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Original Article

Preoperative identification of coronary arterial anatomy in complete transposition, and outcome after the arterial switch operation*

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Abstract *Background:* Perceived correlation between the coronary arterial anatomy in patients with complete transposition, and the outcome of the arterial switch procedure, has made preoperative identification of their patterns standard practice. *Purpose:* Our purpose was to assess the accuracy of preoperative echocardiographic identification of coronary arterial patterns, to evaluate the necessity of preoperative imaging by angiography, and to determine the impact of the coronary arterial anatomy on outcome. *Methods:* We reviewed the medical records of all patients referred for an arterial switch operation between August 1995 and January 2000. The anatomy as described at the time of the operation using the Leiden convention was compared to the preoperative echocardiographic and angiographic findings. *Results:* The procedure had been performed in 67 patients, at a mean age of 9 days, with a range from 3 days to 15 months. In 42 patients, the ventricular septum was intact, while 21 patients had a ventricular septal defect, and the other four had double outlet right ventricle with the aorta anterior and rightward. In 52 patients, the left coronary artery arose from sinus #1, and the right from sinus #2. In 8 patients, the interventricular branch of the left coronary artery arose from sinus #1, with the circumflex coronary artery arising together with the right coronary artery from sinus #2. In three patients, all three coronary arteries arose from sinus #1, while in the remaining individual patients, a large conal branch arose with the left coronary artery from sinus #1, the right coronary and left anterior descending arteries arose from sinus #1, all three coronary arteries took origin from sinus #2, and the left anterior descending and right coronary artery arose from sinus #1 with no circumflex coronary artery identifiable, respectively. In two patients (4%), we identified an intramural coronary arterial course. Echocardiography and angiography were comparable (81% versus 86%) in delineating the coronary arterial anatomy. Patients with a single arterial orifice, or an atypical coronary arterial anatomy, had a slightly longer stay on the intensive care unit, and in the hospital, but showed no difference in mortality. In fact, there was no early mortality (70% confidence limits; 0–2.9%), while two patients died late (2.9%). *Conclusion:* We conclude that complex coronary arterial anatomy does not preclude a successful arterial switch procedure, although patients with a single coronary artery, or other arterial patterns, had a slightly longer hospital course. Preoperative echocardiographic evaluation is comparable to non-selective coronary angiography. Irrespective of complexity, nonetheless, the coronary arteries can successfully be translocated, obviating the need for preoperative coronary angiography.

Keywords: Discordant ventriculo-arterial connections; transposition of great arteries; coronary artery

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The arterial switch procedure has become the standard treatment for patients with concordant atrioventricular and discordant ventriculo-arterial connections, so-called d-transposition of the great arteries, or complete transposition.[1–3] The procedure requires successful re-establishment of flow of blood from the aorta to the coronary arteries, without narrowing or distorting the arteries themselves. Thus, variations in patterns of branching of the arteries have important clinical implications.[4–9] Historically, some patients with complicated coronary arterial anatomy have been denied an arterial switch procedure. In particular, patients with a single coronary artery arising from the right coronary aortic sinus, or the so-called "inverted" pattern, with the circumflex coronary artery arising from the right facing sinus, and the right coronary and left anterior descending arteries arising from the left facing sinus, have been identified as patterns producing an increased risk.[5] The standard of practice has been preoperative identification of the specific anatomy by echocardiography and/or angiography. There have been several systems proposed for the classification of the patterns of branching, but some have expressed concern over this, given that certain patterns are not included in many of these systems.[8] To circumvent this problem, Li et al.[8] proposed a descriptive system which encompasses all potential permutations, hence avoiding pre-existing alphanumeric classifications. The purpose of our study was to assess the accuracy of assessment of coronary arterial anatomy by echocardiography prior to surgery, to determine the necessity of evaluating this anatomy preoperatively by angiography, and to determine the impact of the coronary arterial anatomy on the outcome of the arterial switch operation.

