Single-Stage Neonatal Repair of Taussig-Bing Anomaly

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Taussig-Bing is a form of double outlet right ventricle that is characterized by the presence of subpulmonary ventricular septal defect, double conus and side by side great arteries. Frequently, associated anomalies such as aortic coarctation, arch hypoplasia, subaortic obstruction, and atypical coronary artery anatomy are present; all increasing the complexity of the repair. Advances in perioperative care, perfusion and surgical techniques have allowed the performance of complete repair of Taussig-Bing anomaly with excellent results. Consequently, single stage repair has become the preferred treatment method in most pediatric cardiac centers. Reoperation for right ventricular outflow tract obstruction remains the most common late complication however aggressive resection of the right ventricular muscle bundles creating subaortic obstruction at time of initial repair and reconstruction of the neo pulmonary artery with a generous patch have mitigated that reoperation risk.

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Introduction

The Taussig-Bing anomaly is the second most common type of double-outlet right ventricle. It is defined by the presence of subpulmonary ventricular septal defect, double conus, and side-by-side great arteries. Anatomical repair with the intraventricular baffle has been described and applied in a few selected cases. Nonetheless, the arterial switch operation has become the preferred surgical management strategy in most pediatric cardiac centers. Taussig-Bing is a complex cardiac anomaly; in addition to the subpulmonary ventricular septal defect and side-by-side orientation of the great arteries, a spectrum of malformations involving the aortic arch, coronary arteries, right ventricle, and subaortic area exists. Consequently, surgical treatment of Taussig-Bing anomaly is challenging, and operation is lengthy as the surgeon is required to perform several procedures simultaneously, including the arterial switch operation, ventricular septal defect closure, repair of aortic coarctation and arch hypoplasia, and relief of subaortic right ventricular outflow tract obstruction, which is further complicated by the size mismatch between the pulmonary artery and aorta, and the frequent presence of atypical coronary anatomy. Some authors proposed staged correction of Taussig-Bing anomaly with initial coarctation repair and pulmonary artery banding, followed by a delayed arterial switch and ventricular septal defect closure. Nonetheless, single-stage repair offers several advantages over staged repair, including adequate repair of arch hypoplasia, avoidance of complications related to the pulmonary artery band, and problems related to ventricular hypertrophy, prolonged cyanosis, and pulmonary hypertension. Our policy is to offer single-stage repair to neonates born with the Taussig-Bing anomaly at the age of 1-2 weeks for those who have concomitant arch obstruction and at the age of 2-4 weeks for those who do not have arch obstruction, although some surgeons advocate delaying surgery till 6-8 weeks of age in those with no arch obstruction. Single-stage repair might be challenging in very small neonates, or contraindicated in those who have additional problems such as intracranial hemorrhage, sepsis, lung infection, or significant extracardiac anomalies. In those patients, a staged approach with coarctation repair and pulmonary artery banding, or more recently a hybrid approach with ductal stenting and bilateral pulmonary artery branch banding might be applied. An additional alternative strategy that can be applied in small neonates in whom intracardiac repair can be challenging such as those with multiple ventricular septal defects is completion of extracardiac repair (arterial switch operation with or without arch repair) in addition to banding of the neopulmonary artery with completion of the intracardiac repair at a later stage.

