Influence of coronary anatomy and reimplantation on the long-term outcome of the arterial switch


Children’s Heart Centre, University Medical Centre, Utrecht, The Netherlands

Received 6 September 1999; received in revised form 7 April 2000; accepted 16 May 2000

Abstract

Background: Abnormal coronary artery anatomy is reported to have a significant influence on the outcome of the arterial switch operation. This study examines the impact of coronary anatomy and the occurrence of late coronary obstruction on left ventricular (LV) function and long-term outcome.

Methods: Coronary artery anatomy, of 170 patients after arterial switch operation (1977–1999), was determined based on operative reports and pre-operative aortograms. Current status was evaluated using ECGs, echocardiograms, scintigraphy, and post-operative coronary angiograms.

Results: In 133/170 patients, coronary artery anatomy consisted of an anterior descending (LAD) and circumflex artery (Cx) from the left sinus and the right coronary artery (RCA) from the right or posterior sinus. The left coronary had an intramural initial course in two of these patients. Fifteen patients had the LAD from the left and Cx and RCA from the right sinus; eight had LAD and RCA from one sinus and Cx from the other; and three had three separate ostia. Four patients had complex patterns and four patients had a supra commissural coronary. To date, follow-up angiography was performed in 59 patients. Surgical coronary sequellae were found in five patients. Two patients had an occluded left ostium. Initially, they were asymptomatic but showed polymorphic ventricular extrasystoles on ECG and moderate LV dysfunction with large irreversible perfusion defects on scintigraphy. Both patients developed ventricular fibrillation at the age of 14 years. One patient did not survive. The other patient required implantation of a defibrillator. One patient has an occluded RCA, one patient has stenosis of the right ostium and one patient has multiple tortuous collaterals without obstruction of a major branch. Two patients had an occluded left ostium. Initially, they were asymptomatic but showed polymorphic ventricular extrasystoles on ECG and moderate LV dysfunction with large irreversible perfusion defects on scintigraphy. Both patients developed ventricular fibrillation at the age of 14 years. One patient did not survive. The other patient required implantation of a defibrillator. One patient has an occluded RCA, one patient has stenosis of the right ostium and one patient has multiple tortuous collaterals without obstruction of a major branch. In the latter three patients, coronary sequellae were not suspected on ECG, echo, or scintigraphy and were only found on follow-up angiography. Retrograde collateral flow was noted in all three occluded coronaries. LV dysfunction, with normal coronaries, was noted in three patients. All, of these patients, had peri-operative ischaemia suggesting failure of myocardial protection. Two are now asymptomatic with mild LV dysfunction. One patient continues to have severe myocardial dysfunction and secondary aortic insufficiency. A Ross-like procedure was performed placing the original aortic valve in the neo-aortic root. Coronary artery anatomy did not influence early survival or late coronary sequellae.

Conclusion: Abnormal coronary anatomy was not a determinant of outcome in our study. Surgical coronary obstruction is independent of original anatomy. It can be almost silent and is potentially fatal. Follow-up angiography must be considered in all patients after the arterial switch operation. © 2000 Elsevier Science B.V. All rights reserved.

Keywords: Congenital heart surgery; Arterial switch operation; Coronary anatomy; Long-term outcome

1. Introduction

Re-implantation of the coronary arteries into the neo-aortic root has always been considered the key of the arterial switch operation [1]. Initially, many patients perished due to left ventricular failure frequently caused by impaired myocardial perfusion. With increased surgical experience, mortality decreased significantly but abnormal coronary anatomy can still complicate the task of coronary transfer and is considered a major determinant of operative mortality [2,3]. A recent angiographic follow-up study [4,5] showed an alarming incidence of coronary obstruction, in many cases in asymptomatic patients, but no link was made to the initial coronary anatomy. This study examines the impact of coronary anatomy on outcome after the arterial switch and is aimed both at its impact on early mortality, as well as, at the long-term morbidity due to early or late coronary obstruction. Left ventricular function is included, in the study, as an indicator of coronary perfusion.
2. Methods

From 1977 through 1999, the arterial switch operation has been the treatment of choice for transposition of the great arteries. Included, in the study, were all patients who underwent this operation and who still reside in The Netherlands. Operation reports were studied for descriptions of the coronary anatomy. Preoperative angiograms, usually aorta or right ventricular injections, performed during Rashkind procedure, were studied when available and checked against the surgical findings. Coronary anatomy was classified according to the scoring systems published from the Leiden group [6], as well as, those published by Planche [7] and Yacoub [8]. The medical records were studied for postoperative course. All patients were seen yearly in the outpatient department for 12-lead ECG and detailed echocardiography; 24-h ECG was repeated biannually. Initially, cardiac catheterization was performed when indicated. More recently, we decided to perform elective coronary angiography in all patients over 10 years of age.