Patients and methods

We reviewed the medical records of all patients with concordant atrioventricular and discordant ventriculo-arterial connections, along with those having double outlet right ventricle with the aorta anterior and rightward who were referred for arterial switch repair between August 1995 and January 2000. Preoperative echocardiographic and angiographic analysis of coronary arterial anatomy was documented and compared in all patients. Coronary angiography had been performed at the time of balloon atrial septostomy in 38 patients. We used the determination of coronary arterial anatomy at surgery as our gold standard, and made comparisons with the echocardiographic and angiographic findings.

The origins of the coronary arteries were described as they would be visualized by an observer placed in the pulmonary trunk facing the aorta, with the left hand pointing to the left-facing sinus, and the right hand pointing to the right-facing sinus. We then described the right facing sinus seen from the pulmonary trunk as #2, and the left facing sinus as #1, according to the Leiden convention.[9]

Operative technique

In those with discordant ventriculo-arterial connections, single venous cannulation and full flow cardiopulmonary bypass at 150 cc/kg/min was used. Deep hypothermic circulatory arrest at 22°C was used only for repair of the atrial septal defect. This accounts for less than 10 min of the period required for circulatory arrest. In patients with a ventricular septal defect, and those with double outlet from the right ventricle, either single or double venous cannulation was used. Deep hypothermic circulatory arrest or full flow cardiopulmonary bypass was used for closure of the ventricular and atrial septal defects.

We used previously described methods for transfer of the coronary arteries.[10–12] A standard technique was used to repair two separate coronary arterial orifices (Fig. 1). The coronary arteries were minimally mobilized, and trap door flap incisions were created in the pulmonary trunk. The coronary ostial buttons were anastomosed to the trap door flap, functionally extending the ostium and minimizing axial rotation. In cases of a single orifice, the orifice was left in place, the aorta and pulmonary trunks were sewn together,

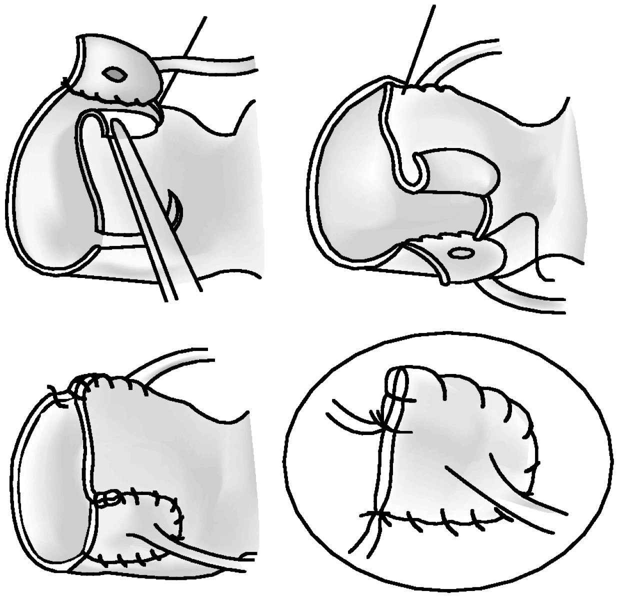

Figure 1.
Standardized translocation of the coronary arteries where the left anterior descending coronary artery and circumflex arise from sinus #1 and the right coronary artery arises from sinus #2. Medially hinged trapdoor flaps are created in the proximal pulmonary trunk, and the coronary arterial cuffs are then sewn into the defects created by the trapdoor flaps without axial rotation, starting at the edge away from the hinged flap (Stark, de Leval: Surgery for congenital heart defects. Second Edition. W.B. Saunders, 1994, 489–492). Reproduced with permission from W.B. Saunders.

