
Patients and results. Group A: Mean age and weight were 1.4±1.3 months and 3.5±0.4 kg. The aortic arch obstruction included: hypoplasia (5/11), interruption (4/11) and discrete coarctation (2/11). Seven patients had a one-stage correction, and four had initial arch repair followed by arterial switch operation. There were no hospital deaths (CL 0–28%). Over a follow-up of 638 patient-months (mean 64±39), there have been no late deaths, and all patients are in New York Heart Association class 1. There have been three cases of recurrent aortic arch obstruction (two requiring reoperation, and one requiring balloon dilation). One patient has been reoperated upon for right ventricular outflow tract obstruction. The actuarial survival and freedom from reoperation rates at 6 years were 100% (CL=66–100%) and 72.9% (CL=38–92%) respectively. Group B: Mean age and weight were 5.9±8.4 months and 5±2.1 kg. All patients had a one-stage operation. There were two early deaths (11.8%, CL=1–36%) and one late death over a follow-up of 678 patient-months (mean 52±31). All survivors are in New York Heart Association class 1 and there have been no reoperations. The actuarial survival and freedom from reoperation rates at 6 years were 81% (CL=56–93%) and 100% (CL=76–100%) respectively.

Conclusions. 1. Aortic arch obstruction has not adversely affected early or late survival (P>0.05) or late functional class. 2. Patients with Taussig-Bing anomaly and aortic arch obstruction may have a higher reoperation rate than those with normal arch anatomy. 3. Taussing-Bing anomaly, with or without aortic arch obstruction, can be repaired with arterial switch operation during the neonatal period with good outcome.

Key words  Cardiac surgery · Congenital heart disease · Taussig-Bing anomaly · Aortic arch obstruction · Arterial switch
Introduction

The Taussig-Bing anomaly (TBA) is a well-known anatomic entity broadly defined as a double-outlet right ventricle (DORV) with subpulmonary ventricular septal defect (VSD) and malalignment of the outlet and main interventricular septa [6, 14, 24, 26]. The septal malalignment may cause aortic arch anomalies due to preferential flow pattern from the left ventricle (LV) to the pulmonary artery [19, 23]. Consequently, aortic arch obstruction (AAO) is a commonly associated feature of TBA [16, 20, 27].

Multivariate analysis of all DORV repairs performed at the Royal Children's Hospital (RCH) suggested that arch obstruction was a risk factor for hospital death, but subpulmonary VSD was not (Table 1). The hypothesis that the coexistence of subpulmonary VSD and AAO could be an incremental risk factor for surgical repair is analysed herein. Our management strategy for TBA during the period of this study has been closure of the VSD (LV to neo-aorta) [12]. In the presence of AAO, both one-stage and two-stage procedures have been used.

Patients and methods

Patient population

Between May 1983 and February 1995, 28 patients with TBA had repair consisting of ASO and baffling of the LV to neo-aorta. Patients were analysed according to the presence or absence of AAO. One patient with TBA and AAO had repair of the aortic arch plus atrial septectomy early in this series, and did not survive to undergo an ASO.

Group A

This group included 11 patients (39%), with TBA and associated AAO. The diagnosis was established with cardiac catheterization (n=3), two-dimensional echocardiography (n=5) or both (n=3). A balloon atrial septostomy was performed in six patients and four required prostaglandin E1 infusion to improve perfusion of the ductus arteriosus. The degree of AAO ranged from localized isthmic coarctation to coarctation plus hypoplastic transverse arch or complete interruption. In our institution, and for this study, a transverse arch diameter (echo or catheter estimate in millimetres) less than the baby's weight (in kg) + 1 is considered to be indicative of hypoplasia. The great vessels were in anterior-posterior relationship (8/11) or side-by-side (3/11). The coronary artery distribution was one left circumflex, two right in 3/11 patients, one left, two right circumflex in 4/11 patients and one left-right, two circumflex in 2/11 patients. The VSD location was subpulmonary in all cases, with septal malalignment. One patient had an additional muscular VSD. Other associated anomalies included: subaortic stenosis (8/11 patients), straddling of the mitral valve (1/11 patients) and moderate right ventricular hypoplasia (1/11 patients). The incidence of subaortic stenosis was similar in patients with or without AAO (P>0.05) (Table 2).

