Can Pediatric Electrophysiologists Safely Perform Electrophysiology Studies on Adults with Congenital Heart Disease?

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ABSTRACT

Introduction: Adults with congenital heart disease (ACHD) are known to have arrhythmias. The purpose of this study was to review the safety and efficacy of a pediatric electrophysiologist performing electrophysiology studies (EPS) on ACHD.

Methods: All ACHD >18 years of age who underwent an EPS performed by a pediatric electrophysiologist between 1995 and 2004 were included. Patient records were reviewed for demographics, cardiac diagnosis, surgery, arrhythmia, and catheterization issues such as vascular access, fluoroscopy time, complications, procedure performed, and success rates.

Results: The study identified 70 patients who underwent 93 EPS: 42 with ventricular tachycardia (VT) and 28 with supraventricular tachycardia (SVT). With respect to safety, vascular access was obtained in 100% of patients, fluoroscopy time averaged 34.2 minutes, and complications occurred in 3 patients (4%) including permanent (1) or transient (1) atrio-ventricular node injury and coagulum on the catheter (1), but no deaths. With respect to efficacy, 30 patients had an attempted radiofrequency ablation that was successful in 21 of 28 (75%) with SVT and 1 of 2 (50%) with VT.

Conclusions: (1) ACHD can safely undergo an electrophysiology study performed by pediatric electrophysiologists, with low fluoroscopy times and few complications. (2) Success rates for ablation in this patient population approach 75%.

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BACKGROUND

Adults with congenital heart disease (ACHD) are prone to arrhythmias (1-4). These arrhythmias may be associated with their structural heart disease (e.g., Wolff-Parkinson-White syndrome with Ebstein's anomaly) or occur as a consequence of previous surgical procedures. Studies regarding the incidence of arrhythmias in ACHD have an occurrence rate as high as 66 % (1,3,4). Arrhythmias in this patient population can have a significant impact on quality of life, as well as morbidity and mortality (1,3,5-7).

When performing an electrophysiology study (EPS) on ACHD, it is important to have a complete understanding of patients' cardiac anatomy, previous surgical repairs, and catheterization procedures, as well as the venous and arterial anatomy. Knowledge of patients' anatomy and previous surgical procedures is important to understand access issues to the chamber involved in their arrhythmia, such as a Mustard with atrio-ventricular nodal re-entry tachycardia, where the atrio-ventricular node may be on the arterial side of the baffle. In addition, many ACHD have undergone a number of catheterizations and surgical procedures, and may have limited venous and arterial access due to either occlusion of vessels or abnormal vascular anatomy.

A majority of the arrhythmias that occur in ACHD require an EPS to either treat or assess risk. Previous reports of EPS on ACHD have reported reasonable success rates with limited complication rates (8,9). The majority of studies on ACHD have been limited in scope, reporting on a single cardiac anomaly (1,3). Additionally, it has not yet been established whether the EPS should be performed by a pediatric or adult electrophysiologist. The purpose of this study was to review the safety and efficacy of an EPS performed by pediatric electrophysiologists on adult congenital heart disease patients.

METHODS

A pediatric electrophysiologist (PEP) at one of two institutions (Yale-New Haven or Texas Children's Hospital) performed all electrophysiology procedures between January 1, 1995, and May 30, 2004. An IRB-approved, retrospective review of the medical records of all patients identified from the hospitals' databases was performed. Patients were included if they had congenital heart disease (CHD), and were >18 years of age at the time of EPS. Patient records were reviewed for information including underlying cardiac diagnosis, previous surgical procedure, form of arrhythmia, vascular access, procedure(s) performed, fluoroscopy time, success rates, and complications.

Safety of the EPS was assessed by considering each of the following: fluoroscopy time utilized, vascular access issues, and the occurrence of complications such as cardiac perforation, hemothorax, atrio-ventricular conduction abnormalities noted during or after EPS, infection, or myocardial infarction. Efficacy was based upon the procedure performed and its success rate.

