Improved results with single-stage total correction of Taussig–Bing anomaly

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Abstract

Objective: The arterial-switch operation (ASO) for management of Taussig–Bing anomaly is associated with important morbidity, mainly related to multiple associated cardiac anomalies. Our surgical management has evolved to suggest a single-stage total repair strategy tailored to address all abnormalities on an individual basis. We examine the efficacy of this treatment approach. Methods: Thirty-three children, (infants \( n = 29 \)), with Taussig–Bing underwent ASO (1979–2005). In our earlier experience (group 1, \( n = 17 \)), initial palliation was performed as needed, including pulmonary–artery banding (9), coarctation repair (6), and atrial septostomy (3) followed by ASO at a later age. In our later experience (group 2, \( n = 16 \)), single-stage total repair was performed; ASO with ventricular septal defect closure and baffling of left ventricle to neo-aorta performed in neonates with arch obstruction (8), or at age 6 weeks in those with no arch obstruction (8). Concomitant relief of right-ventricle outflow-tract obstruction (RVOTO) was performed in 14 patients. Demographics and operative variables affecting outcomes were analyzed. Results: Mean age at operation for group 1 and 2 patients was 312 ± 477 and 42 ± 31 days (\( p < 0.0001 \)). Aortic arch obstruction (52%), sub-aortic RVOTO (61%) and unusual coronary patterns (52%) were similar for both groups. One-year survival for group 1 and 2 patients was 47 ± 5% and 100% (\( p = 0.001 \)). Associated anomalies such as great vessels position, arch obstruction, and unusual coronaries were not significant risk factors for mortality on multivariable analysis. Ten-year freedom from RVOT and arch re-operation was 55 ± 5% and 96 ± 4%. Five-year event-free survival for groups 1 and 2 was 35 ± 6% and 87 ± 1% (\( p = 0.0016 \)). Significant factors affecting event-free survival were group 1 (HR 108, \( p = 0.0005 \)), and larger weight at surgery (HR 1.3, \( p = 0.02 \)). Conclusions: The Taussig–Bing anomaly is complex and often associated with other cardiac anomalies (arch obstruction, RVOTO, unusual coronary pattern). Advances in perioperative care have significantly mitigated mortality. In our experience with single-stage total repair, event-free survival, especially freedom from RVOTO re-operation, has significantly improved.

Keywords: Congenital heart disease; Great vessels anomalies; Reoperation; Arterial switch

1. Introduction

The Taussig–Bing anomaly is the second most common type of double outlet right ventricle (DORV) [1]. Anatomic repair, with connection of the morphologically left ventricle to the aorta and the morphologically right ventricle to the pulmonary artery is the treatment of choice [2,4]. While this can be achieved with interventricular repair in selected cases [4–6], the arterial switch operation (ASO) has become the preferred management strategy in many major centers [7–14].

ASO for the treatment of Taussig–Bing anomaly has been associated with higher morbidity and mortality compared to ASO for D-transposition of the great arteries [15–17]. This was attributed to the increased complexity of the repair due to the frequency of associated anomalies and the increased incidence of residual lesions at completion of the repair [15–17].

Since 1979, we have performed ASO in the management of all patients with Taussig–Bing anomaly. Earlier in our experience, initial palliation procedures were performed such as pulmonary artery banding with repair of aortic coarctation or associated arch obstruction if present, followed by complete correction at a later age during childhood. In our experience, as well as others, that repair strategy was associated with high operative mortality and late morbidity [15–17]. Since 1999, we adopted a single-stage total repair in all patients. Those having arch obstruction are repaired as neonates and those without arch obstruction are repaired around 6 weeks of age.

The present review was designed to explore the efficacy of the change in strategy and to identify risk factors for less than optimal outcome.
2. Patients and methods

2.1. Inclusion criteria

From June 1979 to June 2005, 33 consecutive neonates with Taussig—Bing anomaly underwent ASO at the Hospital for Sick Children in Toronto. The patients were identified using the surgical database. Clinical, operative, and outcome data were abstracted from the medical records. Approval of this study was obtained from the Research Ethics Board at our institution and individual consent was waived.

