Invasive electrophysiology in paediatric and congenital heart disease

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INVASIVE ELECTROPHYSIOLOGY IN PAEDIATRIC AND CONGENITAL HEART DISEASE

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In the last decade invasive electrophysiological studies (EPS) and radiofrequency catheter ablation (RFCA) have become progressively common, driven predominantly by high success and low complication rates, coupled with the inefficacy and side effects of many antiarrhythmic agents. As our understanding of different arrhythmia mechanisms and successful ablative strategies has advanced, RFCA has been applied to increasingly complex substrates and broader clinical indications. Two such examples are young adults with native or surgically repaired congenital heart disease and children with structurally normal hearts. In such situations arrhythmia management should be performed on an individualised basis, considering the potential risks and benefits of different strategies.

In young children the natural history of the arrhythmia, the effects of medication and potential life-threatening complications must all be weighed against the risk of RFCA. In adults with congenital heart disease, arrhythmia management must be closely coordinated between electrophysiologist, cardiologist and cardiac surgeon to optimise haemodynamic performance, potentially performed in conjunction with surgery or other percutaneous interventions. This article aims to review the current indications for EPS and RFCA in both groups of patients, specifically focusing on areas of ongoing interest and debate.

CHILDREN WITH STRUCTURALLY NORMAL HEARTS

Arrhythmia mechanism and presentation

The vast majority of supraventricular tachycardias (SVT) seen in children without congenital cardiac anomalies are atrioventricular reentry tachycardia (AVRT) facilitated by an accessory pathway (AP), atrioventricular nodal reentry tachycardia (AVNRT) and focal atrial tachycardia (FAT). Presentation of AVRT is most common in the neonatal and infant groups, with the vast majority free from symptomatic arrhythmia by the end of the first year of life, although recurrence in later childhood or adolescence is well recognised. Spontaneous remission in those older than 5 years of age at presentation is much less common.[1] Conversely AVNRT is rare in early childhood, becoming more frequent with increasing age, mirroring patterns seen in adult practice. Other less common mechanisms include junctional ectopic tachycardia (JET) and permanent junctional reciprocating tachycardia (PJRT), both of which may be incessant precipitating ventricular dysfunction, and atrial flutter (AFL) which is seldom encountered outside the neonatal age group. Different arrhythmia mechanisms are summarised in fig 1.

Ventricular tachycardia (VT) may be seen at any age throughout childhood with a peak occurring in infancy, although the overall incidence is much lower than that of supraventricular arrhythmias. The outcome is favourable, especially in those who present in infancy with VT originating from the right ventricle. Only nine of 98 patients described in a large retrospective, multicentre analysis of idiopathic childhood VT required RFCA, which was successful in 78%.[2]

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Procedural considerations

Children undergoing invasive EPS with or without RFCA represent a unique population with needs different from their adult counterparts undergoing the same procedure. Careful consideration should be given to the nature of sedation or general anaesthesia, ensuring adequate patient comfort and analgesia with minimum risk. Procedures should be performed in specific facilities with necessary monitoring and safety equipment by a team of experienced electrophysiologists, cardiologists, nursing staff and cardiac physiologists.

