Transposition of the Great Arteries

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Abstract—Many patients with ventriculoarterial discordance have survived to adulthood. Those with complete transposition of the great arteries have often had an atrial switch procedure (Mustard or Senning operation) performed, which leaves the morphological right ventricle (RV) supporting the systemic circulation. RV failure and tricuspid regurgitation are common. Some patients may ultimately require cardiac transplantation. Sinus node dysfunction is increasingly common with longer follow-up, and some patients need pacemaker implantation. Atrial arrhythmias are frequent, and atrial flutter may be a marker for sudden death. Patients with an atrial switch procedure are also surviving to adulthood. Long-term problems include coronary stenoses, distortion of the pulmonary arteries, dilatation of the neoaortic root, and aortic regurgitation. Patients with congenitally corrected transposition have both atrioventricular and ventriculoarterial discordance and therefore also have a morphological RV and delicate tricuspid valve in the systemic circulation. Associated defects, such as abnormalities of the tricuspid valve, ventricular septal defect, and pulmonary stenosis, occur in the majority of patients. Heart block occurs with increasing age. Atrial arrhythmias occur frequently, and their occurrence should prompt a search for a hemodynamic problem. Progressive tricuspid regurgitation occurs with age and is associated with deterioration of RV function. Surgical treatment should be considered at the earliest sign of RV dilatation or dysfunction. All patients should be seen periodically in a center where expertise in the clinical evaluation, imaging, and hemodynamic assessment of adult congenital heart disease is available. (Circulation. 2006;114:2699-2709.)

Key Words: transposition of great vessels ■ heart defects, congenital ■ pediatrics ■ cardiovascular diseases

Many patients with transposition complexes have survived to adulthood. Those with complete transposition (d-transposition) have survived because of prior radical repair and are seen increasingly in cardiology practice. Residua and sequelae are common, and morbidity and mortality are ongoing. In contrast, although some patients with congenitally corrected transposition (C-TGA) have had surgery in childhood, others may present for the first time in adulthood, and the diagnosis is often overlooked. This review will focus on the presentation, evaluation, and management of these 2 complex lesions.

Glossary of Terms

Atrioventricular discordance: Inappropriate connections of the morphological right atrium to the morphological left ventricle and morphological left atrium to right ventricle.

Ventriculoarterial discordance: The pulmonary artery arises from a morphological left ventricle, and the aorta arises from a morphological right ventricle.

Transposition of the great arteries: Refers to ventriculoarterial discordance. The aorta arises from a morphological right ventricle, and the pulmonary artery arises from a morphological left ventricle.

D-loop: Refers to the normal rightward (dextro=D) loop or bend of the embryonic heart tube and indicates that the inflow portion of the right ventricle is to the right of the morphological left ventricle.

L-loop: Refers to a leftward (levo=L) loop or bend of the embryonic heart tube resulting in the inflow portion of the morphological right ventricle being to the left of the morphological left ventricle.

Cardiac chambers: Morphological right and left ventricles refer to the anatomic characteristics of the chambers and not their positions (eg, the morphological right ventricle is on the left in congenitally corrected transposition).

Congenitally corrected transposition: Atrioventricular discordance and ventriculoarterial discordance. The right atrium enters the left ventricle, which gives rise to the pulmonary artery, and the left atrium enters the right ventricle, which gives rise to the aorta. Thus, the circulation continues in the appropriate direction but flows through the “wrong” ventricles.

Complete Transposition of the Great Arteries

In this anomaly, the aorta arises from the morphological right ventricle (RV), and the pulmonary artery arises from the morphological left ventricle (LV) (ie, there is ventriculoarterial discordance). Complete transposition of the great arteries...
is also known as d-TGA; the “d-” refers to the dextroposition of the bulboventricular loop (ie, the position of the RV, which is on the right side). The aorta also tends to be on the right and anterior, and the great arteries are parallel rather than crossing as they do in the normal heart. Because the systemic and pulmonary circulations run in parallel, there has to be a communication between the 2, either with an atrial septal defect, a ventricular septal defect (VSD), or at the great arterial level (patent ductus arteriosus) to support life. These connections allow systemic blood to enter the pulmonary circulation for oxygenation and allow oxygenated blood from the pulmonary circuit to enter the systemic circulation. The most common associated lesions are VSD, which occurs in almost half of the cases, pulmonary outflow tract obstruction, and, less commonly, coarctation of the aorta (≈5%).

