Long-term results after correction of persistent truncus arteriosus in 83 patients

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Abstract

Objective: This study aims to analyse long-term results after correction of truncus arteriosus in all patients operated in one institution over 28 years. Methods: Between 1981 and 2009, 83 patients, median age 54 days, underwent repair of truncus arteriosus. Interrupted aortic arch was present in 14 (17%), severe truncal valve insufficiency in 10 (12%) and non-confluent pulmonary arteries in five (6%) patients. Repair with reconstruction of the right ventricular to pulmonary artery continuity was performed using a valved conduit in 80, and other methods in three patients. At the same time, correction of interrupted aortic arch was done in 14 and truncal valve repair in eight patients. Survivors were repeatedly examined echocardiographically for assessment of residual heart lesions. Results: The early mortality was 19 (23%). Out of 35 patients operated between 1981 and 1996, 17 (46%) died, and out of 48 patients operated between 1997 and 2009, two (4%) died. Operation before 1997 (p = 0.001) and aortic cross-clamping time > 90 min (p = 0.009) were found to be risk factors of death. Eight (10%) patients died late, a median of 68 days after surgery. Fifty-seven (69%) patients were followed for 10.9 years. Three (4%) patients were lost. Twenty-five (30%) patients are alive with their original conduit 7.5 ± 5.2 years after correction. Twenty-eight patients required 41 re-operations for conduit dysfunction with first replacement at mean 5.8 ± 4.1 (range 0.1–14.1 years) years after correction. Nine (11%) patients required 12 truncal valve replacements. Eleven (13%) patients required balloon dilatation or stent for conduit obstruction, pulmonary branch stenosis, aortic arch obstruction or stenosis of vena cava. Recent clinical examination was undertaken in 53 (64%) patients and 49 (59% or 77% of early survivors) are in good/very good condition. Conclusions: Truncus arteriosus remains a challenging heart disease. With growing experience, the early mortality decreased to 4%, but numerous re-interventions for conduit obstruction, pulmonary branch stenosis and truncal valve insufficiency are required. Surgery before 1997 and prolonged cross-clamping were risk factors of death. Pulmonary homografts had the best re-intervention-free survival. Statistically, however, the difference between conduits was not significant. Dysplastic valve and truncal valve insufficiency represent risk factors presenting the need for truncal valve replacement.

Keywords: Congenital heart disease; Persistent truncus arteriosus; Surgery; Long-term results; Re-interventions

1. Introduction

Persistent truncus arteriosus (PTA) represents a rare complex heart disease occurring in less than 3% of all congenital heart diseases [1]. Anatomically, it is characterised by a single arterial trunk arising from the base of the heart supplying coronary arteries, aorta and pulmonary arteries. PTA is associated with ventricular septal defect (VSD). Commonly, other associated heart lesions are also present and interrupted aortic arch (IAA) and truncal valve insufficiency (TRI) are of exceptional clinical and surgical importance [1,2]. The first correction of PTA was performed in 1962 using a non-valved polytetrafluoroethylene (PTFE) conduit [3]. McGoon and colleagues performed the first correction with the use of a valved conduit in 1967 [4]. Initially, the repair of PTA was joined with high mortality [5]. Ebert pioneered correction of PTA during the first 6 months of life and this approach led to improved survival [6]. Introduction of new management protocols oriented towards prenatal echocardiographic diagnosis, non-invasive examination, neonatal repair, introduction of safer methods of perfusion, heart protection and postoperative intensive care resulted in considerable decrease of mortality and improvement of long-term results [2,6–8]. This study analyses mid- and long-term results after correction of PTA in all patients operated on in one institution.