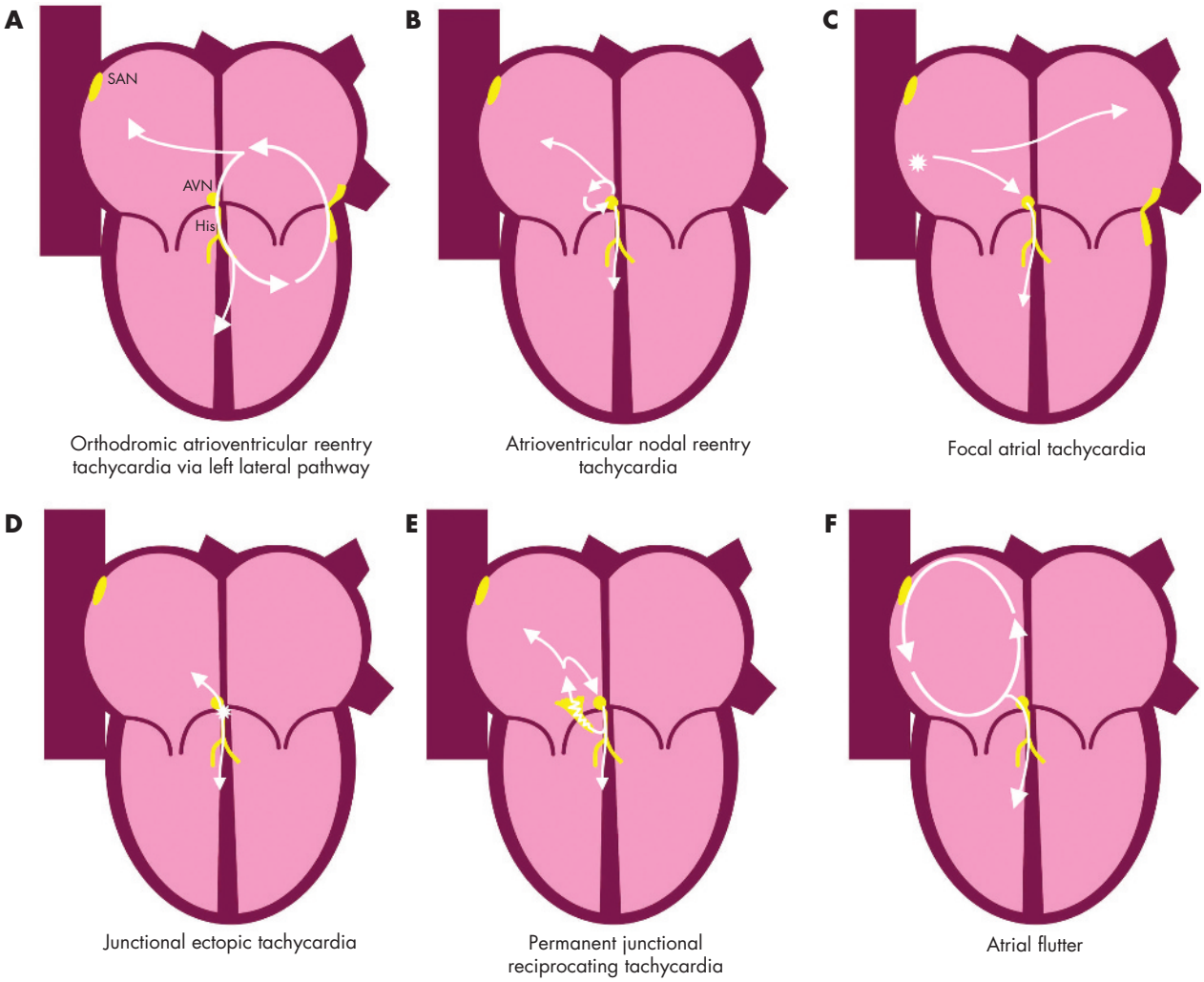

Figure 1 The mechanism of arrhythmia commonly encountered in children. The heart is depicted in a four-chamber view, with the sinoatrial node (SAN), atrioventricular node (AVN) and His bundle (His) depicted in yellow. In all figures white arrows denote activation. (A). Orthodromic atrioventricular reentry tachycardia (AVRT) via a left lateral pathway shows activation progressing anterogradely to the ventricles via the AVN/His bundle and retrogradely to the atria via the left-sided accessory pathway. Antidromic AVRT would be the reverse pattern of activation. (B) Atrioventricular nodal reentry tachycardia (AVNRT) involves the AVN, and two connections between atrial myocardium and node, the ''slow'' and ''fast'' pathways. In typical (slow/fast) AVNRT activation progresses anterogradely in the slow pathway, located in the septal isthmus between the coronary sinus os and compact AVN, before activating the ventricles via the His bundle and the atria retrogradely via the fast pathway. (C) Focal atrial tachycardia is activation of the heart from a source within either the right or left atrium outside the sinus node complex. This is typically manifest by a tachycardia with a 1:1 atrioventricular relationship but an altered P wave morphology, axis and PR interval. (D) Junctional ectopic tachycardia (JET) is most commonly seen in the postoperative period in infants undergoing cardiac surgery, although may rarely occur as an isolated finding. This is an automatic tachycardia originating from the His bundle with anterograde conduction to the ventricles and retrograde conduction to the atria via the AVN. The surface ECG classically shows a narrow complex tachycardia with an irregular RR interval and VA relationship. (E) Permanent junctional reciprocating tachycardia (PJRT) is an incessant arrhythmia facilitated by slow conduction (zig-zag arrow) via an accessory pathway (AP), most commonly located in the posteroseptal region of the right atrium. The ECG shows a narrow complex tachycardia with a long RP interval and abnormal P wave axis dependent on the direction of atrial activation. (F) Atrial flutter is a reentrant circuit within the right atrium rarely seen outside the neonatal period in the normal heart. In typical (counterclockwise) flutter activation progresses up the atrial septum and down the lateral wall and via the isthmus between the tricuspid valve and inferior vena cava.

Diagnostic electrophysiology study

Invasive diagnostic EPS in the catheter laboratory is rarely performed alone without progression to RFCA, although there may be variation between individual institutions and patients depending on co-existing factors. Assessment of arrhythmia mechanism and hence management can largely be guided by 12-lead electrocardiograms (ECGs) and non-invasive assessment. Specific indications for invasive diagnostic assessment may include attempted induction in a patient with recurrent symptoms but no documented arrhythmia, especially if associated with haemodynamic compromise where VT is a potential diagnosis. Invasive EPS in children with AVRT secondary to a concealed AP where atrioventricular (AV) block is unacceptable will allow localisation of the pathway, providing a better prediction of the ablative risk, and will allow risk stratification in patients with asymptomatic

Table 1 Potential indications for diagnostic electrophysiology study

- Diagnosis of arrhythmia in a patient with persistent symptoms and no documented arrhythmia
- Assessment of risk of inadvertent atrioventricular block in a child where long-term pacing is unacceptable
- Assessment of symptoms in patients known to be at risk of life-threatening arrhythmias (eg, VT in tetralogy of Fallot)
- Diagnosis of broad complex tachycardia
- Risk stratification:
  - asymptomatic WPW ECG pattern

ECG, electrocardiogram; VT, ventricular tachycardia: WPW, Wolff-Parkinson-White.

