Double switch for congenitally corrected transposition of the great arteries

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Received 21 September 1998; received in revised form 16 November 1998; accepted 25 November 1998

Abstract

Objective: To evaluate the early and medium term results of operations for congenitally corrected transposition of the great arteries (CCTGA) wherein the left ventricle becomes the systemic ventricle. Methods: Fourteen patients with CCTGA who underwent anatomic repair from 1994 to 1998 were placed in one of two groups: those without pulmonic stenosis (PS) ($N=7$) and those with PS ($N=7$). Main associations in the group without PS were: severe left atrioventricular (AV) valve regurgitation ($N=4$), ventricular septal defect (VSD) ($N=4$), criss-cross AV connection ($N=1$), and hypoplasia of the morphologic RV ($N=1$). Main associations in the group with PS were: VSD ($N=7$), anomalous pulmonary venous drainage ($N=2$) and major aorto-pulmonary collaterals ($N=1$). All patients in the group without PS underwent an arterial switch operation and all except one had a modified senning repair for atrial rerouting for anatomic correction. All seven patients with PS underwent a Rastelli repair and all except one needed a modified senning repair. Results: Early survivorship in both groups was 6/7. Follow-up ranges from 1 month to 48 months. The mean left ventricular ejection fraction (LVEF) in the arterial switch and senning patients was 65% and in the Rastelli and atrial rerouting was 52%. None of the survivors except one have any significant mitral regurgitation (MR). The one exception resulted from iatrogenic damage to a chorda during VSD closure. All except two patients are class I symptomatic and all are in normal sinus rhythm. Conclusion: Satisfactory early and mid term results of anatomic repair support the double switch option as the procedure of choice for patients of CCTGA amenable to biventricular repair. © 1999 Elsevier Science B.V. All rights reserved.

Keywords: Great arteries; Congenitally corrected transposition; Surgical repair

1. Introduction

Disappointing long-term results of ‘classic’ repair of lesions associated with atrioventricular and ventriculoarterial discordance [1–8] provided us with the impetus to explore alternative available options for patients with congenitally corrected transposition of the great arteries (CCTGA).

Encouraging intermediate term follow-up of surgical treatment of CCTGA subsets wherein the morphologic left ventricle becomes the systemic ventricle [9–13] prompted us to attempt the ‘double switch’ for patients of CCTGA presenting to us, wherein a biventricular repair was feasible. This report presents our initial results with this approach.

2. Materials and methods

Between January 1994 and June 1998, 14 patients of CCTGA underwent surgical repair wherein the morphologic left ventricle (LV) was made the systemic ventricle. These patients were divided into two groups depending on the presence or absence of a normal pulmonary valve. There were seven patients in each of the two groups. There were four boys and three girls in both groups. Age ranged from 11 months to 18 years (mean 4 years) in the group with a normal pulmonary valve, and from 18 months to 18 years (mean 7 years 4 months) in the group with pulmonic stenosis (PS). Associated lesions are shown in Table 1.
2.1. Pre-operative characteristics

2.1.1. Patients with a normal pulmonary valve

Five out of seven patients had severe pulmonary arterial hypertension with systemic pulmonary artery pressures (four with ventricular septal defect (VSD) and one with severe left atrioventricular valve regurgitation (LAVVR)). The remaining two had pulmonary artery pressures that were two-thirds of the systemic pressure with no gradients across the pulmonary valve.

Right ventricular (RV) dimensions ranged from 50 to 150% of the predicted normal (mean 90%) [14]. One patient with RV volume that was 50% of the normal also had associated criss-cross atrioventricular (AV) relationship.

Left ventricular (LV) dimensions ranged from 110 to 140% (mean 120%) of the predicted normal [15,16].

One patient had severe biventricular dysfunction with RV ejection fraction (EF) of 20% and LVEF of 40%

All patients had a normal right (morphologically mitral) AV valve.

2.1.2. Patients with a stenotic pulmonary valve

Five out of seven patients had double outlet of the right ventricle (DORV). The single patient with total anomalous pulmonary venous drainage (TAPVD) had mixed pulmonary venous drainage with the left pulmonary veins opening directly into the right atrium, and the right pulmonary veins opening into the coronary sinus in a patient with situs inversus.

