Cardiac Rhythm Management in Patients with Congenital Heart Disease

Standards of Care for patients undergoing catheter ablation and device implantation

Martin Lowe, on behalf of BHRS Council, February 2016

Contents:

1. INTRODUCTION

This document outlines the management of arrhythmias in patients with congenital heart disease (CHD). This coincides with the current review of CHD services and the recommendation that specialist services, including cardiac rhythm management, are centred at a smaller number of high volume institutions, to provide greater expertise in CHD management, and improve outcomes for patients. This model also provides for a multidisciplinary approach to arrhythmia care, with CHD specialists, radiologists and CHD surgeons able to input into patient management. The full range of arrhythmias seen in CHD patients is presented below, along with proposed standards for CHD arrhythmia specialists and CHD centres that perform interventional procedures.

2. BACKGROUND

CHD is the commonest congenital anomaly, with an incidence of nearly 1% of live births world-wide[1]. Surgical advances over the last few decades have resulted in a significant improvement in survival rate, with now >90% of children in the western world expected to reach adulthood[2]. Heart rhythm disorders in this population are prominent and the commonest cause for acute hospitalisation in adults with CHD[3].

Arrhythmias become more prevalent with age in CHD patients and are a leading cause of morbidity and mortality[4-7]. Tachy- and brady-arrhythmias are both common in this population and often co-exist. Supraventricular arrhythmias are most commonly due to intra-atrial reentry tachycardia (IART)[8-10], but may also be due to accessory pathway mediated tachycarda, AV node reentry, twin AV nodes or a focal mechanism[11-13]. The prevalence of atrial fibrillation also increases as the CHD population ages[8,14]. Ventricular arrhythmias are a leading cause of sudden death in CHD, although the absolute incidence is low at approximately 0.1% per year[4,5]. All types of bradyarrhythmias are seen, with sinus node disease, AV block and intra-atrial and His-Purkinje conduction delay common[9,15-18]. Systemic (left ventricular) failure may be seen not only in left sided congenital lesions but also in patients with Fallot's Tetralogy and Ebstein's malformation with adverse right / left ventricular interaction[8,19-21]. Ventricular dys-synchrony secondary to intrinsic conduction delay, or induced by pacing may also give rise to impaired ventricular function.

3. ROLE OF CATHETER ABLATION

Catheter ablation is being used increasingly to treat arrhythmias in the CHD population. The following general principles apply:

- Evaluation by a CHD specialist and when needed data presented for discussion at a multidisciplinary meeting, should be performed prior to electrophysiology intervention. CHD patients may have haemodynamic abnormalities that require correction prior to or instead of focused arrhythmia treatment.
- Pre-procedural assessment should include a detailed review of the patient's anatomy, operation and procedural reports in relation to cardiac surgical and percutaneous procedures, vascular surgical notes, and details of vascular access. All documentation in relation to the target arrhythmia (ECGs, ambulatory recordings, device electrograms, previous electrophysiology studies and ablation) should be reviewed.
- Ablation should be performed by operators experienced in advanced mapping techniques in CHD patients (see below and also BHRS ablation standards document).
- Invasive electrophysiology interventions (electrophysiology studies, ablation and device implantation) that require general anaesthesia should be performed with an anaesthetist experienced with the management of CHD patients.
- Access to cardiothoracic surgical emergency back-up should be available at all times.

ARRHYTHMIA SUBTYPE

Atrioventricular reentry tachycardia (AVRT)

Associated conditions

- Ebstein's anomaly
- Congenitally corrected transposition of the great arteries (ccTGA)

Ebstein's anomaly is commonly associated with the presence of accessory pathways, which are often multiple and may include both manifest and concealed pathways[22]. Fractionated, low amplitude electrograms at the site of the AV groove secondary to atrialisation of the right ventricle and the presence of atriofascicular pathways in some, makes ablation in these patients challenging. A 3D mapping system may be helpful to guide ablation in the presence of multiple accessory pathways.

