Results of biventricular repair of congenital cardiac malformations: definitive corrective surgery?


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Abstract

Objectives: Congenital cardiac malformations are usually corrected in the neonatal period or in early infancy. Corrective surgery may not always be definitive, especially in complex malformations. Long-term morbidity is influenced by reoperations and their risk. Methods: This study analyzes our single-center experience over more than 20 years in a selected group of patients. Data were gathered, with special focus on causes and incidence for reoperations, respectively. Results: Freedom from reoperation after 5, 10, and 15 years for each cardiac malformation was determined. The numbers describe in the following order patient years (y), number of patients (n), and freedom from reoperation at follow-up interval (%), respectively: atrial septal defect (15 864y, n = 1198, 99 ± 0/99 ± 0/99 ± 0), partial atrioventricular septal defect (2506y, n = 234, 95 ± 2/93 ± 2/93 ± 2), total anomalous pulmonary venous connection (742y, n = 141, 93 ± 1/91 ± 0/91 ± 0), complete atrioventricular septal defect (1715y, n = 377, 81 ± 3/76 ± 3/72 ± 4), tetralogy of Fallot <1 year (1503y, n = 197, 94 ± 4/85 ± 1/74 ± 3), transposition of great arteries (1459y, n = 375, 88 ± 2/83 ± 4/73 ± 7), interrupted aortic arch (IAA) (481y, n = 98, 63 ± 6/52 ± 7/45 ± 8), common arterial trunk (CAT) (599y, n = 109, 64 ± 6/24 ± 6/11 ± 5). Conclusions: In most congenital malformations surgical correction is definitive and the rate of reoperations is low. In complex anomalies, such as CAT and IAA, reoperations at long-term are more common. Analysis of such results and recognition of a sometimes inevitable operative morbidity helps to predict long-term outcome and influences the follow-up.

Keywords: Congenital cardiac surgery; Corrective surgery; Reoperation

1. Introduction

For the last 20 years the main issue in congenital cardiac surgery was to lower operative mortality. However, to date mortality is rather low. In the German Heart Center Munich, for example, mortality for all corrective operations in infants less than 1 year of age from January 1999 to September 2000 was 3.3% (n = 332). Therefore, today long-term morbidity and quality of life after correction of congenital malformations becomes the primary focus.

This study represents a long-term evaluation of patients with selected operated cardiac malformations at our institution. Atrial septal defect (ASD), partial atrioventricular septal defect (P AVSD), as well as some complex malformations such as total anomalous pulmonary venous connection (TAPVC), complete atrioventricular septal defect (CAVSD), tetralogy of Fallot (TOF), common arterial trunk (CAT), interrupted aortic arch (IAA), and transposition of great arteries (TGA) were included. Other malformations, such as ventricular septal defects, congenitally corrected transpositions of the great arteries, and the entity of functional single ventricles are in this present study not yet included as we are still awaiting data from our large collectives. Undoubtedly the entity of functional single ventricles plays an important role in regard of multiple (or staged) procedures which lead nowadays finally to completion of a cavopulmonary circulation.

In the early days of cardiac surgery, the term correction came into use to distinguish attempts at reparative operations from the then more common palliative procedures. But in what sense are they corrective? Anatomical correction (e.g. ASD) may differ from physiological correction (e.g. TOF) or hemodynamic correction (e.g. Mustard procedure in TGA). Our findings showed that the likelihood for reinterventions in some complex malformations at long-term
are high. Analysis of such results and recognition of a sometimes inevitable operative morbidity helps to predict long-term outcome, quality of life after correction, and influences the follow-up.

2. Patients and methods

The medical records in regards to the initial clinical features, pathophysiological findings, surgical treatment and hospital mortality were reviewed. Data from outpatients, or those dying after hospital dismissal, were obtained from attending physicians, rehospitalization records, or death certificates. In less than 4% of the patients a late follow-up was retrieved. For statistical analysis the measured values are expressed as mean, standard deviation (SD), range (minimum, maximum) and median. Freedom from reintervention is expressed in Kaplan–Meier curves.

For each malformation the number of patients, the observation period, the male/female ratio and the age at repair are listed in Table 1.