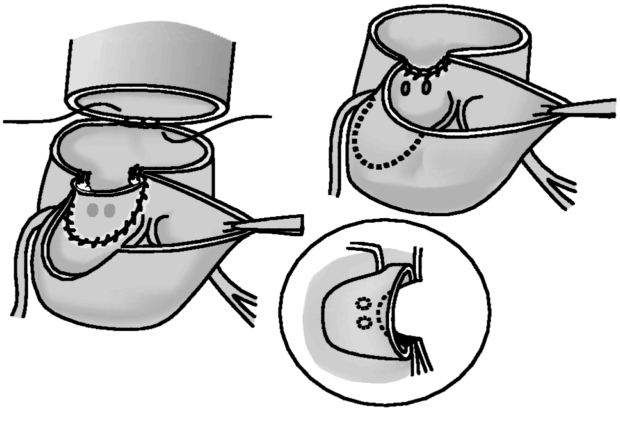

Figure 2.
Repair of a single orifice for the coronary arteries, demonstrating the formation of a hood committing the orifice to the neo-aorta (Stark, de Leval: Surgery for congenital heart defects. Second Edition. W.B. Saunders, 1994, 489–492). Reproduced with permission from W.B. Saunders.

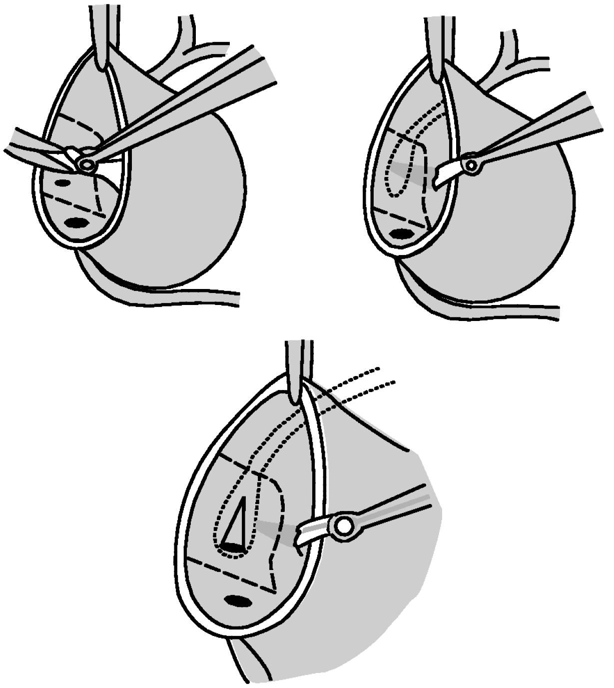

Figure 3.
Repair of an intramural coronary artery, demonstrating unroofing of the coronary arterial orifice (Stark, de Leval: Surgery for congenital heart defects. Second Edition. W.B. Saunders, 1994, 489–492). Reproduced with permission from W.B. Saunders.

and a hood was fashioned to commit the mouth of the artery to the neo-aorta (Fig. 2). During repair of an intramural coronary artery, the commisure was mobilized and the orifice of the coronary artery was unroofed to prevent stenosis (Fig. 3). A modified Lecompte maneuver[13] was used for all patients. Pulmonary arterial reconstruction was performed using two autologous pericardial patches.[14]

Postoperative course and follow-up

A postoperative electrocardiogram was performed on the morning following surgery, or sooner if there was evidence of changes in the ST segments. Follow-up echocardiographic evaluation was performed at the first visit to the outpatient clinic, or sooner if there was evidence of impaired left ventricular function. In all instances, we took care to assess left ventricular function, seek for evidence of myocardial ischemia such as regional abnormalities of wall motion, neo-aortic valvar regurgitation, and stenosis of the pulmonary valve or branches of the pulmonary trunk. We used low threshold Doppler criterions for diagnosis of stenosis at valvar and supravalvar levels or within the branches of the pulmonary trunk.

Definition of mortality

Early mortality was defined as death occurring prior to 30 days postoperatively, with those dying beyond this stage considered to represent late mortality.

