Single centre experience on primary correction of common arterial trunk: overall survival and freedom from reoperation after more than 15 years

Christian Schreiber, Andreas Eicken, Gunter Balling, Michael Wottke, Gebhard Schumacher, Sung Un Paek, Hans Meisner, John Hess, Rüdiger Lange

Department of Cardio-thoracic Surgery, German Heart Centre at the Technical University, Lazarettstrasse 36, 80636 Munich, Germany

Department of Paediatric Cardiology, German Heart Centre at the Technical University, Lazarettstrasse 36, 80636 Munich, Germany

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Abstract

Objective: To identify predictors of early and late outcomes of common arterial trunk (CAT) after primary surgical correction, such as clinical state prior to surgery, age and weight at presentation, implications of truncal valve abnormalities and associated anomalies of this complex congenital malformation.

Methods: A consecutive series of 106 patients, operated between 1976 and 1998, were reviewed retrospectively. Using the ‘Van Praagh’ classification, 59 patients presented as A1, 33 as A2, six as A3, and eight as A4. The mean age at operation was 8.6 months (range, 14 days–8.9 years; median, 4.4 months), and the mean weight was 5.2 kg (range, 2.5–30.8 kg; median, 4.4 kg). At the time of operation, 32 patients had congestive heart failure, five were on ventilator support for less than 1 week, and 21 for more than 1 week.

Results: Between 1976 and 1989, the early mortality was 21%, and between 1990 and 1998, it dropped to 13%. After 1, 10 and 15 years, the overall survival was 66, 61 and 59%. The 1, 5, 10 and 15 year freedom from reoperation was 82, 60, 22 and 10%, respectively. Clinical condition prior to intervention, truncal valvar dysfunction, and coronary anomalies were significantly associated with poor surgical outcome, whereas weight and age at presentation in our series were not.

Conclusions: In the current era of paediatric cardiac surgery, primary surgical repair of CAT can be carried out with reasonable early and late mortality. However, our data suggest that a high incidence of reoperation, mainly due to the outgrowth and failure of the conduit, has to be expected. The patient’s clinical state after diagnosis is decisive for the timing of intervention. © 2000 Elsevier Science B.V. All rights reserved.

Keywords: Common arterial trunk; Truncus arteriosus communis; Truncal valve; Primary surgical repair

1. Introduction

Common arterial trunk (CAT), or truncus arteriosus communis (TAC), is the entity in which one arterial trunk supplies the coronary arteries and both systemic and pulmonary circulation directly [1]. It can be associated with many cardiac or cardiovascular malformations [2], and occurs in 2–4% of all cases of congenital heart disease [3,4]. Patients with CAT usually show signs and symptoms of congestive heart failure in early infancy, and mild to moderate cyanosis before 12 months of life [5,6]. If not operated on in infancy, about 80% of these children die within the first year of life [3,7]. One of the major perioperative risks is pulmonary hypertension (PH) [8,9]. However, residual pathologic complications after the early postoperative period, such as increasing truncal valvar insufficiency, can influence the long-term outcome of these patients.

This study was designed to identify predictors of early and late outcome, and we review our institutional experience over more than 15 years. The likelihood of reoperation within this timespan is pointed out, and the implications of truncal valve abnormalities on both surgical procedure and risk are discussed.

2. Patients and methods

Between 1976 and 1998, 106 patients (62 male, 44 female) underwent primary surgical repair of the CAT. We excluded those who had had prior palliative procedures. The records of all these patients were reviewed and data was gathered concerning the initial clinical features, including catheter or echocardiographic findings. Furthermore, the operative procedures, the immediate postoperative interval,
and the subsequent course of each patient was followed up. Early death was defined as death occurring from the intraoperative period up to 30 days after surgery. Death occurring after that period was defined as late death. Data concerning outpatients or those dying after dismissal were obtained from attending physicians, rehospitalization records or death certificates.

