Electrophysiology assessment and radiofrequency ablation of arrhythmias in adult patients with congenital heart defects: the Christchurch experience

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Abstract

Introduction Adults with congenital heart disease (CHD) frequently have cardiac arrhythmias, many of which are best treated with radiofrequency ablation (RFA). We present our experience in this group.

Methods Retrospective chart based review of diagnosis, arrhythmia type, results of cardiac electrophysiological assessment, and procedural and long-term clinical success of radiofrequency ablation.

Results Forty-five patients were identified with CHD and arrhythmias undergoing RFA; including surgically repaired atrial septal defects (21), Ebstein’s anomaly (12), repaired transposition of the great arteries (3), repaired Tetralogy of Fallot (4), repaired ventricular septal defect (3), repaired coarctation (1) and unrepaired anomalous pulmonary venous anatomy (1).

Arrhythmias were atrial flutter (24), atrial fibrillation (1), atrial tachycardia (3), ativoventricular nodal re-entrant tachycardia (5), and ativoventricular re-entrant tachycardia (12).

Procedural success was ultimately obtained in 36 patients, with 6 having unsuccessful ablation and 3 an undetermined result.

Twelve patients required a repeat procedure. One patient required a third procedure and had insertion of permanent pacemaker and ativoventricular nodal ablation.

With follow-up (range 2–264 months) 31 patients (69%) remained in sinus rhythm, 9 have developed atrial fibrillation, 3 are in atrial flutter or atrial tachycardia, 1 patient reports ongoing palpitations with no documented arrhythmia and 1 patient has died.

Procedural complications were major venous access bleeding (2), transient heart block during slow pathway ablation with late complete heart block (1).

Conclusions The majority of arrhythmias in adult patients with congenital heart defects can be successfully treated with radiofrequency ablation at a relatively low risk.

Historically, many patients with congenital heart defects (CHD) died in childhood owing to the condition’s haemodynamic effect. In the last 50 years, however, the number of patients with CHD surviving to adulthood has increased dramatically due to the development of effective cardiac surgical correction. These patients have a high incidence of cardiac arrhythmias due to their congenital defect or as a
consequence of their cardiac surgery which predisposes to scar mediated re-entrant cardiac arrhythmias.

The management of these arrhythmias is a major issue especially as they may cause haemodynamic embarrassment due to residual cardiac compromise and medical management may be ineffective or poorly tolerated. The majority of these arrhythmias are suitable for cardiac electrophysiological assessment (EPS) and radiofrequency ablation (RFA). These patients provide special challenges during EPS and RFA due to abnormal anatomy; surgical baffles and scarring; and compromised vascular access. However, with advancement in the understanding of the mechanisms of these arrhythmias, and sophisticated mapping system technology it is becoming easier to map and correct these arrhythmias.

We report on the Christchurch experience with electrophysiology study and radiofrequency ablation for the diagnosis and treatment of adults with congenital heart defects, both with and without previous corrective cardiac surgery.

Methods

The study population consisted of all adults with a prior diagnosis of a congenital cardiac defect that underwent EPS and RFA at Christchurch hospital from 1992 to 2013. Patients were identified from our database, and all data was collected retrospectively using patient medical records and procedure reports. Details of arrhythmia diagnosis, congenital heart defect, previous surgery, electrophysiological diagnosis, and the procedural outcome of ablation when performed were collected.

Informed consent was obtained for each procedure. Patients were studied under conscious intravenous sedation with local anaesthetic at the site of vascular access.

Multiple diagnostic and ablating cardiac electrophysiological catheters were placed using fluoroscopy, and 3D mapping systems were incorporated for more complex cases from November 2001. Some patients underwent cardiac computerised tomography scanning before the procedure to provide venous and atrial anatomic details.

Tachycardias and pathways were mapped with conventional mapping techniques, entrainment mapping and, when available, 3D activation maps. Following the diagnostic assessment, radiofrequency ablation was performed targeting the pathways and critical components of re-entrant circuits.

Procedural success was determined by abolition of pathway conduction, block of critical isthmuses and non-inducibility of tachycardia. Clinical success was determined from the clinical follow-up reports.

Results

We identified 45 patients with CHD and arrhythmias undergoing EPS and RFA in our institution. The study population consisted of 46.5% males with ages ranging between 17–76 years.

The patients had the following underlying CHD: atrial septal defects with previous surgical repair (21); Ebstein’s anomaly (12); repaired transposition of the great arteries (3); repaired tetralogy of Fallot (4); repaired ventricular septal defect (3); repaired coarctation (1) and unrepaired anomalous pulmonary venous anatomy (1).

