Congenitally Corrected Transposition of the Great Arteries: Anatomic, Physiologic Repair, and Palliation

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Congenitally corrected transposition of the great arteries (ccTGA) is a lesion that rarely occurs in isolation. The presenting physiology of ccTGA is predominantly secondary to the concurrent cardiac lesions; however, as the child ages, unrepaired ccTGA results in progressive failure of the morphologic right ventricle under the strain of maintaining a systemic pressure. Repair of ccTGA was initially focused on rectification of the underlying physiologic aberrations, but in recent years, the focus of repair has shifted toward anatomic correction to avoid failure of the morphologic right ventricle. This anatomic repair is commonly associated with improved long-term mortality at the cost of increased short-term mortality. Key preoperative considerations such as morphologic left ventricular pressure, tricuspid valve competency, and out flow tract obstructions can assist in determining the optimal repair for individual patients. An alternative, single ventricle, pathway has been proposed for any patient without optimal preoperative anatomy to improve long-term survival. Adjunctive repair options including pulmonary artery banding and one-and-a-half ventricle repairs have also been proposed to augment the survival curves.

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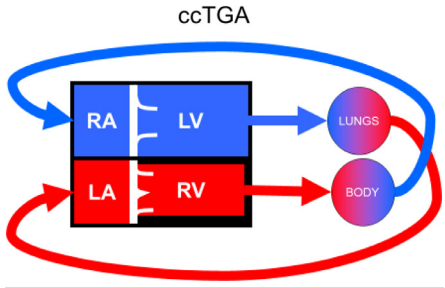

Congenitally corrected transposition of the great arteries.

Central Message

Selection of the appropriate operation, using preoperative patient characteristics, provides the best long-term outcomes for patients presenting at any age with unrepaired congenitally corrected transposition of the great arteries.

INTRODUCTION

Discordance of atrioventricular (AV) and ventriculoarterial connections, commonly referred to as congenitally corrected transposition of the great arteries (ccTGA), was first described and named by Rokitansky in 1875 [1]. ccTGA is exceptionally rare with incidence suggested to be around 1 in 33,000 live births, totaling approximately 0.05% of all congenital heart defects [2]. The anatomy of ccTGA involves systemic venous drainage to the morphologic right atrium (mRA), which is connected via the mitral valve to the morphologic left ventricle (mLV) whose outflow is the pulmonary arterial system. The pulmonary venous drainage arrives at the morphologic left atrium (mLA), which crosses the tricuspid valve into the morphologic right ventricle (mRV) with outflow into the systemic arterial system. In isolation, ccTGA has an in-series circulation without mixing; therefore, the presence or absence of associated defects dictates the patient's clinical presentation. Associated defects are common with ventricular septal defect (VSD) present in 90% of patients, pulmonary valve stenosis or atresia in 65%, and tricuspid valve abnormalities in 46% [3].

NATURAL HISTORY

In a review of 30 patients diagnosed with ccTGA prenatally, there were no events of cardiac decompensation in utero [4]. The risk of congenital heart block in ccTGA is 7% [5], with a 1.3–2% annual risk of complete heart block development [6,7].

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The natural history of unrepaired, isolated ccTGA is progressive failure of the mRV at systemic pressures leading to tricuspid insufficiency [8], though the timing of presentation of symptoms is variable and many patients can live functional lives prior to becoming symptomatic, including successful childbirth [9]. In one review of 44 patients presenting after age 18, 17% were undiagnosed prior to age 60 and 32% have not required surgery at a median follow-up of 53 months. Moderate-to-severe systemic AV valve insufficiency was present in 59%, with mean systemic ventricle ejection fraction of 41% (standard deviation ± 10%), and high-grade AV block in 32% [10].

INDICATIONS FOR INTERVENTION

The indications for surgical intervention are typically dependent on the physiologic consequences of associated defects. Patients diagnosed in utero with ccTGA had a 46% risk of surgery by 32 months old [4]. In a study of 44 adult patients with unrepaired ccTGA, surgical intervention was required in 68% of patients at the same median age as diagnosis. Relative to the patients not requiring surgical intervention, these patients had a higher percentage of systemic AV valve regurgitation and greater mean cardiothoracic ratios by chest radiograph [10].

SURGICAL MANAGEMENT

Surgical management of ccTGA has 2 core philosophical principles focusing on either physiologic repair or anatomic repair. Both of these repairs focus on a biventricular, in-series repair with complete septation of the heart. Neither of these repair techniques has provided ideal long-term outcomes, thus several additional palliative approaches to the surgical management of ccTGA have been proposed including pulmonary artery banding, Fontan palliation, and the addition of a one-and-a-half ventricle repair to the physiologic or anatomic repair. For surgical planning, abnormalities of the conduction system in ccTGA should be noted. In atrial situs solitus, the AV node is anterior in the atrial septum at the junction of the anterior limbus of the atrial septum and the AV valve ring. The penetrating bundle traverses the annulus of the pulmonary valve prior to descent onto the anterior infundibular septum [11]. In atrial situs inversus, the AV node is in its regular posterior position with the course of the penetrating bundle as it would be in AV concordance [12].

PHYSIOLOGIC REPAIR "CLASSIC REPAIR"

Physiologic repair of ccTGA, also referred to as "classical" repair, focuses on repair of concomitant defects to complete septation of the heart for an in-series biventricular repair. In the physiologic repair, the atrioventricular and ventriculoarterial discordances are maintained with the mRV serving as the systemic ventricle (Fig. 1). Concerns with this approach are related to the long-term potential of the right ventricle and tricuspid valve to serve the systemic circulation.

VSD Closure

In physiologic repair, VSD closure is typically performed through the right atrium via a transmitral approach.

