Long-Term Outcome of Ablative Therapy of Postoperative Supraventricular Tachycardias in Patients With Univentricular Heart
A European Multicenter Study

Natasja M.S. de Groot, MD, PhD; Peter Lukac, MD, PhD; Nico A. Blom, MD, PhD; Jan Peter van Kuijk, MD; Anders K. Pedersen, MD, PhD, DMSci; Peter S. Hansen, MD, PhD, DMSci; Etienne Delacretaz, MD; Martin J. Schalij, MD, PhD

Background—Catheter ablation has evolved as a possible curative treatment modality for supraventricular tachycardias (SVT) in patients with univentricular heart. However, the long-term outcome of ablation procedures is unknown. We evaluated the procedural and long-term outcome of ablative therapy of late postoperative SVT in patients with univentricular heart.

Methods and Results—Patients with univentricular heart (n=19, 11 male; age, 29±9 years) referred for ablation of SVT were studied. Ablation was guided by 3D electroanatomic mapping in all but 2 procedures. A total of 41 SVT were diagnosed as intra-atrial reentrant tachycardia (n=30; cycle length, 310±68 ms), typical atrial flutter (n=4; cycle length, 288±42 ms), focal atrial tachycardia (n=6; cycle length, 400±60 ms), and atrial fibrillation (n=1). Ablation was successful in 73% of intra-atrial reentrant tachycardia, 75% of atrial flutter, and all focal atrial tachycardia and focal atrial fibrillation. During the follow-up period of 53±34 months, 2 patients were lost to follow-up, 3 died of heart failure, 2 underwent heart transplantation, and 1 underwent conduit replacement. Of the remaining group, 8 had sinus rhythm and 3 had SVT.

Conclusions—Focal and reentrant mechanisms underlie postoperative SVT in patients with univentricular heart. Successive SVT developing over time may be caused by different mechanisms. Ablative therapy is potentially curative, with a procedural success rate of 78%. In patients who had multiple ablation procedures, the SVT originated from different atrial sites, suggesting that these new SVT were caused by progressive atrial disease. Despite recurrent SVT, sinus rhythm at the end of the follow-up period was achieved in 72%. (Circ Arrhythmia Electrophysiol. 2009;2:242-248.)

Key Words: heart defects ■ congenital ■ tachyarrhythmias ■ mapping ■ ablation

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urvival of patients with univentricular heart (UVH) has improved significantly since the introduction of the Fontan operation.1 As time after cardiac surgery passes, there is an increase in arrhythmia propensity.2 The incidence of supraventricular tachycardias (SVT) in patients with Fontan is high; for example, an incidence of 21% has been reported during a follow-up period of 15 years.3

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Previous studies have demonstrated that development of these late postoperative SVT are related to hemodynamic changes, electrophysiologic alterations, and the surgical techniques applied.1,2,4 SVT in these patients may result in hemodynamic deterioration, increased risk of thromboembolism, and cardiac death.5-7

Management of postoperative SVT in patients with UVH by antiarrhythmic drug therapy or antitachycardia pacing is often not effective and/or hampered by serious side effects of antiarrhythmic drugs.8 In recent years, catheter ablation has proven to be a successful treatment modality in eliminating SVT.9,10 Because of the distortion and dilatation of the atria in patients with UVH, mapping and subsequent ablation of SVT can be difficult. The use of an advanced mapping technique may therefore improve the outcome of ablative therapy.10-13 However, SVT can reappear after successful ablative therapy because of progressive atrial dilatation and fibrosis.

In this multicenter study, we retrospectively evaluated the mechanisms of SVT, the procedural outcome of ablative therapy of late postoperative SVT in patients with UVH, and
studied arrhythmia recurrences after ablative therapy during long-term follow-up.

Methods

Study Population
The study population consisted of 19 patients with UVH and postoperative SVT who were referred for catheter ablation. Patients were treated in 3 different European centers experienced in ablative therapy of SVT in patients with congenital heart defects.

Data regarding complexity of congenital defects, surgical history, and cardiac rhythm during long-term follow-up were obtained from hospital records.

