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# Atrial Fibrillation Ablation in Adults With Repaired Congenital Heart Disease

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**Abstract**

The incidence of atrial fibrillation (AF) in congenital heart disease (CHD) adults has increased in the past decades due to a longer life expectancy of this population where the subjects are exposed to cardiac overflow, overpressure and structural changes for years.

The literature regarding AF ablation in repaired CHD adults emphasizes the importance of intracardiac echocardiography (ICE) to perform the transseptal puncture and the ablation procedure in the left atrium (LA), both effectively and safely. In small case control studies, where the predominant congenital cardiomyopathy was the atrial septal defect, the most common strategy for ablation was antral isolation of the pulmonary veins showing results, at one year follow-up, similar to those in the general population.

The positive results of AF ablation so far, in this specific population, widen the range of therapeutic options for a group of patients whose only chance has been pharmacological treatment, which has proved to be inefficacious in most of the cases and not free from adverse events.

## Introduction

The management of congenital heart diseases (CHD) has much changed during the past few decades. The advances in surgical techniques and treatment of the associated comorbidities have significantly increased the life expectancy of this population. Consequently, the incidence of atrial arrhythmias, and especially atrial fibrillation (AF), has also increased as more patients reach the adulthood.[1] The most studied and prevalent arrhythmia in CHD patients has always been the intra-atrial reentrant tachycardia which is closely related to the surgical procedure , but this is probably about to change as the incidence of AF in these patients is growing independently of the surgery.[2,3,4,5] The prevalence of AF for the CHD population varies in the literature between 3.7 to 15%, a notoriously higher percentage than that of the general population which is around 0.95%.[6,7,8] A different pathophysiology and the conjunction of risk factors (those factors present in the general population and those unique for these patients) could explain the increased prevalence. In a recent multicenter cohort of adults with tetralogy of Fallot, AF was the most prevalent atrial tachyarrhythmia over the age of 55 years.[9] The presence of a higher thromboembolic risk, the higher morbidity and mortality and the increased risk for heart failure in CHD patients deserve special attention.[10,11] Antiarrhythmic drugs have been for a long time the only treatment used in CHD patients, not always with success. Koyak et al. in 2013 investigated the efficacy of antiarrhythmic drugs in 92 CHD patients with new onset supraventricular tachycardia, and 68% of them were AF.[3] Class III drugs were the most effective to prevent recurrences but at the same time they were the drugs with more side effects (dizziness, bradycardia, intolerance) and all patients taking amiodarone presented thyroid toxicity, representing an important limitation for the treatment of this young population. Class I, II and IV antiarrhythmic drugs were not superior in preventing recurrences than no antiarrhythmic therapy. In addition, class I drugs are contraindicated in most cases due to the presence of structural heart disease and ventricular dysfunction. Therefore, sotalol remains the only option of treatment as it is one of the most effective drugs with no extra-cardiac toxicity (2/3 of patients were free from arrhythmia for at least one year follow-up). The limitations of antiarrhythmic drugs pave the way for AF ablation as an alternative of treatment, although the associated technical difficulties prevent its introduction in the majority of the EP labs.

**Key Words:**
Atrial Fibrillation, Ablation, Congenital Heart Disease.

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None.

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## Pathophysiology

The initiation of AF in CHD adults can be the result of several factors: a) left atrial (LA) dilation due to volume overload in the increased right-sided flow CHD or secondary to high systemic blood pressure or left ventricular dysfunction; it is well known the relationship between atrial dilation and the development of

**Table 1:** Summary of the three publications with the largest number of CHD repaired patients who underwent atrial fibrillation ablation

| | Number of CHD patients | Groups | Type of AF | Ablation strategy | Success |
|---|---|---|---|---|---|
| Philip et al. 2012 | 36 | All kind of CHD vs non-congenital structural heart disease (NCSHD) | Paroxysmal (72%) Persistent (27.7%) | PVI +SVC +other LA lines | 84% patients free of symptomatic AF vs 86% in NCSHD at 300 days |
| Santangeli et al. 2011 | 39 | Percutaneous device in ASD due to ostium secundum | Paroxysmal (33%) Persistent (67%) | PVI + SVC in paroxysmal/ PVI+SVC+post wall+septal LA+CFAEs in persistent or long standing AF | 77% patients free of symptomatic and/or documented AF at 14+months |
| Lakkireddy et al. 2008 | 45 | Surgical or percutaneously treated ASD and PFO patients vs age-gender matched controls | Paroxysmal (60%) Persistent (26%) Permanent (14%) | PVI + CTI | 76% patients free of symptomatic and/or documented AF vs 82% in the control group at 15+4 months |

