Anatomic repair for congenitally corrected transposition of the great arteries: A single-institution 19-year experience

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Objective(s): Anatomic repair for congenitally corrected transposition of the great arteries (ccTGA) has been shown to improve patient survival. We sought to examine long-term outcomes in patients after anatomic repair with focus on results in high-risk patients, the fate of the neo-aortic valve, and occurrence of morphologically left ventricular dysfunction.

Methods: We conducted a retrospective, single-institution study of patients undergoing anatomic repair for ccTGA. A total of 113 patients from 1991 to March 2011 were included. Double-switch (DS) repair was performed in 68 patients, with Rastelli-Senning (RS)-type repair in 45. Pulmonary artery banding for retraining was performed in 23 cases. Patients were followed up for survival status, morbidity, and reinterventions. A subgroup of 17 high-risk patients in severe heart failure, ventilated, and on inotropes before repair, were included.

Results: Median age at repair was 3.2 years (range, 25 days to 40 years) and weight was 14.3 kg (3.2-61.4). There were 5 (of 68; 7.4%) early deaths in the DS group and 0 (of 45) in the RS group. Actuarial survivals in the DS group were 87.6%, 83.9%, 83.9% at 1, 5, and 10 years versus 91.6%, 91.6%, 77.3% in the RS group (log-rank: $P = .98$). Freedom from death, transplantation, or heart failure was significantly better in the RS group at 10 years ($P = .03$). There was no difference in reintervention at 10 years (DS, 50.3%; RS, 49.1%; $P = .44$). In the DS group, the Lecompte maneuver was associated with late reinterventions on the pulmonary arteries. Overall survival in the high-risk group was 70.6%. During follow-up, 14.2% patients had poor function of the morphologically left ventricle, all in the DS group, but this was not related to preoperative status or previous banding. The majority of patients after DS had mild aortic incompetence, which appeared well tolerated. Annuloplasty of the aortic root at time of DS reduced the risk of late aortic valve replacement.

Conclusions: There is significant morbidity after anatomic repair of ccTGA, which is higher in the DS than the RS group. Nevertheless, the majority of patients are free of heart failure at 10 years, including high-risk patients in severe heart failure before repair. Aortic annuloplasty may reduce risk of late aortic insufficiency. (*J Thorac Cardiovasc Surg* 2011;142:1348-57)

Supplemental material is available online.

The spectrum of presentation of congenitally corrected transposition of the great arteries (ccTGA) is represented by a heterogeneous group of patients, both anatomically and in terms of clinical symptoms and urgency.¹ Patients may have heart failure or cyanosis in infancy or have balanced circulations with mode of presentation primarily

determined when pulmonary blood flow becomes limiting or associated lesions become manifest. Physiologic-type repairs addressing isolated associated lesions such as pulmonary atresia or ventricular septal defects (VSDs) have achieved acceptable results, although often not in patients with significant tricuspid regurgitation (TR) at the time of repair.² Even with no associated lesions, the natural history of the condition is poor, with only 50% survival at 40 years. As such, the lesion itself is far from “corrected.” True correction, in terms of restoring atrioventricular and ventriculoarterial concordance, has been achieved by anatomic repair. Procedures in this class include atrial–arterial switch (double switch; DS) and the Rastelli-Senning (RS) procedure and its modifications. Late outcomes for these complex procedures are still poorly understood. We present a large, single-institution experience of patients with ccTGA undergoing anatomic repair with particular focus on survival, need for reinterventions, postrepair function of the systemic morphologically left ventricle (mLV), and outcomes in the number of high-risk patients with severe congestive cardiac failure and TR.

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Disclosures: Authors have nothing to disclose with regard to commercial support.

Read at the 91st Annual Meeting of The American Association for Thoracic Surgery, Philadelphia, Pennsylvania, May 7-11, 2011.

Received for publication May 6, 2011; revisions received Aug 1, 2011; accepted for publication Aug 10, 2011; available ahead of print Sept 28, 2011.

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0022-5223/\$36.00

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doi:10.1016/j.jtcvs.2011.08.016

Abbreviations and Acronyms

AI	= aortic incompetence
AVR	= aortic valve replacement
ccTGA	= congenitally corrected transposition of the great arteries
DS	= double switch
ICU	= intensive care unit
mLV	= morphologically left ventricle
MR	= mitral regurgitation
mRV	= morphologically right ventricle
PAB	= pulmonary artery banding
PAB-P	= pulmonary artery banding—protection
PAB-T	= pulmonary artery banding—training
PPM	= permanent pacemaker
RS	= Rastelli-Senning
TR	= tricuspid regurgitation
TV	= tricuspid valve
VSD	= ventricular septal defect

PATIENTS AND METHODS**Follow-up**

We conducted a retrospective, single-institution study of all 113 patients undergoing anatomic repair for ccTGA. Patients from November 1991 to March 2011 were included. In the same period, there were 26 patients who were followed up medically or who had undergone physiologic-type repairs or pulmonary artery banding (PAB) only. Sixty-three patients had undergone PAB placement at either our own or other institutions before definitive repair: these were for purposes of protection (PAB-P) in patients with large, unrestrictive VSDs, or to train the mLV (PAB-T) before DS repair where the mLV had not been conditioned by volume or pressure load. All preoperative and postoperative echocardiographic data were reviewed to assess morphologically right ventricular/left ventricular (mRV/mLV) function, atrioventricular and semilunar valve function, ventricular function, and presence of other associated lesions. One independent cardiologist (O.S.) determined these parameters to eliminate interobserver bias. The study was a retrospective review that had been registered with the Birmingham Children's Hospital Research and Development Board and ethical approval had been waived by the Central Office for Research Ethics Committees owing to use of retrospective, anonymized data.

