interatrial or interventricular baffle obstructions. Specific to the Senning / Rastelli operation, risk factors associated with death include longer

cardiopulmonary bypass and aortic cross-clamp times, and there is an increased risk of complete heart block and ventricular dysfunction if the existing VSD requires enlargement (Gaies et al., 2009; Shin'oka et al., 2007). Nevertheless, intermediate results in a small group of ccTGA patients with VSD and LVOTO who underwent this form of anatomic repair suggest good biventricular function and mild or no AV valve insufficiency up to 17 years post-operatively (Hörer et al., 2007).

An additional variation in the DS for patients with severe RV dysfunction, hypoplasia of the RV, or abnormal right atrial anatomy includes a modified atrial switch termed the "hemi-Mustard/bidirectional Glenn," which is performed in combination with either an arterial switch or a Rastelli procedure. In this operation the interatrial baffle only includes the IVC return, as the SVC is reimplanted into the pulmonary artery to create a cavo-pulmonary Glenn shunt, and the SVC portion of the RA is oversewn. Midterm outcomes from the hemi-Mustard/Glenn as reported by Malhotra et al. (2011) are favorable and hold several advantages over the traditional Senning or Mustard atrial switch. The authors report a prolonged lifespan of the RV to PA conduit due to volume-unloading the RV, increased intra-atrial space for pulmonary venous return (and therefore less risk of pulmonary venous obstruction), and less risk for arrhythmia with the reduction in intra-atrial suture lines. It remains to be seen if the hemi-Mustard / bidirectional Glenn variant of the DS will prove favorable in long-term studies.

8. Outcomes: Physiologic vs. anatomic repair

Alghamdi and associates (2006) published a meta-analysis of 11 nonrandomized studies totalling 124 ccTGA patients and compared in-hospital mortality between physiologic and anatomic repair. Patient age at time of repair ranged from 3 months to 55 years with 41% of patients undergoing definitive repair prior to 1995. Thirty patients underwent physiologic repair, 69 underwent Rastelli-type anatomic repair, and 25 received anatomic repair with arterial switch. The Rastelli-type anatomic repair had significantly lower hospital mortality while era of operation before 1995 demonstrated an increased mortality risk. A large risk analysis performed by Shin'oka et al. (2007) combined ccTGA patients with a group of systemic RV patients with discordant AV connections, (n=189) and compared long-term results of definitive surgical repair with respect to hospitalization, late mortality, and reoperation. Risk factors for hospital death included preoperative moderate TR and intraoperative cardiopulmonary bypass time of over 240 minutes. The presence of TR was also a risk factor for late mortality. Reoperation risks included preoperative cardiomegaly (cardiothoracic ratio of >0.6) and presence of TR, operative need for VSD enlargement, and patient size of <10 kg. Although survival of classic repair in patients without TR was satisfactory in comparison to anatomic repair, patients with ccTGA and discordant AV connections with TR demonstrated improved survival with anatomic repair. More recently Lim and colleagues (2010) report results from a multicenter study including 167 patients who underwent biventricular ccTGA repair. Of the patients studied, 123 underwent physiologic repair (ASD or VSD closure, TV surgery, and/or pulmonary ventricle to PA conduit placement), and 44 underwent anatomic repair (atrial + arterial switch or atrial + interventricular re-routing procedure) over the years 1983 – 2009. Long-term results of biventricular repair revealed an estimated survival of 83.3% ± 0.05% at 25 years. The incidence of complete heart block was lower for the anatomic repair group, and there was a late mortality of 5.9% after physiologic repair in comparison to 0% after anatomic repair.

Freedom from systemic AV valve regurgitation and ventricular dysfunction was significantly higher after anatomic repair. The authors concluded that anatomic is superior to physiologic repair in patients with two adequately sized ventricles. However high risk groups such as those patients with RV dysfunction or the need for LV training warrant careful selection prior to undergoing anatomic repair. Taken together, these outcomes favor anatomic over classic /physiologic repair with careful preoperative assessment of TR for the purpose of risk stratification.