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2. Material and methods

2.1. Data collection and statistics

For this retrospective study, all hospital and outpatient department records were studied. Eight patients with PTA and IAA were reported earlier [9]. The data collection was not complete in three (4%) patients who were lost during follow-up. Early mortality was the mortality during 30 days after surgery and mortality after this time was considered to be late mortality. Variables are expressed as median and/or mean ± standard deviation, as appropriate, and a range of values. Statistical univariate analysis of risk factors for mortality was performed using Fisher’s test (with an application of odds correction) or \( \chi^2 \). Probability of survival and re-operation-free survival were analysed using SigmaStat software, GraphPad InStat (1990 GraphPad Software version 1.12), and Microsoft Office Excel 2003, and expressed using Kaplan–Meier estimation.

2.2. Patients

Between 1981 and 2009, a total of 83 consecutive patients, median age 54 days (range 1 day—15.5 years), underwent repair of PTA in Kardiocentrum, University Hospital Motol, Prague. During the same time PTA was not corrected in any other centre in the Czech Republic. In our database, we found data concerning 23 other patients who were examined in Kardiocentrum and who were found inoperable. Before surgery, parents had to sign an informed consent confirming that they had been informed in detail regarding the risk of the surgery and the need for the use of a valved conduit with its specific risks. In our series, there were 25 (30%) neonates and 37 (45%) infants less than 90 days old. The weight ranged between 1.7 kg and 37.0 kg (median 3.5 kg). According to Collett and Edwards’ classification, PTA was of type I in 45 (54%) and II in 33 (40%) patients. In five (6%) patients, non-confluent pulmonary arteries were found. TRI was present in 58 (70%) patients: it was severe in 10 (12%), moderate in 21 (25%) and mild in 27 (33%) patients. Fourteen (17%) patients had IAA, which was of type A in seven, and type B in seven other patients. Anomalies of coronary arteries were seen in 18 (22%) patients (origin of the left anterior descending artery from the right coronary artery in four, high origin of the left coronary artery in one and single coronary artery in one patient). A number of other cardiovascular malformations were diagnosed: right aortic arch in 19 (23%), left superior cava in six (7%), truncal valve stenosis in four (5%), right pulmonary artery stenosis in four (5%), aberrant right subclavian artery in four (5%), agenesis of the left pulmonary artery with left lung hypoplasia, mitral stenosis, tricuspid stenosis, partial anomalous pulmonary venous connection, unroofed coronary sinus and hypoplastic aortic arch in one (1%) patient each. Severe associated non-heart lesions were found in four (5%) patients: two patients required surgery for oesophageal atresia before repair of PTA, one patient had diaphragmatic hernia and one had choanal atresia. In 16 (19%) patients DiGeorge syndrome was identified. A total of 13 (16%) patients were in severe clinical condition and required intubation with mechanical ventilation prior to surgery.

2.3. Surgical procedure

The surgical procedure for repair of PTA evolved in the course of 28 years and our contemporary approach has already been described [9]. The surgery was performed in cardiopulmonary bypass (CPB) and deep or moderate hypothermia, depending on era and complexity of the surgical procedure. All patients with PTA and IAA were primarily corrected but the method of perfusion and brain protection evolved from deep hypothermic circulatory arrest (six patients) to isolated cerebral perfusion (seven patients). Surgery of PTA repair consisted of VSD closure with a Dacron or pericardial (pre-treated with a glutaraldehyde solution) patch, excision of pulmonary arteries, reconstruction of the aorta and reconstruction of the pulmonary artery using a valved conduit, non-valved conduit or direct anastomosis with a monocuspid patch. For construction of the monocuspid valve, a patch cut from a pulmonary homograft containing one cusp was used in one patient and a native pericardium in the other. In Table 1 the list of conduits, used in our patients, is reported. Aortic and pulmonary homografts represented the most commonly used conduits. If a small homograft was not available, a bicuspid pulmonary homograft was constructed from an adult-sized homograft. The diameter of the conduit was 13.6 ± 2.7 mm on average. Non-confluent pulmonary arteries were connected together by direct anastomosis in three and using a PTFE tube (W.L. Gore & Associates, Inc., Naperville, MN, USA) in one patient. In one patient in whom no left pulmonary artery was found, the conduit was connected only to the right pulmonary artery. In