Follow-up postanatomic repair involved electronic patient records (Heartsuite Database; Systera Inc, Glasgow, United Kingdom) and correspondence or structured telephone interviews with local referring cardiac surgeons or cardiologists for patients living abroad. Follow-up involved assessment of patient New York Heart Association functional status, cardiac rhythm, and echocardiographic findings. This included focus on the atrial pathways and the conduit performance, atrioventricular valve or semilunar valve regurgitation, and ventricular function. All patients in the study had follow-up to within 1 year of the conclusion of the study (March 2011). As such, all patients had been followed up from the time of surgical repair to the end of the study, with none being lost to follow-up (100% complete).

Surgical Technique

Patients for DS repair underwent Senning arterial switch procedures. These were divided into patients with small (ie, restrictive) or no VSDs and those with large (unrestrictive) VSDs. The Lecompte maneuver was generally performed if the vessels were in an anteroposterior relationship,

but not if the vessels were side-by-side. In cases of PAB-T, banding was performed to achieve an mLV/mRV pressure ratio of 0.66:0.75, as previously described.³ Patients undergoing the RS procedure were usually palliated with a systemic–pulmonary artery shunt before definitive repair and 6 underwent a bidirectional Glenn procedure. Techniques have been described in detail previously.⁴ Three patients in the RS group with a double-outlet right ventricle–type arrangement underwent Senning with intraventricular tunneling of the mLV to the aorta.

In the more recent cases, neo-aortic root annuloplasty was performed if the predicted size of the root was 2 mm or more greater than predicted using a 2-layer absorbable purse-string suture placed externally around the root before implantation of the coronary buttons, tied down over the predicted size dilator in the outflow tract.

The Senning procedure was used for the atrial switch, with or without supplementation to the pulmonary venous atrium using autologous (Shumaker modification) or pulmonary homograft patch.⁵ Three patients with unusual venous anatomy underwent an “external” Senning procedure, as described previously.⁶ The systemic venous atrium was not augmented in any cases.

Statistical Analysis

All data were analyzed using the statistical software package R version 2.11.1 (R Foundation, Vienna, Austria). Continuous variables are presented as means with the standard deviation or as medians with the stated range. Comparative univariable analyses were performed using the Kruskal-Wallis test. Binomial or ordinal data are expressed as percentages, and comparative univariable analyses were performed using the 2-sided Fisher exact test. Kaplan-Meier analysis was used for survival with the log-rank test used to determine significant differences.

RESULTS**Patient Demographics, Anatomy, and Surgical Strategy**

A total of 113 patients underwent anatomic repair for ccTGA. Of these, 68 patients underwent DS, with 2 of these as staged procedures. Forty-five patients had undergone RS-type repairs, which included 3 one-and-a-half repairs and 3 double-outlet right ventricle variants with Senning–tunnel procedures.

The majority of patients in the cohort of 113 patients were male (73/113), with median age at repair 3.2 years (range, 25 days–40 years). Median weight at repair was 14.3 kg (3.2–61.4 kg). Morphologic characteristics and associated lesions are shown in Table 1. A variety of concomitant procedures were performed at the time of anatomic repair, including pulmonary venous atrium supplementation, mitral valve repair, and tricuspid valve (TV) repair. Anterior relocation of the pulmonary artery confluence (Lecompte maneuver) was performed in 50 patients in the DS group. Notably, 13 patients in this group also underwent neo-aortic root annuloplasty, with only 1 such patient in the RS group (Table E1). In addition, 15 patients in the DS group had heart block before repair with permanent pacemakers (PPMs) in situ, although none in the RS group. Median conduit size in the RS group was 16 mm (14–22 mm). The surgical strategy algorithm is shown in Figure 1.

Twenty-three patients underwent PAB-T; median age at banding was 2.46 years (0.11–9.82 years), with median

TABLE 1. Patient demographics, morphologies, associated lesions

Characteristic	No. of patients (%)
Demographics	
Total no.	113
Double switch (DS)	68
Rastelli-Senning (RS)	45
Male/female	73/40
Median age at repair (y)	3.2 (25d-40 y)
Median weight at repair (kg)	14.3 (3.2-61.4)
Morphology	
Usual atrial arrangement	103
Situs inversus	10
Azygos continuation of IVC	4
Left atrial isomerism	2
DORV	7
Levocardia	88
Dextrocardia	23
Mesocardia	2
Associated lesions	
VSD	89
Ebsteinoid anomaly of TV	14
Severe TR	6*
Pulmonary hypertension	6*
Coarctation/hypoplasia of aorta	20
Interrupted aortic arch	1
Bicuspid AV/PV	2/3
Quadricuspid PV	1
Double orifice TV	2 (1 with straddling)

IVC, Inferior vena cava; DORV, double-outlet right ventricle; VSD, ventricular septal defect; TV, tricuspid valve; TR, tricuspid regurgitation; AV, aortic valve; PV, pulmonary valve. *All in DS group.

duration of banding 562 days (5-2758 days), and 5 patients required a second banding procedure. Thirteen patients had an unsuccessful PAB-T or were not anatomically suitable

for repair. “Failed” PAB was defined as not meeting the following criteria: good mLV systolic function; adequate mLV free wall thickness; adequate mLV pressure generation of at least 0.66 to 0.75 of mRV (systemic) pressure before repair. We³ have described these criteria in a previous report.

