Late reoperations after neonatal arterial switch operation for transposition of the great arteries

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Received 26 September 2007; received in revised form 3 April 2008; accepted 7 April 2008; Available online 8 May 2008

Summary

The arterial switch operation has become the treatment of choice for neonates with transposition of the great arteries. Currently, the early mortality rate is low as well as the need for early reoperation because of surgical failures; in our experience with 803 neonates, these risks were 3.8% and 1.5%, respectively. The late outcome in terms of survival and functional status is excellent. However, surgical repair is far from anatomical and potential late defects were identified as soon as this procedure was introduced: obstruction of the neo-pulmonary outflow tract, development of obstructions of the reimplanted coronary arteries, dysfunction of the neo-aortic valve, and progressive left ventricular dysfunction. Actually, late reoperations are required in 5—10% of patients (4.5% in our experience with a mean follow-up of 5.8 years). The more frequent indications for reoperation are coronary lesions and right ventricular outflow tract obstructions. Coronary obstructions are, in most cases, detected in patients without any clinical or echocardiographic evidence of myocardial ischaemia. Coronary lesions are progressive and repeated coronary evaluation at regular intervals is necessary. Reoperation is indicated when myocardial ischaemia, at rest or under stress, is demonstrated at myocardial imaging. Satisfactory results can be achieved by surgical coronary patch angioplasty; in selected cases, mammary bypass may be necessary. Right ventricular outflow tract obstruction is related either to inadequate growth of the pulmonary anastomotic site, or to inadequate growth of the whole new right ventricular outflow tract in patients with associated aortic arch obstruction. Reoperation is indicated when significant obstruction (gradient >50 mmHg) is detected at routine echo-Doppler evaluation. Although neo-aortic root dilation and minimal aortic valve insufficiency are common, reoperation for severe neo-aortic valve dysfunction is, to date, very rarely necessary. Whether this will remain the case in the decades to come requires further evaluation. Left ventricular function is maintained in the vast majority of patients. Reoperation may be indicated in some patients for other reasons: mitral valve malformation, tracheo-bronchial compression or pulmonary hypertension.

Keywords: Transposition of the great arteries; Arterial switch operation; Long-term results; Reoperation

1. Introduction

The arterial switch operation (ASO) has become the treatment of choice for neonates with transposition of the great arteries (TGA). Current operative mortality is low and late results are satisfactory [1—6]. However, although called anatomical repair, the result obtained after ASO is far from being truly anatomical. There are many potential late problems. These problems were identified by Sir Magdi Yacoub nearly 30 years ago, at the time of the emergence of this new procedure [7]. As stated, this includes: (1) need to reconstruct a neo-pulmonary trunk, (2) ability of the coronary anastomoses to grow, (3) ability of the neo-aortic valve to function without incompetence, (4) ability of the left ventricle to maintain long-term normal function.

2. Materials and methods

2.1. Arterial switch population

Between January 1987 and June 2007, 803 neonates have undergone arterial switch operation at the Sick Children Hospital, Paris. Five hundred and sixty-seven patients had simple TGA (with associated coarctation in 7) and 236 had TGA with ventricular septal defect (VSD) (with associated aortic arch obstruction in 42). There were 31 (3.8%) in-hospital deaths and 11 (1.4%) late deaths after hospital discharge (all occurring within the first year after surgery).
2.2. Follow-up

Late survivors had an annual examination by the referring cardiologist, which included clinical assessment, ECG and echo-Doppler study; myocardial scintigraphy, stress test, cardiac catheterisation or CT scan were done if deemed necessary by the referring cardiologist. Five of the survivors were lost to follow-up. The remaining 756 late survivors were followed-up for a mean of 5.8 years (range 3 months to 19 years). All reoperations, performed after hospital discharge were recorded. Late reoperations, carried out more than 1 year after ASO, were more specifically studied. The present study has been approved by the local ethics committee.

3. Results

Forty-five patients (5.8% of the study population, 70% confidence limits = 4.9—6.6%) underwent 46 reoperations after hospital discharge. Actuarial survival free from reoperation was 91% at 5 years, 88% at 10 years and 86% at 15 years.