Mapping Procedure
If SVT was not present at the onset of the procedure, it was induced using programmed electrical stimulation. Mapping was performed with a 3D electro-anatomic mapping system (CARTO, Biosense-Webster, Diamond Bar, Calif). A detailed description of the underlying technology of electro-anatomic mapping has been given previously. Only 2 ablation procedures were performed using fluoroscopy only.

Classification of Supraventricular Tachycardias
Based on activation maps, 3 different types of SVT were distinguished: (1) typical atrial flutter (AFL), a single (counter)-clockwise, cavo-tricuspid isthmus dependent macro-reentrant circuit; (2) intra-atrial reentrant tachycardia (IART), a macro-reentrant tachycardia involving scar tissue, suture lines, or prosthetic materials; and (3) focal atrial tachycardia (FAT), electrical activation originating from a small, circumscribed region with spread of activation away in all directions from the site. In addition, atrial fibrillation (AF) was diagnosed according to usual electrophysiological characteristics.

Ablation Procedure
Ablative therapy was applied after mapping. At each site, radiofrequency current was delivered for 60 seconds. In the case of noncooled ablation, tip temperature was set at 70°C and the maximum output at 50 W. During ablation using an irrigated-tip catheter (31%), temperature was limited to 50°C and power to 45 W, with saline flow of 20 mL/h. Each lesion was tagged on the electroanatomic map. Success was defined as (1) AFL, establishment of a line of conduction block over the cavo-tricuspid isthmus, and (2) IART/FAT, termination during ablation and noninducibility.

Long-Term Outcome
After the ablation procedure, patients were seen every 3 to 6 months at the outpatient clinic for clinical follow-up.

Statistical Analysis
All data were summarized as mean±SD or median (range) as appropriate. Statistical significance was defined as P<0.05.

The authors had full access to and take full responsibility for the integrity of the data. All authors have read and agree of the manuscript as written.

Results

Patient Characteristics
Characteristics of the 19 patients (11 male; age, 29±9 years) are given in Tables 1 and 2.

Underlying anatomic diagnoses were tricuspid atresia (n=13), mitral atresia and double-outlet right ventricle (n=3), and double-inlet left ventricle (n=3). Definitive palliative surgical procedures included the construction of a Blalock shunt (n=3), an atroipulmonary Fontan circulation (n=11), or a total cavopulmonary connection with an intra-atrial tunnel (n=5).

| Table 1. Anatomic Diagnoses and Surgical Status of the Study Population |
| --- | --- | --- |
| Patient No. | Sex | Diagnosis | Surgical Status |
| 1 | M | TA | APF |
| 2 | M | TA | TCPC |
| 3 | M | TA | TCPC |
| 4 | M | TA | TCPC |
| 5 | F | DILV | APF |
| 6 | F | TA | APF |
| 7 | F | TA+PS | APF |
| 8 | F | TA+TGA+PS | Blalock shunt |
| 9 | M | MA+DORV+TGA+PS | TCPC |
| 10 | M | TA+TGA+PS | APF |
| 11 | M | TA+TGA+PS | APF |
| 12 | F | TA | APF |
| 13 | M | MA+DORV | TCPC |
| 14 | F | TA+ASD | APF |
| 15 | M | DILV | Blalock shunt |
| 16 | F | TA | Blalock shunt |
| 17 | M | TA+TGA+PS | APF |
| 18 | M | DILV | APF |
| 19 | F | MA+DORV | APF |

TA indicates tricuspid atresia; MA, mitral atresia; TGA, transposition great arteries; PS, pulmonary stenosis; DORV, double-outlet right ventricle; DILV, double-inlet left ventricle; APF, atrio pulmonary Fontan; TCPC, total cavopulmonary connection with an intra-atrial tunnel.

Figure 1 shows age at the first surgical procedure, age when the SVT was first documented, and age at the first ablation procedure for each patient separately. The number of palliative or corrective surgical procedures performed before catheter ablation ranged from 1 to 4 (2±1). Mean age at which the first and last surgical procedures were performed was 5±7 and 14±11 years, respectively. On average, SVT appeared 18±9 years after the first surgical intervention. Ablative therapy was applied 6±5 (0 to 19) years after first documented SVT episode.