2.2. Anatomic characteristics

In 1949, Taussig and Bing defined this anomaly as a subtype of DORV with side-by-side position of the great arteries in which both the aorta and the pulmonary artery arose entirely from the right ventricle and were supported by bilateral coni, the ventricular septal defect (VSD) was located beneath both coni, close to the sub-pulmonary infundibulum and above the trabeculated septum marginal [18]. This original definition of the Taussig—Bing anomaly has been extended in many surgical reports to include all sorts of DORV with sub-pulmonary VSD [1,19,20].

In our current series, DORV definition included patients in whom both great arteries originated from the right ventricle following the 50% rule, with one great artery arising completely from the right ventricle and the second more than 50% from the right ventricle, irrespective of their supporting structure. Patients were identified to have a Taussig—Bing anomaly if they had DORV with sub-pulmonary VSD and no pulmonary stenosis [1].

In the Taussig—Bing anomaly, the pulmonary artery typically arises bi-ventricularly. The aorta may be positioned rightward and slightly anterior (D-transposition), or alongside the pulmonary artery (side-by-side) [1]. These two great vessels are usually parallel; they do not spiral around each other as in the normal heart. In our series, the aorta was positioned side-by-side with the pulmonary artery in 21 patients (64%), and was to the right and slightly anterior to the pulmonary artery in 12 patients (36%).

In the most common coronary pattern, the usual type, the left coronary artery (left anterior descending and left circumflex) arises from sinus 1, while the right coronary artery arises from sinus 2. Sixteen patients in our series (48%) had the usual coronary anatomy pattern, while the remaining 17 (52%) had the less common (unusual) coronary patterns listed in Table 1.

Aortic arch obstruction and other forms of aortic outflow tract obstruction are common in hearts with the Taussig—Bing anomaly. In our series, aortic arch obstruction was common (total n = 17, 52%). This arch obstruction manifested as discrete aortic coarctation (n = 7), diffuse hypoplastic aortic arch (n = 8), or interrupted aortic arch (n = 2).

In addition, sub-aortic right ventricular outflow tract obstruction is commonly present in infants with Taussig—Bing anomaly and was evident preoperatively in 20 patients (61%).

The anatomic characteristics of all patients are listed in Table 1.

2.3. Operative technique

Our operative and perfusion techniques have evolved over time. Our current strategy of single-stage total repair will be briefly described.

All procedures are performed though midline sternotomy. Cardiopulmonary bypass is established via standard aortic and biceaval cannulation. The left ventricle is decompressed by venting through the right superior pulmonary vein. Mild hypothermia (32–34 degrees) is utilized in cases not associated with aortic arch pathology. Antegrade cold blood cardioplegia is used for myocardial protection. Cardioplegia is given into the aortic root before the aorta is opened and directly into the coronary arteries while the neo-aorta is reconstructed and coronary transfer is being accomplished.

After the aortic cross-clamp is applied, the VSD is routinely exposed through the right atrium and pulmonary artery. Only in difficult to expose cases would the VSD be approached through the right ventricle. The VSD is closed utilizing a double velour Dacron patch. After closure of the VSD, the arterial switch is performed. The aorta is transected 3–5 mm above the sino-tubular junction, the coronary artery buttons are excised from their respective sinuses taking generous cuff of aortic sinus tissue. The coronary buttons are adequately mobilized. The pulmonary artery is transected below the bifurcation, the pulmonary arteries are adequately mobilized, and the pulmonary arteries are brought anterior to the aorta. The right ventricular outflow is carefully inspected and, when necessary, muscle resection is performed to prevent right ventricular outflow obstruction (RVOTO) following ASO. The neo-aortic anastomosis is completed after the anterior neo-aortic commissure location is marked from outside. The aortic cross-clamp is released, allowing the aortic root to distend. The proper location for the coronary transfer is selected with a distended neo-aortic arch.
root, incisions are created at those sites, and then the aortic cross-clamp is reapplied. The openings are enlarged to create medi ally hinged trap-door windows for the transfer of coronary buttons. Once coronary artery button transfer is completed, the aortic cross-clamp is removed, and the patient is re-warmed. During the re-warming phase, and on a beating heart, the neo-pulmonary valve is reconstructed with a generous pantaloon-shaped patch of autologous pericardium. In cases with side-to-side great arteries, the reconstruction of the pulmonary artery is done while shifting the opening in the pulmonary artery bifurcation across to the right side to prevent coronary compression.

