Late reoperations for Fontan patients: state of the art invited review

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Summary
Conversion of the atriopulmonary Fontan to a total cavopulmonary extracardiac connection with concomitant arrhythmia surgery and pacemaker placement is a safe and efficacious procedure for this patient population. From 1994 to 2007 a total of 118 patients have undergone this procedure with one (0.8%) early and nine (7.6%) late deaths. During the course of our experience with Fontan conversion our surgical strategy has evolved to include various ablative techniques to treat macro re-entrant atrial tachycardia, focal (automatic) atrial tachycardia, atrioventricular nodal reentry tachycardia, atrial tachycardia due to accessory connections, atrial fibrillation, and ventricular tachycardia. The various mechanisms that we use to treat the underlying atrial arrhythmias are described in this review. We have also encountered patients with variations of the Fontan and other complex anatomic and pathophysiologic aberrations who were not amenable to standard takedown and ablative procedures. We describe those circumstances and the solutions we found to treat those patients.

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1. Introduction
Re-operations in patients with the various forms of extant Fontan connections require innovative procedures to manage anatomic, pathophysiologic, and electrophysiologic complications that may impair the Fontan circulation. We and others have demonstrated that conversion from an atriopulmonary to a total cavopulmonary artery extracardiac Fontan with concomitant arrhythmia surgery and pacemaker placement is a safe and efficacious operation in this increasingly complex patient population [1—15]. The purpose of this invited review is to discuss the various arrhythmia mechanisms, present our current surgical strategy, and highlight the various interventions that are required to treat differing anatomic and arrhythmia substrates.

2. Mechanisms of arrhythmias and surgical therapeutic options
2.1. Macro re-entrant atrial tachycardia (atrial re-entry tachycardia, atrial flutter)
Macro re-entrant atrial tachycardia is a macro re-entrant rhythm disturbance of the right or left atrium (Fig. 1A). The area of interest for this arrhythmia requires unidirectional block and an area of slow conduction in order to establish a re-entrant circuit. A premature atrial contraction can propagate an electrical wave front that encounters unidirectional block near the atrial septum, and is redirected inferiorly by an alternative route, to the right atrial free wall. The electrical wave front encounters an area of slow conduction, usually at the area between the inferior vena cava, tricuspid valve, and the coronary sinus. The electrophysiologic delay allows the area of unidirectional block to recover conduction. The electrical wave front now enters opposite the area of unidirectional block that establishes the re-entrant circuit. Cryoablation lesions are used to interrupt this circuit at the inferior isthmus, defined as the area between the coronary sinus, tricuspid annulus, and the inferior vena cava. The cryoablation lesion, therefore, transforms an area of slow conduction to an area of no
conduction, thereby eliminating the circuit. Multiple areas of slow conduction may exist in patients with congenital heart disease from prior incisions, patches, or wall stress.

2.2. Atrioventricular (AV) nodal re-entry tachycardia

AV nodal reentry tachycardia is a re-entrant circuit in the area between the AV node, coronary sinus, and inferior vena cava associated with unidirectional block in one direction and an area of slowed conduction in the other (Fig. 1B). Atrioventricular conduction encounters unidirectional block in the normal fast pathway fibers superior to the AV node. The electrical wave front is redirected towards the atrial isthmus (between the coronary sinus and tricuspid valve) and encounters the slow pathway fibers of the AV node. After the impulse exits the isthmus, conduction now re-enters the fast pathway, anteriorly and superiorly, and establishes the re-entrant circuit. This accelerated cycle length proceeds to the ventricles, which establishes the tachycardia. Cryoablation of the inferior isthmus region interrupts the circuit.

2.3. Accessory connection-mediated atrial tachycardia

Wolff–Parkinson–White (WPW) syndrome is associated with the delta wave on electrocardiography and occurs during sinus rhythm with pre-excitation. Conduction proceeds simultaneously from the sinus node to the ventricles over two routes, the AV node and the accessory connection. The electrical wave front traverses the accessory connection and depolarizes ventricular tissue first, because of intrinsic slowing of conduction at the AV node. This is the manifest form of accessory connections (WPW). In order for a re-entrant tachycardia to occur, there must be unidirectional block in one pathway with slowed conduction in the other. Fig. 2A shows unidirectional block in the AV node, with conduction proceeding antegrade to the ventricles via the accessory connection. The ventricular muscle is responsible for conduction delay, allowing the wave front to proceed up the AV node (which has had time to regain conduction) to the atrium. This re-entrant circuit is termed antidromic reciprocating tachycardia; the area of interest includes the atria, accessory connection, ventricles, and the AV node.