During the single-stage operation, aortic arch augmentation with a patch is commonly performed. It serves to repair proximal aortic hypoplasia that is often present in patients with Taussig-Bing anomaly and to restore the size mismatch between the dilated pulmonary artery (neoaorta) and the ascending aorta. The ventricular septal defect is usually approached through the dilated pulmonary valve, although a combined exposure through the pulmonary and tricuspid valves is often necessary to close this challenging defect.
Exposure can be facilitated via a right ventriculotomy, although this is rarely necessary. Some exposure can be gained as well through the aortic valve, especially in those with no significant subaortic obstruction. Although there are some concerns about pulmonary valve (neo-aortic valve) injury with ventricular septal defect closure through the pulmonary valve, we have not noted any problems with this approach. The coronary artery anatomy is atypical in more than 50% of the cases, and the incidence of single coronary artery is higher than that in dextrotransposition of the great arteries. In patients with difficult coronary anatomy, the closed technique of coronary transfer is particularly helpful. Finally, reoperation for right ventricular outflow tract obstruction is traditionally the most common late complication following the arterial switch operation for Taussig-Bing anomaly. Nonetheless, this late complication can be mitigated by aggressive resection of the right ventricular muscle bundles, creating subaortic obstruction at the time of repair, and by the use of a generous patch for the reconstruction of the neopulmonary artery (Figs. 1–13).
Figure 1 The heart of a neonate with the Taussig-Bing anomaly. There is double-outlet right ventricle with subpulmonary ventricular septal defect, double conus, and malalignment of the conal septum. In extreme cases, both great vessels arise entirely from the right ventricle, which further complicates intracardiac baffle from the left ventricle to the aorta and usually requires enlargement of the ventricular septal defect to prevent late development of left ventricular outflow tract obstruction. The great arteries are side by side with the aorta to the right of the pulmonary artery. There is significant size mismatch between the pulmonary artery and the aorta, with the pulmonary artery occasionally double the size of the aorta. The aortic arch is hypoplastic, and there is coarctation of the aorta with ductal dependent circulation. The coronary artery pattern is commonly atypical. The malaligned conal septum creates subaortic obstruction that contributes, along with the hypoplastic aortic annulus, to the development of right ventricular outflow tract obstruction following the arterial switch operation. Although the right ventricle is often mildly hypoplastic, biventricular repair is possible in most patients even though a few patients might have significant hypoplasia of the right ventricle that precludes biventricular repair.
Figure 2 Median sternotomy is performed and the thymus is removed. A rectangular patch of the pericardium is excised and preserved wet in a gauze soaked with normal saline for later use to reconstruct the neopulmonary artery. Much of the dissection is performed before cannulation. The innominate vein is mobilized and retracted with a suture to facilitate the exposure and dissection of the brachiocephalic vessels. The aortic arch, brachiocephalic vessels, and ductus arteriosus are all mobilized. The ascending aorta is separated from the pulmonary artery. Before cardiopulmonary bypass, the optimal sites for coronary button transfer on the main pulmonary artery are carefully identified and marked with 7-0 polypropylene marking sutures. In this patient, the right coronary artery and the left anterior descending artery arise from the anterior coronary sinus, whereas the left circumflex artery arises from the posterior sinus. The left and right pulmonary arteries are mobilized beyond their first lobar branches and encircled with vessel loops or heavy silk. After systemic heparinization, a partial occluding clamp is applied at the innominate artery, and an incision is made at the center of the clamp. The anastomosis is performed between the innominate artery and a 3.5 mm polytetrafluoroethylene (PTFE) graft that will be used for arterial cannulation. An 8-F DLP arterial cannula (Medtronic, Minneapolis, MN) is inserted into the graft and secured in place with a silk tie. The superior vena cava and inferior vena cava are directly cannulated using right-angle metal-tip DLP venous cannulas (Medtronic, Minneapolis, MN). Cardiopulmonary bypass is started and the patient is cooled for 30 minutes to a bladder temperature of 18°C. Immediately after the initiation of cardiopulmonary bypass, the branch pulmonary arteries are snared. A 10-F DLP left heart vent catheter (Medtronic, Minneapolis, MN) is placed through a purse string in the left atrium anterior to the entrance of the right superior pulmonary vein and is directed into the left atrium. The remaining dissection is completed while on cardiopulmonary bypass.
