Congenitally corrected transposition of the great arteries

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CONGENITAL HEART DISEASE

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Congenitally corrected transposition of the great arteries (ccTGA) is a rare defect representing approximately 0.5% of all congenital heart disease. ccTGA is characterised by the combination of atrioventricular (AV) discordance and ventriculo-arterial (VA) discordance. Various terms have been used to describe this anatomy: congenitally or physiologically corrected transposition of the great arteries, L-transposition, double discordance and ventricular inversion being the most frequently used. The term 'congenitally corrected transposition' is used to differentiate this condition from 'complete transposition' (d-TGA) in which there is VA discordance but AV concordance.

ANATOMY

There is usually normal drainage of the systemic and pulmonary veins to the right and left atriums respectively. The right atrium connects via the mitral valve to the morphologic left ventricle which supplies the pulmonary artery. The left atrium connects via the tricuspid valve to the morphologic right ventricle which supplies the aorta via a sub-aortic infundibulum. The ventricles are most commonly side by side: in patients with situs solitus the morphologic left ventricle is rightward and the morphologic right ventricle is leftward.

The anatomy is described as congenitally 'corrected' because the flow of blood is normal with the deoxygenated systemic venous blood being pumped to the lungs and the well oxygenated pulmonary venous blood being pumped to the body. Nevertheless, the two wrongs of AV and VA discordance do not make a right, as the morphologic left ventricle (LV) and mitral valve supply the pulmonary circulation and the morphologic right ventricle (RV) and tricuspid valve supply the systemic circulation.

The most common anatomical arrangement is situs solitus with L-looping of the ventricles and the aorta anterior and leftward of the pulmonary artery (S,L,L), found in 19/22 of our cases in Auckland (unpublished data). The aorta was anterior and rightward (S,L,D) in two of 21 cases of ccTGA with situs solitus. In patients with situs solitus the cardiac position is frequently more mesocardic than usual although the apex remains leftward in the majority: in our series of 21 cases there was true mesocardia in one case and dextrocardia in two cases with situs solitus. One of 22 cases (5%) had situs inversus with D-looping of the ventricles and the aorta anterior and rightward (I,D,D).

Coronary artery anatomy

The coronary arteries in ccTGA are inverted. Thus the morphologically left coronary artery (LCA) arises from the patient's right sided sinus and the morphologically right coronary artery (RCA) from the left sided sinus. A variety of coronary arterial patterns are seen. In our series of 19 with ccTGA (S, L,L), five (26%) had single sinus origin of the two main coronary arteries. In four the RCA arose with the LCA either from a single stem or with a separate origin, either with anterior or posterior passage of the RCA. In two of these an additional RV branch arose from the left sided sinus. In the fifth case with commissural malalignment, the RCA and LCA arose from either side of a posterior sinus. Of note we also found 2/18 cases (11%) available for review where the anterior descending coronary artery was hypoplastic, with potential implications for the LV coronary supply after the double switch procedure (see below). Ismat published a morphologic study of coronary anatomy in 20 S,L,L hearts from Philadelphia, with similar variability.[1] Ten of these had ccTGA, of whom one had a single coronary origin from which all three coronary arteries arose, one had the anterior descending artery arising from the RCA, one had the LCA arising directly over the commissure, and one had both coronary arteries arising separately from the left posterior sinus. Careful preoperative delineation of the coronary anatomy is particularly important for the surgeon before the double switch operation (see later).

Conduction system

The anatomy of the conduction system is abnormal in this condition, with the AV node being unable to connect normally to a penetrating AV conduction bundle. There is therefore a second AV node positioned anteriorly which gives rise to an elongated AV bundle.[2] This makes these patients prone to developing heart block and also prone to re-entry tachycardias. Electrophysiological interventions should be performed by those with experience and understanding of congenitally abnormal hearts.

ASSOCIATED ABNORMALITIES

More than 90% of patients have additional anatomical abnormalities. The most frequent are ventricular septal defects, LV outflow tract obstruction, and abnormalities of the tricuspid valve. We have combined the data from our own postmortem series with those from Allwork[3] and

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Van Praagh[4] in order to obtain the data detailed below from a total of 93 cases.