fibrosis and electrical heterogeneity which is the cornerstone for the perpetuation of AF; LA dilation has been observed either in pre and post repaired patients, as surgery cannot prevent the initiation of atrial arrhythmias in most cases,[8,12] b) incisional scarring predisposing to reentrant circuits (percutaneous closure of septal defects has been observed as a protector against AF); this circuits could explain the common coexistence of AF and atrial tachycardia that has been observed in these patients,[13] c) sinus node disease (either primary or post surgery) which facilitates the activation of other atrial triggers, d) blood desaturation in chronic cyanotic patients has been reported as a possible independent factor for the development of AF, although this has not been demonstrated due to its relation to complex CHD and Eisenmenger syndrome with hemodynamic worsening.[8]

## Risk Factors for The Development of Atrial Fibrillation

As previously mentioned, adult CHD patients are subjected to the usual risk factors of the general population (age, hypertension, functional class, obesity, diabetes…) and to some particular risk factors that only affect this group of patients: age at surgery, complexity of the CHD, complexity of surgery, diseases with increased right-sided flow or blood desaturation, among others. Gender seems not to play a role in CHD adults as it does in the general population.

In a study, conditions disproportionately associated with atrial fibrillation were left-sided obstructive lesions, incompletely palliated CHD, and, to a lesser extent, Fontan surgery.[14] Older age, left atrial enlargement, lower left ventricular ejection fraction, and number of cardiac surgeries have been independently associated with AF.[9] Atrial fibrillation is a well-recognized sequela of large, unrepaired atrial septal defects in adults. Early but not late (i.e., >40 years) closure of the atrial septal defect reduces its prevalence postoperatively.[15,16]

## Transseptal Puncture

One reason for many physicians to not include AF ablation in the treatment of CHD patients is the difficulty in performing the transseptal puncture. The modified anatomy makes the usual anatomical references and maneuvers to identify the fossa ovalis for a safe puncture useless.

All publications regarding transseptal puncture in complex CHD or in simple CHD in which the atrial septum and/or its adjacent structures have been modified, are using intracardiac echocardiography (ICE) to access the left atrium. After femoral venous puncture, a phased-array ultrasound imaging catheter is advanced into the right atrium to obtain a direct view of the atrial septum and to localize a safe site for the puncture, as the usual pull back from the superior vena cava and jump into the fossa ovalis are not applicable. The transseptal puncture is performed as usual using a long transseptal sheath and a long needle.

The transseptal puncture technique using ICE has also been described in big detail and proved safe in patients with surgical interatrial patches and closure septal devices, the last ones more and more frequently used due to the widespread of percutaneous techniques. ICE is used to identify the portion of the septal wall not covered by the device, which can be found in a posteroinferior position in a majority of cases as the device is normally anterosuperiorly oriented[17](Fig. 1). When a free portion of the wall is not present, transseptal puncture can be done through the closure device and ICE provides an essential support to directly visualize the appropriate site for the puncture. The technique for the perforation of percutaneous closure devices was first described in 2011 by Santangeli et al.[12] Briefly, once the needle had crossed the device, the 8Fr dilator was removed and an upsized 11Fr dilator was advanced over the wire to dilate the access site across the device. Finally, the transseptal sheath was introduced into the left atrium. No shunt was observed in the follow-up of these patients.

Another handicap when crossing a repaired interatrial septum is its thickness and/or stiffness. In some cases there can be also calcification of the patch or the septum itself. In these cases, perforation of the septum with the usual needle may not be possible. The use of a RF-assisted transseptal needle[18] or a surgical electrocautery pen in the cut mode placed on the proximal hub of the needle while tenting of the septum can solve this problem.