Results

Of the 67 patients undergoing an arterial switch procedure during the period of study, 42 had an intact ventricular septum, 21 had a ventricular septal defect, and four had double outlet right ventricle with a subpulmonary ventricular septal defect with or without abnormalities of the aortic arch. The median age at surgery was 8 days for those with an intact septum, with a range from 3 to 120 days, 21 days for those with a ventricular septal defect, with a range from 6 to 450 days, and 35 days for those with double outlet right ventricle, the range being 10 to 120 days (Table 1).

Coronary arterial anatomy (Table 2)

In 52 patients, the left and right coronary arteries arose from sinus #1 and sinus #2, respectively, with the main stem of the left coronary artery dividing into the left anterior descending and the circumflex coronary arteries. In 48 of these 52 patients, the coronary arterial orifices were in the center of the sinus. Four patients had a markedly eccentric or para-commissural origin of either coronary artery, and four patients had a tangential origin of one coronary artery. One patient also had an accessory left anterior descending coronary artery, which arose from the right coronary artery. In two patients, there was a high take-off from the aorta, with both coronary arteries arising above the sinutubular junction. In one patient with eccentric origin of the right coronary artery, the take-off was directly beneath the aortic valvar commissure near the hingepoint of the valvar leaflet, in other words, a low take-off.

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Table 1. Comparison of the intra and postoperative results in our three groups of patients.

	Intact ventricular septum	Ventricular septal defect	Double outlet right ventricle with anterior and rightward aorta
No. of patients	42 (63%)	21 (31%)	4 (6%)
Median age	8 days (range 3–120)	21 days (range 6–450)	35 days (range 10–120)
CPB	189 min (145–301)	227 min (183–302)	293 min (158–375)
AXC	88 min (70–157)	121 min (85–149)	160 min (128–190)
CA	9 min (0–26)	20 min (0–68)	19 min (0–107)
Extubation	3 days (1–12)	2 days (1–20)	5 days (2–22)
Inotropes*	4 days (0–6)	2.5 days (1–20)	5 days (2–22)
LHS	8 days (5–77)	8 days (6–26)	12 days (9–87)
Arrhythmias	None	3 patients**	None
Re-op	None	1 patient***	2 patients****

Numbers expressed in median and range. *Inotropes include dobutamine, dopamine > renal dose and milrinone; **3 patients had arrhythmia; 2 with junctional ectopic tachycardia and 1 with intra-atrial re-entry tachycardia (IART); ***1 patient developed SVC syndrome requiring surgical exploration; ****2 patients developed pulmonary branch stenosis requiring surgical revision.
Abbreviations: AXC: aortic cross-clamp time; CA: circulatory arrest time; CPB: total bypass time; LHS: length of hospital stay; Re-op: re-operation

Table 2. Distribution of branching patterns, and presence of intramural or tangential origin of the coronary arteries.

Sinus 1	Sinus 2	N pts	Comments
L Cx	R	52	4 tangential/1 intramural
L	R Cx	8	
L Cx R		3	1 tangential
R LADCx		1	Double barrel single ostium – sinus 1
L R		1	No circumflex coronary artery identifiable
L CB	R Cx	1	Large conal branch off left coronary
	L R Cx	1	1 intramural left coronary artery

Sinus origin

Abbreviations: L: left coronary artery; R: right coronary artery; Cx: circumflex coronary artery; LAD: left anterior descending coronary artery; CB: conal branch; N: number; pts: patients

The orifices of the left and right coronary arteries arose from sinus #1 and #2 respectively, but the circumflex coronary artery arose from the right coronary artery, in 8 patients. One of these patients had a double-barreled origin of the single orifice from sinus #2, with early branching of the right coronary and circumflex coronary arteries.

In three patients, all three coronary arteries arose from sinus #1. One patient had two separate coronary arterial orifices arising from sinus #1, with the left anterior descending and circumflex coronaries arising from the main stem of the left coronary artery, which had a double-barrelled lumen. Two patients had a single orifice supplying a single coronary artery which divided into left main and right coronary arteries. One of these patients had a tangential origin of the right coronary artery.

