ARRHYTHMIAS IN ADULTS WITH CONGENITAL HEART DISEASE

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Refinement of surgical techniques for the treatment of congenital heart disease (CHD) has created a new population of young adults with heart disease. In the USA, it is estimated that there are nearly one million CHD patients, 15–20% with disease of severity to warrant surgical intervention. As surgical mortality has fallen, the number of adults living with major congenital heart defects has increased.[1]

Arrhythmias complicate the care of many adults with CHD. Their prevalence and the difficulty of treatment have made arrhythmia a major focus of interest for physicians working in this area. The presence of longstanding CHD in an arrhythmia patient significantly alters the nature and potential severity of the arrhythmia complaint and the safety and feasibility of various treatments. In addition to analysis of the targeted arrhythmia complaint, the physician must have complete and specific knowledge of the patient's cardiovascular anatomy and the consequences of that anatomy and subsequent surgical modifications on cardiovascular function.

The arrhythmogenic substrate in adults with CHD is complex. All arrhythmias prevalent in the normal population may also occur in CHD, and some specific associations are observed—for example, Wolff-Parkinson-White syndrome and Ebstein's anomaly. However, more common are acquired arrhythmias that are rarely seen in normal young adult hearts, and that are associated with longstanding hypertrophy and fibrosis caused by cyanosis, chronic haemodynamic overload, and superimposed surgical scarring. These arrhythmias include re-entrant atrial and ventricular tachycardias, heart block, and sinus node dysfunction. This article will review the evaluation and management of these more common arrhythmia problems in adults with CHD.

CLINICAL BACKGROUND

Although the anatomical classification of congenital heart defects is complex, three major categories constitute a large percentage of adult CHD patients with arrhythmia, because of their frequency and high incidence of arrhythmia (fig 1).

The spectrum of clinical consequences of arrhythmia in adults with CHD ranges from clinically occult arrhythmia to sudden death. Incessant or recurrent arrhythmia may cause gradual haemodynamic deterioration, and vice versa, often resulting in a vicious cycle of clinical decompensation. Thrombosis[2] and thromboembolic events are also associated with tachycardia. Symptoms, frequent need for hospitalisation, and the management of cardiac devices and antiarrhythmic drugs constitute a significant burden on quality of life.

In the era of effective automatic implantable cardioverter-defibrillator (AICD) therapy, assessment of risk of cardiac sudden death is an important component of clinical arrhythmia management. Patients with acquired heart disease can now be segregated into large, relatively homogeneous groups of similarly elevated risk (for example, patients in the first year after myocardial infarction, or with depressed ejection fraction). In contrast, CHD populations are small, anatomically diverse, and have relatively low rates of sudden death, with annual mortality in even "high risk" groups in the range of $< 2\%$.[3 4] Thus, it is difficult to identify specific risk factors in CHD and to measure the effect of interventions on survival. Nonetheless, some clinical markers appear to affect outcomes adversely over a wide range of anatomical defects, including the presence of residual haemodynamic defects, performance of surgical repair later in life, and longer duration of follow up.

At the other extreme of clinical presentation, not all adults with CHD and arrhythmia are symptomatic, and some experience symptoms so subtle that the amount of the day spent in tachycardia is not easily quantified. A major area for future research will be a clarification of the natural history of arrhythmia in these patients, with attention paid to their role in the gradual deterioration of older patients with CHD.

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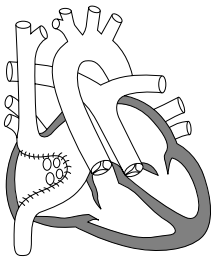
ATRIAL TACHYCARDIAS

Intra-atrial re-entrant tachycardia

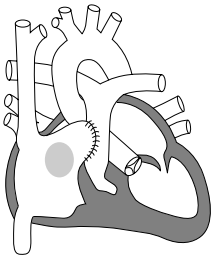
Intra-atrial re-entrant tachycardia (IART) denotes macroreentrant atrial tachycardias other than common atrial flutter occurring in the normal heart. Rare in normal hearts, IART is a common late

Mustard and Senning procedures



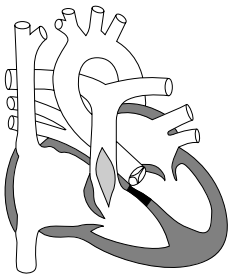
Patients born with transposition of the great vessels in the 1970s through the early 1990s were palliated by the construction of intra-atrial baffles using either synthetic material (the Mustard procedure) or by folding and augmentation of the atrial wall (the Senning procedure). Both redirect caval and pulmonary venous blood to correct cyanosis, utilising the right ventricle as the systemic ventricle. These operations have largely been abandoned in favour of the arterial switch procedure.