2.1. ASD

In 57% of the patients the ASD was closed with a Dacron patch; the remainder underwent direct closure.

2.2. PAVSD

The ‘cleft’ of the left atrioventricular valve (LAVV) was repaired in all but two patients (one required LAVV replacement with a mechanical prosthesis upon primary repair).

2.3. TAPVC

Forty-five percent had a supracardiac, 33% an infracardiac, 15% a cardiac and 7% a mixed type anomaly. The operative technique has varied over the years: In the beginning, a right atrial transverse incision across the atrial septum into the left atrium to the base of the left atrial appendage was used [1]. The transseptal approach via the right atrium only was abandoned during the last decade. The heart was tilted up to the right, giving excellent exposure to the left atrium and the common pulmonary venous sinus. The ASD was closed with a Dacron patch.

2.4. CAVSD

Sixty-six percent of patients had primary repair, 35% were < 6 months. Twenty-two percent of patients underwent palliative surgery 1 month to 11.9 years (mean 3.5 ± 2.9 years) prior to repair in cases with high pulmonary vascular resistance (Rp > 6 U/m²) not reactive to oxygen, hypoplastic left ventricle (left ventricular end diastolic volume < 50% of predicted normal), coexisting coarctation and dystrophic neonates. Between 1974 and 1978 repair was performed using the one-patch technique [2] in 23 (7%) patients Since 1978, the two-patch technique [3] was applied in 304 (93%) patients. Mitral valve anomalies were: double orifice mitral valve (three patients), single left ventricular papillary muscle (four patients), and dysplastic valve with extreme deficiency of mitral tissue (six patients). Down’s syndrome was present in 71% (166 patients).

2.5. TOF

Primary repair was performed in 89% of the patients, whereas 21 patients underwent a two-stage repair, mainly because of small pulmonary arteries or anomalies of the coronary arteries. The VSD was closed with a Dacron patch, using either interrupted, with pledgets reinforced mattress sutures or with an over and over suture. Right ventricular outflow tract obstruction (RVOTO) was relieved by infundibular resection and/or pulmonary valve commissurotomy in 51 patients (26%), RVOT patch enlargement with Gore-Tex or autologous pericardium in 98 (49%), and a transannular patch was implanted in 38 patients (19%). In five patients a right ventricular to pulmonary artery (RV-PA) conduit was implanted; five additional patients had a transatrial repair because of an anomalous coronary artery crossing the RVOT.

2.6. TGA

Two hundred and twenty-six patients had simple TGA, 93 TGA with VSD, and 22 transposition like forms of double

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>n</th>
<th>Observation period</th>
<th>Male/female (%)</th>
<th>Age at repair</th>
<th>Median age</th>
</tr>
</thead>
<tbody>
<tr>
<td>ASD</td>
<td>1198</td>
<td>1974–1998</td>
<td>36/64</td>
<td>1 day–76 years</td>
<td>14 years</td>
</tr>
<tr>
<td>PAVSD</td>
<td>234</td>
<td>1974–1999</td>
<td>49/51</td>
<td>18 days–69 years</td>
<td>5 years</td>
</tr>
<tr>
<td>TAPVC</td>
<td>141</td>
<td>1974–1999</td>
<td>61/39</td>
<td>2 days–13 years</td>
<td>28 days</td>
</tr>
<tr>
<td>CAVSD</td>
<td>377</td>
<td>1974–1998</td>
<td>40/60</td>
<td>30 days–12 years</td>
<td>205 days</td>
</tr>
<tr>
<td>TOF &lt; 1 year</td>
<td>197</td>
<td>1974–2000</td>
<td>60/40</td>
<td>21 days–1 year</td>
<td>240 days</td>
</tr>
<tr>
<td>TGA</td>
<td>375</td>
<td>1983–2000</td>
<td>47/53</td>
<td>2 days–14 years</td>
<td>11 days</td>
</tr>
<tr>
<td>IAA</td>
<td>98</td>
<td>1975–2000</td>
<td>34/66</td>
<td>2 days–175 days</td>
<td>19 days</td>
</tr>
<tr>
<td>CAT</td>
<td>109</td>
<td>1976–2000</td>
<td>57/43</td>
<td>14 days–9 years</td>
<td>135 days</td>
</tr>
</tbody>
</table>