Using the ‘Van Praagh’ classification, 59 patients presented as A1, 33 as A2, six as A3, and eight as A4. The patients were classified in functional classes prior to surgery. Asymptomatic patients were in NYHA I, and patients with tachypnea, failure to thrive and poor feeding were classified as NYHA II. Intubated patients with mildly impaired cardiac function were classified as NYHA III, and the patients intubated for more than 1 week and under increased antiangiogetic medication were in NYHA IV.

According to these criteria, 58 patients were in NYHA II, five were on ventilator support for less than 1 week (NYHA III), and 25 for more than 1 week (NYHA III). All patients under 6 months were on diuretics. Patients with A4 malformation received prostaglandins for patency of the arterial duct. No patient needed dialysis preoperatively. The mean age at operation was 8.6 months (standard deviation (SD), 16.3 months; range, 14 days–8.9 years; median, 4.4 months), and the mean weight was 5.2 kg (SD, 3.8 kg; range, 2.5–30.8 kg; median, 4.4 kg).

Between 1976 and 1987, the mean age at time of operation was 11.6 months, and the mean weight was 5.8 kg, whereas between 1988 and 1998, the mean age was 4.9 months, and the mean weight was 4.5 kg. There were 13 cases of coronary anomalies, 18 of right aortic arches (RAA), 15 of pulmonary branch stenosis (PBS), five of RAA and PBS, one of pulmonary sling, and two of anomalous pulmonary venous drainage. The coronary arteries exhibited a variable pattern. There were seven single coronary arteries, four cases of a high origin of the left coronary artery, and two of a high coronary artery alone. We classified the grade of truncal valve stenosis according to the invasively determined peak to peak gradient (I, <15 mmHg; II, <30 mmHg; III, >30 mmHg), and the grade of insufficiency as trivial, moderate, or severe, according to the Hunt criteria [10]. Most recently, the patients had only an echocardiographic assessment. An echocardiographic gradient up to 25 mmHg was regarded as grade I, a gradient up to 55 mmHg as grade II, and a gradient above 55 mmHg as grade III. Fluorescence in situ hybridization (FISH) was available since August 1993 (Institute for Genetics at the LMU Munich, Professor J. Murken, H. Seidel). A total of 23 FISH analyses were done with five proven microdeletions 22q11.

For statistical analysis, the measured values are expressed as mean, SD, range (minimum, maximum) and median values. Comparison of data was performed using an unpaired t-test or chi-square test. The long-term survival and freedom from reintervention are expressed as Kaplan–Meier curves.

2.1. Operative technique

All patients were operated on via median sternotomy, using extracorporeal circulation. After separation of the pulmonary arteries from the common trunk, a continuity from the right ventricle to the pulmonary arteries was accomplished, and eventual concomitant lesions were repaired. In 35 cases, after resection of the pulmonary arteries, the resulting defect in the common trunk was closed, using a Gore-Tex patch in 30 patients and with glutaraldehyde-treated pericardium in five; otherwise this was carried out by direct suture. A patch was used if it was felt that either the truncal valve or the coronary arteries could get distorted. In 61 patients, an enlargement of the ventricular septal defect (VSD) was performed. This was done in cases of truncal malalignment, in order to restore an unrestricted flow from the left ventricle to the aorta, and to avoid postoperative left ventricular outflow tract obstruction. The unusual high number of VSD enlargements is partly explained by an approach which almost exclusively one particular surgeon preferred during the 1970s and 1980s. The VSD was subsequently closed with either Dacron patch material (46 patients) or Gore-Tex (80 patients), by means of interrupted sutures buttressed with felt pledgets. The pulmonary arteries were connected to the right ventricle by an aortic homograft in 56 patients, by a pulmonary homograft in 11, by a Hancock porcine conduit in 35, and by a Carpentier–Edwards pericardial conduit in four patients. The initial conduit size was 9–12 mm in diameter in 67 patients, and larger than 12 mm in 39 patients. The mean conduit size was 13 mm, with a range from 9 to 24 mm.

