Survival and reintervention after neonatal repair of truncus arteriosus with valved conduit

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Abstract

Objective: Neonatal primary repair has progressively become the treatment of choice for truncus arteriosus with encouraging survival. However, use of valved conduits to reconstruct the right ventricular outflow tract (RVOT) inevitably induces reintervention. This study estimates survival and rate of catheter-interventional and surgical reinterventions.

Methods: Thirty-five consecutive neonates who underwent truncus repair with 27 homografts and 8 Contegras from 1987 to 2007 were studied. Interrupted aortic arch (IAA) was associated in nine patients. Actuarial survival and freedom from reintervention were evaluated according to Kaplan—Meier method.

Results: Five patients died early after repair. Two died late and one death was related to reintervention. Survival was 91.9% /C6 5.4% from postoperative month 2 onwards when IAA was not associated and 41.7% /C6 17.3% from month 4 in IAA presence. During a median follow-up of 68 months (range 1—180 months), 42 reinterventions (of which 17 reoperations) were performed in 21 patients. Rate of reintervention was 2.6 per early survivor per 10 years. RVOT obstruction constituted the main indication: branch pulmonary arteries often being involved (n = 25). Uncommon indication was subaortic stenosis (n = 3), aortic arch obstruction (n = 2) and truncal valve regurgitation (n = 2). At year 10, freedom from first, second and third reintervention was 87.5% /C6 6.8%, 64.1% /C6 10.2% and 39.5% /C6 10.7% at year 1, 3 and 5, respectively. Sixteen first conduits were explanted. Freedom from first conduit replacement was 87.5% /C6 6.8%, 64.1% /C6 10.2% and 39.5% /C6 10.7% at year 1, 3 and 5, respectively. Homografts enjoyed higher durability than Contegras.

Conclusion: Neonatal repair of truncus arteriosus results in high survival, the only risk being IAA association. The rate of reintervention is heavily influenced by stenosis of branch pulmonary arteries.

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Keywords: Truncus arteriosus; Valved conduits; Catheter intervention; Reoperation

1. Introduction

Truncus arteriosus is a highly lethal lesion with about 50% of those born with this anomaly dying in the first month of life [1]. Therefore neonatal repair should be recommended. To reconstruct the right ventricular outflow tract (RVOT), either valved conduits are implanted or techniques using unvalved conduits or apposition of autologous tissues are applied. With early surgery, over the last two decades, patients’ survival has improved. But overall operative mortality rates continue to be higher than for neonatal correction of many congenital diseases: 15.4% (11.4—20.1%, 95% CI) [2]. Moreover the burden of cardiological and surgical reinterventions becomes an increasing concern as the child grows, thus leading to question the suitability of various methods of surgical management.

In 1987 we started a program of early repair using homografts as valved conduits, which brought excellent results [3]. From October 2002, due to shortage of small-diameter allografts, we progressively resorted to the bovine jugular vein 'Contegra'. This study focuses on outcome after neonatal correction of truncus arteriosus over a 20-year period, with particular emphasis on survival, type and rate of reintervention, catheter or surgery.

2. Patients and methods

2.1. Patients

From June 1987 to December 2007, 98 consecutive children underwent repair of truncus arteriosus in our department. A valved conduit was used for RVOT reconstruction in all cases, except the one. Correction was performed at
the age of 30 days or less in 35 patients. Fig. 1 shows increasing incidence of neonatal truncus repair over time (four periods): from a low 7.7% (1/13) rate in our early experience to a high 65.4% (17/26) from year 2001 on.

Some of these 16 girls and 19 boys have been included in previous studies [3, 4]. At the time of operation, median age was 22 days (range 7—30 days) and median weight, 3.06 kg (range 2.10—4.0 kg). Eight patients weighed less than 2.5 kg. According to the Van Praagh classification, it was a common arterial trunk type A1 in 9 cases, type A2 in 17 and type A4 with interrupted aortic arch (IAA) in 9 neonates. Interruption was between the left common carotid artery and the left subclavian artery in eight cases; three among these had an aberrant right subclavian artery (arteria lusoria). The truncal valve was at least grade II incompetent in 11 patients, 5 presenting with severe regurgitation (grade III—IV). Main associated anomalies present in 30 children are listed in Table 1. Complexity according to comprehensive Aristotle score [5] ranged from 11 to 21 points (median: 14.25), with five patients scoring 20 or more.