9. Follow-up and special considerations

Patients with ccTGA require outpatient follow-up every 1-2 years by a pediatric or adult congenital cardiologist. Symptomatology, ventricular function, and valvar insufficiency should further guide frequency of follow-up. It is recommended an ECG be performed at each visit to monitor for AV block with periodic consideration of Holter monitor. Cardiopulmonary exercise testing is performed to assess overall function as well as response to medical or surgical therapy. RV function in the unrepaired or physiologically repaired ccTGA patient must be closely monitored with serial echocardiography even in asymptomatic patients (Bos et al., 2006). Cardiac MRI with cine data used to quantify RV volume, mass, and ejection fraction is the best modality to serially quantify RV function, and should be performed every 3-5 years.

9.1 Pregnancy

Pregnancy in the ccTGA patient is generally well tolerated except in the presence of maternal NYHA class III-IV symptoms, moderate or severe AV valve regurgitation, or poor ventricular function (EF<40%). Evaluation of pregnancy outcome in 22 women with ccTGA revealed 50 live births in 60 total pregnancies (83%). However, the rate of miscarriage in the ccTGA mothers was higher than the general population (Connolly et al., 1999). A recent cohort of patients by Gelson and colleagues (2011) revealed high maternal and neonatal morbidity in women with systemic right ventricles with a significant number of babies born small for gestational age. Although cyanosis in women with ccTGA has been shown to be a risk factor for miscarriage, the women in the cohort of Gelson et al. were normally saturated (Gelson et al., 2011; Thierrien et al. 1999). The risk of congenital heart defects in the offspring of mothers with ccTGA has not been defined.

9.2 Heart transplant

Patients for which heart transplantation may be considered are those with end-stage RV failure, significant LV dysfunction and pulmonary valve abnormalities precluding successful DS operation, or uncontrollable arrhythmia (Duncan & Mee, 2005). For patients undergoing surgical intervention, poor preoperative EF of the systemic ventricle has been shown to predict the eventual need for transplantation (Beauchesne et al., 2002).

10. Conclusions and special considerations

Although debate continues over efficacy and long-term follow-up of physiologic vs. anatomic repair for ccTGA, recent outcomes data favor anatomic correction in which systemic function is restored to the LV. Management considerations specific to this population of complicated patients include type and timing of surgical intervention to

pursue. The age and eligibility of pulmonary artery banding for LV retraining is yet to be standardized, and as pulmonary banding for maintenance of LV function in the asymptomatic infant is further evaluated, individualized decisions such as these are sure to produce much debate.

11. References

Alghamdi, A.A., McCrindle, B.W., & Van Arsdell, G.S. (2006). Physiologic Versus Anatomic Repair of Congenitally Corrected Transposition of the Great Arteries: Meta-Analysis of Individual Patient Data. *Annals of Thoracic Surgery*, Vol. 81, No. 4, (April 2006), pp. 1529-1535, ISSN 0003-4975.

Anderson, R.H., Becker, A.E., Arnold, R., & Wilkinson, J.L. (1974). The Conducting Tissues in Congenitally Corrected Transposition. *Circulation*, Vol 50, (1974), pp. 911-923, ISSN 1524-4539.

Beauchesne, L.M., Warnes, C.A., Connolly, H.M., Ammash, N.M., Tajik, A.J., & Danielson, G.K. (2002). Outcome of the Unoperated Adult Who Presents With Congenitally Corrected Transposition of the Great Arteries. *Journal of the American College of Cardiology*, Vol. 40, No. 2, (July 2002), pp. 285-290, ISSN 1558-3597.

Bogers, A.J., Head, S.J., de Jong, P.L., Witsenburg, M., & Kappetein, A.P. (2010). Long term follow up after surgery in congenitally corrected transposition of the great arteries with a right ventricle in the systemic circulation. *Journal of Cardiothoracic Surgery*, Vol. 5, Article No. 74, (September 2010), ISSN 1749-8090.

Bos, J.M., Hagler, D.J., Silvilairat, S., Cabalka, A., O'Leary, P., Daniels, O., Miller, F.A., & Abraham, T.P. (2006). Right Ventricular Function in Asymptomatic Individuals with a Systemic Right Ventricle. *Journal of the American Society of Echocardiography*, Vol. 19, No. 8, (August 2006), pp. 1033-1037, ISSN 0894-7317.

Bottega, N.A., Kapa, S., Edwards, W.D., Connolly, H.M., Munger, T.M., Warnes, C.A., & Asirvatham, S.J. The cardiac veins in congenitally corrected transposition of the great arteries: Delivery options for cardiac devices. *Heart Rhythm*, Vol. 6, No. 10, (October 2009), pp. 1450-1456, ISSN 1547-5271.