In cases requiring repair of aortic arch hypoplasia or aortic coarctation, the aortic arch, proximal descending aorta and head vessels are dissected during the cooling phase. The arch repair is performed under deep hypothermic circulatory arrest (DHCA) at 18 degrees. We currently utilize selective cerebral perfusion. Aortic coarctation is repaired with resection and then, most commonly, patch augmentation of the undersurface of the arch (pulmonary homograft of glutaraldehyde treated pericardium) in order to achieve size resection and then, most commonly, patch augmentation.

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Concomitant procedures

Interrupted arch repair
Coarctation/Arch repair
Sub-aortic muscle resection
Closure of multiple ventricular septal defects
Debanding
Patch angioplasty of pulmonary artery
Cardiopulmonary bypass duration (min)
Ischemic duration (min)
Circulatory arrest time (min)

2.4. Follow-up

Late outcomes were determined from recent office visits at the Hospital for Sick Children or from written correspondence with patients’ community cardiologist. The mean follow-up duration was 4.07 ± 1.04 years (range 0 day—25.6 years) and was complete. Follow-up for group 1 and 2 patients was different, 5.7 ± 7.5 years versus 2.1 ± 2.2 years, respectively, (p < 0.0001).

2.4.1. Statistical analysis

All the data were analyzed with the SAS software program (version 9; SAS Institute, Inc., Cary, NC). Data are presented as frequency, median with range, or mean ± SD as appropriate, with the number of non-missing values indicated. Unrelated two-group comparisons were done with unpaired, 2-tailed t-tests for continuous variables and χ² or Fisher’s exact test for categorical data. Predictors of perioperative mortality were identified using multivariable logistic regression analysis. Estimates for long-term survival or freedom from re-operation were made by the Kaplan—Meier method. Differences between survival curves were evaluated with the log-rank statistic. Cox regression was used to determine the independent predictors of late outcomes.

3. Results

3.1. Patient cohort

During the study period, 33 children with Taussig—Bing anomaly underwent ASO. There were 20 males (61%) and 13 females (39%).

Our treatment strategy has evolved with time. In our earlier experience, in group 1 (n = 17), initial palliative surgery was performed at early infancy as needed: pulmonary artery band (n = 9), coarctation/arch repair (n = 6) and atrial septostomy (n = 3). Complete repair with ASO and VSD closure baffling the left ventricle to the neo-aorta was performed at a later stage during childhood (Table 2). In group 2 (n = 16), since 1999, our management strategy has changed to single-stage total correction in infancy with ASO, VSD closure, arch reconstruction and relief of sub-aortic RVOTO as needed (Table 2).

The mean age of the entire cohort was 183 ± 366 days, the mean weight was 5 ± 3.6 kg, and the mean body surface area was 0.28 ± 0.02. Patients in group 2 were considerably younger (42 ± 31 days vs 316 ± 477 days in group 1, p < 0.0001) and their weight was significantly smaller, (3.8 ± 0.6 kg vs 6.1 ± 4.8 kg in group 1, p < 0.0001).

3.2. Early and late survival

There were eight deaths in the series, none since the single-stage approach was initiated in 1999. Five deaths were shortly following ASO for failure to wean off cardiopulmonary bypass, bleeding, or persistent low cardiac output state. Two additional patients expired following re-operation during the same initial admission for residual RVOTO (n = 1), or residual arch obstruction (n = 1). One additional infant had a sudden death at home 2 weeks following discharge. There has
been no mortality beyond that early postoperative period and the overall 1-year and 5-year survival was $72 \pm 8\%$. One-year survival for group 1 and 2 patients was $47 \pm 12\%$ and $100\%$, respectively, ($p = 0.001$).