* ASD, atrial septal defect; PAVSD, partial atroioventricular septal defect; TAPVC, total anomalous pulmonary venous connection; CAVSD, complete atroioventricular septal defect; TOF, tetralogy of Fallot; TGA, transposition of great arteries; IAA, interrupted aortic arch; CAT, common arterial trunk.
outlet right ventricles (DORV)/Taussig–Bing anomaly (TB). In all but four patients with side-by-side great vessels, the Lecompte maneuver was used. Coronary transfer was initially undertaken following the guide Lines of Quaege-beur [4], whereas after 1990 the trap-door technique was primarily used.

Three different surgical techniques were employed for pulmonary artery reconstruction. From 1983 to 1989, direct pulmonary artery anastomosis without patch material was employed in 40 patients [5]. Between 1984 and 1994, reconstruction with two free pericardial patches was employed in 101 patients, and since then the ‘pantaloan patch’ technique [6] has been applied.

2.7. IAA

All patients were operated through a median sternotomy, except for five patients who were operated between 1974 and 1979 via a lateral thoracotomy. Usually, a direct anastomosis of the arch could be accomplished. In nine cases, either a prosthesis interposition or a patch augmentation was performed. The etiologic substrate of IAA is often associated with the presence of a left ventricular outflow tract obstruction (LVOTO), most often due to a malalignment VSD to the left and posterior. In the case of a documented relevant LVOTO, a transaortic approach was used in two cases, a transatrial in three, a right ventricular in one, and a combined approach in one case for relief.

2.8. CAT

In 36 cases, after resection of the pulmonary arteries, the resulting defect in the common trunk was closed using a Gore-Tex patch, in 30 with glutaraldehyde-treated pericardium and otherwise by direct suture. In 61 patients, an enlargement of the VSD was performed. The VSD was subsequently closed with either Dacron patch or Gore-Tex, by means of interrupted sutures buttressed with felt pledgets. The pulmonary arteries were connected to the RV by a homograft in 69 patients, by a Hancock porcine conduit in 35 patients and by a Carpentier–Edwards pericardial conduit in four patients The initial conduit size was 9–12 mm in diameter in 68 patients and larger than 12 mm in 41 patients. Truncal valve reconstruction was performed in 38 patients.

3. Results

For freedom from reoperation for the selected cardiac defects at 5, 10, and 15 years, see Table 2.

3.1. ASD

The main indication for reoperation was recurrent ASD, 20 days to 9 years after primary correction. Three patients additionally required a permanent pacemaker system. There were no deaths at reoperation.

3.2. PAVSD

Nineteen (8%) patients required 27 reoperations. The time interval was 38 days to 27 years (mean 4.0 ± 7.7 years) after primary repair. Main indication for reoperation was considerable LAVV incompetence as reported by El Najdawi et al. [7]. Refixation and repair was possible in most cases, except in nine, requiring valve replacement. Other indications were subaortic stenosis, recurrent ASD and tricuspid valve incompetence. Nine (4%) patients additionally required a permanent pacemaker system at long-term. There were no deaths at reoperation.

3.3. TAPVC

Indication for reoperation was given in 12 (9%) patients, 1 day to 9.2 years (mean 128 days ± 3.5 years) after primary correction. Main causes for reoperation were restenosis at the anastomosis in five patients, as found in equally large collectives [8–10]. In one patient, functional restenosis of the anastomosis was due to hypoplastic pulmonary veins. In this patient, extensive surgical revision and enlargement-plasty could not prevent late death due to progressive pulmonary hypertension. Three patients were operated due to a residual atrial shunt. Two patients, because of superior vena cava thrombosis on the first postoperative day. One patient because of an additional membrane in the left atrium, which was not identified during the first operation. Another patient required a pacemaker 16 years after initial correction because of total atroventricular block.