Ventricular septal defect

A ventricular septal defect (VSD) was present in 84% of cases, although clinical series have suggested VSD to be present in 60–75% of cases. This is most commonly a perimembranous outflow (subpulmonary) defect and is frequently large. VSDs of other types are also described including doubly committed subarterial (conal) defects, muscular defects and AV canal type defects. Perimembranous defects may become at least partially obstructed by aneurysmal membranous septum tissue or by overriding or straddling tricuspid valve tissue.

Left ventricular outflow obstruction

There was LV outflow obstruction in 53% of cases, including 16% with pulmonary atresia. The LV outflow obstruction may comprise subpulmonary stenosis and/or pulmonary valve stenosis. Subpulmonary stenosis may be due to a fibromuscular membrane, tunnel-like hypoplasia, or may be related to an aneurysm of the membranous septum. Alternatively it may be caused by accessory mitral or tricuspid valve tissue or indeed by systolic anterior motion of the mitral valve. LV outflow obstruction is frequently associated with a VSD, especially at the extreme end of the spectrum with pulmonary valve atresia.

Tricuspid valve abnormalities

The tricuspid valve has been reported to be abnormal in the great majority of ccTGA patients at autopsy, but not all will be clinically important. The nature of the abnormality is variable, but is frequently Ebstein's anomaly. There is a wide spectrum of severity with the worst representing severe Ebstein's. In the combined series the tricuspid valve was abnormal in 94%, with Ebstein's anomaly in 53%. The impact in terms of tricuspid valvar regurgitation may be greater in this population due to the fact that the RV is working at systemic pressure and also that the geometry of the tricuspid annulus is altered due to the abnormal septal position resulting from the systemic pressure RV.

Mitral valve abnormalities

Mitral valve abnormalities are less frequent than those of the tricuspid valve, but were nevertheless present in 55% of pathological specimens in the series published by Gerlis.[w1] These included abnormal numbers of cusps, abnormalities of the subvalvar apparatus, cleft mitral valve, and mitral valve dysplasia.

PRESENTATION AND DIAGNOSIS

With increasing frequency the diagnosis is made antenatally. Sharland published a series of 34 antenatal diagnoses over a 10 year period at Guy's Hospital.[5] In the fetal four chamber echocardiographic view the normal apical offsetting of the tricuspid valve is reversed, thus alerting the sonographer to the fact that the right atrium connects to a ventricle of LV morphology. It is worth noting, however, that with large VSDs with inlet extension, especially with straddling or overriding of the tricuspid valve, the normal offsetting is not present. If not diagnosed antenatally, young children with ccTGA are likely to be asymptomatic if there are no associated abnormalities. In the presence of any of the associated abnormalities described above it is likely that a murmur and/or symptoms will be present which may lead to the diagnosis being made echocardiographically. Older children and adults may present with symptoms related to systemic right ventricular dysfunction. Cardiac conduction defects may cause symptoms related to bradycardia which leads to cardiac investigations being performed. There are also reports in the literature of the diagnosis of isolated ccTGA being made in previously asymptomatic adult patients—for example, during investigation of coronary events—even in the sixth or seventh decades of life.

Investigations

The ECG is characteristic, at least in patients with situs solitus, with left axis deviation and Q waves in the right precordial leads but not the left precordial leads. Echocardiography is the mainstay of diagnosis (figure 1). The sonographer needs to take a detailed sequential approach to the cardiac anatomy, starting with the visceral atrial situs and working through to the great arteries. The presence of associated abnormalities should be defined and the function of the systemic RV and tricuspid valve assessed. Cardiac magnetic resonance imaging (MRI) has taken on an increasing role in patients with ccTGA (figure 2), particularly for quantitative assessment of the function of the systemic RV. Nevertheless even MRI assessment of RV function is challenging due to the complex shape of the RV and the multiple coarse trabeculations in the context of the RV hypertrophy associated with systemic pressure functioning. This issue has been recently studied by a multicentre Dutch group headed by Barbara Mulder, who recommended an approach of tracing RV contours outside the trabeculations as being both more time efficient and more reproducible.[6] In our own practice we would use cardiac MRI as our primary method of tracking RV size and function in patients without pacemakers, usually performing MRI approximately every 2–3 years in clinically stable patients. In paced patients echocardiography is the primary modality, but radionuclear ventriculography should also be considered and has been shown by Hornung *et al* to correlate well with MRI in the related Mustard and Senning groups.[w2]