2.1. Statistical analysis

Analysis, using all subgroups of coronary anatomy, was impossible because of the small number of patients in each group except for the most common anatomy. Analysis was repeated using two groups (common vs. other). Chi-square analysis was used for ordinal data and Student's t-test for continuous variables. Multivariate analysis was performed using logistic regression.

3. Results

One hundred and seventy patients were included in the study. Mean age at follow-up was 8.1 years (range 0.3–22 years). One hundred and three patients had simple transposition, 46 had transposition with VSD and 20 had double outlet right ventricle with transposition and sub-pulmonary position, 4 had transposition with VSD and 20 had double outlet right ventricle with transposition and coarctation. He had a single coronary artery and in the other two ran between the great arteries. One additional patient had double outlet right ventricle with transposition and coarctation. He had a single coronary ostium from the right cusp, with the left coronary coursing between the great arteries before dividing in LAD and circumflex branches. It was deemed impossible to transfer the coronary. The coarctation was repaired and the pulmonary artery was banded with a view to later Rastelli operation. This is the first patient in our experience deemed un-switchable due to coronary anatomy. Since no arterial switch was performed, this patient is not included in the study.

LV failure was the most common cause of early mortality. Coronary anatomies of the patients who died and causes of death are listed in Table 2. It is not always clear, especially in the older patients, whether LV failure was due to imperfect coronary transfer, poor myocardial protection, or loss of LV mass prior to operation. Unexpected LV outflow obstruction, caused the LV to fail in two patients. One patient with moderate LV dysfunction thought due to the failure of myocardial protection arrested in the ICU with a tamponade. He was resuscitated but subsequently treatment was discontinued because of adverse neurological prognosis. Other causes of death included septicemia in one patient, prematurity and the consequences of severe respiratory distress syndrome in one, and pulmonary bleeding or embolus in three patients. There was no significant statistical relation between coronary anatomy and early operative mortality ($\chi^2 = 0.256$, $P = 0.61$). This was true regardless

<table>
<thead>
<tr>
<th>$N = 170$</th>
<th>Coronary anatomy</th>
</tr>
</thead>
<tbody>
<tr>
<td>131</td>
<td>1LAD,Cx,2R</td>
</tr>
<tr>
<td>15</td>
<td>1LAD,2R,Cx</td>
</tr>
<tr>
<td>4</td>
<td>1LAD,R,2Cx</td>
</tr>
<tr>
<td>3</td>
<td>1Cx,2R,LAD</td>
</tr>
<tr>
<td>3</td>
<td>3 separate ostia</td>
</tr>
<tr>
<td>3</td>
<td>Single right ostium</td>
</tr>
<tr>
<td>3</td>
<td>Supracommissural ostium + small extra</td>
</tr>
<tr>
<td>1</td>
<td>Supracommissural ostium from the right cusp, with the left coronary coursing between the great arteries before dividing in LAD and circumflex branches. It was deemed impossible to transfer the coronary. The coarctation was repaired and the pulmonary artery was banded with a view to later Rastelli operation. This is the first patient in our experience deemed un-switchable due to coronary anatomy. Since no arterial switch was performed, this patient is not included in the study.</td>
</tr>
<tr>
<td>5</td>
<td>Complex anatomy</td>
</tr>
</tbody>
</table>

$^a$ Abbreviations: LAD, left anterior descending artery, Cx, circumflex artery, RCA, right coronary artery.
$^b$ The coronary that was above the commisure.

The coronary artery anatomy is described in Table 1. Single right ostium was encountered in three patients, in one of whom the left coronary looped around the pulmonary artery and in the other two ran between the great arteries. One additional patient had double outlet right ventricle with transposition and coarctation. He had a single coronary ostium from the right cusp, with the left coronary coursing between the great arteries before dividing in LAD and circumflex branches. It was deemed impossible to transfer the coronary. The coarctation was repaired and the pulmonary artery was banded with a view to later Rastelli operation. This is the first patient in our experience deemed un-switchable due to coronary anatomy. Since no arterial switch was performed, this patient is not included in the study.