The mean cross-clamp time was 75 min (SD, 17.8 min; range, 25–135 min; median, 74 min). In 74 patients, circulatory arrest was used (mean, 57 min; SD, 21 min; range, 10–114 min; median, 65 min). The mean bypass time was 124 min (SD, 39.2 min; range, 44–271 min; median, 118 min). During the initial operation, a valvuloplasty of the truncal valve was performed in 19 cases, and one truncus valve was replaced by a homograft. A commissural resuspension was performed in 11 cases, a commissurotomy in six, and in two cases, an annuloplasty was performed, placing single horizontal sutures externally at the sinotubular junction. On inspection, 29 truncal valves were found to be myxomatous thickened; two with two leaflets, and 16 with four leaflets.

3. Results

3.1. Survival and risk factors

Early death was defined as death occurring from the intraoperative period up to 30 days after surgery. Death occurring after that period was defined as late death (Table 1). Between 1976 and 1989 (mean age, 11.6 months;
mean weight, 5.8 kg), the early postoperative mortality was 21%, and between 1990 and 1998 (mean age, 4.9 months; mean weight, 4.5 kg), it dropped to 13%. There were six intraoperative deaths. In two of these, the cause of death was related to elevated pulmonary pressures and consecutive right ventricular failure, in one, a possible coronary flow impairment due to a compression of a conduit, and in three, a distortion of the anomalous coronary arteries was postulated. From the remaining 13 early deaths, among them were pulmonary hypertensive crisis, sepsis, arrhythmia, and sudden bleeding, and an impairment of flow in anomalous coronary arteries was presumably the reason for circulatory failures in another two patients. There were 23 late deaths. Truncal valvar dysfunction led eventually to severe left ventricular impairment, and prolonged ventilation was necessary due to bronchus and/or trachea compression. The clinical state prior to surgery proved to be significantly correlated with mortality (clinical state I/II vs. III/IV; $P = 0.007$). After 1, 10 and 15 years, the overall survival was 66, 61 and 59%, as depicted in Fig. 1. No significant correlation between the two collectives was found regarding survival and weight at operation, age, X-clamp time, cardiac arrest time and bypass time. Severe truncal dysfunction, mainly a combination of stenosis and insufficiency, must also be regarded as a risk factor for surgery ($P = 0.007$), as well as the presence of a coronary anomaly (CA; $P = 0.018$).

3.2. Reoperations

The time related freedom from reoperation is depicted in Fig. 2. A total of 64 reoperations were performed. Reoperation was necessary after a mean time of 4.9 years (SD, 5.0 years; range, 20 days–13.0 years; median, 3.1 years). Five patients were reoperated within 30 days after primary repair, and seven within 3 months. Three had a residual VSD, four had a truncal valvar dysfunction, three had a bronchus and/or trachea obstruction, one had an aneurysm at the site of the right ventricular outflow tract, and two had a complete atrioventricular block. The indication and incidence for reinterventions is depicted in Table 2. The reasons for reintervention were mainly due to outgrowth and failure of the conduit. At the time of first reoperation, 45 conduits had to be replaced (allograft, $n = 41$; Hancock, $n = 4$). The graft had to be replaced after a mean time of 6 years (range, 0.6–13.4 years; median, 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 9.2 years. These findings were of no statistical significance, as the distribution of each conduit type at time of primary correction was too unequal. At reoperation for conduit dysfunction (right ventricle to pulmonary artery), only valved conduits were used (Hancock conduits or homografts). Another four truncal

Table 1
Causes for early and late death

<table>
<thead>
<tr>
<th></th>
<th>Early</th>
<th>Late</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myocardial failure</td>
<td>11&lt;sup&gt;a&lt;/sup&gt;</td>
<td>7&lt;sup&gt;b&lt;/sup&gt;</td>
</tr>
<tr>
<td>Arrhythmia</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>Sepsis</td>
<td>3</td>
<td>6</td>
</tr>
<tr>
<td>Prolonged ventilation (pneumonia)</td>
<td>–</td>
<td>4</td>
</tr>
<tr>
<td>Bleeding</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Sudden death (unknown)</td>
<td>–</td>
<td>3</td>
</tr>
</tbody>
</table>

<sup>a</sup> Of the 11 cases, there were five due to CA, five due to PH and one due to left ventricular heart failure (LVHF).