3.1. Early reoperations

Eleven reoperations were performed within the first year after ASO and were considered as early reoperations. The indications are listed in Table 1. One patient died following reoperation for coronary obstruction. Most early reoperations (8/11) were needed for surgical failures (residual or recurrent aortic arch obstruction, inadequate coronary transfer leading to coronary obstruction).

3.2. Late reoperations

Late reoperations were defined as reinterventions performed more than 1 year after ASO.

Thirty-four patients underwent 35 late reoperations. There were 28 boys and 6 girls. Initial diagnosis was simple TGA in 22 (2 of them had associated coarctation) and TGA with VSD in 12 (4 with associated aortic arch obstruction). Reoperations were performed after a mean interval of 6.5 years (range 16 months to 15.2 years). At the time of reoperation, the mean age of the patients was 6.5 years (range 1.3—15.4 years). Late reoperations were more frequent in TGA with aortic arch obstruction (with or without VSD) (5/39, 12.8%) but the incidence was similar in simple TGA (21/532, 3.9%) or TGA/VSD without aortic arch obstruction (8/186, 4.3%). Most late reoperations were performed within the first 9 years following ASO (29/35, 83%); only six reoperations (17%) were necessary after 10 years up to 19 years (Fig. 1). The causes of late reoperations are listed in Table 2. As shown in Fig. 1, the causes were equally distributed throughout the follow-up period.

3.3. Reoperations for coronary lesions

Seventeen patients underwent reoperations for coronary lesions. Coronary lesions were detected after a mean interval of 33 months. Six patients had ECG and/or echocardiographic findings suggestive of myocardial ischaemia. In the last 11 patients, coronary lesions were detected at the time of routine coronary angiography. Surgical revascularisation was indicated in patients with coronary lesions in whom thallium myocardial perfusion imaging showed myocardial ischaemia at rest or under stress (dipyridamole injection). Reoperations were performed at a mean age of 6.4 years (range 1.9—10.9 years).

Fourteen patients underwent surgical coronary patch angioplasty using, in most cases, a saphenous vein patch. Coronary angioplasty involved the right coronary artery in two cases, the left anterior descending artery in three and the left main coronary artery in the remaining nine patients. One patient with a long atretic segment of the left main stem underwent implantation of a short saphenous vein bypass between the ascending aorta and the distal left main coronary artery. Two patients had a mammary bypass using the left mammary artery.

There was no postoperative or late mortality. One patient with a residual stenosis of the left anterior descending artery underwent a second reoperation (mammary bypass) 2 years after the first reoperation. After a mean follow-up of 6.5 years, all patients were leading a fully active, symptom-free life. At myocardial perfusion imaging, myocardial perfusion was normal in 15 patients and showed minimal residual perfusion defects in 2.

Table 1
Causes of early reoperations (within the first postoperative year)

<table>
<thead>
<tr>
<th>Cause</th>
<th>n</th>
</tr>
</thead>
<tbody>
<tr>
<td>Coronary obstruction</td>
<td>4</td>
</tr>
<tr>
<td>Aortic coarctation (residual or recurrent)</td>
<td>4</td>
</tr>
<tr>
<td>Right ventricular outflow tract obstruction</td>
<td>2</td>
</tr>
<tr>
<td>Pace-maker implantation</td>
<td>1</td>
</tr>
</tbody>
</table>

Table 2
Causes of late reoperations (after the first postoperative year)

<table>
<thead>
<tr>
<th>Cause</th>
<th>n</th>
</tr>
</thead>
<tbody>
<tr>
<td>Coronary obstruction</td>
<td>18</td>
</tr>
<tr>
<td>Right ventricular outflow tract obstruction</td>
<td>11</td>
</tr>
<tr>
<td>Left ventricular outflow tract lesion</td>
<td>3</td>
</tr>
<tr>
<td>Tracheal compression</td>
<td>1</td>
</tr>
<tr>
<td>Pulmonary hypertension (Potts anastomosis)</td>
<td>2</td>
</tr>
</tbody>
</table>
3.4. Reoperations for right ventricular outflow tract (RVOT) obstruction

Eleven patients underwent reoperation for RVOT obstruction that was detected at a routine echo-Doppler evaluation. At the time of reoperation, the mean gradient was 77 mmHg (range 60–100 mmHg). Reoperation was performed at a mean age of 6.8 years (range 1.3–15.4 years). The obstruction was supravalvular (located at the anastomotic site) in eight cases and involved the whole RVOT (including valvular and subvalvular areas) in three patients.