<table>
<thead>
<tr>
<th>Method/type of conduit (manufacturer)</th>
<th>Diameter (mm)</th>
<th>n</th>
<th>%</th>
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<tbody>
<tr>
<td>Aortic homograft</td>
<td>12–25</td>
<td>28</td>
<td>34</td>
</tr>
<tr>
<td>Pulmonary homograft</td>
<td>8–25</td>
<td>16</td>
<td>19</td>
</tr>
<tr>
<td>Bicuspid pulmonary homograft</td>
<td>13–17</td>
<td>15</td>
<td>18</td>
</tr>
<tr>
<td>Hancock bioprosthetic valved conduit — Dacron tube, porcine nonstented aortic valve (Medtronic, Inc., Minneapolis, MN, United States)</td>
<td>12–22</td>
<td>12</td>
<td>15</td>
</tr>
<tr>
<td>Carpentier—Edwards conduit — Dacron tube, pericardial valve (Edwards Lifesciences, Irvine, CA, United States)</td>
<td>12–20</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>Shelhigh pulmonic valve conduit no-react treated — conduits bearing porcine pulmonic valves (Shelhigh Medical Devices, Union, NJ, United States)</td>
<td>11</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>Polystan valved conduit (Polystan, Vaelrose, Denmark)</td>
<td>16</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Contegra pulmonary valved conduit — bovine jugular vein (Medtronic, Inc., Minneapolis, MN, United States)</td>
<td>12</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Non-valved PTFE conduit (W.L. Gore &amp; Associates, Inc. Naperville, MN, United States)</td>
<td>12</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Monocuspid transannular patch</td>
<td>NA</td>
<td>2</td>
<td>2</td>
</tr>
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8 out of 10 patients with severe TRI, valvuloplasty was performed. In individual patients, valve repair consisted of a combination of different methods: commissuroplasty was performed in four patients, complete suture of one commissure in four, partial suture of a wide commissure in two, partial resection of the wall of one sinus in two, commissurotomy in one and excision of fibrous nodes in one patient. In three patients, the wall of a large truncus above the valve was partially excised. Mean times of CPB and aortic cross-clamping were 171 ± 49 min and 70 ± 30 min, respectively. In 37 (45%) patients, the chest was left opened and it was closed at median 3 days (range 1–23 days) after surgery. In one patient with signs of severe low cardiac output syndrome, conventional CPB was restarted and continued for 4 h for myocardial recovery. In no patient, however, were extracorporeal membrane oxygenation or left ventricular assist device used.

2.4. Follow-up

After discharge from the hospital, all patients were repeatedly examined by a paediatric cardiologist in our centre using two-dimensional echocardiographic examination, flow velocity measurement with Doppler and colour-flow mapping. Ventricular function, the truncal and the pulmonary valve anatomy and function, degree of insufficiency, residual defects, flow characteristics across the aorta and the conduit and pulmonary arteries were assessed. Gradients showing pressure in the right ventricle of more than 70% of the systemic pressure usually represented an indication for re-operation. Recent clinical and echocardiographic examinations were obtained in 53 patients.

3. Results

3.1. Mortality

The early mortality rate in our series was 19 (23%). The deaths occurred at mean 4.1 ± 7.0 days (median 1 day, range 0–28 days). The causes of early death were heart failure (11 patients), multiple organ failure (three patients), sepsis (two patients), pulmonary hypertensive crisis (two patients) and sudden death (one patient). Out of 35 patients operated between 1981 and 1996, 17 (46%) patients died, but out of 48 patients operated between 1997 and 2008, only two patients (4%) died (p = 0.001). In the whole series, the early mortality was higher in patients with IAA (36% vs 20%) but it decreased to 13%. Out of three older patients with previous pulmonary artery banding, two died. However, the early mortality was not increased in patients with severe TRI and also in patients with non-confluent pulmonary arteries. Both patients died after previous surgery for oesophageal atresia. By a univariate analysis of risk factors, longer duration of aortic cross-clamping was detected as the second-most important risk factor of death. Out of 20 patients with aortic cross-clamp time >90 min, 11 (55%) died, and out of 63 patients with aortic cross-clamp time <90 min, eight patients died (13%) (p = 0.009). The mortality was also higher in patients who had a larger conduit implanted. The mortality was 33% when the diameter was larger than 13 mm, but it was 14% when the conduit was in the range 8–13 mm (p = 0.096). As for the type of valved conduit, the mortality was lowest when a pulmonary homograft was used (13%). In the group of patients with aortic homografts, the mortality was 39% and in the group with xenografts, it was 38%. The difference was not statistically important.

