Atrial Fibrillation Ablation in Adults With Repaired Congenital Heart Disease

Marta Acena MD, Ignasi Anguera MD PhD, Paolo D. Dallaglio MD, Marcos Rodriguez MD, Xavier Sabaté MD PhD

Heart Disease Institute, Bellvitge Biomedical Research Institute-IDIBELL, Bellvitge University Hospital.

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 overload, 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. 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. 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%. 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. 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. 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. 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.

Disclosures:
None.

Corresponding Author:
I. Anguera
Cardiology Department
Bellvitge Hospital, L’Hospitalet
Barcelona, Spain.
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,11 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, un repaired 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 (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 RF 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 needle15 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
isolation was performed in all patients. Additional lines were done to repair, 22 surgically and 23 percutaneously. Pulmonary vein antral isolation was performed in 45 patients with atrial septal defect or patent foramen ovale (ASD) defects. In 2008 Lakkireddy et al. reported problems to reach the LA with the ablation catheter. In a patient with transposition of the great vessels due to technical difficulties, ICE was used during the transseptal puncture and to guide the position of the catheters during ablation. Therefore, the changes in the anatomical distortion in the cardiac anatomy that makes the usual position and maneuver of the catheters unhelpful. 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 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 presence of a persistent 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.

The presence of persistent left superior vena cava has been described as a trigger for AF. Its isolation has been performed in small case series 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.

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 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.

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 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.
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 anterolateral 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. 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, 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, but, again, information on the long-term risk 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.

References