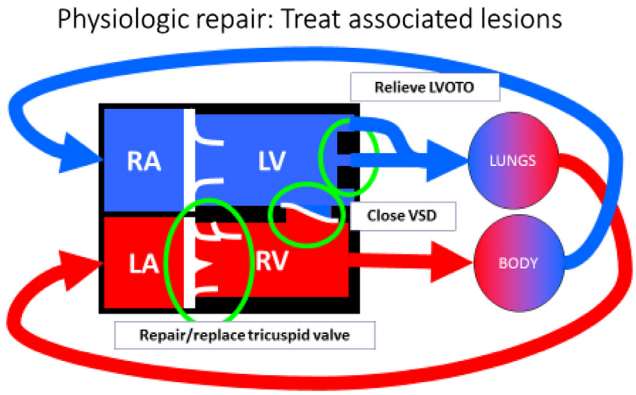

Figure 1 Schematic representation of the physiologic repair in ccTGA by restoring normal physiology by repairing the associated lesions. LA, left atrium; LV, left ventricle; LVOTO, left ventricular outflow obstruction; RA, right atrium; RV, right ventricle; VSD, ventricular septal defect.

Alternatively, a ventriculotomy can be used to access the VSD when a left ventricle to pulmonary artery conduit is planned. Using either approach, avoidance of injury to the conduction system is an important consideration. In ccTGA, the conduction system is located in the anterosuperior margin of the defect on the mLV side as we mentioned earlier. The VSD sutures along the anterosuperior margin are taken along the opposite (eg, right ventricular) side of the septum [5,6]. In a series of 111 patients with AV discordance, including 68 with ccTGA, VSD closure was performed in 91%, mLV to pulmonary artery conduit placed in 59%, and atrial septal defects were closed in 58%. Overall, early mortality was 16%; however, there was an era effect with early mortality decreased to 3% in patients receiving operation after 1986. Prior operation, higher preoperative NYHA class, and nonsinus preoperative rhythm were associated with late mortality [13]. Overall, 5-year mortality has been reported across series as 77−78% [13,14], 10-year mortality as 60.7−95.5% [13−15], and 20-year mortality as 63−90.2% [15−17].

Left Ventricular Outflow Tract Obstruction Repair

A VSD is associated with morphologic left ventricular outflow tract obstruction (mLVOTO) in 46% of all patients [5]. Depending on the extent of LVOTO or pulmonary valvar stenosis, patients may either be cyanotic, overcirculated, or fortuitously balanced. The primary mechanisms of mLVOTO include septal muscular hypertrophy, accessory AV valvar tissue, or aneurysmal dilatation of the membranous ventricular septum. At the time of VSD closure, the muscle of septal hypertrophy can be resected if the heart is situs inversus; however, muscular resection would be ill-advised in the setting of situs solitus due to the presence of the conduction system in the mLVOT. If the mLVOTO is secondary to accessory AV valvar tissue, this can be resected if not of critical importance to the function of the AV valve. AV valvar regurgitation represents a significant source of morbidity and reoperation following physiologic repair. Aneurysmal dilatation of the membranous

ventricular septum can be resected with minimal consequence. Some patients require a Rastelli procedure to provide an mLV to pulmonary artery conduit and relieve mLVOTO in the setting of an unrepairable anatomic obstruction. A retrospective cohort study evaluated the outcomes of 32 patients with the mRV functioning as the systemic ventricle by comparing patients with and without a Rastelli procedure. A Rastelli procedure modestly improved 20-year survival (67% vs 62%). This improvement in survival came at a price of higher reintervention (79% vs 53%) 20 years postoperatively [16].

Tricuspid Valve Repair/Replacement

Systemic AV valve regurgitation is a significant source of need for reoperation and a marker for systemic ventricular dysfunction leading to late mortality. The need for systemic AV valve replacement or repair has been reported to range from 31% to 39% at mean follow-up of 11.4–16.9 years [13,14]. Systemic AV valve replacement has been associated with improved survival and functional status with NYHA class I or II status ranging from 88% to 96% at median times of 4.7–16.9 years following replacement [14,15]. A low threshold for recommending systemic AV valve replacement at the earliest signs of progressive systemic ventricular dysfunction is supported by data suggesting that preoperative systemic ventricular ejection fraction greater than 44% was associated with improved survivorship [15]. Once systemic ventricular failure occurs, mechanical support and/or orthotropic heart transplantation are long-term options [8].

In addition to systemic AV valve replacement or repair, overall freedom from reintervention following physiologic repair or ccTGA is low. A series of 32 patients reported a 20-year freedom from reintervention of 32% [16]. The high rate of reintervention with suboptimal long-term survival has led to a search for alternative repair techniques to protect the mRV or remove it from the systemic circulation [16].

ANATOMIC REPAIR

The anatomic repair strategy is based upon the objective to remove the mRV from the systemic circulation and was initially described by Ilbawi in 1990 [18]. The anatomic repair aims to restore blood flow paths through the "normal" sequence of anatomic structures, specifically with the systemic venous drainage to the mRA then mRV to the pulmonary circulation and from the pulmonary venous drainage to the mLA then mLV to the systemic circulation. The anatomic repair is not a single surgical technique, but rather represents a collection of repairs that accomplish this aim. The most frequently performed iteration is the combined atrial switch and arterial switch procedure, often called the "double switch operation" (Fig. 2). When pulmonary valve stenosis is present, arterial switch operation is not feasible and alternatives for the arterial switch component include the Rastelli procedure (Fig. 3), the Nikaidoh procedure, and Réparation à l'Etage Ventriculaire (REV) [19]. When hypoplasia of right ventricular structures are present, a bidirectional cavopulmonary anastomosis can be added (discussed in further detail below). Da Silva et al [20] recommended pulmonary root transfer with 0% in-hospital mortality and 12.5% long-term mortality in patients with ccTGA.

