



# Diagnostic and Therapeutic Approach to Arrhythmias in Adult Congenital Heart Disease

*Karine Guerrier, DO, MPH\**  
*Benjamin Hendrickson, MD*  
*B. Rush Waller, MD*  
*Glenn T. Wetzels, MD, PhD*

## Address

\*Le Bonheur Children's Hospital, The Heart Institute, 49 N Dunlap, 3rd Floor,  
Memphis, TN, 38103, USA  
Email: kguerrie@uthsc.edu

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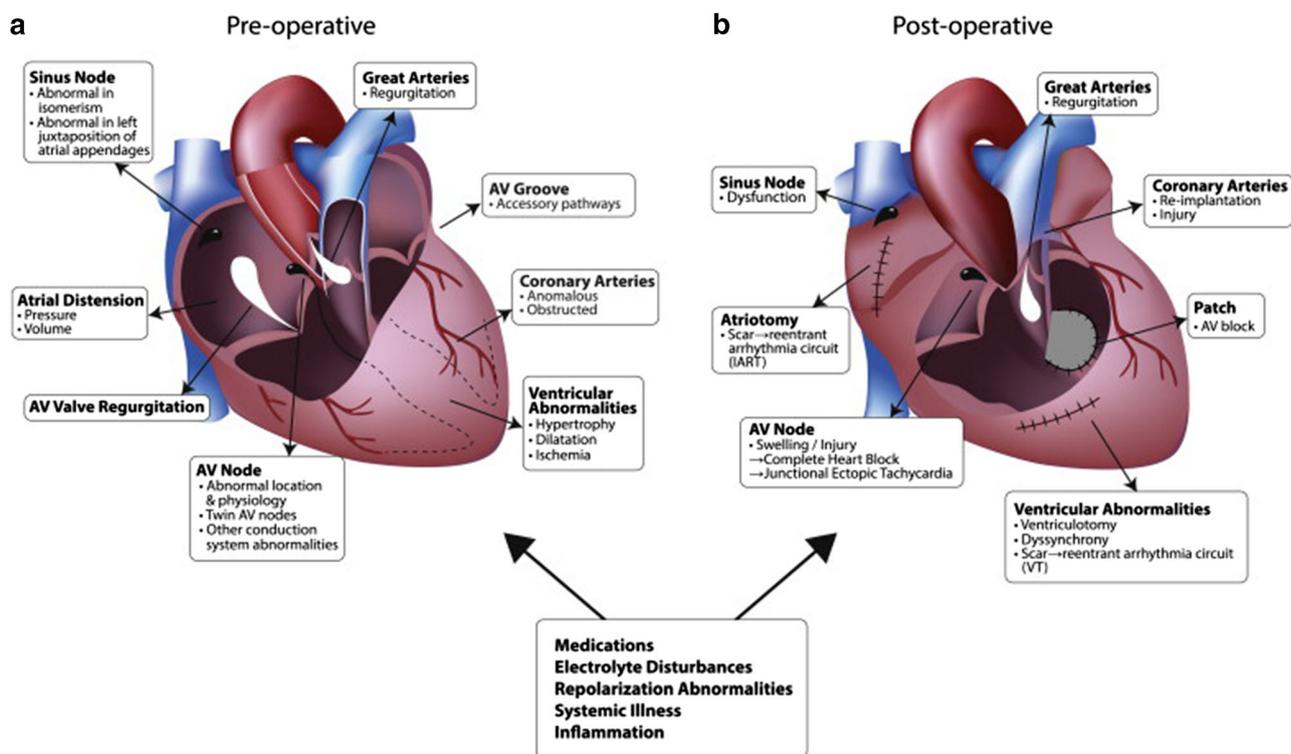
**Keywords** Arrhythmia · Adult congenital heart disease · Bradycardia · Management · Tachycardia

## Abstract

*Purpose of review* Adult survivors of congenital heart disease (CHD) are at increased risk of arrhythmia. The goal of this review is to outline diagnostic and therapeutic approaches to arrhythmia in adult CHD patients.

*Recent findings* Macro-reentrant atrial tachyarrhythmia is the most common arrhythmia encountered in adults with CHD. Approximately 25% of hospitalizations associated with arrhythmia. The risk of ventricular arrhythmia is estimated as high as 25–100 times that for the general population and increased after two decades.

