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Review

Navigating Arrhythmias in Tetralogy of Fallot Throughout the Lifespan: A Case-based Review

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ABSTRACT

Arrhythmias are a common complication associated with tetralogy of Fallot (ToF), one of the most prevalent forms of congenital heart disease. As illustrated by this case-based review, various forms of arrhythmias can be encountered across the lifespan of patients with ToF, from infancy to older adulthood. These include atrioventricular block, junctional ectopic tachycardia, and atrial and ventricular arrhythmias. Arrhythmias have important implications on the health and quality of life of patients with ToF and require treatment by caregivers with dedicated expertise. The choice of pharmacologic and/or interventional therapies to alleviate symptoms, avoid complications, and mitigate risks depends in part on the type, severity, and frequency of the arrhythmia, as well as on the particularities of individual clinical scenarios. Preventing, monitoring for, and managing arrhythmias are an integral component of the care of patients with ToF throughout their lifespan that is critical to optimizing health outcomes.

RÉSUMÉ

L'arythmie est une complication fréquemment associée à la tétralogie de Fallot (TF), l'une des cardiopathies congénitales les plus courantes. Dans le présent article de synthèse basé sur des études de cas, nous illustrons les différentes formes d'arythmie tout au long de la vie des patients atteints de la TF, de la petite enfance à l'âge adulte avancé. Les formes d'arythmie décrites incluent le bloc atrioventriculaire, la tachycardie jonctionnelle ectopique et les arythmies auriculaire et ventriculaire. L'arythmie a des répercussions importantes sur l'état de santé et sur la qualité de vie des patients atteints de la TF, et elle requiert un traitement par des personnes dotées d'une expertise particulière. Le choix d'un traitement (pharmacologique, interventionnel ou les deux) pour soulager les symptômes, éviter les complications et réduire les risques dépend du type, de la sévérité et de la fréquence de l'arythmie, ainsi que des particularités de chaque tableau clinique. La prévention, la surveillance et la prise en charge de l'arythmie font partie intégrante des soins pour les patients atteints de la TF tout au long de leur vie, et elles sont cruciales pour optimiser les résultats cliniques.

Tetralogy of Fallot (ToF) is the most common type of cyanotic heart disease and accounts for approximately 3%-5% of all congenital heart malformations, with an incidence of 0.28-0.34 per 1000 live births.[1,2] Since the first description of ToF by Nicolas Steno in 1673 and the advent of surgical correction in the 1950s by Lillehei et al.,[3] remarkable progress has been made in the management of this condition, allowing up to 85% of patients with surgically repaired ToF (rToF) to reach adulthood.[4] However, the surgical interventions themselves can have arrhythmic consequences. In particular, junctional ectopic tachycardia (JET) is frequently seen in the early postoperative period, whereas atrioventricular (AV) block is a well-recognized complication after ToF repair. A surgical scar, by virtue of its proarrhythmic potential, brands an individual as being at risk for arrhythmias throughout their lives. In one multicentre study of adults with rToF who had an average age of 37 years, 43% had experienced at least 1 sustained arrhythmia or arrhythmia intervention.[5]

Although the nature and pattern of arrhythmias encountered in patients with ToF differ across the lifespan, arrhythmias affect patients of all ages and require lifelong medical surveillance. The mechanisms of arrhythmias are diverse and may involve a combination of structural, electrical, and haemodynamic factors. The choice of therapy should reflect underlying causes, along with the type, severity, and frequency of arrhythmias, and associated clinical factors. Risk stratification plays a critical role in identifying patients who

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are susceptible to developing severe ventricular arrhythmias in order to prevent sudden cardiac death. The comprehensive approach to managing arrhythmias in patients with rToF generally includes addressing risk factors and predisposing conditions and tailored arrhythmia-directed therapies that may include antiarrhythmic drugs, electrical cardioversion, catheter or surgical ablation, and cardiac implantable electronic devices.[6,7]

In this article, we will review various types of arrhythmias that patients with rToF may experience from birth to adulthood through illustrative case presentations. In so doing, monitoring strategies and treatment options will be discussed, along with relevant literature. The hope is that, collectively, these case presentations will provide an overview of the various arrhythmias encountered throughout the lifespan of patients with ToF, impart diagnostic insights, and offer practical management tips.

Case 1: Postoperative JET

An 11-month-old boy with an antenatal diagnosis of ToF was electively admitted for surgical reintervention. His medical history was remarkable for a right ventricular outflow tract (RVOT) transannular patch, infundibular myomectomy, and left pulmonary angioplasty at 14 days of life. At 3 months of age, percutaneous stenting of the left pulmonary artery was performed for severe left pulmonary artery stenosis.

At admission, he weighed 9.7 kg and his resting oxygen saturation was 94%. Surgery consisted of closing a ventricular septal defect (VSD), enlarging the RVOT, and tricuspid valvuloplasty. A small atrial septal defect was left open. Bypass and aortic cross-clamping times were 89 and 72 minutes, respectively. The postoperative transesophageal echocardiography showed no residual VSD, no RVOT gradient, free pulmonary regurgitation, and moderate biventricular dysfunction (left ventricular ejection fraction [LVEF] evaluated by the Simpson method of 37%; tricuspid annular plane systolic excursion of 4 mm [normal 8 mm]). Postoperatively, he was admitted to the paediatric intensive care unit (ICU) intubated, with 20 ppm of inhaled nitric oxide and with epinephrine inotropic support (maximum 0.06 μg/kg/min). Two hours after his surgery, he developed poorly tolerated rapid JET at 195 bpm. Blood tests were normal (including haemoglobin and electrolytes), and cardiac function on imaging had normalized. He was sedated and treated with dexmedetomidine (2 μg/kg/h). His body temperature was controlled to remain under 37.0°C. He was subsequently treated with intravenous amiodarone. His JET slowed to 160 bpm (Fig. 1A), allowing temporary atrial pacing at 170 bpm. This strategy achieved haemodynamic stability. He remained in JET for 40 hours before reverting to sinus rhythm with normal AV conduction and right bundle branch block (Fig. 1B). Amiodarone was discontinued. At discharge, he remained in sinus rhythm.