LV failure was the most common cause of early mortality. Coronary anatomies of the patients who died and causes of death are listed in Table 2. It is not always clear, especially in the older patients, whether LV failure was due to imperfect coronary transfer, poor myocardial protection, or loss of LV mass prior to operation. Unexpected LV outflow obstruction, caused the LV to fail in two patients. One patient with moderate LV dysfunction thought due to the failure of myocardial protection arrested in the ICU with a tamponade. He was resuscitated but subsequently treatment was discontinued because of adverse neurological prognosis. Other causes of death included septicemia in one patient, prematurity and the consequences of severe respiratory distress syndrome in one, and pulmonary bleeding or embolus in three patients. There was no significant statistical relation between coronary anatomy and early operative mortality ($\chi^2 = 0.256$, $P = 0.61$). This was true regardless
of the classification system used to describe coronary anatomy. Two patients died late. One patient died of pulmonary vascular occlusive disease. The other patient is described below.

Follow-up cardiac catheterization was performed in 59 patients. Mean age at catheterization was 10.1 years (0.1–18.4 years). Only right-sided catheterization was performed or the coronaries were not adequately visible on the aortogram in four patients. The coronaries were visualized in 55 patients, in 17 patients on biplane aortogram, in 34 patients on selective injections in both coronaries, and in four patients on aortogram with selective injection in one coronary. The coronary anatomy of the 55 patients was 1LAD,Cx:2R in 43 patients, 1LAD:2R,Cx in two, 1LAD,R:2Cx in four and 1Cx:2R,LAD in one. Two patients had three separate ostia, two patients had complex anatomy and one patient had supra-commissural coronary ostium. Coronary sequelae were found in five patients.

Two patients had an occluded left ostium with immediate post-operative ischaemia/infarction. Both had the most usual coronary anatomy. On follow-up, they had polymorphic ventricular extrasystoles and impaired LV function. Technetium scintigraphy demonstrated large irreversible defects in the left ventricle, and 12-lead and 24-h ECG showed polymorphic ventricular extra systoles. The left coronary ostium was completely occluded with the right coronary arising from the right cusp. There was collateral perfusion of the left coronary artery system (Fig. 1).

Both patients were asymptomatic with good exercise tolerance. However, they both developed ventricular fibrillation at the age of 14 years. One was successfully resuscitated and a defibrillator was implanted. This patient continues to do well. The defibrillator terminated one episode of ventricular fibrillation during strenuous exercise. At the time, we discussed an operation to re-perfuse the coronary, but this was declined, as the remaining left coronary was tiny. The
events, of this patient, prompted further investigations in the second patient with occluded left coronary. An operation to re-perfuse the coronary was discussed but was declined, at the time, because there was no area of reversible ischaemia around the infarct. Implantation of a defibrillator was discussed, however, the parents refused based on the fact that the patient had no symptoms and on their religious background. The patient arrested on her way to the library. She was resuscitated but brain death was diagnosed upon her arrival in the hospital.

One patient had an occluded right coronary artery. She had complex anatomy with circumflex branches from both ostia. Retrograde collateral flow was noted to perfuse the area of flow of the right coronary artery and perfusion scintigraphy was normal. Monomorphic ventricular extra systoles were seen in some but not all of the 24-h ECG studies. This patient required three re-operations to relieve pulmonary stenosis. Right-sided cardiac catheterization was performed a number of times but no arterial catheterization had been performed. The previous experience led to the decision to repeat catheterization with selective coronary angiography. The VES have disappeared after the pulmonary valve was replaced with a homograft. This patient is now 17 years old and asymptomatic.

Asymptomatic stenosis of the right coronary ostium was documented in one patient (Fig. 2). Another patient has multiple tortuous collaterals without evident coronary obstructions (Fig. 3). Both patients had the most frequent anatomy. They have normal ECG, normal left ventricular function, and normal scintigrams.

No relationship was found between initial coronary anatomy and the risk of abnormal coronary findings on follow-up angiography. Logistic regression analysis demonstrated an increased risk of abnormal findings with patients operated earlier in our experience. Again no influence of coronary anatomy could be demonstrated.

Five patients had impaired LV function on echocardiography, confirmed by angiography. Coronary anatomy was 1Cx:2R,LAD in one patient and 1LAD,Cx:2R in four. Two, of these patients, were the patients with occluded left ostium. No coronary abnormalities were found in the remaining three patients. The left ventricular dysfunction was preceded by peri-operative ischaemia suggesting failure

Fig. 1. Angiograms of a patient with obstructed left coronary ostium. Panel A: aortogram with filling of the right coronary. The left coronary artery is not visible. Panel B: late frame of selective right coronary injection. Note filling of the (tiny) left coronary artery (arrow) via collaterals (arrowheads).

