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Arrhythmia Management in the Fontan Patient

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Abstract With longer duration of follow-up, as many as 50% of Fontan patients will develop atrial tachycardia, usually in association with significant hemodynamic abnormalities. Arrhythmia management in the Fontan patient is reviewed. The incidence and type of arrhythmia occurrence are examined, including macro-reentrant rhythm which involves the right atrium, reentrant rhythm localized to the pulmonary venous atrium (seen in patients with lateral tunnel procedures), and atrial fibrillation. Risk factors for development of these arrhythmias are considered, and short- and long-term therapeutic options for medical and surgical treatment are discussed. Surgical results are presented for 117 patients undergoing Fontan conversion and arrhythmia surgery (isthmus ablation (9), modified right atrial maze (38) or Cox-maze III (70)). Operative mortality is low (1/117, 0.8%). Seven late deaths occurred, and include two patients who died shortly following cardiac transplantation (2/6, 33%) after Fontan conversion and arrhythmia surgery. Overall arrhythmia recurrence is 12.8% during a mean follow-up of 56 months. Fontan conversion with arrhythmia surgery can be performed with low operative mortality, low risk of recurrent tachycardia, and marked improvement in functional status in most patients. Because the development of tachycardia is usually an electromechanical problem, attention to only the arrhythmia with medications or ablation may allow progression of hemodynamic abnormalities to either a life-threatening outcome or a point at which transplantation is the only potential option. Because cardiac transplantation in Fontan patients is associated with high early mortality, earlier consideration for surgical intervention is warranted.

Keywords Atrial fibrillation · Atrial reentry tachycardia · Fontan procedure · Fontan revision

The incidence of atrial arrhythmias following Fontan-type surgery increases steadily with the postoperative interval, with at least 50% of patients experiencing problematic atrial tachycardia by 20 years of follow-up [22, 47, 62]. Although the arrhythmias are generally believed to be a consequence of surgical interventions, electron microscopy has demonstrated that the atria of patients with tricuspid atresia show a distinctly abnormal atrial fiber array compared with normal hearts [50]; this unusual fiber orientation may predispose the atria to the slowing of conduction necessary for reentrant rhythms. In addition, in natural history studies of unoperated adults with tricuspid atresia, at least 40% of patients experienced tachycardia by the fourth decade [58, 59]. In postoperative Fontan patients, supraventricular tachycardia is usually a macro-reentrant rhythm involving the right atrium, although in lateral tunnel-type repairs the reentrant rhythm may be localized to the pulmonary venous atrium [16]. Slowed conduction with reentry is facilitated by anatomic barriers, such as the orifices of the inferior and superior vena cavae, the os of the coronary sinus, and the atrial septal defect, further compounded by extensive atrial suture lines of either the atriopulmonary anastomosis or the lateral tunnel repair. Residual hemodynamic abnormalities (or simply the dissipation of energy forces within the atria) result in marked

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distention with fibrosis, which is complicated by sinus node dysfunction; an irregular atrial rhythm serves as a trigger for the onset of tachycardia.

Risk Factors

Surgical variables identified as risk factors for the development of tachycardia include an older age at the time of initial repair, early postoperative arrhythmias, sinus node dysfunction, and double-inlet left ventricle [10, 20, 22, 60]. Modifications of the atriopulmonary anastomosis to the lateral tunnel repair were developed in an effort to improve hemodynamics and limit late arrhythmias. Although initial reports suggested a decreased incidence of arrhythmias following the lateral tunnel technique [6, 10, 22, 46], this difference has become less significant with longer follow-up [19]. Limitation of atrial suture lines by performance of the extracardiac total cavopulmonary anastomosis has a theoretical advantage in limiting the development of tachycardia, which is supported by the early follow-up report of Amodeo et al. [3] showing a 5-year arrhythmia-free rate of 92%.

Significant hemodynamic abnormalities are present in many Fontan patients with recurrent atrial tachycardia. Marked atrial dilatation with stasis of atrial flow and limited forward output predisposes patients to thrombus development and the risk of stroke or pulmonary embolus. The dilated right atrium may impinge on pulmonary venous return and distort ventricular anatomy sufficiently to depress systolic function. Anti-arrhythmic medications frequently exacerbate bradycardia and further depress ventricular function. Thus, it is essential to view the presence of atrial tachycardia in most cases as an electromechanical problem, which is not adequately addressed by either arrhythmia therapy or Fontan revision in isolation. Additionally, ventricular function may be further depressed by the development of chronic atrial tachycardia or atrial fibrillation, and repeated attempts at catheter ablation may allow a window of opportunity to pass for surgical intervention with low risk.