3.4. CAVSD

Thirty-nine (11%) patients required 50 reoperations. Time interval was 7 days to 8.0 years (mean 1.2 ± 2.0 years) after repair. Time-related freedom from reoperation is shown in Fig. 1. Main indication for reoperation was considerable LAVV incompetence. Refixation and repair of LAVV was possible in most cases, except in 11 patients requiring valve replacement. Other indications were subaortic stenosis, recurrent VSD and tricuspid valve incompetence. Eighteen patients additionally required a permanent pacemaker system, in two cases because of congenitally
acquired AV-block. Five patients died early after reopera-
tion. The trend towards primary repair revealed that the 30-
day mortality could be reduced from 17.7% in 1974–1979 to

3.5. TOF

Twenty-four reoperations (12%) had to be performed 3 
months to 15.7 years (mean 4.2 ± 4.9 years) after repair.
Time-related freedom from reoperation is shown in Fig. 2. 
Causes for reoperations were residual VSD in six patients,
recurrent RVOTO in four, peripheral LPA stenosis in four,
RVOT aneurysm in three and implantation or replacement
of a RV-PA conduit in six patients There were no deaths at
reoperation. Among the three patients who developed an
aneurysm of the RVOT, there was neither a relation between 
the type of material used to enlarge the RVOT (autologous 
pericardium or Gore-Tex), nor had the pressure ratio RV/LV 
been more than 0.7 at repair. Equal findings after early 
correction have been reported by Hennein et al. [12] and 
Caspi et al. [13].

3.6. TGA

Reoperations were required in 33 patients; seven under-
went a second reoperation. Time-related freedom from
reoperation is shown in Fig. 3. Reoperations were signifi-
cantly less common in patients with simple TGA (1 month
to 3.2 years; mean 44.2 ± 47.5 months), whereas 16 patients
with TGA and VSD (15%) required reintervention after a 
mean time of 22.1 months (±34.1 months) and 8 patients
with DORV/TB (27%) required reoperation after a mean 
time of 16.5 months (±40.7 months). The main reason for 
reoperation was associated with stenosis of the RVOT, with-
out influence of the various techniques on pulmonary artery 
reconstruction, as found in other equally large collectives 
[14–16]. A second reoperation had to be performed for resi-
dual valvular and subvalvular stenosis; one of them under-
went patch annuloplasty of the RVOT and in the remaining
two a RV-PA conduit was necessary. In one of these 
patients, additional tricuspid insufficiency required annulo-
plasty. One had LVOTO resection, one required a second 
angioplasty of the right and left pulmonary artery and one 
patient underwent aortic valve replacement. Almost 2 years 
after mitral valve annuloplasty, mitral valve replacement 
was necessary due to persistent severe regurgitation in one 
patient Six patients with complete heart block after VSD 
closure and one patient with sick sinus syndrome required 
additional permanent pacemaker implantation. There were 
no deaths at reoperation.

Forty-six patients (17%) underwent follow-up cardiac 
catheterization with coronary angiography. In two patients 
coronary occlusion could be detected. While one patient 
underwent coronary revascularization with the internal 
mammary artery, the other had sufficient retrograde 
collateralization of the left coronary artery with only mild 
myocardial dysfunction under stress conditions and did not 
require any intervention.

3.7. IAA

Recurrent arch obstruction (20 patients) was the most 
common indication for reoperation [17]. Upon revision, 
incomplete resection of the ductal tissue and/or undue 
tension were assumed to be causes for restenosis. Time 
interval was 1 day to 21 years (mean 4.8 ± 5.8 years). 
Time-related freedom from reoperation is shown in Fig. 4.
A residual LVOTO required resection through a transaortic approach in two, a ventriculotomy in one, and an enlargement of a VSD patch in one. A further two transaortic resections were performed at the time of the second reoperation. In two cases, a homograft was replaced for previous CAT. Bronchial compression was found in nine cases and surgically relieved by aortopexy (one patient), aortic arch augmentation (four patients), pulmonary artery mobilization and patch augmentation (two patients), resection of a tracheal stenosis (one patient), bronchial wall suspension by a ring enforced prosthesis (one patient). There was one early death after relief of bronchial compression. A residual VSD needed closure in three cases. Consecutive diaphragmatic plication was necessary in five patients due to paresis. A complete atrioventricular block after VSD closure occurred twice, requiring the additional implantation of a permanent pacemaker in one case (the other patient died early).