*Summary* Routine ambulatory monitoring is important for arrhythmia risk assessment in adults with CHD. There are limitations, potential adverse effects, and risk of recurrence with antiarrhythmic drugs, catheter ablation, and surgical approaches. Adults with CHD suffer various forms of arrhythmia, are at increased risk of sudden death, and require special consideration for medical and interventional therapy.



**Fig. 1.** Schematic of factors leading to arrhythmias in **a** pre- and **b** post-operative congenital heart disease. AV atrioventricular. Reprinted from *Canadian Journal of Cardiology*, Vol 29 (7), Carolina Escudero MD, FRCPC, Paul Khairy MD, PhD, FRCPC, Shubhayan Sanatani MD, FRCPC, *Electrophysiologic Considerations in Congenital Heart Disease and Their Relationship to Heart Failure*, Pages 821–829 (2013) with permission from Elsevier.

## Introduction

Adults with congenital heart disease (ACHD) comprise a heterogeneous and burgeoning global population with unique risks for major cardiovascular complications, especially arrhythmia, that confront health care providers and patients with complex and difficult challenges [1, 2]. Advancements in surgery, interventional procedures, and medical care are credited for the increased survival and have resulted in a growing and aging population with adults outnumbering children with congenital heart disease (CHD) in developed nations [1]. Treatment has shifted mortality in CHD towards older ages with a distribution closer to that of the general population [3]. North American epidemiologic data for the last decade report growth in the ACHD population around 60% with substantial increases in the most complex diagnoses [4, 5]. In the USA, there are more than 1.4 million adults with CHD, outnumbering children nearly one- and one-half-fold,

with an estimated prevalence up to 6.5 per 1000 [5, 6]. The growth of this population is expected to plateau around the year 2050 and carries implications for the provision of health care given the complex needs and increased risk for complications, including arrhythmia and sudden death [7].

Arrhythmia is increasingly burdensome to patients, providers, and health systems with implications for planning and delivering services to a growing population. Adults with CHD presenting to emergency departments of children's hospitals have increased, with twice as many having complex diagnoses, and associated with greater risk of admission and death [8]. Figure 1 illustrates factors that contribute to arrhythmic sequelae in CHD including anatomic substrate, malformed or displaced conduction systems, surgical scars, abnormal hemodynamics, fibrosis, and hypoxia [9–11, 12•]. ACHD populations suffer various forms and recurrence

of arrhythmia, are victim to sudden death, and require special consideration for medical and interventional therapy [13].

## Bradyarrhythmia

Bradyarrhythmia, whether related to sinus node dysfunction or atrioventricular (AV) block, is common in certain subsets of CHD [9, 12•, 13]. The interaction of congenital anatomic variations, progressive fibrosis, and physical damage during surgery plays a role in the development of bradycardia. Sinus node dysfunction or conduction abnormalities are seen in patients with heterotaxy syndromes, abnormal segmental connections, and post-atrial or ventricular surgery [9, 12•, 13]. Recurrent AV block is recognized as a potential late complication of intervention, particularly following certain ventricular septal defect (VSD) repairs and left ventricular outflow tract operations. Chronotropic incompetence or loss of AV synchrony can cause significant hemodynamic disturbances in patients with atrial baffles or Fontan circulations, leading to symptoms or exercise intolerance, and is poorly tolerated.

### Sinus node dysfunction

Congenital heart defects and surgical intervention can contribute to sinus node dysfunction and associated bradycardia [9, 12•]. In heterotaxy syndrome, the sinus node is commonly atypical in morphology, number, or location. Left atrial isomerism with polysplenia heterotaxy is associated with an absent or hypoplastic sinoatrial node that is displaced posterior-inferiorly. In this setting, ectopic atrial bradycardia and junctional rhythms are commonly encountered. Surgical palliation itself can result in direct trauma to the sinoatrial node or artery as in the case of Mustard, Senning, and Fontan procedures [14, 15]. Chronotropic incompetence can result in significant symptoms, negatively impact hemodynamics, and reduce quality of life.

### AV block

Displacement of the conduction system is associated with several congenital heart defects such as heterotaxy syndrome, congenitally corrected-transposition of the great arteries (CC-TGA), and endocardial cushion defects [12•]. Absence of a penetrating connection and displacement from the usual course surrounding the triangle of Koch lead to abnormal functional properties of the conduction system. Patients with CC-TGA are at increased risk of congenital and post-surgical heart block with 20% of patients estimated to develop AV block by adulthood [16].