Characteristics of Postoperative Atrial Tachyarrhythmias
A total of 41 SVT were mapped and subsequently ablated during 38 procedures (Figure 2).

At the onset of the ablation procedure, 10 (24%) of the SVT were incessant or persistent. In the other cases (n=31, 76%), SVT were induced using programmed electric stimulation. Isoproterenol was not required to induce SVT in any of the patients.

Mechanisms of SVT included IART (n=30), AFL (n=4), FAT (n=6), and AF (n=1). Representative examples of activation maps of the different SVT mechanisms and cycle length (CL) of all SVT are plotted in Figure 3.

Multiple SVT occurred in 11 patients (2 SVT: n=7, 3 SVT: n=2, 4 SVT: n=1, 9 SVT: n=1). In 6 patients, SVT were caused by different mechanisms. In 3 patients, multiple SVT were targeted during 1 ablation procedure; the mechanism of these successive ablated SVT were different in 2 of them. In 1 patient, a FAT developed spontaneously after
elimination of focal AF; this ablation procedure has recently been described in detail. In the case of both a right and left atrium, most SVT originated from the right atrium; only 1 IART and 1 FAT arose from the left atrium.

Procedural Outcome
Mean procedural and fluoroscopy time was 258±110 and 49±27 minutes, respectively.

Tachycardia termination during ablation was achieved in 32 (78%) of all SVT. The outcomes for the different types of SVT according to their mechanism are shown in Figure 2. A bidirectional conduction block was established in 75% of the patients with AFL. Twenty-two (73%) IART terminated during ablation. In 2 patients, SVT converted to another regular SVT (n=1) or AF (n=1) during ablation. In the case of an unsuccessful ablation procedure (n=6), the SVT was electrically cardioverted to sinus rhythm (n=5) or terminated by overdrive pacing (n=1).

In patients with IART, critical conduction isthmuses that were successfully ablated were located between areas of scar tissue (n=18) or between scar tissue and anatomic structures (n=3). In all patients with more than 1 reentry circuit, critical pathways of conduction of successive SVT were located at different sites in the atrium.

All FAT (n=6) terminated during ablation. They originated from the right atrial free wall (n=1), the interatrial

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Age (First Ablation Procedure)</th>
<th>Diagnosis of SVT</th>
<th>Successful Ablative Therapy</th>
<th>Long-Term Outcome</th>
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<tr>
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<td>IART, A-F, FAT, D(+)</td>
<td>yes, yes, yes, yes</td>
<td>AF, died</td>
<td></td>
</tr>
</tbody>
</table>

IART indicates intra-atrial reentrant tachycardia; SR, sinus rhythm; C-SVT, conversion to another SVT; C-AF, conversion to AF; HTX, heart transplantation; D(+), usage of antiarrhythmic drugs; D(−), no antiarrhythmic drug usage.

![Figure 1](http://circ.ahajournals.org/figure/1)

**Figure 1.** Age at the first surgical procedure (224), diagnosis of the SVT (●), and ablation procedure (224). Patients are ranked according to increasing age at the first diagnosis of SVT.
septum (n=4), or the midleft atrial posterior wall (n=1). All areas of earliest activation were embedded within low-voltage regions.

In the patient with AF, encircling an area of continuous electric activity at the interatrial septum resulted in elimination of AF. The ablation procedure in this patient is described in detail in a previous article.16

On hospital discharge, all patients had sinus rhythm. Procedural complications were not observed in any of the patients.

Long-Term Outcome
The follow-up period was 53±34 months. Two subjects were lost to follow-up. Three patients died of progressive heart failure; rhythm before death was AF in 2 of them. One had heart failure after a surgical procedure consisting of banding a dilated Blalock shunt as a last attempt to improve hemodynamics. Two subjects underwent heart transplantation; 1 had AF and 1 had sinus rhythm.