Carey, L.S. & Ruttenberg, H.D. (1964). Roentgenographic features of congenital corrected transposition of the great vessels: A comparative study of 33 cases with a roentgenographic classification based on the associated malformations and hemodynamic states. *American Journal of Roentgenology, Radium Therapy, and Nuclear Medicine*, Vol. 92, No. 3, (September 1964), pp. 623-651, ISSN 0002-9580.

Caso, P., Ascione, L., Lange, A., Palka, P., Mininni, N., & Sutherland, G.R. (1998). Diagnostic value of transesophageal echocardiography in the assessment of congenitally corrected transposition of the great arteries in adult patients. *American Heart Journal*, Vol. 135, No. 1, (January 1998), pp. 43-50, ISSN 0002-8703.

Chang, H-Y., Yin, W-H., Hsiung, M-C., & Young, M-S. (2009). A Heart Reversed Triply: Situs Inversus Totalis with Congenitally Corrected Transposition of the Great Arteries in a Middle-Aged Woman. *Echocardiography: A Journal of Cardiovascular Ultrasound and Allied Techniques*, Vol. 26, No. 5, (May 2009), pp. 617-621, ISSN 1540-8175.

Connolly, H.M., Grogan, M., & Warnes, C.A. (1999). Pregnancy among women with congenitally corrected transposition of the great arteries. *Journal of the American College of Cardiology*, Vol. 33., No. 6, (May 1999), pp. 1692-1695, ISSN 1558-3597.

Daliento, L., Corrado, D., Buja, G., John, N., Nava, A., & Thiene, G. (1986). Rhythm and Conduction Disturbances in Isolated, Congenitally Corrected Transposition of the Great Arteries. *The American Journal of Cardiology*, Vol. 58, No. 3, (August 1986), pp. 314-318, ISSN 0002-9149.

Diller, G. P., Okonko, D., Uebing, A., Ho, S. Y., & Gatzoulis, M. A. (2006). Cardiac resynchronization therapy for adult congenital heart disease patients with a systemic right ventricle: analysis of feasibility and review of early experience. *Europace*, Vol. 8, No. 4, (April 2006), pp. 267-272, ISSN 1099-5129.

Dodge-Khatami, A., Tulevski, I.I., Bennink, G.B., Hitchcock, J.F., de Mol, B.A., van der Wall, E.E., & Mulder, B.J. (2002). Comparable Systemic Ventricular Function in Healthy Adults and Patients With Unoperated Congenitally Corrected Transposition Using MRI Dobutamine Stress Testing. *Annals of Thoracic Surgery*, Vol. 73, No. 6, (June 2002), pp. 1759-1764, ISSN 0003-4975.

Dore, A., Houde, C., Chan, K-L., Ducharme, A., Khairy, P., Juneau, M., Marcotte, F., & Mercier, L-A. (2005). Angiotensin Receptor Blockade and Exercise Capacity in Adults with Systemic Right Ventricles: A Multicenter, Randomized, Placebo-Controlled Clinical Trial. *Circulation*, Vol. 112, No. 16, (October 2005), pp. 2411-2416, ISSN 1524-4539.

Duncan, B.W. & Mee, R.B. (2005). Management of the failing systemic right ventricle. *Seminars in Thoracic and Cardiovascular Surgery: Pediatric Cardiac Surgery Annual*, Vol. 17, No. 2, (Summer 2005), pp. 160-169, ISSN 1092-9126.

Fratz, S., Hager, A., Busch, R., Kaemmerer, H., Schwaiger, M., Lange, R., Hess, J., & Stern, H.C. (2008). Patients After Atrial Switch Operation for Transposition of the Great Arteries Can Not Increase Stroke Volume Under Dobutamine Stress as Opposed to Patients with Congenitally Corrected Transposition. *Circulation Journal*, Vol. 72, No. 7, (July 2008), pp. 1130-1135, ISSN 1347-4820.

Friedberg, D.Z. & Nadas, A.S. (1970). Clinical profile of patients with congenital corrected transposition of the great arteries: A study of 60 cases. *The New England Journal of Medicine*, Vol. 282, No. 19, (May 1970), pp. 1053-1059, ISSN 1533-4406.