Direct trauma to the conduction system during surgical or catheter-based intervention is a common cause of AV block in CHD patients, occurring in 1–3% of cases [17, 18]. Ventricular septal defect repair, whether isolated or in conjunction with other congenital heart defects, is frequently associated with post-surgical AV block [17]. Post-surgical complete heart block is reported to occur in 4–8% of CHD repairs involving VSDs [19]. The subsequent need for chronic right ventricular pacing can lead to inter-ventricular dyssynchrony,

remodeling, and systolic dysfunction [20].

## Tachyarrhythmia

Macro-reentrant atrial tachyarrhythmia is the most common arrhythmia encountered in adults with CHD [21, 22]. Atrial arrhythmia in CHD patients is three times more prevalent than in the general population, with a cumulative lifetime risk near 50%, higher with greater age and CHD complexity, and associated with adverse outcomes including heart failure, death, and need for intervention [23]. Intra-atrial reentrant tachycardia (IART) is the most common atrial arrhythmia, with the prevalence of atrial fibrillation (AF) surpassing IART at age 50, and increasingly permanent at older ages [24]. The risk of developing AF in setting of CHD is 22 times higher than the general population, greatest with the most complex diagnoses, and increased 3.6-fold with history of surgery [25]. Cavotricuspid isthmus (CTI)-related IART is most frequently encountered, although non-CTI IART is often seen concomitantly or alone and related to CHD complexity [26]. Adult CHD patients with atrial arrhythmia have a 2-fold increased risk of death and a 4-fold increased risk of heart failure [27]. Atrial arrhythmia with 1:1 AV conduction may contribute to and is poorly tolerated in setting of ventricular dysfunction.

The incidence of ventricular arrhythmia in ACHD patients is estimated at 0.1–0.2% per year [13]. The primary mechanism is reentry involving ventriculotomy incision or scar tissue [28]. Ventricular arrhythmias are thought to be responsible for most sudden cardiac deaths (SCD) in ACHD, for which the risk is estimated as high as 25–100 times that for the general population and increased after two decades [29]. SCD is cited as the most frequent cause of mortality in ACHD [30–32]. Recent studies estimate that 23% of deaths in ACHD patients are due to sudden cardiac death [30, 31]. Guideline recommendations exist for management of ventricular arrhythmia [13, 33, 34, 35]. It is recognized that the clinical prognostic utility of ventricular ectopy and non-sustained or sustained ventricular tachycardia (VT) varies depending on the underlying CHD, with increased risk in patients with Eisenmenger syndrome, CC-TGA, D-TGA post-atrial switch, and Fontan circulation [36].

Several studies suggest an association of atrial arrhythmias and QRS prolongation with increased risk of SCD [36–38]. Additional predictive factors include ventricular dysfunction [37], AV valve replacement, and post-bypass Fontan pressure > 20 mmHg [38]. In patients with pulmonary arterial hypertension in the setting of CHD, a prior history of arrhythmia, predominately supraventricular arrhythmia, was associated with death [39].

## Diagnostic evaluation

Non-invasive tests remain critical to the evaluation of arrhythmia in ACHD patients. The 2018 ACHD guidelines provide direction in the frequency recommended for such evaluation [35]. The 12-lead ECG remains an important diagnostic tool that can also direct subsequent therapy. Routine ambulatory monitoring is recommended in asymptomatic patients that are increased risk of conduction disease and arrhythmia. Disease-specific recommendations exist for routine surveillance in patients with Tetralogy of Fallot (ToF), CC-TGA, D-TGA,

and Fontan physiology [35•]. Event monitors may provide additional information in symptomatic patients.

Continued monitoring with implantable loop recorders (ILR) are particularly useful in patients with syncope, recurrent palpitations, or episodes that have been unable to be captured on traditional monitors. Recent data demonstrate that ILRs provide useful clinical information in over half of the patients evaluated for syncope with a median time to diagnosis of 4.5 months [40].

