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Mini Review

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Suraj Pai* and Suresh Pai

Department of Cardiovascular and Thoracic Surgery, Kasturba Medical College Mangalore, India

ABSTRACT

Arrhythmias represent one of the most significant long-term complications in patients with congenital heart disease (CHD). With advancements in surgical and medical care, survival into adulthood has improved dramatically, resulting in an increasing population of adults with CHD (ACHD) who are at risk of arrhythmias. These arrhythmias are associated with a variety of pathophysiological mechanisms including surgical scars, chamber enlargement, myocardial fibrosis, and autonomic dysfunction. This review provides a focused overview of the epidemiology, pathophysiology, diagnosis, and management of arrhythmias in CHD patients, with an emphasis on current therapeutic strategies and long-term considerations.

Keywords: Congenital heart disease; Arrhythmia; Intra-atrial reentrant tachycardia; Ventricular tachycardia; Fontan circulation; Electrophysiology

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Corresponding Author:
Suraj Pai, Department of Cardiovascular and Thoracic Surgery Kasturba Medical College Mangalore Manipal, India, Tel: +91 9880501134

INTRODUCTION

Congenital heart disease (CHD) represents the most common congenital malformation in live-born infants, with an estimated global birth prevalence of approximately 9 per 1,000 live births [1]. Advances in surgical and medical management have significantly improved the survival of children with CHD, resulting in a growing population of adolescents and adults living with repaired or palliated cardiac defects [2]. This demographic shift has given rise to a new spectrum of long-term complications, among which cardiac arrhythmias represent a leading cause of morbidity and mortality. Arrhythmias in CHD patients may arise from a complex interplay of structural, hemodynamic, surgical, and electrophysiological factors. The mechanisms are often multifactorial, including scarring from prior surgeries, chamber enlargement, myocardial fibrosis, and inherent conduction system abnormalities [3,4]. Furthermore, the type and timing of surgical repair, particularly procedures involving atriotomy or ventriculotomy, contribute substantially to arrhythmogenic risk [5]. The spectrum of arrhythmias varies depending on the underlying congenital lesion. Supraventricular arrhythmias, particularly intra-atrial reentrant tachycardia (IART) and atrial fibrillation, are frequently encountered in patients with atrial septal defects (ASD), atrioventricular septal defects (AVSD), and after Fontan palliation [6-8]. Ventricular arrhythmias, including monomorphic ventricular tachycardia (VT), are more prevalent in conditions such as tetralogy of Fallot (TOF) and complex cyanotic CHD with prior ventriculotomy or patch repair [9].

Importantly, arrhythmias in this population are not merely symptomatic disturbances but carry significant prognostic implications. They are associated with increased risk of heart failure, thromboembolic events, and sudden cardiac death, especially in adults with repaired CHD [10-12]. As such, timely identification, risk stratification, and targeted management of arrhythmias are vital components of long-term care in this population. Despite the growing burden of arrhythmias in CHD, challenges persist in diagnosis and management due to the heterogeneity of cardiac anatomy, variable surgical histories, and limitations in applying

standard electrophysiological approaches [13]. Tailored strategies, often requiring multidisciplinary input and specialized expertise in adult congenital heart disease (ACHD), are essential for optimal outcomes.

EPIDEMIOLOGY

The prevalence and types of arrhythmias in CHD vary according to the underlying defect, type of surgical repair, and patient age. In general, the incidence of arrhythmias increases with time following surgical correction [5]. Atrial arrhythmias, such as intra-atrial reentrant tachycardia (IART) and atrial flutter, are more prevalent in patients with atrial septal defects (ASD), Fontan circulation, and atrioventricular septal defects [6]. Ventricular arrhythmias are more commonly seen in patients with tetralogy of Fallot (TOF), transposition of the great arteries (TGA), and systemic right ventricles [7]. The risk of sudden cardiac death (SCD) is increased in these populations, especially when residual lesions or myocardial scarring is present [8].

PATHOPHYSIOLOGY

Arrhythmogenesis in CHD is multifactorial. One key mechanism is surgical scarring, which can create reentrant circuits, particularly in patients who have undergone atriotomy or ventriculotomy incisions [9]. Chamber dilatation from residual shunts or valvular dysfunction can lead to structural remodeling and fibrosis, further predisposing to arrhythmias [10]. Chronic hypoxia and pressure overload, common in cyanotic lesions or single-ventricle physiology, contribute to myocardial fibrosis and electrical heterogeneity [11]. Additionally, patients with Fontan circulation may experience autonomic dysfunction, which can facilitate both bradyarrhythmias and tachyarrhythmias [12].