Case 1 discussion

Postoperative JET is a well-recognized complication after surgical repair of ToF. It is believed to be an automatic tachycardia originating from the AV junction. The arrhythmia has been reported to occur in 8%-30% of patients after corrective surgery for ToF, which is far higher than the 1% incidence observed in a mixed cohort with congenital heart surgery.[8] Postoperative JET typically occurs within the first hours and days after surgery and has been associated with a longer stay in the ICU and a prolonged need for inotropic support and mechanical ventilation.[9] Several risk factors for postoperative JET in rToF have been identified, including younger age at surgery, higher preoperative heart rate, longer cardiopulmonary bypass time, postoperative inotrope use, and hypomagnesaemia.[8,9] The mechanism underlying postoperative JET remains poorly understood, but it is thought to be related to direct or indirect mechanical trauma of the conduction system precipitating automaticity of the bundle of His.[8]

The treatment for postoperative JET in patients with rToF involves a multifaceted and staged approach that includes careful management of risk factors, nonpharmacologic and pharmacologic therapy, and potential reintervention[10] (Table 1). This approach has evolved throughout the years and has consistently been reported to be effective in most cases. The strategy is primarily geared towards achieving rate control as opposed to rhythm control. The rationale is to lower the heart rate sufficiently to allow atrial pacing to restore AV synchrony. The preferred first-line antiarrhythmic drug for postoperative JET varies across centres, with intravenous amiodarone and procainamide being common choices.[11,12] More recently, oral ivabradine has emerged as a reasonable alternative, with noninferiority to intravenous amiodarone demonstrated in a small open-label randomized trial.[13] Studies have also explored prophylactic antiarrhythmic management to decrease the incidence of postoperative JET, primarily using perioperative dexmedetomidine, magnesium,[14] body temperature control, or esmolol.[15] However, identifying the children most likely to benefit from such pre-emptive approaches remains challenging.[8]

In conclusion, postoperative JET is a common arrhythmia after surgical repair of ToF that can cause significant morbidity and mortality if left untreated. A thorough understanding of the risk factors and underlying mechanisms is critical for effective management. Further research is required to identify high-risk patients for targeted prophylactic therapy.

Case 2: Atrial Arrhythmias

A 13-year-old boy had corrective surgery for ToF at 2 years of age consisting of pulmonary valve–sparing enlargement of the RVOT and VSD closure. He was referred to our centre for severe left ventricular systolic dysfunction in the context of an asymptomatic atrial arrhythmia of unknown duration with predominantly 2:1 conduction (Fig. 2). Transthoracic echocardiography revealed moderate left ventricular dilation with an LVEF of 10%-20%, no residual shunt, and no significant valvar regurgitation. Transesophageal echocardiography showed thrombus in his right atrium. Heart failure therapy was commenced, along with anticoagulation and an initial rate control strategy (metoprolol and digoxin). One month later, he remained in atrial tachycardia with repeat imaging showing no improvement in left ventricular function and stabilization of the right atrial thrombus. Amiodarone was initiated. Electric cardioversions failed to maintain normal sinus rhythm for more than a few consecutive minutes.

A catheter ablation procedure was performed under deep sedation with continuous monitoring, a 3-dimensional