Gaies, M.G., Goldberg, C.S., Ohye, R.G., Devaney, E.J., Hirsch, J.C., & Bove, E.L. (2009). Early and Intermediate Outcome After Anatomic Repair of Congenitally Corrected Transposition of the Great Arteries. *Annals of Thoracic Surgery*, Vol. 88, No. 6, (December 2009), pp. 1952-1960, ISSN 0003-4975.

Gelson, E., Curry, R., Gatzoulis, M.A., Swan, L., Lupton, M., Durbridge, J., Deans, C., Steer, P., & Johnson, M.R. (2011). Pregnancy in women with a systemic right ventricle after surgically and congenitally corrected transposition of the great arteries. *European Journal of Obstetrics and Gynecology and Reproductive Biology*, Vol. 155, No. 2, (April 2011), pp. 146-149, ISSN 0301-2115.

Giardini, A., Lovato, L., Donti, A,. Formigari, R., Oppido, G., Gargiulo, G., Picchio, F.M., & Fattori, R. (2006). Relation Between Right Ventricular Structural Alterations and Markers of Adverse Clinical Outcome in Adults with Systemic Right Ventricle and Either Congenital Complete (After Senning Operation) or Congenitally Corrected Transposition of the Great Arteries. *The American Journal of Cardiology*, Vol. 98, No. 9, (November 2006), pp. 1277-1282, ISSN 0002-9149.

Graham, T.P., Bernard, Y.D., Mellen, B.G., Celermajer, D., Baumgartner, H., Cetta, F., Connolly, H.M., Davidson, W.R., Dellborg, M., Foster, E., Gersony, W.M., Gessner, I.H., Hurwitz, R.A., Kaemmerer, H., Kugler, J.D., Murphy, D.J., Noonan, J.A.,

Morris, C., Perloff, J.K., Sanders, S.P., & Sutherland, J.L. (2000). Long-Term Outcome in Congenitally Corrected Transposition of the Great Arteries: A Multi-Institutional Study. *Journal of the American College of Cardiology*, Vol. 36, No. 1, (July 2000), pp. 255-261, ISSN 1558-3597.

Graham, T.P. & Markham, L.W. (2010). Congenitally corrected transposition of the great arteries, In: *Diagnosis and Management of Adult Congenital Heart Disease*, M.D. Gatzoulis & G.D. Webb (Eds.), pp. 371-377, Churchill Livingstone, ISBN 978-0-7020-3426-8, Edinburgh.

Graham, T.P., Markham, L., Parra, D.P., & Bichell, D. (2007). Congenitally Corrected Transposition of the Great Arteries: An Update. *Current Treatment Options in Cardiovascular Medicine*, Vol. 9, No. 5, (October 2007), pp. 405-413, ISBN 1092-8464.

Hoffman, J.I.E. (2009). Congenitally Corrected Transposition of the Great Arteries, In: *The Natural and Unnatural History of Congenital Heart Disease*, Julien I.E. Hoffman (Ed.), pp. 206-217, Wiley-Blackwell, ISBN 978-1-4051-7927-0, Oxford.

Hörer, J., Schreiber, C., Krane, S., Prodan, Z., Cleuziou, J., Vogt, M., Holper, K., & Lange, R. (2008). Outcome after Surgical Repair/Palliation of Congenitally Corrected Transposition of the Great Arteries. *The Thoracic and Cardiovascular Surgeon*, Vol. 56, No. 7, (October 2008), pp. 391-397, ISSN 1439-1902.

Hörer, J., Haas, F., Cleuziou, J., Schreiber, C., Kostolny, M., Vogt, M., Holper, K., & Lange, R. (2007). Intermediate-term results of the Senning or Mustard procedures combined with the Rastelli operation for patients with discordant atrioventricular connections associated with discordant ventriculoarterial connections or double outlet right ventricle. *Cardiology in the Young*, Vol. 17, No. 2, (April 2007), pp. 158-165, ISSN 1047-9511.

Hornung, T.S. & Calder, L. (2010). Congenitally corrected transposition of the great arteries. *Heart*, Vol. 96, No. 14, (July 2010), pp. 1154-1161, ISSN 1468-201X.