Fontan procedures



Many congenital heart defects have in common anatomical features that preclude surgical septation of the ventricles resulting in univentricular physiology. The common end point of staged surgical palliation is the Fontan procedure, which utilises the single ventricle as the systemic ventricle and sends blood directly from the systemic veins to the pulmonary arteries. Several approaches to this have been used; currently, an anastomosis between the superior vena cava and pulmonary artery is created, with an intercaval connection effected by tube graft or intra-atrial baffle.

Repaired Tetralogy of Fallot (TOF)



The prevalence of TOF, its potential for survival through childhood without operation, and the early date at which reparative surgery became available have resulted in large group of patients with relatively homogenous clinical experience. Repair involves closure of the ventricular septal defect (VSD) and relief of right ventricular obstruction, often requiring both ventriculotomy and atriotomy.

Figure 1 Congenital heart malformations commonly associated with arrhythmia.

complication in many varieties of CHD. Like atrial flutter, it tends to have a stable cycle length and P wave morphology, suggesting that it is organised by a fixed substrate. Its prevalence among patients who have undergone surgical procedures involving extensive atrial dissection and repair indicates a particular dependence on surgical injury,[5] and animal models explicitly patterned after surgeries associated with IART (for example, the Mustard and Fontan procedures) result in tachycardias similar to those observed clinically.

Frequently identified risk factors for IART include older age at operation and longer follow up. About half of those patients with "old-style" Fontans—connection of the entire right atrium to the pulmonary artery by anastomosis or conduit—will develop IART within 10 years of surgery,[6] while construction of a lateral right atrial tunnel and cavopulmonary connection are at lower risk.[7] It is anticipated that the extracardiac Fontan, performed using an intercaval tube graft, may also be low risk, but arrhythmia has been reported in early follow up of those patients.[8] Survivors of the Mustard and Senning procedures are at risk for the development of sinus node dysfunction and IART, often concurrently. IART is more prevalent in patients with repaired tetralogy of Fallot (TOF) than ventricular tachycardia, and more likely to be associated with symptoms.[9]

The first large follow up study of IART after CHD surgery revealed a mortality rate over 6.5 years of 17%, with 10% experiencing sudden death. More recently, a group of patients with atrial tachycardias and a prior surgical history of Fontan, Mustard or Senning procedures reported sudden cardiac death in 6% over an average follow up of three years. The clinical factors associated with sudden death were ongoing and/or poorly controlled tachycardia episodes and overall poor clinical status.[5]

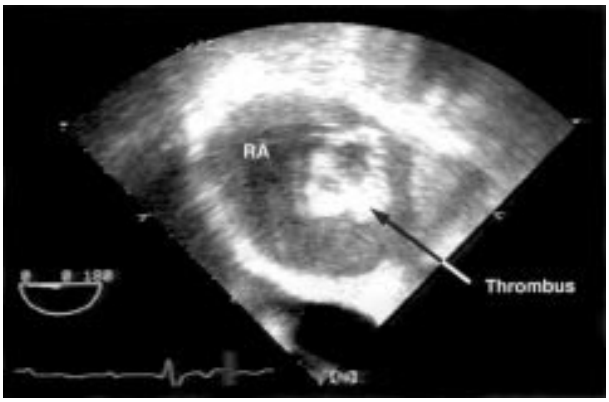


Figure 2 Echocardiographic image of a large thrombus identified in the giant right atrium of a patient with an "old-style" Fontan procedure using an atriopulmonary anastomosis.

Reports of stroke after cardioversion of IART in CHD patients are rare. However, intravascular and intracardiac thromboses are associated with IART, and a prevalence of intracardiac thrombi in 42% of patients undergoing echocardiography before cardioversion has been reported (fig 2).[2] It is not clear whether atrial tachycardias actually promote such events, or are merely a concomitant problem occurring in patients with sick, prematurely aging hearts.