The more common form of tachycardia in association with an accessory connection is called orthodromic reciprocating tachycardia. Fig. 2B illustrates conduction block in the accessory connection, thus resulting in a lost (concealed) delta wave. Conduction proceeds normally through the AV node to the ventricle. The delay encountered in the AV node allows the accessory connection to regain electrical function. The electrical impulse then enters the atria from the opposite direction. This area of interest is similar to antidromic reciprocating atrial tachycardia except that the electrical pathways are reversed and concealed.

2.4. Focal (automatic) atrial tachycardia

Focal atrial tachycardia is a localized area of electrical impulse generation. This automatic mechanism fires repeatedly, rapidly, and independently of normal sinus function, which is inhibited (Fig. 2C). Electrical impulse conduction is propagated over the body of the atrial, which results in stimulation of the AV node and ventricles. Cryoablative therapy and resection is used to obliterate or isolate this localized discrete area.

Re-entrant atrial circuits tend to be multiple in patients with varying anatomy and complex congenital heart disease. This led to the development of the modified right atrial maze procedure and the modified Cox maze III procedure (Figs. 3 and 4) [16]. Fig. 3A demonstrates the possible lines of ablation that can be used to treat macro re-entrant atrial tachycardia in the presence of various atrial anomalies. These include juxtaposition of the atrial appendages, total anomalous pulmonary venous connection, separate atrial entry of the hepatic veins, persistent left superior vena cava, and absence of the coronary sinus. The most common anatomic variances of single ventricle that require adaptation of the right-sided maze procedure are tricuspid atresia, mitral atresia, and common atrioventricular valve. Fig. 4A–C show the cryoablation lesions that are applied to areas of slow conduction in these patients [6].

Atrial fibrillation is characterized by uncoordinated atrial activation with consequent deterioration of atrial mechanical function. On ECG, consistent P waves are replaced by rapid oscillations or fibrillatory waves that vary in amplitude, shape, and timing. This is associated with an irregular, frequently rapid ventricular response when atrioventricular conduction is intact. The Cox maze III procedure was
developed to treat atrial fibrillation and has been modified for patients with single ventricle, as noted in Fig. 3B.

### 2.5. Ventricular tachycardia

Ventricular tachycardia is an abnormal ventricular rhythm with a rate greater than 120 bpm in adolescents or adults, or a rate greater than 150 bpm in children. This arrhythmia arises from various locations in either the left or right ventricles. Since there is no specific localized origin of the tachycardia, treatment with cryoablative lesions or endocardial/epicardial resection must be individualized to the patient.

### 3. Resternotomy

At the time of Fontan conversion, adhesions from previous operations, dilated right atria, and long-standing abnormal physiology of the atropulmonary circuit all contribute to the difficulty of sternal re-entry. In rare cases, the femoral vessels can be dissected before the resternotomy for rapid cannulation and institution of cardiopulmonary bypass when there is high risk for cavitary entry. Early identification of the aorta and atrium is achieved so that rapid institution of cardiopulmonary bypass can be accomplished in the event of unwanted mishaps during dissection. To avoid injury to the phrenic nerves the dissection plane should remain medial to the pericardium and entry into the cardiac chambers is to be avoided to reduce the risk of paradoxical air embolus occurring through residual intracardiac shunts. Careful preoperative planning and extreme caution during the resternotomy should insure a safe outcome for the patient [17].