## Atrial Fibrillation Ablation Strategy and Results

Publications about AF ablation in CHD adults are scarce. With

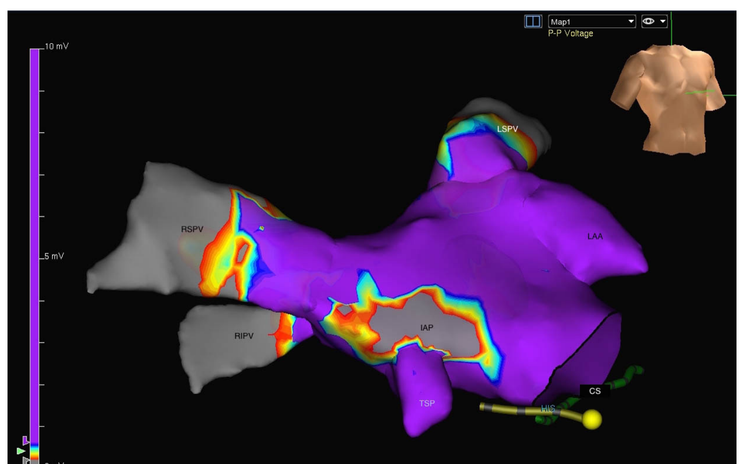


**Figure 1:** Left atrium voltage map in a right anterior oblique view. A surgical interatrial patch (IAP) can be seen as a grey spot as it has no electrical conduction. The transseptal puncture was performed posteriorly to the patch where the native tissue was thinner and easier to puncture. RSPV: right superior pulmonary vein; RIPV: right inferior pulmonary vein; LSPV: left superior pulmonary vein; LAA: left atrial appendage; IAP: interatrial patch; TSP: transseptal puncture; CS: coronary sinus

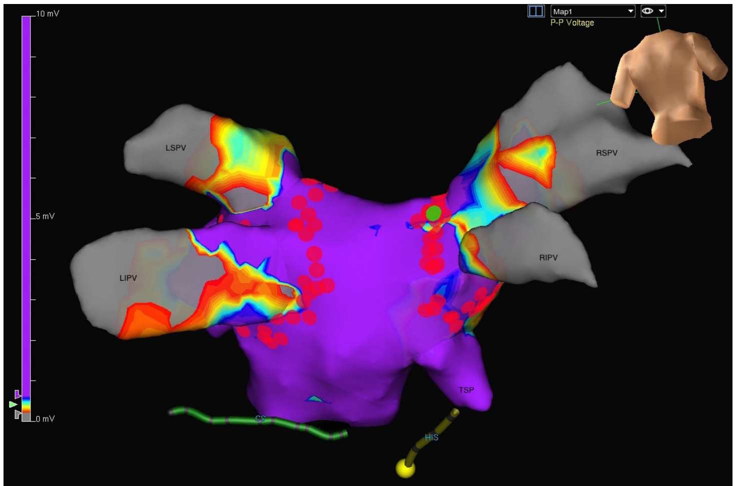


Figure 2: Left atrium voltage map in a posteroanterior view. The red dots represent the lines of radiofrequency ablation in a circular antral modality around the pulmonary veins ostia. RSPV: right superior pulmonary vein; RIPV: right inferior pulmonary vein; LSPV: left superior pulmonary vein; LIPV: left inferior pulmonary vein; TSP: transseptal puncture; CS: coronary sinus

respect to catheter ablation of AF, operators have largely mimicked and adapted standard strategies, including isolation of pulmonary venous antra (Fig. 2), connecting lesion sets to the left-sided AV annulus, and cavotricuspid isthmus ablation.[1] These complex procedures require careful anatomic planning and ideally utilize techniques for real-time and/or registered volume imaging of the heart to facilitate visualization of relevant anatomy.

There are only three reports with a considerable number of patients which are summarized in Table 1. The largest report was published in 2012 by Philip et al. who included all kind of CHD (61% of patients had an atrial septal defect) and reviewed prospectively 36 patients.[19] A total of 34 patients had undergone some kind of reparation, a surgical intervention in the majority of them. These patients were compared to a control group of 355 patients with non-congenital structural heart disease (57% valvular heart disease). The last group was slightly older and had a higher prevalence of diabetes, hyperlipidemia and hypertension. Paroxysmal AF was the predominant type of AF in both groups. ICE was used during the transseptal puncture and to guide the position of the ablation catheter during ablation. An 8mm tip RF ablation catheter (70W, 55°C) was used to isolate the four pulmonary veins targeting the pulmonary vein antrum in all cases. The second site most approached was the superior vena cava. Other sites were the LA septum, the mitral isthmus or the coronary sinus. Intravenous heparin was administered during the procedure to maintain an activated clotting time of 350 to 400 seconds. After a mean of 300 days of follow-up, 84% of patients in the CHD group and 86% of patients in the non-congenital structural heart disease group were free of symptomatic AF under antiarrhythmic therapy. After 4 years, the proportion was 61 and 69%, respectively. The procedure failed in the only patient with Tetralogy of Fallot and also in a patient with transposition of the great vessels due to technical problems to reach the LA with the ablation catheter.