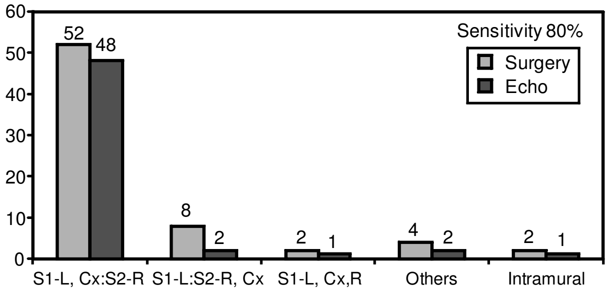

Figure 4.
Graph demonstrating the correlations between the intraoperative and echocardiographic findings concerning coronary arterial anatomy.

Four patients had additional patterns of branching. In one patient, both the right coronary artery and left anterior descending coronary artery arose from a single orifice in sinus #1, with the circumflex coronary artery arising from sinus #2. In another patient, both the left and right coronary arteries arose from sinus #1, and there was no circumflex coronary artery. In still another patient, the left interventricular coronary artery and a large conal branch arose from sinus #1, and the right and circumflex coronary arteries arose from sinus #2. In the last patient, the left interventricular, right, and circumflex coronary arteries all arose from sinus #2, with the left interventricular and right coronary arteries passing between the aorta and the pulmonary trunk.

One patient had cardiac catheterization and atrial septostomy performed without echocardiography at our institution, having had a prior diagnostic echocardiogram in the referring center. There was correlation between the echocardiographic and intraoperative findings in 53 of 66 patients (81%) (Fig. 4), and angiographic correlation in 31 of 36 patients (86%)

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Table 3. Accuracy of echocardiography and angiography in diagnosis of different types of coronary arterial anatomy.

	S1-L, Cx S2-R	S1-L S2-R, Cx	S1-L, Cx, R	Others	Intramural
No. of patients, n = 67	52	8	3	4	2
Echo S, n = 66 (85%)	48/52 (92%)	2/8 (25%)	1/2 (50%)	2/4 (50%)	1/2 (50%)
Angio S, n = 36 (86%)	28/29 (97%)	2/4 (50%)	1/1 (100%)	0/2 (0%)	0/2 (0%)

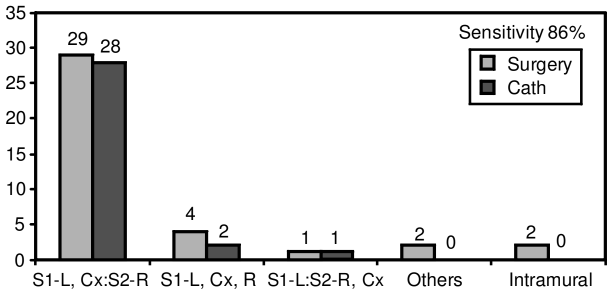

Figure 5.
Graph demonstrating the correlations of intraoperative and angiographic findings concerning coronary arterial anatomy.

(Table 3, Fig. 5). Excluding catheterizations which had been undertaken at outside institutions, the accuracy increased to 90%, 19 of 21 patients. The coronary arterial anatomy was assessed from ventricular angiography or ascending aortograms rather than selective coronary angiography in the majority of patients, including the two patients with unsuspected intramural coronary arteries.

Outcome

The arterial switch operation was successfully performed in all 67 patients, with no intra-operative or 30 day mortality. No patient was denied an arterial switch procedure based upon their pre- or intra-operative coronary arterial anatomy. One patient with a single coronary arterial orifice required a right internal mammary arterial bypass to the right coronary artery because of arterial obstruction noted during an initial trial to wean the patient from cardiopulmonary bypass. Two patients required revision of the new pulmonary trunk within the first 2 weeks, both having double outlet right ventricle with the aorta anterior and rightward.