<sup>b</sup> Of the seven cases, there were three PH and four LVHF.

Table 2
Incidence and indication for reoperations

<table>
<thead>
<tr>
<th>Reoperations</th>
<th>ReOp1</th>
<th>ReOp2</th>
</tr>
</thead>
<tbody>
<tr>
<td>Conduit dysfunction (CD)</td>
<td>31</td>
<td>4</td>
</tr>
<tr>
<td>Residual VSD closure</td>
<td>5</td>
<td>–</td>
</tr>
<tr>
<td>Aortic insufficiency (AI)</td>
<td>–</td>
<td>2</td>
</tr>
<tr>
<td>AI and CD</td>
<td>6</td>
<td>2</td>
</tr>
<tr>
<td>AI and residual VSD closure</td>
<td>4</td>
<td>–</td>
</tr>
<tr>
<td>AI and CD and residual VSD closure</td>
<td>2</td>
<td>–</td>
</tr>
<tr>
<td>Bronchus obstruction (BO)</td>
<td>5</td>
<td>2</td>
</tr>
<tr>
<td>BO and CD</td>
<td>–</td>
<td>1</td>
</tr>
<tr>
<td>Pacemaker implantation</td>
<td>2</td>
<td>–</td>
</tr>
</tbody>
</table>

Fig. 1. Time related survival.

Fig. 2. Freedom from reoperation.
valves were replaced (St. Jude 25, Biocor 21, Hancock 22, Carbomedics 27).

3.3. Follow-up and clinical state of the survivors

The follow-up for 105/106 patients was complete. One patient moved to Iran and no further follow-up was available, so this patient has been excluded from the evaluation. Sixty-four patients survived. The mean follow-up time for all patients was 6 years (median, 4.4 years; 641 patient years), and for the surviving 64 patients, this was 9.1 years (SD, 4.8 years; range, 0.4–16.6 years; median, 10.5 years; 583 patient years). Using the ability index of Perloff [11], 95% of the patients are in functional class 1 or 2, and can participate in normal daily activities; three are in functional class 3, and one is in functional class 4. Forty-seven patients are on no medication; six patients are only on ACE inhibitors, seven on diuretics, and four on a combination of digitals and diuretics. The two patients with mechanical prostheses are on a regimen of warfarin sodium, aiming to maintain the INR between 2.5 and 3.5. Absent or mild truncal valve insufficiency has been diagnosed in 44 patients, being moderate in 16 and severe in two. On X-ray of the chest, the mean cardio-thoracic index is 0.59 (SD, 0.05; range, 0.5–0.72; median, 0.57). The mean Doppler gradient across the conduit in the right ventricular outflow is 31 mmHg (SD, 21 mmHg; range, 5–100 mmHg; median, 25 mmHg). The weight according to age dependent percentiles is below P 10 in 8/62, and length in 12/62 patients. The neurological development is mildly retarded in two, retarded in five and severely retarded in two patients. Of these, one has a VACTERL association, and another has a speculated chromosomal deletion. Two had a prolonged cardiac arrest time of more than 85 min. All five patients with proven microdeletion 22q11 show developmental retardation.

4. Discussion

Although the alphanumeric classification as proposed by Van Praagh [12] of the entity of CAT is widely used, a descriptive approach for a full diagnosis of this complex malformation seems to be more reasonable. Anderson [13] points out, that the entity of CAT can be associated with other malformations (i.e. double aortic arch, categorization of pulmonary arterial pathways), and should be described as such.

First attempts, dealing with the complex congenital malformation of the CAT, date back more than 30 years [14,15]. Reports from Ebert [8], Marceletti [7] and Applebaum [16] favoured repair in infancy, as older surgical candidates were more likely to develop a progressive pulmonary obstructive disease. Neonatal repair has become more common at many centres in the 1980s and 1990s [6,17–20]. Patients with major associated anomalies show an often rapid clinical deterioration, and must be regarded as high risk patients. Repair within the first 6 months of life is aimed at trying to avoid morbidity related to the pulmonary circulation [9]. This tendency towards earlier intervention was also found at our institution in recent years. Following this strategy, the early mortality at our institution was lowered to 13% between 1990 and 1998. Close monitoring of the patients prior to surgery, an increased operative experience, and improved postoperative management in the intensive care unit, have led to a drop in operative risk.