The median number of follow-up visits was 15 (0 to 39); the mean interval between successive visits was 4±3 months.

Ten patients underwent multiple ablation procedures (n=2, 7 patients; n=3, 2 patients; n=10, 1 patient). Patient 7 had multiple successful ablation procedures (n=10) for SVT caused by various mechanisms; she finally underwent cardiac surgery, during which an extracardiac conduit was placed between the superior caval vein and the pulmonary artery.

Recurrent SVT after successful ablative therapy were observed in 9 patients. In Figure 4, intervals between the first ablation procedure and successive recurrences are plotted. In addition, the last documented rhythm at the end of the follow-up period is also demonstrated. Averaged time between ablative therapy and recurrence of the AT ranged from 2 to 72 (15±17) months. Despite recurrences, sinus rhythm was achieved in 55% of these patients.

Before ablation, 1 patient used aspirin and 12 patients coumarin type-oral anticoagulants. One patient died of progressive heart failure 6 years after ablative therapy. At that time, he had been using oral anticoagulants for 8 years. During postmortem examination, a large thrombotic mass was found in the free wall of the right atrium. Two other patients used oral anticoagulants only during the first 6 months after ablative therapy.

The long-term outcome for the different SVT or specific combinations separately is summarized in Figure 5. At the end of the follow-up period, the majority of the remaining patients (72%) who did not undergo cardiac surgery (n=11) were in sinus rhythm (n=8), and antiarrhythmic drugs were used by only 2 of them.

Discussion
This study reports on the mechanisms, procedural success, and long-term outcome of ablative therapy of late postoperative SVT in patients with UVH. Earlier series included a small number of patients with UVH and did not analyze specifically the substrate and mechanism. Late postoperative SVT in this study population is most often due to
IART related to areas of scar in the right atrium, but they also have various mechanisms, including FAT, AFL, and AF. In some patients with multiple SVT, recurrent SVT were caused by different mechanisms. Even in this population of patients with complex anatomy and severely enlarged atria, catheter ablation is a potentially curative treatment with a procedural success rate of 79%. However, patients may have new tachycardia mechanisms over time, caused by progressive atrial dilatation and fibrosis and need repeat procedure. Overall, despite recurrent SVT requiring multiple ablation procedures, sinus rhythm at the end of the follow-up was achieved in 8 of 11 of the patients who did not undergo additional cardiac surgery (Table 2).

Atrial Tachyarrhythmia Mechanism

Consistent with other reports on the mechanism underlying postoperative SVT in patients with a variety of congenital heart defects, IART and AFL were also frequently observed in patients with UVH.17,18

Electrophysiological studies in patients after modified Fontan repair for functional single ventricle have demonstrated prolongation of the atrial refractory period.19 This might be the result of right atrial hypertension developing after Fontan repair. Local dispersion of atrial refractoriness combined with chronic bradycardia due to sinoatrial node dysfunction and areas of intra-atrial conduction delay provides the substrate for reentrant tachyarrhythmias. In addition, the presence of conduits, long suture lines, or scar tissue increase the likelihood of an IART because they can serve as barriers of the reentrant circuits.13,20

Atrial tachyarrhythmias of focal origin were also frequently observed. We previously demonstrated that in patients with congenital heart defects, FAT arise mainly from areas where conduction is abnormal.16,21 The atria of patients with UVH contain areas of fibrotic tissue giving rise to local dissociation in conduction, which favors development of FAT as electric uncoupling allows focal activity to become manifest.22–24

Kirsh et al5 demonstrated that AF is not an uncommon SVT in patients with congenital heart defects. However, reports on the mechanism underlying AF in this population are rare.

Experimental mapping studies have demonstrated that an area of focal activity gives rise to fibrillatory conduction and hence AF on the surface ECG. In line with these experiments, we also found that delineation of an area of focal activity resulted in termination of AF.16

Persistent AF during long-term follow-up developed in patients who were either in sinus rhythm or in SVT, indicating that there is a difference in the mechanism giving rise to persistence of AF. Hence, further studies in larger populations are required to gain insight into the mechanism of AF in this patient group.