Fig. 2. Injection in the right coronary cusp showing stenosis of the right coronary ostium.
of myocardial protection. Two patients had normal ECG and normal scintigraphy. Both are asymptomatic with moderate LV dysfunction. The third patient developed myocardial dysfunction and secondary aortic insufficiency. A Ross-like procedure was performed placing the original aortic valve in the neo-aortic root [9]. After a brief initial improvement, recurrent aortic insufficiency was caused by dilation of the new aortic root. The aortic valve was replaced using a St Jude (18 mm) prosthesis. The left ventricle remains severely dilated and exercise tolerance is limited.

4. Discussion

When our institution opened in 1977, our preference went to the arterial switch over the atrial redirection. This was based upon the assumption that the long-term outcome would be more favourable than after atrial redirection surgery. The proof of this assumption is yet undetermined. Favourable long-term follow-up studies of patient-populations after the atrial redirection surgery have been published [10,11]. Previous reports, on the arterial switch, have indicated that overall mortality figures have dropped to very acceptable standards [12]. Morbidity has been reported pre-dominantly in the area of pulmonary artery stenosis [13,14]. A few studies have demonstrated late coronary occlusion [5,15]. Previously, reported arrhythmias were limited to a small contingent of rather innocent findings [16]. This previous report has to be amended since two of the patients in the previous study with polymorphic ventricular extra systoles have now developed ventricular fibrillation. Prompted, by these two patients, all previous aortograms were re-evaluated. It was possible to judge the coronary ostia in 16/21 aortograms but smaller more distal abnormalities would be missed, such as, the multiple collaterals found in one patient. Selective coronary angiography, as advocated previously [4], is now performed in all patients. Coronary angiography is not a risk-free procedure in children, since it can provoke ischaemia leading to arrhythmia or low output.

The overall early and late mortality was 16%. This includes the initial learning curve when the arterial switch was still very much uncharted territory. The current (1996–2000) operative mortality is 2%. Although no relationship was found between anatomy and risk of peri-operative mortality, there are some forms of coronary anatomy that are generally recognized as more difficult, such as, intramural coronary artery. If recognized timely, this anatomy could be dealt with successfully. Patients with single right coronary ostium, with interarterial course of the left coronary can pose insurmountable problems. In our series, one patient was switched successfully, one patient died and one patient was not switched.

The study, by Bonhoeffer [4,5], includes a number of patients in whom a one-patch coronary transfer technique was used. This technique was never used in our institution. However, a similar number of late coronary sequelae was encountered (5/55). In two of these five patients the findings were completely unexpected, very similar to the 3% reported by Tanel [15].

Patients with coronary stenosis or occlusion can be asymptomatic and have normal ECG and Echo findings as demonstrated in two patients. The patient, with the occluded right coronary, currently has a normal 24-h ECG while the obstruction is still present. Scintigraphy as a method of screening for coronary obstructions is not very helpful, in our experience, as three of five patients with coronary sequelae had normal findings. Scintigraphy detects ischaemia and excellent collateral coronary circulation prevents ischaemia even during chemical stress. The same is
expected to be true for stress-echocardiography and PET scanning, though we have no personal experience with these methods in transposition patients.

Coronary angiography is, therefore, the only tool to detect these silent obstructions. A therapeutic strategy dealing with obstructions still needs to be developed. An area with reversible ischaemia on stress-scintigraphy might provide an arrhythmogenic focus and an attempt should be made to re-perfuse the coronary by opening the obstructed ostium or performing some kind of bypass surgery. At the time, there was doubt if this would be beneficial in the surviving patient with left coronary obstruction. Currently the discussion regarding this patient is reopened as we have received various comments strongly advising that an attempt should be made.

In absence of reversibility, the benefits of re-operation may not outweigh the risks. If scintigraphy is normal the decision is difficult. If re-perfusion is easily and permanently achieved, such as in the patient with stenosis of the right ostium, we feel that it should be done. It is difficult to judge the risks and benefits in the asymptomatic patient with obstructed right coronary since she had already three repeat surgeries for pulmonary stenosis. The coronary is occluded over a distance of several centimeters and the only option seems to be some form of bypass. This, in essence, provides extra collateral circulation to the coronary with may impede the development of the collaterals that are currently providing adequate circulation even during exercise and chemical stress. The debate will continue until the collective experience increases and provides a more definite answer. Only very long-term studies can clarify the influence of ageing in these patients.

Finally, there is a small group of patients with postoperative ischaemia and normal coronary perfusion. This suggests failure of myocardial protection, which led to dilatation and secondary aortic insufficiency in one of these patients. The patient, 2 years old at the time, needed aortic valve replacement and a Ross-like procedure seemed to be the best choice, incidentally placing the original aortic valve in the neo-aortic root. The result was dilatation and subsequent recurrence of regurgitation.