Medical Treatment of Atrial Tachycardia

Early Postoperative Tachycardia

A small subset of patients develops atrial tachycardia during the acute postoperative period following Fontan completion. At that time, it is important to determine whether tachycardia is a consequence of inotropic medications used to maintain cardiac output or related to either significant residual anatomic problems or marked sinus node dysfunction. Persistent atrial arrhythmias in the absence of inotropic medications are usually related to structural hemodynamic sequelae; attention to these abnormalities by early diagnosis and possible emergent reoperation is of critical importance and should not be delayed. Acute management after structural hemodynamic residua have been excluded includes removal of inotropic agents and providing a regular atrial rhythm with pacing, as necessary. Intravenous amiodarone is highly effective for rhythm control in this setting; diltiazem may be used for acute rate control as needed.

Late Postoperative Tachycardia

For patients developing late postoperative arrhythmias, the mean time to presentation is usually 6–11 years post-repair [16, 52]. Following the initial presentation with tachycardia, many patients will experience isolated episodes of tachycardia during the next few years [16, 36]. However, as time progresses, tachycardia recurrences become more frequent and prolonged, with the potential development of atrial fibrillation after several years [32].

Recognition of the acute onset of tachycardia is confounded by the usual presence of 2:1 atrioventricular conduction resulting in ventricular rates of 110–130 beats per minute (bpm); such patients may present with vague symptoms of unexplained fatigue or respiratory symptoms. In contrast, patients with rapid 1:1 atrioventricular conduction of tachycardia frequently experience syncope or rarely experience cardiac arrest. The vagal response during syncope may terminate tachycardia, creating uncertainty as to the cause of symptoms. In either setting, patients with single ventricle physiology do not tolerate persistent tachycardia and may develop profound congestive failure within 12–36 hours of the onset of persistent heart rates greater than 100 bpm. In addition to depressed ventricular systolic function and atrioventricular valve regurgitation, tachycardia increases the risk of developing atrial thrombi resulting in pulmonary or cerebral emboli. Therefore, the acute termination of tachycardia within 24 hours of presentation is of high importance; delaying termination of tachycardia for 4–6 weeks of anticoagulation, as is recommended for tachycardia of uncertain but prolonged duration in adult patients with two ventricles, may have life-threatening consequences. In the setting of prolonged tachycardia, echocardiograms (usually transesophageal) are performed to determine the presence of atrial thrombi and establish the risk of acute emboli with cardioversion. Although this is the most sensitive technique for detecting atrial thrombi, our experience with transesophageal echocardiograms immediately preceding Fontan conversion suggests that this technique is neither sensitive nor specific

for the detection of thrombi in the dilated right atria [31]. The risk of cardioversion with embolic phenomena must be weighed against the risk of delaying cardioversion and developing profound ventricular dysfunction or sudden cardiac arrest.

Acute Therapy

Therapeutic choices for acute cardioversion include pharmacologic agents, transesophageal or intra-atrial pacing, and synchronized direct-current cardioversion. Data regarding the efficacy of acute pharmacologic cardioversion are not available, and there are only anecdotal reports of efficacy [22]. In the hemodynamically stable patient, a trial of small doses of intravenous calcium channel blocking medication, such as diltiazem, may be effective in terminating tachycardia or at least slowing the ventricular response. Ibutilide, a class III anti-arrhythmic medication, may be acutely successful but generally is avoided in patients with electrolyte abnormalities or those receiving other medications such as sotalol or amiodarone, which may prolong the QT interval. Transesophageal pacing conversion is frequently effective, particularly in younger patients or patients with lateral tunnel-type repairs [8]. Placement of cardioversion pads in the anterior–posterior configuration is useful to facilitate cardioversion of the massively dilated atria, and energy doses of 1 or 2 J/kg, or 200 J in adult patients, are used.