Surgical treatment. Our ASO for TBA was similar to that used in our institution for other neonates with transposition of the great arteries [11–13]. We generally employed bicaval cannulation and hypothermic full flow (150 ml/kg per min) cardiopulmonary bypass at 22°C. Alphastat pH strategy was employed during cooling. For one stage procedures, the arch repair was performed using a period of circulatory arrest at 18°C. We tended to employ the Lecompte manoeuvre earlier in this series, and did not survive to undergo an ASO.

Table 1 Multivariate analysis of the outcome for 193 DORV repairs at the Royal Children’s Hospital. Arch obstruction was a risk factor for operative mortality, but the presence of a subpulmonic VSD was not.

<table>
<thead>
<tr>
<th>Variable</th>
<th>Odds ratio</th>
<th>P</th>
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<tbody>
<tr>
<td>Subpulm VSD</td>
<td>0.48</td>
<td>ns</td>
</tr>
<tr>
<td>Uncommitted VSD</td>
<td>0.85</td>
<td>ns</td>
</tr>
<tr>
<td>Multiple VSD</td>
<td>5.6</td>
<td>0.024</td>
</tr>
<tr>
<td>Coarctation</td>
<td>4.7</td>
<td>0.041</td>
</tr>
<tr>
<td>AVSD</td>
<td>0.82</td>
<td>ns</td>
</tr>
<tr>
<td>Hypoplastic LV/RV</td>
<td>5.1</td>
<td>0.052</td>
</tr>
<tr>
<td>Year (pre 1985)</td>
<td>0.466</td>
<td>0.006</td>
</tr>
<tr>
<td>Age (&lt;1 month)</td>
<td>0.57</td>
<td>0.008</td>
</tr>
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</table>

Table 2 Age and weight distribution for infants with TBA+AAO. The patients with AAO were operated earlier in life, but did not have a higher prevalence of subaortic stenosis (SAS) at the time of surgery.

<table>
<thead>
<tr>
<th>Group A (AAO)</th>
<th>Group B (no AAO)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>24</td>
<td>79</td>
</tr>
<tr>
<td>Weight</td>
<td>3.4</td>
<td>4.5</td>
</tr>
<tr>
<td>SAS</td>
<td>8/10</td>
<td>7/17</td>
</tr>
</tbody>
</table>

AAO = aortic arch obstruction; ASO = arterial switch operation; LV = left ventricle; LV/RV = left/right ventricle; SAS = subaortic stenosis; VSD = ventricular septal defect.
The mean aortic cross-clamp time was 86±10 min and the mean period of cardiopulmonary bypass was 158±39 min. In eight patients, circulatory arrest was necessary, its mean duration being 33±14 min. A delayed sternal closure (2 days) was necessary in two patients.

**Group B**

Seventeen patients had isolated TBA. The diagnosis was established with a cardiac catheterisation (n=6), echocardiography (n=2) or both (n = 9). A balloon atrial septostomy was performed in 12 patients and 9 patients received prostaglandin E2 infusions. The great vessel relationship was anterior-posterior (8/17) or side-by-side (9/17). Coronary artery anatomy was one left circumflex, two right in 6/17 patients, one left, two right circumflex in 5/17 patients, and one right-left anterior descending, two circumflex in 6/17 patients. At operation the mean age, weight and body surface area were 5.9±8.4 months, 5±2.1 kg and 0.27±0.08 m² respectively. Eleven patients underwent repair before 3 months of age. One patient had received a palliative pulmonary artery banding 2.3 years before the complete repair. Associated anomalies include subaortic stenosis in seven patients (41%), a straddling tricuspid valve in two patients, and a left juxtaposition of atrial appendages in two patients.