Pre-operative saturation ranged from 60 to 80% and one patient had a prior Blalock shunt.

RV dimensions ranged from 60 to 110% of the predicted normal (mean 90%) and LV dimensions from 90 to 110% (mean 95%) of the normal.

2.2. Operative technique

Standard cardiopulmonary bypass (CPB) with ascending aortic and bi-or tricaval cannulation was utilized with cooling to 24°C. No period of circulatory arrest was used. Myocardial protection was achieved by blood cardioplegia administered every 30–40 min into the aortic root. An exception occurred during the arterial switch when the time on single cardioplegia exceeded 60 min. The first dose of cardioplegia was preceded by a bolus of 6 mg of Adenosine given into the aortic root to achieve immediate cessation of all electrical activity.

Total perfusion time ranged from 210 to 360 min (mean 270 min), and aortic cross-clamp time ranged from 168 to 180 min (mean 170 min).

2.3. Modified Senning procedure

Because of the generally small size of the right atrium (RA) and overhanging position of the morphological LV in CCTGA, the right atriotomy was made about 8 mm from and parallel to the right AV groove, so that almost the entire free wall of the RA went towards formation of the systemic venous baffle. When present, the coronary sinus (CS) was left draining into the pulmonary venous chamber. A CS receiving a left superior cava was unroofed and the left cava directed into the tricuspid valve by placing the atrial septal flap posterior to its opening. An in situ pericardial flap was utilized to complete the pulmonary venous chamber and almost the entire wall of this chamber from the opening of the pulmonary veins to the fringe of the RA next to the right AV groove was constituted of pericardium at the completion of the Senning procedure.

Two patients did not require a Senning procedure, as the relative position of the pulmonary venous openings and the mitral valve orifice were such that an intraatrial baffle sufficed to direct pulmonary venous inflow into the mitral valve, leaving the rest of the atrial cavity for systemic venous drainage towards the tricuspid valve.

2.4. VSD closure

In the group without PS, one patient had VSD closure from the aorta, while the other three were closed through the right atrium. A single continuous suture starting on the left of the superior septum was used running along the ridge so raised till the lower edge of the VSD was reached. Here the suture line was brought to the right side of the septum. A dacron patch was utilized for VSD closure in all patients. In the group where a Rastelli repair was part of the double switch, the VSD was closed through a right ventriculotomy. The ventriculotomy was made in the mid-portion of the RV and suturing started from the tricuspid annulus posteriorly. The pulmonary outflow was included in the leftward aspect of the patch which was taken around the aortic valve annulus. One patient required enlargement of the VSD which was done in the posterior-inferior region. Again, a single continuous suture was utilized for VSD closure. This was reinforced with a few interrupted pledgeted mattress sutures.

2.5. Arterial switch operation

Coronary artery anatomy was identical in all seven patients undergoing the arterial switch operation (Fig. 1). The origin of the coronary arteries differed from those seen generally in D-transposition of the great arteries in that (i) the ostia were located deeper in the aortic sinuses and (ii) there was high incidence of early bifurcation of both the left coronary artery and the right coronary artery. Also, the right coronary ostium was often close to a pulmonary valve commissure [17]. These factors acted to reduce the leeway available for rotating the right coronary button and suturing to the rightward and posterior main pulmonary artery (MPA). All coronary buttons were sutured into punched out defects or excised wedges in the MPA or by utilizing medially based trapdoor flaps. While the anterior left coronary artery (LCA) button was generally not a problem, the posterior right coronary artery RCA origin has been recently tackled...
routinely by utilizing a direct suture of the posterior edge of the button to the posterior wall of the punch hole, and using a small pericardial patch between the PA wall and the anterior edge of the button, so as to allow the RCA to lie in the orientation it had held when it originated from the aorta (Fig. 1).