NATURAL HISTORY

The natural history of patients with ccTGA is largely dictated by the function of the systemic RV and by the presence or absence of associated abnormalities. In the series of antenatal diagnoses from Guy's Hospital, where the pregnancy was

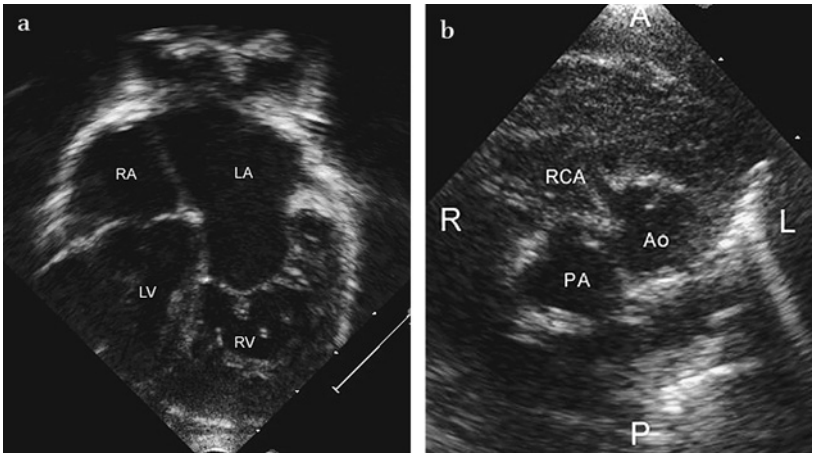

Figure 1 (A) Echocardiographic apical four chamber view demonstrating atrioventricular discordance. There is increased apical offsetting of the septal leaflet of the tricuspid valve indicative of Ebstein's anomaly. (B) Echocardiographic parasternal short axis view showing anterior and leftward aorta. A, anterior; Ao, aorta; L, left; LA, left atrium; LV, left ventricle; P, posterior; PA, pulmonary artery; R, right; RA, right atrium; RCA, right coronary artery; RV, right ventricle.

continued, 19 of 23 babies survived to the end of infancy. As mentioned previously, there are case reports in the literature describing patients with isolated ccTGA and good RV function who have been diagnosed incidentally in the fifth to seventh decades of life. It is therefore possible for the RV to supply the systemic circulation for decades with normal function; however, it appears that this situation is the exception rather than the rule. Most patients will develop progressive RV dysfunction, in some cases as early as the first decade of life. A multicentre study of 182 adults with ccTGA published by Graham showed that important systemic RV dysfunction and congestive cardiac failure symptoms are common by the third decade of life and have an increasing frequency with advancing age, both in patients with and without associated abnormalities.[7] In this study 67% of patients with associated abnormalities had clinical heart failure by the age of 45 years, compared to 25% of those without associated abnormalities. The study showed moderate or severe RV dysfunction in 56% of patients with associated abnormalities and 32% of those without associated abnormalities. A single centre database review from the Toronto Congenital Cardiac Centre for Adults of 131 patients with ccTGA showed a mortality rate of 21% in patients >18 years of age, with a mean age at death of 33 years.