Drug treatment

Although some small studies have suggested otherwise, clinical experience generally has shown that antiarrhythmic drug

Abbreviations

AICD: automatic implantable cardioverter-defibrillator
AV: atrioventricular
IART: intra-atrial re-entrant tachycardia
CHD: congenital heart disease
TOF: tetralogy of Fallot
VT: ventricular tachycardia

treatment is unlikely to suppress recurrences of IART. Experimental models of atrial re-entry have given us a good understanding of the potential salutary effects of class 1C and class 3 drugs, and symptomatic arrhythmias can sometimes be suppressed in individual patients using these agents. However, proarrhythmia and adverse effects on ventricular and nodal function may limit their value. Novel antiarrhythmic drugs with pure class 3 activity have not been widely used in IART, and may prove useful.

The frequent occurrence of thrombosis in adult patients with CHD and atrial tachycardia suggests that warfarin or other potent anticoagulant treatment is indicated in most of these patients. Atrioventricular (AV) nodal blocking drugs may also be used, but are often difficult to titrate because of the relatively slow cycle length and fixed conduction ratios often seen in IART.

Pacemaker therapy

Atrial antibradycardia pacing alone sometimes results in symptomatic improvement and decreased tachycardia frequency.[10] In patients with sinus node dysfunction, this may be the result of improved haemodynamics with appropriately timed atrial activation. Automatic antitachycardia pacing has also been of value for some patients. The overall efficacy of atrial pacing is variable, and there are significant technical difficulties associated with lead placement in these patients. Few endocardial or epicardial sites are generally available and able to generate sensed electrograms of sufficient quality to ensure reliable atrial sensing. Endovascular placement of atrial leads may also increase risk of thrombosis. The potential of other innovative device therapies currently being developed for treatment of atrial fibrillation, such as dual site pacing and the atrial defibrillator, has not been explored in CHD.

Catheter ablation

A proposed curative approach to IART has been to extend or create lines of conduction block, using catheter based and/or surgical techniques. This anatomical approach to treatment involves the design of a lesion or lesions based on an understanding of the relation of macroreentrant circuits to the underlying cardiac anatomy. It has precedents in the catheter and surgical ablation procedures for ventricular tachycardia (VT) and the maze procedure for atrial fibrillation.

Acute success rates reported for radiofrequency catheter ablation for IART range from 55–90%.[11] Catheter ablation procedures usually target individual macroreentrant circuits, seeking a vulnerable site for application of a radiofrequency lesion. Review of IART ablation experience has shown that, in patients with a right AV valve, the isthmus between that valve and the inferior vena cava commonly supports IART, similar to common atrial flutter.[12] When this isthmus is present, as is the case in patients with Mustard and Senning procedures, TOF, and other biventricular repairs, techniques developed for atrial flutter may be used to perform and assess the effectiveness of the ablation. Even in these familiar anatomies, however, the observation of multiple IART circuits is common, and other anatomical or surgical features relevant to ablation may be difficult to locate fluoroscopically. It may also be difficult to generate the large and confluent lesions sometimes needed to interrupt these circuits. Application of recently introduced mapping and ablation techniques, such as advanced activation mapping technologies, and application of irrigated radiofrequency lesions, is associated with improved acute success rates. Longer term follow up after ablation has revealed that arrhythmia symptoms and quality of life are improved in most patients after IART ablation, but recurrences are documented in almost half of these patients.[13] Further advances in our understanding of the arrhythmia substrate and the technology available to visualise and modify it will be necessary to improve this important clinical outcome (fig 3).