The Lecompte manoeuvre was performed as part of all arterial switches. A generous piece of autologous untreated pericardium was used to fill up the gaps created in the proximal aorta after excision of the coronary buttons, but before implanting the coronaries into the proximal MPA. The distal anastomosis with the PA bifurcation was then made, taking care to avoid any twist of the neo-PA.

2.6. Conduit insertion

In patients where a conduit was placed, after ligating the proximal MPA, a pulmonary arteriotomy was made, and a suitably sized aortic homograft sutured in place using a direct anastomosis distally and a pericardial hood proximally.

2.7. Follow-up

Patients were followed-up after hospital discharge, clinically and echocardiographically. Two patients in each group have undergone cardiac catheterization at a mean follow-up of 1 year.

3. Early results

The early outcome of patients is shown in Table 2.

3.1. Arterial switch and atrial rerouting

There was one operative mortality in a patient (No. 3) who could not be weaned from CPB. There was no other early mortality. All patients had primary sternal closure in the operating room.

Time spent on ventilator was 48–120 h (mean 52 h). All patients except one were in sinus rhythm at the time of discharge. This patient had ectopic atrial tachycardia which reverted spontaneously to sinus rhythm on follow-up.

Two patients had global left ventricular dysfunction (echocardiographically) with low left atrial pressures in the intensive care unit. However, they could be extubated and weaned off inotropes without difficulty.

Hospital stay ranged from 15 days to 2 months (mean 20 days).

3.2. Rastelli repair and atrial rerouting

There was one early death, in a patient requiring concomitant ligation of major aorto pulmonary collaterals and enlargement of a restrictive VSD. Complete AV block and severe low cardiac output led to his demise. There was no other hospital death. All patients had primary closure of their sternotomies.

Time on ventilator ranged from 24 to 72 h (mean 36 h). One patient developed disruption of the VSD patch and had to be reoperated 21 days following his first surgery from which he recovered uneventfully.

Hospital stay ranged from 15 days to 2 months (mean 40 days). Five patients needed to remain in hospital because of prolonged pleural effusions. However, no haemodynamic cause could be detected by echocardiography.

All survivors were discharged in sinus rhythm.

3.3. Follow-up

Follow-up ranges from 1 month to 48 months (mean 21 months) for the patients undergoing an arterial switch and atrial rerouting, and from 1 month to 24 months (mean 13 months) for the group undergoing the Rastelli procedure with atrial rerouting, and is 100% complete in both groups.

There has been no late death in either group.

3.4. Arterial switch and senning

3.4.1. Clinical

One patient developed iatrogenic avulsion of a mitral valve chorda during VSD closure with unsuccessful reimplantation at the same operation. She continues to have at
least moderate mitral regurgitation and needs decongestive medication for the same. Another patient suffers from episodes of atrial tachycardia, probably as a result of the senning component of the repair and needs antiarrhythmic medication. The rest of the survivors are in excellent condition and off all medication.

3.5. Echocardiographic data

All patient have undergone transthoracic echocardiogram at regular intervals following repair. The two patients who had global left ventricular dysfunction immediately following repair have recovered normal left ventricular function. LVEF of the entire group ranges from 60 to 70% (mean 65%).

Patients who had LAVVR preoperatively have only mild residual tricuspid incompetence following conversion of this valve’s role to the pulmonary circuit. One patient has moderate mitral incompetence. All others survivors have no mitral valve abnormality.

None of the patients has more than mild semilunar valve in competency.

There is no incidence of obstruction to either systemic or pulmonary venous baffle.

3.6. Cardiac catheterisation

There is no residual VSD in the two patients who were catheterized. Biventricular function is normal and pulmonary artery pressures have normalized.

3.7. Rastelli repair with senning

3.7.1. Clinical

Signs of right heart failure persisted for a mean of 3 months following hospital discharge. Patients have been kept on decongeoative medication for 4–6 months following surgery. There has been no recurrence of pleural effusions following discharge. All patients continue to be in sinus rhythm.

3.7.2. Echocardiography

LVEF ranges from 45 to 55% (mean 52%). There is no mitral or significant tricuspid incompetence in any of the survivors.