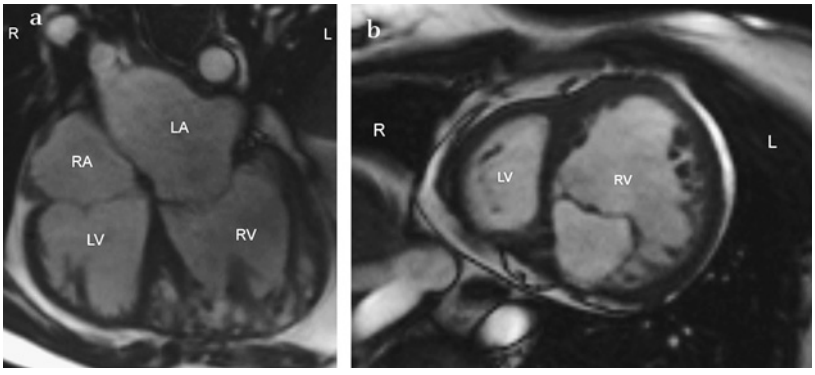

Figure 2 (A) Magnetic resonance imaging (MRI) four chamber view demonstrating atrioventricular discordance. The morphological right ventricle is clearly identifiable by virtue of the apical trabeculations. (B) MRI ventricular short axis view showing a hypertrophied morphological right ventricle and bowing of the septum towards the left ventricle. L, left; LA, left atrium; LV, left ventricle; R, right, RA, right atrium, RV, right ventricle.

Functional outcomes

In the multicentre study published by Graham,[7] approximately 60% of patients with associated abnormalities were in Warnes–Somerville functional class 1, with the majority of the remainder being in class 2. In patients without associated abnormalities approximately 70% were in class 1. Nevertheless, when measured objectively, most patients with ccTGA have impaired functional capacity. Fredriksen published the results of cardiopulmonary exercise testing in a group of 41 adult patients from Toronto.[8] This showed a notably reduced maximal oxygen uptake of 22 ml/kg/min in the 19–29 year age group and 21 ml/kg/min in the 30–39 year age group (both were approximately half that of normal controls). In the 40–55 year age group this dropped to 11 ml/kg/min, less than one third that of the healthy controls. The reasons for this are multifactorial, but important factors include impaired ventricular function, limited chronotropic response to exercise and abnormal lung function, particularly in patients who have had previous surgery.

Pregnancy

Connolly published the results of a retrospective review of the Mayo clinic experience of 60 pregnancies in 22 women with ccTGA over a 17 year period.[9] Forty-nine of these pregnancies resulted in live births, one of which was a premature delivery at 29 weeks. The mean birth weight was 3.2 kg. One woman had 12 pregnancies, of which 10 were successful: she had various complications including congestive heart failure, endocarditis and myocardial infarction. Only one other patient developed cardiac complications—congestive heart failure related to tricuspid valve regurgitation necessitating tricuspid valve replacement at 2 months postpartum. Therrien published a series of 45 pregnancies in 19 patients from the Royal Brompton Hospital, with cardiac complications in five, including heart failure, worsening cyanosis and cerebrovascular accident.[10]

Although pregnancy may be well tolerated by some patients in this group, detailed pre-pregnancy assessment should be performed and women with functional class 3 or 4, more than mild ventricular dysfunction or significant tricuspid regurgitation should be counselled against pregnancy. There are some data to suggest that function of the systemic RV may deteriorate, potentially irreversibly, during pregnancy and this concern should be discussed with patients.[w3] The final factor that should be

discussed during pre-pregnancy counselling is the uncertain maternal longevity in patients with ccTGA.

MECHANISMS OF SYSTEMIC RV DYSFUNCTION

Although the structural differences between the morphological LV and RV are clear, both at a macroscopic and microscopic level, the exact reasons for the development of RV dysfunction remain the subject of debate. The coronary supply to the RV differs from that of the LV at a microvascular level. Hornung with the Sydney group published nuclear medicine data using sestamibi myocardial perfusion scanning that suggests that impaired myocardial perfusion and RV myocardial fibrosis is frequent in subjects with this diagnosis and is associated with regional hypokinesis (figure 3).[11] However, recent attempts to confirm this finding using magnetic resonance delayed enhancement imaging have produced conflicting results.[w4, w5] Hauser from Munich published positron emission tomography data which demonstrated that although resting myocardial blood flow to the systemic RV does not differ from the systemic LV in normal subjects, coronary flow reserve to the systemic RV is substantially reduced (2.5 compared to 4.0) in the ccTGA group.[12] It is possible, therefore, that chronic low grade coronary insufficiency may contribute to progressive RV dysfunction in this population.