In eight cases, supravalvular pulmonary stenosis was repaired by enlargement using a heterologous pericardial patch. In three patients with diffuse obstruction, a transannular patch was inserted. One patient had a concomitant coronary artery bypass for coronary obstruction.

There was no postoperative or late mortality. After a mean follow-up of 6.8 years, the mean residual gradient was 23 mmHg (range 10–45 mmHg). No further reoperation was necessary.

3.5. Reoperations for left ventricular outflow tract (LVOT) lesions

In three patients (all with TGA/VSD), LVOT lesions developed and needed reoperation at a mean age of 5.8 years (range 2.6–11.6 years). One patient had aortic coarctation repair 3 years after ASO. One patient developed severe aortic regurgitation with aneurysmal dilatation of the ascending aorta. The native pulmonary valve was bicuspid. This patient underwent a Bentall procedure with a mechanical valve 11.6 years after ASO. In the last patient, reoperation was performed for subaortic stenosis 4.1 years after ASO.

There was no mortality and the three patients were alive and well without residual lesions after a mean follow-up of 4.7 years (range 2.5–8.8 years).

3.6. Other late reoperations

One patient with right aortic arch and abnormal retrooesophageal left subclavian artery developed symptomatic tracheal compression 16 months after ASO; symptoms were relieved by section of the left subclavian artery.

Two patients had severe pulmonary hypertension with suprasystemic pulmonary artery pressure. Due to the high risk of sudden death, the decision was taken to construct a shunt between the left pulmonary artery and the descending aorta (Potts anastomosis). Respectively 6 and 4 years after reoperation, both patients were alive and doing well.

4. Discussion

4.1. Reoperations after ASO: incidence and outcome

All reported series show that the need for reoperation after ASO is between 5% and 10% [1–6]. Most reoperations are performed during the first years following ASO. This initial hazard phase is followed by a period during which the risk of reoperation is very low. One study has suggested that there may be a slightly ascending late phase due to the increasing need for neo-aortic valve surgery [2]. It is usually said that the risk of reoperation is increased in complex TGA; this is not the case in our experience, except for patients with associated aortic arch obstruction (with or without ventricular septal defect). Reoperations can be carried out with a very low operative risk. Survival and functional outcome are not affected by the need for reintervention.

4.2. Reoperations for RVOT obstruction

RVOT obstruction represents, in most series, the most frequent cause for reoperation [1–6,8]. In our series, late reoperation for RVOT obstruction was necessary in 1.5% of survivors. The risk is higher during the first postoperative years but there is a persistent constant low risk thereafter [2]. Most obstructions are located at the neo-pulmonary anastomotic site and are clearly related to inadequate growth. In our experience, percutaneous dilatation has usually been ineffective and reoperation with patch angioplasty is advocated.

Several technical factors are probably important to reduce the incidence of this complication: (1) use for the reconstruction of the pulmonary trunk of a generous pantaloon-shaped patch of fresh autologous pericardium, tailored so as to correct the discrepancy in size between the neo-pulmonary root and the distal pulmonary bifurcation, (2) extensive mobilisation of the pulmonary arteries to allow tension-free anastomosis after Lecompte manoeuvre, (3) use of absorbable suture material. It should be pointed out that observing a mild gradient (around 20 mmHg) is common; in most patients, careful observation does not show any change in pressure gradient over time indicating adequate anastomotic enlargement [1].

In patients with TGA, VSD and associated aortic arch obstruction, late RVOT may involve the subvalvular and the valvular area [8,9]. The presence of an initial small aortic root is probably a major risk factor. Routine resection of subaortic obstruction at the time of ASO may promote growth of the neo-pulmonary annulus [9]. Once established, relief of RVOT obstruction requires placement of a transannular patch, or of an extracardiac conduit if coronary anatomy precludes transannular patching.