Trans-conduit gradients are less than 20 mmHg in all patients. Two patients have inconsequential residual VSDs.

3.7.3. Cardiac catheterization

Both patients who have undergone catheterization have no residual VSD, no gradient across the extracardiac conduit and have good biventricular function.

4. Discussion

Suboptimal function of the systemic right ventricle either at rest or on exercise is well-established. This has been extensively investigated and elaborated in lesions with ventriculo arterial discordance with or without atrioventricular discordance [1,2,18,19]. Coexistence of abnormalities of left AV valve structure and function in CCTGA [20] predispenses the patient of CCTGA to more rapid clinical deterioration following classic repairs than the patient with simple transposition of the great arteries undergoing atrial level repair.

Attractive as the concept of anatomic repair in these double discordance lesions is, there are certain issues that need to be addressed for satisfactory outcome following repair.
4.1. Suitability of the morphologic LV for taking on the systemic after load

In two of the patients without PS, LV pressure was sub-systemic and was found to be two thirds of RV pressure at preoperative catheterization. However, LV free wall thickness comparable with normal in age-matched controls [21], gave us the confidence to proceed with a single stage senning and arterial switch without preparatory pulmonary artery banding.

4.2. Existence of ventricular hypoplasia of different degrees

Extreme degree of hypoplasia would mitigate against use of a two ventricular approach. However, 50% of RV size (with near normal tricuspid valve annulus diameter) is probably sufficient especially when accompanied by an increased pulmonary blood flow physiology (i.e. VSD alone and no PS). In the two such patients in this series, the baffle between the pulmonary and systemic venous compartments of the senning repair was fenestrated by a 4 mm punch hole to allow for pretricuspid right to left shunting to maintain adequate cardiac output.

4.3. Routability of the VSD

This is applicable only to patients with PS. A prominent conal septum would necessitate excision to make the VSD routable to the aorta. In the absence of a sizeable conal septum, a restrictive VSD would need to be enlarged in the posterio-inferior region. In the single patient requiring VSD enlargement in our series, the outcome was one of severe low cardiac output accompanied by complete AV block. The location of the bundle in the anterior superior edge of the VSD on the left ventricular aspect of the septum is fairly constant in AV discordance lesions with situs solitus, but is not that clear in association with situs inversus [22–24].

The length of the VSD patch used to connect the VSD to the aortic annulus must be exact. In some hearts with DORV in association with CCTGA, the aortic valve may be fairly distant from the interventricular septum, and a patch of inadequate length predisposes to recurrence of VSD as happened in one patient.

4.4. Atrial rerouting

A small right atrium with an overhanging morphologic LV is quite common in hearts with CCTGA, especially when there is associated dextrocardia or mesocardia. In contrast to groups who have preferred a Mustard operation in patients with small right atria [9–12], we have uniformly utilized the senning approach with the modification mention above. Follow-up has failed to reveal any venous inflow problems with use of this technique.

Sometimes a formal Senning procedure may not be necessary as in two cases in this series. One patient had coexistent mixed intracardiac TAPVD with the two left sided pulmonary veins opening in the right atrium and the two right sided veins opening into the coronary sinus. An intraatrial baffle could easily direct the left pulmonary veins and the coronary sinus into the mitral valve.

The other patient had criss-cross AV relationship and the presence of the mitral valve in a posterior plane allowed atrial redirection by simple atrial septectomy, followed by a generous patch covering the pulmonary veins and the mitral valve.

4.5. Coronary anatomy

Normal myocardial perfusion following completion of repair is a sine-qua-non for successful outcome of repairs when the pulmonary valve is normal. Coronary anatomy in this group was fairly uniform in all seven patients. In the first two patients of this experience, a simple transfer of coronary buttons to appropriate sites on the proximal PA achieved satisfactory coronary perfusion. In the third patient, however, failure to wean from CPB was probably due to an unrecognized coronary problem. This possibility was retrospectively recognized when after completion of surgery, utilizing direct anastomosis of coronary buttons to excised wedges/punch holes in the PA, patient 4 also failed to wean from CPB. Direct inspection of the RCA revealed evidence of non-perfusion and revision of this Anastomosis utilizing an anterior pericardial patch augmentation of the button (Fig. 1) resulted in uneventful recovery. We have subsequently employed this modification in all posterior RCA to PA anastomoses in CCTGA with good result.