### 2.2. Surgical management

Except for the first patient of this series who had aortic arch reconstruction through a left thoracotomy 2 weeks before truncus repair, all other 34 children underwent one-stage complete correction. The technique used in our unit for repair of truncus arteriosus and to prevent pulmonary hypertensive crises has been already described [3]. The operation was performed with deep hypothermic arrest in 18 patients. Table 1 gives mean cardio-pulmonary bypass data. Care was taken to separate the pulmonary arteries from the truncal vessel without injuring the coronary arteries. The ventricular septal defect was closed with a patch usually by means of buttressed interrupted sutures. The right ventricular outflow tract was reconstructed with either cryopreserved homograft \((n = 27)\) or Contegra \((n = 8)\). Choice was dictated by availability of small-diameter allografts. Alternatively, as of October 2002, Contegra was used. The type and size of implanted conduits are displayed in Table 2. Their median z-value was 3.65, range 1.04—5.57. Miyamoto et al. [4] details surgical techniques applied in case of aortic arch interruption with, from 1997, arch reconstruction on the beating heart and under cerebral perfusion. The insufficient (grade III—IV) truncal valve was primarily repaired in three patients. It was not surgically addressed in the two other cases: the first died perioperatively, the second underwent truncal valve replacement by a homograft 2 weeks after initial repair. Commissurotomy and valvotomy were performed to manage the three severe stenotic truncal valves. To facilitate chest re-entry in view of the inevitable reoperation for conduit failure, we routinely approximated the pericardium and covered the heart with an artificial membrane. In 14 cases, the sternum was closed secondarily.

To prevent sudden rises in right ventricular afterload, sedation and paralysis were maintained with a continuous infusion of fentanyl, midazolam and pancuronium in the first 24 postoperative hours. Three patients suffered from pulmonary hypertensive crises. There were six early reoperations for bleeding. The median duration of endotracheal intubation for 29 early survivors was 5 days, range 2—18 days. One patient was transferred under mechanical ventilation to
another hospital where a tracheostomy was performed. He was later (6 months) referred back to us for surgical reintervention, still with open tracheostomy.

2.3. Follow-up. Indication for reintervention

Echo Doppler studies were used to evaluate heart and conduit function at least every 6 months for the first 2 years and once a year thereafter. If a problem was detected or suspected, cardiac catheterization with angiography was performed. Catheter intervention was considered when peak echo Doppler was $\geq 40$ mmHg at any RVOT site. Indication for conduit replacement was grade III/IV regurgitation with dilatation of right ventricle, or RVOT obstruction with right ventricle pressure at least 75% of the systemic pressure. Reintervention was directed to residual cardiovascular lesions as necessary.

2.4. Data analysis

Data were collected retrospectively for the homograft group and as part of a protocol after implantation for the Contegra patients. The actual assessment took place from December 2007 to March 2008. Kaplan–Meier curves for actuarial survival and freedom from intervention were calculated and log-rank test applied (variables comparison) using the GraphPad Prism (San Diego, CA, USA). Patients were censored at the time of death or most recent follow-up. The significance level is set at a $p$ value of $\leq 0.05$. Medians and means are given with range and standard deviation. Early reintervention for secondary chest closure and for bleeding is not included in the study. Early survivors are patients who survived surgical repair at least 30 days.

3. Results

3.1. Survival and functional status

Five patients died early after repair: operative mortality of 14.3% (4.8–30.3%, 95% CI). Four among them had associated IAA and a comprehensive Aristotle score $\geq 20$. The fifth neonate to die succumbed to iterative pulmonary hypertensive crises. Two patients died later. One death was related to interventional dilatation of the distal aortic arch, 4 months after repair of a truncus A4. The other late death (2 months) was due to aspiration. Overall survival was 79.2% $\pm$ 7.0% from month 4 onwards. Survival was significantly higher ($p = 0.0012$) when IAA was not associated (truncus type A1 and A2): 91.9% $\pm$ 5.4% from postoperative month 2, versus 41.7% $\pm$ 17.3% from postoperative month 4 when IAA was present (truncus A4), and when comprehensive Aristotle score was below 20 ($p = 0.0014$). There was no difference in survival according to period of surgery (1987–1997 vs 1998–2007), weight at time of repair ($<2.5$ kg vs $\geq 2.5$ kg) and type of valved conduit implanted.