Diagnostic electrophysiology (EP) study is useful in risk assessment in CHD such as repaired ToF thought to be at risk for VT and SCD and should be considered in several scenarios in the ACHD population including unexplained syncope and palpitations suggestive of tachyarrhythmia [13]. In CHD patients with ventricular pre-excitation, studies are recommended for risk stratification even if asymptomatic. Patients who may be undergoing cardiac surgery, especially if catheter access will be limited post-operatively, should be considered for EP study pre-operatively.

## Therapeutic options

A series of guidelines for the evaluation and treatment of ventricular arrhythmias and prevention of sudden cardiac death have been published, most recently by a collaborative effort of the American Heart Association, American College of Cardiology, and Heart Rhythm Society [33] and by the European Society of Cardiology [34]. Similar guidelines have been published for the management of supraventricular tachycardia [41]. While comprehensive in terms of the broad spectrum of arrhythmias and available treatments, these guidelines include little information specific to CHD in adult patients. A collaborative statement by Pediatric and Congenital Electrophysiology Society and HRS in 2014 directly addressed arrhythmia management in ACHD patients [13].

### Medical therapy

Choice of antiarrhythmic therapy in adults with CHD should be made in consideration of co-existing conduction disease, ventricular dysfunction, and non-cardiac comorbidities [13]. Class IC antiarrhythmic drugs (flecainide and propafenone) may facilitate VT by decreasing conduction and increasing spatially heterogeneous action potential duration in setting of incisional scarring and fibrosis. Due to their proarrhythmic effects, class IC antiarrhythmics are contraindicated in patients with conduction abnormalities, structural heart disease, and ventricular dysfunction [42, 43]. In a recent retrospective study, flecainide was found to be equally effective in pediatric patients with and without CHD with no difference in the rate of adverse events [44]. There is sparse data regarding the safety and efficacy of flecainide in ACHD patients.

QT prolongation and ventricular arrhythmias including torsades de pointes are important side effects of class III antiarrhythmic drugs. Amiodarone has been associated with other significant adverse effects, including thyroid dysfunction that can in turn exacerbate arrhythmia [45]. While amiodarone is highly effective, toxicity and intolerance lead to a high rate of discontinuation. Low-dose amiodarone ( $\leq 200$  mg/day) was found to be effective for control of

tachyarrhythmia with a low incidence of adverse effects in a select group of CHD patients [46]. Similar to amiodarone, sotalol has been shown to be effective in controlling arrhythmia in adults with CHD, but nearly 20% of patients discontinue sotalol secondary to adverse effects. Patients with Fontan physiology are particularly affected by significant bradycardia, similar to that seen with amiodarone [45, 47]. For acute pharmacologic conversion of IART, ibutilide has been shown to successfully convert > 70% of cases [48]. Dofetilide also demonstrates adequate rhythm control, but is associated with significant adverse events involving ventricular arrhythmia [49].

With consideration of potential adverse effects, overall data would suggest use of amiodarone or dofetilide in setting of complex CHD or systemic ventricular dysfunction [13, 50]. Dofetilide requires close monitoring of concomitant medications, QT interval, and renal function as it is contraindicated in patients with renal failure. Amiodarone requires close monitoring of thyroid, hepatic, and pulmonary function and should be used with caution in patients with cyanotic heart disease, low body mass index, or QT prolongation [13]. The addition of AV nodal blocking medications such as beta blockers or non-dihydropyridine calcium channel blockers (verapamil, diltiazem) may help to prevent rapid ventricular rate, associated symptoms, and cardiac impairment.

## Catheter ablation

Adverse effects of antiarrhythmic medications, their limitations, and patient choice for a drug-free lifestyle contribute to increased utility of catheter ablation. Catheter ablation is a common treatment for arrhythmias with an average acute success rate of 75% [51, 52]. While acutely effective, several studies demonstrate a higher recurrence rate in adults with CHD [53, 54] ranging from 12 h to 4–5 year [55]. Body mass index  $\geq 30$  and Fontan circulation have been associated with arrhythmia recurrence post-ablation [55].

While IART is the most common arrhythmia encountered overall in adults with CHD, AF has emerged as the most common arrhythmia in those aged greater than 50 years [24] with an incidence of 4–15%, which is considerably higher than in the general population. Incisional scars that may predispose to re-entrant circuits, atrial dilation and subsequent fibrosis, and activation of different atrial sites in the setting of sinus node dysfunction are proposed mechanisms that may facilitate initiation and perpetuation of AF. Catheter ablation of AF is acutely successful in 72% of cases [56] with at least partial long-term freedom from AF in follow-up regardless of CHD complexity [57]. Ablation of IART demonstrates similar acute success rate of 75–78% [58, 59]. A recent study suggests that acute success is a predictor of freedom from recurrence [60], which has been associated with previous AF, non-CTI IART, and complex CHD [58, 59].