Adventitial purse string sutures are placed around the ductus using 6-0 polypropylene sutures and the ductus is divided between those sutures. The branch pulmonary artery snare is released, and additional mobilization is performed if necessary. After completion of all the necessary dissection, the aortic cross clamp is applied and antegrade blood-buffered cardioplegia using the Del-Nido solution is administered through the aortic root. Although single-dose cardioplegia is typically used in neonates who undergo the arterial switch operation or those who undergo aortic arch reconstruction including the Norwood operation, an additional dose of cardioplegia is commonly used in patients with Taussig-Bing anomaly who require concomitant arch reconstruction with the additional dose usually given following the closure of the ventricular septal defect and enlargement of the right ventricular outflow tract and before arch reconstruction and coronary transfer. The second cardioplegia dose is given directly into the mobilized coronary buttons using a 3-mm olive-tip DLP arteriotomy cannula (Medtronic, Minneapolis, MN). The aorta is divided approximately 0.5 cm above the sinotubular junction. After inspection of the coronary ostia, the coronary artery buttons are excised with a generous rim of aortic sinus tissue and are mobilized to the extent deemed adequate to allow a tension-free anastomosis.
Figure 4 The aortic cusps are retracted to allow inspection of the right ventricular outflow tract. Septal myectomy is often needed and is performed sharply using an 11-blade scalpel and sharp scissors to excise the redundant muscles causing obstruction medially and anteriorly underneath the aortic valve annulus. Rarely, adequate relief of the right ventricular outflow tract obstruction might require a small ventriculotomy and patch augmentation. In that case, this should be accomplished very carefully given the frequent presence of important coronary arteries crossing the right ventricular outflow tract. Very rarely, a right ventricle to pulmonary artery conduit over the crossing coronary artery is necessary.
Figure 5  (A) The main pulmonary artery is divided below its bifurcation. The pulmonary valve cusps are retracted to allow exposure of the ventricular septal defect. Occasionally, the ventricular septal defect needs to be enlarged to prevent late development of left ventricular outflow tract obstruction. The ventricular septal defect is closed using a double velour Sauvage Dacron patch (C.R. Bard, Murray Hill, New Jersey) that is secured to the edges of the ventricular septum with a running 6-0 polypropylene suture. Retraction of the pulmonary valve cusps facilitates exposure and placement of the superior sutures to the muscle rim underneath the pulmonary valve cusps to baffle to left ventricular outflow tract to the pulmonary valve (neoaortic valve).
It is not uncommon that additional exposure to the ventricular septal defect is required through the right atrium with retraction of the tricuspid valve leaflets to facilitate suture placement. Following completion of the ventricular septal defect closure and the tunneling of the left ventricle outflow tract to the pulmonary valve, the atrial septal defect is closed primarily with a running 6-0 polypropylene suture.
By the time the ventricular septal defect is closed, the patient has been cooled for more than 30 minutes to a bladder temperature of 18°C. Yasargil temporary occlusion clips are applied on the innominate artery proximal to the PTFE graft, and distally on the remaining 2 brachiocephalic arteries. A vascular clamp is applied on the descending aorta as distally as possible with care not to include the left recurrent laryngeal nerve with the clamp. The flow is decreased to continuous selective cerebral perfusion at 30-40 mL/kg/min. During the period of selective cerebral perfusion, near-infrared spectroscopy is monitored to manage the flow and PCO₂. The aorta is divided proximal and distal to the ductal insertion points, and the entire residual ductal tissue is excised from both ends. The arch is opened beginning at the medial aspect of the divided ascending aorta and extending across the inferior aspect of the transverse aortic arch all the way to the divided distal end at the ductus insertion site. A longitudinal cutback incision for almost 0.5 cm is created into the posterior left lateral descending aorta.
The opened distal arch is next interdigitated into the proximal descending aorta cutback and the posterior walls are sutured together using a running 7-0 polypropylene suture. Following that, a second longitudinal cutback incision for at least 1 cm is created into the anterior descending aorta. An arrowhead-shaped pulmonary homograft patch is used to complete the aortic arch reconstruction and creation of the neoascending aorta. The suture line of the patch is begun at the apex of the anterior cutback incision in the descending aorta and is completed using a running 7-0 polypropylene suture. Following the completion of the suture line, the reconstructed aortic arch is deaired by releasing the descending aortic clamp, and an aortic cross clamp is applied on the neoascending aorta. The Yasargil clips are removed, and cardiopulmonary bypass is increased to full flow. Patient rewarming is started.
Figure 7 (Continued)
Figure 8 Medially placed flaps are created on the main pulmonary artery (neoaorta) with the guidance of the original marking sutures that were placed to mark the site for the coronary transfer before cardiopulmonary bypass. The coronary artery buttons are sutured into the neoaorta using running 7-0 or 8-0 polypropylene sutures. Not uncommonly, the implantation site for one of the coronaries is high and crosses the suture line between the neoaorta and neoascending aorta.