3.8. CAT

A total of 64 reoperations were performed after 20 days to 13.0 years (mean 4.9 ± 5.0 years). Time-related freedom from reoperation is shown in Fig. 5. Five patients were reoperated within 30 days after primary repair, and seven within 3 months. Three had a residual VSD, four a truncal valvar dysfunction, three a bronchus and/or trachea obstruction, one an aneurysma at the site of the RVOT, and two a complete atrioventricular block. Reasons for reintervention were mainly due to outgrowth and failure of the conduit [18]. At time of first reoperation, 45 conduits had to be replaced (allograft n = 41, Hancock n = 4) after 6 months to 13.4 years (mean 6 ± 6.2 years). Aortic homografts had to be replaced after a mean of 4.9 years, pulmonary after a mean of 3.1 years, Hancock after a mean of 4.4 years, and Carpentier–Edwards after a mean of 9.2 years. Four truncal valves were replaced (St. Jude 25, Biocor 21, Hancock 22, Carpentier–Edwards 27).

4. Discussion

Nowadays, the operative mortality for the very early correction of a great diversity of even very complex cardiac defects has become considerably low. Therefore, in recent years, the focus of attention shifted from the issue of pure survival to the issue of the quality of life after surgical correction.

The present investigation shows that the long-term fate of children after primary correction of selected cardiac defects in early infancy is considerably different for various cardiac malformations. For some defects, such as ASD and PAVSD, the reoperation rate is trivial, so that those malformations can be considered ‘cured’ after primary surgical correction. Other cardiac defects, such as CAVSD, TGA and TOF, require a somewhat higher rate of reoperative procedures, also dependent on the individual morphologic features. After correction of IAA, and even more so of CAT, the reoperation rate is considerably high.

‘Corrective’ does not mean necessarily ‘curative’. Primary repair of congenital cardiac defects is only successful and hence curative if it provides a permanent correction, leading to a normal heart function and a normal life span. Early repair of defects such as CAVSD, TOF, CAT, IAA, or TGA are now considered routine, and in most instances the results of correction in infancy surpass those of staged procedures. Still, residual defects or sequelae of the primary operation are predictable. Primary corrective operations may leave behind certain ‘abnormalities’, of which some could be avoided by even more thorough attention during the primary repair. These ‘avoidable’ residual defects are recurrent shunts between the ventricles or the atria, as in all operations with closure of ventricular and atrial septal defects. Likewise, postoperative AV-block, incomplete infundibular resection as in TOF repair, missing ‘cleft’-closure as in AVSD repair, incomplete resection of ductal tissue and therefore undue tension on
the anastomosis as in IAA repair, compression of adjacent anatomical structures and incomplete LVOTO relief as in IAA repair, restenosis at the anastomotic site as in TAPVC repair, stenosis of the reconstructed pulmonary arteries as in TGA repair, or distortion of anomalous coronary arteries as in CAT repair, could sometimes be avoided if the multitude of pathological diversities did not challenge even the most experienced surgeons. Therefore ‘avoidable’ in this case is theoretical, since it will hardly be possible to master all technical challenges with a 100% success rate. However, increasing awareness of the potential residual defects upon primary repair may enhance the surgeon’s awareness and thereby further his technical skills. This was one of the intentions of our retrospective investigation.

On the other hand, some reoperative procedures are inevitably predicted by the pathology of the cardiac malformation. For example, in patients with CAEVD, dysplastic AV valves and severe AV valve regurgitation prior to repair predicts the likelihood of postoperative failure of valve reconstruction. Likewise, the need for a transvalvar patch in TOF, the presence of hypoplastic pulmonary veins in TAPVC, and the presence of a significantly malformed trunval valve in CAT also predict almost certainly the need for further reoperations. In general, the use of allografts or valved conduits still predicts the necessity for multiple replacements [19,20]. Also in the entity of functional single valve in CAT also predict almost certainly the need for multiple reoperations. Various staged and primarily palliative procedures can only lead to partial or complete cavopulmonary circulation.