Atrioventricular nodal reentry tachycardia (AVNRT)

Associated conditions

- Mustard and Senning repair for Transposition of the Great Arteries (TGA)
- AV canal repair

- Tricuspid atresia with Fontan circulation

AVNRT is seen less frequently than AVRT in the CHD population but has been associated with patients following Mustard repair for TGA, AV canal repair, and tricuspid atresia[23-27]. Anatomically the compact AV node may be displaced inferiorly, increasing the risk of complete heart block with ablation of the slow pathway.

Atrial tachyarrhythmias

Associated conditions

- Atrial septal and AV septal defect
- Ebstein's anomaly
- TGA with Mustard or Senning procedure
- Tetralogy of Fallot
- Left sided valvular abnormalities
- Single ventricular physiology with Fontan circulation
- Eisenmenger syndrome

The most common atrial arrhythmia in clinical practice is intra-atrial reentrant tachycardia (IART), due to reentry within the atrial musculature, around areas of scar, atriotomy sutures and surgical baffles, and often utilising the cavo-tricuspid isthmus[3,28]. Ectopic atrial tachycardias are also seen[29-33], often with an origin from low voltage sites within the atria, with micro-reentry being a likely mechanism[33].

Ablation strategies centre around cavotricuspid isthmus ablation, elimination of scar related tachycardia by creating linear lesions to connect scar to a fixed anatomical obstacle such as the vena cavae and in the case of ectopic tachycardia focal ablation. A 3D mapping system is best deployed in these patients because of the complexity and number of circuits in many[33-37].

In those patients who require surgical correction of haemodynamic abnormalities concomitant arrhythmia surgery (right atrial MAZE procedure, pulmonary vein isolation and / or left atrial compartmentalisation) may be utilised.

Atrial fibrillation

Associated conditions

- Atrial septal and AV septal defect
- Ebstein's anomaly
- TGA with Mustard or Senning procedure
- Tetralogy of Fallot
- Left sided valvular abnormalities
- Single ventricular physiology with Fontan circulation
- Eisenmenger syndrome

Atrial fibrillation is seen increasingly in the CHD population. Ablation strategies have centred around pulmonary vein isolation, complex fractionated potential ablation and linear ablation to compartmentalise the

left atrium. In patients with an atrial septal defect and atrial fibrillation amenable to ablation this should be performed prior to ASD closure[14]. Arrhythmia surgery for atrial fibrillation should be performed at the time of surgical ASD closure, in those unsuitable for a percutaneous septal closure device.

AV nodal ablation has been used infrequently in the CHD population because of the concern regarding the deleterious effects of ventricular pacing in patients with complex anatomy and systemic ventricular dysfunction.

Ventricular tachycardia

Associated conditions

- Fallot's Tetralogy
- TGA with Mustard or Senning repair
- CHD syndromes with systemic or right ventricular dysfunction

Sustained monomorphic ventricular tachycardia is seen in 0.1-0.2% of CHD patients per year[38]. The role of ventricular stimulation testing in the general CHD population is unclear. It has been used in the Fallot population[20 39], for risk stratification but in other CHD conditions such as TGA inducibility of ventricular tachycardia does not correlate with risk[7].

In patients with Tetralogy of Fallot, the right ventricular outflow tract is the usual source of macroreentrant VT[40-42]. Mapping studies have identified critical isthmuses bounded by the pulmonary valve, annular patch or ventricular incision site, and less commonly VSD patch[43]. The use of a 3D mapping system is helpful to identify each isthmus and guide successful ablation in these patients. Epicardial ablation may be performed at the time of surgery for those who require surgical pulmonary valve replacement during follow-up.