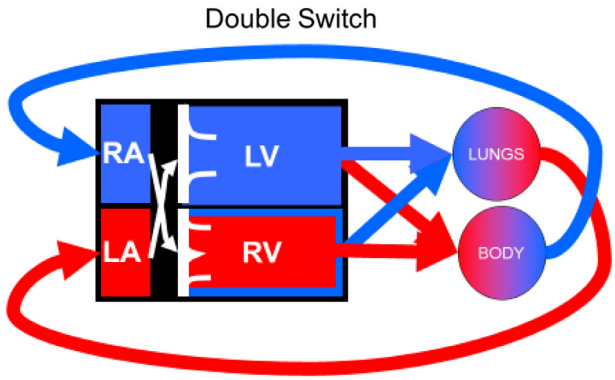

Figure 2 Schematic representation of the anatomic repair in the form of a double switch operation by restoring flow in the normal arrangement, specifically with the systemic venous drainage to the morphologic right atrium then the morphologic right ventricle to the pulmonary circulation and from the pulmonary venous drainage to the morphologic left atrium then the morphologic left ventricle to the systemic circulation. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

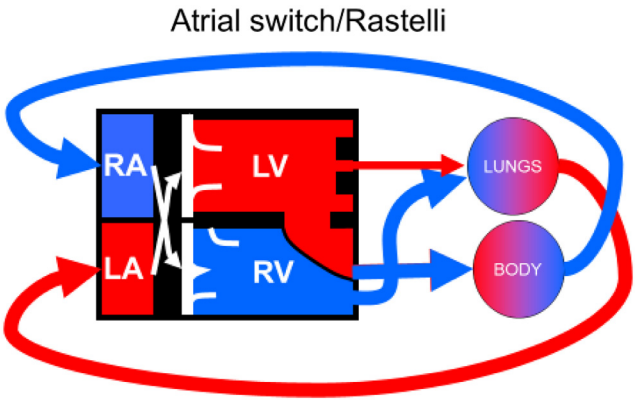

Figure 3 Schematic representation of the anatomic repair in the form of an atrial switch with Rastelli as the arterial switch component when pulmonary stenosis is present. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

Technique for Senning Procedure

The Senning procedure is an atrial switch technique initially published by Senning in 1959 [21]. The key tenets of the Senning procedure are a 3-incision technique with a 3-layer closure. Using only autologous tissue a Y-in-C arrangement of the atria is created with the 2 Y-shaped arms of the systemic venous pathway encircled by the C-shaped pulmonary venous pathway. The first incision is made parallel to Waterston's groove and extends from approximately 1 superior vena cava (SVC) diameter below the SVC to 1 IVC diameter above the IVC (Fig. 4A). The second incision extends superiorly beneath the limbus with a full thickness incision laterally through the

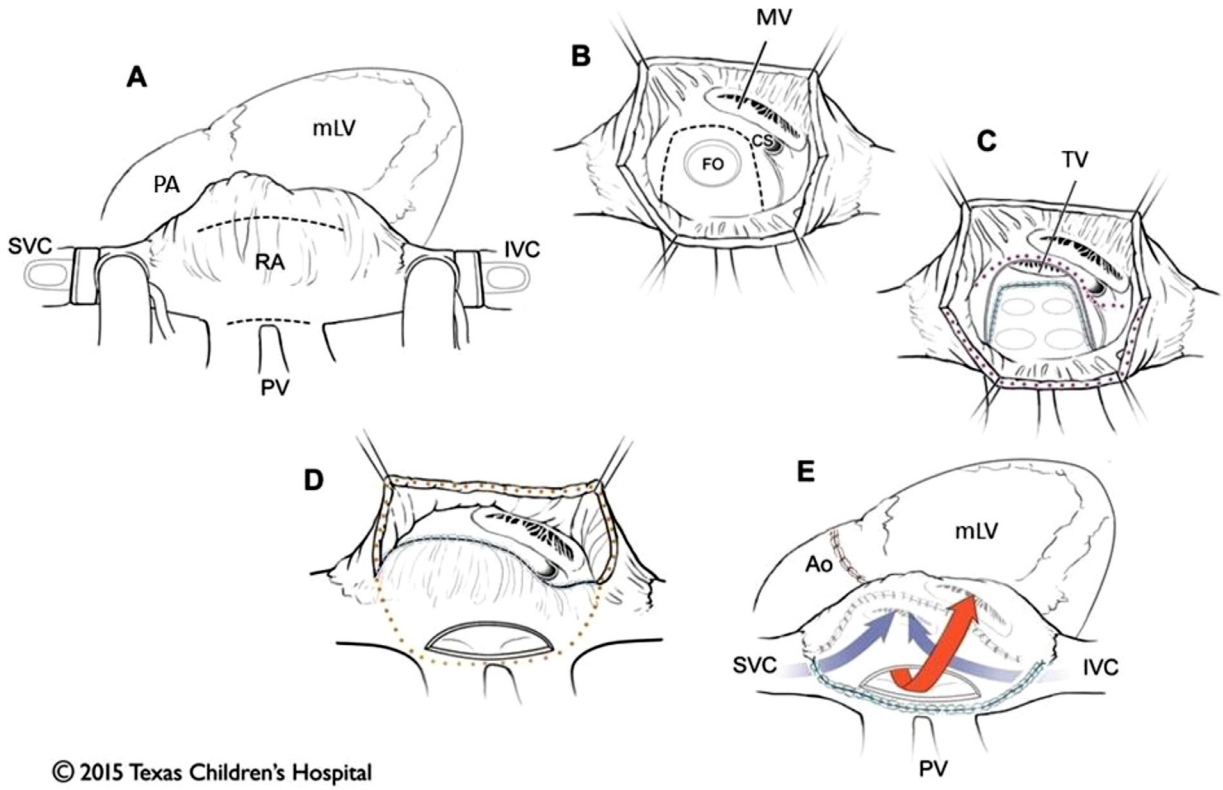

Figure 4 The Senning procedure. (A) Incision sites for the first and third incisions. The first incision extends from the superior vena cava (SVC) to the inferior vena cava (IVC). The third incision creates a lateral opening in to the pulmonary venous (PV) chamber. (B) The second incision extends superiorly beneath the limbus through the limbus to the root of the SVC. Inferiorly, the incision is through the fossa ovalis (FO) toward the lateral wall. (C) The first layer closure brings the hinged flap of interatrial septum deep across the heart. (D) The second layer brings the superior free edge of the right atrial flap to the lateral opening in Waterston's groove. (E) The final Y-in-C configuration of the Senning procedure. Ao, aorta; mLV, morphologic left ventricle; PA, pulmonary artery; RA, right atrium; TV, tricuspid valve. "Printed with permission from Texas Children's Hospital."

limbus to the root of the SVC. Inferiorly, the incision is carried through the medial aspect of the fossa ovalis and across the floor of the atrium toward the lateral wall (Fig. 4B). When performed correctly, this should create a flap of atrium comprising the floor of the fossa ovalis hinged on its attachment to the lateral wall. The third incision can be created by passing a right angle instrument deep to the hinge of the flap such that it can be seen through the thin atrial tissue created by the development of Waterston's groove. By pushing the right angle instrument through this tissue to create an opening, the third incision is created, which can be carried inferiorly, deep to the hinge, in the plane of tissue along Waterston's groove, and thus creating a lateral opening into the pulmonary venous chamber.