On multivariable analysis, multiple factors were examined including: great vessels position, unusual coronary anomaly, age, weight, group, and arch involvement. None of those variables was found to be a significant predictor for survival.

3.3. Time-related freedom from morbid events

During the follow-up period, 8 patients underwent 9 cardiac re-operations. Two re-operations were early during the same admission following ASO. One was for residual arch obstruction and the other was for residual sub-aortic RVOTO. Both patients expired during the same hospital stay.

Additional re-operations included relief of sub-valvar and supra-valvar RVOTO ($n = 5$), resection of an aneurysm of the right ventricular outflow tract ($n = 1$), mitral valve repair ($n = 1$), and aortic valve replacement ($n = 1$). In addition, 1 patient with persistent compression of the left main bronchus following total repair and arch reconstruction required suspension of the left pulmonary artery and the aortic arch to relieve his airway compression.

Ten-year freedom from re-operation for RVOTO and from arch re-operation for the entire cohort was $55 \pm 5\%$ and $96 \pm 4\%$, respectively. Five-year event-free survival for groups 1 and 2 was $21 \pm 10\%$ and $88 \pm 11\%$, respectively, ($p = 0.0016$) (Fig. 1).

On multi-variable Cox regression analysis, significant factors affecting event-free survival were group 1 (HR 108, $p = 0.0005$), and larger weight at surgery (HR 1.3, $p = 0.02$).

4. Discussion

Taussig—Bing anomaly is a complex cardiac disease that includes, in addition to the presence of sub-pulmonary VSD, a spectrum of malformations involving the position of the great arteries, coronary artery anatomy, presence of sub-aortic RVOTO and various forms of aortic arch obstruction [1].

Anatomic repair, with connection of the morphologically left ventricle to the aorta and the morphologically right ventricle to the pulmonary artery is the treatment of choice [2—4]. While this can be achieved with inter-ventricular repair re-routing the left ventricular flow through a patch tunnel across the VSD to the aorta with resection of infundibular septum in selected cases; [4—6] arterial switch operation has become the preferred management strategy in many major centers [7—14].

4.1. Survival

In many series studying outcomes following the ASO, Taussig—Bing anomaly was associated with a higher rate of mortality compared to D-transposition of the great arteries [15—17]. This increased risk was attributed to several risk factors such as complex associated arch and right ventricle outflow tract anomalies [17], the position of the great arteries [9], and the increased likelihood of unusual coronary pattern [12].

In our current series, associated anomalies such as aortic arch obstruction (52%), sub-aortic RVOTO (61%), side-by-side position of the great arteries (64%), and unusual coronary distribution patterns (52%) were frequent. All those variables were examined and none was found to be a significant factor for early or late mortality.

This diverse identification of risk factors from different series and institutions including ours can be explained by the rarity of this disease and the small number of patients in each series. Nonetheless, our results indicate that in the current era, with increased experience in ASO and arch reconstruction, improved perfusion strategies and most importantly with advances in postoperative care, complex cardiac reconstructions that require prolonged cardiopulmonary bypass and ischemic times can be undertaken with excellent survival rates provided complete repair is performed and residual lesions are eliminated.

In the current series, despite more complex repair in the later stage of our experience (group 2), we noted that the required cardiopulmonary bypass time has decreased reflecting increased operative experience in the ASO.

Another controversy is related to single-stage versus two-stage management of associated cardiac anomalies, most importantly associated arch obstruction.

A single-stage total repair approach addresses all malformations associated with Taussig—Bing anomaly, reduces residual lesions following ASO, and may avoid problems related to delaying surgery such as development of ventricular hypertrophy, development of pulmonary hypertension, effects of prolonged cyanosis, and complications related to the pulmonary artery banding. In recent years, there have been multiple reports of successful repair of complex transposition anomalies associated with arch obstruction with excellent early and late outcomes [10,11,22,23].