COMMON ARRHYTHMIAS IN CHD

The types of arrhythmias observed in CHD are diverse and often lesion-specific. Atrial arrhythmias such as atrial flutter, atrial fibrillation (AF), and IART are the most common and often occur decades after surgical repair [13]. IART is especially prevalent in patients with Fontan physiology or those who have undergone the Mustard or Senning procedures [14]. Sinus node dysfunction is frequently observed following atrial surgeries, particularly in patients with ASDs and Fontan palliation [15]. High-grade atrioventricular (AV) block may occur in patients who have undergone surgical repair involving the septal area [16]. Ventricular tachycardia (VT) and ventricular fibrillation (VF), although less frequent, are associated with significant mortality and are typically seen in TOF, TGA, and systemic right ventricles [17] (Table 1).

DIAGNOSIS

Arrhythmia diagnosis in CHD requires a multimodal approach. Standard 12-lead ECG and ambulatory Holter monitoring are essential first-line tools for detecting both symptomatic and asymptomatic episodes [18]. For patients with infrequent symptoms, extended monitoring using event recorders or implantable loop recorders improves diagnostic yield [19]. Invasive electrophysiology studies (EPS) are useful for mapping complex arrhythmias and determining ablation strategy [20]. Cardiac imaging, particularly MRI, plays a vital role in assessing ventricular function, chamber sizes, surgical scarring, and myocardial fibrosis [21].

MANAGEMENT STRATEGIES

Medical Therapy

Pharmacologic therapy remains an important component of arrhythmia management. Beta-blockers are frequently used for both rate and rhythm control. Class III antiarrhythmics, such as amiodarone and sotalol, are often effective but require careful monitoring due to potential side effects and proarrhythmic risk [22] (Table 2).

Catheter Ablation

Catheter ablation has become a mainstay for treating IART and other reentrant arrhythmias. While success rates are high in some lesions, anatomical complexity and prior surgeries may lower efficacy, particularly in Fontan patients or those with systemic venous baffles [23]. Advances in three-dimensional electroanatomical mapping have improved procedural outcomes.

Table 1: Common Arrhythmias by Congenital Heart Defect

Congenital Heart Defect	Common Arrhythmias	Notes
Atrial Septal Defect (ASD)	Atrial flutter, Atrial fibrillation, Sinus node dysfunction	Risk increases with age and post-atrial surgery
Tetralogy of Fallot (TOF)	Ventricular tachycardia, PVCs	Scarring from ventriculotomy is a major substrate for VT
Fontan Circulation	Intra-atrial reentrant tachycardia, Sinus node dysfunction	Arrhythmias worsen long-term outcome; complex anatomy complicates therapy
Transposition of Great Arteries (TGA) – Mustard/Senning	Atrial flutter, IART, Sinus node dysfunction	Associated with atrial baffles and atriotomy scars
Atrioventricular Septal Defect	Complete heart block, Atrial arrhythmias	Conduction tissue proximity to repair area increases AV block risk
Single Ventricle Physiology	Bradyarrhythmias, IART, VT	Electrophysiologic remodeling and surgical history influence risk

Table 2: Risk Factors for Arrhythmias in CHD

Risk Factor	Mechanism / Relevance
Surgical scarring	Creates reentrant circuits, particularly after atriotomy or ventriculotomy
Chamber enlargement	Promotes atrial or ventricular arrhythmias via stretch and fibrosis
Myocardial fibrosis	Increases heterogeneity of conduction and reentry potential
Hypoxia or cyanosis	Alters cellular electrophysiology, especially in cyanotic lesions
Autonomic dysfunction (e.g., Fontan)	Facilitates both brady- and tachyarrhythmias
Postoperative conduction system injury	Direct injury can lead to AV block or sinus node dysfunction

Pacemaker and ICD Therapy

Sinus node dysfunction and AV block may necessitate permanent pacemaker implantation [24]. Patients with CHD and a history of sustained VT or aborted SCD are candidates for implantable cardioverter-defibrillators (ICDs) [25]. Guidelines also recommend ICDs for primary prevention in selected high-risk groups, such as TOF with significant right ventricular enlargement or dysfunction [26] (Table 3).