Wolff-Parkinson-White (WPW) ECG pattern. Complications are rare, although in a recently reported series Pappone *et al* performed 165 diagnostic EPS in children aged 5–12 years with an asymptomatic WPW ECG pattern, documenting a complication rate of 6%, half of which were related to anaesthesia.[3] Potential indications for diagnostic EPS are summarised in table 1.

A less invasive study using a bipolar oesophageal electrode allows determination of a wide variety of electrophysiological parameters to better define the mechanism in the vast majority of cases (for example, differentiating AVRT from AVNRT) and hence management. Cardioversion by rapid atrial pacing may also be performed via an oesophageal electrode and is successful in virtually all cases of paroxysmal SVT and 73% of cases of intra-atrial reentry/AFL.[4]

Radiofrequency catheter ablation

The indications for RFCA in children were recently reported by an expert consensus conference and are documented in table 2.[5] The evolution, success and complications of RFCA in children has been documented by members of the Pediatric Electrophysiology Society who have submitted data to a central registry since its inception in 1990.

To compare early (1991–1995) and later (1996–1999) eras Kugler *et al* reported the success rates related to arrhythmia substrate (AVRT, AVNRT and FAT) in 7600 children undergoing RFCA during the study periods.[6] There was a significant improvement in overall procedural success from 90.4% to 95.2%, with improved outcome seen in all substrates except anteroseptal AP and FAT. More recently (1999–2003) Van Hare reported a 95.7% success rate for AVRT and AVNRT, although also noted an important adjusted recurrence rate of 10.7% at 12 months in a prospective assessment after successful RFCA. Recurrence rate varied by location and mechanism, most frequently seen after RFCA of right sided septal pathways and least common after AVNRT.[7]

Concomitant with the rise in success rates, there has been a documented fall in procedural complications. While RFCA is largely safe, major complications are well described including injury to epicardial coronary arteries, cardiac perforation, systemic thromboembolism, permanent AV block and death. Between 1991–1995 and 1996–1999 there was an overall fall in the complication rate from 4.2% to 3%, but no change in major complications or procedural-related death, although the power of this analysis was limited by the low incidence of adverse events.[6] Latterly (1999–2003) the overall complication rate was reported as 4.2% and 4.0% for EPS and RFCA, respectively, with no mortality, which was suggested by the authors to reflect an improvement in operator skill, available technology and case selection.[7]

Due to the requirement for long-term pacing with all its inherent sequelae, AV block remains an important major complication in children undergoing RFCA, most commonly seen in association with AVNRT and septal pathways. The incidence of AV block as documented by the Pediatric RFCA Registry was 0.88% (1991–1995), 0.56% (1996–1999) and 1.2% (1999–2003). Mortality associated with RFCA is rare, reported in 5/4092 children (0.12%) and associated with left sided

Table 2 Indications for radiofrequency catheter ablation procedures in paediatric patients

Class I
1. WPW syndrome following an episode of aborted sudden cardiac death
2. The presence of WPW syndrome associated with syncope when there is a short pre-excited R-R interval during atrial fibrillation (pre-excited R-R interval, 250 ms) or the anterograde effective refractory period of the AP measured during programmed electrical stimulation is <250 ms
3. Chronic or recurrent SVT associated with ventricular dysfunction
4. Recurrent VT that is associated with haemodynamic compromise and is amenable to catheter ablation

Class II A
1. Recurrent and/or symptomatic SVT refractory to conventional medical therapy and age 4 years
2. Impending congenital heart surgery when vascular or chamber access may be restricted following surgery
3. Chronic (occurring for 6–12 months following an initial event) or incessant SVT in the presence of normal ventricular function
4. Chronic or frequent recurrences of IART
5. Palpitations with inducible sustained SVT during electrophysiological testing

Class II B
1. Asymptomatic pre-excitation (WPW pattern on an ECG), age 5 years, with no recognised tachycardia, when the risks and benefits of the procedure and arrhythmia have been clearly explained
2. SVT, age >5 years, as an alternative to chronic antiarrhythmic therapy, which has been effective in control of the arrhythmia
3. SVT, age <5 years (including infants), when antiarrhythmic medications, including sotalol and amiodarone, are not effective or associated with intolerable side effects
4. IART, 1–3 episodes per year, requiring medical intervention
5. AVN ablation and pacemaker insertion as an alternative therapy for recurrent or intractable IART
6. One episode of VT associated with haemodynamic compromise and which is amenable to catheter ablation

Class III
1. Asymptomatic WPW syndrome, age <5 years
2. SVT controlled with conventional antiarrhythmic medications, age <5 years
3. Non-sustained, paroxysmal VT which is not considered incessant (ie, present on monitoring for hours at a time or on nearly all strips recorded during any 1 hour period of time) and where no concomitant ventricular dysfunction exists
4. Episodes of non-sustained SVT that do not require other therapy and/or are minimally symptomatic