Chronic Therapy

The choice of anti-arrhythmic therapy is determined by the severity of symptoms associated with tachycardia and also the reliability of detection of tachycardia and delivery of prompt medical care. Patients presenting with symptoms of palpitations and seeking care at the earliest onset of infrequent episodes of hemodynamically stable tachycardia may be treated with periodic cardioversion, without chronic anti-arrhythmic drugs beyond beta-blockers or digoxin. Once tachycardia becomes recurrent, medications such as procainamide, sotalol, and propafenone are initially effective, but with time tachycardia usually becomes refractory to medications. In our experience, a combination of sotalol with low-dose beta blockade has been the most effective chronic pharmacologic treatment. Due to the high incidence of pulmonary and thyroid side effects with chronic use, amiodarone is not recommended for Fontan patients other than as a bridge to more definitive therapy. Based on the risk of thrombosis, patients with atrial tachycardia should receive chronic anticoagulation therapy with warfarin.

Hemodynamic Assessment

Following the initial presentation with tachycardia, allowing for a period of recovery of ventricular function, a careful assessment is performed to detect hemodynamic abnormalities. Patients presenting with severe symptoms, such as congestive failure, syncope, or cardiac arrest, undergo invasive cardiac catheterization with electrophysiology testing and progress to more definitive therapy. Noninvasive evaluation including chest radiograph, echocardiogram, and exercise testing is performed for patients with milder symptoms and physical examinations consistent with acceptable hemodynamics. The presence of cardiomegaly, decreasing exercise tolerance, peripheral edema or lower leg discoloration, ascites, protein-losing enteropathy, or progressive cyanosis with resting saturations less than 92% should prompt aggressive evaluation. Patients with Fontan physiology typically do not complain of exercise intolerance until advanced stages of failure; therefore, exercise testing provides objective documentation of declining maximal oxygen consumption, desaturation, or arrhythmia occurrence with exertion [49].

The transthoracic echocardiogram frequently underestimates the degree of atrial dilatation or areas of obstruction to atrial flow, which are more accurately assessed by cardiac catheterization with angiography. Cardiologists may be misled by assessing hemodynamic status on the basis of right atrial pressure measurement alone; an otherwise acceptable right atrial pressure of 15 or 16 mmHg may be associated with angiographic evidence of marked atrial dilatation with poor forward flow from the atria to the pulmonary arteries. In this setting of passive filling of the pulmonary circulation, a right atrial-to-pulmonary artery gradient as low as 2 mmHg may represent significant obstruction. Full hemodynamic assessment in addition to pressure measurements includes angiographic estimation of right atrial emptying; right-to-left atrial shunting; kinking and distortion of the pulmonary arteries, particularly of the right pulmonary artery; and the degree of pulmonary arteriovenous malformations. Obstruction of pulmonary venous return is assessed by both angiography and measurement of the pressure difference between bilateral wedge pressures and ventricular end diastolic pressure. Ventricular angiograms should assess atrioventricular valve regurgitation and provide a qualitative assessment of contractility, in addition to catheterization for measurement of end diastolic pressure and cardiac index.

Catheter Ablation

Catheter ablation is recommended in patients without significant hemodynamic abnormalities or in patients

Table 1 Ablation results in Fontan patients

Reference	No. of patients	Acute success (%)	Mean follow-up (months)	Recurrence (%)
Triedman et al. [54]	6	33	—	50?
Kalman et al. [27]	4	50	17	50
Chinitz et al. [12]	3	33	12	100
Lesh et al. [35]	3	33	—	—
Betts et al. [9]	5	60	6	66
Chan et al. [11]	1	100	—	100
Collins et al. [13]	43	72	—	32
Nakagawa et al. [44]	6	100	10	33
Triedman et al. [53]	63	43	25	52
Kannankeril et al. [28]	15	—	38	53
Weipert et al. [62]	30	83	18	76[a]

[a] Arrhythmia recurrence not specifically stated; 76% receiving anti-arrhythmic medications at follow-up

considered very high-risk surgical candidates due to ventricular dysfunction or multiorgan disease. In our series of Fontan patients undergoing electrophysiologic mapping of right atrial macro-reentry, three primary exit sites of atrial tachycardia circuits identified are the low lateral right atrial wall, the perimeter of the atrial septal defect patch, and the inferomedial right atrial isthmus [16]; others have reported similar findings [4, 18, 63]. The atriopulmonary anastomosis is a less common site of exit. In patients with a lateral tunnel-type Fontan repair, the tachycardia circuit may be partitioned to the pulmonary venous atrium; these patients often have negative P waves in lead I [16]. Infrequently, focal atrial tachycardia may be present.