There are two other studies focused in adult patients with repaired atrial septal (ASD) defects. In 2008 Lakkireddy et al.[17] reported 45 patients with atrial septal defect or patent foramen ovale repair, 22 surgically and 23 percutaneously. Pulmonary vein antral isolation was performed in all patients. Additional lines were done if required (cavotricuspid isthmus in 6 patients and a figure of 8 flutter ablation in one case). The ablation catheter used was either an 8-mm non-irrigated or a 3.5-mm irrigated catheter. Warfarin was not discontinued for the procedure and additional enoxaparin was administered when the international normalized ratio (INR) was under.[2] The mean follow-up duration was 15+ 4 months. Failure was defined as any documented and/or symptomatic atrial arrhythmia. The repaired ASD patients were compared to an age-gender-AF type matched controls. Failure between 3 and 12 months after the procedure was slightly higher in ASD patients than in the control group (24% vs 18%, P=0.7). Recurrence rates were also higher in the non-paroxysmal group compared with the paroxysmal patients (33% vs 19%, P≤0.4) as in the general population.

In 2011, Santangeli et al.[12] published a study including 39 patients with percutaneous atrial septal closure devices due to ostium secundum defects. ICE was used as a guide for the transseptal puncture as previously described. In paroxysmal AF patients pulmonary vein antrum isolation and isolation of the superior vena cava were performed. In persistent and long-standing persistent AF patients applications on the entire posterior wall to the coronary sinus and the left side of the septum and ablation of complex fractionated atrial electrograms in the left atrium and the coronary sinus were added to PV and superior vena cava isolation. At the end of the procedure an infusion of isoproterenol was administered to show vein reconnection or extra pulmonary vein firing. Warfarin was not discontinued and intravenous heparin was administered during ablation to reach an activated clotting time >300. At mean follow-up of 14 + months, 77% of patients were free from atrial arrhythmia recurrence (defined as any atrial arrhythmia lasting for at least 30 seconds).

The presence of a persistent left superior vena cava has been described as a trigger for AF. Its isolation has been performed in small case series[20] by advancing a circular mapping catheter into the left superior vena cava and eliminating all fractionated signals inside the vein by pulling back an irrigated RF catheter. Isolation of the left superior vena cava has been also performed with cryothermal energy when the diameter of the proximal coronary sinus allows the introduction of a cryoballoon.

## Summary

The prevalence of AF in CHD adults is increasing due to longer life expectancy, the presence of more cardiovascular risk factors (hypertension, diabetes, obesity) in older ages and intracavitary pressure changes and cardiac remodeling. Antiarrhythmic drugs are less efficacious to prevent recurrences in CHD patients and drugs have side effects which are not desirable in this young population who need a long-term treatment. AF ablation can be an option to help improving the efficacy of antiarrhythmic drugs or even as a single treatment.

The major difficulty to perform the AF ablation procedure is the distortion in the cardiac anatomy that makes the usual position and maneuver of the catheters unhelpful. The changes in the anatomical references are especially relevant during the transseptal puncture and to guide the position of the catheters during ablation. Therefore, the use of ICE to have a direct visualization of the anatomical structures is almost imperative. An additional barrier is the presence of interatrial septal patches or closure devices. Again, ICE and the use of radiofrequency energy to perforate the septum can help overcoming these obstacles.

There are only few studies with small samples or case reports about AF ablation in CHD adults. The majority of patients in these publications have repaired atrial septal defects, who have less anatomical changes than major congenital heart defects, and paroxysmal AF. The standard strategy for ablation in all cases was antral PV isolation but the additional applications or atrial lines were heterogeneous between the different studies. Although CHD patients are more vulnerable for presenting a thromboembolic event and therefore the strategy for anticoagulation during ablation was aggressive, no complication related to the procedure was reported in any case.

The clinical results reported from the available publications are promising, with similar success rates when compared to the general population. However, the fact that the authors are all members of experienced teams should be kept into account. A review of eight prospective randomized trials in the general population, comparing AF ablation with antiarrhythmic drug therapy or rate control agents alone, reported a success rate of 77.8% in the AF ablation arm.[21] In line with these results, the success rate for AF ablation in CHD patients ranged from 76% to 84%.

It has been observed a higher incidence of recurrences in the long-term follow-up of postoperated CHD patients after atrial flutter ablation,[4,5] however, information about the outcomes during the long-term follow-up after AF ablation for CHD patients is missing. For patients with drug refractory AF or those not suitable for catheter ablation, AV nodal ablation might be considered. AV nodal ablation with post ablation ventricular pacing in patients with CHD has been reported in a small series,[22] but, again, information on the long term is very limited.

In conclusion, the use of AF ablation in the CHD population looks promising and safe, nevertheless, more studies are needed to provide further learnings and conclusions.

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