Class I: There is consistent agreement and/or supportive data that catheter ablation is likely to be medically beneficial or helpful for the patient.
Class II: There is a divergence of opinion regarding the benefit or medical necessity of catheter ablation. **II A:** The majority of opinions/data are in favour of the procedure. **II B:** There is clear divergence of opinion regarding the need for the procedure.
Class III: There is agreement that catheter ablation is not medically indicated and/or the risk of the procedure may be greater than benefit for the patient.
AP, accessory pathway; AVN, atrioventricular node; ECG, electrocardiogram; IART, intra-atrial reentrant tachycardia; SVT, supraventricular tachycardia; VT, ventricular tachycardia; WPW, Wolff-Parkinson-White.
Reprinted from Friedman *et al*[5] with permission from Blackwell Publishing.

procedures in all cases. Death was related to myocardial perforation, coronary and cerebral thromboembolism and arrhythmia, but not access to the left heart or the site of lesion placement, suggesting that left-sided RFCA carries a small but identifiable risk.[8] The application of radiofrequency energy close to coronary arteries highlights the possibility of injury to the coronary vasculature, which is classically associated with RFCA of the AP at the atrioventricular junction, but has also been described in association with slow pathway ablation for AVNRT.[w1] In younger children where the epicardial structures are closer to the catheter tip this risk is theoretically increased. The fact that coronary injury may be easily missed and the potential for expansion of lesion size applied to the maturing myocardium suggests coronary stenosis presenting late after RFCA may be seen in the future.[w2]

Radiofrequency catheter ablation in infants

Traditionally RFCA in infants <15 kg has been associated with an increased complication rate. Because of the natural history of the predominant tachycardia mechanism in this age group, RFCA is generally deferred unless specific indications exist, usually refractory or life threatening arrhythmia with associated ventricular dysfunction. Specific procedural modifications may be used to facilitate RFCA in smaller patients, including the use of 5 French ablation catheters and single catheter technique in a smaller chamber. The application of 5–10 second "test" lesions and reduced power and temperature settings potentially alleviates immediate and long-term myocardial injury.

A retrospective analysis reviewed the outcome and complication rates of RFCA in infants <18 months compared to older children.[9] No difference was found between the two groups either for success rates in all tachycardia substrates or the incidence of major complications. Secondary analysis of all children weighing <15 kg and older than 18 months did find a significant association between weight and major, but not total, complication rate. The authors concluded that RFCA performed by experience operators is a powerful tool in the management of drug refractory and life threatening arrhythmias in younger patients. Long-term follow-up addressing the implications of RFCA in the maturing human myocardium are awaited.

Asymptomatic Wolff-Parkinson-White ECG pattern

Palpitation due to AP-mediated AVRT is the most common reason patients with WPW come to the attention of cardiologists and electrophysiologists. The unexpected finding of anterograde atrioventricular conduction over a manifest AP, termed WPW ECG pattern, poses an interesting question related to management. Patients with this ECG pattern may remain asymptomatic for many years while some may go on to develop symptomatic AVRT. A small proportion of both adults and children are at risk of life-threatening arrhythmias predisposing to sudden cardiac death, which may be the first manifestation of the disease. The accepted mechanism is degeneration of AVRT into atrial fibrillation (AF), which initiates ventricular fibrillation (VF) by anterograde conduction over an AP with rapid conduction velocity and a short refractory period. Identifying this small section of the WPW population has been the cause of much interest and investigation over many years.

Abbreviations

- **AERP:** anterograde effective refractory period
- **AF:** atrial fibrillation
- **AFL:** atrial flutter
- **AP:** accessory pathway
- **ASD:** atrial septal defect
- **AV:** atrioventricular
- **AVNRT:** atrioventricular nodal re-entrant tachycardia
- **AVRT:** atrioventricular reentry tachycardia
- **IART:** intra-atrial reentrant tachycardia
- **EPS:** electrophysiological studies
- **ERP:** effective refractory periods
- **FAT:** focal atrial tachycardia
- **JET:** junctional ectopic tachycardia
- **MEA:** multi-electrode array
- **PJRT:** permanent junctional reciprocating tachycardia
- **PVS:** programmed ventricular stimulation
- **RFCA:** radiofrequency catheter ablation
- **SPRR:** shortest pre-excited R-R interval
- **SVT:** supraventricular tachycardias
- **VF:** ventricular fibrillation
- **VT:** ventricular tachycardia
- **WPW:** Wolff-Parkinson-White

Electrophysiological parameters that may render individuals at risk of VF are the shortest pre-excited R-R interval (SPRR) and the AP effective anterograde refractory period (AERP). Measurements of SPRR in patients with documented VF and those who were asymptomatic showed considerable overlap between the two groups, emphasising that no one factor has sufficient specificity or positive predictive value to accurately delineate those at highest risk.[w3]

Due in part to the complexity of identifying an at risk group and the rarity of life threatening arrhythmias in patients with WPW ECG pattern, routine electrophysiological testing has not to date been recommended as part of clinical guidelines. Todd *et al* reviewed the data from numerous electrophysiological and population-based studies, finding only two episodes of sudden cardiac death in 780 patients with follow-up ranging from 1.8 to 21.8 years. In the longest running of these analyses only 15% of cases progressed to symptomatic AVRT and there were no sudden deaths.[10] [w4]