ELECTROPHYSIOLOGICAL ISSUES

Patients with ccTGA have a 2% per annum risk of developing complete heart block. This risk is increased in patients requiring intracardiac repair of associated anomalies.[7] In the context of the systemic right ventricle, chronic ventricular pacing is likely to further negatively impact on long term ventricular function.

There is also a risk of tachyarrhythmias, both re-entry type arrhythmias related to the abnormal conducting system anatomy or to accessory pathways, and also atrial and ventricular tachycardias which may be related to ventricular dysfunction and fibrosis.

MEDICAL MANAGEMENT

Drug treatment

There are very limited data available regarding the drug management of ventricular dysfunction in the context of a systemic RV. In patients with symptomatic heart failure or more than mild systemic RV dysfunction, most centres have used conventional systemic LV protection strategies. Small studies of the use of angiotensin converting

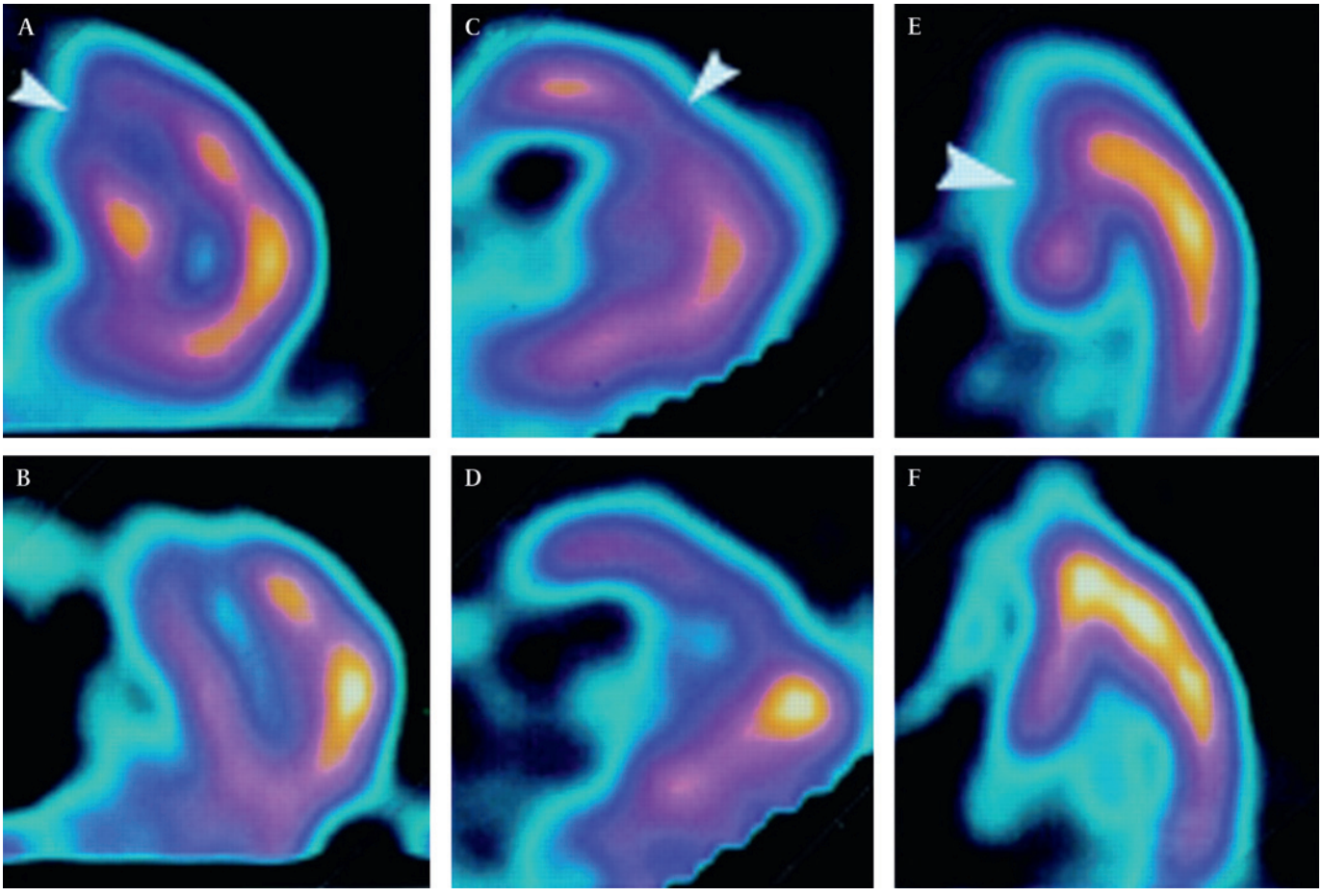

Figure 3 Sestamibi myocardial perfusion scan showing a fixed defect in the anterior wall (small arrowheads) and reversible ischaemia in the septum (large arrowhead). Transaxial views at mid ventricular to basal level at (A) stress and (B) rest. Vertical long axis views at (C) stress and (D) rest. Horizontal long axis views at stress (E) and rest (F).

enzyme (ACE) inhibitors and angiotensin receptor blockers have suggested little or no benefit in the related postoperative Mustard and Senning populations with d-TGA, but have been and will continue to be hampered by small patient numbers.[w6] β-blocker use has not been studied in the ccTGA population, but there is some suggestion of benefit in the Mustard and Senning groups.[w7]

Resynchronisation therapy

A substantial number of ccTGA patients will eventually require permanent pacing for complete heart block, with the ventricular lead being placed in the morphologic LV. This may negatively impact on the systemic RV due to the resulting interventricular dyssynchrony. Resynchronisation approaches to the systemic RV have been used in small numbers of patients with severe ventricular dysfunction, a coronary sinus lead being used to pace the RV.[w8] Although numbers are small, this approach appears to have produced a symptomatic improvement in some patients with class III–IV symptoms.