D-TGA is one of the most common cyanotic defects seen in newborns, and when the ventricular septum is intact, it is usually cyanotic in the first day of life. If circulatory mixing occurs via a patent ductus, physiological closure of the ductus causes abrupt cyanosis and clinical deterioration. Cyanotic babies may be treated percutaneously with a Rashkind atrial balloon septostomy to create a more sizable atrial septal defect, which may dramatically improve their oxygenation until definitive surgery can be performed. Almost all patients with this defect who reach adulthood have had prior reparative cardiac surgery, although some with a large VSD and pulmonary vascular disease sometimes survive with Eisenmenger physiology.

**Atrial Switch Procedure**

The first atrial switch procedure was performed by Senning in 1958 and involves the creation of an atrial baffle from autologous tissue to direct the venous return to the contralateral atrioventricular (AV) valve and ventricle. Thus, deoxygenated blood from the vena cavae is directed to the mitral valve and LV and thence to the pulmonary artery, and pulmonary venous blood is directed into the morphological RV and into the aorta. An alternative operation was subsequently developed by Mustard, who excised the atrial septum and used synthetic material to create the baffle (Figures 1 and 2). Sometimes an atrial switch is preceded by either the Rashkind balloon atrial septostomy or surgical atrial septectomy (Blalock-Hanlon). Both atrial switch procedures provide excellent midterm clinical results but in the long term are associated with important sequelae.

**Arrhythmias**

Late development of both atrial bradyarrhythmias and tachyarrhythmias is a recognized late complication of atrial baffle surgery and is more likely to occur with longer follow-up. Sinus node dysfunction is common in adults, and the cause is probably damage to the sinus node and atrial conduction tissue or interruption of sinus node blood flow at the time of operation. In the series of 534 children by Gelatt et al, sinus rhythm was present in 77% at 5 years and only 40% at 20 years. Fifty-three patients (11%) needed pacemaker implantation. Pacemaker implantation in this setting can be technically challenging because of the complex anatomy. Atrial flutter was also fairly common, occurring in 14% of the survivors, and, as others have reported, is a probable marker for sudden death. There were 77 late deaths (16%), with unexpected sudden death occurring in 31 patients at a median of 3.2 years after the operation. Sudden presumed arrhythmic death and myocardial failure were the most frequent causes of late death, a finding that agrees with other reports. Atrial arrhythmias can precipitate a significant deterioration in systemic ventricular function, and, in general, sinus rhythm should be restored when possible. Judicious doses of antiarrhythmic drugs should be used because aggressive therapy may precipitate heart block. Radiofrequency ablation for atrial arrhythmias is technically more challenging because of the complex anatomy, atrial scar, and presence of artificial tissue. The reported success
rates are ≈70%. Ventricular arrhythmias are uncommon in the absence of severe ventricular dysfunction.

Systemic Ventricular Dysfunction and Tricuspid Regurgitation

Late RV dysfunction is a recognized outcome after the Mustard or Senning procedures because the RV is still the systemic ventricle. Tricuspid regurgitation often coexists, and both lesions increase the propensity for atrial arrhythmias. Mild to moderate tricuspid regurgitation is very common in adult survivors and tends to progressively worsen. Occasionally the tricuspid valve apparatus may be intrinsically abnormal or may have been damaged at the time of prior VSD repair or by endocarditis. In this circumstance, tricuspid valve replacement may be warranted, but in most cases the regurgitation is secondary to annular dilatation from RV failure, and tricuspid valve replacement is not helpful.

The treatment of systemic ventricular dysfunction is challenging. Although it is tempting to extrapolate the use of angiotensin-converting enzyme inhibitors and β-blockers from trials of acquired heart disease to patients after an atrial switch procedure, convincing data that they increase exercise time, ventricular function, or survival do not exist. Nonetheless, angiotensin-converting enzyme inhibitors are often used empirically. Caution should be exercised with the use of β-blockers, which might exacerbate AV block and precipitate bradycardia.