In our experience, survival improved significantly in our later era with no early or late mortality following complete single-stage repair compared to a considerably higher mortality in our early experience in which a staged approach was primarily utilized. We believe that this improvement is a reflection of the advances in operative and perfusion
techniques, and the improvement in postoperative care rather than a solid evidence of superiority of the single-stage approach. This decreased operative mortality paralleled improvement in our outcomes following ASO for D-transposition of the great arteries at our institution. Postoperative cardiac mechanical support may still be required following aggressive total repair. In our series, two neonates required postoperative extracorporeal membrane oxygenator (ECMO) support for failure to wean off cardiopulmonary bypass support and both successfully survived their hospital stay and are asymptomatic on latest follow-up. This incidence of required ECMO implies a significant morbidity for the single-stage approach which could lead to mortality as the series expands with time.

On the other hand, staged treatment of Taussig–Bing anomaly with initial arch repair ± pulmonary artery banding, followed by ASO at a later stage in childhood remains a valid option and continues to be utilized as described in recent publications from experienced centers reporting good outcomes [6,12,14].

4.2. Late morbidity

The most frequently reported late morbidity following ASO is RVOTO [8,10–14]. Several preoperative anatomic indicators have been identified as risk factors for the development of late RVOTO, such as associated aortic arch obstruction, preoperative presence of sub-aortic RVOTO, side-by-side position of the great arteries, mismatch between the size of the pulmonary artery and the aorta, unusual coronary artery distribution, and the presence of malaligned VSD [13,24]. Therefore, it is natural that patients with Taussig–Bing anomaly have a high risk of developing RVOTO following ASO compared to patients undergoing ASO for D-transposition of the great arteries. RVOTO can be in the form of supra-valvar, valvar or sub-valvar stenosis [8,10–14,24].

In our series, five patients underwent six re-operations of RVOTO, all of them had supra-valvar stenosis and three had additional sub-valvar and valvar stenosis. Subsequent surgical management of RVOTO included patch angioplasty of the pulmonary arteries (n = 5), RV-PA conduit (n = 2), infundibular muscle resection (n = 1), and infundibular patch (n = 1). Overall ten-year freedom from re-operation for RVOTO was 55%. There have been no re-operations for RVOTO in the single-stage total repair group patients. Although the follow-up duration for group 2 patients is shorter, we believe that the various technical modifications that we have adopted will contribute to decreased incidence of RVOTO development.

Several technical factors have been identified resulting in increased risk of supra-valvar RVOTO development. Those include insufficient mobilization of the pulmonary arteries, inadequate size or form of the pericardial patch used to reconstruct the neo-pulmonary trunk, and pericardial patch shrinkage [8,10–14,24]. Technical modifications have been applied in our practice to reduce the risk of supra-valvar pulmonary stenosis such as the use of a generous size autologous pericardial patch in the reconstruction of the neo-pulmonary trunk, and complete mobilization of the pulmonary arteries prior to performance of the Lecompte maneuver. We continue to perform the Lecompte maneuver in all cases. In patients with side-by-side great vessels position, the Lecompte maneuver is performed and the neo-pulmonary trunk is reconstructed as usual. Following that, the pulmonary anastomosis is shifted to the right pulmonary artery to prevent compression of the coronary circulation and distortion of the pulmonary valve.

More importantly, we have recognized the importance of vigorous resection of the sub-aortic right ventricular outlet during the ASO. With this technique, none of our group 2 patients had sub-valvar and valvar RVOTO on follow-up echocardiogram suggesting that a low incidence of problems in the RVOT can be achieved. We expect that future follow-up will confirm those favorable results, as valvar and sub-valvar pulmonary stenosis following ASO is usually evident early in the postoperative period. Other groups have reported similar encouraging results with comparable aggressive sub-aortic conal muscle resection approach [10,13].