Eight (10%) patients died late, median 68 days after surgery (range 33–692 days). All deaths but one occurred during the first year after surgery. The most common cause of late death was heart failure in three patients. Two patients died from sepsis and two died from respiratory failure. There was no late death between early survivors after repair of PTA with IAA. There were two late deaths between patients with severe TRI and one late death between patients with non-confluent pulmonary arteries.

3.2. Re-operations for conduit dysfunction

A total of 57 (69%) patients were followed up for 10.9 ± 6.7 years (median 10.6 years, range 0.3–23.9 years). During the follow-up, 30 (36%) patients required a total of 53 re-operations (Table 2) and 11 (13%) patients required 13 catheter interventions (Table 3). Conduit dysfunction represented the most common indication for re-operation. A total of 41 conduit replacements were performed in 28 (34%) patients, in whom one to three conduit replacements were done. The first re-operation for conduit dysfunction was performed at mean 5.8 ± 4.1 years (range 0.1–14.1 years) from the first operation. The second re-operation was required in nine patients at mean 6.6 ± 3.6 years (range 0.9–13.4 years) after the first re-operation and the third re-operation in four patients at mean 3.7 ± 1.9 years (median 4.0, range 0.7–6.4 years) from the second re-operation. Out of the 25 patients who are alive with their original conduit, eleven were followed up for more than 10 years. Fig. 1 compares the actuarial survival without conduit replacement in patients with aortic homografts, pulmonary homografts and xenografts. Concomitantly with conduit replacement, the truncal valve was replaced seven times in five patients.

3.3. Re-operations for TRI

Fourteen re-operations were required in 10 patients with TRI at mean 4.7 ± 4.1 years (range 6 days–14.1 years) after repair of PTA. In two small infants, after unsuccessful truncal valve repair, replacement of the truncal root with an aortic
homograft was required 6 days and 11 days, respectively, after correction. Both children died in the course of 2 months after root replacement— from heart failure the first and from aspiration the second. In all seven older children, mechanical bileaflet valves St. Jude Medical (St. Jude Medical, Inc., St. Paul, MN, USA) or Carbomedics (Carbomedics, Sorin Group, Saluggia, Italy) were used for truncal valve replacement.

3.4. Catheter interventions

Eleven patients required a total of 13 catheter interventions and, in some of them, several procedures were performed at the same time (Table 3). Balloon dilatation for a conduit or pulmonary branch stenosis was indicated most often. It was successful to relieve localised conduit and pulmonary branch stenosis. The duration of improvement after balloon dilatation was usually limited to several months or few years and a second catheter or surgical re-intervention was usually required.

3.5. Follow-up and last examination

A total of 57 (69%) patients were followed up for 10.9 ± 6.7 years (median 10.6, range 0.3—25.9 years). The actuarial survival of patients after repair of PTA using a Kaplan—Meier estimation is graphically demonstrated in Fig. 2. During the follow-up, only 26 (31% of operated patients) patients remain without re-operation. Recent clinical examination was available in 53 patients (64% of operated patients): 24 (45%) were in New York Heart Association (NYHA) class I, 25 (46%) in class II and five (9%) in class III. At time of the last examination, nine patients had truncal valve replaced, seven patients had TRI grade 3 and 15 patients had TRI grade 2. Doppler pressure gradient across the conduit of more than 75 mmHg was detected in three and a gradient between 50 and 75 mmHg was found in 11 patients. Pulmonary insufficiency grade 3 was found in 12 patients. Twenty-two patients are without medication, 22 are on digitalis and nine patients with mechanical valves are on warfarin. Severe neurological deficit was found in two patients and mild-to-moderate psychomotor retardation in seven patients. DiGeorge syndrome was proved in five of them. In two patients, the neurological deficit could be attributed in part to a complicated postoperative period with prolonged low cardiac output. In none of them, however, could it be attributed to deep hypothermic circulatory arrest or other perfusion or surgical method per se.