In children without complex associated lesions before the age of 2 months, preoperative diagnosis is performed by echocardiography (catheterization is not mandatory). Recognition of the proximal coronary arterial anatomy is essential, as it must be regarded as a risk factor [21,22]. A high incidence of coronary ostial and arterial abnormalities are well described [23]. There is the potential risk of distortion of the coronary ostia or proximal coronary segments during reconstruction. For unrestricted flow in the coronary arteries, direct suturing of the defect of the common trunk after disconnection of the pulmonary arteries should be avoided. The right ventriculotomy must be placed away from the right coronary artery, or further down, in case of a crossing in front of the truncal root.

Dealing with severe truncal dysfunction remains a challenging surgical dilemma. A moderate truncal valve stenosis may be mostly flow related and less severe postoperatively, and can be addressed by a valvotomy. A moderate truncal valve insufficiency can be addressed by valvuloplasty or annuloplasty, and is usually well tolerated postoperatively after closure of the VSD, and thus, a reduction of volume overload. However, both severe stenosis and severe regurgitation, or a combination of the two, are more complicated to deal with. Truncal valve replacement at the initial repair carries a high risk [9,22]. The choice of surgical technique in these cases is still controversially discussed. McElhinney et al. [24] believe that mechanical prostheses seem to be more favourable for late truncal valve replacement than the use of allografts. Addressing truncal valve dysfunction at a later stage requires close monitoring of the patient.

A microdeletion 22q11 is known to be associated in patients with conotruncal defects. The frequency of microdeletion is 34.5% in patients with CAT, as found by Goldmuntz et al. [25]. FISH analysis was performed in 23 patients with suspected microdeletion since August 1993, and was positive in five patients. All five patients show developmental retardation.

In conclusion, the 10–15 year survival after primary repair is good, and the majority of the children are in good clinical condition. Predictably, conduit replacement and/or truncal valve dysfunction were the main indications for reoperations.

References


Appendix A. Conference discussion

Dr A. Corno (Lausanne, Switzerland): I have two questions. The first is how do you manage the truncal valve regurgitation from a surgical point of view? Do you try to reduce the number of cusps, do you try to do any special plasty of the valve?

Second question. Regarding the bronchial obstruction, it is generally reported as being associated with truncus and aortic arch interruption or the presence of right aortic arch. Do you have the same experience? And if not, how do you prevent or try to prevent the bronchial obstruction?

Dr Schreiber: May I just answer first your second question. In one case, we put a ring in Gore-Tex prosthesis in, in order to avoid that the conduit would get squashed. We didn’t ever try to reduce the valvar ring. To your first question, we just tried to resuspend the valves in order to make the truncal valve less insufficient.

Dr T. Tlaskal (Prague, Czech Republic): Perhaps I have not seen it well, but you did not mention the subset of patients with concomitant interrupted aortic arches. Would you kindly comment on your experience with truncus arteriosus with interrupted aortic arch; that is the first question. Could you comment on the necessity of reintervention for conduit obstruction on the size of conduit which was used during original repair? How did the necessity of reoperation for homograft or conduit obstruction depend on the size of the conduit used?

Dr Schreiber: Well, I can’t exactly answer this question, for the fact that these children did not just only grow out of their conduit which had been placed on the right side, but also, as we all know, due to calcification. So, not only a stenotic component had to be addressed, but also the conduit became insufficient. That is why we had to take the children back to theatre to replace the right-sided conduit.

Dr Tlaskal: If I understand, you were not investigating specifically the risk of redo according to the size of the conduit on primary repair.