Hraska, V., Duncan, B.W., Mayer, J.E., Freed, M., del Nido, P.J., & Jonas RA. (2005). Long-term outcome of surgically treated patients with corrected transposition of the great arteries. *The Journal of Thoracic and Cardiovascular Surgery*, Vol. 129, No. 1, (January 2005), pp. 182-191, ISSN 0022-5223.

Huhta, J.C., Maloney, J.D., Ritter, D.G., Ilstrup, D.M., & Feldt, R.H. (1983). Complete atrioventricular block in patients with atrioventricular discordance. *Circulation*, Vole. 67, No. 6, (June 1983), pp. 1374–1377, ISSN 1524-4539.

Ismat, F.A., Baldwin, H.S., Karl, T.R., & Weinberg, P.M. (2002). Coronary anatomy in congenitally corrected transposition of the great arteries. *International Journal of Cardiology*, Vol. 86, No. 2-3, (December 2002), pp. 207-216, ISSN 0167-5273.

Janousek, J., Tomek, V., Chaloupecky, V. A., Reich, O., Gebauer, R. A., Kautzner, J., & Hucin, B. (November 2004). Cardiac resynchronization therapy: a novel adjunct to the treatment and prevention of systemic right ventricular failure. *Journal of the American College of Cardiology*, Vol. 44, No. 9, (November 2004), pp. 1927-1931, ISSN 1558-3597.

Jennings, H.S., Primm, R.K., Parrish, M.D., Prager, R.L., & Graham, T.P. (1984). Coronary arterial revascularization in an adult with congenitally corrected transposition. *American Heart Journal*, Vol. 108, (September 1984), pp. 598-600, ISSN 0002-8703.

Jonas, R.A. (2004). Congenitally corrected transposition of the great arteries, In: *Comprehensive Surgical Management of Congenital Heart Disease*, Richard. A. Jonas (Ed.), pp. 483-496, Hodder Arnold, ISBN 978-0-3408-0807-8, London.

Kordybach, M., Kowalski, M., & Hoffman, P. (2009). Heart failure in a patient with corrected transposition of the great arteries. When is biventricular pacing indicated? *Acta Cardiologica*, Vol. 64., No 5., (October 2009), pp. 673-676, ISSN 0001-5385.

Kral Kollars, C.A., Gelehrter, S., Bove, E.L., & Ensing, G. (2010). Effects of Morphologic Left Ventricular Pressure on Right Ventricular Geometry and Tricuspid Valve Regurgitation in Patients with Congenitally Corrected Transposition of the Great Arteries. *The American Journal of Cardiology*, Vol. 105, No. 5, (March 2010), pp. 735-739, ISSN 0002-9149.

Lim, H-G., Lee, J.R., Kim, Y.J., Park, Y-H., Jun, T-G., Kim, W-H., Lee, C-H., Park, H.K., Yang, J-H., Park, C-S., & Kwak, J.G. (2010). Outcomes of Biventricular Repair for Congenitally Corrected Transposition of the Great Arteries. *Annals of Thoracic Surgery*, Vol. 89, No. 1, (January 2010), pp. 159-167, ISSN 0003-4975.

Ly, M., Belli, E., Leobon, B., Kortas, C., Grollmüss, O.E., Piot, D., Planché, C., & Serraf, A. (2009). Results of the double switch operation for congenitally corrected transposition of the great arteries. *European Journal of Cardio-thoracic Surgery*, Vol. 35, No. 5, (May 2009), pp.879-884 , ISSN 1010-7940.

Malecka, B., Bednarek, J., Tomkiewicz-Pajak, L., Klimeczek, P., Zabek, A., Pasowicz, M., & Lelakowski, J. (2010). Resynchronization therapy transvenous approach in dextrocardia and congenitally corrected transposition of great arteries. *Cardiology Journal*, Vol. 17, No. 5, (2010), pp. 503-508, ISSN 1897-5593.

Malhotra, S.P., Reddy, V.M., Qui, M., Pirolli, T.J., Barboza, L., Reinhartz, O., & Hanley, F.L. (2011). The hemi-Mustard/bidirectional Glenn atrial switch procedure in the double-switch operation for congenitally corrected transposition of great arteries: Rationale and midterm results. *The Journal of Thoracic and Cardiovascular Surgery*, Vol. 141, No. 1, (Janurary 2011), pp. 162-170, ISSN 0022-5223.