However, recent publications from Pappone *et al* have prompted reassessment of the way in which we manage children with asymptomatic WPW pattern.[3] In a prospective analysis of the benefits of routine electrophysiological testing to risk stratify, the authors found that arrhythmia induction (AVRT and/or AF) at the time of EPS as opposed to AP AERP more accurately identified those who subsequently became symptomatic.[w5] This criterion was then applied to a large cohort of 165 children aged between 5–12 years found to have asymptomatic WPW pattern on routine ECG testing. Inducibility identified 60 high risk patients, who were randomly assigned to RFCA or continued observation. Prophylactic ablation resulted in a high rate of arrhythmia freedom (95%), while the incidence of arrhythmia in the control group was 44% with all events occurring within a 12-month period. Of specific interest was the finding of asymptomatic pre-excited AF with rapid ventricular rates in five children, of whom two later developed documented VF and

Invasive electrophysiology in paediatric heart disease: key points

- Radiofrequency catheter ablation (RFCA) is a safe and effective procedure in children with outcomes comparable to the adult population
- Considering the limited success and side effects of anti-arrhythmic medication with the need for strict compliance, RFCA may be considered first line therapy in many situations
- RFCA in infants is equally associated with high success and low complication rates when performed by experienced operators, and should be considered in cases of drug refractory and/or life-threatening arrhythmias
- The management of asymptomatic Wolff-Parkinson-White ECG pattern remains debated

one died suddenly, although the rhythm was not documented. This report has important implications for the management of children with asymptomatic WPW pattern, although whether the adoption of routine EPS and RFCA becomes accepted by paediatric electrophysiologists and incorporated into clinical guidelines remains to be seen. A survey published before Pappone's report suggested that the majority of paediatric electrophysiologists would perform a diagnostic EPS, progressing to RFCA based on the SPRR rather than inducibility of AVRT.[11]

ADULT CONGENITAL HEART DISEASE

In the adult population arrhythmia complicates many variants of native and surgically repaired congenital heart disease, which provide an ideal substrate for different reentrant and focal mechanisms. Multiple factors are undoubtedly contributory, including abnormal haemodynamic loading, repeated surgical intervention, distortion of chamber anatomy and sinus node dysfunction. The limited success and frequent cardiovascular and other systemic side effects of many antiarrhythmic agents has limited their use, such that RFCA plays an important part in the arrhythmia management of these complex patients. Despite the success of this approach there are still many areas where the optimum treatment strategy is debated, and the use of both EPS and RFCA as either prognostic or therapeutic tools remains uncertain. While the prevalence of arrhythmia in adult congenital heart disease is unquestioned, overall the numbers are small, which hinders large-scale prospective trials to help identify a strategy with maximum benefit.

Atrial septal defect

In multiple longitudinal analyses, atrial arrhythmias are well recognised as a long-term complication in patients with an atrial septal defect (ASD).[W6] Due to the significant morbidity associated with AF—specifically that related to thromboembolic disease—prompt restoration of sinus rhythm in a relatively young population is highly desirable. In a 40-year-old man who, on investigation for recurrent, self-limiting palpitations, is found to have a large ASD, what is the appropriate management regarding his arrhythmia?

The benefits of ASD closure on life expectancy and heart failure are clear, although early studies comparing medical management with surgery failed to demonstrate any anti-arrhythmic benefit of intervention. Restoration of sinus rhythm cannot be achieved in the majority of those who undergo surgical closure after the development of sustained arrhythmias, while an additional 8% have been shown to develop new arrhythmias over a mean follow-up period of 3.8 years.[12] In this particular study, age >40 years and preoperative arrhythmias were predictive of postoperative AFL or AF. Similarly an increased propensity to arrhythmia following a percutaneous approach has been reported in those with previously documented paroxysmal or persistent AF or AFL, although the age limit associated with arrhythmia freedom in the medium term was 55.[13] Longer follow-up after ASD closure, either surgical or interventional, is required to assess the continued arrhythmia risk.

The different mechanisms of atrial arrhythmia in patients with an ASD have not been clearly delineated, with many studies simply focusing on their presence or absence. Electrophysiological remodelling of the right atrium associated with ASD can be demonstrated, specifically prolongation of atrial effective refractory periods (ERP), sinus node dysfunction and conduction block at the crista terminalis, most likely a result of chronic atrial stretch.[14] Persistent conduction block at the crista, which has been implicated in typical/atypical AFL and AF, is demonstrable after successful closure.[14] The evolution of transcatheter implantation of ASD devices may reduce the incidence of incisional macro-reentry circuits, although clearly significant electrophysiological changes and incomplete volume reduction persist; this suggests that closure may delay rather than eliminate the arrhythmia risk, and that a proportion of these patients may be prone to AFL and/or AF at an earlier age than the normal population. If an empiric ablative strategy, either percutaneous or surgical, is to be adopted, a better understanding of the evolution and source of different arrhythmias is necessary to ensure timely and appropriate intervention.

Tetralogy of Fallot

Despite excellent results for surgical correction of tetralogy of Fallot for over 40 years, VT and sudden cardiac death remain significant complications, documented at 4.2% and 2.0%, respectively, in a recent multicentre study conducted over a 10-year period.[15] Numerous markers have been proposed to identify those at risk, including QRS duration >180 ms, older age at repair, and rapid progression of QRS duration. Persistent pulmonary regurgitation secondary to the use of a transannular patch at the time of initial surgery is the haemodynamic lesion most associated with VT and sudden cardiac death.