Acute success rates for ablation of atrial tachycardia in Fontan patients range from 33% to 100% [9, 11–13, 25, 27, 35, 44, 53, 63]; however, recurrence of tachycardia during short-term follow-up is reported as 33%–100% (Table 1). Acute and long-term results of catheter ablation in Fontan patients are significantly worse than results achieved in most other forms of congenital heart disease. Improvement in acute success rates is expected with the use of three-dimensional mapping and larger tipped, irrigated catheters with more powerful energy delivery. The technique of scar mapping of the right atrium with ablation lines delivered to eliminate channels between scars has provided promising results [44]. Limitations to the catheter technique specific to Fontan patients include the multiplicity of tachycardia circuits, restricted catheter access, distorted anatomy, hemodynamic instability, and the inability to deliver lesions of sufficient depth to achieve transmural penetrance. In our experience with more than 100 patients undergoing repeat surgery, right atrial wall thicknesses as measured in the operating room average 12 mm, extending to 20 mm,

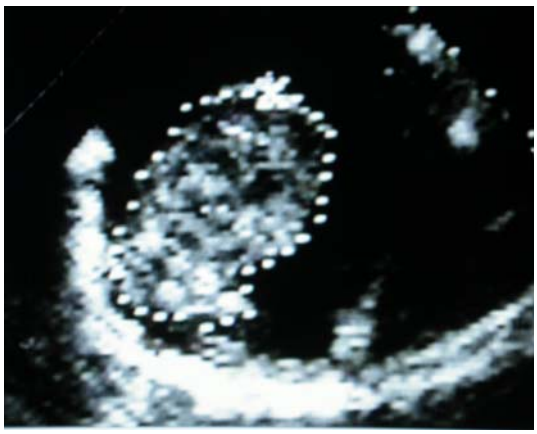

Fig. 1 Image of thrombus obtained in an adolescent months after a partially effective ablation procedure

indicating the difficulty of completion of transmural lines of block with lesion depths of 3–8 mm. Thrombogenicity of extensive ablation lesions in the stagnant right atrium is of concern, as evidenced by the image of thrombus obtained in an adolescent months after a partially effective ablation procedure (Fig. 1). In addition, focal wall thinning due to extensive ablation may produce an additional area of slowed conduction, promoting tachycardia. The most significant concern regarding ablation in Fontan patients is the possibility of overlooking significant hemodynamic problems and allowing ventricular dysfunction, atrial thrombosis, or protein-losing enteropathy to develop and/or progress to either a life-threatening condition or beyond the possibility of surgical intervention.

Surgical Treatment of Arrhythmias

Fontan Revision

Because the majority of Fontan patients with arrhythmias have significant associated structural/hemodynamic problems, a number of centers have performed Fontan revisions without arrhythmia surgery, resulting in improved hemodynamic outcomes [7, 29, 34, 41, 48, 51, 55, 57]. In a meta-analysis of 77 reported cases of Fontan revisions (Table 2), tachycardia recurred in 76% of patients during short-term follow-up. The inability to eliminate tachycardia by only improving hemodynamics led our group to attempt to incorporate ablation techniques into the operative revision of the Fontan circulation. Initially, we performed limited ablation of the inferomedial right atrial isthmus, with implantation of an atrial anti-tachycardia pacemaker [17]. Due to recurrent tachycardia in several of these patients, a modified right atrial maze procedure was developed to eliminate all right atrial macro-reentrant circuits,

Table 2 Results of Fontan revision without arrhythmia surgery

Reference	No. of patients	Mortality (%)	Transplant (%)	Arrhythmia recurrence (%)
Balaji et al. [6]	3	33	—	0
Kao et al. [29]	3	0	—	66
Vitullo et al. [57]	9	11	—	100
McElhinney et al. [41]	7	14	14	100
Kreutzer et al. [34]	8	12.5	12.5	66
Scholl et al. [51]	12	8.3	—	100
van Son et al. [55]	18	11	11	69
Petko et al. [48]	13[a]	7.7	—	—
Sheikh et al. [52]	4	0	0	75
Total	77	9	6	76

[a] Five of 13 patients underwent arrhythmia surgery; arrhythmia recurrence not stated

essentially eliminating tachycardia recurrence during medium-term follow-up [16]. The surgical technique of the Fontan conversion to a total cavopulmonary extracardiac connection has been extensively reported by our group [15–17, 37–40].