Intra-operatively, the duration of cardiopulmonary bypass, aortic cross-clamping, and circulatory arrest time were longest in patients with double outlet right ventricle. Post-operatively, the time to extubation, duration of inotropic support, and total length of stay were also longer for those with double outlet right ventricle (Table 1). Significant arrhythmias developed only in 3 patients with normal coronary arterial arrangement, but transient changes in the ST segments were noted in 7 patients, all with different patterns. Interestingly, neither of the two patients with an intramural course had changes in the ST segments or arrhythmias. The median length of hospital stay was 8 days for those with an intact septum, 8 days for those with a ventricular septal defect, and 12 days for those with double outlet right ventricle. Those with both coronary arteries arising from a single orifice, or with atypical coronary arterial anatomy, had a longer duration on inotropic support, longer time to extubation, and a slightly longer stay in the hospital (Table 4).

Follow-up and late mortality

There were two late deaths (2.9%). One 2 kg premature infant with bronchopulmonary dysplasia developed septicemia and died on the 94th day following surgery. A second patient developed superior caval venous syndrome with chronic chylothorax, and died on the 77th postoperative day. Both patients had good ventricular function documented by echocardiography, and no history of arrhythmia.

All patients underwent echocardiographic assessment on follow-up. The mean duration of follow-up was 5 months, with a range from 0.1 to 32 months. All patients had good left ventricular function, and no evidence of myocardial ischemia. Pulmonary valvar stenosis was noted in 1 patient (1.9%), supravalvar stenosis in 3 patients (5.7%), and stenosis of the pulmonary arterial branches in 7 patients (13.5%). Multiple levels of stenosis were seen in one patient (1.9%). Surgical revision of the pulmonary arteries was needed in two patients in the first two weeks after the arterial switch. No patients required interventional catheterization, nor did any develop significant neo-aortic regurgitation. Echocardiographic evidence of mild aortic regurgitation was present in 10 of 67 patients, in whom 5 had a major discrepancy in size between the aortic and pulmonary valvar orifices.

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Table 4. Influence of branching patterns on postoperative course.

Coronary arterial pattern	S1-L, Cx S2-R	S1-L S2-R, Cx	S1-L, Cx, R	Others	Intramural
No. of patients	52	8	3	4	2
ST segment changes	2	2	1	1	None
Arrhythmia	3	None	None	None	None
Extubation	2 days	2.5 days	3 days	3 days	3.5 days
Inotropes	3 days	4 days	5 days	5 days	2 days
Discharge	8 days	8 days	10 days	10 days	9 days

The median duration is given, S1, S2 – sinus 1 and sinus 2

Discussion

Since the first successful arterial switch procedure in 1975 by Jatene,[15] and its further modification by Lecompte,[13] anatomic correction has been widely accepted as the procedure of choice for patients with concordant atrioventricular and discordant ventriculo-arterial connections. The critical importance of establishing unimpeded flow of blood from the aorta to the coronary arteries following the switch became very clear following the earliest unsuccessful attempts in the 1950s.[16,17] The multiple variations known to occur in the origins and distribution of the coronary arteries in hearts with concordant atrioventricular and discordant ventriculo-arterial connections have been well documented.[4–9] Historically, some patients with complex coronary arterial patterns were denied an arterial switch operation, with a single right coronary artery, or the left anterior descending arising from the right coronary artery and the circumflex arising from a separate aortic sinus, being identified as patterns associated with high risk.[4] Recent studies have reported encouraging outcomes following the arterial switch procedure, with a 94% survival and 78% probability of freedom from re-operation at 10 years following surgery even in patients with challenging coronary arterial anatomy.[1,18] To our knowledge, there have been no recent reports to support the need to perform coronary angiography prior to an arterial switch procedure.