The role of invasive programmed ventricular stimulation (PVS) in congenital heart disease to predict those at high risk of VT or sudden cardiac death has been limited by a low incidence of clinically significant events in comparatively small groups of heterogeneous patients. Due to its relative prevalence, tetralogy of Fallot has been more extensively studied than other lesions associated with VT such as transposition of the great arteries following the atrial switch procedure, varying types of left ventricular outflow tract obstruction, and ventricular septal defect repair. Patients are commonly referred for PVS based on clinical findings, arrhythmia symptoms or the presence of sustained ventricular arrhythmias on non-invasive assessment. In a large cohort of patients with a variety of congenital cardiac lesions who underwent PVS on clinical grounds, a positive

study was associated with a six-fold increase in mortality and a three-fold increase in serious arrhythmic events using multivariate analysis. The combination of symptoms, patient and ECG characteristics and anatomy together with a positive PVS predicted mortality, with a sensitivity of 87% and positive predictive value of 24%, although the study reported a significant false negative rate (37%).[16]

The predictive value of PVS for VT and sudden cardiac death in tetralogy of Fallot was recently assessed in a large multicentre study, performed in 159 of 252 patients for clinical symptoms and/or documented arrhythmia and 93 as routine screening.[17] The induction of both monomorphic and polymorphic VT predicted future VT and sudden cardiac death, with inducible sustained VT an independent risk factor for further events. As previously noted by Alexander, sustained polymorphic VT should not be disregarded as non-specific.[16] Arrhythmia inducibility was also found to independently predict future VT and sudden cardiac death in those screened routinely, although the clinical features of these patients are not reported. The positive and negative predictive values were, respectively, 25% and 99% in those who underwent PVS as routine screening, as opposed to 67% and 86% in those with clinical indications. The benefit of specific non-invasive indicators identifying those who should proceed to invasive PVS is currently under active investigation.[17] Ultimately the timing and outcome of PVS, coupled with other clinical features such as patient age, ECG parameters, right ventricular size and function may enable better risk assessment and treatment, specifically as regards the use of implantable cardioverter-defibrillators.

MAPPING AND ABLATION SYSTEMS

Conventional electrophysiological mapping techniques employ multipolar catheters placed at specific locations within the heart under fluoroscopic visualisation, which record the cardiac activation sequence both at baseline and in response to predetermined pacing protocols. Quadripolar catheters are traditionally positioned in the high right atrium and right ventricular apex, and decapolar catheters across the right atrioventricular valve to record the His bundle and in the coronary sinus to record left-sided activation. A deflectable quadripolar mapping catheter is moved around the chamber of interest to select appropriate sites for radiofrequency ablation based on the character of local intracardiac electrograms. This technique remains the gold standard for the vast majority of EPS in children with a structurally normal heart, although recent technological advances which permit non-fluoroscopic imaging and annotation of specific anatomical sites may prove beneficial in select cases. The structural complexity of the heart in adults with surgically corrected congenital heart disease ensures inconsistent anatomical and surgical landmarks coupled with areas of low-voltage myocardium and scarring, making conventional electrophysiological mapping challenging and necessitating long procedure and fluoroscopy times. Three-dimensional mapping using either electroanatomic or non-contact mapping systems has significantly facilitated EPS and RFCA in these challenging patients.

EnSite NavX

EnSite NavX (St Jude Medical, St Paul, Minnesota, USA) is a three-dimensional visualisation, navigation and mapping technology, which uses standard intracardiac electrophysiological catheters to sense the electrical signals transmitted between three pairs of surface electrode patches. A three-dimensional geometry of the chamber of interest can be created and catheter location is superimposed onto this model (fig 2). Specific points of interest, local activation times and radiofrequency lesions can be annotated to the geometry to facilitate with diagnosis. No reports exist to date of the use of EnSite NavX specifically in the paediatric population, although the system has a number of attractions, specifically minimising the procedural radiation dose and allowing accurate electrophysiological and anatomical annotation during ablation of septal APs and AVNRT where inadvertent AV block is an important consideration.