4.3. Reoperations for coronary lesions

In reported large series, reoperations for late coronary lesions are rare. This is not the case in our experience in which coronary lesions represent the main cause for reoperation. It has been clearly established that most patients with coronary lesions do not show any clinical, electrocardiographic or echocardiographic evidence of myocardial ischaemia [10]. Only routine systematic evaluation (by coronary angiography or multislice computed tomographic angiography) can detect coronary lesions. Our policy is to perform coronary evaluation: (1) early in the presence of any electrocardiographic or echocardiography finding, even minimal, suggestive of myocardial ischaemia, (2) during the first postoperative year in patients with unusual coronary patterns or intraoperative difficulties in coronary transfer and (3) routinely before 5 years of age.
Early coronary lesions are more frequent in patients with unusual coronary patterns [11] and various technical factors have been incriminated (inadequate coronary transfer, excessive use of fibrin glue, abnormal early fibrosis). The pathophysiology of late coronary lesions remains much more obscure. It has been postulated that reimplantation of the coronary arteries may produce subite flow abnormalities, which, in turn, alter shear stress and induce progressive intimal thickening. Late coronary lesions may occur in all coronary patterns and they are progressive, thus requiring repeated coronary evaluation at regular intervals during follow-up.

To date, our policy has been to perform revascularisation only in patients in whom myocardial ischaemia at rest or under stress could be demonstrated. Whether revascularisation should be carried out even in the absence of evident myocardial ischaemia remains to be determined.

The optimal management of coronary lesions is controversial. Percutaneous transluminal coronary angioplasty, mammary bypass and surgical coronary angioplasty have been used. Our own experience shows that surgical coronary angioplasty can be performed with a very low operative risk and a high patency rate and that this procedure restores normal myocardial perfusion in most cases [12].

4.4. Reoperations for LVOT lesions

LVOT obstruction may develop in a few patients with TGA/VSD. Trivial aortic regurgitation is common. Regurgitation grade II or more is rare but increases in frequency with time [1,3,13]. Aortic root dilatation is also common but usually stabilises without significant progression over time [14]. All these LVOT lesions are more frequent in TGA with VSD. Several factors may be implicated: initial discrepancy in size between pulmonary and aortic roots, modification in neo-aortic root anatomy by coronary reimplantation or alteration in valve anatomy by VSD closure.

To date, reoperation for aortic valve surgery (repair or replacement) with or without replacement of the ascending aorta have been very rare in all reported series. However, the increasing incidence of significant aortic regurgitation with time, although slow, is disturbing. The very long-term fate of the neo-aortic valve remains unknown. Close observation is warranted.

4.5. Other reoperations

Mitral valve reoperations are rarely necessary for ischaemic mitral regurgitation or residual mitral valve malformation (mitral cleft) [2].

Tracheobronchial compression by vascular structures (aorta or pulmonary arteries) may develop and require surgical relief [1].

The association of TGA with pulmonary vascular disease and pulmonary hypertension is rare but severe [15]. In an attempt to reduce the risk of sudden death in patients with suprasystemic pulmonary artery pressure, a Potts anastomosis may be constructed [16]. This has been successful in two of our patients although the very late prognosis remains poor.

4.6. Left ventricular function

In the absence of myocardial infarction or severe aortic regurgitation, left ventricular function is normal [1,2,5]. It remains however to determine if this will be the case in the very long-term.

5. Conclusions

Late after neonatal ASO, the outcome in terms of survival and functional status is very satisfactory. Reoperations for coronary lesion or RVOT obstruction are not rare. The operative risk is low and late results are good. Careful observation, especially in the period of rapid somatic growth during adolescence, is necessary to detect and treat these lesions. To date, neo-aortic valve function and left ventricular function are maintained in a great majority of patients. Whether this will remain the case in the decades to come requires further evaluation.

Acknowledgement

We gratefully acknowledge the secretarial assistance of Miss Corinne Pasquet.

References