The Lecompte maneuver is performed, bringing the pulmonary artery bifurcation anterior to the neoascending aorta. The aortic cross clamp can be reapplied on the neoascending aorta to keep the pulmonary artery bifurcation behind it after performing the Lecompte maneuver (not shown). The proximal neoaorta is sutured to the reconstructed neoascending aorta using a running 7-0 polypropylene suture. Following left ventricular deairing maneuvers, the aortic cross clamp is released and that anastomosis suture is tied. The left heart vent catheter is placed back on gentle suction and the remaining part of the procedure is performed on a beating heart. The suture lines are inspected and additional repair sutures are placed if necessary. The coronary arteries are inspected to ensure proper placement and orientation and satisfactory perfusion to all areas.
Figure 10: Alternatively, the closed coronary transfer technique is applied. This technique is especially helpful in patients with unusual coronary artery pattern such as single or intramural coronary arteries, commonly present in Taussig-Bing. In this closed technique, the location of the superior end of the facing commissure between the 2 potential sites for the coronary transfer is marked externally with a 7-0 polypropylene suture. The anastomosis between the main pulmonary artery (neoaoorta) and the reconstructed neoascending aorta is performed before making the incisions for the coronary transfer. The aortic cross clamp is released to allow distention of the aorta and closure of the neoaoortic valve. This maneuver helps in precisely determining the site for the coronary button transfer on the distended neoaoorta. Following this, incisions are made in the neoaoorta and medially based flaps are created for the coronary buttons with the guidance of the external marking sutures. The aortic cross clamp is reapplied and the coronary buttons are sutured to the corresponding openings on the neoaoorta.
Figure 11  A vent is placed across the tricuspid valve and is placed on gentle suction. This allows good exposure during reconstruction of the neopulmonary artery on a beating heart. A pantaloon-shaped patch of autologous pericardium is used to reconstruct the neopulmonary artery using a 7-0 polypropylene suture. If the commissure between the coronary arteries was detached during the coronary button excision, this is resuspended to the patch.
Figure 12 In patients 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. The opening in the pulmonary artery bifurcation is enlarged toward the right pulmonary artery. The left end of the original pulmonary artery bifurcation is closed with the use of a small patch of autologous pericardium. The anastomosis between the neopulmonary artery and that shifted opening is completed using a 7-0 polypropylene suture.
The right atrium is closed with a running 6-0 polypropylene suture. The left heart vent catheter is removed and a left atrial monitoring line is inserted through the same site. Following rewarming to a bladder temperature above 35°C, the patient is weaned off cardiopulmonary bypass. After completion of modified ultrafiltration and confirmation of satisfactory repair with transesophageal echocardiogram, heparin reversal is achieved with protamine sulfate and hemostasis is secured. The cannulas are removed, and the graft that was used for arterial cannulation is clipped and divided. A right atrial line is placed via a purse string suture in the right atrial appendage. Temporary pacing wires are placed on the right ventricle and right atrium. Overall, 2 15-F round silicone Blake drains (Ethicon, Somerville, NJ) are used to drain the pleural and pericardial cavities and a 10-F round Jackson-Pratt drain (Cardinal Health, Dublin, OH) is used to drain the peritoneal cavity. Commonly, the sternum is left open. A silastic patch is sutured to the skin edges, and a transparent dressing (Tegaderm, St. Paul, MN) is applied on top of the silastic patch for further security.
Results

Despite surgical complexity, results of the arterial switch operation for Taussig-Bing anomaly are good. Operative mortality in the current era is between 0% and 7%. Atypical coronary artery anatomy and concomitant arch obstruction were identified in some series to be associated with higher operative mortality on univariate analysis although those factors might have been neutralized in the current era. Survival at 10 years ranges between 80% and 100%, and the majority of children are in New York Heart Association class I. Late reoperations continue to be most commonly needed for right ventricular outflow tract obstruction followed by recurrent arch obstruction. Freedom from reoperation at 10 years ranges between 60% and 80%. In a series from Toronto, reoperation risk for right ventricular outflow tract obstruction was mitigated by an aggressive approach to relief subaortic obstruction at the time of initial surgery.

Conclusions

Single-stage repair of Taussig-Bing anomaly and related lesions is associated with low operative mortality and good late outcomes. Staged repair is reserved for the very small neonates and those with additional major extracardiac morbidity. Methods to decrease the risk of late reoperation for recurrent arch obstruction and right ventricular outflow tract obstruction include the interdigitating coarctation repair technique, arch augmentation with a patch, aggressive resection of the obstructive subaortic muscles, and reconstruction of the neopulmonary artery with a generous patch.

References