Typically, when performed as part of a double switch operation (DSO), the initial incisions are performed, followed by the arterial switch component, with the Senning layers closed last. The first layer closure brings the hinged flap of interatrial septum deep across the heart, isolating the pulmonary veins from the tricuspid valve and atrial appendage (Fig. 4C). To accomplish this, the suture line should begin at the left atrial appendage and extend superiorly and inferiorly to separate the 2 sides. Rarely is additional tissue required to augment this flap. Due to the dog-legged nature of the SVC pathway, care must be taken to avoid narrowing the pathway by using approximately two-thirds of the patch to complete the superior portion of the suture line as deep as possible. Given the enlargement of the mLA due to chronic tricuspid regurgitation and the course of the IVC, it is unlikely that the IVC pathway will become obstructed when using one-third of the flap perimeter to create the inferior suture line to the atrial floor. At this time, the right atrium should be divided by the first incision and the Y-shaped systemic venous pathway should be completed. The lateral portion of the right atrial wall should be folded into the heart, suturing the midpoint to the remnant of the atrial septum, immediately deep to the mitral valve annulus. The suture line should be carried superiorly and inferiorly to meet the caval origins. In contrast to when the Senning procedure is used for d-TGA, in the setting of ccTGA, the inferior suture line can be allowed to travel anteriorly to incorporate the coronary sinus, as the AV node is anteriorly displaced in ccTGA with situs solitus. Naturally, in situs inversus when the

AV node remains in its usual location, the inferior suture line cannot deviate anterior to incorporate the coronary sinus, leaving it in the pulmonary venous pathway. The procedure is completed by bringing the superior free edge of the right atrial flap to the lateral opening in Waterston's groove, thereby closing the C-shaped pulmonary venous pathway (Fig. 4D) [22–24].

Technique for Mustard Procedure

The Mustard procedure is an atrial switch technique initially published by Mustard in 1964 [25]. In contrast to the Senning procedure, the Mustard procedure utilizes a pericardial patch to create the atrial baffle. In the absence of suitable pericardium, synthetic patch material, such as Gore-Tex, may be used. The shape of the patch should be that of a pair of pants using size estimates based on the diameters of the cavae, as suggest by Brom [26]. The angle between the legs of the pants should measure 30° and the length of the front rise of the pants should be equal to the mean of the width of both legs. After initiation of bicaval cardiopulmonary bypass, an oblique right atriotomy is performed from the root of the IVC across the atrial appendage (Fig. 5A). Alternatively, a transverse atriotomy may be made to extend from the junction of the right atrial appendage and right pulmonary veins to the base of the right atrial appendage. At this time, the atrial anatomy is inspected and any remnant atrial septum should be excised (Fig. 5B and C). Resection of the atrial septum anterior to the coronary sinus places the AV node at risk and should be avoided. The coronary sinus may require cutback in the setting of a short distance between the coronary sinus and the right lower pulmonary vein or when a remnant left SVC drains to the coronary sinus. Again, care should be taken to avoid the AV node; therefore, the scissors should be placed into the coronary sinus and directed posteriorly into the left atrium when making this incision. Next, the patch is sewn into the heart with the middle of the waist of the pants sewn between the left-sided pulmonary veins (Fig. 5D). The inferior suture line travels around the left lower pulmonary vein and below the right lower pulmonary vein. The superior suture line goes around the left upper pulmonary vein toward the upper edge of the orifice of the