McEwing, R.L. & Chaoui, R. (2004). Congenitally corrected transposition of the great arteries: clues for prenatal diagnosis. *Ultrasound in Obstetrics and Gynecology*, Vol. 23, No. 1, (January 2004), pp. 68-72, ISSN 1469-0705.

Metton, O., Gaudin, R., Ou, P., Gerelli, S., Mussa, S., Sidi, D., Vouhé, P., & Raisky, O. (2010). Early prophylactic pulmonary artery banding in isolated congenitally corrected transposition of the great arteries. *European Journal of Cardio-thoracic Surgery*, Vol. 38, No. 6, (December 2010), pp. 728-735, ISSN 1010-7940.

Mongeon, F-P., Connolly H.M., Dearani, J.A., Li, Z., & Warnes, C.A. (2011). Congenitally Corrected Transposition of the Great Arteries: Ventricular Function at the Time of Systemic Atrioventricular Valve Replacement Predicts Long-Term Ventricular Function. *Journal of the American College of Cardiology*, Vol. 57, No. 20, (May 2011), pp. 2008-2017, ISSN 1558-3597.

Oechslin, E. (2009). Physiologically "Corrected" Transposition of the Great Arteries, In *Echocardiography in Pediatric and Congenital Heart Disease: From Fetus to Adult*, W.W. Lai, L.L. Mertens, M.S. Cohen & T. Geva, (Eds.), pp. 439-455, Wiley-Blackwell, ISBN 978-1-4051-7401-5, Oxford.

Orchard, E.A., Ormerod, O., Myerson, S., & Westaby, S. (2010). Congenitally Corrected Transposition of the Great Arteries Presenting in a Nonagenarian. *Circulation*, Vol. 122, No. 9, (August 2010), pp. e441-444, ISSN 1524-4539.

Paladini, D., Volpe, P., Marasini, M., Russo, M.G., Vassallo, M., Gentile, M., & Calabro, R. (2006). Diagnosis, characterization and outcome of congenitally corrected transposition of the great arteries in the fetus: a multicenter series of 30 cases.

Ultrasound in Obstetrics and Gynecology, Vol. 27, No. 3, (March 2006), pp. 281-285, ISSN 1469-0705.

Piacentini, G., Digilio M.C., Capolino, R., De Zorzi, A., Toscano, A., Sarkozy, A., D'Agostino, R., Marasini, M., Russo, M.G., Dallapiccola, B., & Marino, B. (2005). Familial Recurrence of Heart Defects in Subjects with Congenitally Corrected Transposition of the Great Arteries. *American Journal of Medical Genetics,* Vol. 137, No. 2, (August 2005), pp. 176-180, ISSN 1552-4833.

Poirier, N.C., Yu, J.H., Brizard, C.P., & Mee, R.B. (2004). Long-term results of left ventricular reconditioning and anatomic correction for systemic right ventricular dysfunction after atrial switch procedures. *The Journal of Thoracic and Cardiovascular Surgery*, Vol. 127, No. 4, (April 2004), pp. 975-981, ISSN 0022-5223.

Presbitero, P., Somerville, J., Rabajoli, F., Stone, S., & Conte, M.R. (1995). Corrected transposition of the great arteries without associated defects in adult patients: clinical profile and follow up. *British Heart Journal*, Vol. 74, No. 1, (July 1995), pp. 57-59, ISSN 0007-0769.

Prieto, L.R., Hordof, A.J., Secic, M., Rosenbaum, M.S., & Gersony, W.M. (1998). Progressive Tricuspid Valve Disease in Patients With Congenitally Corrected Transposition of the Great Arteries. *Circulation*, Vol. 98, No. 10, (September 1998), pp. 997-1005, ISSN 1524-4539.

Quinn, D.W., McGuirk, S.P., Metha, C., Nightingale, P., de Giovanni, J.V., Dhillon, R., Miller, P., Stumper, O., Wright, J.G., Barron, D.J., & Brawn, W.J. (2008). The morphologic left ventricle that requires training by means of pulmonary artery banding before the double-switch procedure for congenitally corrected transposition of the great arteries is at risk of late dysfunction. *The Journal of Thoracic and Cardiovascular Surgery*, Vol. 135, No. 5, (May 2008), pp. 1137-1144, ISSN 0022-5223.