Of note, 17 patients were classified as high-risk before anatomic repair, defined as requiring intensive care unit (ICU) admission, ventilation, and inotropic support preoperatively. All had signs of severe congestive cardiac failure, 6 had pulmonary hypertension, and 6 had severe TR. An additional 4 patients had severe TR in the group requiring PAB-T.

Intraoperative Data, Survival, and Secondary Outcomes

Median cardiopulmonary bypass and crossclamp times were similar in DS and RS groups (Table 2). Median deep hypothermic circulatory arrest time in the DS group was 15 minutes (0-66 minutes) and 38 minutes (1-74 minutes) in the RS group. There were 5 (of 68; 7.4%) early deaths (within 30 days) in the DS group and 0 (of 45; 0%) in the RS group. There were 5 and 7 late deaths in the 2 groups, respectively (Table 2). One-, 5-, and 10-year Kaplan-Meier survivals in the DS group were 87.6%, 83.9%, and 83.9%. Respective survivals for the RS group were 91.6%, 91.6%, and 77.3%. There was no significant difference between groups (Figure 2, A; log-rank, $P = .98$).

There were 3 unexpected deaths in our series. None of these patients were in the high-risk group before repair. Two had undergone RS at 13 months and 5 years of age and died after initial good progress at 10 and 14 years of age, respectively. The third patient underwent DS at 2 years

FIGURE 1. Surgical strategy algorithm. PA/VSD, Pulmonary atresia/ventricular septal defect; PS/VSD, pulmonary stenosis/ventricular septal defect; PA, pulmonary artery; CP, cavopulmonary.

TABLE 2. Outcomes for patients undergoing anatomic repair

Outcome	DS	RS
Mortality		
No. of patients (n)	68	45
Early deaths (n)	5 (7.4%)	0 (0%)
Late deaths (n)	5 (7.4%)	7 (15.5%)
High-risk group	17	
Early deaths	3 (17.5%)	0
Late deaths	2 (11.7%)	0
Perfusion and ischemic times*		
CPB time (min)	176 (102-492)	132 (66-218)
Crossclamp time (min)	134 (69-258)	114 (44-169)
Circulatory arrest time (min)	15 (0-66)	38 (1-74)
Secondary outcome measures		
Median ICU length of stay (d)	3.2 (0.1-47.8)	5.2 (1.8-48.9)
Median hospital LOS (d)	14.0 (0-86.0)	20.5 (0-84.0)
ECMO	4 (1 late)	0
Transplanted	3	0
Neurologic event	1	3
Tachyarrhythmias after repair	3	1
PPM insertion		
Operative	10	5
Preoperative	15	0

DS, Double switch; RS, Rastelli-Senning; CPB, cardiopulmonary bypass; ICU, intensive care unit; LOS, length of stay; ECMO, extracorporeal membrane oxygenation; PPM, permanent pacemaker. *Times stated as median with range.

of age and died in the operating room. The patient had repeated episodes of ventricular fibrillation just before ICU transfer and, despite reinstitution of cardiopulmonary bypass, the heart persisted in ventricular fibrillation. No structural reason for this outcome could be found. The single patient who died after DS procedure owing to persistent ventricular fibrillation had received a PAB for protection for unrestrictive VSD and not for training the mLV. There were no further data available concerning the mechanism of death for the 2 patients who died late unexpected deaths in the RS groups, 1 of whom was followed up abroad.

In terms of secondary outcome measures, 3 patients from the DS group underwent late transplantation and 4 patients in this group required postoperative extracorporeal membrane oxygenation: 2 for postoperative cardiac support, of whom both survived with good function, and 1 late readmission requiring aortic valve replacement (AVR) who survived but has poor mLV function; the final case was for respiratory support, having developed respiratory syncytial virus–positive bronchiolitis late postoperatively (this patient was a late death). There were 4 patients who had significant neurologic injury after surgery: 1 in the DS group and 3 in the RS group (2 of whom were late deaths). The median ICU and hospital lengths of stay were similar (Table 2).

Postoperative PPM insertion for heart block was required in 10 DS patients and 6 RS patients. Three of these patients went on to have an upgrade to biventricular pacing systems to achieve better resynchronization, 1 with dramatic

improvement in mLV function. There was a low incidence of tachyarrhythmias overall. Preoperatively, 3 DS patients and 1 RS patient had supraventricular tachycardia; 1 of the 3 DS patients had Wolf-Parkinson-White syndrome. After repair, tachyarrhythmia developed in 4 patients: 3 DS and 1 RS (Table 2). All 3 of the DS patients required electrophysiologic studies and catheter ablations (1 for ventricular tachycardia; 2 for supraventricular tachycardia). Overall, 54 of the 58 patients in the DS group and all surviving 38 RS patients are in New York Heart Association class I/II.