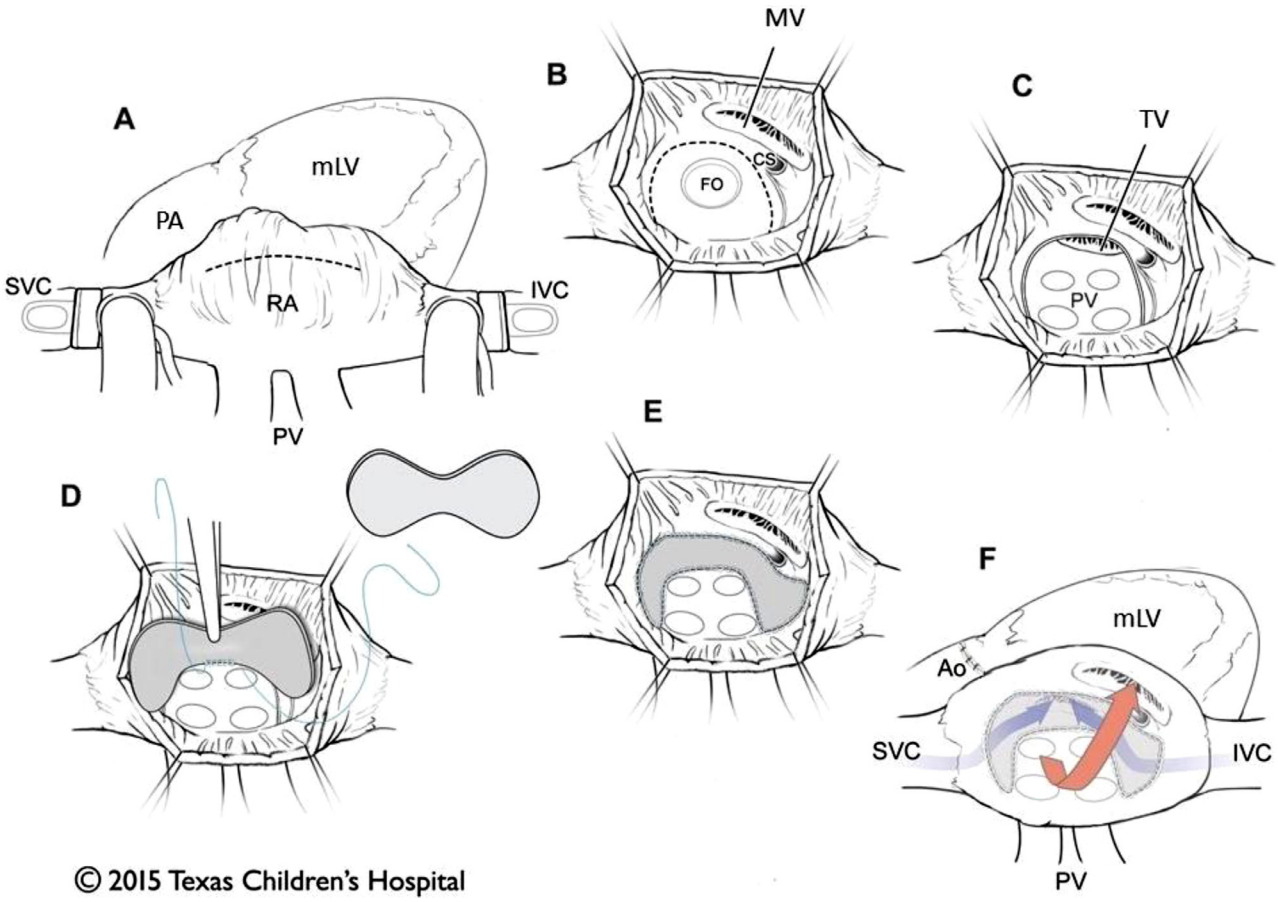

Figure 5 The Mustard procedure. (A) An oblique right atriotomy is performed from the root of the inferior vena cava (IVC) across the atrial appendage. (B) Any remnant atrial septum should be excised. (C) View into the left atrium following excision of the atrial septum. (D) The patch is sewn into the heart with the middle sewn between the left-sided pulmonary veins (PV). (E) Completion of the patch suture lines, incorporating both vena cavae into the systemic pathway with exclusion of the pulmonary veins. Ao, aorta; FO, fossa ovalis; mLV, morphologic left ventricle; MV, mitral valve; PA, pulmonary artery; RA, right atrium; SVC, superior vena cava; TV, tricuspid valve. "Printed with permission from Texas Children's Hospital."

right upper pulmonary vein. Great care should be taken with this suture line as carrying the patch too low in the atrium can tent the left pulmonary veins leading to obstruction, whereas deviating too high on the atrium, toward the cut edge of the septum, can lead to obstruction of the SVC pathway. The patch should not place tension on the left atrium as the suture lines being too close to each other or too far apart can lead to pulmonary venous obstruction. The 2 legs of the patch diverge toward their respective cavae. The crotch point of the patch should then be approximated to the edge of the atrial septum and the lower edge of the patch is sewn to the cut edge of the atrial septum and across the floor of the coronary sinus (Fig. 5E). If the coronary sinus was not opened, it is excluded from the systemic venous pathway. Again, care should be taken to avoid placing sutures anterior to the coronary sinus as this can damage the AV node, particularly in situs inversus. If concern for obstruction of the pulmonary venous pathway exists after completion of the patch, or if the mRA is known to be of low volume, the mRA can be enlarged by placing a wide pericardial or synthetic patch across the initial atriotomy incision with the intent to create space between the upper and lower pulmonary veins. When performed in conjunction with a bidirectional cavopulmonary anastomosis, the design of the patch is simplified to create a baffle between the IVC and tricuspid valve. This reduces the risk of SVC and pulmonary venous obstruction as the SVC limb of the baffle is the Achilles' heel of the Mustard procedure [23].

Technique for Arterial Switch Operation

The arterial switch operation was initially reported by Jatene in 1976 [27]. While atrial cannulation may be performed for d-TGA, in the setting of ccTGA where an atrial switch procedure will be performed, bicaval cardiopulmonary bypass will be established. If not previously performed, the Patent Ductus Arteriosus (PDA) or ligamentum arteriosum should be ligated and divided. The pulmonary arteries are dissected to the level of the hilar branches. The anticipated location of the coronary artery translocation onto the proximal main pulmonary artery should be determined and marked with 2 marking stitches. The ascending aorta is divided across from the pulmonary bifurcation, approximately at its midpoint. The coronary arteries are excised with an arterial button, often with the majority of the respective sinus of Valsalva attached. The proximal several millimeters of each coronary artery should be mobilized with care to preserve all branches until the buttons reach the anticipated reimplantation sites tension-free. The main pulmonary artery is then divided immediately proximal to the bifurcation. It is also at this time that any surgically relievable mLV outflow tract obstruction is resected. With adequate mLV outflow, the decision should be made to pursue either 2 medially based trap door flaps or 2 U-shaped resections at the location of the original coronary artery implantation sites. If trapdoor incisions are created, the required rotation of the coronary artery pedicles will be decreased at the cost of increasing the already large circumference of the neoaortic root. Alternatively, the U-shaped incisions are created in such a way that the bottom of the incisions are in line with the crests of the neoaortic valve commissures; however, extreme caution must be taken to avoid any rotation or twisting of the coronary buttons. After assuring a water-tight suture line without distortion of the coronary ostia, the ascending aorta can be anastomosed to the neoaortic root in a running continuous fashion. Mattress sutures should be used to fortify all points of overlapping suture lines. The neopulmonary root sinus defects created by the coronary artery transfer are closed with either fresh or treated pericardium and the pulmonary artery to neopulmonary root anastomosis is completed in a running fashion. While this anastomosis would be commonly performed during the rewarming phase of an arterial switch operation for d-TGA, bypass can be continued if completion of the atrial switch component is required [23,24].