Patients considered for arrhythmia surgery undergo preoperative electrophysiology studies with mapping of atrial macro-reentrant circuits; epicardial mapping of the right atrium is then repeated in the operating room in an attempt to correlate the findings of the endocardial studies with observed anatomic findings. Careful review of the tachycardia history and tracings is performed because the presence of clinical atrial fibrillation indicates that more extensive arrhythmia surgery in the left atrium will be necessary. Details of the original surgical Fontan procedure are reviewed because lateral tunnel-type repairs, incisions in the roof of the left atrium, or deviation of the atrial septal patch to the left may partition the anatomic right atrium, and the reentrant circuit, to the pulmonary venous chamber. In addition, focal atrial tachycardia is not addressed by the right atrial maze and requires direct elimination of the tachycardia focus.

Right Atrial Reentry Tachycardia

The modified right atrial maze surgery involves an incision from the superior vena cava to the inferior vena cava, with resection of a large portion of the anterior right atrial wall and the right atrial appendage. Linear cryoablation lesions are delivered as follows: (1) from the os of the coronary sinus to the transected inferior vena cava; (2) from a right-sided atrioventricular valve orifice, when present, to the transected inferior vena cava; (3) from the posterior rim of the atrial defect to the lateral rim of the resected atrial wall; (4) from the base of the resected right atrial appendage to the superior rim of the atrial defect; and, more recently, (5) an additional lesion delivered from the inferior rim of the atrial defect to the posterior border of the coronary sinus. Cryoablation lesions were originally delivered using a circular probe (Frigitronics) at –60°C for 90 seconds each; currently, a malleable 12-Fr linear probe is used to deliver temperatures of –155°C for 60 seconds (CryoCath Technologies, Montreal, Quebec). The total time necessary to complete the ablation lesions is 5 or 6 minutes. Care is taken to avoid the conducting system near the atrioventricular node. Based on the findings of the electrophysiology testing and the operative history, additional lesions may be necessary in the partitioned left atrium. Patients with atrial reentry tachycardia undergo postoperative atrial pacing studies after cessation of inotropic support to assess the efficacy of surgery and to program atrial anti-tachycardia pacemakers, if necessary. Patients subsequently receive beta-blocker medications for a period of 3 months postoperatively.

Atrial Fibrillation

The presence of atrial fibrillation mandates performance of the Cox maze III procedure in the left atrium; right atrial surgery decreases but does not eliminate atrial fibrillation [5, 23, 24]. In addition to the modified right atrial maze procedure reported previously, a left atrial maze procedure is performed as described by Cox et al. [14]. Lesions were initially delivered by incision, but the use of linear cryoablation lesions with a malleable probe has significantly decreased the operative cross-clamp time.

The pulmonary veins are isolated with an encircling lesion. The left atrial appendage is excised, with a cryoablation lesion connecting the base of the resected appendage to the encircling pulmonary vein lesion. A cryoablation lesion is delivered from the inferior pulmonary veins to the mitral valve annulus; a corresponding lesion is placed epicardially on the coronary sinus. Performance of the left atrial maze requires an additional 30–45 minutes using the cryoablation technique and more than 60 minutes using incisions, with attendant increased risk of

renal failure and myocardial dysfunction. In patients with multiple prior surgeries and markedly distorted anatomy, adequate exposure of the left atrial appendage is difficult and may not be possible. In these circumstances, a circular cryoablation lesion can be placed at the base of the left atrial appendage within the atrium to isolate the left atrial appendage without resection. Patients with atrial fibrillation receive intravenous amiodarone postoperatively, followed by oral amiodarone for a period of 3 months postoperatively; atrial pacing studies are not performed prior to hospital discharge.

Pacemaker Implantation

Of our initial 13 patients, atrial anti-tachycardia pacemakers were implanted in 12 (Intertach II, Intermedics). Because this pacemaker became unavailable, and due to the decreased incidence of tachycardia with the more extensive right atrial maze and the need for rate responsiveness, atrial rate-responsive pacemakers (Medtronic Kappa SR, Medtronic, Inc., Minneapolis, MN) were implanted in the subsequent 31 patients. More recently, dual-chamber anti-tachycardia pacemakers (Medtronic Gem III AT) have been implanted due to the potential need for ventricular pacing during late follow-up and to avoid the risk of reoperation; most of these pacemakers are programmed for atrial pacing only. Atrial pacing rates are programmed to consistently maintain a regular atrial rhythm faster than the intrinsic rhythm, usually 70–80 beats per minute during late follow-up. One patient received an epicardial defibrillator in addition to multisite ventricular pacing; this patient had a prior cardiac arrest secondary to ventricular tachycardia, and ventricular dysfunction associated with left bundle branch block.