Prolonged chest tube drainage has been recognized by us as a common occurrence in patients undergoing a Rastelli and Senning procedure (4/6 patients) and has been in the absence of any significant residual anatomic defects. The explanation probably lies in the RV dysfunction that follows a right ventriculotomy. A fenestration of the senning septation may help in reducing the problem as it has in univentricular repairs. We have been reluctant to attempt a valve-less right ventricle to pulmonary artery connection with or without a unicusps, for fear of further aggravating right heart failure in these patients. All patients have therefore received aortic valve homografts for RV to PA connection.

Anatomic repair of CCTGA seems to offer many advantages over previously available classic repairs (AV valve repair/replacement and VSD closure with or without conduit placement between LV and PA). It, however, does suffer from the disadvantage that it may not be applicable to all VSD locations (when associated with PS) or in patients with marked imbalance of ventricular sizes. Here a choice between the classic repair and a univentricular repair would be appropriate [25,26].

So far, left ventricular function in the medium term is well preserved and is accompanied by normal mitral valve
function in 11 out of 12 survivors. Longer follow-up is essential to determine whether this trend continues in the long-term, and whether myocardial perfusion continues to be supported by the newly constructed coronary anastomoses.

References


Appendix A. Conference discussion

Dr J. Moll (Lodz, Poland): How did you prepare the patient without pulmonary stenosis? Because you have to have some criteria to operate this patient. You didn’t say anything. Maybe I didn’t understand it. But could you tell us?

Dr Sharma: Since this was early in our experience, we selected patients who already had elevated pulmonary artery pressures. Five out of seven had systemic pulmonary artery pressures either as a result of severe AV valve regurgitation or because of a large VSD. The other two patients had a two third systemic pressure. However, on the basis of left ventricular wall thickness compared with normal age and weight-matched controls, we decided to go ahead with a single stage double-switch operation. If in future, we have a patient in whom the left ventricle is unprepared, we would do a pulmonary artery band to prepare the left ventricle for a subsequent double-switch operation.

Dr Moll: Then you did some bands?

Dr Sharma: No, we did not do any pulmonary artery bands. They were all primary double-switch operations, as all patients had a prepared left ventricle.

Dr R. Neirotti (Grand Rapids, MI, USA): Do you think that you change the natural history of patients with corrected transposition by doing this double-switch operation? I have recently put a pacemaker in a patient that had this kind of surgery elsewhere because of late development of AV block.
Dr Sharma: It is certainly too early to say if we are changing the natural history of this condition. But I think it is certainly a preferable option than the earlier classic repairs, in which we can have development of systemic atrioventricular valve regurgitation as a secondary feature of closure of the VSD alone even if it did not exist earlier. Post operatively, we have good ventricular function and a functionally normal mitral valve on mid-term follow-up, and hopefully these advantages will hold out in the long-term results.

Dr J. Quaegebeur (New York, NY, USA): Would you maybe comment on the coronary artery anatomy in the patients who underwent an arterial switch operation?

Dr Sharma: All the patients that we encountered had the standard coronary artery anatomy with coronary arteries arising from facing sinuses. The only difference was, as mentioned in Dr. Anderson’s paper recently in the Journal of Thoracic and Cardiovascular Surgery, that the coronary artery origins were located deeper in the sinuses. Therefore, the coronary artery buttons had to be mobilized more than in the switch operation for D-transposition of the great arteries. The left coronary artery is usually not a problem, and it can be implanted with a little bit of stretch. But the right coronary artery comes off from the anterior aorta with its sinus of origin facing directly anteriorly and it really likes to maintain the same position following transfer to the posterior and rightward pulmonary artery. For this reason, we had to put an anterior pericardial patch on the coronary button with a posterior direct anastomosis, and we have done it in the last three patients without any problem.