### 4. Conduct of the operation

After the sternum is opened we proceed to aortic and direct vena cava cannulation using right-angle cannulas inserted high in the superior vena cava and straight cannulas in the inferior vena cava. After the commencement of cardiopulmonary bypass the inferior vena cava is transected and anastomosed to a 24 mm Gore-Tex (W.L. Gore & Associates, Flagstaff, AZ) tube graft. A vent is placed in the right superior pulmonary vein, the aorta is cross-
clamped, and cold blood cardioplegia is delivered. With the heart arrested, an atrial septectomy and resection of the right atrial appendage are performed. Linear cryoablation lesions for the right-sided maze procedure are placed with a Surgifrost device (CryoCath Inc., Kirkland, Quebec, Canada) applied at −160 °C for 1 min. The lesions extend from the base of the right atrial appendage to the cut edge of the atrial septal defect, across the crista terminalis from the cut edge of the atrium to the atrial septal defect, and from the transected inferior vena cava to the coronary sinus. In addition, if a tricuspid valvar orifice is present, a lesion is created from the inferior vena cava to the annulus of the tricuspid valve. We also place a lesion to prevent macro re-entrant circuits extending from the coronary sinus to the edge of the atrial septal defect. In patients with atrial fibrillation, we perform the Cox maze III procedure. The left atrial appendage is excised and an encircling pulmonary venous isolation is performed using cryoablation incisional techniques. A cryoablation lesion is placed from the cut portion of the excised left atrial appendage to the confluence of the pulmonary veins. An additional lesion is placed from the encircling atriotomy lesion to the annulus of the mitral valve, directing it at the infero-medial scallop. A lesion, created for 2 min, is placed on the epicardial surface of the coronary sinus directly opposite that involving the mitral valve. We also place a lesion to prevent macro re-entrant circuits extending from the coronary sinus to the edge of the atrial septal defect. In patients with atrial fibrillation, we perform the Cox maze III procedure. The left atrial appendage is excised and an encircling pulmonary venous isolation is performed using cryoablation incisional techniques. A cryoablation lesion is placed from the cut portion of the excised left atrial appendage to the confluence of the pulmonary veins. An additional lesion is placed from the encircling atriotomy lesion to the annulus of the mitral valve, directing it at the infero-medial scallop. A lesion, created for 2 min, is placed on the epicardial surface of the coronary sinus directly opposite that involving the mitral valve. At the conclusion of the Cox maze III procedure, the left atrium is closed with running polypropylene, the heart de-aired, and the cross-clamp removed.

5. Pacemaker implantation

Because of scarring from previous operations epicardial pacemaker leads can present significant challenges. We have found that the atrial lead can be placed at the dome of the left atrium beneath the ascending aorta or near the right atrioventricular groove. For the ventricular pacemaker lead we place it on the diaphragmatic surface of the heart where there are fewer adhesions. Evolution of pacemaker technology has provided more options for postoperative arrhythmia management. Recently, we have implanted atrioventricular antitachycardia dual-chamber pulse generators, multisite ventricular pacing for dyskinetic ventricles, and automatic cardiac defibrillators for ventricular tachycardia [14].

In our experience with Fontan conversion we have encountered a number of patients with complex connections established at the original Fontan operation that resulted in an anatomic configuration that required individual correction for optimal Fontan physiology. Following are some examples of these cases [13].

6. Anatomic and electrophysiologic variations of Fontan conversion

6.1. Takedown of right atrial-to-right ventricular Bjork modification

Conversion of a Bjork–Fontan modification (right atrial to right ventricle connection) to total cavopulmonary artery extracardiac connection presents the surgeon with two dilemmas: (1) takedown of the Bjork–Fontan anastomosis, and (2) management of the right ventricle. A posterior reversed right atrial flap and an anterior prosthetic patch usually forms the right atrial to right ventricular connection in most patients without a bioprosthetic valve. To accomplish right atrial wall reduction and epicardial pacemaker lead placement careful dissection of the right atrioventricular groove is required. The dissection begins near the ascending aorta where an undissected plane is readily achieved and continues as far as possible toward the posteroseptal area as long as the dissection plane allows. Care is taken to perform this dissection with a low electrocautery setting to avoid unwanted injury to the right coronary artery. In two patients in our series the right coronary artery was entered. In the first patient the injury was caused by electrocautery dissection. We initiated cardiopulmonary arrest and performed Gore-Tex arterio-plasty. The second injury was caused by sharp dissection and was repaired with cardiopulmonary arrest and interrupted suture technique. Both injuries were repaired without sequelae.

The dilemma of what to do with the right ventricle poses a challenge because disconnecting the main pulmonary artery would leave the right ventricle without an egress of blood flow that would eventually lead to right ventricular dilatation and leftward interventricular septal compression of the left ventricle. We have approached this dilemma by preserving right ventricular to main pulmonary artery continuity. We have found that the developed right ventricular pressure is low under these circumstances because of decreased right ventricular pre-load and does not affect the non-pulsatile blood flow established by the total cavopulmonary extracardiac connection. Fig. 5A–C demonstrate conversion of a Bjork–Fontan modification to a total cavopulmonary extracardiac connection.

