Prophylactic Atrial Arrhythmia Surgical Procedures With Congenital Heart Operations: Review and Recommendations

Constantine Mavroudis, MD, John M. Stulak, MD, Niv Ad, MD, Allison Siegel, MSSA, Alessandro Giamberti, MD, Louise Harris, MD, Carl L. Backer, MD, Sabrina Tsao, MD, Joseph A. Dearani, MD, Nihal Weerasena, MD, and Barbara J. Deal, MD

Johns Hopkins Children's Heart Surgery, Florida Hospital for Children, Orlando, Florida; Division of Cardiovascular Surgery, Department of Surgery, Mayo Clinic, Rochester, Minnesota; Inova Fairfax Hospital, Cardiac Surgery Inova Heart and Vascular Institute, Falls Church, Virginia; Pediatric and Adult Congenital Heart Surgery, IRCSS Policlinico San Donato, San Donato M.se (MI), Italy; Division of Cardiology, Peter Munk Cardiac Centre, Toronto Congenital Cardiac Centre for Adults, Toronto, Ontario, Canada; Cardiovascular-Thoracic Surgery and Division of Cardiology and the Department of Pediatrics, Ann & Robert H. Lurie Children's Hospital of Chicago, Chicago, Illinois; and Division of Cardiothoracic Surgery, The General Infirmary, Leeds, United Kingdom

Specific congenital heart anomalies significantly increase the risk for late atrial arrhythmias, raising the question whether prophylactic arrhythmia operations should be incorporated into reparative open heart procedures. Currently no consensus exists regarding standard prophylactic arrhythmia procedures. Questions remain concerning the arrhythmia-specific lesions to perform, energy sources to use, need for atrial appendectomy, and choosing a right, left, or biatrial Maze procedure. These considerations are important because prophylactic arrhythmia procedures are performed without knowing if the patient will actually experience an arrhythmia. This review identifies congenital defects with a risk for the development of atrial arrhythmias and proposes standardizing lesion sets for prophylactic arrhythmia operations.

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Patients with congenital heart disease (CHD) are at increased risk for the development of late arrhythmia, which adversely impacts ventricular function, physical well-being, and long-term survival [1, 2]. Arrhythmias are the leading source of morbidity requiring hospitalization in adulthood after repair of CHD [3, 4]. Efficacy of catheter and surgically based treatment for specific atrial arrhythmia substrates has been well established; whether these therapeutic tenets can be extended to the prevention of arrhythmia development is unknown and untested. Prophylactic arrhythmia operations are proposed for patients with specific anatomic substrates as an effort to reduce the development of late arrhythmias. If effective, one would expect a significant decrease in late morbidity and perhaps a reduction in the risk of sudden death from arrhythmias.

There is no unanimity of opinion as to what constitutes a standard therapeutic arrhythmia procedure, let alone a prophylactic intervention [5–9]. Application of therapeutic lesion sets with demonstrated efficacy may have unwanted consequences that pose concerns when used as prevention. Although the aim of the original Cox-Maze procedure was to treat atrial fibrillation (AF) with low morbidity, concerns for sinus node dysfunction led to modification of the lesion sets that was designed to minimize potential harm to the sinus node. Numerous technical considerations are paramount, including optimal prophylactic lesion sets, type of energy source, proximity of lesions to important structures, need for left atrial (LA) appendectomy, and application to 1 or both atria. These complex issues are balanced by the fact that prophylactic lesions are performed without advanced knowledge that arrhythmias will actually develop postoperatively. Invocation of bioethical principles of nonmaleficence, beneficence, patient autonomy, and justice are relevant and apply.

The purpose of this article is to identify patients undergoing CHD repair who are at significant risk for the development of late atrial arrhythmias, review the literature of concomitant prophylactic and therapeutic arrhythmia ablation during CHD repair, and offer standardized lesion sets for future interprogram comparisons. A meeting, *Arrhythmia Surgery in Patients with Congenital Heart Disease*, was called in May 2013 with all the authors to present his or her vast experience in arrhythmia prevention and ablation in patients with CHD. After the presentations, a discussion took place to develop a set of lesions to be used in these patients in an attempt to prevent arrhythmias. It was determined that the anomalies to be discussed would include Ebstein's anomaly

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Address correspondence to Dr Mavroudis, 2501 N Orange Ave, Ste 540, Orlando, FL 32804; e-mail: constantine.mavroudis.md@flhosp.org.