Said, S.M., Burkhardt, H.M., Schaff, H.V., & Dearani, J.A. (2011). Congenitally Corrected Transposition of Great Arteries: Surgical Options for the Failing Ventricle and/or Severe Tricuspid Regurgitation. *World Journal for Pediatric and Congenital Heart Surgery*, Vol. 2, No. 1, (January 2011), pp. 64-79, ISSN 2150-1351.

Scardi, S., Knoll, P., & Pandullo, C. (1999). Corrected Transposition of the Great Vessels and Situs Inversus Viscerum in a 65-Year-Old Oligosymptomatic Woman. *Circulation*, Vol. 100, No. 7, (August 1999), pp. 777, ISSN 1524-4539.

Schmidt, M., Theissen, P, Deutsch, H.J., Dederichs, B., Franzen, D., Erdmann, E., & Schicha, H. (2000). Congenitally corrected transposition of the great arteries (L-TGA) with situs inversus totalis in adulthood: findings with magnetic resonance imaging. *Magnetic Resonance Imaging*, Vol. 18, No. 4, (May 2000), pp. 417-422, ISSN 0730-725X.

Shima, Y., Nakajima, M., Kumasaka, S., & Migita, M. (2009). Prenatal diagnosis of isolated congenitally corrected transposition of the great arteries. *Archives of Gynecology and Obstetrics*, Vol. 279, No. 4, (April 2009), pp. 557-559, ISSN 1432-0711.

Shin'oka, T., Kurosawa, H., Imai, Y., Aoki, M., Ishiyama, M., Sakamoto, T., Miyamoto, S., Hobo, K., & Ichihara, Y. (2007). Outcomes of definitive surgical repair for congenitally corrected transposition of the great arteries or double outlet right ventricle with discordant atrioventricular connections: risk analyses in 189 patients. The Journal of thoracic and cardiovascular surgery, Vol. 133, No. 5, (May 2007), pp. 1318-28, ISSN 0022-5223.

Takemoto, M., Nakashima, A., Muneuchi, J., Yamamura, K-I., Shiokawa, Y., Sunagawa, K., & Tominaga, R. (2010). Para-Hisian Pacing for a Pediatric Patient with a

Congenitally Corrected Transposition of the Great Arteries (SLL). *Pacing and Clinical Electrophysiology*, Vol. 33, No. 1, (January 2010), pp. e4-7, ISSN 1540-8159.

Tay, E.L., Frogoudaki, A., Inuzuka, R., Giannakoulas, G., Prapa, M., Li, W., Pantley, G., Dimopoulos, K., & Gatzoulis, M.A. (2011). Exercise intolerance in patients with congenitally corrected transposition of the great arteries relates to right ventricular filling pressures. *International Journal of Cardiology*, Vol. 147, No. 2, (March 2011), pp. 219-223, ISSN 0167-5273.

Teo, L.L. & Hia, C.P. (2011). Advanced cardiovascular imaging in congenital heart disease. *International Journal of Clinical Practice*, Vol. 65, Suppl. 171, (April 2011), pp. 17-21, ISSN 1742-1241.

Thierrien, J., Barnes, I., & Somerville, J. (1999). Outcome of Pregnancy in Patients with Congenitally Corrected Transposition of the Great Arteries. *The American Journal of Cardiology*, Vol. 84, No.7, (October 1999), pp. 820-824, ISSN 0002-9149.

Van der Bom, T., Zomer, A.C., Zwinderman, A.H., Meijboom, F.J., Bouma, B.J., & Mulder, B.J. (2011). The changing epidemiology of congenital heart disease. *Nature Reviews Cardiology*, Vol. 8, No. 1, (January 2011), pp. 50-60, ISSN 1759-5002.

Van Praagh, R., Papagiannis, J., Grunenfelder, J., Bartram, U., & Martanovic, P. (1998) Pathologic anatomy of corrected transposition of the great arteries and surgical implications. *American Heart Journal*, Vol 135, No. 5, (May 1998), pp. 772-785, ISSN 0002-8703.