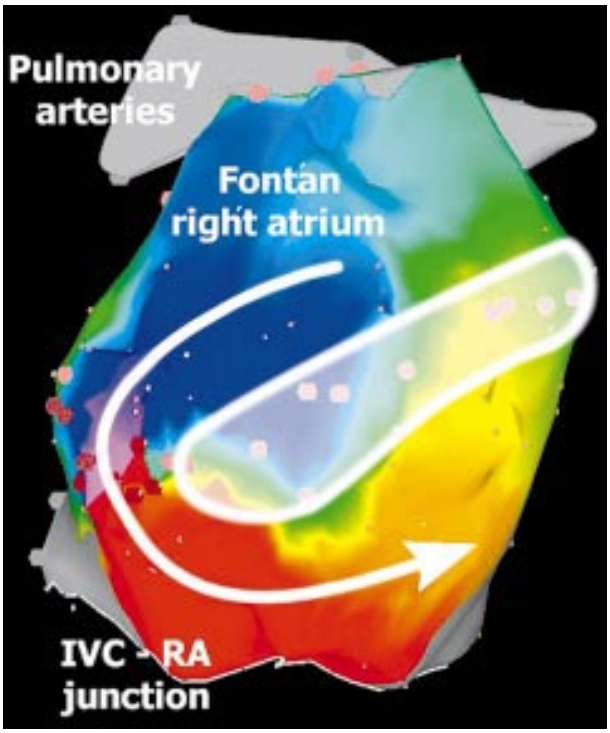


Figure 3 Electroanatomical map in right anterior oblique view of an intra-atrial re-entrant tachycardia circuit constructed in a patient with an older variant of the Fontan procedure. Activation times are colour coded to indicate the movement of the wavefront in this tachycardia, indicated by the white arrow. The white shaded area indicates an area of scarring and conduction block, inferred from characteristics of electrograms recorded from that region.

Surgical treatment

Attempts to revise "old style" Fontan patients to cavopulmonary type connections for haemodynamic reasons are associated with perioperative mortality in the region of 10%,[14] and in the absence of specific intervention for arrhythmia do not reliably prevent arrhythmia recurrence. More recent reports of right atrial maze procedures performed with surgical and/or cryoablative techniques and employing an empiric set of lesions have shown promising results, with no clinically significant arrhythmia recurrence in the majority of patients.[15] This suggests that maze revision of Fontan procedures can be performed at a reasonable surgical risk and may greatly reduce recurrence of postoperative IART. Additional follow up studies are needed to ascertain long term haemodynamic and arrhythmia benefit.

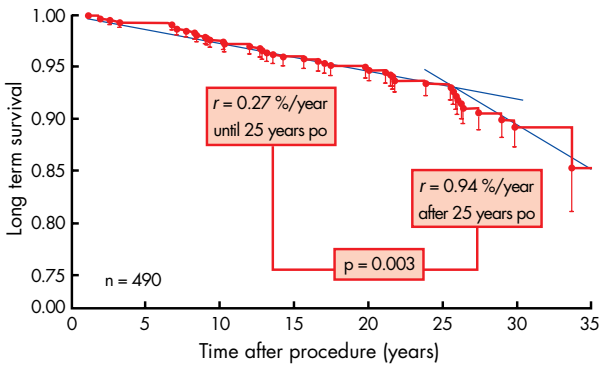


Figure 4 Survival curve in late follow up of adult patients with tetralogy of Fallot. Most deaths were sudden; increased mortality in late decades of follow up has also been observed in other series. Reproduced from Nollert *et al*[4] with permission of the publisher.

Atrial fibrillation

Atrial fibrillation occurs in as many as 25–30% of patients with CHD and atrial tachycardia. The limited information available on these patients suggests that those with residual left sided obstructive lesions or unrepaired heart disease are more prone to atrial fibrillation. Principles of management are drawn from the general adult population, including anticoagulation and rate control. Risk of thromboembolism is presumably elevated. Sinus rhythm is haemodynamically preferred in CHD, and cardioversion, prophylactic antiarrhythmic drugs, and atrial pacing are used to prevent the establishment of permanent atrial fibrillation if possible. The occurrence of atrial fibrillation in patients who also have IART reduces the likelihood that ablation will be beneficial, and may prompt consideration of a surgical maze procedure, though the efficacy of this approach to atrial fibrillation in CHD has not yet been reported. Use of internal atrial defibrillators and ablation of focal atrial fibrillation in the pulmonary veins have not been explored in CHD.