Congenital displacement and anomalous course of the conduction system in certain CHD lesions can disrupt the traditional location of the AV node relative to the triangle of Koch, which is important in the catheter ablation of AV nodal reentry tachycardia (AVNRT). Recent studies report a success rate of 86–100% of AVNRT cases [61, 62]. Complex CHD was associated with longer procedure time and increased risk of AV block [61]. It is reasonable to defer if AV nodal location is uncertain.

Ventricular arrhythmia is not uncommon in adults with CHD and the prevalence is higher in patients with VSD patch closure or ventricular incision [63]. The primary mechanism is reentry involving a critical isthmus [64, 65]. In ventricular arrhythmia, catheter ablation is successful in greater than 80% of cases, with a recurrence rate of 14% [66]. Confirmation of conduction block is predictive of preventing recurrence [66]. High-density mapping, contact force, irrigated tip catheters, and intracardiac echocardiography are additional strategies that may help to improve ablation outcomes.

## Device therapy

There is an increasing need for implantable cardiac devices in ACHD patients due to the high prevalence of congenital and post-operative sinus node dysfunction and AV block. Chronic isolated sinus node dysfunction or loss of AV synchrony contributes to impaired hemodynamics. Symptomatic sinus node dysfunction and symptomatic bradycardia with concomitant AV block are class I indications for pacing in ACHD patients [13]. As sinus bradycardia increases the interval during which a premature atrial complex may initiate IART, consideration can be made for pacemaker implantation to prevent recurrent tachyarrhythmia [67]. Devices with antitachycardia pacing have been found to be effective in terminating IART in ACHD patients [68, 69].

Lead position is an important aspect of pre-procedural planning. Traditional atrial appendage placement may not be feasible in certain post-repair cases. In addition, compared to appendage pacing, atrial septal position may decrease the frequency of unnecessary ventricular pacing [70]. While there is no evident single optimal endocardial ventricular pacing site, several studies have demonstrated that the right ventricular free wall should be avoided [71, 72].

Sudden cardiac death accounts for 20–25% of deaths in CHD patients post-surgery [30, 73], with the majority related to arrhythmia [73]. Patients at higher risk include those with CC-TGA, D-TGA post-Mustard or Senning, and ToF [29, 73]. There are additional elements in ToF that have been associated with increased risk including increased QRS duration, non-sustained ventricular tachycardia, and inducible sustained ventricular tachycardia [21, 74], particularly in those with history of ventriculotomy. In this population, there is an 8–10% annual rate of appropriate ICD shocks [74].

Whether in pacing or ICD systems, transvenous leads are to be avoided in the setting of intracardiac shunts [75]. The subcutaneous ICD (S-ICD) has emerged as an alternative to the transvenous ICD system. The electrode is placed in the left parasternal position and the pulse generator in the left mid-axillary line. The S-ICD implantation method offers the advantage of minimizing procedure-related complications, preserving venous structures, and providing an alternative route when transvenous access to the ventricular is precluded by abnormal systemic venous pathways as in the setting of complex CHD. Eligibility ECG screening is performed to avoid oversensing of the T-wave. Abnormal ECG findings associated with ineligibility include T-wave inversion, prolonged QRS duration, and insufficient R:T wave ratio [76, 77].

Approximately 75% of ACHD patients meet eligibility criteria with at least one suitable vector [78, 79], with potential increase in eligibility rate with right parasternal lead position [80]. As the S-ICD system does not allow for conventional pacing or ATP, it can be considered reasonable in ACHD patients that do not have bradycardia or require ATP functionality.