Reinterventions After Anatomic Repair

Overall, there were 41 reinterventions in 13 DS patients and 34 interventions in 16 RS patients during follow-up. These reinterventions included both surgical and catheter procedures, although excluding insertion of PPMs and the single patient who was readmitted and required extracorporeal membrane oxygenator support. The most frequent intervention in the RS group was for right ventricular–pulmonary arterial conduit ballooning or exchange (14 patients; Table 3). Of note, 6 AVRs were required in the DS group for progressive aortic incompetence (AI), although only 1 in the RS patients. Reinterventions on the Senning pathways were required in 12 patients; 5 were operative (4 pulmonary venous obstruction and 1 superior vena cava pathway) and 7 were catheter interventions (all superior vena cava pathways). Four patients in the RS group required relief of mLV outflow tract obstruction, although in 2 cases the obstruction was not severe and was relieved as a concomitant procedure at the time of necessary conduit replacement. When censoring deaths and right ventricular–pulmonary arterial conduit interventions/changes in the RS group, the Kaplan-Meier freedom from reintervention at 10 years in both groups was very similar: DS, 50.3%; RS, 49.1% (Figure 2, B; log–rank, $P = .44$).

During follow-up, the incidence of mild to moderate AI in the DS group was 40 (of 58; 70%) compared with 8 (of 38; 21%) in the RS group, all of which were mild. An additional 6 patients in the DS cohort had severe AI during follow-up and underwent AVR, compared with 1 AVR for severe AI in the RS cohort. Aortic root annuloplasty was performed in 13 patients owing to the finding of a dilated annulus and/or prolapsing aortic leaflets at time of DS. Follow-up of these patients showed less AI in this cohort than in the whole DS group, but the difference did not reach statistical significance (Figure E1). None of these patients has required AVR.

Mild to moderate mitral regurgitation (MR) occurred during follow-up in 31% of the DS group and 14.3% in RS group. Mild to moderate TR occurred in 27.6% in the DS group compared with 40% in the RS group. No patients in either group had severe MR or TR. The stated incidence of MR represents a new incidence of MR after anatomic repair, although the precise timing of this valvular

FIGURE 2. A, Survival (death) after ccTGA repair. B, Freedom from reintervention after ccTGA repair. C, Freedom from death, transplantation, or poor mLV function. ccTGA, Congenitally corrected transposition of the great arteries; mLV, morphologically left ventricle.

dysfunction could not be clearly determined. Detailed data concerning the preoperative incidence of MR were not available and thus we have been unable to relate this to the postoperative incidence of MR. In fact, the occurrence of new MR after repair was only mild in the majority of cases (total incidence of new MR in DS group, 31%-22.4% mild; total incidence of new MR in RS group, 14.3%-11.4% mild). There was no significant difference in incidence of postoperative MR in those with/without pulmonary venous atrium supplementation, and as such it would be difficult to attribute the development of new MR to the Senning technique. Finally, only qualitative data on mLV function late after repair were available, although it would be interesting to obtain more quantitative data on mLV cavity size and geometry late postoperatively to ascertain whether these were related to the incidence of late MR.

The Lecompte maneuver was performed in 50 of the 68 DS group, and 5 of these patients required 8

reinterventions on the pulmonary arteries. This compares with zero reinterventions in the 18 patients in whom the pulmonary arteries were left in the native position (log-rank, $P = .2$; Figure E2).

Outcomes in High-Risk DS Patients and Those With Poor mLV After Repair

There were 17 high-risk patients, all in the DS group. These patients had severe heart failure and required ICU admission, ventilation, and inotropic support preoperatively; 6 of 17 patients had severe TR and 6 of 17 pulmonary hypertension. All these patients fell into the DS group and 11 of 17 were first banded with or without arch repair (Figure 1). The remaining 6 patients had DS procedures without prior banding. There were 3 early and 2 late deaths in the high-risk group (70.6% survival). Two of these early deaths were in patients who had been banded.

TABLE 3. Reinterventions for DS and RS patients

Reintervention	DS	RS
AVR	7*	1
MV repair	1	0
TV repair	3	0
RF ablation for atrial flutter	4	0
Multisite resynchronization	3	0
Aortic angioplasty	0	1
Residual VSD	3	0
LVOTO resection	0	4
SV pathway	3 (3 balloon)	5 (4 balloon; 1 stent)
PV pathway	4	0
Senning takedown	0	1
PA confluence reconstruction	8	3 (2 balloon)
RPA intervention	3 (1 balloon; 2 stents)	2
LPA intervention	0	3 (balloon)
RVOT enlargement	2	0
Balloon to RV-PA conduit	n/a	2 (balloon)
RV-PA conduit change	n/a	12
Total No. of reinterventions	41	34

DS, Double switch; RS, Rastelli-Senning; AVR, aortic valve replacement; MV, mitral valve; TV, tricuspid valve; RF, radiofrequency; VSD, ventricular septal defect; LVOTO, left ventricular outflow tract obstruction; SV, systemic venous; PV, pulmonary venous; PA, pulmonary artery; RPA, right pulmonary artery; LPA, left pulmonary artery; RVOT, right ventricular outflow tract; RV-PA, right ventricular–pulmonary arterial; n/a, not applicable. Catheter interventions are in parentheses. *Includes 1 additional AVR at the time of initial surgical repair.

If the high-risk group is compared with the remainder of the DS group (ie, elective or low-risk DS), the early mortality is 17.5% compared with 3.9% ($P = .06$). In terms of ventricular function, 5 of the 17 high-risk patients had poor mLV function after repair and 4 of these died (2 early deaths; 2 late deaths). The corollary of this is that, among the early survivors of the high-risk group, 80% have well preserved mLV function at follow-up (Table E2).