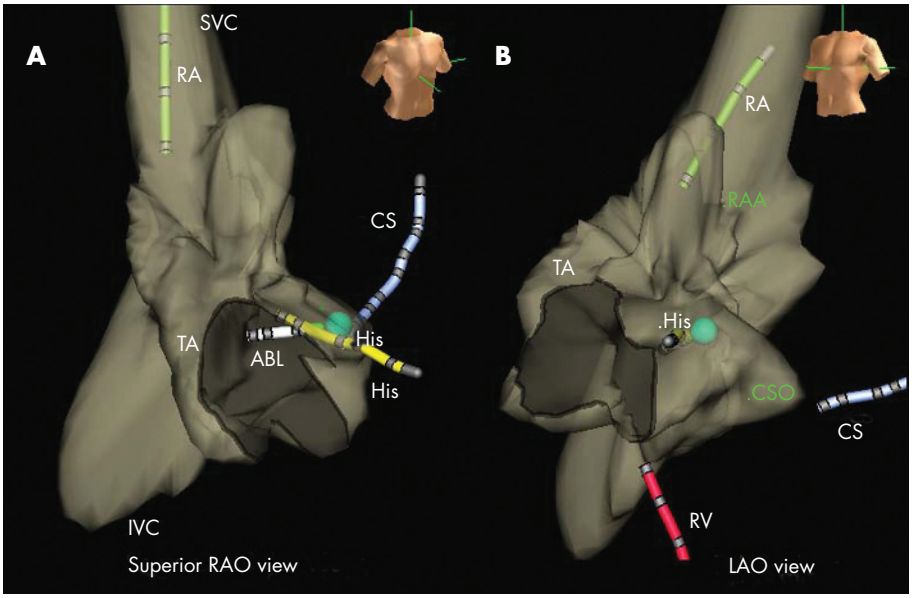

Figure 2 Radiofrequency ablation of an anteroseptal accessory pathway using EnSite NavX and cryoablation. A partial reconstruction of the right atrium is shown from (A) a right anterior oblique (RAO) view canted superiorly and (B) a left anterior oblique view (LAO), (torso indicates orientation). The tricuspid annulus (TA) has been cut away, and the positions of the right atrial appendage (RAA), coronary sinus os (CSO) and His bundle (His) annotated on the geometry. Catheters shown are right atrial (RA) in green, coronary sinus (CS) in blue, right ventricle (RV) in red and His bundle (HIS) in yellow. A cryocatheter (ABL) is shown in white approximating the HIS catheter and the position of the successful lesion is shown by a green dot. Image courtesy of Dr Simon Sporton, St Bartholomew's Hospital, London.

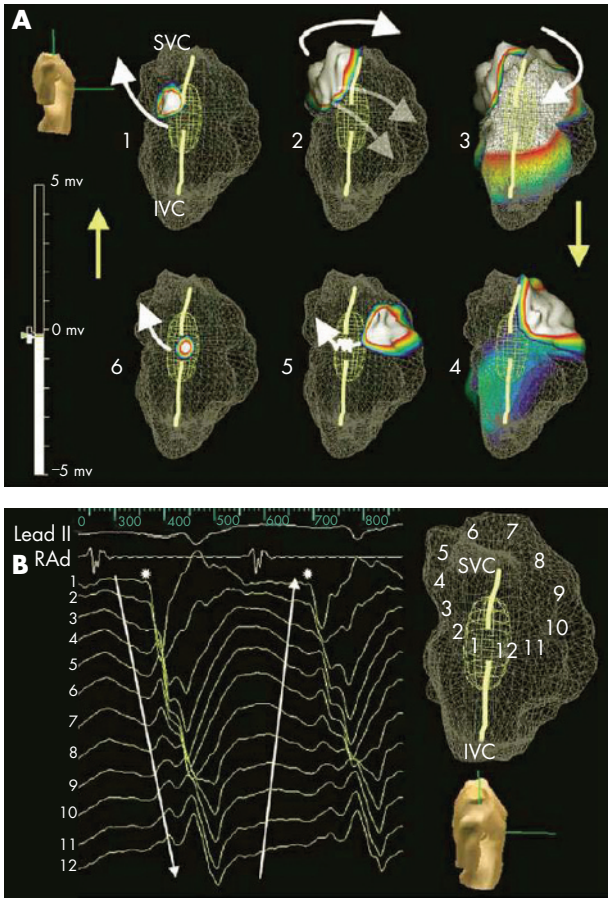

Figure 3 (A) Upper loop re-entry circuit with the superior vena cava and functional conduction block along the crista terminalis acting as the central obstacle. A computer generated reconstruction of the right atrial (RA) endocardium (''virtual endocardium'') with isopotential maps superimposed, recorded during atrial tachycardia using non-contact mapping (EnSite, St Jude Medical). The right atrium is seen from the right lateral view in all frames (torso indicates orientation) and the inferred position of the crista terminalis (CT) depicted by a yellow line. During sinus rhythm activation progressed across the upper part of the CT from anterior to posterior. Conduction block was demonstrated by programmed atrial stimulation from the medial aspect of the chamber at a cycle length of 300 ms. Isopotential maps generated from non-contact unipolar electrograms during arrhythmia demonstrate activation (white arrow) exiting from a gap in the CT and ascending the posterior wall superiorly towards the superior vena cava and medially to the atrial septum (frame 1). The wave front proceeds around to the anterior wall (faded white arrow), with the orifice of the superior vena cava (SVC) acting as the superior turning point, before returning to the gap in the crista terminalis (frames 2–4) which acts as a narrow corridor of slow conduction (zig-zag arrow) (frame 5), prior to re-emerging on the posterolateral wall to complete the circuit (frame 6). (B) Non-contact unipolar electrograms depicting atrial activation during tachycardia. Electrograms from lead II, the distal bipole of a right atrial catheter (RAd) and virtual unipolar electrograms recorded during the tachycardia shown in panel A. The right atrium is seen from a right superolateral aspect (torso indicates orientation) to expose the SVC orifice and demonstrate the upper turning point of the circuit. Electrograms recorded at 12 points (1–12) around the circuit are demonstrated. Activation onset at point 1 is depicted by QS morphology on the unipolar electrogram (*) and rotates in an anti-clockwise direction to point 12. The orifice of the SVC and the upper part of the CT appear to act as the central obstacle. Reprinted with permission from: Abrams D, Schilling R. Mechanisms and mapping of atrial arrhythmia in the modified Fontan circulation. *Heart Rhythm* 2005;**2**(10):138–44.