It is worth noting, however, that in our own Auckland series of 22 postmortem ccTGA cases the coronary sinus was absent or not identified in five cases (23%), with the coronary veins having separate entries to the right atrium or to the ipsilateral atria. In Uemura's publication from 1996, the RV coronary drainage was all to the coronary sinus in 87% and partial or total to the left atrium in 11%, whereas the coronary venous drainage from the LV was completely to the coronary sinus in only 14%, direct to the RA and coronary sinus in 63%, and direct to the RA in 20%.[13] These anatomical variations would clearly have implications for placement of a coronary sinus lead to pace the RV and would likely make this approach impossible, necessitating a more invasive epicardial approach.

SURGICAL MANAGEMENT

Conventional repair

Until recently, patients with associated abnormalities underwent conventional physiological surgical repair—for example, isolated VSD repair or VSD repair with insertion of an LV to pulmonary artery conduit for pulmonary valve atresia or stenosis. After this type of repair the RV remains the systemic ventricle and the problem of progressive RV dysfunction may be exacerbated by the negative impact of bypass surgery on the myocardium. Long term results of this type of surgical repair are disappointing. In an early paper, Lundstrom reported the results of 111 patients from the Hospital for Sick Children and the National Heart Hospital, London, of whom 51 underwent surgical repair, with 11 early deaths. Termingon reported the experience from Laennec Hospital, Paris of 52 patients with 15% operative mortality. The 10 year survival was 55% for patients with VSD and LV outflow obstruction and 71% for patients with VSD alone.[w10] Hraska published the Boston experience of 123 patients with survival of 70% at 10 years for patients undergoing repair of VSD ± pulmonary stenosis or atresia.[14] Freedom from RV dysfunction was approximately 40% at 15 years with factors predicting RV dysfunction being Ebstein's anomaly, tricuspid valve replacement and postoperative complete heart block. Yeh published the Toronto experience of 127 patients (including nine double switch procedures).[15] Operative mortality was 6%. At 20 years, survival was 48% and reoperation was required in 56%. Survival did not differ according to type of abnormality requiring repair.