VENTRICULAR ARRHYTHMIAS

Considerable data are available on the natural history data of ventricular arrhythmias and clinical outcomes among patients with TOF, because of its prevalence in the adult CHD population and elevated incidence of ventricular arrhythmia. Mapping studies have shown that, similar to IART, VT in TOF involves a macroreentrant circuit dependant on an anatomical obstacle, in this case the right ventricular outflow tract patch and/or the conal septum.[16]

The long term prognosis for patients with repaired TOF is excellent, with nearly 90% survival at 30 years.[4] Sudden death and VT occur with a reported incidence of 1–2% over five years for young adults and an overall prevalence of sudden cardiac death of 3–6% (fig 4).[4 17] Although clinical presentation of adult TOF patients with sustained monomorphic VT is uncommon, such VT is inducible by programmed stimulation in 15–30% of patients,[18 19] and half have frequent and complex ventricular ectopy on ambulatory ECG.[20] Sinus node dysfunction and IART occur in 20–30% of patients with repaired TOF, and in up to 50% of symptomatic patients,[9] often mimicking VT symptoms and/or causing wide complex tachycardias. These issues make it difficult to apply standard diagnostic tools to screen individuals with clinical arrhythmia symptoms for increased risk of sudden death.

Although patients with Mustard, Senning, and Fontan procedures experience atrial tachycardias and premature mortality, they do not appear to be particularly prone to VT. Data on VT prevalence in other defects are limited. Patients with valvar aortic stenosis, pulmonary stenosis, and ventricular septal defect have been noted to have frequent ventricular ectopy. Aortic stenosis has the highest risk of sudden death among these lesions, but mortality in this defect is characterised by severity of outflow tract obstruction, rather than arrhythmia.

Risk stratification

Simple models of risk stratification for sudden death (for example, ejection fraction) do not exist for adult CHD patients. Assessment of the risk of sudden death caused by ventricular arrhythmia requires an understanding of the limited predictive values of commonly used diagnostic tests in this population. Although Holter, exercise testing, and programmed ventricular stimulation are useful for provoking and/or recording clinically documented arrhythmias, their value as screening tests is unclear. Risk assessment is further complicated by the occurrence of atrial tachycardias, which may also cause symptoms and sudden death.

Several clinical features are associated with VT and sudden death in adult CHD patients, including older age, older age at repair, and poorer haemodynamic status. Electrocardiographically, pronounced prolongation of QRS duration and prolongation dispersions of the QT and JT intervals—poorly understood indices of ventricular repolarisation—are associated with cardiomegaly, mortality, and inducible sustained VT in TOF patients.[21] These findings identify a more arrhythmogenic myocardium and suggest that both depolarisation and repolarisation are abnormal in high risk TOF patients. Because of the ubiquity of lower grades of ventricular ectopy in this population, ambulatory ECG is often abnormal, and in the absence of significant runs of VT it may be of limited value in discriminating patients at elevated risk.

The value of programmed ventricular stimulation in patients with CHD is unclear. In one large series evaluating programmed stimulation in patients with a variety of defects, inducibility of VT predicted subsequent cardiac arrest and mortality after adjustment for covariate clinical factors, but also emphasised the importance of careful selection of patients for study on the basis of those clinical features.[22] In another study of adults with TOF, no patients who subsequently died suddenly had inducible VT.[18] Both false positive studies[19] (inducible VT in patients without VT or mortality on follow up) and false negative studies (non-inducibility of patients with documented sustained VT) occur with appreciable frequency.

Management

Minimally symptomatic patients with non-sustained ventricular ectopy must be evaluated to determine whether an associated evolution of underlying abnormal haemodynamics or metabolism has occurred. If not, periodic clinical monitoring and non-invasive assessment (ECG, echo, and Holter monitoring) are probably sufficient. Event monitoring may be useful for investigation of arrhythmia symptoms. More ominous arrhythmia presentations such as syncope, near syncope with palpitation or non-sustained VT should trigger more comprehensive inquiry, including catheterisation with haemodynamic assessment and programmed atrial and ventricular stimulation. Patients with negative studies, minimal symptoms, and good haemodynamics are managed without treatment, or by using drugs with a favourable side effect

profile (such as β blockers) to suppress symptomatic ectopy. Supraventricular tachycardia is treated with ablation when possible, and severe bradycardia managed with pacing. Patients with severe symptoms or inducible VT are considered for more aggressive antiarrhythmic drug treatment and AICD placement (fig 5).