4. CARDIAC DEVICE IMPLANTATION

The following general principles apply to patients with CHD undergoing device implantation:

- Evaluation by a CHD specialist and when needed data presented for discussion at a multidisciplinary meeting, should be performed prior to device implantation. CHD patients may have haemodynamic abnormalities that require correction prior to lead deployment and an epicardial system may be more appropriate.
- Pre-procedural assessment should include a detailed review of the patient's anatomy, operating reports in relation to cardiac surgical, percutaneous procedures and vascular surgery. Knowledge of vascular access and the presence of venous abnormalities, e.g. persistent left SVC, is crucial to guide lead placement.

- Access to the ventricle may not be possible e.g. in patients post Fontan procedure for tricuspid atresia. Under these circumstances an epicardial device is indicated. Baffle patency is particularly important to assess in the Mustard / Senning population, as systemic venous pathway stenosis may prevent access to atrium and ventricle.
- The presence or absence of intracardiac shunts should be noted and hence the requirement for anticoagulation peri and post procedurally.
- Device implantation should be performed by an operator experienced in lead placement in patients with CHD (see requirements section below and also BHRS device standards document).
- Device implantation that requires general anaesthesia should be performed with an anaesthetist familiar with CHD patients.

BRADYARRHYTHMIAS AND PACEMAKERS

Sinus node disease

Associated conditions

- Senning or Mustard procedure for TGA
- Fontan procedure
- Glenn shunts
- Repaired Ebstein's anomaly
- Post-operative repair of ASD, TAPVD, and TOF

Sinus node disease is usually seen following cardiac surgery through injury to the SA node[9] [44]. The resulting junctional rhythm may be associated with an increased risk of atrial arrhythmias, AV valve regurgitation and thromboembolism.

Pacemaker implantation is recommended for sinus node dysfunction if there are symptoms related to bradycardia, either resting or with exercise, loss of AV synchrony, or worsening AV valve regurgitation leading to ventricular impairment. In those with tachy-brady syndrome pacing may allow more effective control of IART pharmacologically, although many of these patients will also be candidates for catheter ablation. All patients should undergo implantation of an atrial based pacemaker and in those with evidence of AV conduction disease in addition a dual chamber or cardiac resynchronisation device is appropriate.

AV block

Associated conditions

Native:

- Atrioventricular septal defect
- Congenitally corrected TGA

- Left atrial isomerism
- Anomalous left coronary artery arising from the pulmonary artery

Acquired:

- Post cardiac surgery

The AV node may be congenitally displaced and vulnerable to progressive disease in patients with AV septal defects, congenitally corrected TGA, and left atrial isomerism. In many the AV node is displaced inferiorly, rendering it susceptible to damage during surgery or catheter ablation[45]. Dual chamber or biventricular pacing is recommended in these patients in the presence of higher degree AV block.

AV block occurs in 1-3% of patients undergoing CHD surgery, with patients undergoing septal defect closure, LV outflow tract and left sided valve surgery at greatest risk. In those whose heart block is not transient, implantation of a dual chamber pacemaker or biventricular pacemaker is recommended[46].

ICD implantation

The incidence of sudden death in the CHD population overall is less than 0.1% per year[4-7], reflecting the benign nature of many CHD subtypes. There are, however, a number of conditions associated with an increase in arrhythmia-related mortality namely Fallot's tetralogy, TGA with Mustard or Senning procedure, Ebstein's anomaly and Eisenmenger syndrome. Sudden death in these patients is not exclusively secondary to ventricular arrhythmias, with AV block, and rapidly conducted atrial arrhythmias being other recognised mechanisms of haemodynamic deterioration, leading to cardiac arrest.

Implantation of an ICD for secondary prevention is indicated in those with CHD and aborted sudden cardiac death, or ventricular tachycardia with haemodynamic compromise. Primary prevention risk stratification is more problematic as outside the setting of conventional criteria the evidence in favour of implantation is based on small case series, rather than large randomised trials.