4. Discussion

The first successful correction of PTA using an aortic homograft was performed in 1967 [4]. Initially, the surgery was joined with a high mortality rate [5]. Nowadays, however, in some centres the early mortality has decreased to 5—10% [8,10]. Presence of severe TRI, coronary artery anomalies and IAA were determined as the main risk factors.
for death [2]. Different surgical tactics and techniques were used for reconstruction of the right ventricular–pulmonary artery continuity, improvement of the truncal valve function and aortic arch reconstruction [7–12]. In the literature, there are not many long-term follow-up studies after PTA repair in the current era [11–16]. Our analysis has shown high early mortality during the initial 15-year period, which was influenced especially by learning curve, late indication for surgery and bad preoperative condition of patients which parallel the experience from other centres [14–16]. The early mortality in our series decreased from 46% to 4%, and we identified surgery before 1997 (p = 0.001) and aortic cross-clamp time >90 min (p = 0.009) to be risk factors of death in this series. We were not able to prove any influence of age, body weight, length of CPB and type of a conduit, to mortality and postoperative morbidity. In patients with IAA, the early mortality decreased from 66% to 13%. Sano and Jahangiri reported larger series of patients with PTA and IAA in which the repair was done without mortality but, in most studies, the mortality rate in this subset of patients with PTA is still high [10,17,18].

Late deaths in our series occurred in eight patients and all but one died in course of the first year after repair. The majority of patients who died had haemodynamically significant residual lesions or complications, which were observed in other series, too [13]. In one series 50% of late deaths were related to re-intervention [19]. In contrast to this experience, no patient in our series died in association with re-operation.

Low cardiac output syndrome and heart failure are the most common causes of early and late death after repair of PTA. The right ventricular function may be unfavourably influenced by pulmonary hypertension, development of conduit obstruction and pulmonary branch stenosis. The left ventricular function deteriorates with progression of TRI and in presence of significant residual lesions [1,13,19]. The function of both ventricles may be also unfavourably influenced by longer ischaemia, inadequate myocardial protection and by injury or compression of coronary arteries with a conduit or at re-operation. The risk of coronary artery injury is increased, especially in presence of coronary artery anomaly [2].

Selection of the optimal method of reconstruction of the right ventricle–pulmonary artery continuity is considered to be one of the key problems of PTA repair because it may basically influence the long-term outcome, frequency of reoperations and the quality of life [8,11,12,20]. A number of different valved conduits were used, but the pulmonary homograft remains the conduit of choice because of its good function and prolonged durability [13–16,20–22]. Small homografts less than 12 mm in diameter are joined with an early occurrence of obstruction and need for replacement at 3 years after implantation, but sometimes earlier [21,22]. Several studies identified young recipient age, small conduit size and pulmonary hypertension to be risk factors for homograft failure [12,13]. This is in accordance with our experience. Pulmonary and aortic homografts or xenografts less than 12 mm in diameter were used in 11 patients, and six (55%) of them required conduit replacement or angioplasty 0.5–3 years after the repair. This is a much shorter interval in comparison with an average time to the first conduit replacement in the whole group, which was 5.8 ± 4.1 years. However, the use of small pulmonary homografts is limited by the lack of donors in this age category. Aortic homografts represent another option though their durability is usually shorter due to earlier degeneration. Bicuspidal adult-size pulmonary homografts proved to be a suitable alternative to small pulmonary homografts, though their long-term function is usually suboptimal and there is a tendency to conduit dilatation joined with pulmonary insufficiency. Pulmonary branch stenosis may also develop and progress with time. Therefore, bicuspid pulmonary homografts are not suitable in patients with very small or distorted pulmonary branches and in patients with pulmonary hypertension, cardiomegaly or extreme dilatation of the truncal root. The pulmonary homograft suitable for construction of a bicuspid conduit for a neonate must not be too large — not more than 18–22 mm in diameter. Below the conduit valve, a hood made from a glutaraldehyde-treated pericardium or PTFE patch should be constructed to prevent valve distortion.