Antiarrhythmic drugs may be useful for suppression of symptomatic ventricular arrhythmias, but have not been shown to prolong survival in CHD. AICD therapy is feasible in many patients with CHD, and its use is increasing. Catheter ablation of VT has been successful in small series of patients with CHD, and may be appropriate for patients with sustained, monomorphic VT that is haemodynamically tolerated.[23] When patients warrant surgery for haemodynamic reasons, attempts to resect potential critical zones for VT may be considered. Recently, indications have broadened for pulmonary valve replacement in patients with symptoms and/or signs of right heart failure and pulmonary regurgitation—many of whom also have prolonged QRS duration on ECG. The effect such surgical intervention may have on ventricular arrhythmia is unknown.

BRADYCARDIA

Sinus node dysfunction

Gradual loss of sinus rhythm occurs after the Mustard and Senning and all varieties of Fontan procedures.[24] Patients with heterotaxy syndromes, particularly left atrial isomerism, may also have congenital abnormalities of the sinus node independent of the effects of their surgical procedures. Paroxysmal atrial tachycardias are frequently associated with sinus node dysfunction, and loss of sinus rhythm appears to increase risk of sudden death.

Electrophysiological study of patients with the Mustard procedure have identified a variety of abnormalities of atrial electrophysiology, including prolonged sinus node recovery times, intra-atrial conduction times, and atrial refractoriness.[25] Direct surgical injury to the sinus node has been proposed as a cause of observed abnormalities of sinus node function. However, the progressive loss of sinus rhythm observed over extended follow up implies additional ongoing pathophysiological processes related to chronic haemodynamic abnormality.

AV block

Interventricular conduction abnormalities, particularly right bundle branch block, are very common after surgery for CHD. Complete postoperative heart block is caused either by direct surgical injury to the specialised conduction system or by indirect damage due to inflammatory response. It is typically associated with surgical manipulation of the ventricular septum. Patients at highest risk are those undergoing surgery for left ventricular outflow tract obstructions and patients with ventricular inversion (L-transposition of the great arteries), but it is also common after ventricular septal defect and TOF repairs. Review of clinical outcomes before cardiac pacing systems appropriate for CHD patients were available showed that postoperative heart block had a high mortality rate, even in the presence of an escape rhythm.

Complete heart block also occurs spontaneously in patients with certain structural heart defects, especially endocardial cushion defects and ventricular inversion. This may be caused by aberrant anatomy of the AV node and His bundle in these patients, rendering them vulnerable to injury. Although some of these patients present with heart block at birth, it may progress at any stage of life.

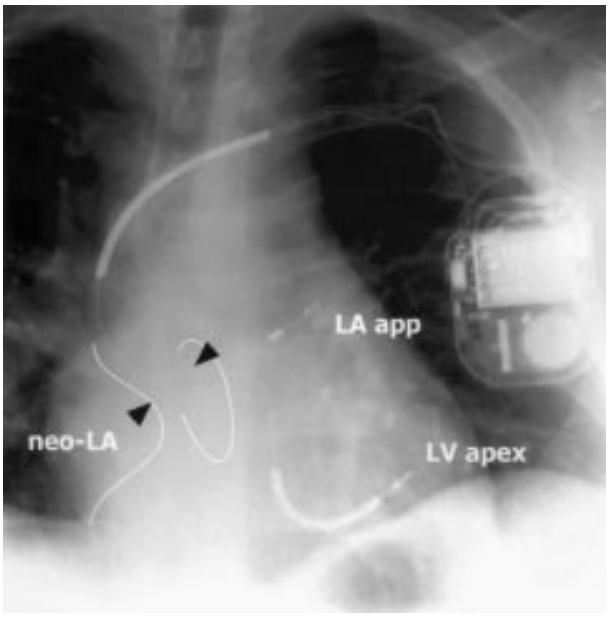


Figure 5 Radiograph illustrating a variety of technical issues with pacemaker placement in a patient who has undergone the Mustard procedure. The ventricular pacing lead is located in the apex of the left ventricle (LV apex), and the atrial lead in the mouth of the left atrial appendage (LA app). Both traverse the superior limb of the Mustard baffle between the superior vena cava and the left atrium, which was stenotic and required stenting (black arrows) to relieve obstruction before lead placement. The presumed locations of the lateral margin of the intra-atrial baffle, defining the pulmonary venous atrial channel (neo-LA) and the communication between left and right atria, are highlighted in white.