Many young patients have survived to their 20s and 30s, and an actuarial survival of 80% at 20 years has been reported from a single-center experience. Late attrition from RV failure and arrhythmias is an ongoing issue, however, that warrants continued follow-up. The long-term prognosis must be guarded, and in later decades it is possible that many will require cardiac transplantation.

The assessment of RV function is challenging, and the most commonly used modality is still 2-dimensional echocardiography, albeit qualitative rather than quantitative because of the complex geometry of the RV. Other echo-Doppler parameters can also be followed, such as the index of myocardial performance and the dP/dT with the use of the tricuspid regurgitant velocity. More recently, a tissue Doppler measurement of myocardial acceleration during isovolumic contraction, “isovolumic myocardial acceleration,” has been reported and may be a sensitive, noninvasive method of assessing RV contractility, which is less load-dependent than other parameters. Other imaging modalities include radionuclide methods, angiography, or magnetic resonance imaging (MRI).

Although the functional status of many young adults is good, reduced exercise capacity is the norm. This may relate to chronotropic incompetence, impaired systemic ventricular function, or impaired capacitance and conduit function of the intra-atrial conduit, which may be fibrotic or calcified and produce fixed ventricular filling rates.

Late Surgical Strategies

As an alternative to medical therapy for the failing RV, consideration has been given to the restoration of the RV to the subpulmonary position in the hope that, in the face of reduced afterload, its function will improve. This involves “retraining” of the LV, however, before it can assume function as a systemic pump, and therefore the pulmonary artery must be banded to facilitate hypertrophy of the LV. If successful, the atrial baffle may subsequently be taken down and an arterial switch operation performed. This approach has been successful in some young patients but is more problematic in adults. In addition to the technical challenges of the extensive surgery, the LV appears less able to respond to the increased afterload, and ventricular failure occurs after the pulmonary banding. In a series of 35 patients with pulmonary banding, 10 failed retraining, and this was more common in patients aged >12 years. Whether a slower, more physiological banded process might be more effective in retraining the LV in adults remains to be seen.

Because pulmonary banding increases the LV pressure, a rightward “septal shift” may occur with increased coaptation of the systemic AV valve and reduction in systemic AV valve regurgitation. Some authors have advocated pulmonary banding as a therapy for the failing RV in its own right, but whether this is an effective palliation has yet to be determined. In many centers, these surgical approaches have been abandoned in adults in favor of cardiac transplantation.

Atrial Baffle Obstruction andLeaks

Scarring and narrowing of the atrial baffle are infrequent late complications but should always be investigated. Obstruction of the superior vena cava is more common than that of the inferior vena cava and may produce “SVC syndrome.” Imaging of the superior limb before the implantation of an endocardial pacing system will ensure that the lead will not cause obstruction and possible thrombosis. This imaging can...
be accomplished by transthoracic or transesophageal echocardiography, angiography, or MRI. Inferior vena cava obstruction is less common but in rare cases may cause hepatic congestion or cirrhosis. Both limbs can often be assessed by transthoracic Doppler echocardiography. Mild stenosis can be treated conservatively, but significant stenosis may need percutaneous balloon and stent placement or even surgical intervention.

Pulmonary vein stenosis is much less common but may cause pulmonary hypertension. It can be treated percutaneously or surgically. Small baffle leaks are more common than obstruction and are often visualized only by transesophageal echocardiography. They are usually small and hemodynamically insignificant but pose a risk of paradoxical embolus and cerebrovascular event in the setting of tachyarrhythmias or an endocardial pacemaker. It is the author’s preference either to close the defect before implantation of a pacemaker or to place the leads epicardially because stroke may occur even with concomitant anticoagulation. Patients with a large shunt will require closure, performed either percutaneously or surgically.