Adults with CHD are at increased risk for complications related to implantable cardiac devices including infection, device malfunction, lead malfunction, and venous occlusion [81–83] that may lead to a need for lead abandonment or extraction. Up to 30% of ACHD patients will have at least one or more abandoned transvenous lead [84]. Transvenous lead extraction has inherent risks and complications in patients with structurally normal hearts [85]. In the ACHD population, 40% of lead extractions are successfully performed with simple extraction [84] and greater than 90% are completely extracted with advanced extraction techniques including those with lumenless pacing leads [86, 87]. While transvenous lead extraction has been performed with similar complication rates as that in adult patients without CHD [84, 88], careful consideration of the risks and benefits must be weighed regarding whether to abandon or remove the lead. While intended to be life-saving, implanted cardiac rhythm devices can be associated with depression, increased anxiety, and decreased quality of life [89, 90], highlighting the potential utility of appropriate screening prior to implantation.

## Surgical therapy

Many ACHD patients require reoperation for deterioration of implanted prostheses or residual hemodynamic sequelae, creating an opportunity to surgically address substrates for arrhythmia [13, 91–93]. Surgical techniques for arrhythmia emerged prior to the development of transcatheter options, were pioneered for supraventricular arrhythmias, and were initially utilized in patients without CHD [94, 95]. Early approaches were developed for treatment of accessory pathways, atrial flutter, and then atrial fibrillation using cut-and-sew, energy ablation, or combined techniques (e.g., Cox-Maze procedure). Operative anti-arrhythmic strategies have been adapted for CHD and refined for use across the spectrum of anatomic and electrical heterogeneity [91, 92]. Recommendations have been developed for operative management of arrhythmia in ACHD, yet the scope is not comprehensive and approaches to treatment and prophylaxis continue to evolve [13, 35, 96].

Atrial arrhythmia is most prevalent in certain right heart anomalies, univentricular hearts, and following atrial surgery [13, 96]. Substrates for AF frequently reside in the left atrium, and the aim of left atrial surgery is isolation of the pulmonary veins and disruption of critical isthmuses for reentry (left AV valve, coronary sinus, and Bachmann's bundle), often including left atrial appendage ligation [97]. Right atrial macroreentrant tachycardias (IART, AF) can occur around conduction barriers that are common from prior operation (e.g., atrial incisions, patches, baffles) and often involve the CTI. Successful operative approaches must create transmural lines of conduction block that completely isolate arrhythmic foci or areas of slow conduction; otherwise, any gap may

serve as a nidus for recurrent arrhythmia [91, 93, 96].

Arrhythmia surgery in ACHD is safe and effective with favorable results [98–102]. Cox-Maze type procedures (right, left, and biatrial) were predominantly utilized with modifications to account for specific anatomic or prior surgical factors, and complications or mortality related to the arrhythmia surgery were very low or absent. High success rates and freedom from recurrence of up to 75% at 6 years are reported, although age, arrhythmia type, and duration are related to recurrence (e.g., persistent AF) and may represent targets for earlier or modified intervention [98–102]. Specifically considering Fontan conversion surgery, durable long-term freedom from late recurrence of atrial tachycardia is 77% at 10 years with no recurrence of atrial fibrillation after biatrial arrhythmia surgery [103].

There is considerably higher risk of ventricular arrhythmia and SCD in patients with ToF, systemic right ventricle, and left heart obstruction [13]. Intraoperative treatment of VT most often accompanies surgery for hemodynamic lesions, with best practices still evolving. Operative techniques range from cryoablation to endocardial/epicardial resection, with success rates from 50 to 85% and frequently combined with defibrillator implantation given the risks and consequences of treatment failure [13]. Although risk scoring has not been prospectively validated, recent data has emerged regarding the use of perioperative EP studies in patients with ToF at risk for VT after pulmonary valve replacement (PVR) [104, 105]. In an unselected adult population undergoing PVR, 49% had inducibility before surgery, 47% remained inducible afterward and underwent defibrillator implantation, and 21% of those received appropriate ICD shocks for symptomatic VT [104]. These results highlight the high rate of inducibility in patients with ToF, how surgical ablative therapy may reduce the rate of potentially malignant arrhythmia, and underscore the importance of patient selection in surgical ablation.

## Conclusion

Adult survivors of CHD are vulnerable to hemodynamic compromise and decreased quality of life secondary to paroxysmal and at times life-threatening arrhythmia. Routine monitoring and advanced therapeutic options may serve to decrease morbidity and mortality in these patients.

## Compliance with Ethical Standards

### Conflict of Interest

The authors declare that they have no conflict of interest.

### Human and Animal Rights and Informed Consent

This article does not contain any studies with human or animal subjects performed by any of the authors.

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