6.2. Pulmonary atresia with intact ventricular septum after atriopulmonary Fontan

Two patients in our series had pulmonary atresia and intact ventricular septum that were treated with an atriopulmonary Fontan procedure and main pulmonary artery disconnection. Over time the right ventricle became hypertrophied and the patient developed supra-systemic right ventricular pressure. Because of the significant tricuspid regurgitation, the patient developed right atrial hypertension and dilatation with atrial arrhythmias. An extracardiac total cavopulmonary connection without addressing the tricuspid valve would increase the resultant left common atrial pressure and cause circulatory compromise. We performed the modified right-sided maze procedure and isolated the tricuspid valve with a fenestrated (8 mm) Gore-Tex patch, which restricted blood flow into the right ventricle and decreased developed pressure by volume unloading. The fenestration also allowed blood egress into the common atrium during diastole. In these patients the developed right ventricular pressure was significantly reduced and had no effect on the common left atrial pressure (Fig. 6A).

6.3. Takedown of tricuspid valve isolation patch for right-sided maze procedure

In patients who had a previous Fontan procedure for double inlet ventricles, criss-cross hearts, or straddling
atrioventricular valves, the smaller right-sided atrioventricular valve was oftentimes isolated with a patch when the left-sided atrioventricular valve was large enough and functional. The right-sided maze procedure cannot be effectively performed in this setting because landmarks such as the coronary sinus and the tricuspid valve are not exposed. The patch must be removed so that the tricuspid valve annulus and coronary sinus can be properly identified and the cryoablation lesion can be placed. If the valve is not competent it can be isolated again with a Gore-Tex patch anchored to the leaflets of the tricuspid valve near the annulus to prevent unwanted atrioventricular block (Fig. 6B–D).

6.4. Right atrial cannulation in the presence of a right atrial clot

A small percentage of patients who undergo Fontan conversion have right atrial clots. In this setting Fontan conversion is hazardous because of compromised cardiac output during sternotomy, the risk of clot dislodgement with cannulation, and the potential for venous catheter occlusion. To assist in safe atrial cannulation, we have used magnetic resonance imaging, computerized tomography, and epicardial echocardiography to evaluate clot size and location preoperatively. Often times it is preferable to perform aorto-right atrial bypass to establish adequate cardiopulmonary bypass and improve perfusion while the dissection is completed that can lead to bicaval cannulation (Fig. 7A).

6.5. Distended left superior vena cava causing left pulmonary vein stenosis

Two patients had distended left superior vena cava resulting in left pulmonary vein stenosis. In the first patient this was caused by congenital orifice atresia in the right atrium and previous surgical ligation at the innominate vein connection and in the second patient it was caused by coronary sinus occlusion due to lengthy radiofrequency catheter ablation. Both patients had enlarged cardiac chambers caused by blood accumulation without egress anterior to the course of the left pulmonary veins, producing stenosis. During Fontan conversion the coronary sinus was

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Fig. 5. (A) Illustration of a Bjork—Fontan modification showing a right atrial to right ventricular non-valved connection with a prosthetic graft roof. (B) Illustration of completed electrocautery dissection of the entire atrioventricular groove. The amount of atrium that is freed during this maneuver will determine the extent of atrial reduction and the amount of unscarred atrial tissue for the atrial pacemaker leads that will be placed at the end of the Fontan conversion. (C) The completed extracardiac connections are shown. Note the long atrial suture line indicating right atrial wall reduction and closure. A right ventricular patch maintains right ventricular to main pulmonary artery continuity, thus insuring outflow of the thebesian venous flow and avoidance of right ventricular dilatation. (Reprinted with permission. Mavroudis et al. [13].)