Taking into account all 113 patients with anatomic repairs, there were 16 (14.2%) patients with poor mLV function detected during follow-up. All were in the DS cohort and only 5 of the 16 were high-risk patients in heart failure before repair. Of the 11 remaining patients, 7 had been banded for mLV training. Taking these 7 patients with PAB-T who developed poor mLV function after repair, median age at banding was 3.03 years (0.29-9.82 years), with a median duration of banding of 579 days (5-939 days), and 3 of these 7 patients had required 2 PAB-T procedures. In comparison, for the 15 other patients with PAB-T who did not have poor mLV function postoperatively, median age at banding was 2.1 years (0.11-7.97 years; $P =$ not significant). Median duration of banding was 544 days (25-2758 days; $P =$ not significant), but none required more than 1 banding procedure. Whereas neither the age at banding nor duration of banding exhibited a statistically significant

difference between groups who did/did not develop late mLV dysfunction, the numbers of patients were relatively small. Further, analysis of Kaplan-Meier survival at 1, 5, and 10 years did not demonstrate any significant difference among DS patients who had undergone PAB-T compared with those who had not (log-rank, $P = .31$).

Despite a high incidence of mild to moderate AI in the DS group, there was no association between this and late mLV dysfunction (Table E2). Similarly, only 4 of these 16 patients had mild to moderate MR. When taken together as a combined end point, freedom from death, transplantation, or poor mLV function was significantly worse for the DS group at 1, 5, and 10 years when compared with the RS group (log-rank, $P = .03$; Figure 2, C). This is in contrast to the raw survival as shown in Figure 2, A.

DISCUSSION

This study has demonstrated that in a large cohort of patients with ccTGA, there is a heterogeneous mix of anatomic and functional subtypes as well as a wide variability in preoperative clinical status. Despite overall encouraging early mortality for anatomic repair, we have highlighted that there is a significant late morbidity in these patients, particularly in the DS group.

Other than the requirement for conduit replacement, the ccTGA patient with large VSD and pulmonary stenosis/pulmonary atresia (ie, the RS group) have excellent early and late outcomes and appear to be much more likely to remain free of late ventricular dysfunction or AI. We observed a low incidence (4 patients) of late subaortic obstruction in the RS group. The Senning component of the procedure is not without late morbidity, particularly pathway obstruction, although the majority of cases can be successfully treated with catheter intervention. Four of 113 patients sustained significant neurologic injury. The observed neurologic injuries did not occur in the setting of multiple cardiac arrests, although they were, we believe, partly attributable to low cardiac output after repair. Cranial imaging in these patients did not suggest that there had been embolic events, nor was there any suggestion that these events had been related to complexities of intraoperative cannulation or perfusion.

The incidence of tachyarrhythmias in our series was encouragingly low, given the concerns that have been raised with long-term follow-up of the atrial switch procedures.^{2,7} Freedom from reintervention was the same for both the DS and RS groups at 5 and 10 years, and overall reintervention rates are similar to other reported series.^{2,8,9} Despite the requirement for conduit replacement in the RS group, the overall reintervention rates are similar for both groups, predominantly owing to the increased need for aortic valvular and pulmonary artery reinterventions in the DS group.

The TV, Pulmonary, and Systemic Venous Pathways

In this series, 12.4% of patients had an Ebsteinoid anomaly of the TV. TV repair was performed at the time of repair in 13.2% of DS patients, and in over half the cases this involved closure of a cleft in the septal leaflet. Our intraoperative strategy was to briefly examine the TV in all cases. Only patients in whom there was an evident gap or cleft in the valve underwent repair at the time of DS or RS procedure. Postoperative incidence of mild to moderate TR was 27.6% in the DS group and 40% in the RS group. The high incidence in the latter may reflect an approach that could be more aggressive in terms of treating milder forms of TV dysplasia at the time of initial repair. In many cases, it was anticipated that restoration of the TV to the low-pressure pulmonary circulation would result in improvement in the TR after repair, as has been reported by other investigators.

Restoring the mRV to the low-pressure pulmonary circulation has been shown to markedly reduce TR just by virtue of mechanical unloading of the mRV and septal shift with restoration of the mLV to the systemic circulation.^{4,10} Although this is clearly important, our results would suggest that there may still be a role in assessing the TV carefully with a low threshold for repair at a time of surgery.

Senning pathway reintervention rate was 11%, which was higher than we had seen in shorter follow-up studies.⁴ Although the majority of patients were successfully treated by interventional catheterization, pulmonary venous pathway obstruction required surgical relief in all 4 patients. Others have reported similar venous pathway reintervention rates, and although the Senning is the most widely reported technique, there remains no consensus as to whether the Mustard or Senning procedure provides better long-term results in this setting.^{7,10} Our results did not show that augmentation of the pulmonary venous baffle at the time of initial surgery affected reintervention rates.