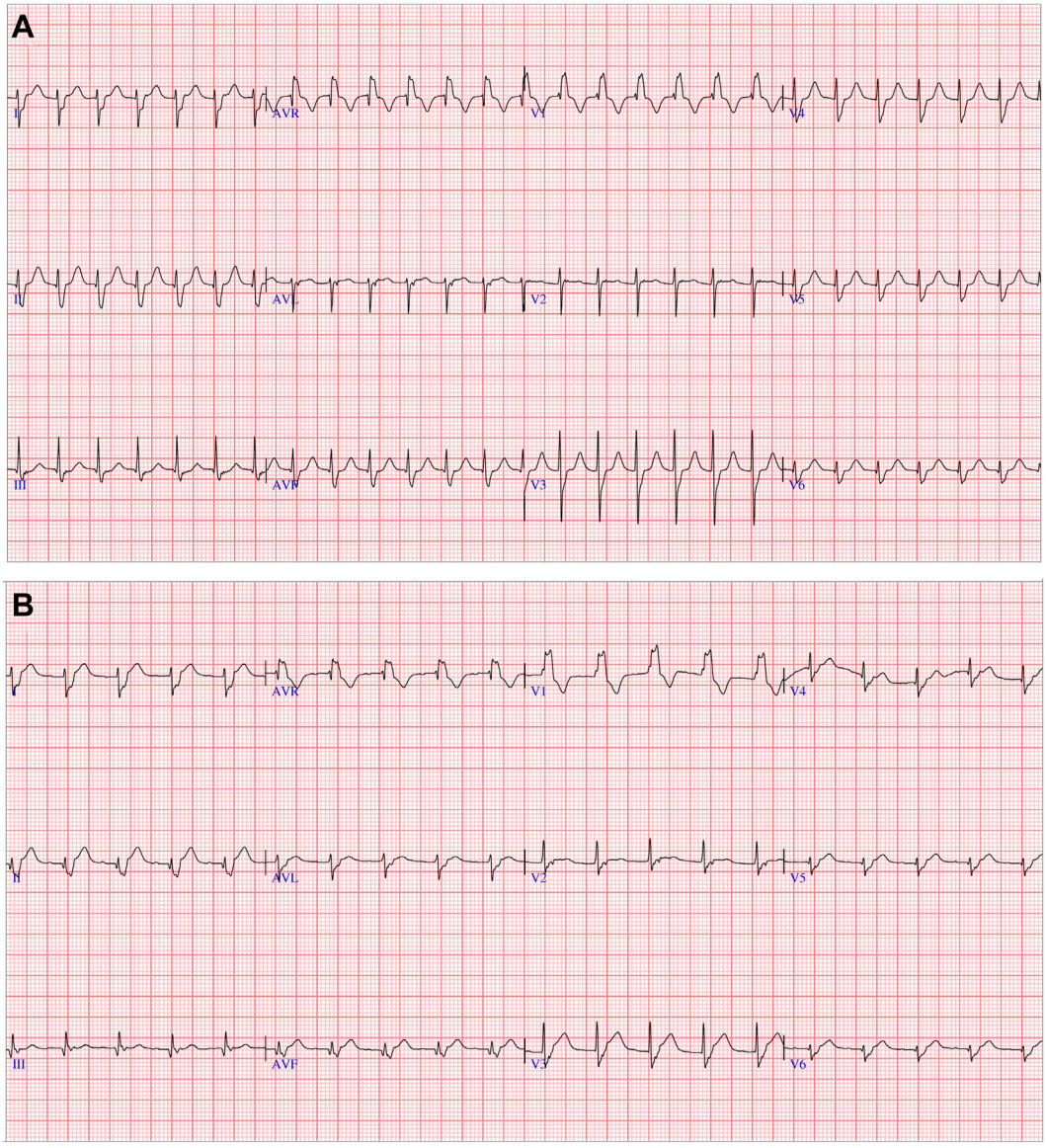

Figure 1. Junctional ectopic tachycardia (JET). (**A**) Postoperative JET at a rate of 160 bpm. (**B**) Restoration of sinus rhythm with intact AV conduction and an underlying right bundle branch block after amiodarone therapy. AV, atrioventricular.

electroanatomic mapping system (Carto3; Biosense Webster, Inc, Johnson & Johnson, Irvine, CA), and robotic magnetic navigation (Niobe platform; Stereotaxis, St. Louis, MO). Four arrhythmias were mapped and ablated during the procedure. An intra-atrial re-entry tachycardia (IART) with a cycle length of 300 milliseconds that propagated around a lateral atriotomy was interrupted by an ablation line connecting the atriotomy to the inferior vena cava. The remaining 3 arrhythmias were nonautomatic focal atrial tachycardias (NAFATs) in various locations on the posterior wall of the right atrium, all of which were successfully ablated by focal lesions. Nevertheless, there were at least 2 other inducible arrhythmias after ablation that were unmappable owing to self-termination, transitioning from one to another, and/or degeneration into atrial fibrillation (AF). It was, therefore, decided to pursue aggressive pharmacologic therapy (including amiodarone, metoprolol, ivabradine, and rivaroxaban) in the hopes of stabilizing his arrhythmogenic substrate with plans to subsequently return for a second ablation procedure.

The patient remained arrhythmia-free for 2 weeks, after which poorly tolerated atrial arrhythmias recurred. Several electrical cardioversions were performed that allowed sinus rhythm to be maintained for days to weeks. By 6 months after ablation, his LVEF had improved to 35%-40%. A repeat electrophysiological study (EPS) revealed a NAFAT located at the anterior portion of the junction between the inferior vena cava and right atrium (Fig. 3). Cavotricuspid-dependent IART was ruled out by entrainment mapping. Sinus rhythm was restored during the first ablation lesion. Consolidation lesions were added and the cavotricuspid isthmus was prophylactically ablated with bidirectional block achieved. The prior ablation line between the atriotomy and inferior vena

Table 1. Staged approach for postoperative JET in patients with ToF

Therapeutic strategy	Management options
1. Control of fluids and electrolytes	• Avoid hypovolemia and • Avoid ionic imbalances • Magnesium sulphate bolus
2. Reduction in adrenergic stimuli	• Sedation (dexmedetomidine as a first-line agent) • Body temperature control and/or therapeutic hypothermia • Weaning of inotropic support if feasible
3. Cardiac pacing to restore AV synchrony	• AAI mode • DDD mode • AVT mode[40]
4. Antiarrhythmic drugs	• Amiodarone • Procainamide • Ivabradine • β-Blockers
5. Extracorporeal membrane oxygenation	

AV, atrioventricular; JET, junctional ectopic tachycardia; ToF, tetralogy of Fallot.

cava was verified and found to be intact. No other arrhythmia was inducible. The patient was discharged in sinus rhythm on amiodarone, β-blockers, and anticoagulation. Amiodarone was discontinued 6 months later, and the patient has remained arrhythmia-free with normalization of his left ventricular function at his follow-up appointment 18 months after ablation.