Technique for Rastelli

The Rastelli technique for arterial switch in the setting of pulmonary stenosis was published by Rastelli in 1969 [28]. The patient should not be a candidate for an arterial switch operation with unresectable mLV outflow tract obstruction. If an interventricular conduit is not feasible in this setting, a single ventricle palliation strategy should be used. It is therefore of critical importance that the ability to revert to a bidirectional cavopulmonary shunt or Fontan procedure is not negated until the intracardiac anatomy is confirmed and septation can be achieved. The patient is cannulated for bicaval cardiopulmonary bypass with the SVC cannula at or in the innominate vein and the IVC cannula as inferior as possible in preparation for the atrial switch portions of the procedure. The mRA is opened in the fashion appropriate for the atrial switch operation, as described above, to be utilized later. Through the mitral valve, the VSD is inspected for the presence of AV valve tissue that may preclude formation of an interventricular baffle. Once the procedure is determined to be feasible, a high, oblique mRV ventriculotomy is performed. Any large muscle bundles noted at the site of ventriculotomy will require resection to avoid delayed obstruction of the neopulmonary outflow tract. Through this ventriculotomy, the VSD is baffled to the aorta using treated pericardium. If adequate pericardium is unavailable due to prior operations or needed as the baffle in a Mustard operation, a synthetic baffle, such as Dacron, may be used. At this time, the atrial switch operation of choice should be performed. An aortic or pulmonary homograft or alternative valved conduit is thawed and prepared for anastomosis. Caution should be taken to avoid creating a lengthy conduit, which will be at risk for kinking when filled with blood. The main pulmonary artery should be divided and the pulmonary valve oversewn and the main pulmonary artery is closed in 2 layers. The distal end of the thawed homograft is anastomosed in an end-to-end fashion to the distal pulmonary artery. In the setting of levocardia, the homograft should lay to the left of the aorta; however, this may not be achievable with dextrocardia or mesocardia. The proximal end of the pulmonary homograft

is anastomosed to the superior aspect of the ventriculotomy. The anastomosis should be completed only after the left heart is allowed to fill and the ventriculotomy is used as a vent. A concomitant Damus-Kaye-Stansel anastomosis has been proposed [29] in adjunct to the Rastelli procedure for patients with variance in size between the VSD and aortic annulus. This additional carries a reported 100% survival in the modern era [23,28].

Technique for Nikaidoh

The posterior aortic translocation was first described by Nikaidoh in 1984 for transposition in the setting of pulmonary stenosis [30]. The Nikaidoh procedure begins with transection of the aorta and pulmonary artery above their respective roots. The aorta does not require complete transection if root rotation is not required. In cases of incomplete transection, the aorta can remain attached in the area under the left coronary artery. In both cases of complete or incomplete transection, the coronary arteries can remain attached to the aortic root with enough mobilization, predominantly of the right coronary artery, to perform the posterior translocation. Depending on the coronary artery anatomy, excision of coronary arteries with a button of aortic root may be required to preventing twisting or kinking of the coronary arteries in a manner analogous to that of the arterial switch operation, explained previously. The aortic root and valve are then excised from the mRV outflow tract. The pulmonary root and valve are subsequently excised from the mLV outflow tract. At this point, any required excision of superfluous tissue from the ventricular outflow tracts should be performed. The conal septum is routinely excised including the superior margin of the VSD, if present. The aortic root is translocated to the mLV outflow tract and anastomosed to the ventriculotomy. If the coronary arteries required excision prior to root transfer, they should be reattached. Regardless of whether the coronary arteries required revision or were left in situ, the ostia and proximal extent of both coronary arteries should be probed to assure patency. The aortic root-ascending aorta continuity is then re-established in an end-to-end fashion. Patch closure of the VSD can be performed using pericardium or synthetic material as preferred. If the pulmonary valve is suitable for use, the pulmonary root is anastomosed to the mRV outflow tract in the portion of the ventriculotomy aligned with the mRVOT. If the pulmonary valve is not suitable for use, an aortic or pulmonary homograft or valved xenograft can be used in the same manner as in a Rastelli procedure. With the main pulmonary artery anastomosis completed, attention can be returned to the atrial switch procedure of choice [19,23].

Technique for REV

The REV procedure was introduced in 1982 by LeCompte et al as an alternative technique for arterial switch with pulmonary stenosis that does not require a prosthetic conduit [31]. After establishment of bicaval cardiopulmonary bypass, the aortic root and main pulmonary artery are transected. The pulmonary valve is oversewn with the main pulmonary artery root closed in 2 layers. The aortic root to ascending aorta continuity is re-established in an end-to-end fashion. An infundibular morphologic right ventriculotomy is developed with care to avoid the infundibular branch of the right coronary artery. Through this ventriculotomy, an interventricular pericardial or synthetic patch is used to create the neoleft ventricular outflow tract, thereby closing the VSD to the mRV. The hallmark of the REV operation is the direct anastomosis of the main pulmonary artery to the right ventricle, though alternatives including the use of a prosthetic conduit or portion of the ascending aorta to augment the anastomosis have been proposed. In a classical REV procedure, the distal main pulmonary artery is directly anastomosed to the infundibular incision beginning at the posterior aspect of the artery. A second pericardial or synthetic patch is used to augment the anastomosis, creating an open morphologic right ventricular outflow tract [19].

Results

Survival rates following anatomic repairs were variable across studies reporting actuarial survival. Survival at 1 year was 81−100% [32−38], 5-year survival was 80.5−100% [32,34,36,37,39−41], 10-year survival was 62−100% [32,34,36−38,41,42], 15-year survival was 95% [43], and 20-year survival was 70.2−100% [29,32,44]. The DSO had a 0−14% in-hospital mortality [32,36,37,42,45,46]. The critique of the DSO is its poor early mortality; however, the long-term survival is acceptable with 20-year survival of 83.3−100% [32,44]. Several studies directly compared long-term survival between DSO and atrial switch with Rastelli. In these comparisons, long-term survival ranged from no different [33,42] to better with Rastelli [36,40] to improved early survival with Rastelli, but better late survival with DSO [37].