Neo-Aortic Incompetence After DS

The majority (70%) of patients after DS repair had at least mild AI at follow-up. Six of the surviving DS patients had severe AI requiring AVR, with 1 additional patient undergoing AVR at the time of repair. It is clear, however, that mild AI is well tolerated by the majority of DS patients and it did not correlate with late mLV dysfunction. There are potentially anatomic and surgical substrates for the development of AI in these patients.^{1,11} The neo-aortic root dilatation observed in the DS patients occurred primarily at the level of the ventriculoarterial junction, below the level of the coronary arteries. The purse-string suture placed was 4-0 PDS (Ethicon, Inc, Somerville, NJ) at this level as a double running suture. Clinical observations from our early experience with the DS procedure suggested that postoperative AI was associated with anatomic repair, and as a result

we examined more critically the need for reduction of the neo-aortic annulus with a 2-layer annuloplasty stitch placed before reimplantation of the coronary buttons. As such, our approach to minimize AI represents an evolution in surgical technique over time, and we now routinely perform an objective assessment of the neo-aortic valve/root at the time of repair. In the present series, a repair was performed in 13 patients in the DS group with encouraging outcomes and minimal AI at follow-up. It is possible, however, that a more aggressive approach to tailoring of the neo-aorta at the time of anatomic repair might reduce the incidence of AI, and this is an area for potential improvement in the DS procedure.

Reinterventions for Right Ventricular–Pulmonary Arterial Conduits and Pulmonary Arteries

The commonest reinterventions in the RS group were for conduit changes. This reflects the natural history of these conduits as well as natural outgrowth.¹¹⁻¹³ Malhotra and associates⁹ report improved conduit performance using their strategy of DS with one-and-a-half-type repair with cavopulmonary shunts, attributing conduit longevity to reduced forward flow. This is an attractive strategy but must be balanced by the potential concerns over the one-and-a-half-type repair (reduced functional status, problems in the setting of high pulmonary vascular resistance, and loss of access for pacing intervention).

In the DS patients, all 8 reinterventions (in 5 patients) on the pulmonary arteries occurred in patients who had undergone the Lecompte maneuver. No reinterventions were necessary in the (smaller) group of patients in whom the pulmonary arteries had been left in their native position. The Lecompte maneuver is not always straightforward in the DS inasmuch as the great vessels are frequently side-by-side; furthermore, the older age and previous procedures in these children often make the pulmonary arteries much less mobile and flexible than in a regular neonatal arterial switch. The evidence would suggest that the Lecompte maneuver may produce particular problems in the setting of the DS and that it may be more appropriate to leave the pulmonary arteries posteriorly, even if this requires supplementing the main pulmonary artery to allow more length. Our current practice is to only perform the Lecompte if the vessels are in an anteroposterior relationship and can be freely mobilized.

We did not observe any instances in which acute coronary compression resulted in early mortality. Importantly, however, in 3 patients after DS procedure, there was difficulty in weaning from cardiopulmonary bypass that appeared to be related to the geometry of the pulmonary artery reconstruction after the Lecompte maneuver. The main pulmonary artery was therefore lengthened with a subsequent uneventful postoperative course. It would be interesting, however, to determine whether any degree of chronic

coronary compression occurs long-term in these patients, and this would be an area for further study.

High-Risk Patients Undergoing DS and Poor mLV Function After Repair

In this study we focused in particular on outcomes achieved in high-risk patients with ccTGA who had severe heart failure with associated moderate to severe TR and pulmonary hypertension in some cases. Significant TR and impaired mRV function before repair have been widely documented to be risk factors for mortality after physiologic and anatomic repair for ccTGA.^{2,7,14,15} All of these high-risk patients fell into the DS group in terms of underlying anatomy, and it is important to note that the VSD/pulmonary stenosis group of patients do not present with uncontrolled heart failure or severe TR. The poor preoperative status of this group of patients was reflected in the relatively high early mortality (17.5%), but it was encouraging to note that the long-term outcome for this group was good, being similar to that of the low-risk DS and with no increased incidence of poor mLV function.

During follow-up, 14.2% of patients had poor mLV function, all of whom were in the DS group; an incidence of 18% to 20% has been reported by others, and this may be both systolic and diastolic dysfunction.^{7,10,16} In terms of the etiology of this marked impairment of function, possible factors to consider are the following: PAB-T duration and age at training; presence of neo-AI; presence of MR after repair; heart block requiring PPM insertion; and preoperative heart failure.¹⁶

The need to train the mLV before DS has previously been cited as a risk factor for late outcome.¹⁷ However, the use of PAB-T was not associated with poor outcome in this analysis. Furthermore, we found that neither age at banding nor duration influenced development of late mLV dysfunction. One aspect of our strategy that has evolved over time is the tendency to apply PAB-T in younger patients inasmuch as they appear to have a more favorable response, perhaps related to hyperplasia of cardiac myocytes as opposed to a hypertrophic response.¹⁸

Despite mild AI being common, our results did not suggest that either AI or MR was correlated with deterioration in mLV function, as only 4 of 16 patients with poor mLV function had associated moderate to severe AI and only 4 of 16 had associated mild to moderate MR. Further, only 3 of 16 patients required postoperative PPM insertion, although data on electrocardiographic QRS complex duration, predictive of mLV dysfunction in this context, were not available.¹⁶

It is difficult, therefore, to attribute mLV failure to any single factor in our patient cohort. The cause is likely to be multifactorial, although it was notable that late mLV dysfunction occurred exclusively in the DS group. One further observation is the presence of mild mLV dilatation in the

majority of patients, although this is difficult to quantify objectively. Early, reproducible, standardized assessment of such subtle changes in mLV size, geometry, and function might identify patients who would benefit from alteration of their medical therapy to perhaps modulate any potential adverse remodeling of the myocardium and improve long-term clinical outcomes and functional status. In our series, 3 patients received multisite pacing for resynchronization therapy, and an earlier role for this in the deteriorating mLV may prove valuable.¹⁶ The role of pharmacologic modulation of mLV function, such as with angiotensin-converting enzyme inhibitors, was not formally evaluated in this study.

