Rescuing the Late Failing Fontan: Focus on Surgical Treatment of Dysrhythmias

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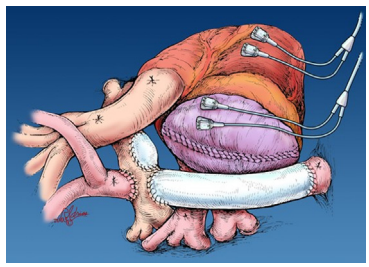
Patients with a failing Fontan frequently have dysrhythmias. This review will focus on the treatment of those dysrhythmias based on the 140 patients who have had a Fontan conversion with arrhythmia surgery at Ann & Robert H. Lurie Children's Hospital of Chicago (Chicago, IL). Important technical points to be emphasized are the following:

- **Atrial fibrillation is extremely common in patients with a failing Fontan and strong consideration should be given to a Cox-maze III during any Fontan conversion**
- **Sinus node dysfunction is common post-conversion and all patients should receive a dual-chamber epicardial pacemaker**
- **Careful preoperative imaging with either computed tomographic or magnetic resonance imaging is needed to predict re-entry problems at the time of repeat sternotomy**
- **If the patient has protein-losing enteropathy, ascites, or a right or indeterminate ventricular morphology, strong consideration should be given to heart transplantation rather than a Fontan conversion and arrhythmia surgery.**

Freedom from cardiac death or transplant for patients undergoing Fontan conversion with arrhythmia surgery is 84% at 10 years. The effects of atrial arrhythmia operations are durable in most patients. There is increasing evidence that earlier referral of patients with an atriopulmonary Fontan for Fontan conversion will improve long-term results. Fontan conversion should be performed at centers with the institutional expertise to care for these complex patients. Note that this is an eroding patient population. The time to act may be now.
Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 20:33-37 **© 2017 Elsevier Inc. All rights reserved.**

Introduction

The Fontan conversion procedure was originally introduced by Dr. Hilel Laks and colleagues.[1] They described conversion of an atriopulmonary to a cavopulmonary anastomosis for the management of late arrhythmias and atrial thrombosis in patients having a failing Fontan. The surgical strategy used was a lateral tunnel-type cavopulmonary Fontan. At Ann & Robert H. Lurie Children's Hospital of Chicago (Chicago, IL) we recommend using an extracardiac Fontan approach with a Gore-tex (W.L. Gore & Assoc., Flagstaff, AZ) graft from the inferior caval vein to the pulmonary artery rather than a lateral tunnel. In addition, patients were evaluated and, when required, had arrhythmia surgery (nearly all patients). Arrhythmia surgery consisted of a right atrial maze procedure for patients who had re-entry atrial tachycardia and a Cox-maze III procedure for patients with atrial fibrillation or left-sided atrial



Completed Fontan conversion. (Reprinted with permission from Backer et al. Cardiol Young 2006;16[suppl 1]: 85-91.)

Central Message

Most Fontan conversion patients will require a Cox-maze III and dual chamber epicardial pacemaker for surgical treatment of their dysrhythmia. Earlier referral will improve long-term results.

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http://dx.doi.org/10.1053/j.pcsu.2016.09.012
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Table 1 Patients undergoing Fontan conversion at Ann & Robert H. Lurie Children's Hospital from 1994 to 2012 (n = 140)

Median age at initial Fontan	5.6 yrs (range, 1.1 – 34.9)
Median age at Fontan conversion	23.2 yrs (range, 2.6 – 47.3)
Interval from initial Fontan to conversion	16.7 yrs (range, 1.2 – 33.6)
Right atrial tachycardia	48 patients
Left atrial tachycardia	21 patients
Atrial fibrillation	67 patients

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re-entry tachycardia. The purpose of this review is to evaluate the outcomes of that treatment strategy and, based on that analysis, make recommendations for individual failing Fontan patients.

Many of the surgical recommendations in this article are based on the recent review published in 2016.[2] That review focused on the first 140 Fontan conversions with arrhythmia surgery performed at our institution. These patients were analyzed for predictors of cardiac death or transplant and the incidence of arrhythmia recurrence. Characteristics of this patient population are shown in Table 1. Many of these patients develop a massively dilated right atrium. An example of magnetic resonance imaging showing the greatly enlarged right atrium is shown in Figure 1.