Similar to all neonates and infants with aortic coarctation or hypoplastic aortic arch, re-operation may be required for recurrent arch obstruction. In our series, the incidence of arch re-operation requirement was low and was not different between single-stage or staged approaches. Total repair through a median sternotomy allows more proximal extension of the incision into the ascending aorta and the construction of a larger anastomosis with augmentation of the entire aortic arch with a patch when necessary. Patients with diffuse arch hypoplasia or interrupted aortic arch require midline sternotomy for correction of their arch anomalies. On the other hand, in patients with discrete coarctation, staged approach, with initial coarctation repair followed by later second stage ASO is performed in many centers with excellent early and late results [6,12,14].

The optimal approach for neonates and infants with intracardiac anomaly associated with discrete coarctation remains controversial. In a previous report from our institution, the risk of recurrent coarctation and arch intervention following surgery via midline sternotomy or left thoracotomy was not found to be different following single-stage or staged repair of the aortic arch in 141 neonates with ventricular septal defect and coarctation of the aorta [25]. A potential benefit of the staged approach is eliminating the need for DHCA and prolonged cardiopulmonary bypass duration during the vulnerable neonatal period. While the risk of increased cardiopulmonary bypass in the current era seems to have been neutralized, as evident in our series as well, the issue of adverse effect of DHCA on future neurological development remains unanswered. There has not been a direct comparison of neurological injury or long-term neurological development between patients undergoing combined repair with the use of cardiopulmonary bypass and DHCA and patients undergoing initial coarctation repair alone. Moreover, although we tend to use selective cerebral perfusion during arch reconstruction, available studies have failed so far to demonstrate significant benefit in neurological outcome. A prospective study with formal and thorough periodic neurological assessment may be needed to determine any benefit of that approach.

4.3. Study limitations

This case series is subject to the limitations inherent in all retrospective observational studies such as selection bias and...
References


Appendix A. Conference discussion

Dr E. Belli (LePlessis-Robinson, France): An important information is missing, crucial for us. What was the cause of mortality, early or intermediate mortality in both groups?

Dr Alsoufi: All mortalities were in the perioperative period, with one additional death at home few weeks following discharge. There was no mortality beyond that period. The cause of preoperated persistent low cardiac output state. Two additional patients expired following early reoperation during the same initial admission for residual right ventricular outflow tract obstruction in one patient and residual arch obstruction in the other patient.

We don’t think that the improved results we have achieved in the later part of our series are necessarily reflective of the superiority of the single-stage total correction approach; our study extends over 25 years and there are many variables that are involved, including changes in our perfusion and surgical strategies, our philosophy towards initial palliation versus single-stage repair, the way we handle right ventricular outflow tract obstruction, and most importantly the improvement in postoperative care in the intensive care unit. All those variables likely contributed to the superior results in the current era.

Dr H. Karusawa (Tokyo, Japan): We surgeons know that this particular anomaly, Taussig—Bing anomaly, has a very wide variation in terms of the lack of randomization. Additionally, the small cohort size due to the rarity of this disease, and the multiple variables in this series that reflect a development of different surgical approaches related to management of arch reconstruction, relief of sub-aortic RVOTO, policies regarding initial palliative versus single-stage total correction, changes in perfusion strategies, and most importantly changes in postoperative intensive care all preclude sophisticated statistical analyses and limit the power of the study to identify clinically significant risk factors.
anatomy. Have you had an experience in doing the so-called intracardiac repair called the Kawashima operation in this particular anomaly? If you have such experience, how do you choose the surgical option, the Kawashima or arterial switch operation for this particular disease?

Dr Alsoufi: Historically, our institution has adopted the arterial switch operation for the management of children with Taussig–Bing anomaly. We don’t have a large experience with the Kawashima procedure at our hospital. Despite that Kawashima operation is a well-known treatment option for Taussig–Bing patients with side-by-side vessels, the available literature is limited in regards to that surgery. My review of the literature revealed three papers with a total of 20 patients who have undergone the Kawashima operation. The reported outcomes have been good although there were some associated problems such as tunnel stenosis and RV outflow tract obstruction.