Patients with Fallot's tetralogy are at increased risk of ventricular arrhythmias[4 47-49]. Markers of increased risk include increased QRS duration, non-sustained VT, systemic ventricular dysfunction, history of ventriculotomies and prior palliative shunts. Inducibility of monomorphic VT has been shown to predict a five-fold increase in clinical VT or sudden cardiac death during follow-up[39]. Outside the setting of Fallot's tetralogy the role of invasive electrophysiology testing is much less clear.

Cardiac resynchronisation therapy (CRT)

CHD patients may benefit from CRT if their systemic ventricular ejection fraction is < 35%, QRS duration > 150ms, and NYHA status is 2-4 [50 51]. Multisite pacing of the right ventricle has been shown to improve right

ventricular function in selected patients with Fallot's tetralogy in short term studies, but long term benefit remains uncertain[50].

In patients with AV block who require long term pacing, biventricular pacing may help to protect against pacing induced systemic ventricular dysfunction[52].

Lead extraction

Lead extraction indications for patients with CHD are similar to those without CHD and include infection, arrhythmias related to a retained lead fragment, the need to create a passage for new lead(s) in patients with venous occlusion and non-functioning leads. Leads greater than 6 months old should be extracted with the availability of locking stylets and mechanical or laser sheaths, as in the general device population[53].

5. REQUIREMENTS FOR PERFORMING CATHETER ABLATION AND CARDIAC DEVICE IMPLANTATION IN CHD PATIENTS

(see also BHRS standards for interventional EP study and ablations in adults and implantation of CRM devices)

Cardiologists – training requirements

- Trainees should have achieved a CCT in Cardiology and completed at least 2 years EP sub-speciality training
- Advanced interventional EP training in CHD should be undertaken in a high volume centre performing at least 50 CHD CRM interventions per year
- Trainees should have been actively involved in 20 CHD ablations, with 10 as first operator
- Trainees should perform a minimum of 25 new pacemakers per year, and have gained additional experience in ICD and CRT implantation preferably with training specifically in CHD pts

Cardiologists – maintenance of competence

- There should be at least one and preferably two interventional electrophysiologists per centre with expertise in CHD
- Complex ablation should not be performed in single operator centres
- Interventional electrophysiologists should perform at least 50 catheter ablations per year, with a minimum of 25 complex cases. Of these, at least 20 cases per year should be in patients with CHD
- Interventional electrophysiologists should perform at least 35 new pacemaker implants per year, and a minimum of 30 complex device cases. Of these, at least 10 cases per year should be in patients with CHD.

- CHD ablation results should be submitted to NICOR, and electrophysiologists should audit their results and complications and patient reported outcome measures.