**Pulmonary Hypertension**

Pulmonary hypertension is a serious complication of atrial switch repairs for d-TGA and occurs in approximately 7% of those who survive to adulthood. The exact cause is not completely clear, but patients appear more likely to develop pulmonary vascular disease when undergoing operation at >2 years of age. Other risk factors include having had shunts at the ventricular or great artery level before repair. Patients with mild elevation of pulmonary pressures at early postoperative catheterization appear at increased risk for developing pulmonary vascular disease. Pulmonary hypertension may also be caused by pulmonary venous baffle obstruction, which should always be excluded.

**Pregnancy**

Pregnancy may be well tolerated after an atrial switch procedure, but careful prepregnancy counseling and evaluation, including a frank discussion about the patient’s long-term prognosis, are critical. Attention should be paid to the function of the systemic RV and degree of tricuspid regurgitation because pregnancy poses a significant volume load that may increase RV dimensions and is sometimes irreversible. Pregnancy poses a definite risk of deterioration of functional class, even if the patient is reportedly asymptomatic before pregnancy. In 1 series, 2 of 16 pregnant women required heart failure therapy during or soon after pregnancy. The recurrence risk of congenital heart disease in the offspring, in the absence of a family history, is probably <5%, although no large series have been reported.

**Arterial Switch Procedure**

This operation restores the normal anatomic arrangement of the circulation and, as such, is a more attractive physiological long-term option. From its first description by Jatene in 1976, it has steadily become the procedure of choice when the anatomy is appropriate and is usually performed in the first month of life. It involves transection of the great arteries above the sinuses and detachment of the coronary arteries along with a “button” from the aortic wall. The great arteries are then switched into their new position, with the pulmonary artery brought forward anterior to the aorta and the coronary buttons sutured into the “neo-aorta.” Adult survivors of this procedure are only now appearing in adult congenital heart disease clinics. Complications include distortion of the RV outflow tract and pulmonary arteries and dilatation of the neo-aortic root with aortic regurgitation. Coronary stenoses may also occur and cause sudden death or myocardial infarction. Other coronary stenoses may require surgical or catheter intervention.

**Rastelli Procedure**

This operation is used when d-TGA coexists with a large subaortic VSD and pulmonary stenosis. A patch is placed to direct blood from the LV into the aorta. The pulmonary valve is oversewn, and a valved conduit is inserted from the RV to the pulmonary artery to bypass the pulmonary stenosis. Reproduced from Hornung et al with permission from Elsevier.
Arrhythmias should prompt a detailed evaluation of the hemodynamics.

Follow-Up

All patients, regardless of the type of prior surgical repair, should have a clinical evaluation yearly or every 2 years. Imaging by echocardiogram and/or MRI permits an anatomic and hemodynamic assessment, and exercise testing facilitates detection of arrhythmias, occult coronary disease, and serial functional capacity. Periodic Holter monitoring allows detection of sinus node disease and atrial arrhythmias, although it may be of little help in identifying those at risk of sudden death. Most patients need endocarditis prophylaxis unless they have had an atrial switch procedure and have no residual valve dysfunction or outflow tract disturbance.

Congenitally Corrected Transposition

C-TGA is a rare anomaly and comprises <1% of all forms of congenital heart disease. In adults, it represents a very heterogeneous patient population, some of whom may not have been identified until adulthood. Surprisingly, the diagnosis is often overlooked. In this anomaly, the right atrium enters the morphological LV, which gives rise to the pulmonary artery, and the left atrium communicates with the morphological RV, which gives rise to the aorta. Thus, AV and ventriculoarterial discordance exist, and although blood flows in the normal direction, it passes through the “wrong” ventricular chamber (Figure 4). Hence, this “double discordance” results in the term C-TGA, which is, in essence, a misnomer. It is also called L-transposition because the morphological RV is in the levoposition. The aorta is also usually, but not universally, anterior and to the left, and the great arteries may be side by side. Because the tricuspid valve always enters a morphological RV, it too is on the left side in the systemic circulation and is more appropriately termed the systemic AV valve (Figure 5).

Associated Anomalies

Most patients have 1 or more associated cardiac anomalies, and the presence or absence of these markedly alters the natural history.