During the study period, similar catheter types were utilized in all radiofrequency ablations and electrophysiology studies. The standard catheters utilized were ablation (6 or 7 French, 4 millimeter tipped), coronary sinus (6 French deflectable octapolar), His (6 French quadrapolar), and ventricle (5 or 6 French quadrapolar). For a ventricular stimulation protocol, three catheters were inserted, one in the atrium (6 French non-deflectable), and the His and ventricular catheter. All patients with ventricular tachycardia (VT) underwent programmed electrical stimulation utilizing two unique locations (right ventricular apex and outflow tract). The protocol for VT studies was giving a single, double, or triple ventricular extra stimuli after a paced drive train down to a cycle length of 180 milliseconds or ventricular muscle effective refractory period. The ventricular catheter was moved to a new location and the protocol was repeated. After this, the VT study was repeated on an isoproterenol drip.

This is a descriptive study of the safety and efficacy of EPS in ACHD. Normative data for these outcomes in this patient population are not available. Statistical testing was not performed since comparisons within or between groups were not made.

RESULTS

This study identified 70 ACHD, 36 male and 34 female, >18 years of age with a mean age of 26.2 + 7.2 years (range 18-47.6), who had an EPS. A total of 93 EPS were performed on these patients (average 1.3 per patient, range 1-3).

In the single ventricle patients, access to the ventricle was obtained retrograde from the femoral artery; otherwise, femoral venous access was considered the usual access route. In 90 procedures (96.8%), access was obtained utilizing femoral puncture(s). In the other three procedures (3.2%), notations were made in previous catheterization reports regarding difficult vascular access. These patients had vascular access obtained from the right internal jugular (1), right cephalic vein (1), or both (1).

The second safety measure was the fluoroscopy time. The average fluoroscopy time was 34.2 + 8.9 minutes (range 0.1-152). In three patients, each of whom had complex CHD, fluoroscopy time was greater than 120 minutes. Two of these patients had Ebstein's anomaly of the tricuspid valve, one of whom also had ventricular inversion with l-transposed great arteries and a ventricular septal defect. The third patient had tetralogy of Fallot and underwent attempted ablation of intra-atrial re-entry tachycardia prior to the advent of 3-D mapping.

The third marker for safety was complications of the procedure. This study identified three patients (4%) with complications: one developed transient atrio-ventricular nodal injury, a coagulum was noted on the tip of the ablation catheter in the second, and the third developed complete atrio-ventricular block during radiofrequency ablation. The patient who developed complete atrio-ventricular block had d-transposition of the great arteries and had undergone a prior Mustard procedure. An ablation was attempted to modify the slow pathway for atrio-ventricular node re-entry tachycardia using a retrograde catheter approach from the aorta when the complication was noted. No other complications were noted. Specifically, no infections, cardiac perforations, myocardial infarctions, or deaths occurred.

VT was the indication for performing a total of 64 EPS on 42 patients. Of these 42 patients, 36 had non-sustained VT and the remaining six sustained episodes (lasting >30 seconds). VT was detected on routine Holter monitoring in 36, four presented to the Emergency Department in VT, and two patients experienced syncope with exercise. VT was induced during EPS utilizing programmed electrical stimulation with and without isoproterenol in 26 (62%) of the 42 patients; 21 (58%) of those with VT on Holter, four (100%) from the emergency room, and one (50%) with syncope during exercise. An ablation was attempted in two of the Emergency Department patients with long-term success documented in one. The other 24 patients received an implantable cardioverter-defibrillator (ICD). Of these, 18 received transvenous

Table 1. Arrhythmia substrate by anatomic diagnosis.

Diagnosis	Ventricular tachycardia (n = 42)	Supraventricular tachycardia (n = 28)
Tetralogy of Fallot	10	3
d-transposition of the great arteries	9	5
l-transposition of the great arteries/ ventricular inversion	3	5
Single ventricles	7	8
Other	13*	7†

* Myocarditis (3), anomalous left coronary artery (2), partial (2) and complete (2) AV canal defect, Shone's complex (mitral stenosis, aortic stenosis, coarctation), ventricular non-compaction, ventricular septal defect, sub-aortic stenosis.