Centre requirements

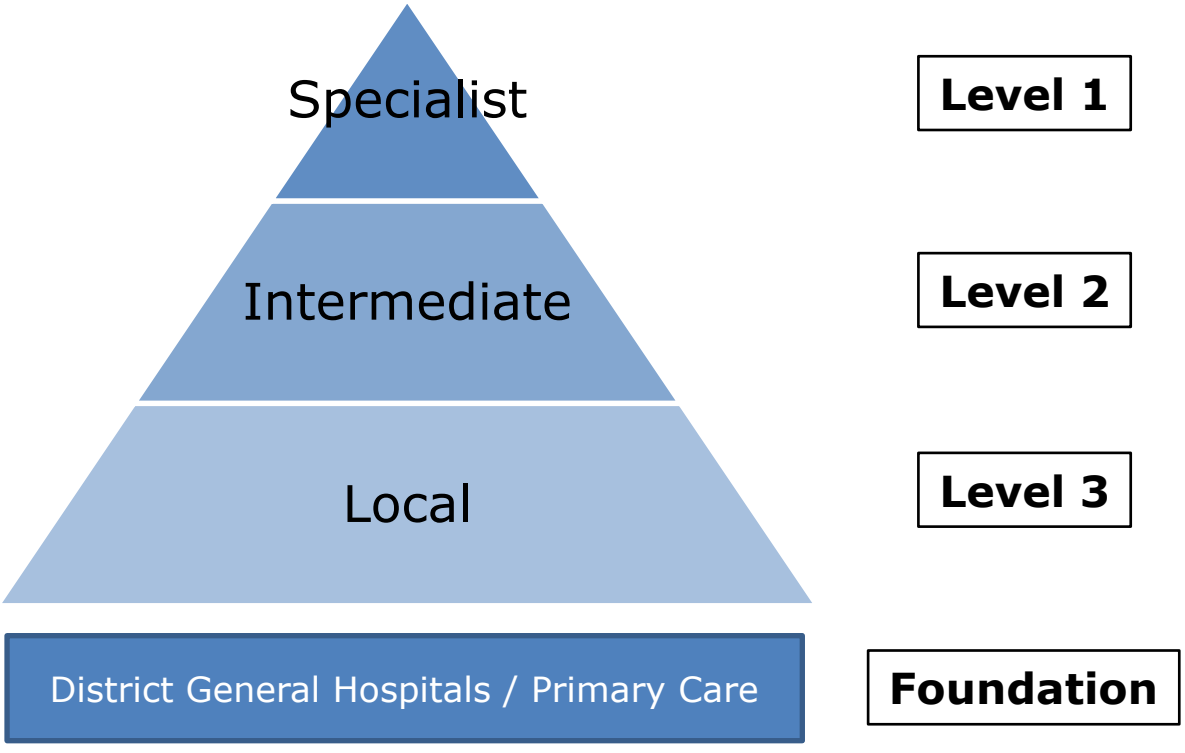
- In addition to the basic requirements for catheter ablation and device implantation, the following should be available for CHD patient procedures:
  - Electroanatomic mapping hardware and software.
  - Anaesthetist trained in cardiac anaesthesia in CHD patients.
  - Intensive therapy unit for high risk cases (Eisenmenger physiology, univentricular heart etc.).
  - Immediate access to cardiothoracic surgery.

6. PROPOSED CHD MODEL OF CARE

The current NHS review of Congenital Heart Disease services proposes a model of care based on 3 levels – Specialist, Intermediate and Local – on a congenital cardiac network basis.

Within each network, Level 1 is used to refer to the Specialist level, with onsite CHD surgery, Level 2 Intermediate and Level 3 Local level.

It has been proposed that CHD CRM intervention should in general be performed in a Level 1 unit, although 'simple' procedures could be carried out at a Level 2 hospital (see below). All patients should have their case reviewed by a CHD specialist before cardiac rhythm intervention.



'Complex' and 'simple' CRM procedures have yet to be fully defined. A guide is offered below:

Complex CRM procedures

Ablation
ASD repair with atriotomy and patch atrial flutter / IART
AVSD repair atrial flutter / IART
Ebstein's anomaly WPW, flutter or VT
Mustard / Senning for TGA with IART or VT
Fontan circulation IART, focal AT
Tetralogy of Fallot flutter IART VT
Unrepaired single ventricular physiology atrial flutter, atrial tachycardia

Devices
Senning or Mustard TGA with SA node disease
Fontan SA node disease
Repaired Ebstein's anomaly SA node disease or AV block (CS lead may be more appropriate than RV lead)
Post-operative Fallot SA node disease
Glenn shunts, TAPVD SA node disease
AV septal defect AV block
Congenitally corrected TGA AV block (CRT may be most appropriate)
Left atrial isomerism AV block
Fallot's tetralogy ICD insertion for VT

Simple CRM procedures

Ablation
CTI flutter ablation post ASD device closure
Isolated mild left sided valve abnormalities e.g. cleft MV and atrial flutter, atrial tachycardia, atrial fibrillation

Devices
Post-operative ASD SA node disease (although pacemaker may be required in the immediate post-operative period in the level 1 centre)
Post-operative AV block for isolated left sided valvular lesions (but, as above)

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