Case 2 discussion

Atrial arrhythmias are common in patients with rToF and can lead to increased morbidity and mortality. In a multicentre retrospective study, over 20% of young adults with rToF had developed at least 1 atrial arrhythmia.[5] The most common atrial arrhythmia is IART with the most frequent circuits implicating either the cavotricuspid isthmus or lateral right atrial wall. NAFATs typically border scar tissue in the right atrium and are thought to be due to microreentry. Both IART and NAFAT have been associated with a larger right atrial size, multiple surgeries, and tricuspid regurgitation.[5,16] In contrast, factors associated with AF include older age, larger left atrial size, lower LVEF, and hypertension.[5,16] Thus, as the population with rToF ages, the prevalence of AF increases.

The pathophysiology of atrial arrhythmias in rToF is believed to be multifactorial, involving both structural and functional changes in the atrial myocardium. Chronic volume overload due to residual pulmonary regurgitation and/or right ventricular dysfunction can lead to tricuspid regurgitation and atrial dilation, fibrosis, and electrical remodelling. Furthermore, surgical scarring may also contribute to arrhythmia substrates. Treatment strategies for atrial arrhythmias in rToF mirror those in patients without congenital heart disease and include heart rate control, rhythm control, and anticoagulation. However, some unique considerations in the management of patients with rToF and atrial arrhythmias include the following:

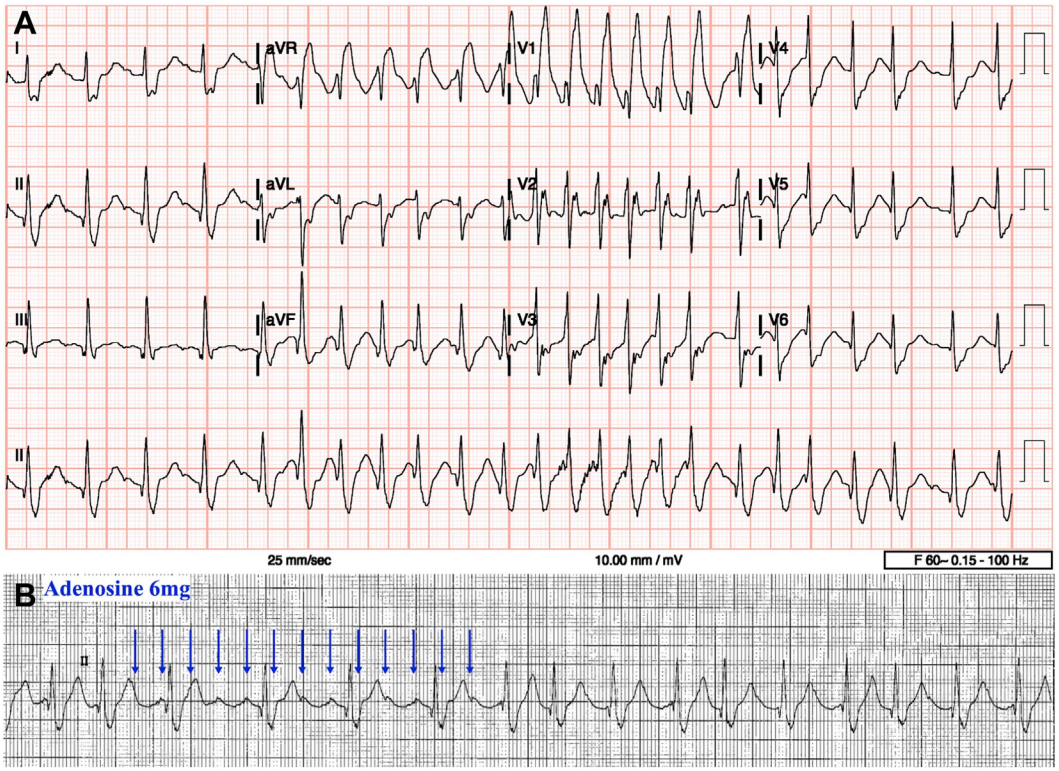

Figure 2. Intra-atrial re-entrant tachycardia (IART). (**A**) An IART with predominant 2:1 conduction. Complete right bundle branch block is noted along with right ventricular hypertrophy. (**B**) After the administration of 6 mg of intravenous adenosine, transient 3:1 conduction allows atrial activity (**blue arrows**) to be better appreciated, confirming a cycle length of approximately 300 milliseconds.