Surgical Results

Since 1994, 117 patients have undergone arrhythmia surgery with Fontan conversion at Children's Memorial Hospital; the surgical outcome is summarized in Table 3. There was one early death (0.8%), occurring in an adolescent male with heterotaxy syndrome and marked ventricular diastolic dysfunction. Late mortality has been 5.9% due to intractable heart failure (1), coronary artery disease (1), discontinuation of renal dialysis (1), injuries from a motor vehicle collision (1), and suddenly after sedation administration (1). Two additional patients died following cardiac transplantation 7 and 10 months after Fontan conversion.

Overall arrhythmia recurrence is 12.8% during a mean follow-up of 56 months. In patients with atrial reentry tachycardia, we initially performed isthmus ablation on 9 patients. Since initiation of the modified right atrial maze procedure, 3/34 patients (8.8%) have had atrial tachycardia recurrence. These surgical arrhythmia results have been duplicated and reported by other centers with similar outcomes; variations in arrhythmia recurrence may be due to the absence of preoperative characterization of the arrhythmia circuits, variable lesions, and the use of radiofrequency versus cryoablation [1, 33, 52, 56, 61]. In the group of patients with atrial fibrillation who underwent the Cox maze III procedure, no patient has had late recurrence of atrial fibrillation. However, the late development of isolated episodes of atrial reentry tachycardia has occurred in 9/70 patients (12.8%) at a mean postoperative period of 36 months; recurrences are usually controlled with beta-blocking medications alone. The occurrence of late atrial reentry tachycardia following catheter ablation procedures for atrial fibrillation is reported to be 11%–43% [2, 25, 28]. The possibility of atrial conduction superiorly near Bachmann's bundle or via atrial fibers in the coronary sinus is speculated.

Functional classification improved dramatically in most patients, as demonstrated by exercise testing [30]. However, a small number of patients experienced progressive decline in ventricular dysfunction, usually associated with significant atrioventricular valve regurgitation. Six patients have undergone cardiac transplantation at a mean postoperative interval of 12 months; one patient died awaiting transplantation, as noted previously. Overall, the incidence of late transplantation (5.1%) is similar to that reported in other series for Fontan patients [55]; in certain patients, Fontan conversion may substantially delay the timing of transplantation.

Cardiac Transplantation

Of note, early mortality following transplantation for Fontan patients is higher than for other forms of congenital heart disease and is reported to be 33%–48% [21, 26, 42, 43]. In our center, 12 Fontan patients were referred initially for consideration for cardiac transplantation: 7 underwent Fontan conversion for potentially correctable hemodynamic abnormalities, and 5 were listed for transplantation. Of the 7 undergoing surgery, all survived, and 5 are alive and considered to be in New York Heart Association class I or II; 2 patients went on to cardiac transplantation 23 months postoperatively. Patients with systemic right ventricles, heterotaxy syndrome, severe atrioventricular valve regurgitation, and protein-losing enteropathy appear to be at the highest risk for poor outcome. Of the 5 patients listed for transplantation, 2 patients died waiting, and 3 patients underwent transplantation, 2 of whom died of rejection and

Table 3 Results of Fontan conversion with arrhythmia surgery at Children's Memorial Hospital, 1994–2007