† Atrial septal defect (2), Ebstein's anomaly (2), dilated cardiomyopathy, Shone's complex, supra-valve aortic stenosis.

systems and six required epicardial patches. In each epicardial system, the generator was implanted in the patient's abdomen.

In the 16 patients who were studied more than once for VT, it was induced in two, both during their third EPS, resulting in subsequent implantation of an ICD. In patients with VT, the most common repaired CHD were 10 with tetralogy of Fallot (24%), 9 with d-transposition of great arteries (21%) and 7 with forms of single ventricle (17%) (Table 1).

Supraventricular tachycardia (SVT) was the indication for referral to the electrophysiology laboratory for 29 procedures in 28 patients. In those cases in which an ablation was attempted, 21 (68%) had immediate success. The eight unsuccessful ablations included five with intra-atrial reentry tachycardia, one with a para-Hissian accessory pathway, and one with atrio-ventricular nodal reentry. The failed atrio-ventricular node reentry occurred in a patient who had undergone a prior Mustard procedure and who subsequently returned to the EP lab, where the slow pathway was successfully modified. Successful implantation of the coronary sinus catheter was achieved in 17 of 18 patients (94%). The one unsuccessful implant was in a patient with l–transposition of the great arteries with ventricular inversion. A trans-septal procedure was successfully performed in nine patients without any complications. In patients with SVT, the most frequently encountered cardiac disorders were forms of single ventricle (29%), l-transposition of the great arteries with ventricular inversion (18%), and d-transposition of great arteries (18%) (Table 1).

DISCUSSION

Arrhythmias are common in adults with congenital heart disease. In this study, we identified 70 ACHD who required an EPS. The two reasons for performing an EPS study on this patient population were ventricular tachycardia, which was the indication in 60% of the patients, and supraventricular tachycardia, constituting the other 40%. This study was done not only to determine whether pediatric electrophysiologists are capable of safely and effectively performing EPS on ACHD, but also to determine if they should be the ones performing these procedures.

To the best of our knowledge, there are no reports published by either adult or pediatric electrophysiologists concerning safety measures with regard to EP procedures performed on ACHD. The first safety concern analyzed for this study was vascular access. The majority of ACHD have undergone a number of previous catheterizations and surgical interventions, and these patients may have spent a significant amount of time in the intensive care unit with invasive arterial and venous lines. All of these factors can lead to thrombosis of arteries and veins, limiting future vascular access. In addition, a significant number of ACHD have limited access to the desired cardiac chambers, such as Fontan patients with ventricular tachycardia and Mustard patients with atrio-ventricular node re-entry tachycardia. In these patients, access to the ventricle or slow pathway can be obtained retrograde from the artery or after a trans-baffle puncture. In this study, access was an issue in only 3% of the patients. These issues were known in two of the three patients, so no time was lost attempting femoral access. Therefore, planning for the number and location of catheters needed for the EP study can be done prior to arrival to the laboratory.

The second safety concern was patients' exposure to fluoroscopy during their EP study. In general, these patients have undergone a number of cardiac catheterizations in the past, which could increase their risk for the development of radiation toxicity and cancer. The limited fluoroscopy times encountered in this study again relay to a thorough knowledge of the anatomy, physiology, arrhythmia, and access to affected chamber(s) prior to the initiation of the study. Additional factors, such as the use of a three-dimensional mapping system, have helped to limit patients' exposure to fluoroscopy time.

The third, and most important, of the safety issues reviewed was that of complications. During this study, only three complications (3.2%) occurred in the EPS attempted. The observed complication rate in three large multi-center studies—two adult and one pediatric—was approximately 4% (ranging from 3.8–4.4%) (10–12). The number of complications from this study is slightly better then the overall safety of an EP study performed by either an adult or a PEP on traditional patients.