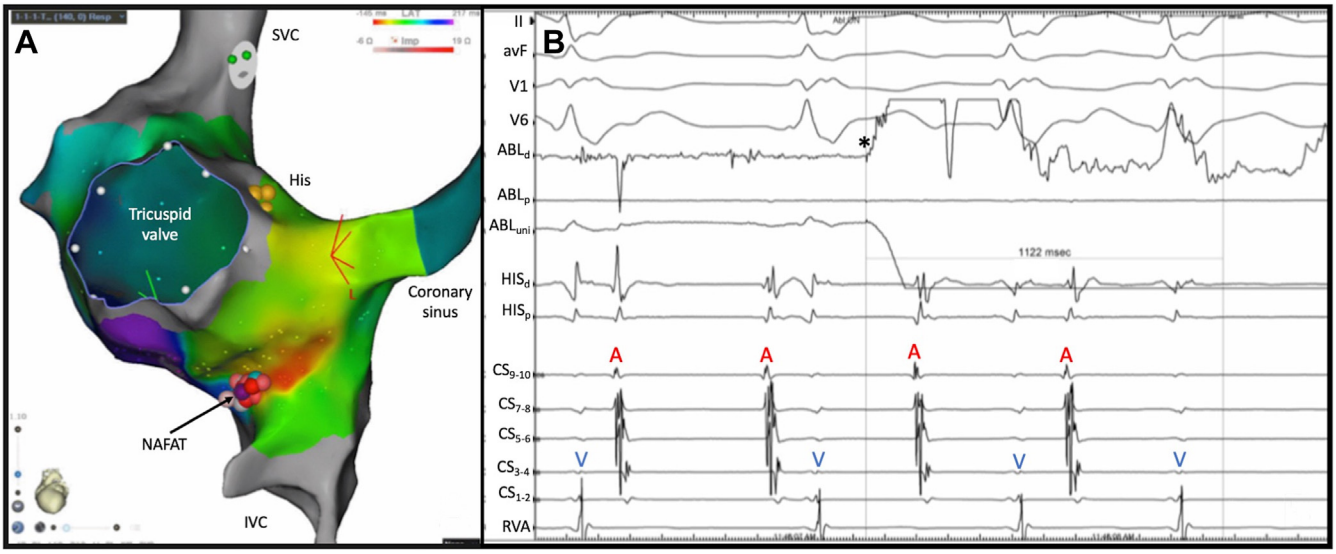

Figure 3. Nonautomatic focal atrial tachycardia (NAFAT). (**A**) A 3-dimensional electroanatomic activation map in a left anterior oblique view with a caudal tilt. Activation times are color-coded, from red (earliest) to purple (latest). Radial spread of activation from a focal source at the anterior margin of the inferior vena cava (IVC) is observed. The **yellow spheres** indicate the position of the His bundle. (**B**) Termination of the NAFAT during the first radiofrequency application (≤1.1 seconds). Shown are surface leads II, avF, V1, and V6, and intracardiac electrograms recorded from distal (ABL_d) and proximal electrode pairs of the ablation catheter (ABL_p), distal (HIS_d) and proximal (HIS_p) His bundle catheter, proximal (CS_{9-10}) to distal (CS_{1-2}) coronary sinus, and right ventricular apex (RVA). The red As and blue Vs indicate atrial and ventricular electrograms, respectively. SVC, superior vena cava.

- *Anticoagulation:* regardless of the $CHADS_2$ or CHA_2DS_2-VASc score, anticoagulation is generally indicated in patients with sustained atrial arrhythmias and rToF (or other moderate or complex forms of congenital heart disease) given the higher risk of thromboembolic events.[17]
- *Rhythm vs rate control:* as this case illustrates, the particularities of each patient must be carefully considered. Nevertheless, as a guiding principle, whenever feasible an initial rhythm-control strategy is generally preferred over rate-control.[6,18] Atrial arrhythmias often have longer cycle lengths in patients with congenital heart disease, which can predispose to rapid AV conduction that can be poorly haemodynamically tolerated and/or lead to tachycardia-induced cardiomyopathy.[18]
- *Catheter ablation:* catheter ablation is an effective treatment option for many patients with rToF and symptomatic or refractory atrial arrhythmias.[6,18] Multiple arrhythmias are frequently present, sometimes with complex or double-loop circuits, such that a systematic approach with detailed mapping is required to maximize success rates.[19] Contemporary high-density multielectrode catheters that have closer interelectrode spacing (be they in multispline, basket, or grid configurations) allow for quicker mapping with a higher spatiotemporal resolution.[20] However, large atrial volumes can result in incomplete maps.[21] Technologies such as robotic magnetic navigation can provide stable catheter tip-to-tissue contact regardless of the size of the chamber, with a favourable safety and efficacy profile.[22] The role of catheter ablation in the setting of AF remains to be defined.

In summary, atrial arrhythmias are common in patients with rToF and can have significant clinical implications including thromboembolic complications, haemodynamic instability, and tachycardia-induced cardiomyopathy. Understanding the complex pathophysiology and individualizing treatment strategies based on patient-specific factors are key to optimizing outcomes. Anticoagulation is a cornerstone of therapy, and arrhythmia interventions should be performed by electrophysiologists with expertise in congenital heart disease.

Case 3: Pacemaker Implantation in rTOF

A 10-month-old girl with an antenatal diagnosis of ToF-type double outlet right ventricle, additional muscular VSD, persistent left superior vena cava, and right aortic arch was electively admitted for complete repair. She had a right Blalock-Taussig shunt at 10 days of age in the context of severe cyanosis. Her genetic testing was unremarkable.

