Cardiac Electrophysiologic Study in Children

Preliminary Report from Turkey

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SUMMARY

Eighteen children (10 females, 8 males; age range 5.5 to 17 years, median: 10 years) who suffered from recurrent syncope, drug refractory supraventricular tachycardia and atrioventricular block or bradycardia were evaluated by cardiac electrophysiologic study between August 1988–April 1990. During the study, basal intervals were measured and the conduction system and sinus node functions were investigated. In some patients the mechanism of the tachycardia was investigated and drug-electrophysiologic studies also performed.

Two of 6 cases with recurrent syncope had positive electrophysiologic findings and adequate treatment resolved the symptoms. The others have been followed. The mechanism of the tachycardia was determined in 5 cases with drug refractory supraventricular tachycardia and drug-electrophysiologic studies were done in all of them. The site of block was determined in 7 patients with atrioventricular block and pacemaker implantation was done in 5 patients.

The results emphasized the usefulness of cardiac electrophysiologic study in childhood arrhythmias.

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CARDIAC electrophysiologic studies have proved valuable in the diagnosis and treatment of cardiac rhythm disturbances.¹⁾ During the past 20 years, cardiac electrophysiologic studies have been used widely in adult patients. Some pediatric cardiologists are interested in this method and

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many reports have been published recently.²⁾⁻⁴⁾ In future, the method will be an excellent diagnostic and therapeutic tool in childhood arrhythmias.

No previous cardiac electrophysiologic studies have been carried out in children in Turkey. During the past 2 years, we have applied this method in children for various purposes, and determined the accuracy, safety and therapeutic usefulness of this technique.

MATERIALS AND METHODS

During a 21 month period ending April, 1990, 18 children and adolescents suffering from various rhythm disturbances had cardiac electrophysiologic studies (EPS) performed at Hacettepe Children's Hospital Pediatric Cardiology Unit. Their ages ranged from 5.5 years to 17 years (median: 10 years). Indications for EPS included: syncope in 6 cases, refractory supraventricular tachycardia in 5 and bradycardia with atrioventricular block in 7.

Diazepam (0.01 mg/kg) was administered intravenously to each patient 2 min before cardiac catheterization. All cardioactive drugs were stopped at least five half-lives before performing EPS.

Three to four bipolar, tripolar and quadripolar catheters were inserted percutaneously into one or both femoral veins and were positioned in one or more of the following areas: (1) high right atrium near the sinoatrial node; (2) across the tricuspid valve orifice to record the low septal right atrial and His depolarizations; (3) right ventricular apex; (4) left atrium via the patent foramen ovale.

In patients with syncope, baseline intervals, sinus node and atrioventricular node functions were determined, and atrial and ventricular stimulation protocols were each carried out using techniques previously reported by various centers. The patients with supraventricular tachycardia were investigated as above; in addition a drug-electrophysiologic study was carried out in all patients. In these patients, after the baseline EPS was carried out, a drug was chosen which could be used potentially as a chronic therapeutic agent or would assist in further delineating the mechanism of the arrhythmia. The drug was administered intravenously during EPS. If a chronic antiarrhythmia drug proved acutely successful in controlling the arrhythmia and had no severe adverse effects, it was then used chronically. If the first antiarrhythmia drug was unsuccessful, a second drug was then tested following a sufficient interval to allow clearance of the unsuccessful drug.

Atrioventricular block was investigated with baseline interval measurements. In some patients secondary pacemaker recovery time, sinus node

functions and ventriculoatrial conduction were determined.

RESULTS

Listed in Table I are the ages, diagnoses, study protocols and treatments of the 18 patients.

In cases with recurrent syncopal attacks, the diagnosis was made by noninvasive methods in 3 patients (Cases 1, 5, 6). The first case with exercise-induced ventricular tachycardia (Case 1) was diagnosed as idiopathic long QT syndrome after the measurement of the baseline interval recordings. In the other case with exercise-induced ventricular tachycardia (Fig. 1), EPS revealed no abnormalities and the ventricular stimulation protocol failed to generate tachycardia (Fig. 1). These 2 cases have been on oral pindolol and are symptom free. The other case with syncope, whose surface electrocardiogram showed sinus arrest, atrioventricular dissociation and bradycardia, was diagnosed as sick sinus syndrome by noninvasive methods. In EPS sick sinus syndrome was proven and atrioventricular and intra-ventricular conduction disturbances detected as well (Figs. 2A and 2B). A VVIR endocardial pacemaker was inserted. In the remaining 3 cases EPS were within normal limits. These cases have been followed at close intervals.

Listed in Table II are the results of drug-electrophysiologic study.