Mean follow-up for early survivors was 74 $\pm$ 63 months, median: 68, range 1–180 months. At last follow-up, 18 of the 28 late survivors (64%: 18/28) are in the functional NYHA class I, 9 (32%) in class II and the remainder in class III. Echo Doppler shows grade I truncal valve regurgitation in most cases (50%: 14/28) and normal function in 40% (11/28) of patients. It exhibits mild valvular stenosis in two cases (7%: 2/28), and mild stenosis and insufficiency of the homograft, which replaced the truncal valve in the remaining survivor.

3.2. Reintervention

A total of 42 reinterventions was performed in 21 patients. Rate of reintervention was 2.6 per early survivor per 10 years. Table 3 summarizes incidence, timing and kind of catheter ($n = 25$) and surgical ($n = 17$) reinterventions. Indication was in the great majority of cases ($n = 20$) RVOT obstruction. Involved was the site of pulmonary artery branches, or the implanted conduit. Reintervention consisted of dilatation and stent insertion when indicated, or replacement of the failed graft. Multiple procedures were usually performed at surgery (Table 3B). It is to be noted that in three patients, bilateral pulmonary stenosis caused aneurismal graft dilatation, which compressed left main bronchus. This was managed by conduit replacement and anterior translocation of pulmonary arteries (LeCompte maneuver). Other reasons to reintervene were truncal valve insufficiency ($n = 2$), residual septal defects ($n = 3$), subaortic stenosis ($n = 3$) and aortic arch obstruction ($n = 2$). As aforementioned, one patient died after aortic arch dilation. No death occurred after reoperation.

Freedom from first reintervention was 76.2% $\pm$ 8.5%, 49.3% $\pm$ 10.4% and 35.9% $\pm$ 10.1 at year 1, 3 and 5, respectively. At year 10, freedom from first, second and third reintervention (Fig. 2) was 17.9% $\pm$ 8.1%, 46.1% $\pm$ 10.6% and 81.9% $\pm$ 9.5%, respectively. Patients operated upon in period 1997–2007 and those in whom Contegra was implanted had higher incidence of reintervention (Table 4). Freedom from first reintervention according to type of valved conduit used is

<table>
<thead>
<tr>
<th>Procedure Number</th>
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<tbody>
<tr>
<td>LPA/RPA dilatation 19</td>
</tr>
<tr>
<td>With stent implantation 6</td>
</tr>
<tr>
<td>Graft dilatation 6</td>
</tr>
<tr>
<td>+ Stent implantation 1</td>
</tr>
<tr>
<td>Aortic arch dilatation 2</td>
</tr>
<tr>
<td>Graft explantation and replacement 17</td>
</tr>
<tr>
<td>+ LPA/RPA plasty 6</td>
</tr>
<tr>
<td>+ Infundibular resection 3</td>
</tr>
<tr>
<td>+ Subaortic stenosis resection 3</td>
</tr>
<tr>
<td>+ LeCompte maneuver 3</td>
</tr>
<tr>
<td>+ Truncal valve repair or replacement 2</td>
</tr>
<tr>
<td>+ ASD closure 2</td>
</tr>
<tr>
<td>+ VSD closure 1</td>
</tr>
</tbody>
</table>

LPA: left pulmonary artery; RPA: right pulmonary artery.

Table 3

Catheter and surgical reinterventions: incidence and procedures

<table>
<thead>
<tr>
<th>Procedure</th>
<th>First</th>
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<td>2</td>
<td>17</td>
</tr>
<tr>
<td>Total</td>
<td>21</td>
<td>15</td>
<td>6</td>
<td>42</td>
</tr>
</tbody>
</table>

A. Incidence

B. Type of reintervention

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depicted in Fig. 3, the most favorable graft being the aortic homograft with freedom of 30.0% at year 10.

There were 12 instances of first catheter intervention directed to branch pulmonary artery stenosis. Interventional dilatation failed or was not effective in five cases. It allowed postponing surgery (n = 4) and catheter procedure (n = 1) for a median duration of 6 months: range 5—14 months. The two remaining patients do not still require any reintervention after 82 and 89 months.