Ventricular Septal Defect

A VSD occurs in 70% of patients, usually in the perimembranous location. Patients with a large VSD usually present in infancy or childhood with congestive heart failure.

Pulmonary Stenosis

Pulmonary stenosis occurs in ~40% of patients and is commonly subvalvular. It may result from an aneurysm of the interventricular septum or may be associated with fibrous tissue tags or a discrete ring of tissue in the subvalvular area. Associated valvar pulmonary stenosis also occurs. Patients who have both a VSD and pulmonary stenosis may be hemodynamically well balanced or have varying degrees of cyanosis.

Abnormalities of the Systemic (Tricuspid) AV Valve

Some abnormality of the systemic AV valve occurs in up to 90% of patients. Ebstein’s anomaly of the systemic AV valve

Figure 4. Schematic drawings to demonstrate normal AV and ventriculoarterial relationships (left) and those in C-TGA (right), in which both AV and ventriculoarterial discordance exist. In C-TGA, the normal great arterial relationships are lost, and the pulmonary trunk (PT) is no longer anterior to the aorta. Ao indicates aorta; LA, morphological left atrium; LV, morphological LV; RA, morphological right atrium; and RV, morphological RV.

Figure 5. Necropsy specimens showing AV concordance (left) and AV discordance (C-TGA, right). The crux anatomy facilitates recognition of AV morphology because the tricuspid valve is always lower (arrow) than the mitral valve and always enters a morphological RV. Although the ventricular morphology may be suggested by the more trabeculated pattern of the RV, this is not always consistent or easily identified. Reproduced, with kind permission of Springer Science and Business Media, from Seward JB, Tajik AJ, Edwards WD, Hagler DJ. Two-Dimensional Echocardiographic Atlas, Vol 1: Congenital Heart Disease. New York, NY: Springer-Verlag; 1987.
also occurs but is different from the typical right-sided Ebstein’s anomaly, the only real similarity being that the valve is displaced inferiorly closer to the cardiac apex.51 No large “saillike” anterior leaflet exists, adherence of the septal and posterior leaflets is limited, and the atrialized portion of the RV inflow is also relatively small. In addition, this systemic tricuspid valve can rarely be surgically repaired with success. These delicate valves are inherently vulnerable to developing regurgitation, which is an additional burden for the systemic morphological RV. Sometimes the tricuspid valve may straddle the ventricular septum across a VSD, making a biventricular repair much more difficult and sometimes impossible. Such patients may need single ventricle repair or palliation with a Fontan operation or bidirectional cavopulmonary anastomosis.

**Conduction System**
The AV node and His bundle have an unusual position and course, and many patients have dual AV nodes.52,53 The second anomalous AV node and bundle are usually anterior, and the long penetrating bundle is vulnerable to fibrosis with advancing age. This makes the conduction system somewhat tenuous, with a progressive incidence of complete AV block occurring at ≈2% per year. Tricuspid valve or VSD surgery may also precipitate heart block.

**Diagnostic Evaluation**

**Chest Radiograph**
With mesocardia or levocardia, the diagnosis may be suspected from the chest radiograph. The vascular pedicle appears abnormally straight because the normal arterial relationships are lost. The ascending aorta is not visible on the right side, and the convexities from the descending aortic knob and pulmonary artery are absent on the left side. The ventricular border on the left may also appear more vertical than usual (Figure 6A). C-TGA is one of the most common anomalies associated with dextrocardia and should be suspected when there is abdominal situs solitus (gastric bubble on the left) and dextrocardia (Figure 6B).

**Electrocardiogram**
The ECG may be misinterpreted as inferior myocardial infarction. Because of the ventricular inversion, the right and left bundles are also inverted, causing septal activation to occur from right to left. This produces Q waves in the right precordial leads and absent Q waves in the left precordial leads (Figure 7). Varying degrees of AV block are also common.