CONCLUSIONS

Although the majority of patients remain well after anatomic repair of ccTGA, there is significant late morbidity. Long-term surveillance of patients is required to identify any deterioration in systemic mLV, atrioventricular valve, and neo-aortic valve function, as well as to detect Senning pathway obstructions. The DS group is at significantly higher risk for the development of late mLV dysfunction and AI than the RS group. Careful assessment of the aortic valve at the time of DS and avoidance of the Lecompte maneuver in side-by-side vessel arrangements may help to improve long-term outcomes. Anatomic repair has encouraging results for symptomatic patients, in particular those with significant TR and severe heart failure.

We acknowledge our colleagues in the Department of Cardiology (Dr J. DeGiovanni, Dr J. Wright, Dr P. Miller, Dr R. Dhillon, Dr A. Chikermane, and Dr T. Desai) for help with the care of all patients, as well as the ICU staff and Departments of Perfusion and Anesthesia at Birmingham Children's Hospital. We are also grateful to Dr Sara Thorne and Dr Paul Clift for their help in obtaining follow-up data for patients and to Mr Joe Eurell for assistance in data collection.

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Discussion

Dr Richard A. Jonas (Washington, DC). I have no disclosures.

Dr Barron, many thanks for sending me the manuscript. This is obviously a complex area and you have a huge amount of data, so it was very helpful to be able to look at the manuscript ahead of time.

You are certainly to be congratulated on this additional, extremely important contribution regarding the challenging problem of ccTGA. It was just over 20 years ago that Dr Michel Ilbawi introduced the concept of the double switch (DS) procedure and held out hope for this difficult problem of ccTGA. Until that time the results of traditional surgery, which left the right ventricle as the systemic ventricle, resulted in dismal long-term outcomes, as was so ably documented by Dr Viktor Hraska when he was with our group in Boston. Although encouraging early results were reported for the DS by Professor Imai's group at Tokyo Women's Medical College and Dr Roger Mee in Melbourne, as well as Dr Brawn and your group in Birmingham, this most recent analysis

will surely lead all of us to rethink which patients are best managed by this procedure.

I would like to highlight some of the disturbing points that Dr Barron has just presented to us. First, late left ventricular dysfunction is not uncommon. Recent reports had already raised that suspicion, but what is of greater concern is that late left ventricular dysfunction cannot be accurately predicted. I think we were all hopeful that training of the left ventricle at an older age, such as greater than 12 or 15 years, or the presence of preoperative heart failure or the presence of postoperative valve dysfunction might have been predictive of late left ventricular dysfunction. Regrettably, none of this has been documented by the analysis we have just heard. In fact, this large experience raises the question whether the fundamental premise that the child with ccTGA must always be better off with a systemic mLv and a biventricular circulation is correct.

What is of equal concern to the high incidence of late left ventricular dysfunction is the high incidence of late need for reoperation and reintervention. The DS patients not only had a higher incidence of late left ventricular dysfunction relative to the RS patients, but in addition they had a surprisingly high rate of late aortic valve regurgitation and pulmonary artery problems, particularly after a Lecompte maneuver. These problems were less frequently seen in the RS patients, but surely this latter option is in other respects a less desirable option than a true DS since it condemns patients to a lifetime of conduit changes. Conduit changes can be a particular problem when the patient has dextrocardia, which applied to more than 20% of your patients in this series and in many other series has applied to as many as one third of all patients. And for both groups reoperations were needed for the Senning pathways, both systemic and pulmonary, which leads to my questions:

First, is there ever a place for a prophylactic DS in the child with an intact septum and no TR? What sort of associated problems like a mild degree of Ebstein-like malformation of the TV or congenital heart block might lead you to do a prophylactic DS?

Dr Barron. Thank you very much for your very fair comments. I completely agree. It has raised concerns in all our minds about the long-term management of these patients. Absolutely, it is patient selection—who needs what operation at what time—that is the key.

I am not sure whether many people would ever have suggested that doing a prophylactic DS in a child who is completely well, has no TR, and a good functioning systemic right ventricle, that putting them through this major maze operation, would be the right thing to do. So no, I do not think there is any place for prophylactic DS.

Now, if they have a degree of TR, then it starts to get a bit trickier because the natural history is bad. As soon as they have moderate or greater TR, they are on a bad survival curve, and that is more difficult. The first thing I would say is that I think there is a place for a PAB in that situation, because that in itself may treat the TR and buy some time. You are setting them on that track toward DS at that stage. However, that may well be their best outcome despite what we are saying here.

Dr Jonas. Would it be fair to say that the bar is shifted slightly against the DS in terms of these patients who do not have a VSD and who have an intact septum?

Dr Barron. Yes, I think so. That's a fair comment. Of course, we are all conscious of the patients in whom symptoms develop. The more difficult problem is the ones who do not have symptoms but whose right ventricle does not look good or who have some TR.

Dr Jonas. Second, how important is dextrocardia as a contraindication to an RS procedure in view of the risk of proximal conduit compression by the sternum and need for frequent conduit changes?