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Abbreviations and Acronyms

AF = atrial fibrillation
AFL = atrial flutter
ASD = atrial septal defect
AV = atrioventricular
CHD = congenital heart disease
EA = Ebstein's anomaly
IVC = inferior vena cava
LA = left atrial
RA = right atrial
RF = radiofrequency
SVC = superior vena cava
SVT = supraventricular tachycardia
TGA = transposition of great arteries
TOF = tetralogy of Fallot

(EA), atrial septal defect (ASD), Fontan physiology, and tetralogy of Fallot (TOF). All authors provided their personal libraries and presentations for the purpose of providing guidance for physicians working with this patient population. After the meeting, a search was completed on PubMed for all articles published after 2000 using the following list of search terms: arrhythmia in CHD, prophylactic arrhythmia, sudden death in CHD, lesion sets, EA, TOF, ASD, single-ventricle physiology (Fontan, univentricular), atrial flutter (AFL), AF, surgical ablation, and energy sources with surgical ablation arrhythmias. Because the authors are from all over the world, communication continued through e-mail, telephone conferences, and in-person review as the article was developed and completed.

Lesion-Specific Literature Review

Large clinical CHD database analyses with long-term follow-up have demonstrated the prevalence of late arrhythmias and identified the anatomic substrates at highest risk [2, 10–15]. Between 1985 and 2000, the prevalence of severe CHD in adults increased by 85%, and now approximately 49% of severe CHD exists in adults rather than children [2]. Bouchardy and colleagues [14] evaluated the impact of atrial arrhythmias in their adult database and found that in patients with no reported arrhythmias by 18 years of age, greater than 50% experienced atrial arrhythmias by 65 years. The impact of atrial arrhythmias increased the risk of early mortality by 50%, more than doubled the risk of stroke or heart failure, and caused a 3-fold increase in cardiac interventions. The anatomic diagnoses with the highest prevalence of supraventricular tachycardia (SVT) in the Bouchardy and colleagues study were EA (33%), transposition of the great arteries (TGA) status post atrial switch (28%); single-ventricle physiology (24%), ASD (18.9%), and TOF and truncus arteriosus (15.5%) [14].

Ebstein's Anomaly

The arrhythmia concerns associated with EA are right-sided accessory connections (15%–30%), the late development of AFL/AF presumably related to atrioventricular (AV) valve regurgitation and atrial dilatation (65% of SVT episodes), and an increasing awareness of the high risk of sudden death in this population (7%–15%) [16–18]. Catheter ablation of right-sided accessory connections in EA is associated with technical challenges and high arrhythmia recurrence owing to right atrial (RA) dilatation, AV annulus distortion, and muscle continuity between the right atrium and right ventricle [18–21]. In a large follow-up series of patients with EA after tricuspid valve repair, more than 60% were rehospitalized for cardiac causes at 20 years, the most common of which was arrhythmia [16]. The cause of the increased risk of sudden death is not clear and could be a consequence of rapid atrial arrhythmia resulting in low cardiac output or primary ventricular arrhythmia [18, 22–24]. Efforts to reduce the occurrence of atrial arrhythmias in patients with EA are thus critical [16–19].

Transposition of the Great Arteries

Atrial switch repairs for TGA are associated with loss of sinus node function in 50% of patients at 20-year follow-up and the development of AFL in approximately one quarter of survivors [25, 26]. The atrial switch operation for TGA has been abandoned except in patients with congenitally corrected TGA in whom a double switch operation is performed. If future studies demonstrate prophylactic arrhythmia operation efficacy in other diagnostic substrates, incorporation of cavotricuspid isthmus ablation during the primary reparative procedure for TGA variants undergoing atrial baffle procedures may be considered.

Single-Ventricle Physiology

Perhaps the most challenging patients with CHD are those with Fontan physiology in whom atrial arrhythmias develop in more than 50% of cases [27–32], usually associated with significant RA dilatation and hemodynamically important lesions. Arrhythmias include multiple RA reentrant circuits, focal atrial tachycardia, and an increased incidence of AF. Modification of the Fontan operation has decreased the incidence of late atrial tachycardia from more than 60% in earlier atriopulmonary connection repairs [27, 28] to 13% to 60% in lateral tunnel connections [33–35] and approximately 8% to 15% in extracardiac connections [30–32]. Late arrhythmias in patients with modified connections can be expected to rise with longer follow-up. Catheter ablation in the patient with Fontan physiology has acute success rates of 50% to 70%, with at least 70% recurrence of tachycardia within 2 years [36–38]. Catheter access to the right atrium in patients with extracardiac connections is limited to the transhepatic or transthoracic approach, with attendant morbidity. Certainly patients with previous Fontan operations who are undergoing reoperations should be considered for prophylactic atrial lesion sets.

Atrial Septal Defect

As many as 50% of patients undergoing ASD repairs after 40 years of age [39, 40] were found to experience late