The detrimental effect of arrhythmias on adult patients who have undergone an atriopulmonary Fontan was reported by Diller and colleagues in 2010.[3] Patients with arrhythmia have a much higher incidence of death or transplant (Figure 2). Patients with an arrhythmia have a six times higher incidence of death or need for heart transplantation with a 3-year mortality of 25% and a 6-year mortality of >40%. We currently believe that all patients status-post atriopulmonary Fontan who have developed atrial arrhythmias should be carefully evaluated and most likely referred for Fontan conversion.

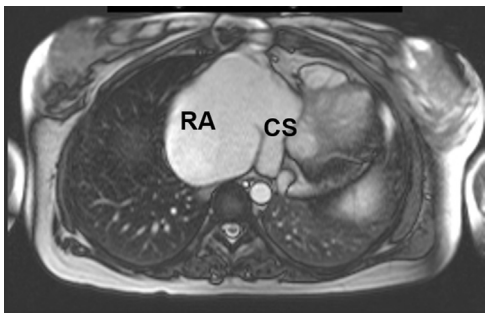

Figure 1 Axial image of magnetic resonance imaging demonstrates severely enlarged right atrium after an atriopulmonary connection. Note the very dilated coronary sinus. CS, coronary sinus; RA, right atrium.

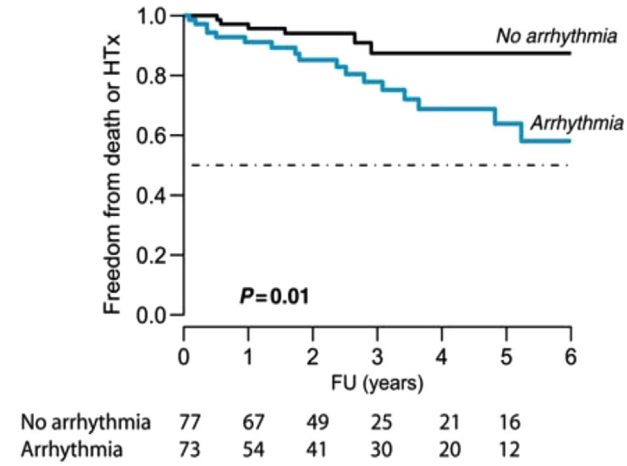

Figure 2 The impact of onset of arrhythmia in adult patients who have undergone an atriopulmonary Fontan. The top line shows freedom from death or transplant for patients without arrhythmia. The lower line shows the freedom from death or transplant in patients with an arrhythmia. (Reprinted from Diller, et al. *Eur Heart J* 2010;31:3073-3083, by permission of Oxford University Press.[3])

Surgical technique

After initiation of cardiopulmonary bypass with cannulation of the high superior caval vein and direct cannulation of the inferior caval vein, cardiopulmonary bypass is initiated (Figure 3). After cross-clamping the aorta and delivering cardioplegia, the following steps are performed:

1) resection of the enlarged right atrium
2) atrial septectomy
3) right atrial maze or biatrial maze with cryoablation (-160°C × 1 minute)

Cross-clamp off

4) extracardiac Gore-tex graft (inferior caval vein to pulmonary artery)
5) bidirectional superior cavopulmonary anastomosis
6) placement of an epicardial dual-chamber pacemaker with bipolar steroid eluting leads.

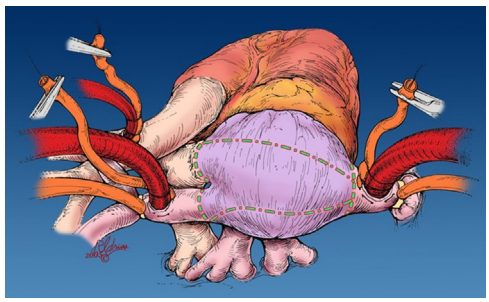

Figure 3 This illustration shows the cannulation strategy for a failing Fontan with an atriopulmonary connection. The high superior caval vein and inferior caval vein have been cannulated directly. The dotted lines indicate the portion of right atrium that will be resected, and where the inferior caval vein and superior caval vein are transected. (Reprinted from Mavroudis et al. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu* 1999;2:143-56, with permission from Elsevier.)

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