Ablative Therapy

Most ablation procedures performed in this study were guided by a 3D electro-anatomic mapping system. Dorostkar et al25 were the first to demonstrate the usefulness of electroanatomic mapping for defining the arrhythmia circuit, facilitating diagnosis and guiding ablative therapy of postoperative SVT in patients with congenital heart dis-
ease. Later, Delacretaz et al26 demonstrated that 3D electroanatomic mapping combined with entrainment mapping allowed accurate identification of critical isthmuses of complex reentry circuits as targets for ablation of postoperative SVT.

Triedman et al27 demonstrated the beneficial effect of an electro-anatomic mapping system over a conventional fluoroscopy–based mapping technique to the outcome of ablative therapy. Comparable to their ablation results, in our study 27% of the IART also did not terminate during ablation despite the use of a 3D electro-anatomic mapping technique. This outcome emphasizes that ablation of IART is difficult and should be performed in experienced centers. In most IART, critical reentry isthmuses were located between areas of scar tissue, indicating necessity of accurate delineation of low-voltage areas.

In patients who had multiple ablation procedures, target sites for ablation of successive SVT were located at distinct atrial sites. Hence, it is likely that new SVT were caused by progression of atrial cardiomyopathy instead of arrhythmogenicity of prior ablation lesions.

Another interesting finding is that in some patients with UVH and multiple SVT, mapping revealed different mechanisms underlying the SVT; for example, an IART during the first ablation procedure and a FAT in the next procedure. To our knowledge, the presence of different mechanisms underlying consecutive SVT in patients with UVH has not been reported so far.

AFL did not terminate during ablation in 1 patient despite entrainment demonstrating cavo-tricuspid isthmus dependent conduction. This could be due to insufficient lesion depth in fibrotic areas intermingled with myocardial fibers often present in patients with UVH.

The success rate of ablative therapy of FAT in this study population is high, which is in line with other reports on the outcome of ablative therapy of FAT in patients without previous cardiac surgery.

Study Limitations
Because this study was performed retrospectively, Holter monitoring was not consistently performed in every patient to determine the incidence of SVT after ablative therapy. However, most patients with UVH and SVT present symptoms caused by acute hemodynamic deterioration.

During the mapping procedure, crucial pathways of reentrant circuits were mainly selected by analyzing electro-anatomic activation maps. Entrainment techniques in patients with Fontan are known to be nonspecific because concealed entrainment can be demonstrated over large areas. We have attempted to entrain some IART, but pacing in low-voltage areas was difficult and often resulted in conversion to another SVT.

Conclusion
Focal and reentrant mechanisms underlie late postoperative SVT in patients with UVH. Successive SVT developing over time may be caused by different mechanisms. Ablative therapy is a possible curative treatment modality with a procedural success rate of 78%. In patients who had multiple ablation procedures, SVT originated from different atrial sites, suggesting that these new SVT were caused by a progressive atrial cardiomyopathy instead of arrhythmogenicity of prior ablative lesions. Despite recurrent SVT, sinus rhythm during long-term follow-up was achieved in 72%.

Disclosures
None.

References


**CLINICAL PERSPECTIVE**

Catheter ablation has evolved as a possible curative treatment modality for supraventricular tachycardias in patients with univentricular heart. The long-term outcome of ablation procedures is still unknown. In this study, we evaluated the procedural and long-term outcome of ablative therapy of late postoperative supraventricular tachycardias in patients with univentricular heart. These supraventricular tachycardias were due to either a focal or reentrant mechanisms. In patients who had multiple ablation procedures, supraventricular tachycardias originated from different atrial sites, suggesting that these new supraventricular tachycardias were caused by a progressive atrial cardiomyopathy instead of arrhythmogeneity of prior ablative lesions. Successive supraventricular tachycardias developing over time were caused by different mechanisms. The procedural success rate was 78%. Despite recurrent supraventricular tachycardias, sinus rhythm during long-term follow-up was achieved in 72%.
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