Clinical arrhythmias diagnosed prior to EPS included atrial flutter (24); atrial fibrillation (1); atrial tachycardia (3); atrioventricular nodal re-entrant tachycardia (5); and atrioventricular re-entrant tachycardia (12).

Of the 45 patients in the study group, procedural success was ultimately obtained in 36 patients; 6 had an unsuccessful ablation and 3 had an undetermined result. One patient underwent diagnostic EPS initially and was found to have multiple atrial
tachycardias. This patient subsequently received a permanent pacemaker and atrioventricular nodal ablation rather than undergoing an attempted ablation of the atrial tachycardias.

In our group, 12 patients required a repeat study. Of the 12 patients that re-presented with arrhythmias following an initial RFA attempt, 6 were found to have a different arrhythmia and 6 had the original arrhythmia. Of the six with the original arrhythmia 4 had previous acute success and 2 had previous failed ablations. One patient required a third procedure, and had an insertion of a permanent pacemaker and subsequent AV nodal ablation. The outcomes from the 13 repeat procedures in 12 patients were 9 successful, 1 partially successful, 2 undetermined, and 1 unsuccessful.

Overall, of the 45 patients, 37 (82%) had their arrhythmias successfully treated.

Atrial septal defect patients—In the 21 patients with atrial septal defects, all had previous surgical repair. Fifteen had atrial flutter, of which 3 had typical tricuspid annular flutter only and 11 had incisional atrial flutter, including 4 with figure of 8 circuits.

The majority of atypical and figure of 8 flutters were around the atriotomy scar on the lateral wall of the right atrium, with the figure of 8 circuits having the second limb through the tricuspid isthmus (Figure 1). For tricuspid annular-dependent circuits a line of ablation from the tricuspid annulus to the inferior vena cava was performed and block across the line was checked with differential pacing. Ablation for atypical circuits involved lines from scar tissue anchored to inferior vena cava, superior vena cava or tricuspid annulus.

Figure 1. CARTO3 maps of right atrium showing activation of figure of 8 atrial flutter

Note: Red shows early signals through to purple being latest signals. The red bar depicts where early signals meet late signals, indicating a reentrant circuit. Arrows show direction of wave fronts: around tricuspid annulus and around the superior vena cava. The wave fronts meet anteriorly and split into the two circuits at the point of the atriotomy scar (pink dots). Green dots show area of entrainment, black
dots show annular points and blue dots show points of double potentials. (a) Right anterior oblique projection (b) Left anterior oblique projection.

For patients with figure of 8 tachycardias, 2 or more line sets were required for success (Figure 2). Block across these lines was checked with differential pacing. Of the 15 patients with atrial septal defects and atrial flutter, procedural success was achieved in 11 patients, though 4 patients required a repeat procedure to achieve success. One patient with two previous ablation attempts was unable to be ablated and subsequently underwent AV nodal ablation and insertion of a permanent pacemaker.

Figure 2. CARTO3 maps of right atrium showing lines of ablation (red dots) performed to terminate the figure of 8 atrial flutter

Note: Lines were performed from tricuspid annulus to inferior vena cava, superior vena cava to lateral scar and lateral scar to tricuspid annulus. Grey dots depict the area of lateral atriotomy scar, purple and pink dots depicts interesting and fractionated signals, blue dots show double potentials and green dots show areas of entrainment. The yellow dot shows the point at which flutter terminated to sinus rhythm.

Two patients had multiple focal atrial tachycardias. One received AV nodal ablation and a pacemaker; the other had an undetermined result from their ablation procedure.

Three patients had atroventricular nodal re-entrant tachycardia treated with slow atrioventricular nodal pathway ablation, two had procedural and clinical success and 1 had an unsuccessful procedure.

The sole patient with permanent atrial fibrillation underwent an atrial fibrillation ablation with total pulmonary vein isolation and ablation to fractionated left atrial signals, roof line between right and left superior pulmonary veins, a mitral isthmus line and a tricuspid isthmus line.

With long-term follow-up (range 2–180 months) 11 patients remain in sinus rhythm, 2 patients have recurrent atrial flutter, 1 patient is deceased, 6 patients have developed atrial fibrillation and the patient with atrial fibrillation remains in chronic atrial fibrillation.
Ebstein’s anomaly patients—Twelve patients had Ebstein’s anomaly, of which two patients had previous surgery for Wolff Parkinson White syndrome. Clinical arrhythmias were atrioventricular tachycardia associated with ventricular pre-excitation (Wolff Parkinson White syndrome) in 11 patients; one of these patients also had typical atrial flutter; and one patient with multiple incisional atrial flutters from previous cardiac surgery.