Dr Barron. In our series I would not say it has been a major concern, and we certainly would not regard it as a contraindication to the DS. Certainly dextrocardia makes the operation technically more difficult to do, but we can be reassured by the fact that the operative results for the RS have been very good. If you look at the incidence, we have only had to actually replace 12 conduits in the whole population over the 19 years of this study. So the commitment to lots of operations for conduit replacement does not seem to be too bad.

We have not analyzed it specifically to look at whether the dextrocardia patients are those who have needed more conduit replacement, so I could not answer that.

Dr Jonas. Third, I have a question about PAB. To whom do you now offer left ventricular preparation by PAB? Do you see a role for PAB as destination therapy to induce septal shift to treat TR in the patient with an intact septum and low left ventricular pressure?

Dr Barron. I do not think we would consider PAB unless the patients had evidence that they were running into trouble, that they had moderate TR, or that they had some dysfunction or some signs of dilatation of the mRV. Those are the patients we would select for banding at that point. I quite agree that the banding itself will cause septal shift and may reduce the TR and symptoms in many of the patients.

Whether that will be the destination, I do not think I could answer. I think each patient must be evaluated individually. Once PAB is done, you are making a commitment that you would be expecting to take them through a DS. If they remain well with the PAB on, they should be safe and there is no reason not to just keep watching them, indefinitely if need be. But in our experience, with the majority of patients, once TR and right ventricular dilatation have developed, they are going to ultimately get systemic ventricular failure and they will need something.

Dr Jonas. You can certainly make the argument that the left ventricle is going to manage the banded pressure over the longer-term. If the TR is stable and the septum remains shifted across, perhaps in the long-term that would be a more satisfactory outcome than some of the multiple reinterventions that you have documented.

Final question: What is the role of the Fontan operation versus the DS in light of the results that you have presented?

Dr Barron. There are certain populations within this group of patients for whom the Fontan would be a good and a valid alternative. I do not think there is any role for it in the DS-type population, because you have not removed the concern of the mRV failing or the TR becoming a problem. If you take them through a Fontan program, that risk is still there.

I guess the Fontan is more appropriate in the group of patients who are in the RS group, in whom you would avoid the whole problem of having to place a conduit, perhaps the risk of heart block and the risk of leaving a left ventricle-aortic tunnel. And you know that the Fontan is a pretty safe operation.

We are all kind of hard-wired to think that if the patients have 2 ventricles, it is a sin to take them through a Fontan circulation, and that is something we can debate. However, I think the functional performance and the functional result the patients get with a biventricular repair, which in this case means RS versus the Fontan, is better with an RS. Thus I would still put my vote for a biventricular repair with an RS, and I think that is supported by these outcomes. We know that the Fontan, although safe, has a long-term attrition. I would argue it might be worse than we are going to see in the RS group. Certainly there are patients in the RS group who have an uncommittable VSD, and in those patients I think there is a very good argument for doing a Fontan rather than a conventional physiologic repair.

Dr Jonas. Thank you. Once again, congratulations, this is going to be a really important contribution.

Dr Rodolfo A. Neirotti (*Cambridge, Mass*). Congratulations for your large number of patients and for being honest in presenting the data in which you have shown some long-term problems that can be useful for others trying to use the same approach.

I would like to expand on Richard Jonas's question, and that is, the place of the Fontan operation. In patients with ccTGA and large VSDs, the ventricular dysfunction appears usually after the septation. In this cohort of patients, after the Fontan operation, the 2 ventricles function as a large single chamber, with a single outlet, an anatomy and physiology that may result in a better ventricular function than when you do the septation and the DS.

Dr Barron. No, I think that is a fair comment. If you use the Fontan approach, they will fall into the better series of Fontan outcomes. However, I still believe that a biventricular repair provides a better functional result. The results here would support the fact that so long as they have a suitable anatomy where the Rastelli is readily performed, the left ventricle can be committed back to the aorta without causing any obstruction within the ventricle. I think the functional outcome for those patients would still be superior than to do a Fontan.

Dr V. S. Reddy. David, I think you can discuss it later because we are way over time.

FIGURE E1. Follow-up after aortic root annuloplasty.

FIGURE E2. Follow-up after the Lecompte maneuver.

TABLE E2. Development of poor mLV function during follow-up

	Moderate/good mLV	Poor mLV
Double switch	42/63 (75%)	16/63 (25%)
Rastelli-Senning	45/45 (100%)	0/45 (0%)
High-risk group	12/15 (80%)	3/15 (20%)
≥Moderate AI or AVR	7/92 (6%)	3/16 (19%)
Previous PAB	53/92 (58%)	7/16 (44%)
Duration PAB	554 d	579 d

Early deaths were excluded. *mLV*, Morphologically left ventricle; *AI*, aortic insufficiency; *AVR*, aortic valve replacement; *PAB*, pulmonary artery band.

TABLE E1. Concomitant surgical procedures at time of anatomic repair

Group	Concomitant procedure				
	Aortoplasty/neo-AV repair	MV repair	TV repair	PV atrium supplementation	Lecompte maneuver
DS (n = 68)	13	3	9	19	50
RS (n = 45)	1	0	0	17	0

Figures are numbers of patients. *AV*, Aortic valve; *MV*, mitral valve; *TV*, tricuspid valve; *PV*, pulmonary venous; *DS*, Double switch; *RS*, Rastelli-Senning.