Neoaortic regurgitation is a long-term complication of the arterial switch procedure. When compared to patients with d-TGA, patients with ccTGA had lower long-term survival, more neoaortic regurgitation, the neoaortic regurgitation was more significant, and they required more aortic valve replacements. Additionally, a sinotubular junction reconstruction was shown to have benefit in patients with large discrepancies between their aortic and pulmonary valves [47].

As described, tricuspid regurgitation is a significant risk factor for late mortality in the ccTGA cohort. Patients with significant preoperative tricuspid regurgitation or an Ebsteinoid tricuspid valve were evaluated, comparing patients who had a tricuspid valvuloplasty during anatomic repair to patients who did not have a tricuspid valvuloplasty during anatomic repair. Patients who had a tricuspid valvuloplasty had an absolute risk reduction for late tricuspid regurgitation of 16.7% in the tricuspid regurgitation group and 22.2% in the Ebsteinoid group at a median of 32 months following repair [48].

PULMONARY ARTERY BANDING

In ccTGA, the mLV is in the subpulmonary position and, in the absence of outflow tract obstruction, will be exposed to

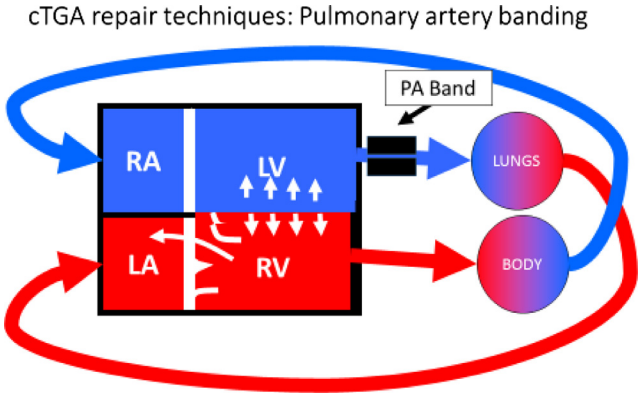

Figure 6 Schematic representation of pulmonary artery banding to train the morphologic left ventricle in anticipation for a future double switch operation. LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle.

low pulmonary blood pressures with eventual decline in the ability to achieve pressures equivalent to aortic pressure. Consequently, pulmonary artery banding has been suggested as a method to train the mLV to accommodate aortic pressures prior to an arterial switch [48] (Fig. 6). Prophylactic pulmonary artery banding has been suggested for mLV retraining and improvement or stabilization of tricuspid valve, aortic valve, mRV, and mLV function [49–52]. The mLV loses potential for successful training as the patient ages, although the threshold at which mLV cardiomyocyte adaption from hyperplasia to hypertrophy disappears is not well defined. Winlaw et al [52] identified patients older than 16 years as unlikely to be suitable for arterial switch due to training failure. Poirier et al [53] found a lower cutoff at 12 years old. Myers et al [50] described a 67% prevalence of mLV dysfunction at 21 months postrepair for patients aged 2–23 years; however, the prevalence decreased to 0% in patients who had a pulmonary artery band placed when younger than 2 years old. Helvind et al [54] recognized failure of the mLV to be retrained with pulmonary artery banding as an important risk factor associated with mortality.

The protocol for pulmonary artery banding varies between centers. Some centers recommend prophylactic banding at diagnosis, citing the potential for banding as a long-term palliation strategy in addition to retraining [49,51,55], while others recommend pulmonary artery banding for patients with mLV pressure less than 75% systemic [50,52,56]. Once the band is in place, there have been a variety of management modalities suggested, including dilatable bands to avoid additional operations [49,57], surgical replacement prior to repair [56], or use of B-type natriuretic peptide to predict myocardial tolerance of pulmonary artery banding [58]. Pulmonary artery banding in ccTGA is associated with significant mortality (5.3%) [49–53,56,57,59]. In patients undergoing DSO after pulmonary artery banding for mLV retraining, operative mortality ranged from 0% to 17% [49,52,53,56,57,59,60]. Late mortality in patients who had prerepair pulmonary artery banding for mLV retraining ranged from 76% to 91% [49,52,53,60].

Several proponents of palliative pulmonary artery banding exist, citing a stabilization in tricuspid valve, aortic valve, mRV, and mLV function [50–52]. In the series of 20 patients by Cools et al [51] looking at palliative pulmonary artery banding in patients with ccTGA and VSD or with d-TGA after atrial switch, there were no mortalities and patient's functional status remained stable or improved. Ma et al [55] compared patients with deconditioned mLV who had a pulmonary artery band initially placed that subsequently either maintained the palliative pulmonary artery band or went on to have an anatomic repair. Patients who maintained the palliative pulmonary artery band had better overall survival, greater proportion in NYHA class I or II, and higher mean mLV ejection fraction than the anatomic repair group. Age at pulmonary artery banding was associated with late postrepair mortality and mLV dysfunction after anatomic repair. Based on these findings, the authors suggested that pulmonary artery banding may be the ideal procedure for patients with deconditioned mLV. Winlaw et al [52] noted that while the pulmonary artery band improved the function, it did not reduce tricuspid regurgitation in patients prior to repair.

The results of pulmonary artery banding were not always positive. In a retrospective cohort study comparing patients after anatomical repair with prior pulmonary artery banding compared to patients who did not undergo pulmonary artery banding, Brawn et al [61] found no difference between groups in actuarial freedom from death or transplant; however, the group that had mLV retraining prior to repair had greater actuarial transplant, death, or moderate-to-severe mLV dysfunction. In another retrospective cohort study comparing the same groups, Quinn et al [60] identified greater actuarial freedom from death or transplant with good mLV function in patients who did not have prior mLV retraining before anatomic repair. Sun et al [62] found incremental decreases in mean mLV ejection fraction 1 week following pulmonary artery band placement and after anatomic repair (median 21 months following pulmonary artery band placement) with associated decreases in mean global strain and mean lateral wall strain. A fourth study designed to compare multiple procedures identified a 79% transplant-free survival in patients with palliative pulmonary artery banding at 10 years, the lowest among all groups [63].