In our study, no patient was denied the arterial switch based upon their pre- or intra-operative coronary arterial anatomy. There was no intra-operative or 30-day post-operative mortality (70% confidence limits; 0–2.9%), with only two patients dying later, although the size of our cohort was limited. This is in accordance with several recent reports based on medium or large populations, which have showed in recent years that coronary arterial pattern has no impact on outcome.[19,20] Earlier studies reported the presence of an intramural course, or a single coronary artery, to be the most significant risk factors for mortality.[21] Mayer et al.[22] considered 12 of their 20 hospital deaths to be attributable to problems with inadequate coronary arterial flow, with the highest mortality found in those with a single right coronary artery, or the left anterior descending arising from the right coronary artery and the circumflex arising from a separate orifice. The institutional cardiac registry in Toronto similarly reported a 41% death rate with the arterial switch from 1985 to 2000 in those patients having a single coronary artery.[6] They further reported the implanting of a well-mobilized coronary arterial button into a previously anastomosed neoaorta as the key prophylactic step in reducing their mortality, with no deaths encountered in the three years following its introduction. One large recent study also found single or "inverted" coronary arterial anatomy not to be predictive of mortality. Rather, it was associated with an increased incidence of delayed sternal closure and prolonged mechanical support.[20]

A morphologic study which studied necropsy specimens in 88 patients, in addition to 189 children who had undergone the arterial switch, found the origin of the coronary arteries at or above the sinutubular junction in 20% of cases, the radial origin to be paracommissural in 3%, and the angle of origin not orthogonal in 7%.[8] This abnormal orientation of the coronary artery, including the presence of a paracommissural or high-take-off, may pose further challenges to surgical transfer. Our population had a 4% incidence of intramural coronary artery, a 6% incidence of tangential origin of one coronary artery, 4 cases with eccentric or para-commissural origin of a coronary artery, but only 2 cases with the coronary arteries arising above the sinutubular junction. This contrasts markedly with the incidence of 20% reported by Li et al.[8] None of these patterns had an adverse effect on our surgical outcome. Nevertheless, our experience highlights potential factors other than the branching patterns themselves which may complicate the arterial switch procedure.

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The accuracy of echocardiography and angiography in delineating the coronary anatomy prior to arterial switch were comparable, at 81% and 86%, respectively. Previously reported sensitivities of echocardiography in detecting coronary arterial anatomy have ranged between 80% and 95%.[23,24] The higher sensitivity may be attributed to selection bias, since only studies with adequate images were included. In our series, we included all studies except one which was unavailable. No patient in our cohort developed left ventricular dysfunction when assessed at medium-term follow-up by echocardiographic evaluation. Impaired myocardial blood flow, and coronary flow reserve, nonetheless, correlate poorly with left ventricular function at long term follow-up.[25] Both late deaths in our series had well preserved myocardial function documented by echocardiography, and no history of arrhythmia. Previous studies have, however, demonstrated normal function by echocardiography and scintigraphy in patients having orificial stenosis, or even coronary arterial occlusion, revealed by angiography.[4]

Limitations

Firstly, this is a retrospective study of a relatively small number of patients with a short duration of follow-up. Data regarding the coronary arterial anatomy was retrospectively obtained from the surgical operative reports. Secondly, it is difficult to draw meaningful conclusions correlating coronary arterial anatomy and adverse outcome when no patient died at operation or in the early postoperative period. Finally, the number of patients with unusual coronary arterial anatomy was limited in our population, and it is this population with unusual patterns which is more likely to encounter adverse outcomes.

Conclusion

Preoperative assessment of coronary arterial anatomy was comparable using transthoracic echocardiography and coronary angiography. The coronary arterial anatomy did not influence morbidity or mortality in our cohort of patients, although the prevalence of unusual coronary arterial patterns was limited among our patients. The arterial switch procedure can be performed with an excellent outcome, irrespective of the coronary arterial anatomy, obviating the need for preoperative coronary angiography.

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