Pacemaker issues

While heart block is a clear indication for permanent cardiac pacing in CHD, others are less well substantiated. Many patients with CHD tolerate chronic bradycardia well, but pacing may alleviate symptoms such as fatigue, dizziness, or syncope in some patients with junctional escape rhythms, severe resting bradycardia, chronotropic incompetence, and/or prolonged pauses. Pacing may also be necessary to permit treatment with antiarrhythmic drugs.

Cardiac pacing in adults with CHD presents a variety of special challenges (fig 5). Congenital and acquired cardiovascular abnormalities and shunting may limit opportunities for endocardial lead placement and necessitate an epicardial or even a hybrid approach. Examples include patients with old transvenous lead systems who may have associated acquired vascular abnormalities, and Fontan patients, in whom the ventricular cavities and much or all of the atrial myocardium are surgically excluded from systemic venous pathways. Patients with the Mustard and Senning procedures may receive transvenous dual chamber pacing systems, and even AICDs, but the leads must navigate the superior limb of the intra-atrial baffle, which is prone to obstruction. Atrial lead placement in unusual sites may be difficult and must avoid inadvertent stimulation of the phrenic nerve. Because asynchronous atrial pacing may provoke IART, careful lead site selection resulting in excellent sensing of atrial electrical activity is important.

Clinical experience shows the value of AV synchrony and favours implantation of a system capable of providing a physiological heart rate response. However, the specific value of rate responsive and dual chambered pacing as compared to

Arrhythmias in adults with CHD: key points

- Although long term survival and clinical outcomes for adults with congenital heart disease (CHD) are generally good, arrhythmias are a significant cause of morbidity and mortality in this group of patients, especially in later decades of follow up
- Strategies for individual risk assessment are limited, but groups at particular risk for arrhythmia include patients with the Mustard and Senning procedure for transposition of the great vessels, patients with the Fontan procedure, and patients with repaired tetralogy of Fallot
- In most forms of CHD, atrial tachycardias appear to be more prevalent than ventricular tachycardias, frequently symptomatic, and associated with an increased risk of thrombosis and death
- Interventional strategies are currently in development for treatment of atrial and ventricular tachycardia in patients with CHD, and include innovative applications of catheter based and surgical ablative procedures, and antitachycardia and defibrillator device therapies

simpler pacing modalities is not well established in CHD. Practical limitations often require that the choice of system be adapted to patient specific problems faced with lead placement and maintenance. Exploration of the potential utility of new device technologies in CHD, such as dual site pacing for ventricular resynchronisation and atrial defibrillators, will further challenge the inventiveness of physicians caring for these patients.

CONCLUSION

Our understanding of arrhythmia in adults with CHD has progressed rapidly, through increased appreciation of the extended natural history of these patients and innovative application of treatments designed for and tested in patients without CHD. Patients with these tachycardias have poor outcomes, but the small size and anatomical diversity of this group make it difficult to determine which patients are most at risk and whether arrhythmia control will lead to measurable gains in longevity and health. Animal models and the application of evolving therapeutic technologies have provided us with valuable insights into the anatomical substrates of arrhythmia in this group, and helped to understand some of the problems with preventing their recurrence. Development of a more complete picture of the underlying pathophysiological changes in the myocardium that lead to these arrhythmias will help to focus further efforts to improve our current therapeutic outcomes.

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20 **Cullen S**, Celermajer DS, Franklin RC, *et al.* Prognostic significance of ventricular arrhythmia after repair of tetralogy of Fallot: a 12-year prospective study. *J Am Coll Cardiol* 1994;**23**:1151–5.

21 **Gatzoulis MA**, Till JA, Redington AN. Depolarization-repolarization inhomogeneity after repair of tetralogy of Fallot: the substrate for malignant ventricular tachycardia? *Circulation* 1997;**95**:401–4.

▶ **This and earlier reports from the same group associate prolongation and variability in resting ECG intervals with increased risk of ventricular arrhythmia and death, and propose possible pathogenetic mechanisms to link the two.**

22 **Alexander ME**, Walsh EP, Saul JP, *et al*. Value of programmed ventricular stimulation in patients with congenital heart disease. *J Cardiovasc Electrophysiol* 1999;**10**:1033–44.
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23 **Gonska BD**, Cao K, Raab J, *et al*. Radiofrequency catheter ablation of right ventricular tachycardia late after repair of congenital heart defects. *Circulation* 1996;**94**:1902–8.
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