Dr Schreiber: No. And if I just may address the first question, we had only seen eight children with a concomitant interrupted aortic arch. As we know from the literature, you should actually see more children with this complex malformation. Most likely, those children just have not had a surgical correction. So we only had eight patients, but it was not statistically significant that those had a worse outcome than the others.

Dr F. Lacour-Gayet (Le Plessis Robinson, France): Your conclusion is focused on coronary anomalies, and you conclude that this is a major risk factor. However, in our experience, real coronary anomalies that have an impact on the reconstruction of truncus arteriosus were exceptional; the main difficulty being that the left coronary ostium can be very close to the left pulmonary branch. I would like you to comment on the definition and the description of coronary anomalies in your paper.

My second comment relates to pulmonary branch stenoses following repair of truncus arteriosus that has really become a difficult problem to manage. Have you met this complication?

Dr Schreiber: We know that in patients with CAT, not only the coronary anomalies have a higher take-off, but also in many cases, the right coronary artery just comes across higher than in other cases or is just a dominant coronary. I assume that as we are looking back 25 years, this problem wasn’t always addressed properly, and in too few cases a patch was used in order to reconstruct the trunk after excision of the pulmonary arteries.

Most recently, almost in all cases, we patched it, in order to avoid a distortion, which could have most likely led into intraoperative problems, long bypass, and finally, to impaired output and cardiac failure. So, we...
believe that one should probably always, even if it doesn’t look like it, put a patch in, in order to restore the shape of the trunk.

**Dr P. Burczynski (Warsaw, Poland):** I would like to ask you about the postoperative period, because some of your patients were, during the operation, older than 6 months of age, and I wonder if you have some observation with pulmonary artery hypertensive crises, and how did you manage, if you have observed it?

**Dr Schreiber:** For the last years, our policy has changed. Patients ought to be operated on earlier in life, knowing that hypertensive crises are more likely to be avoided in children under the age of 100 days. A PA line is an extremely helpful tool in order to manage the patients intraoperatively and postoperatively, as, for instance, the application of nitric oxide can be monitored adequately.

**Dr B. Maruszewski (Warsaw, Poland):** Dr Andreas Urban, a year ago, was responsible for collecting the truncus data for the European Congenital Heart Surgeons’ Foundation, and we would like very much for Dr Urban to comment on that, because this was probably the largest series ever collected.

**Dr A. Urban (St. Augustin, Germany):** I have a comment and two questions. The first comment. Congratulations on your series of the German Heart Centre in Munich. This is a large series of a selected patient population and with a low mortality. The Congenital Heart Surgeons Club had conducted a study in 1998 with 238 patients of 14 centres in 11 countries in Europe. The early mortality was 27% for the time frame between 1990 and 1997. The patients were probably not comparable because there were only infants in the European study, and the interrupted arch was found in 11% of those patients.

In another multicentric study, the Pediatric Cardiac Care Consortium of USA published their series of 49 hospitals, where the early mortality, in the time frame from 1985 to 1994, for primary correction was in the region of 44%. I think your results are a very good achievement.

I like to comment in support of coronary artery anomalies. I think one reason for high operative mortality in truncus is, that surgeons are not aware of coronary artery anomalies. If they would look for them, the incidence of minor and major coronary artery anomalies would rise up to 50%, as in pathological series. In my personal series of 63 truncus arteriosus, we found an incidence of 41% of major and minor coronary artery anomalies.

My first question regarding coronary anomalies is: do you close the truncotomy directly or with a patch when you have a coronary ostium which is arising at an ectopic site, over the commissure?

My second question is: you had an 11% incidence of residual or recurrent VSDs. Why is that, how do you close your VSDs?

**Dr Schreiber:** In all the patients, single buttressed sutures were placed once the residual VSD was identified. The former patch did not have to be removed once. As I have mentioned earlier, we do always intend to patch the trunk. Routinely, we use Gore-Tex, and only in two cases, autologous pericardium was used.

**Dr Maruszewski:** To finish this very interesting topic, may I ask the audience, who of you believes nowadays that truncus repair is a neonatal surgery?

**Audience:** (Show of hands).

**Dr Maruszewski:** Okay, thank you very much.