Catheter Ablation for Atrial Fibrillation in Adults With Congenital Heart Disease
Lessons Learned From More Than 10 Years Following a Sequential Ablation Approach

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ABSTRACT

OBJECTIVES This study aimed to evaluate the impact, safety, and success of atrial fibrillation (AF) ablation in adults with congenital heart disease (ACHD) transferring ablation strategies established in normal hearts.

BACKGROUND AF is an emerging arrhythmia in ACHD.

METHODS Fifty-seven consecutive ACHD (median age 51.1 ± 14.8 years) with drug-refractory AF were analyzed who underwent catheter ablation between 2004 and 2017. CHD was classified according to its complexity into mild (61.4%), moderate (17.5%), and severe (21.1%) lesions. AF ablation was performed in 104 procedures following a sequential ablation approach.

RESULTS Of the 57 patients, 30 underwent corrective surgery, 6 underwent palliative surgery, 5 had catheter interventions, and 16 were natural survivors. Follow-up was available for all patients (median 41 ± 36 months). The median duration of cyanosis was 9.2 ± 19.7 years, and the time of volume or pressure overload prior to corrective surgery or intervention was 26.1 ± 21.2 years and 18.1 ± 15.8 years, respectively. The Kaplan-Meier estimate for arrhythmia-free survival following the index ablation procedure was 63% for 1 year and 22% for 5 years. Performing subsequent ablation procedures (2.0 ± 0.5), the Kaplan-Meier estimate significantly improved, with 99% for 1 year and 83% for 5 years (p < 0.01). Five patients died during follow-up due to their underlying CHD condition or underwent transplantation.

CONCLUSIONS AF ablation strategies established in normal hearts can be transferred to ACHD. The treatment is safe and effective with acceptable long-term results. Varying anatomical pre-conditions and the heterogeneous population itself are challenging and contribute toward a higher reablation rate. Therefore, AF ablation in ACHD should be reserved for dedicated and highly specialized teams. (J Am Coll Cardiol EP 2018;–:––) © 2018 by the American College of Cardiology Foundation.
Society expert consensus document on the recognition and management of arrhythmias in ACHD recommends that AF ablation might be considered after failure of trials of cardioversion with pharmacologic rhythm control (1). There is lacking evidence regarding individual risk stratification, ablation strategies, and long-term follow-up after AF ablation in ACHD. Predominantly, operators have largely transferred ablation approaches including pulmonary vein isolation (PVI), connecting linear lesion sets to the left-sided mitral isthmus, and cavotricuspid isthmus ablation (1), and no reliable data currently exist for substrate-based ablation approaches. In addition, individual disorders and cardiac lesions contribute to the arrhythmogenesis and perpetuation of cardiac arrhythmias and therefore increase the severity of AF ablation in ACHD. Recently, Tilz et al. (2) reported their findings following a sequential ablation strategy for persistent AF (PERS) in patients with structurally normal hearts. This approach included basically PVI, linear lesion sets, and complex fractionated atrial electrogram (CFAE) ablation, and leads to a long-term maintenance of sinus rhythm (SR) with a rate of 45% after multiple ablation procedures (2). These findings are in line with the data from O’Neill et al. (3) demonstrating that a stepwise ablation approach focusing on termination of AF during ablation leads to improved outcomes in patients with PERS without CHD. AF ablation in the setting of ACHD remains challenging and therefore, this study aimed to evaluate the impact, safety, and mid- to long-term success of AF ablation in ACHD transferring AF ablation strategies from patients without cardiac anomalies to ACHD.

**METHODS**

**STUDY POPULATION.** Between 2004 and 2017, a total number of 67 consecutive ACHD (median age 50 years; range: 17.5 to 76.3 years) with drug-refractory AF underwent a total of 150 left atrial (LA) ablation procedures for AF at our institution. Ten ACHD with preceding LA ablation attempts at another center as well as those candidates with previous surgical treatment for AF were excluded. Consequently, 57 ACHD with a primary interventional approach for drug-refractory AF at our institution remained, in whom 104 procedures were performed and analyzed in this observational single-center study. Previous catheter ablation for right atrial tachycardia (AT) has been performed in 17 of 57 patients (29.8%) focusing on re-entrant tachycardia based on either macro re-entrant and/or micro re-entrant focal mechanism. The congenital cardiac anomalies were classified according to the 32nd Bethesda consensus document (4) into their levels of complexity: mild (61.4%), moderate (17.5%), and severe (21.1%) (Table 1). Ninety percent of patients underwent previous cardiac surgery and 21% had a history of cyanosis. Paroxysmal AF (PAF) and PERS were defined according to the recommendations of the 2012 Heart Rhythm Society/European Heart Rhythm Association/European Cardiac Arrhythmia Society Expert Consensus Statement on Catheter and Surgical Ablation of Atrial Fibrillation (5).

**ELECTROPHYSIOLOGICAL STUDY.** Prior to each procedure, LA thrombus formation was excluded. In patients on vitamin K antagonists, oral anticoagulation was stopped 3 days before ablation and replaced by intravenous heparin to maintain a partial thromboplastin time of 2 to 3 × the normal value or bridged with low-molecular-weight heparins. Since 2012, ablation was performed under therapeutic international normalized ratio values of 2 to 3. Direct oral anticoagulants were stopped the day before and continued after the procedure when pericardial effusion had been ruled out. The intervention was performed under deep sedation utilizing midazolam, fentanyl, and a continuous infusion of propofol. One standard catheter was positioned inside the coronary sinus. Two SL1 sheaths (St. Jude Medical, Minneapolis, Minnesota) were advanced to the LA using a modified Brockenbrough technique (6). After transseptal catheterization, intravenous heparin was administered, targeting an activated clotting time of >300 s. Three-dimensional electroanatomical reconstruction using the CARTO system (Biosense Webster, Diamond Bar, California) and ablation were performed using a 3.5-mm-tip catheter (ThermoCool Navi-Star, Biosense Webster) (2,6,7). Low voltage for abnormal atrial areas was defined as an amplitude of ≤0.5 mV. In case of right atrial ablation, the anatomic course of the phrenic nerve was identified using pace mapping. The location of the esophagus was visualized using an 8-F nasogastric tube (Nutritub, B. Braun, Melsungen, Germany). Before AF intervention, a baseline electrophysiological study was performed to evaluate and, if necessary, eliminate coexisting tachycardia. Written informed consent was obtained from each patient before the procedures after intensive elucidation with special respect to the individual CHD condition. The study was approved by the Institutional Review Board.
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