In two of our patients, reconstruction of the right ventricular–pulmonary artery continuity by direct anastomosis with a ‘transannular’ patch and monocuspid valve was performed. The long-term result is favourable without the need for re-operation at 9 and 5 years, respectively, after implantation, though one of them has a well-tolerated grade 3 pulmonary insufficiency. This experience is very limited but promising as a useful alternative for patients with PTA with good pulmonary arteries and without TRI. Relatively large and promising experience with direct anastomosis of pulmonary arteries was presented by Danton and colleagues from Birmingham [12]. According to some authors, there is, however, an increased risk of life-threatening complications after direct implantation of pulmonary arteries in patients with suboptimal anatomy or pulmonary hypertension [12,20]. The general use of direct anastomosis between pulmonary arteries and the right ventricle remains therefore controversial and in the majority of patients, it is recommended to use a valved conduit [8,10,14,20].

Homografts or Hancock bioprosthetic valved conduits were most often used for conduit replacement, and this policy is similar in other centres, too. Theoretically, the second or the third conduit should have a prolonged durability. Peripheral pulmonary stenoses, increased pulmonary resistance, severe TRI with cardiomegaly and/or truncal root dilatation may cause conduit dilatation, valve distortion and/or conduit and pulmonary branch compression. In three of our patients after repair of PTA with IAA, the enlarged root of the truncus compressed the conduit and lead to an accelerated conduit obstruction. Under these circumstances, it is important to resolve surgically eventual dysfunction of the truncal valve by valvuloplasty or valve replacement, and at the same time to perform a plastic reconstruction of the truncus. It is probably wise to use a Hancock conduit in these patients instead of a homograft, because the risk of conduit compression and valve distortion is not so high. Balloon dilatation of an obstructed conduit or pulmonary branches proved to be a useful method, which may prolong the interval between conduit replacements [12,20]. However, in some instances of calcified or severely scarred conduit obstructions, balloon dilatation is not effective [19,20,21].
The function of the truncal valve represents one of the most important factors influencing the long-term outcome after repair of PTA and, in many studies, severe TRI was found to be a risk factor of mortality [2,5,12,15,16]. Massive insufficiency may lead to acute heart failure, chronic heart failure and/or accelerated obstruction of the conduit which may be compressed by the dilated heart or the truncus itself. Henaine and colleagues proved that presence of moderate or severe TRI at the time of PTA repair represented an important risk factor for TRI progression and need for valve replacement in the future [23]. Dysplastic truncal valve, severe TRI at the time of PTA repair, association with IAA and defect of the fibrous tissue formation may unfavourably influence function of the truncal valve but, in our experience, the function of the truncal valve has a tendency to deteriorate with time in nearly all patients with PTA [2,5,12,15,16,23]. In our study, new occurrence or progression of TRI was observed in 44% of all operated patients, and even in 38% of patients who did not have any regurgitation at the time of repair. Intervention on a severely malformed and incompetent truncal valve is therefore mandatory. The timing and the most appropriate type of procedure remains, however, controversial.