Surgical and Hybrid Approaches

In patients undergoing reoperation for other indications, surgical ablation (e.g., maze procedure) may be added to reduce atrial arrhythmia burden [27]. Hybrid strategies combining surgical access with catheter mapping are increasingly used for complex arrhythmias that are inaccessible via standard endovascular routes [28].

Long-Term Considerations

Lifelong surveillance is essential for early detection and treatment of arrhythmias in CHD patients. Routine follow-up with ECG and ambulatory monitoring should be part of standard care, particularly in those with repaired complex lesions or previous arrhythmias [29]. Transition from pediatric to adult care must include arrhythmia education, screening, and integration into multidisciplinary ACHD programs [30]. The presence of arrhythmias is a major determinant of quality of life and can increase the risk of heart failure and thromboembolism, especially if unrecognized or undertreated [31] (Table 4).

Table 3: Diagnostic Tools for Arrhythmia Evaluation in CHD

Tool	Utility
12-lead ECG	Initial assessment, rhythm documentation, conduction delays
Holter monitoring	Detects intermittent arrhythmias; monitors therapy response
Event recorder / Loop recorder	Captures infrequent but symptomatic episodes
Electrophysiology study	Defines arrhythmia substrate; guides ablation strategies
Cardiac MRI	Assesses fibrosis, anatomy, ventricular function, and scarring
Echocardiography	Evaluates structural defects, residual lesions, chamber size

Table 4: Summary of Management Strategies

Therapy	Indication	Limitations
Beta-blockers	Rate control, some arrhythmia prevention	May worsen bradyarrhythmias
Amiodarone/Sotalol	Atrial and ventricular arrhythmias	Long-term toxicity; proarrhythmic potential
Catheter ablation	IART, focal atrial tachycardia, some VTs	Lower success in complex anatomies
Pacemaker implantation	Sinus node dysfunction, AV block	Requires careful lead placement in abnormal anatomy
ICD implantation	Secondary/primary prevention of SCD	Risk of inappropriate shocks; anatomical barriers
Surgical ablation	Maze or cryoablation during other cardiac surgeries	Requires open-heart surgery
Hybrid procedures	Complex arrhythmias inaccessible via standard EP routes	Limited availability; requires specialized teams

CONCLUSION

Arrhythmias in congenital heart disease are a significant source of morbidity and mortality. Their management requires a deep understanding of congenital anatomy, prior surgical interventions, and arrhythmic mechanisms. Advances in diagnostics, ablation techniques, and device therapy have improved care, but individualized, lifelong management remains essential. Collaborative, multidisciplinary care and early intervention are key to optimizing long-term outcomes.