In the 11 patients with pre-excitation, all pathways were right-sided and 4 patients had multiple pathways. Ten patients had procedural and clinical success, although 2 required a repeat procedure for recurrence of the same pathway. One patient had a failed procedure and has not represented for a repeat study.

The patient with typical atrial flutter underwent successful cavotruncuspid isthmus ablation in addition to successful pathway ablation.

One patient, with previous successful pathway ablation, returned with atypical atrial flutter. This was found to be an incisional flutter around the atriotomy scar and was successfully ablated.

The patient with multiple incisional atrial flutters underwent two separate ablations of multiple circuits. Whilst they had inducible arrhythmia at the end of the procedure, they have had overall clinical success.

With long-term follow-up (range 2–264 months), no patient who underwent pathway ablation has had pathway recurrence. Of the two patients receiving ablations for atypical flutter, one remains in sinus rhythm, the other has ongoing multiple atrial tachycardias.

Tetralogy of Fallot patients—Four patients had surgically-repaired tetralogy of Fallot. Two patients had atrial flutter; one typical, one incisional figure of 8; both had procedural success. One patient had a right atrial focal tachycardia with procedural success. However, they subsequently developed atrial flutter and were found to have both typical tricuspid flutter and incisional flutter. At repeat EPS and RFA the typical flutter was successfully ablated, with block achieved for the tricuspid line, but block across the lateral line was not achieved. One patient had atrioventricular re-entrant tachycardia with ventricular pre-excitation and procedural success.

With long-term follow-up (range 23–97 months) two patients remain in sinus rhythm, one patient has recurrent atrial flutter and atrial fibrillation and one patient has ongoing palpitations without documented arrhythmia.

Transposition of the great arteries patients—Three patients had Mustard atrial baffle repairs of transposition of the great arteries (Figure 3) and atrial flutter. One patient had cavotruncuspid isthmus flutter; one had micro re-entrant flutter in the systemic atria; and one had multiple atrial flutter circuits, including cavotruncuspid isthmus flutter and upper and lower-loop circuits in the systemic venous atria. In patients with cavotruncuspid isthmus-dependent flutter, isthmus block was achieved by ablation from the inferior vena cava to baffle, then completing the isthmus line via a retro-aortic approach to the tricuspid end of the isthmus.

Procedural and clinical success was obtained in all, though 1 patient required a second procedure for success.
With long-term follow-up (range 10–38 months) all patients currently remain in sinus rhythm.

**Figure 3. Computerised tomographic images of atria in a patient with Mustards repair of transposition of the great arteries**

![Computerised tomographic images of atria](image)

**Note:** Red shows the pulmonary venous atrium, which feeds into the right ventricle and pumps oxygenated blood to the body. Blue shows the systemic venous atrium. The inferior vena cava and superior vena cava have been redirected to feed into the left ventricle and pump deoxygenated blood to the lungs. (a) Right anterior oblique projections (b) Left anterior oblique projection (c) Posterior-anterior projection.

**Other patients**—These patients included ventricular septal defect (2); ventricular septal defect with coarctation (1); surgically repaired coarctation with bicuspid aortic valve (1); and unrepaired anomalous venous anatomy (1). The arrhythmias were atrioventricular nodal re-entrant tachycardia (2) with procedural and clinical success in both, though one patient required a repeat procedure. Three patients had atrial flutter; one typical, one both typical and atriotomy dependent flutter. The patient with anomalous venous anatomy had atypical cavotricuspid isthmus dependent flutter. All 3 patients had procedural and clinical success.

With long-term follow-up (range 13–180 months), 4 patients remain in sinus rhythm and one patient has developed atrial fibrillation.

**Procedural duration**—The procedure duration was 191±92 minutes (mean±standard deviation), (range 30–450 minutes). The screening time was 45±28 minutes (range 5–117 minutes). The radiation dose was 36±39 Gy.cm$^2$ (range 2–161 Gy.cm$^2$).

**Procedural complications**—Of the 45 patients undergoing a total of 58 procedures, three procedural complications occurred. Two patients had major venous access bleeding, with one requiring a blood transfusion. One patient had transient AV block during slow pathway ablation for AVNRT and subsequently re-presented with complete heart block and required a permanent pacemaker.

**Discussion**

Patients with congenital heart disease are now surviving into adulthood due to effective cardiac surgical correction.$^1$ These patients are at increased risk of cardiac
Arrhythmia due both to a higher incidence of pathway mediated arrhythmias, and also due to the occurrence of arrhythmias that are a consequence of the cardiac surgery.\(^2\)

Arrhythmia management can be especially challenging in this group of patients. The arrhythmias may be poorly tolerated, due to the associated residual cardiac defects but also the nature of the arrhythmias. In particular, atriotomy dependent atypical atrial flutters are difficult to manage with drug therapy, can become incessant and may be highly symptomatic.