**Two-Dimensional Echocardiography**
Echocardiography can be difficult for those without experience of congenital heart disease, particularly because dextro-
cardia and mesocardia may occur. Subcostal imaging facilitates detection of atrial situs and position of the cardiac apex and thus determines the presence or absence of dextrocardia or mesocardia. Ventricular morphology can best be determined from short-axis and apical 4-chamber views. The morphological RV is on the patient’s left (I-loop), and although it has prominent trabeculations, it may be mistaken for a LV. Because the tricuspid valve is always more inferior (closer to the cardiac apex) than the mitral valve and always enters the morphological RV, an examination of the cardiac crux facilitates detection of the anomaly (Figure 8). Mitral-pulmonary fibrous continuity can be seen on the long-axis and 4-chamber views along with the abnormal ventriculoarterial connections. A high parasternal short-axis view may show the abnormal arterial relationships, with the aorta usually anterior and to the left of the pulmonary artery.

Figure 8. Apical 4-chamber view of a patient with C-TGA in the same tomographic plane as Figure 5. Pulmonary veins (dashed arrows) enter the left atrium (LA). The cardiac crux has a mirror-image appearance with the right-sided AV valve clearly inserting higher than the left-sided valve (arrows). The lower valve is the tricuspid valve, which enters the morphological RV, which has prominent trabeculations (arrowheads). AS indicates atrial septum; LV, morphological LV; and RA, right atrium. Reproduced, with kind permission of Springer Science and Business Media, from Seward JB, Tajik AJ, Edwards WD, Hagler DJ. Two-Dimensional Echocardiographic Atlas, Vol 1: Congenital Heart Disease. New York, NY: Springer-Verlag; 1987.

Cardiac Catheterization
Cardiac catheterization permits evaluation of systemic AV valve regurgitation and systemic ventricular function. A hemodynamic assessment of associated anomalies can be performed, including measurement of left and right heart pressures and pulmonary resistance. In older patients, coronary angiography is necessary before any operative intervention.

MRI/Radionuclide Imaging
MRI may help to define ventricular function and volumes, although it requires some expertise in imaging congenital heart disease. Because some patients have cardiac pacemakers, it is not always a feasible imaging modality. Radionuclide angiography may also be used.

Natural History/Long-Term Sequelae
The minority of patients may be relatively normal from a functional standpoint, and survival to the seventh and eighth decades has been reported when no associated anomalies exist. Failure of the systemic ventricle is much more common earlier in life, usually with concomitant tricuspid regurgitation.

Systemic Ventricular Failure and AV Valve Regurgitation
There has been much debate about the cause of systemic ventricular failure. The coronary anatomy is concordant, and therefore the morphological RV is perfused by a single right coronary artery. In such a situation, there may be limitations of myocardial perfusion and a mismatch between oxygen supply and demand. In a study of 20 patients in which exercise sestamibi imaging was used, all 20 had perfusion defects at rest, and in 17 these defects worsened with exercise. In a study in which positron emission tomography imaging was used, myocardial blood flow in the systemic ventricle was normal at rest, but the hyperemia resulting from adenosine-induced vasodilatation was significantly less than in the controls, suggesting that coronary flow reserve is greatly attenuated.

A retrospective multi-institutional study clearly demonstrated an increasing incidence of systemic ventricular dysfunction and clinical congestive heart failure with advancing age. Even in patients with C-TGA and no significant associated lesions, more than one third had congestive heart failure by the fifth decade. In patients with significant associated defects and prior open heart surgery, two thirds of patients had congestive heart failure by the age of 45 years. In addition to the morphological RV’s inherent vulnerability to failure, it has a complex relationship with systemic AV valve regurgitation, and controversy exists as to which is the “chicken” and which is the “egg.” In general, however, it appears that primary RV failure, while uncommon, is a frequent sequel to systemic AV valve regurgitation. Thus, when systemic ventricular function deteriorates, AV valve regurgitation should be considered the explanation until proof of a different explanation can be provided, and surgery should...
be considered early, before irreversible ventricular function ensues.