**Surgical treatment.** The ASO technique was similar to that described for group A. The Lecompte manoeuvre was performed in all cases. The VSD was closed (LV to neoaorta) via the right atrium in all patients. In two patients with multiple VSDs, the closure was approached through the pulmonary artery in one and via the right ventricle in the other. A subaortic resection was necessary in five patients. In four cases this was performed via the right atrium, and in one case via the aorta. One patient required a section and reposition of a tricuspid papillary muscle. One patient required a lung biopsy.

The mean aortic cross-clamp time was 100±13.9 min and the mean period of cardiopulmonary bypass was 165.3±24.5 min. Circulatory arrest was necessary in five patients, its mean duration being 34±10 min.

**Results**

In Group A, there were no hospital deaths (CL=0–28%). Over a follow-up time of 638 patient-months, there have been no late deaths, but three patients have required reoperation. Two children needed a second arch reconstruction for recurrent AAO. In one, a homograft aortic onlay patch was used to enlarge the previous end-to-end anastomosis. The second patient had an end-to-end anastomosis to relieve an initially inadequate PTFE patch repair (performed elsewhere). One patient had moderate residual AAO and required a balloon angioplasty. A third patient received a residual VSD with right ventricular outflow tract obstruction (RVOTO) (60 mmHg gradient).

The actuarial survival and freedom from reoperation rates (Figs. 1 and 2) at 6 years were 100% (CL=66–100%) and 72.9% (CL=38–92%). The functional status of all patients is NYHA class 1, and all patients are in sinus rhythm. In Group B, there were two hospital deaths (11% CL=1–36%) and one late death over 678 patient-months follow-up. The hospital mortality was due to a small left ventricle in one case and in the other to probable coronary spasm with myocardial ischemia. The single late death occurred 6 months following operation, and was related to LV failure of unknown cause (the patient resided overseas and could not return for further treatment). There have been no reoperations in this group. All survivors are in NYHA class 1, maintaining sinus rhythm. The actuarial survival and freedom from reoperation rates at 6 years (Figs. 1 and 2) were 81% (CL=56–93%) and 100% (CL=75–100%). Early and late mortality and reoperation probability are summarised in Table 3.
Discussion

A good anatomic study should begin with a precise definition, but in the case of TBA this presents difficulties. Classically the TBA refers to a form of DORV with the aorta lying more or less directly to the right of the pulmonary trunk, which is closely related to a VSD over which it tends to straddle. There is usually a sagittal orientation of the infundibular septum, which is itself not part of the main intraventricular septum. The presence of a subaortic conus is the rule, but a subpulmonary conus is variably present [6, 28]. There is anatomic overlap between transposition of the great arteries with VSD, TBA and DORV with uncommitted VSD. Part of the difficulty in defining this entity arises in the use of the term “subpulmonic VSD” for patients with a bilateral muscular conus. Taking a simplified surgical view, we might say that TBA is a DORV in which the VSD can be more conveniently and directly connected to the pulmonary artery than to the aorta. This is the definition used for our study.

Taussig-Bing anomaly with AAO has been reported as a common association that complicates about 50% of the cases [12, 16]. Aortic arch obstruction, by way of comparison, is present in only 6% of cases of TGA with VSD. The anatomic characteristics of TBA may influence this correlation, as septal malalignment produces a preferential flow from LV to pulmonary artery [13]. Also, the subaortic muscular conus may act as a partial obstruction tending to divert flow away from the aorta [10]. The flow into the ascending aorta would therefore be limited and flow across the isthmus reduced, with resultant AAO [19]. The natural history of the TBA is very unfavourable, influenced by severe congestive heart failure and early development of pulmonary vascular disease. This prognosis is even worse when AAO is present. Patients with TBA and AAO present for operation earlier in life than infants without AAO, in keeping with the more acute natural history of the former association (24 vs 79 days, P=0.02) (Table 2).