In conclusion, abnormal coronary anatomy was not a statistically significant determinant of mortality in our study. However the study is limited in numbers and follow-up duration. Incidental patients with coronary anatomy that are very difficult or even impossible to transfer, but mortality can be avoided in experienced hands. Surgical coronary obstruction is independent of original anatomy. It can be almost silent and is potentially fatal. Follow-up coronary angiography is strongly advised although it carries a certain risk.

References


Appendix A. Conference discussion

Dr F. Lacour-Gayet (Le Plessis Robinson, France): I have a couple of questions to ask you. I enjoyed your paper, but there is in your group of transposition a striking difference with most of the series is that, if I had well understood, you don’t have patients with so-called intramural course of coronary artery. As you know, it is probable one of the weak points of your experience. Now, you have been fortunate, because all of us who have been facing this problem have had difficulties, and the mortality in this
group initially in our hands was up to 40%. Now it is reduced probably to 5%. So I don’t say that you can conclude what you conclude in regards to your group of patients. Therefore I would like to ask you if it is a selective group of patients that you have operated, and if you decided not to perform arterial switch in patients with an intramural coronary?

Now, my second question is that we have already identified those patients that you named type C in your classification, namely, patients who have the right coronary coming from the anterior descending with side-by-side vessels, the anterior button after relocation can be compressed by the pulmonary artery. This has been corrected by lining up on the right the pulmonary trunk, but this has been, in our experience, a difficult problem. So my question is, have you selected your patients and do you really believe that type C is not statistically significant in your experience?

**Dr Bennink:** Well, I am not so long in paediatric heart surgery as you are, but my colleague, Professor Hitchcock, is, and it is not a selection, and we did not meet any intramural coronary anatomies until recently when we moved to the new hospital, and in the last eight switches we did, I had two switches with intramural anatomy, and both did very well, luckily, but they are not included in this study. So it is not a selection, and it is pure coincidence.

**Dr A. Urban (St. Augustin, Germany):** My question to you is a little similar to Francois Lacour-Gayet’s question. Normally the distribution of coronary arteries in transposition is such that 63% are Type I, but you had about 82 or so percent of Type I. The other, more difficult types are less common but still they are very much less common in your series. So, again, the question, why is that?

One of your conclusions is that coronary anatomy has no impact on early and late outcome. You did not tell us about early outcome. We do not know your early mortality, and I think only if we know that we can follow your conclusion. And from our personal series of 364 arterial switch operations we are aware that if you have to relocate intramural coronary arteries, the mortality is higher. Intramural course of coronary arteries e.g. has a significant impact on early and late outcome. So I do not agree with your conclusion.

**Dr Bennink:** Well, I only can say that recently we did two intramurals and the early outcome was good, so they were both discharged after 10 or 12 days, if I remember correctly. What you do see is that there was no selection in this series, and I don’t know why we have this distribution of coronary anatomy. And if I read the literature, I also thought that there is definitely an impact on early and late outcome depending on coronary pattern, but if you look at the numbers, they are relatively small in comparison with a big series. Of course if you look at it, I think it was in 1990, Wernovsky (Circulation) wrote a big series, and there, the coronary pattern did not have an impact on the outcome mortality but mainly on the morbidity.

Our early mortality, I quickly mentioned that out of the last 50 switches, we lost one, and before that 25 patients were lost in the whole series, but that includes the early days since 1977 with the two-stage repair. So our early mortality now is 2%.

**Dr P. Pohlner (Brisbane, Australia):** I was very interested in what you are able to do with the coronaries once you have defined that they are of no value. I think our experience has been, and most of the patients in fact have come from other centres, we don’t see much of this problem in our institution, but we do have problems revascularizing these patients long term, and I just wonder what your experience and perhaps others has been about that. We do find, again, a range of abnormalities of single coronary arteries, intramural coronaries in addition to the ones you have already described. They are very difficult.

We have to use a variety of strategies to establish a good coronary position with switching. Our results are really very good and equivalent to your last stated results. But I do have some uncertainty about what we can do about the coronaries which do not remain patent.

**Dr Bennink:** I think it is a very good question. We are dealing with the same problem. I hope someone can give the answer, but we are not sure what to do especially with asymptomatic patients with no reversible defects in certain areas. There is no reason to do a bypass, for example.

We discussed this 14-year-old patient, and we thought that because he had a lot of ventricular extra systoles that he was best off by using an AICD. Of course we also were thinking about doing a bypass, but since the experience in that area is very limited, and there were irreversible defects, we decided to go the easier way of implanting an AICD. One of our reasons for presenting the problem of occluded coronaries is to discuss what to do with all those patients.