	RA surgery	Cox maze III	Total
No. of patients	47	70	117
Age at initial Fontan, years (median)	6.12 ± 3.72 (5.7)	7.62 ± 5.85 (5.75)	7.02 ± 5.13 (5.7)
Age at Fontan conversion, years (median)	18.14 ± 6.40 (17.4)	25.89 ± 7.40 (24.4)	22.78 ± 7.96 (21.6)
Mean post-Fontan interval, years (median)	11.24 ± 4.29 (10.6)	16.70 ± 4.68 (16.7)	14.51 ± 5.25 (14.7)
New York Heart Association			
Class II	13	22	35
Class III	26	36	62
Class IV	8	12	20
Mean age at onset SVT, years (median)	11.81 ± 6.19 (11.0)	17.08 ± 7.99 (15.8)	15.11 ± 7.77 (14.0)
Mean No. of anti-arrhythmic medications (median)	2.6 ± 1.7 (2)	3.1 ± 1.7 (3)	2.9 ± 1.7 (3)
Hemodynamic abnormalities			
Massive RA	18	45	63
End-diastolic pressure > 12 mmHg	6	4	10
RA–PA gradient > 2 mmHg	8	5	13
Pulmonary vein obstruction	5	10	15
Right to left shunting	10	5	15
Mean cardiac index (L/min/m^2)	2.58 ± 0.71	2.66 ± 0.85	2.62 ± 0.78
Mortality			
Early	0	1	1
Late	2[a]	5[b]	7
Duration of follow-up, months (median)	87 ± 39 (85)	36 ± 30 (24)	56 ± 42 (50)
Recurrent SVT	6	9[c]	15
Heart transplant/listed for transplant	3/1	3/1[d]	6/2

PA, pulmonary artery; RA, right atrium; SVT, supraventricular tachycardia

[a] Automobile accident in one; post-sedation administration in one patient

[b] Two deaths after cardiac transplantation subsequent to Fontan conversion

[c] No recurrence of atrial fibrillation

[d] One Cox maze III patient awaiting cardiac transplantation died 7 months postoperatively of multiorgan failure

1 is alive and well. These results indicate that earlier intervention to correct residual hemodynamic and arrhythmia problems is an important adjunct to improve overall survival.

Prophylactic Surgical Techniques

Prior to the performance of the initial Fontan repair, atrial tachycardia may be induced in as many as 20% of patients [45]. As described previously, avoidance of intra-atrial suture lines by performance of the extracardiac total cavopulmonary connection may decrease the incidence of arrhythmias during medium-term follow-up. Based on studies in animal models showing the importance of atrial reentry between the inferior atriotomy and the tricuspid valve annulus, centers performing the lateral tunnel-type repair have delivered prophylactic incisions from the atriotomy across the anterior atrial wall to the right-sided valve annulus to decrease arrhythmia development [45]. However, in postoperative patients, multiple centers have documented similar areas of tachycardia reentry: the isthmus between the inferior vena cava and the coronary sinus, the low lateral right atrial wall, and the superior rim of the atrial defect, sometimes extending to the superior vena cava [44]. Lesions in the inferior isthmus may be effective for at least one of these circuits. The loss of a regular sinus rhythm was shown by Fishberger et al. [20] to be the most important predictor of the subsequent development of atrial tachycardia; placement of an atrial pacemaker at the initial Fontan repair may thus delay the development of tachycardia.

Conclusions

With longer durations of follow-up, as many as 50% of Fontan patients will develop atrial tachycardia, usually in association with significant hemodynamic abnormalities. Catheter ablation in Fontan patients has a high risk of

recurrent tachycardia during short-term follow-up and allows underlying hemodynamic problems to progress. Fontan revision without arrhythmia surgery results in improved hemodynamic status but recurrent tachycardia. When tachycardia recurrence becomes frequent or is associated with significant symptoms and/or when significant hemodynamic problems exist, Fontan conversion with arrhythmia surgery can be performed with low operative mortality, low risk of recurrent tachycardia, and marked improvement in functional status in most patients. Because the development of tachycardia is usually an electromechanical problem, attention to only the arrhythmia with medications or ablation may allow progression of hemodynamic abnormalities to either a life-threatening outcome or a point at which transplantation is the only potential option. Because cardiac transplantation in Fontan patients is associated with high early mortality, earlier consideration for surgical intervention is warranted.

By limiting atrial incisions and suture burden, the extracardiac total cavopulmonary connection may decrease the incidence of late atrial arrhythmias. In addition, "prophylactic arrhythmia surgery" should be considered in patients undergoing initial lateral tunnel-type Fontan repairs or Fontan conversion for structural/hemodynamic problems without problematic arrhythmias, based on the likelihood of developing tachycardia over time and the lack of future transvenous access to the right atrium. Performance of the modified right atrial maze lesions requires less than 6 minutes and does not increase the morbidity of the surgical procedure. Finally, we speculate that earlier atrial pacing with lead implantation performed at the time of the initial Fontan procedure may delay the appearance of atrial tachycardia by avoiding irregular and slow atrial rhythms.

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