From a therapeutic point of view, there are a number of operations currently in use for TBA. These can be grouped according to the method of VSD closure, either directly to the pulmonary artery, or to the aorta by way of an intraventricular tunnel (Figure 3). Intraventricular tunnel repair, combined with an extensive infundibular resection, is attractive considering that the native aortic valve is preserved and coronary dissection and translocation are avoided [10, 22, 28]. However, this technique requires an adequate tricuspid-pulmonary valve annular distance in order to avoid subaortic tunnel obstruction [11, 21]. For this reason the intraventricular repair of the TBA may be difficult or impossible in some hearts. However, operations in this category have been successfully undertaken by many surgeons, and certainly continue to have an important role in the treatment of the TBA. In the past it has been demonstrated that an atrial switch is an unsatisfactory technique in TBA, with poor early and late results [1, 12, 25]. Such repairs should therefore be avoided except under extraordinary circumstances. The ASO with a LV to neoaoa rct is an excellent way to deal with the intracardiac features of TBA and to establish normal origin of the great vessels and coronary arteries [12, 14]. Over the preceding decade, we employed the ASO in all Taussig-Bing patients for whom biventricular repair was a possibility, irrespective of intracardiac features, great vessel size/position or coronary anatomy.

Several procedures have been used for the treatment of AAO. A large direct anastomosis seems to be the most satisfactory, and is nearly always possible after complete resection of the ductal tissue, adequate mobilisation of the ascending and descending aorta, and an adequate incision of the aortic arch. We believe that an effective arch reconstruction is best achieved by an anterior approach when the degree of obstruction is more complicated than discrete coarctation [9]. If discrete coarctation is present, with a normally sized transverse arch, then initial repair via left thoracotomy is a useful option in some patients. In this way one might reduce the total cardiac ischaemic time required for repair of the TBA, which is performed as a second stage without interim pulmonary artery banding. However, if there are doubts regarding the adequacy of the transverse arch, then a one-stage repair by median sternotomy is more appropriate [5, 7, 17, 18].

For 28 patients treated with a direct ASO, we have an early mortality probably of 7.1% (CL=1–23%), with no deaths occurring in the last decade. There was no significant difference in hospital mortalit y or 6-year actuarial survival between groups A and B, although this is a small series (Table 3). In a comparative mortality analysis of some related anatomic groups treated with ASO, there were also no statistical differences in the early outcome for the TBA (with or without AAO), all DORV biventricular repairs and

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**Fig. 3** Surgical options for TBA grouped according to type of VSD closure. In our own practice, we have employed the ASO option for all TBA patients who were suitable for biventricular repair, irrespective of intracardiac features, great vessel position or coronary anatomy. (REV=Réparation à l’étage ventriculaire, comprising intracardiac features, great vessel size/position or coronary anatomy.)
all infants having arch repairs combined with intracardiac repair (Table 4). The ASO combined with VSD closure could therefore be considered a good choice of procedure for the TBA anomaly with a risk similar to other complex neonatal operations in which a reasonable outcome has been achieved.

It has been demonstrated that a neonatal one-stage repair prevents progression of the subaortic gradient commonly seen in TBA [5, 18]. In the presence of a hypertrophied infundibular septum, a direct muscle resection appears to provide effective relief, although VSD closure may itself tend to reduce the gradient [3]. The use of two large autologous pericardial patches for neopulmonary artery sinus reconstruction is a key technique to avoid post ASO RVOTO. However, in a comparative study of post ASO right ventricle to pulmonary artery mean gradients, we found that the TBA group has ranged higher (TBA: 18.5 mm, transposition with intact ventricular septum: 6.2 mm, transposition with VSD: 9.2 mm, P<0.01). These gradients rarely increase and reoperation is usually not indicated. In our experience only one patient with a gradient needed surgical treatment (concurrent to closure of a residual VSD).

From this analysis, we conclude that the presence of arch obstruction need not adversely affect long- or short-term survival for patients with TBA who undergo ASO. The presence of AAO may influence the ultimate need for reintervention. The TBA repair with ASO carries a risk similar to that of other complex repairs in the newborn. The ASO performed in early infancy (birth to 12 weeks) is therefore a very appropriate operative strategy for both groups of TBA patients, irrespective of other anatomic features.