Surgical management of systemic tricuspid valve regurgitation

Progressive tricuspid regurgitation is common and often goes hand in hand with progressive RV dysfunction. Van Son published the results of a series of 40 patients from the Mayo Clinic undergoing tricuspid valve surgery, with valve replacement in 39 cases.[16] Preoperatively 68% were New York Heart Association (NYHA) functional class III or IV. Early mortality was 10%. With a median follow-up of 4.7 years (maximum 26 years) there were eight late deaths from congestive cardiac failure related to systemic RV dysfunction. Five year survival was 78% and 10 year survival 68%. Although the era of surgery appeared to be a major factor in outcome, preoperative RV ejection fraction also correlated closely with survival. The survival in patients with RVEF <44% was 49% at 5 years and 20% at 10 years, whereas in patients with RVEF ≥44% the survival was 100%. The authors concluded that surgery for significant tricuspid regurgitation should be considered at the earliest sign of progressive RV dysfunction. Scherptong recently published a multicentre Dutch experience of tricuspid valve surgery in 16 patients with a systemic RV (including patients with d-TGA after the Mustard and Senning operations).[w11] Eight patients underwent repair and eight underwent replacement. This study showed a trend towards decreased survival after valve repair compared to replacement and although there was initial improvement in those undergoing repair, a steady increase in the degree of regurgitation was seen over the first postoperative year. This supports the impression that an approach of primary tricuspid valve replacement should be taken in this group.

Anatomical repair

More recent surgical approaches have focused on so called 'anatomic' repairs which return the LV to the systemic circulation and may therefore result in improved long term prognosis. These operations represent major and technically challenging surgical procedures. Patients without LV outflow obstruction in whom the pulmonary valve is suitable to function as a neo-aortic valve may be approached using the double switch procedure: this combines (1) the Senning atrial baffle procedure, which redirects blood flow at the atrial level, with (2) the arterial switch procedure where the great arteries

are transected and reversed. Successful outcome after this procedure is dependent on the LV being adequately prepared for functioning at systemic arterial pressure. In the presence of a large VSD the LV will have been functioning at systemic pressure, but in the absence of associated abnormalities it will have been functioning at pulmonary artery pressure. In this latter context the LV can be retrained in younger children by the placement of a pulmonary artery band to increase LV pressure, although this approach has met with mixed success. Patients with VSD and LV outflow obstruction can be approached using the Rastelli–Senning procedure, where the Senning operation is combined with the Rastelli procedure, which involves channelling the LV outflow across the VSD to the aorta and placing an RV to pulmonary artery valved conduit.

Series of double switch and Rastelli–Senning procedures have been published by several groups. Surgical mortality is generally of the order of 0–10%. Duncan from the Cleveland Clinic published the results of 46 patients undergoing anatomical repair, with median age at surgery of 28 months, the oldest patient being 16 years old.[17] There were no hospital deaths and one late cardiac death during a median follow-up of 24 months. Shin'oka published a series of 84 anatomical repairs from Tokyo, with longer follow-up: the 15 year survival was 75% in the double switch group and 80% in the Rastelli–Senning/Mustard group.[18] Langley published the Birmingham series of 54 patients: the 7 year survival was 85% in the double switch group and 95% in the Rastelli–Senning group.[w12]

Anatomic repair: patient selection

Most groups would now regard anatomical repair as being preferable to conventional surgery in patients requiring repair of associated defects when the LV has been functioning at high pressure—for example, when there is a large VSD. In those patients without associated abnormalities who do not have a prepared LV but have RV dysfunction or significant tricuspid regurgitation, retraining of the LV with pulmonary artery banding should be considered. Most centres would not recommend a prophylactic double switch procedure for patients without associated abnormalities in whom RV and tricuspid valve function is normal.