Freedom from first catheter reintervention decreased over years to 80.2% ± 7.9%, 61.6% ± 10.2% and 51.3% ± 10.8%, respectively at year 1, 3 and 5, to stabilize at 45.6% ± 11.0% from month 78. This freedom was statistically similar (p = 0.37) after repair of truncus type A1 and type A2. But it was a low 33.3% ± 19.2% from month 4 for patients in whom Contegra graft was implanted and much higher (p = 0.0002) for those who received homografts: 94.4% ± 5.4% and 72.2% ± 10.6%, respectively at 1 and 3 years, and 53.5% ± 12.2% from month 78.

Sixteen original grafts were explanted: three Contegas and one pulmonary homograft because of regurgitation and dilatation, and because of stenosis for eight aortic and three pulmonary calcified homografts. They were replaced by 11 Contegas, 3 aortic and 2 pulmonary allografts, size 16—22 mm, median z-value of 4.4, range: 3.1—7.0. Freedom from first conduit explantation was 87.5% ± 6.8%, 64.1% ± 10.2% and 39.5% ± 10.7% at year 1, 3 and 5, respectively. Homografts, aortic in particular (58.3% ± 14.2% at year 5), enjoyed higher durability than Contegas as evidenced in Fig. 4. A second aortic homograft failed and was replaced by a Contegra. So far none of the 11 patients with Contega as second valved conduit needed reoperation during a median interval of 33 months: range 4—56 months. But catheter reintervention for branch pulmonary artery stenosis was necessary on five occasions.

4. Comment

4.1. Preamble

Because of the inherent high mortality, truncus arteriosus warrants early surgery. Delaying repair exposes those patients who present in stable condition to unpredictable
episodes of cardio-pulmonary decompensation and increases the risk of pulmonary hypertensive disease. We are of the opinion that all types of common arterial trunks should be repaired in the neonatal period, unless a specific problem opposes. This study therefore selected newborns from the cohort of 98 patients who underwent truncus repair in our unit, excluding young infants. Results are intended to document what can be expected when neonatal truncus correction is applied, by focusing on two important matters: survival and rate of reintervention and, in that way, to encourage pediatric cardiologists to early refer newborns with truncus arteriosus.

4.2. Discussion

Following the early reports of Hanley et al. and Bove et al. [6,7], neonatal primary repair has progressively become the treatment of choice for truncus arteriosus. In particular, presence of major associated anomalies, such as IAA and severe truncal valve regurgitation dictates diligent surgery regardless of age or weight. This experience (Fig. 1) illustrates increasing awareness of referring cardiologists, so that nowadays the great majority of our patients are operated upon in the first month of life. Early and late survival remained high (91.9% ± 5.4%) all over the period when IAA was not associated, therefore supporting the policy of truncus repair in neonatal period. However concern persists for newborns with truncus arteriosus and IAA. The poor prognosis for this group of patients has been confirmed by a multi-institutional Congenital Heart Surgeons Society study with a 10-year survival of 31% and high risk of reinterventions in survivors [8]. We recently analyzed our unsatisfactory results for truncus A4 according to Aristotle scoring and we concluded that patients with a score under 20 are likely to survive surgical repair [4].

Use of small-diameter homografts, and more recently of the 12 mm Contegra for RVOT reconstruction inevitably leads to reintervention as these conduits do not grow and can prematurely degenerate. In this series graft durability was 64.1% ± 10.2% at year 3, which matches the 66% ± 6% lifespan at the same interval recently reported by the largest single comparative study of allograft and Contegra in infants with truncus arteriosus [9]. Few of these small implants were still functioning after 10 years (Fig. 4), the great majority having been replaced after 5 years. It appears that interventional dilatation could postpone replacement of failing graft only for a short period: usually 6 months. We did not experience better behavior from pulmonary allografts as reported by other authors [9–12]. Contegra durability was lower, but the number of Contegras implanted is too small (n = 8) and the time of follow-up too short (maximum 29 months) to allow definitive conclusions. Nevertheless, results in this study were not as promising as recently reported by us [13] and by the aforementioned multi-institutional study [9]. It is to be noted that these two reports do not supply conduit survival beyond three postoperative years. Longer-term outcome for small grafts are needed. We are currently reviewing the outcome of all 12 and 14 mm Contegras implanted in our unit from October 2002 to December 2007 (n = 37) and preliminary results indicate that their lifespan is limited to 4 years. The fate of second valved conduits, Contegras in particular, should be observed scrupulously as, given their size, they are presumed to last a long time.