Late referral is common. In the Mayo Clinic series of 44 patients aged 20 to 79 years with no prior cardiac surgery, 26 patients (59%) were referred with significant (>3/4) AV valve regurgitation.49 Of 30 patients who needed systemic AV valve replacement, 16 were referred late with clinical ventricular dysfunction >6 months. The mean preoperative ejection fraction was 39%, which would be considered unacceptably late in patients with normal ventricular anatomy and mitral regurgitation. Although there was no early mortality, 4 patients ultimately needed cardiac transplantation for poor ventricular function, and the only marker for poor survival was a poor preoperative ejection fraction.49

Because complete heart block is common, it should be noted that implantation of an endocardial pacemaker may also precipitate deterioration in systemic ventricular function and worsening of AV valve regurgitation. This procedure presumably alters the position of the ventricular septum, inducing “septal shift” and failure of tricuspid valve coaptation64 (Figure 9).

Surgical Repair

Systemic AV valve replacement is relatively common in the adult age group. Surgery can be accomplished with acceptable perioperative mortality in experienced centers. The outcomes of 40 patients aged 5 months to 70 years who underwent systemic AV valve replacement at the Mayo Clinic between 1964 and 1993 were reported by van Son.59 Of these 40 patients, 36 had severe regurgitation, and 29 (72.5%) had associated cardiac anomalies. The preoperative ejection fraction ranged from 20% to 60% (mean, 48%). The in-hospital mortality in this high-risk group was 10%, but with follow-up to 26 years (mean, 4.7), another 8 patients died. The survival was 78% at 5 years and 61% at 10 years. The cause of death in all 12 patients in this series was systemic ventricular failure, an outcome which emphasizes the need for early operation before the development of ventricular dysfunction. Functional status improved in the 28 survivors, with 27 patients in New York Heart Association class I or II. Survivorship correlated with a preoperative ejection fraction >44%

Rutledge et al65 reported a pediatric series of 121 patients seen at Texas Children’s Hospital between 1952 and 1999 with a median age at diagnosis of 1 month. Asymptomatic patients had isolated C-TGA, a small VSD, or mild pulmonary stenosis, or they were hemodynamically well balanced with a VSD and pulmonary stenosis. In contrast, patients with heart failure had a large VSD, a VSD with mild pulmonary stenosis, or a regurgitant systemic AV valve. Those with cyanosis had a VSD and either pulmonary stenosis or pulmonary atresia. In a median follow-up of 9.3 years, the 5-, 10-, and 20-year survival rates were 92%, 91%, and 75%, respectively. There were 20 deaths (16.5%) at a median age of 13.2 years and 5 cardiac transplants (4%). Surgery was performed in 86 patients with an operative mortality of 2.5%. Biventricular repair was performed in 47 patients and varied according to the underlying cardiac anatomy. Risk factors for mortality included age at biventricular repair, moderate or severe systemic AV valve regurgitation, and poor RV function. In this study, as in others, most patients undergoing surgical repair had worsening systemic AV valve regurgitation to a moderate or severe degree. Implicated factors include the “insult” of cardiopulmonary bypass leading to annular dilatation, annular distortion caused by VSD closure, changes in the position of the ventricular septum, and annular dilatation after relief of pulmonary stenosis.68,69 In addition, poor tolerance of the systemic AV valve to systemic pressure after VSD closure is possible because afterload is increased, and postoperative complete AV block may compound the adverse hemodynamics.69–72 The observation of progressive systemic AV valve regurgitation after operative intervention has prompted some authors to recommend systemic AV valve replacement at the time of intracardiac repair if the systemic AV valve regurgitation is more than mild at the time of surgery.66

Hraska et al72 recently reported the outcomes of 123 patients with C-TGA who underwent a variety of surgical procedures. The 10-year survival was only 67%, and systemic ventricular dysfunction occurred in 44% of patients. Ventricular dysfunction was again closely linked to systemic AV valve regurgitation, particularly when Ebstein’s anomaly of the systemic AV valve was present. The authors also speculated that the change in RV geometry precipitated more tricuspid regurgitation, which in turn led to more annular dilatation and then more tricuspid regurgitation. They too were disappointed with the results of classic biventricular...
repair and suggested that alternative approaches should be explored, such as banding the pulmonary artery to increase LV pressure, which might maintain the geometry of the morphological RV and prevent progression of systemic AV valve regurgitation.