During the childhood years the subpulmonary LV has been successfully retrained using a strategy of progressive pulmonary artery banding in order to provide an increased pressure load on the LV and promote LV hypertrophy. Cardiac catheterisation is used to assess LV pressure, together with echocardiography and MRI to assess LV wall thickness, mass and function. The Cleveland Clinic group recommend that in childhood an LV systolic pressure of at least 70–80% of systemic pressure, and in adolescence an LV systolic pressure closer to 100% systemic pressure should be attained before restoring the LV to the systemic circulation. The Paris group recommend an LV mass: LV volume ratio >1.5. Others recommend an LV wall thickness and mass that is normal for a systemic LV using established echocardiographic or MRI values. The LV must be monitored carefully and development of LV dysfunction should be regarded as a failure of LV retraining. One of the best predictors of failure of retraining is age, with most successfully retrained patients being <10 years of age. Devaney from Ann Arbor published a series of 23 patients who were felt to be candidates for a Senning plus arterial switch protocol.[19] Pulmonary artery banding was performed in 15 patients, in 11 cases for the sole purpose of LV retraining. The two oldest, aged 12 and 14 years, had LV failure necessitating removal of the band. A further patient who underwent banding with subsequent double switch at age 7 years required heart transplantation for severe LV failure with diastolic dysfunction.

Double switch outcomes: a word of caution

Systolic dysfunction of the LV is relatively common after the double switch procedure, with Quinn of the Birmingham group demonstrating moderate-severe LV dysfunction in six of 11 patients who had required LV retraining and six of 33 patients who did not require retraining.[20] Bautista-Hernandez published the results of the Boston experience of 44 patients, of whom 18% developed LV dysfunction, the requirement for permanent pacing being a predictor for LV dysfunction.[21]

It remains to be seen what the longer term incidence of LV dysfunction will be, let alone other complications such as progressive aortic dilatation and regurgitation as seen after the standard arterial switch procedure for complete transposition. Although short-medium term results are promising, long term follow-up after the double switch is not yet available and it is important to be cognisant of this fact when counselling patients and their families.

FOLLOW-UP

All patients with ccTGA should be followed by a paediatric cardiologist or adult cardiologist with specific expertise in congenital heart disease. Most patients can be seen every 1–2 years, although those with complicating factors such as severe systemic right ventricular dysfunction may require more frequent visits. Patients should of course be advised regarding maintaining a healthy lifestyle and avoiding smoking. Regular moderate exercise should be encouraged, although many cardiologists would advise avoidance of strenuous or endurance exercise. Pregnancy counselling should be provided as discussed above. Because of the risk of progressive heart block, an ECG should be performed at each visit, with consideration of 24 h Holter monitoring. Patients should also undergo regular assessment of functional capacity with cardiopulmonary exercise testing. Regular assessment of ventricular function is important: the 'gold standard' imaging modality for the systemic RV is cardiac MRI, and as this becomes more widely

Congenitally corrected transposition of the great arteries: key points

Frequent associations
- Structural lesions
  - Ventricular septal defect
  - Left ventricular outflow obstruction
  - Tricuspid valve anomalies
- Rhythm abnormalities
  - Heart block
  - Re-entry tachyarrhythmias

Requirements for double switch operation
- LV pressure ≥70—80% systemic
- LV mass: LV volume ratio >1.5
- LV wall thickness normal for systemic LV
- Normal LV function after pulmonary artery banding

Follow-up
- Should be performed by cardiologists experienced in the care of patients with congenital heart disease
- Regular assessment of systemic RV function, preferably by cardiac MRI
- Ongoing review of cardiac conduction to exclude development of progressive heart block
- Regular cardiopulmonary exercise testing
- Pre-pregnancy advice and planning

available it should be performed at least every 3 years.

CONCLUSIONS

Patients with ccTGA represent a rare and complex group for whom outcomes have traditionally been largely dependent on function of the systemic right ventricle. Surgical management has changed in the last two decades from physiological to 'anatomical' repair, which returns the LV to the systemic circulation. Longer term follow-up will dictate whether the early promise offered by this approach will translate into improved long term survival.

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