She weighed 6.4 kg at admission and had a resting oxygen saturation of 80%. Surgery consisted of Dacron patch closure of the main VSD, tunnelling left ventricular outflow towards the aorta, direct suture closure of the additional VSD, subpulmonary myomectomy, infundibular patch, valve-sparing pulmonary valvuloplasty, and enlargement of the main and right pulmonary arteries. Bypass and aortic cross-clamping times were 180 and 98 minutes, respectively. Postoperative transesophageal echocardiography showed no residual VSD, no RVOT gradient, no significant pulmonary regurgitation, moderate biventricular dysfunction, and right ventricular hypertrophy. She was admitted to the paediatric ICU intubated and on inotropic support with dobutamine (maximum 10 μg/kg/min) and norepinephrine (maximum 0.1 μg/kg/min). Her postoperative course was remarkable for right ventricular

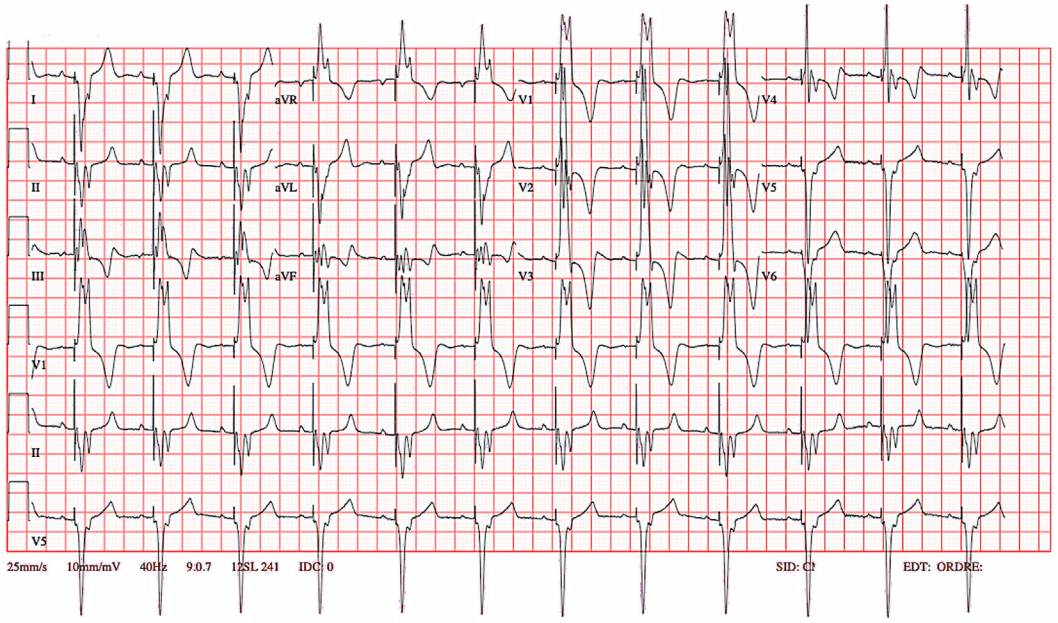

Figure 4. Postoperative AV block. The electrocardiogram shows AV sequential pacing (ie, atrial sensing with ventricular pacing) in a patient with a dual chamber epicardial pacemaker. The lead placed on the left ventricle produces a typical paced right bundle branch block pattern. AV, atrioventricular.

dysfunction responsible for a capillary leak syndrome and chylothorax. Moreover, she had complete AV block without an escape rhythm and required pacing by means of temporary epicardial wires. The complete AV block persisted, prompting implantation of an epicardial dual chamber pacemaker on postoperative day 14 (Fig. 4). The battery-depleted pacemaker generator was subsequently changed 8 years later, and new pacemaker leads were implanted due to increasing pacing thresholds.

Case 3 discussion

Persistent complete AV block is a well-recognized complication after surgery for congenital heart disease with an incidence of approximately 1% for all paediatric congenital heart surgeries combined.[23,24] Risks are higher in rToF, with a reported incidence of postoperative complete AV block ranging from 1.8% to 3.7%.[24,25] ToF and its variants, including ToF-type double outlet right ventricle, account for approximately 7% of postoperative complete AV blocks that require pacemaker implantation in patients with congenital heart disease.[23,24] It has been attributed to postoperative perinodal inflammation and/or direct surgical trauma to the conduction system. The length of hospital stay and hospital-related expenditures are significantly higher when the postoperative course is complicated by transient, and even more so by persistent, complete AV block.[25] Postoperative complete AV block that persists for 7-10 days is considered a class I indication for pacemaker implantation.[26]

Although the true incidence of late-onset complete AV block is unclear in patients with rToF, the association between VSD closure and later AV block is well described.[27,28] The combination of right bundle branch block and left anterior hemiblock after ToF repair was once believed to confer increased risk for late-onset AV block, but subsequent longitudinal studies along with electrophysiological assessments demonstrating normal HV intervals and AV nodal properties have called this association into question.[29] Sinus node dysfunction can also occur in patients with rToF and frequently coexists with atrial tachyarrhythmias, presumably reflecting shared predisposing factors such as right atrial dilation and scarring and/or linked pathophysiological mechanisms (eg, sinus node dysfunction secondary to atrial arrhythmia-induced electrical remodelling). Pacemaker indications should follow standard management guidelines.[30] However, before transvenous lead implantation, residual intracardiac shunts should be carefully ruled out given their association with stroke and systemic embolization in the presence of an intracardiac lead.[31] Other potential indications for pacing in rToF that require larger confirmatory studies before widespread adoption include right ventricular resynchronization for severe right ventricular dysfunction.[32]

In conclusion, complete permanent AV block, be it in the acute postoperative setting or during later follow-up, is a less frequent complication in patients with rToF than atrial arrhythmias despite the high prevalence of right bundle branch block with or without associated left anterior hemiblock. Much remains to be learned about the incidence of late-onset complete AV block and its associated risk factors.