These arrhythmias are suitable for treatment with curative intent by RFA\(^3\); however most of these procedures are performed in large centres, with large volumes of adults with congenital heart disease.

We present the experience of our small general arrhythmia service with a low volume of cases with congenital heart disease. In our series of adults with congenital heart disease and arrhythmias the majority of these patients treated with radiofrequency ablation had both procedural and long-term clinical success, with a low risk of complications. Procedure durations were often long, but the procedures were well tolerated.

There are a number of special considerations that need to be taken into account when performing ablations in adults with congenital heart disease.

In patients who have had an atriotomy for surgical correction of their congenital defect, incisional atrial flutter was the commonest mechanism of supraventricular tachycardia.

Incisional atrial flutter was the commonest arrhythmia in our series, occurring most commonly in patients that had undergone closure of an atrial septal defect, but also in patients with atrial baffle repairs for transposition of the great arteries, and surgical correction of accessory pathways in Ebstein’s anomaly. These atrial flutters are difficult to manage with medical therapy, are often incessant, and can usually be cured by ablation,\(^3\) making this the treatment of choice for most patients.

The atrial flutter circuit is determined by the previous surgery, and in the majority of cases, is dependent on areas of scaring and slow conduction on the anterior wall of the right atrium, in the area of the surgical atriotomy.\(^4\)–\(^6\) However, as reported in other series, in none of our patients were the atrial flutters confined to the area of septal repair.\(^6\)

Many patients also had simultaneous tricuspid annular atrial flutter resulting in a “dual loop figure”, also known as a figure of 8 circuit.\(^7\)\(^8\) (Figure 1). Patients with figure of 8 flutters required both a cavotricuspid isthmus ablation line and a second line between the atriotomy scar and adjacent anatomical boundary (Figure 2).

Furthermore, as these patients often have extensive right atrial scarring and multiple potential circuits, non-clinical tachycardias may be inducible following successful ablation of the clinical arrhythmia, but do not predict clinical failure as we saw in our patients.

Patients with atrial baffle repairs for transposition of the great arteries provide special challenges. The atrial anatomy is greatly distorted by the surgical repair, making access to the pulmonary venous atrium difficult (Figure 3). We observed both incisional and cavotricuspid dependent atrial flutter in our patients. Cavotricuspid
dependent atrial flutter is the most commonly observed arrhythmia in these patients and requires ablation in both the systemic venous atria from the inferior vena cava to the baffle, with the completion of the line from the baffle to the tricuspid valve in the pulmonary venous atria. We chose to perform this via a retro-aortic approach, though others have described accessing the pulmonary venous atria via a baffle puncture.

Ebstein’s anomaly is associated with the Wolff Parkinson White syndrome and pathway mediated atrioventricular tachycardia in 6 to 36% of cases. These pathways are almost exclusively right sided and approximately half the patients will have multiple pathways as seen in our series.

These pathways can be challenging to ablate, with lower procedural success rates and higher recurrence rates than seen with other pathways due to the distortion of the tricuspid annulus, difficulty in achieving catheter stability and the frequent occurrence of multiple pathways. We found that a number of patients with Ebstein’s anomaly required a second ablation attempt to achieve long-term clinical success.

Adults with congenital heart disease and arrhythmias require different approaches to other patients undergoing ablation. Preparation prior to the case will facilitate the procedure and outcomes. In particular, detailed knowledge of the patients’ anatomy and the details of the surgical procedure greatly facilitate a successful outcome.

Knowledge of the surgical incisions and atrial anatomy will guide the likely site of any incisional arrhythmia. In selected cases, prior CT scanning helps define the anatomy (Figure 3), whilst 3D mapping can facilitate defining atypical flutter circuits (Figure 1). We used CT scanning to define atrial anatomy in the 3 patients with prior transposition of the great arteries repair, and 3D mapping in all the recent cases of atrial flutter.

In conclusion, we report our results with the cardiac electrophysiological assessment and ablation of arrhythmias of adults with congenital heart disease. With appropriate preparation, and knowledge of the patients’ anatomy and surgical details the majority of these patients can undergo successful treatment of their arrhythmias.

Limitations—This report is retrospective and is therefore subject to the inherent biases in data recording and retrieval that arise from investigations of this nature. We report on a small number of patients in a single centre and our findings may not be able to be generalised to the wider population.

Conclusions

We report our results for ablation of arrhythmias in adults with congenital heart disease. The majority of these arrhythmias can be successfully treated at a relatively low risk.

Competing interests: Nil.

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