Van Son, J.A, Danielson, G. K., Huhta, J. C., Warnes, C. A., Edwards, W. D., Schaff, H. V., Puga, F. J., & Ilstrup, D. M. (1995). Late results of systemic atrioventricular valve replacement in corrected transposition. *The Journal of Thoracic and Cardiovascular Surgery*, Vol. 109, No. 4, (April 1995), pp. 642-653, ISSN 0022-5223.

Wan, A.W., Jevremovic, A., Tierney, E.S., McCrindle, B.W., Dunn, E., Manlhiot, C., Wald, R.M., Kingdom, J.P., Jaeggi, E.T., & Nield L.E. (2009). Comparison of Impact of Prenatal Versus Postnatal Diagnosis of Congenitally Corrected Transposition of the Great Arteries. *The American Journal of Cardiology*, Vol. 104, No. 9, (November 2009), pp. 1276-1279, ISSN 0002-9149.

Warnes, C.A. (2006). Transposition of the Great Arteries. *Circulation*, Vol. 114, No. 24, (December 2006), pp. 2699-2709, ISSN 1524-4539.

Winlaw, D.S., McGuirk, S.P., Balmer, C., Langley, S.M., Griselli, M., Stümper, O., De Giovanni, J.V., Wright, J.G., Thorne, S., Barron, D.J., Brawn, W.J. (2005). Intention-to-Treat Analysis of Pulmonary Artery Banding in Conditions With a Morphologic Right Ventricle in the Systemic Circulation With a View to Anatomic Biventricular Repair. *Circulation*, Vol. 111, No. 4, (February 2005), pp. 405-411, ISSN 1524-4539.

Winter, M.M., Bouma, B.J., Groenink, M., Konings, T.C., Tijssen, J.G., van Veldhuisen, D.J., & Mulder, BJ. (2009). Latest insights in therapeutic options for systemic right ventricular failure: a comparison with left ventricular failure. *Heart*, Vol. 5, No. 1, (June 2009), pp. 960-963, ISSN 1468-201X.

Zhang, Y., Cai, A., Sun, W., & Zhao, Y. (2011). Prenatal diagnosis of fetal congenitally corrected transposition of the great arteries. *Prenatal Diagnosis*, Vol. 31, No. 6, (June 2011), pp. 529-535, ISSN 1097-0223.
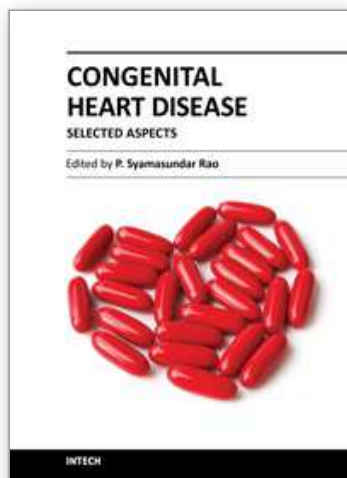
Congenital Heart Disease - Selected Aspects

Edited by Prof. P. Syamasundar Rao

There are significant advances in the understanding of the molecular mechanisms of cardiac development and the etiology of congenital heart disease (CHD). However, these have not yet evolved to such a degree so as to be useful in preventing CHD at this time. Developments such as early detection of the neonates with serious heart disease and their rapid transport to tertiary care centers, availability of highly sensitive noninvasive diagnostic tools, advances in neonatal care and anesthesia, progress in transcatheter interventional procedures and extension of complicated surgical procedures to the neonate and infant have advanced to such a degree that almost all congenital cardiac defects can be diagnosed and "corrected". Treatment of the majority of acyanotic and simpler cyanotic heart defects with currently available transcatheter and surgical techniques is feasible, effective and safe. The application of staged total cavo-pulmonary connection (Fontan) has markedly improved the long-term outlook of children who have one functioning ventricle. This book, I hope, will serve as a rich source of information to the physician caring for infants, children and adults with CHD which may help them provide optimal care for their patients.

How to reference

In order to correctly reference this scholarly work, feel free to copy and paste the following:

English C. Flack and Thomas P. Graham (2012). Congenitally Corrected Transposition of the Great Arteries, Congenital Heart Disease - Selected Aspects, Prof. P. Syamasundar Rao (Ed.), ISBN: 978-953-307-472-6, InTech, Available from: http://www.intechopen.com/books/congenital-heart-disease-selected-aspects/congenitally-corrected-transposition-of-the-great-arteries

INTECH
open science | open minds