Case 4: Arrhythmia Management Peripulmonary Valve Replacement (PVR)

A 41-year-old woman with ToF had corrective surgery at 2 years of age consisting of pulmonary valvotomy, a non-transannular RVOT patch, and VSD closure. She experienced progressive dyspnoea on exertion with no palpitations or syncope. She achieved 7.9 METS on exercise testing in

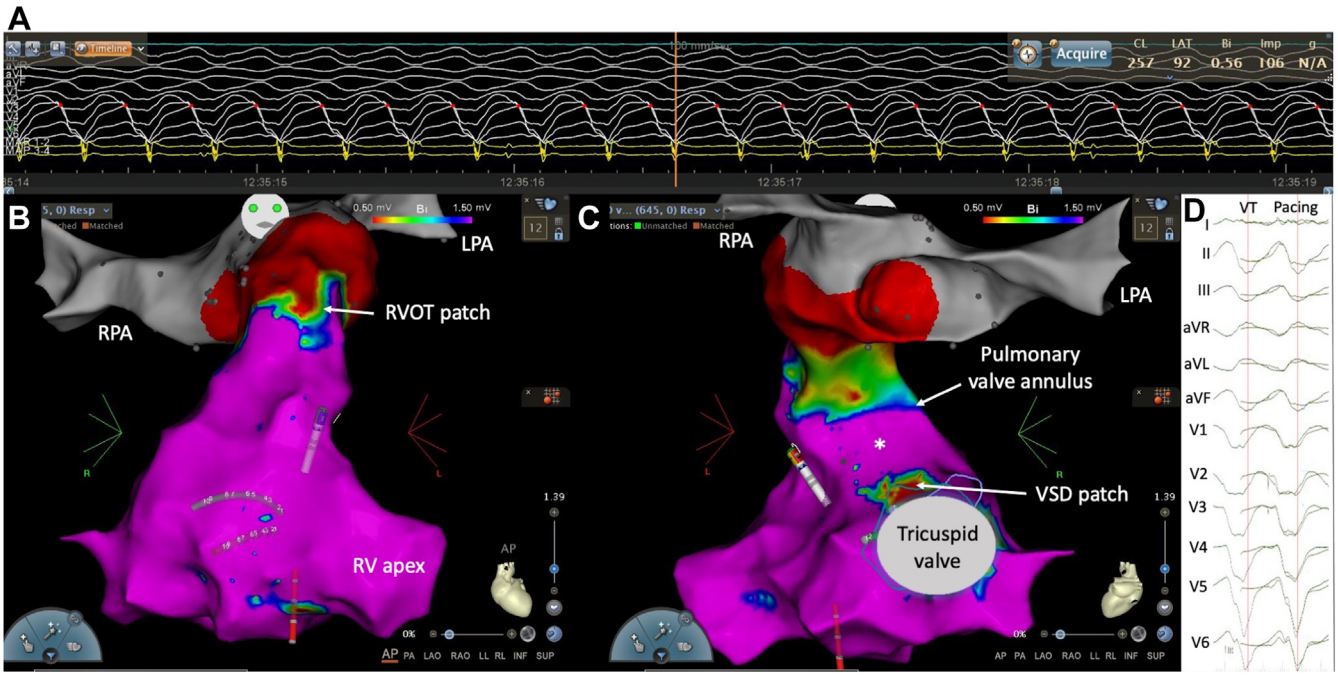

Figure 5. Electrophysiology study before peripulmonary valve replacement. (**A**) Monomorphic ventricular tachycardia (VT) at a cycle length of 257 milliseconds is induced. Activation mapping could not be performed in VT due to haemodynamic instability, prompting antitachycardia overdrive pacing. Three-dimensional electroanatomic maps in sinus rhythm are shown in anteroposterior (**B**) and posteroanterior (**C**) views. Voltages ≥1.5 mV are represented in purple (normal tissue), whereas scar (≤0.5 mV) is color-coded red. Right (RPA) and left (LPA) pulmonary arteries, right ventricular outflow tract (RVOT) patch, right ventricular (RV) apex, pulmonary valve annulus, ventricular septal defect (VSD) patch, and the tricuspid valve are identified. Pace mapping was performed at the potential critical isthmuses for macroreentrant VT. The best pace map (**D**; 92% match) was obtained at the level of the asterisk along the isthmus between the VSD patch and pulmonary valve annulus.

contrast to 13.6 METS 2 years prior. Her electrocardiogram revealed normal sinus rhythm with a right bundle branch block, normal axis, QRS duration of 148 milliseconds, and mild QRS fragmentation (3 contiguous leads). Infrequent ventricular ectopy was noted on Holter monitoring, with no runs of nonsustained ventricular tachycardia. Cardiac magnetic resonance imaging showed preserved biventricular systolic function, severe pulmonary insufficiency (grade 4/4, regurgitant volume 64 mL, and regurgitant fraction 55%), and severe right ventricular dilatation, with an end-diastolic volume of 173 mL/m^2.

On a routine preoperative EPS, sustained haemodynamically unstable monomorphic ventricular tachycardia with a cycle length of 257 milliseconds was induced with 3 extrastimuli delivered from the right ventricular apex at a drive train of 600 milliseconds (Fig. 5A). The tachycardia was poorly tolerated haemodynamically prompting successful overdrive pacing. Potential critical isthmuses for the sustained ventricular tachycardia were identified by 3-dimensional electroanatomic voltage mapping in sinus rhythm (Carto3; Biosense Webster, Inc) to delineate surgical scars (VSD patch and RVOT patch) and the position of tricuspid and pulmonary valves (Fig. 5, B and C). At each potential critical isthmus, pace mapping was performed to assess similarities to the induced ventricular tachycardia. The implicated critical isthmus was between the VSD patch and pulmonary annulus, where pace mapping revealed a 92% match (Fig. 5D). During PVR surgery, concomitant cryoablation of the identified critical isthmus was performed.