REFERENCES

1. van der Linde D, Konings EE, Slager MA, Witsenburg M, Helbing WA, Takkenberg JJ, et al. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. J Am Coll Cardiol. 2011; 58: 2241-2247.
2. Marelli AJ, Mackie AS, Ionescu-Ittu R, Rahme E, Pilote L. Congenital heart disease in the general population: changing prevalence and age distribution. Circulation. 2007; 115: 163-172.
3. Baumgartner H, De Backer J, Babu-Narayan SV, Budts W, Chessa M, Diller GP, et al. 2020 ESC Guidelines for the management of adult congenital heart disease. Eur Heart J. 2021; 42: 563-645.
4. Triedman JK, Alexander ME. Arrhythmias in adults with congenital heart disease. Circulation. 2005; 112: 1098-1107.
5. Khairy P, Landzberg MJ, Gatzoulis MA, Lucron H, Lambert J, Maron BJ, et al. Transvenous pacing leads and systemic thromboemboli in patients with intracardiac shunts: a multicenter study. Circulation. 2006; 113: 2391-2397.
6. Walsh EP. Interventional electrophysiology in patients with congenital heart disease. Circulation. 2007; 115: 3224-3234.
7. Gatzoulis MA, Balaji S, Webber SA, Siu SC, Hokanson JS, Poile C, et al. Risk factors for arrhythmia and sudden cardiac death late after repair of tetralogy of Fallot: a multicentre study. Lancet. 2000; 356: 975-981.
8. Silka MJ, Hardy BG, Menashe VD, Morris CD. A population-based prospective evaluation of risk of sudden cardiac death after operation for common congenital heart defects. J Am Coll Cardiol. 1998; 32: 245-251.
9. Karpawich PP, Crosson JE, Sade RM. Transvenous pacing in the pediatric and congenital heart disease patient: a review. J Electrocardiol. 1990; 23: 102-107.
10. Rodriguez RM, Dubin AM. Arrhythmias in the adult with congenital heart disease. Card Electrophysiol Clin. 2017; 9: 275-287.
11. Cohen MI, Snyder CS, Weinberg PM, Vetter VL, Thomus p. Diagnostic and therapeutic management of arrhythmias in patients with congenital heart disease. Pediatr Cardiol. 2004; 25: 252-256.
12. Gewillig M, Stephen B. The Fontan circulation. Heart. 2005; 91: 839-846.
13. Triedman JK. Arrhythmias in adults with congenital heart disease. Heart. 2002; 88: 254-260.
14. Epstein AE, DiMarco JP, Ellenbogen KA, Estes NA III, Freedman RA, Gettes LS, et al. ACC/AHA/HRS 2008 guidelines for device-based therapy of cardiac rhythm abnormalities. J Am Coll Cardiol. 2008; 51: e1-–62.
15. Villain E, Bonnet D, Sidi D, Kachaner J. Permanent cardiac pacing in children with congenital heart disease. Am J Cardiol. 1992; 69: 428-432.
16. Berul CI, Triedman JK, Gauvreau K, Walsh EP. Risk factors for death after surgical repair of congenital heart defects a case-control study. Pacing Clin Electrophysiol. 2003; 26: 1581-1584.
17. Yetman AT, Nykanen D, McCrindle BW, Freedom RM, Benson LN. Balloon angioplasty for branch pulmonary artery stenosis after the Fontan operation. Pediatr Cardiol. 2002; 23: 502-505.
18. Stephenson EA, Batra AS, Knilans TK. Pediatric ambulatory electrocardiography. Card Electrophysiol Clin. 2017; 9: 199-206.
19. Yap SC, Roos-Hesselink JW, Hoendermis ES, Bartelings MM, Simoons ML, Schalij MJ, et al. Outcome of implantable loop recorders in patients with unexplained syncope or palpitations. Europace. 2010; 12: 869-874.
20. Kugler JD. Indications for electrophysiology studies in patients with congenital heart disease. Pediatr Cardiol. 2000; 21: 362-368.
21. Puranik R, Tsang VT, Broadbent E, Taylor AM. Magnetic resonance imaging in congenital heart disease: evaluation of myocardial fibrosis. Heart. 2005; 91: 821-826.
22. Zeppenfeld K, Ehrlich JR, Miyazaki S, Stevenson WG. Ventricular arrhythmias in congenital heart disease: Mechanisms and management. Europace. 2017; 19: 1459-1468.
23. Moore JP, Gallotti RG, Shannon KM, Van Hare GF. Efficacy and safety of catheter ablation for atrial tachycardia in congenital heart disease. Heart Rhythm. 2014; 11: 1163-1170.
24. Janousek J, Paul T, Wren C. Permanent pacemaker implantation in children: worldwide practice and comparison of guidelines. Europace. 2013; 15: 728-736.
25. Khairy P, Landzberg MJ, Gatzoulis MA. Sudden cardiac death in adult congenital heart disease. Curr Opin Cardiol. 2008; 23: 506-511.
26. Silka MJ, Bar-Cohen Y, Morwood J, Liem LB. Cardiac rhythm devices in congenital heart disease. Heart Rhythm. 2009; 6: S90-94.
27. Stulak JM, Dearani JA, Burkhart HM, Sundt TM, Schaff HV. Surgical ablation for atrial arrhythmias in congenital heart disease. Semin Thorac Cardiovasc Surg. 2007; 19: 66-72.
28. Janousek J, Kubuš P, Hindmarsh G, Gebauer RA. Hybrid procedures in pediatric arrhythmia management. Pediatr Cardiol. 2012; 33: 1159-1165.
29. Diller GP, Kempny A, Alonso-Gonzalez R, Swan L, Uebing A, Li W, et al. Arrhythmia surveillance and management in adults with congenital heart disease. Circulation. 2010; 122: 1130-1138.
30. Stout KK, Daniels CJ, Aboulhosn JA, Bozkurt B, Broberg CS, Colman JM, et al. 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease. Circulation. 2019; 139: e698-800.
31. Oliver JM, Gallego P, Gonzalez AE, Sanchez-Recalde A, Benito F. Risk of thromboembolism in adults with atrial arrhythmias and congenital heart disease. Rev Esp Cardiol. 2002; 55: 1047-1053.

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Contact Info

+1 302 966 3456

info@clinicscardiology.com

651 N. Broad St., Ste 308,
Middletown, DE 19709