This study indicates (Table 3) that conduit failure was not the first reason to reintervene, but stenosis of one or both branch pulmonary arteries. How should RVOT be reconstructed to minimize this problem? In our experience, incidence of RVOT obstruction at this site was similar for the two anatomical A1 and A2 truncus types. Higher occurrence in Contegra bearers would suggest a graft-narrowing effect on adjacent pulmonary arterial tree, even if no fibrous pannus at distal anastomosis was found at surgery in the three patients in whom Contegras were explanted. Noteworthy is the finding from the already cited multi-institutional study [9] that the risks of all-cause intervention and conduit for branch pulmonary stenosis were not different between allograft and Contegra. In this report, transcatheter intervention to pulmonary arteries was performed in 31% (40/107) of patients. This might be compared to the 43% (12/28) cases undergoing first catheter intervention in our study, what would be not significantly different (p = 0.66).

This site of RVOT obstruction was mainly managed by transcatheter balloon dilatation with or without stenting. It was also alleviated at reoperation by patch augmentation (n = 6) and by anterior translocation (n = 3) of the pulmonary arteries. This technique (LeCompte maneuver) is particularly indicated and efficacious when the right pulmonary artery is compressed by a large ascending aorta. It should be considered more often for application as the primary repair in those cases with hypoplastic pulmonary artery branches.

Surgery was rarely undertaken for residual septal defects and truncal valve dysfunction contrary to the findings of McElhinney et al. [10] and Henaine et al. [14]. Rate of reintervention was estimated at 2.6 per early survivor per 10 years. Half of patients have already undergone at least one reintervention at the age of 3 years, and at least two at the age of 10. No literature is available for comparison, as publications either include all ages of patients at time of truncus repair, or mention only reoperations, or embraces a limited follow-up term. But there were 121 reinterventions performed in 81 patients (1.5 reintervention per patient) in long-term period in the series of McElhinney et al. [10], which also includes young infants. There was 1.4 (42/30) reintervention per patient in this study comprising newborns only. Therefore our rate of reintervention should effectively reflect the expected burden of catheter-interventional and surgical procedures after early repair of truncus arteriosus with valved conduits.

Could the rate of reintervention be reduced by reconstructing RVOT without valved conduits? Techniques using unvalved conduits or apposition of autologous tissues were pioneered by Reid et al. [15] and popularized by Barbero-Marcial et al. [16]. Earlier reports acknowledged higher operative mortality than after correction with valved grafts as a consequence of acute right ventricle failure secondary to severe pulmonary regurgitation; but some recent publications [17–19] demonstrate improved early survival, so that the question of reintervention incidence may be asked. In the series of Chen et al. [18], the need for any reintervention was similar between the two groups of patients with either conduit RVOT reconstruction or direct anastomosis. Honjo et al. [19] summarized and compared published literature.
about freedom from reintervention for the two strategies. Freedom in Honjo’s own publication was 50% (19–81%, 95% CI) at 5 years, in direct anastomosis technique. It was 68% ± 14% in Danton et al. series [17]. These figures are to be compared to the 35.9% ± 10.1% rate, at the same interval, in this study. Stenosis of pulmonary arteries was again the most frequent reason to reintervene. Honjo also advocates LeCompte maneuver to tackle this problem.

5. Conclusion

Neonatal primary complete repair of truncus arteriosus with the use of valved conduits in pulmonary position results in high survival, the only risk being a combination of associated aortic arch interruption and a comprehensive Aristotle score of 20 or more points. This study adds data to support the policy of neonatal repair of any type of truncus arteriosus. Lack of graft growth obligatory leads to reintervention and reoperation for conduit failure. Stenosis of the pulmonary arteries constitutes a frequent risk for reintervention in the early and long term. It has to be observed whether similar survival with lower reintervention rate can be achieved with direct anastomosis techniques for RVOT reconstruction.

References