Repeat electrophysiological testing before discharge on postoperative day 5 revealed the absence of inducible ventricular tachycardia despite an aggressive stimulation protocol (ie, 2 drive trains [400 and 600 milliseconds], 2 pacing sites [right ventricular apex and outflow tract], up to 3 extrastimuli with a minimum coupling interval of 180 milliseconds, with and without an isoproterenol infusion titrated to increase the heart rate by 30%-50%). The patient was discharged the following day without an implantable cardioverter-defibrillator (ICD). She remains arrhythmia-free at 6 years of follow-up.

Case 4 discussion

Sudden cardiac death of presumed arrhythmic etiology is the leading cause of death in patients with rToF.[5] Surgical scars and anatomic obstacles (ie, tricuspid and pulmonary valves) create barriers to conduction, which combined with structural and electrical right ventricular remodelling provide the substrate for macroreentrant ventricular tachyarrhythmias.[33] Factors associated with sudden death include repair at an older age, presence of a transannular RVOT patch, systolic and diastolic ventricular dysfunction, prolonged QRS duration, nonsustained ventricular tachycardia, and markers of ventricular scarring such as extent of gadolinium enhancement and QRS fractionation.[16,34] Programmed ventricular

stimulation carries important prognostic information for clinical ventricular tachycardia and sudden cardiac death independent of standard clinical, haemodynamic, and electrocardiographic parameters.[35]

A few studies have suggested that addressing the haemodynamic substrate alone, such as with PVR in the setting of severe pulmonary regurgitation, provides insufficient protection against sudden cardiac death. Attention has, therefore, turned towards perioperative electrophysiological management for patients undergoing PVR in order to optimize outcomes.[36-38] In one report, an EPS with transcatheter ablation before PVR rendered 8 of 9 (88.9%) patients with inducible ventricular tachycardia no longer inducible.[38] During a median of 13 months of follow-up, none of the successfully ablated patients experienced a sustained ventricular arrhythmia.[38] In another study, 34 of 70 patients (49%) had inducible ventricular tachycardia before surgical PVR.[36] Concomitant surgical cryoablation was performed. Ventricular tachycardia remained inducible in 14 patients postoperatively prompting ICD implantation. Three patients with ICDs received appropriate shocks over a median follow-up of 6.1 ± 3.2 years. No patient rendered noninducible by surgical cryoablation died from an arrhythmic cause. However, 2 patients had ICDs implanted, one for sustained monomorphic ventricular tachycardia and the other for recurrent bouts of nonsustained ventricular tachycardia. Finally, a third study assessed a heterogeneous cohort in whom 17 ablations were performed on 18 patients with inducible ventricular tachycardia preoperatively.[37] Ablation of the isthmus responsible for ventricular tachycardia was performed either surgically (2/17), by a catheter (5/17), or using both approaches (10/17). Among these patients, no patient remained inducible on postoperative electrophysiological testing, but 5 patients underwent ICD implantation for primary ($N = 4$) or secondary ($N = 1$) prevention. On follow-up, 3 initially inducible patients experienced ventricular tachycardia (2 of whom had prior implantation of primary prevention ICDs), whereas the third had recurrent ventricular tachycardia after having been rendered noninducible by ablation. He underwent a second catheter ablation procedure along with ICD implantation.

In conclusion, an argument could be made for routine electrophysiologic studies before PVR given that (1) sudden cardiac death is the leading cause of mortality in patients with rToF, (2) patients who require PVR have adverse haemodynamic conditions that can predispose to ventricular arrhythmias, (3) substrates for ventricular arrhythmias are identifiable and ablatable in most patients with rToF, (4) concomitant surgical ablation offers the opportunity for the critical isthmus to be addressed from both endocardial and epicardial surfaces with the potential to create truly transmural lesions, and (5) the PVR itself may impede future attempts at ablation by covering tissue critically involved in the ventricular tachycardia circuit, particularly in the context of transcatheter valves.[39] Further studies are required to draw definitive conclusions about the merits of such an approach.

Conclusions

A diverse range of arrhythmias can be observed in patients with rToF across the lifespan, substantially impacting morbidity and mortality. Bradyarrhythmias, particularly postoperative complete AV block, are a common complication that poses long-term challenges associated with a lifetime of pacing. JET can likewise complicate the postoperative course. Collectively, atrial arrhythmias, namely IART, NAFAT, and AF, are the most frequent arrhythmias encountered. Ventricular arrhythmias are responsible for the majority of sudden cardiac deaths in patients with rToF such that screening for, and appropriately managing, high-risk patients requires diligent attention. Haemodynamics and arrhythmias are intricately linked. New-onset or worsening arrhythmias should incite a thorough assessment for altered cardiac physiological conditions, and, conversely, haemodynamic interventions should prompt careful consideration of arrhythmic issues. Outcomes are likely to be optimized by a multidisciplinary team- and evidence-based approach that considers all facets of care, incorporates patient preferences, and is focused on long-term health span.

Ethics Statement

This is a review article, not a research study. No research is reported.

Patient Consent

The authors confirm that patient consent is not applicable to this review article that retrospectively incorporates de-identified data in the various clinical scenarios presented.

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