Interest has recently been expressed in a more anatomic repair that permits the morphological LV to function in the systemic circulation. This strategy would have the theoretical advantage of relieving the hemodynamic burden on the RV and tricuspid valve, potentially improving surgical results and longevity. Anatomic repair can be accomplished by using a venous switch procedure (either the Mustard or Senning technique), and, in those with a normal LV outflow tract, the arterial switch operation (Figure 10). For patients with a VSD, a patch can be inserted to tunnel the LV flow into the aorta, and the morphological RV is connected to the pulmonary artery via a conduit (Rastelli technique). Before anatomic repair, the LV needs to be “prepared” to function as a systemic ventricle, and therefore, in the absence of pulmonary stenosis that would have “trained” the LV to hypertrophy and function at a higher pressure than a venous ventricle, pulmonary banding may be performed. Concomitant tricuspid valve surgery can also be performed if necessary. Langley et al. reported the results of this approach in 54 patients aged 7 weeks to 40 years (median, 3.2 years) and noted a surgical mortality of 5.6% and 2 late deaths. Those with tricuspid regurgitation, however, still tended toward a poorer outcome: Of 18 patients with moderate to severe tricuspid regurgitation at the time of operation, 2 died early, 2 died late, and 1 had cardiac transplantation. Given the complexity of these operations, however, the early mortality appeared encouraging (and better than many other centers have experienced), but several reinterventions had to be performed. Seven patients with the Rastelli procedure needed a repeat conduit, aortic valve replacement, or transplantation. Percutaneous balloon dilatations for baffle obstruction or pulmonary stenosis were also needed. Survival at 9 years was 77%. Although most survivors are in New York Heart Association class I, aortic regurgitation in those with a double switch and residual LV dysfunction are among the ongoing concerns. In addition, atrial arrhythmias are common after venous baffle procedures and increase with longer follow-up. Whether older patients are suitable candidates for these procedures remains to be determined because the idea that the LV can be “trained” after childhood remains controversial. Some authors have reported failure of the LV after pulmonary banding even before adolescence. Late follow-up will be necessary to determine whether this management strategy confers a survival advantage.

Pregnancy

Successful pregnancy can be achieved in most women with C-TGA, but careful prepregnancy evaluation is mandatory. A comprehensive cardiovascular examination should include an assessment of cardiac rhythm, ventricular function, AV valve regurgitation, associated lesions, and postoperative sequelae. In general, those women with a systemic ventricular ejection fraction <40% or significant systemic AV valve regurgitation should be counseled against pregnancy because the added volume load of pregnancy will not be well tolerated. Because angiotensin-converting enzyme inhibitors are contraindicated in pregnancy, the assessment of ventricular function should be performed with the patient not taking these drugs. Patients should be advised that the rate of fetal loss and maternal cardiovascular morbidity is increased. In the series of Connolly et al., 22 women had 60 pregnancies resulting in 50 live births (83%). None of the offspring had congenital heart disease. There were no pregnancy-related deaths, but 1 woman with important AV valve regurgitation developed congestive heart failure, and another woman who had 12 pregnancies had multiple complications, including endocarditis and congestive heart failure.

If pregnancy is undertaken, the patient should be monitored throughout, and delivery should be managed by a multidisciplinary healthcare team including obstetricians, cardiologists, and obstetric anesthesiologists, preferably in a center where the cardiologists have expertise in congenital heart disease and high-risk pregnancy care.

Follow-Up

All patients, whether operated on or not, should have a periodic evaluation by a specialist in adult congenital heart disease. The evaluation should include a detailed imaging study by echocardiography and/or MRI. Declining systemic ventricular function should prompt a search for worsening AV valve regurgitation. Exercise testing facilitates detection
of subtle deterioration in exercise capacity in patients who may believe that they are asymptomatic and permits exercise guidelines to be prescribed. Symptoms or signs of arrhythmia warrant detailed investigation and a review of underlying hemodynamic problems. Most patients require endocarditis prophylaxis unless they have no valvular dysfunction, outflow obstruction, or VSD.

Disclosures

None.

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