The safety of a PEP performing an EPS on ACHD is evident. The reason for the limited number of complications and safety issues is most likely multifactorial. Before an electrophysiology study is performed on ACHD, it is of utmost importance to have a complete understanding of the patient's underlying cardiac anatomy, previous surgical repairs, catheterization procedures, and venous and arterial anatomy. First, the chart must be reviewed with specific attention paid to all previous surgeries. It is also important to understand the nuances of the surgical procedure and location of suture lines, vascular folds, appendages, etc. After this, all previous catheter reports must be reviewed, looking for access difficulties to allow for the appropriate selection of catheters. Angiograms are reviewed to further delineate intra-cardiac anatomy and physiology, and to plan catheter course and access to the chamber involved in the arrhythmia. This is especially important to know prior to the beginning of the case, if the patient has a patch leak or fenestration or if a trans-patch puncture will be necessary.

The success of the ablation attempts was quite encouraging in this difficult group of patients. In those ACHD where an ablation was attempted for either VT or SVT, the success rates were 73%. These results compare favorably with previously published reports on individual congenital heart defects in this population, as well as those on pediatric patients with congenital heart disease (8,9). The unsuccessful ablations occurred in patients with difficult arrhythmia mechanisms such as intra-atrial reentry tachycardia, para-Hissian accessory pathway, or ventricular tachycardia around a surgical scar.

CONCLUSIONS

In conclusion, this study yields sufficient evidence to illustrate that electrophysiology studies can be performed on adults with congenital heart disease. In spite of their complex intra-cardiac anatomy, these patients can safely undergo electrophysiology studies with low fluoroscopy time, no vascular access problems, and few complications. In addition, the long-term success rates for ablation in this population of patients approaches 75%. We feel that pediatric electrophysiologists should be called upon to either perform or aid in all EP studies on this unique patient population.

REFERENCES

1. Puley G, Siu S, Connelly M, et al. Arrhythmia and survival in patients >18 years of age after the Mustard procedure for complete transposition of the great arteries. Am J Cardiol 1999;83:1080-1084.
2. Li W, Somerville J. Atrial flutter in grown-up congenital heart (GUCH) patients. Clinical characteristics of affected population. Int J Cardiol 2000;75:129-137.
3. Roos-Hesselink J, Perlroth MG, McGhie J, Spitaels S. Atrial arrhythmias in adults after repair of tetralogy of Fallot. Correlations with clinical, exercise, and echocardiographic findings. Circulation 1995;91:2214-2219.
4. Peters NS, Sommerville J. Arrhythmias after the Fontan procedure. Br Heart J 1992;68:199-204.
5. Gatzoulis MA, Balaji S, Webber SA, et al. Risk factors for arrhythmia and sudden cardiac death late after repair of tetralogy of Fallot: a multicentre study. Lancet 2000;356:975-981.
6. Nakazawa M, Shinohara T, Sasaki A, et al. Arrhythmia late after repair of tetralogy of fallot: a Japanese Multicenter Study. Circ J 2004;68:126-130.
7. van den Bosch AE, Roos-Hesselink JW, Van Domburg R, Bogers AJ, Simoons ML, Meijboom FJ. Long-term outcome and quality of life in adult patients after the Fontan operation. Am J Cardiol 2004;93:1141-1145.
8. Wiepert J, Noebauer C, Schrieber C, et al. Occurrence and management of atrial arrhythmia after long-term Fontan circulation. J Thorac Cardiovasc Surg 2004;127:457-464.
9. Triedman JK, Bergau DM, Saul JP, Epstein MR, Walsh EP. Efficacy of radiofrequency ablation for control of intraatrial reentrant tachycardia in patients with congenital heart disease. J Am Coll Cardiol 1997;30:1032-1038.
10. Van Hare GF, Javitz H, Carmelli D, et al. Prospective assessment after pediatric cardiac ablation: Demographics, medical profiles and initial outcomes. J Cardiovasc Electrophysiol 2004;15:759-770.
11. Scheinman MM. Catheter ablation for cardiac arrhythmias, personnel, and facilities. Pacing Clin Electrophysiol 1992;15:715-721.
12. Hindricks G. The Multicentre European Radiofrequency Survey (MERFS): complications of radiofrequency catheter ablation of arrhythmias. The Multicentre European Radiofrequency Survey (MERFS) investigators of the Working Group on Arrhythmias of the European Society of Cardiology. Eur Heart J 1993;14:1644-1653.