FONTAN PALLIATION

For patients with ccTGA who do not have a septatable heart due to ventricular size, ventricular function, or associated defects, single ventricle palliation with a Fontan procedure is often the only viable option. It has been noted that patients have relatively favorable outcomes following the Fontan procedure piquing interest in single ventricle palliation for patients traditionally believed to be candidates for biventricular repair. At least 7 studies [7,63–67] have compared the outcomes of patients with Fontan palliation to physiologic and anatomic repairs. In 119 patients identified in these studies, the 10-year actuarial survival ranged from 90.9% to 100% [63–66],

including 100% in 2 of the 4 studies. Additionally, where studied, Fontan palliation was protective from the high reintervention rates of the other repair pathways, with a 91.3% freedom from reintervention ratio. Based on their experience with all repair techniques, Hsu et al [65] recommended Fontan for any patient with complex anatomy, even if traditionally believed to be a biventricular candidate. Talwar et al [67] evaluated outcomes of 23 patients with ccTGA, VSD, and pulmonary stenosis or atresia, finding an 81.8% event-free 10-year survival with NYHA class I status in 85.7% of survivors, concluding that Fontan may be the optimal procedure for this challenging subset of patients. Given the high rate of heart block in patients with ccTGA, Simmons et al [7] compared univentricular palliation to biventricular repair and isolated ccTGA, finding that patients in the Fontan pathway were less likely than the other cohorts to have complete heart block; however, the rate of spontaneous heart block remained uniform across all groups.

ONE-AND-A-HALF VENTRICLE PALLIATION

The atrial switch component of the anatomic repair, through Senning or Mustard procedure, carries risk of long-term complication from systemic venous obstruction. One of the proposed techniques to minimize risk of systemic venous obstruction is to perform a bidirectional cavopulmonary anastomosis at the time of anatomic repair. There is an additional benefit in this technique in that there is avoidance of volume loading of the mRV, theoretically decreasing mRV dilation, tricuspid insufficiency and trans-septal strain of the mLV in the setting of increased mLV afterload. Two studies [45,68] with a combined cohort of 39 patients investigated the use of bidirectional cavopulmonary anastomosis in conjunction with a hemi-Mustard procedure at anatomic repair with a combined 5% in-hospital mortality, but zero late mortalities or transplants at 4.5 and 4.9 years of follow-up. Complications of the bidirectional cavopulmonary anastomosis occurred in 5% of the survivors, without sinus nodal or intracardiac baffle complications. A third study [69], including 19 patients with bidirectional cavopulmonary anastomosis with either a physiologic or anatomic repair concluded that the addition of a bidirectional cavopulmonary anastomosis increased the pool of candidates for biventricular anatomic repair with acceptable long-term outcomes. One-and-a-half ventricle palliation is a reasonable option in patients with inadequate mRV for complete septation; however, longer term follow-up and larger study populations are required before benefit over single ventricle palliation can be truly delineated.

COMPARISON OF PROCEDURES

There have been several attempts made at comparing outcomes from the various repair options. The outcomes for physiologic and anatomic repairs are comparable. Shin'oka et al [66] is the only paper to identify a trend toward survival benefit to anatomic repair compared to physiologic repair, with lower tricuspid regurgitation rates in the anatomic repair group making it the more favorable repair. In spite of the physiologic repairs' greater than 25% survival benefit over anatomic repair, even after adjustment for early mortalities, Hsu et al [65] recommend anatomic repair for highly favorable anatomy with all other patients receiving a Fontan repair. Lim et al [69] advised for anatomic repair with bidirectional cavopulmonary anastomosis in patients with an inadequate mRV.

Two studies found that the physiologic and anatomic repairs were equivalent in the 10-year transplant-free survival ranging 83.3−93% in the physiologic cohort and 77.3−86% in the anatomic repair cohort [63,64]. Systemic ventricle dysfunction and systemic AV valve insufficiency were identified as risk factors for death or transplant with equivalent rates between repairs [63]. Lower rates of reoperation were identified in the anatomic cohort at the price of worse ventricular function and AV valve competency compared to the physiologic cohort [60].

One study [7] was specifically designed to assess the incidence of complete heart block following the various repairs. Of 64 patients with isolated ccTGA, biventricular ccTGA, or univentricular ccTGA, 21.9% of patients developed complete heart block at a rate of 1.3% per 100 person-years of follow-up. As previously discussed, the rate of spontaneous complete heart block did not vary between groups; however, a Fontan palliation strategy had a lower overall incidence of complete heart block.

Alghamdi et al [3] recognized the inadequacy of these comparisons through meta-analysis due to the observational nature of these studies. Patient cohorts divided by the various procedures differed significantly in era of operation, age, right-sided abnormalities, and preoperative arrhythmia status, skewing comparison. The meta-analysis of observational studies comparing repair strategies concluded that the various repairs may each have exclusive patient cohorts who would be ideal candidates for the technique of repair. An editorial commentary by Tweddell et al [70] suggested a repair strategy of physiologic repair or Fontan palliation for patients with good mRV, no tricuspid regurgitation, and a nonrestrictive VSD at repair. Patients with tricuspid regurgitation or mRV dysfunction, but with a good mLV would benefit from anatomic repair. For patients with anatomy that is too complex for biventricular repair, Fontan physiology remains a reasonable option for palliation. Regardless of repair technique, surgical techniques require refining to improve outcomes for patients with ccTGA [71].

CONCLUSION

The optimal surgical repair of patients with ccTGA remains largely unclear. Key preoperative considerations such as morphologic left ventricular pressure, tricuspid valve competency, and outflow tract obstructions can assist in determining the optimal repair for individual patients that would provide the best long-term outcomes. An alternative, single ventricle, pathway has been proposed for any patient without optimal preoperative anatomy to improve long-term survival. Adjunctive repair options including pulmonary artery banding and one-and-a-half ventricle repairs have also been proposed to augment the survival curves.

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