Different methods of truncal valve reconstruction have been recommended [10,24,25]. Though, in several centres, valvuloplasty and valve remodelling were successfully applied [10,24,25], our experience as well as the experience of others is more pessimistic [23]. The anatomy of the truncal valve is very variable and often the valve is quadricuspid, dysplastic and potentially stenotic. In 8 of the 10 patients with severe TRI in our series, valvuloplasty was performed at the time of PTA repair and, in the other two patients, it was done at re-operation. Though, in most of the patients, the immediate result was favourable, the effect of valvuloplasty was always temporary. We do recommend, therefore, truncal valve plasty at the time of PTA repair only in patients with severe insufficiency. Persistent insufficiency is usually well tolerated and we would follow-up the patient and try to do a plasty or to replace the truncal valve with a mechanical valve at the time of conduit replacement. If it was not possible to use a mechanical valve, the valve replacement should be postponed. The truncal valve replacement with an aortic homograft should be considered critically because the risk of early or late death after valve replacement with a homograft is higher [19,20,21].

Actuarial probability of survival after repair of PTA in our series is 75% at 2 years, 75% at 10 years, and 75% at 20 years, respectively, after the repair. A recent clinical examination was available in 53 patients. Only about 30% of patients remain without re-operation 10 years after PTA repair. Today, more than 90% of patients are in good or very good clinical condition, but the proportion of patients in NYHA class I decreases with time from surgery. More than 50% of patients have significant residual lesions. Some residual lesions occur in combinations and aggravate the haemodynamics and the clinical picture. The long-term results after repair of PTA depend on the era of surgery, patient’s age, associated heart lesions, evolution of truncal valve function and the type of conduit used for pulmonary reconstruction. Though good mid- to long-term survival rate can be reached with an optimal management protocol, the long-term outcomes are far from ideal. They are unfavourably influenced by numerous re-interventions and risk of severe complications. Further research will be therefore required so as to improve the long-term survival and the quality of life of patients with PTA by prolonging the durability of a valved conduit and improving the methods of reconstruction of the truncal valve.

5. Conclusions

Primary repair of PTA can be done with a relatively low mortality and acceptable long-term results. Surgery before 1997, cross-clamp time longer than 90 min and presence of IAA were diagnosed as risk factors of death. The main long-term problems after repair of this lesion are associated with the development of conduit obstruction and progression of TRI. The use of pulmonary homografts probably has the best long-term results with a prolonged re-intervention-free survival. The superiority of pulmonary homografts, however, was not confirmed statistically. The direct anastomosis of the pulmonary artery to the right ventricle with a monocuspid valve represents a promising method, which requires further studies. TRI has a tendency to progression and truncal valve replacement is required in the majority of patients who originally had severe TRI, dysplastic valve and/or IAA.

References

Appendix A. Conference discussion

Dr E. Belli (Paris, France): The dramatic improvement of your results is in concordance with congenital Database curves concerning the outcome in newborns who underwent the procedure.

Your study addresses all specific angles of truncus arteriosus and the usual associated lesions, and your conclusions are in agreement with that shared with other recently published series.

Let me just emphasise that it would probably be more accurate to state both early, or 30-day, and in-hospital mortality distinctly.

I have two questions. If the clinical status gives you the opportunity to wait, what is, in your opinion, the ideal age for repairing a truncus arteriosus without any associated lesions?

And the second, you did not mention to observe any bronchial compression following truncus and interrupted aortic arch repair. We and others did. Can you comment on the technical precautions to avoid this complication?

Dr Tlaskal: For the first question, today the optimal age at time of surgery for a patient with truncus arteriosus would be about 1 month. I would not do the surgery within the first several days, though we operated on one neonate with truncus arteriosus with interrupted aortic arch on the first day after birth. This child is doing well 11 years after surgery. In simple truncus I would prefer to wait until about 4 weeks of age. The neonates with interrupted aortic arch are, however, operated at the age of 7 days on average.

As for the second question, we have seen 3 patients in whom there were signs of bronchial compression. In one of them severe compression of the left bronchus and the right pulmonary artery was diagnosed. We reported our experience with this patient about 10 years ago. In this patient with truncus and interrupted aortic arch we had to prolong the ascending aorta so as to make wider the window where the left bronchus passes.

In another patient aortopexy was required about 2 weeks after the repair. In a third patient signs of bronchial compression were mild and the pathology did not require surgical intervention. So in general, we did not see many problems with bronchial compression.