Fig. 6. (A) Right atrial view of a patient with pulmonary atresia and intact ventricular septum who had a prior atrio-pulmonary Fontan and developed arrhythmias and suprasystemic right ventricular pressure. Note the cryoablation lines are placed for the modified right-sided maze procedure and the fenestrated tricuspid valve isolation patch. (B) Right atrial view of a patient with double inlet ventricle who had an atrio-pulmonary Fontan with tricuspid valve isolation and atrial septal defect closure. After the removal of the isolation patch the tricuspid valve and coronary sinus are uncovered to facilitate the right-sided maze procedure. (C) Right atrial view showing the results of isolation patch removal and atrial septal defect creation. The cryoablation lesions are shown after proper identification of the tricuspid valve annulus and coronary sinus. The cryoablation lesion from the base of the right atrial appendage to the anterior tricuspid annulus is optional. (D) Right atrial view of Gore-Tex tricuspid valve isolation after cryoablation. The suture line for attachment is placed in the valve tissue near the annulus in order to avoid injury to the atrioventricular node. (Reprinted with permission. Mavroudis et al. [13].)

Fig. 7. (A) Right atrial cannulation in the presence of a right atrial clot. (B) Distended LSVC causing left pulmonary vein stenosis. (C) Coronary sinus was unroofed thus establishing proper coronary sinus drainage. (D) Reconstruction of discontinuous pulmonary arteries using polytetrafluoroethylene graft. (Reprinted with permission. Mavroudis et al. [13].)
unroofed in the first patient and occlusive fibrous tissue/thrombus was resected relieving the pathway obstruction in the other (Fig. 7B and C).

### 6.6. Discontinuous pulmonary arteries

Modification of the extracardiac Fontan may be used to convert patients with discontinuous pulmonary arteries resulting from the combination of a right classic Glenn procedure and a right atrial-to-left pulmonary artery connection. Aortic homografts have a natural favorable curve to accomplish an end-to-end anastomosis between the distal homograft and left pulmonary artery, a side-to-side anastomosis between the homograft and the classic Glenn, and an end-to-end anastomosis between the proximal homograft and the inferior vena cava. Recognition of the possible need for future cardiac transplantation led us to modify our reconstructive procedures to avoid the use of allograft material, which has been shown to increase panel-reactive antibody (PRA) levels and interfere with optimal immunosuppression. We now favor polytetrafluoroethylene grafts for reconstruction of discontinuous pulmonary arteries (Fig. 7D).

### 6.6.1. Patient population

From December 1994 until August 2007, 118 patients (mean age 23.0 ± 8.1 years) have undergone 119 procedures for conversion from an atriopulmonary to a total cavopulmonary artery extracardiac Fontan with concomitant arrhythmia surgery and pacemaker placement at our institution. All patients underwent preoperative cardiac catheterization with hemodynamic assessment and angiography for anatomic definition. Most underwent preoperative intracardiac electrophysiologic mapping; patients with atrial fibrillation and poor vascular access were omitted. In the first nine procedures in our series isthmus cryoablation was used for arrhythmia control. The next 110 consecutive cases had variations of the modified right atrial maze for macro re-entrant atrial tachycardia and the Cox maze III procedure for atrial fibrillation and left macro re-entrant atrial tachycardia.

### 7. Results

There was one (1/118, 0.8%) early and nine (9/118, 7.6%) late deaths. Six patients (6/118, 5.1%) required cardiac transplantation one week and 6, 8, 10, 11, and 33 months after Fontan conversion with arrhythmia surgery. One patient (0.8%) underwent re-exploration for postoperative bleeding, mediastinitis occurred in two patients (2/118, 1.7%), and seven patients (7/118, 5.9%) had acute renal failure. The mean age at the original Fontan operation was 7.0 ± 5.1 years. Sixteen patients had a prior Fontan revision and the mean interval from the original Fontan operation to the total cavopulmonary artery extracardiac Fontan conversion was 14.6 ± 5.3 years. The mean duration of chest tube placement was 9.1 ± 4.9 days. The mean postoperative hospital stay was 13.7 ± 11.9 days. The Kaplan–Meier curve (Fig. 8) shows freedom from death in 118 patients who underwent Fontan conversion with arrhythmia surgery from 1994 to 2007.

### 8. Conclusion

Fontan conversion to total cavopulmonary artery extracardiac Fontan with concomitant arrhythmia surgery and pacemaker placement is excellent therapy for patients whose Fontan repair has failed. Complex reconstructive techniques ensure unobstructed pathways. Evolution of pacemaker technology has provided more options for postoperative electrophysiologic management. Fontan conversion does not preclude cardiac transplantation which will continue to be an important mode of therapy for these patients.

### References