Acknowledgements Patients in this study have been operated on by Mr. Roger Mee, Mr. William Brawn, and Dr. Tom Karl. We acknowledge the role of our anaesthetists, perfusionists, intensive care specialists, and nurses in the care of these babies.

Table 4 Comparative risk for surgical repair of TBA and related problems. Included are all patients having biventricular repair of DORV, patients with TGA and VSD, all infants with TGA, and all infants undergoing concurrent aortic arch and intracardiac repairs.

<table>
<thead>
<tr>
<th></th>
<th>Mortality (95% CL)</th>
<th>P</th>
<th>n</th>
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<tbody>
<tr>
<td>Taussig-Bing (ASO)</td>
<td>0.07 (0.01–0.23)</td>
<td>*</td>
<td>28</td>
</tr>
<tr>
<td>All DORV (bivent)</td>
<td>0.07 (0.01–0.12)</td>
<td>0.18</td>
<td>143</td>
</tr>
<tr>
<td>TGA, VSD (ASO)</td>
<td>0.05 (0.02–0.12)</td>
<td>1</td>
<td>93</td>
</tr>
<tr>
<td>All TGA (ASO)</td>
<td>0.03 (0.01–0.05)</td>
<td>0.7</td>
<td>245</td>
</tr>
<tr>
<td>Arch + cardiac repair</td>
<td>0.09 (0.04–0.18)</td>
<td>1</td>
<td>75</td>
</tr>
</tbody>
</table>

References


**Discussion**

**Dr. J. Van Son (Leipzig, Germany)**

*Dr. Comas*, whenever there is a Taussig-Bing anomaly with aortic arch obstruction, one has to be highly alert with regard to the presence of associated subaortic obstruction. As a rule of thumb, whenever the diameter of the subaortic area is less than the weight of the baby in kilograms I would be very hesitant to perform an arterial switch operation. In such a case I prefer an intraventricular reouting, in which, in addition to resection of the conal septum, a pathway is constructed from the left ventricle to the aorta, in combination with a Norwood reconstruction of the aorta (including anastomosis of the proximal aorta to the main pulmonary artery), and creation of an extracardiac valved homograft pathway from the right ventricle to the pulmonary artery confluence. I notice that you had two deaths in your series. Were these in any way related to subaortic obstruction?

**Dr. Comas** The patient who had right ventricular outflow tract obstruction had a 60 mmHg gradient, but he had, too, a residual VSD and it probably was increased for that. Our policy for the subaortic stenosis is direct removal of the muscular tissue, and we think there are no problems. One of the differences that I see compared to the other groups, is that in Melbourne when we reconstruct the pulmonary artery we use two pericardial patches, not only one, as a patch for this changes something, but in our experience, it looks like this changes something.

**Dr. E. Lacour-Gayet (Paris, France)** I would like to congratulate Juan Comas for this very good presentation and the group from Melbourne for their beautiful results. We have observed the same in our experience in Paris. Similarly, the Taussig-Bing anomaly by itself was not an incremental risk factor for early mortality and, as well, the risk for reoperation for pulmonary stenosis was important. I have a question regarding RVDI obstruction following arterial switch in Taussig-Bing. In order to prevent late stenosis, do you believe that it could be useful to implant, at the end of the operation, an additional conduit between the right ventricle and the pulmonary artery to bypass either a small aortic, which became pulmonary, annulus or some degree of residual subaortic stenosis?

**Dr. Comas** Thank you, Francois, for your question. I am in agreement with you with this problem. In fact, in a complete follow-up of all the patients who had arterial switch in Melbourne, we see the postoperative gradient between the right ventricle and the PA, the top of the patients who had a bigger gradient is the Taussig-Bing. I think it is double. I think the median is 18 mmHg and in TGA, VSD only, it is 9, and in TGA, intraventricular septum, it is only 5 mmHg. Another technique that you propose probably is a good technique. I am sure, because you propose that, but we didn’t use it. Well, it is possible.