Electroanatomic mapping

Electroanatomic mapping (CARTO, Biosense Webster, Diamond Bar, California, USA) allows the creation of a three-dimensional reconstruction of any cardiac chamber by the serial acquisition of endocardial points. The navigation system locates the catheter tip in three dimensions within the heart to within 1 mm using a location magnet. The serial acquisition of endocardial points allows the construction of a three-dimensional geometry. Cardiac activation acquired from local electrogram timing in reference to a stable catheter is colour-coded and superimposed onto the anatomical reconstruction. Anatomical and/or surgical structures can be specifically identified and added to the geometry, as may electrophysiologically important points such as double potentials or areas of electrical silence (scar), which may be relevant to arrhythmia mechanism. The use of electroanatomic mapping in congenital heart disease has been found to be a positive predictive factor of a long-term favourable outcome, as defined by an improvement in a multi-scale index of clinically relevant arrhythmia activity at more than three months following ablation.[18] Electroanatomic mapping has provided powerful information on arrhythmia mechanism in congenital heart disease; however, because of the time taken to create individual activation maps, its use in the mapping of non-sustained arrhythmias or those associated with haemodynamic instability may be limited.

Non-contact mapping

Non-contact mapping (EnSite, St Jude Medical) uses a multi-electrode array (MEA) placed within the chamber of interest and the position of any catheter can be located by the system with respect to the MEA allowing the creation of a three-dimensional geometry (virtual endocardium). Raw far-field electrograms are detected on the endocardium by the 64 electrodes on the non-contact MEA, and are enhanced by applying an inverse solution to Laplace's equation, to reconstruct over 3000 unipolar electrograms superimposed onto the virtual endocardium to produce colour coded isopotential maps (fig 3). The EnSite system allows rapid analysis of endocardial activation and beat-to-beat variation, which has great benefits in non-sustained or haemodynamically unstable rhythms, and provides important information regarding the onset, termination and mechanism of arrhythmia. Potential disadvantages include the limited ability of unipolar electrograms to identify low-amplitude signals and areas of scar, and poor electrogram identification with increasing distance from the balloon—a factor which may be pertinent in grossly dilated chambers such as the right atrium following the atriopulmonary Fontan procedure. Clinical results with the non-contact system are encouraging, having been used successfully following the Fontan, atrial switch procedures and repair of atrioventricular septal defects, as well as to ablate right ventricular outflow tract tachycardia in the structurally normal heart. Preliminary results suggest a reduced fluoroscopy dose. A potential clinical limitation of the system is the size of the sheath required to deliver the MEA (9 French) although it has been used without complication in children of 17 kg.[19]

Cryoablation

Cryoablation (Cryocath, Kirkland, Canada) acts by removing heat from local tissues at varying amounts and rates, allowing

Invasive electrophysiology in adult congenital heart disease: key points

- Arrhythmia management in adult congenital heart disease requires a closely coordinated approach between electrophysiologist, cardiologist and surgeon
- Success of RFCA has become increasingly successful in adult congenital heart disease, related to a better understanding of arrhythmia mechanism coupled with technological advances, although remains inferior to many more simple substrates
- The approach to patients with atrial septal defect remains limited by a poor understanding of the arrhythmia mechanism and natural history in the long-term
- ECG, functional, electrophysiological and surgical risk factors may ultimately guide successful risk stratification of ventricular arrhythmias in older patients with at risk lesions

for both reversible and permanent effects to the underlying tissue. This technique has numerous attractive applications in paediatric arrhythmia management, specifically the ability to deliver a reversible test lesion in areas of potential atrioventricular block (cryomapping), enhanced catheter stability by adherence to the tissue during the application of energy (cryoadhesion), and the creation of smaller lesions in younger patients, thereby limiting damage to surrounding structures. To date cryoablation has been found to be a safe alternative to RFCA, although acute success and recurrence rates are somewhat below those currently achieved using radiofrequency energy.[20 21] This may reflect the smaller lesion size created by the cryocatheter or a learning curve in the use of this new technology.

Irrigated-tip catheter ablation systems

The inability to create transmural lesions in thickened atrial myocardium has been proposed as a possible reason for reduced success rates in congenital heart disease. Saline irrigation of the catheter tip allows greater radiofrequency power delivery and hence deeper and larger lesions. A retrospective analysis found the introduction of irrigated-tip catheters was associated with statistically higher success rates, recently confirmed in a prospective analysis by the same author.[18 22] Irrigated ablation appears to be safe in congenital heart disease and may potentially reduce the risk of coagulum and thrombus formation associated with standard RFCA.

CONCLUSIONS

Paediatric patients and those with congenital heart disease by their very nature represent unique challenges to the interventional electrophysiologist. While no specific studies have addressed the question, the results and complication rates of RFCA in children appear to be highly comparable with large adult series. The advent of both mapping and ablative technologies, coupled with a better understanding of arrhythmia mechanism, has improved ablative outcome of patients with congenital heart disease. An increasing awareness of the natural and surgical history of the electrophysiological substrate, and complimentary different techniques to modulate this, suggest continued improvement is a realistic future goal.

Additional references appear on the *Heart* website—http://www.heartjnl.com/supplemental

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