In an editorial in 1986, de Leval [21] already remarked that “in future the attitude towards the treatment of complex cardiac malformations in neonates will be based on the quality of adult life by patients operated on today”. Stark remarked in a guest lecture in 1989 [22] that “Ideally, an operation for congenital heart defects should be considered corrective if (1) normal function is achieved and maintained, (2) life expectancy is normal, and (3) further medical or surgical treatment is not necessary” and pointed out that “the major discoveries will probably not be in the surgical knitting and stitching but in finding the causative mechanisms of congenital heart defects”. Kirklin stressed in 1990 [23] the fact that “every patient care decision is based on a prediction of outcome for the patient in terms of time-related events, and a comparison of these predicted outcomes after one form of treatment with that after another form of treatment”. It is therefore one of the responsibilities of both the cardiac surgeon and the cardiologist to provide the patient or the patient’s parents with thorough information about the implications of the individual cardiac malformation on possible surgical intervention, operative risks, long-term morbidity and quality of life. The present investigation was considered to further our understanding about quality of surgical repair in the long-term, i.e. the 20–25 years that are available today.

References


Appendix A. Conference discussion

Dr W. Daenen (Leuven, Belgium): This was a very nice paper with very careful follow-up. I suppose that the completeness of your follow-up was nearly over 90%.

Dr Lange: 96%.

Dr Daenen: Did you have any idea what the incidence was for catheter reintervention in these patients?

Dr Lange: I have no idea. That’s only because when we got a new cardiologist four years ago, the catheter interventions really started in our center. Before there were very little interventions, but I don’t have the numbers.

Dr Daenen: I asked this because I presume that the incidence would be high.

Dr Lange: Yes.

Dr B. Maruszewski (Warsaw, Poland): Your paper is very informative. I wonder why you didn’t include the most frequent condition, which is VSD. I think demographically, for sure it is the most frequent condition. And I think we would like to know, is it a curative operation and corrective operation? What is the long run?

Dr Lange: You are absolutely right, and I can give only one conclusive answer, that we just didn’t get it done. It was a large number of patients, and we wanted to include them, but we didn’t finish it. But it’s very important, of course.

Dr G. Ziemer (Tubingen, Germany): I just may pick on one of your diagnoses, namely truncus arteriosus. First question: Is this 10% bronchial obstruction in truncus only in those patients with interrupted aortic arch, the Van Praagh type A4 patients?

The second question is with regard to your notion that reoperation in truncus was kind of a programmed one because you used conduits in the original repairs. Is there any newly developed consequence out of your experience, like just forgetting about conduits in truncus and repair it like a tetralogy?

Dr Lange: The first question, you’re right, those are the patients with the truncus 4. Now, to the second question, we haven’t done any attempts to get rid of the conduit yet, but it’s certainly something very serious to consider.

Dr A. Urban (St. Augustin, Germany): I have two questions. Was the rate of reoperation related to the original population of the series or was it related only to the survivors of the first operation?

The second question is, the reoperation for AVSD was mainly in the very early postoperative period. Was that because the cleft was not closed, and, if so, did you recently change your policy in closing the cleft and has the rate of reoperation then gone down?

Dr Lange: The mortality, the patients who died perioperatively I excluded from the calculated reoperation rate. Now, in the German Heart Center the cleft is always to be closed. I said this because, like in any other center, we have reoperations from other hospitals, too, and those, we frequently find that the cleft was not closed. In those patients who have been reoperated early after complete AV canal, it’s mainly dysplastic valves, tearing of valve construction, technical problems.

Dr W. Daenen: I have another question on the truncus group. One of the most difficult problems I conceive is the timing for replacement or repairing the truncal valve, because many of these patients have some truncal valve insufficiency and sometimes you have to replace a conduit, and then the question that arises is should we do something to the truncal valve as well despite the fact that it’s well tolerated?

Dr Lange: You mean primary replacement of the valve? Our policy is to avoid primary replacement?

Dr Daenen: I mean reintervention. When you replace the conduit and you have some truncal valve insufficiency, when do you repair or replace a truncal valve?

Dr Lange: We still have the policy even when we replace the conduit to repair the valve, if possible, and not to replace it, and we know from the St. Augustin group that the primary replacements have an extremely high mortality.

Dr Urban: They are difficult but we had no mortality as yet in this subgroup of patients with truncus arteriosus.