Fig. 1. Exercise induced ventricular tachycardia. Top panel shows bradycardia and nodal escape beats at rest; second panel was recorded after vigorous exercise and shows bigeminy ventricular extrasystoles; third panel demonstrates ventricular tachycardia at recovery; fourth panel was recorded after pindolol therapy during exercise testing.

Table I. Clinical and

Case	Age Sex	Cardiac disease	Diagnosis before EPS	Diagnosis after EPS	Exercise testing
1	9M	—	Exercise induced VT	Idiopathic long QT syndrome	+
2	15 F	—	Syncope	Normal	+
3	9 F	—	Syncope	Normal	+
4	12M	—	Syncope	Normal	+
5	12M	—	Exercise induced VT	Normal	+
6	11M	—	Sick sinus syndrome	Sick sinus syndrome, AV conduction disturbance	—
7	17 F	—	PSVT	PSVT, AV reentry, left accessory way	—
8	12 F	—	Bradycardia-tachycardia syndrome	Sick sinus syndrome, AV nodal reentry	—
9	9M	Dilated cardiomyopathy	Non-paroxysmal SVT	AV nodal reentry	—
10	6 F	—	PSVT	AV nodal reentry	—
11	17 F	Operated secundum ASD	PSVT	PAFF, intraventricular conduction disturbance, nodal pacemaker	—
12	9 F	Operated primum ASD	Complete AV block	Infrahisian block	—
13	5.5 F	Operated VSD	Complete AV block	AV nodal block	—
14	6M	Operated FT	Type II 2° AV block?	2° AV nodal block	—
15	8 F	—	Complete AV block	AV nodal block	—
16	9 F	—	AV dissociation	AV nodal block, retrograde His activation	—
17	7M	—	Sick sinus syndrome	Vagotonia, AV conduction disturbance	—
18	11M	Operated FT	Sinus bradycardia	2° infrahisian block	—

EPS=electrophysiologic study; VT=ventricular tachycardia; HRA=high right atrium; RVA=right ventricular apex; ERP=effective refractory period; AV=atrioventricular; VA=ventriculoatrial; PSVT=paroxysmal supraventricular tachycardia; SVT=supraventricular tachycardia

Study Data of Patients

Holter monitori- zation	Electrodes	Procedures	Treatment
+	HRA, His, RVA	Sinus functions; atrial, AV nodal and ven- tricular ERP; ventricular stimulation	Pindolol
+	HRA, His, RVA	Basal measurement, sinus functions; atrial, AV nodal and ventricular ERP; atrial and ventricular stimulation	—
+	HRA, His, RVA	Basal measurement, sinus functions; atrial, AV nodal and ventricular ERP; atrial and ventricular stimulation	—
+	HRA, His, RVA	Basal measurement, sinus functions; atrial, AV nodal and ventricular ERP; atrial and ventricular stimulation	—
+	HRA, His, RVA	Basal measurement, sinus functions; atrial, AV nodal and ventricular ERP; atrial and ventricular stimulation	Pindolol
+	HRA, His, RVA	Basal measurements, sinus functions, VA conduction	VVIR pacemaker
—	HRA, His, RVA, left atrium	Basal measurements, determining tachy- cardia mechanism, drug-electrophysio- logic study	Propafenone
+	HRA, His, RVA CS	Basal measurements, determining tachy- cardia mechanism, drug-electrophysio- logic study	VVIR pacemaker, propafenone
+	HRA, His, RVA, left atrium	Basal measurements, determining tachy- cardia mechanism, drug-electrophysio- logic study	Amiodarone
+	HRA, His, RVA	Basal measurements, drug-electrophysio- logic study	Verapamil
+	HRA, His, CS, RVA	Basal measurements, atrial and ventricular stimulation, drug-electrophysiologic study	Amiodarone
—	HRA, His, RVA	Basal measurements	—
—	HRA, His, RVA	Basal measurements	VVIR pacemaker
—	HRA, His, RVA	Basal measurements	VVIR pacemaker
—	HRA, His, RVA	Basal measurements, secondary PRT	—
—	HRA, His, RVA	Basal measurements, sinus functions	VVIR pacemaker
—	HRA, His, RVA	Basal measurements, sinus functions, AV conduction	VVIR pacemaker
—	HRA, His, RVA	Basal measurements	VVIR pacemaker

cardia; PAFF=paroxysmal atrial flutter-fibrillation; CS=coronary sinus; ASD=atrial septal defect; VSD=ventricular septal defect; FT=Fallot's tetralogy; PRT=pacemaker recovery time.

Fig. 2A. AH interval prolongation (AH=130 msec) obtained from His electrocardiogram. HRA=high right atrium; LRA=low right atrium; HIS=His electrocardiogram; V=right ventricular apex electrocardiogram; S=surface electrocardiogram; a=atrial depolarization; h=His bundle depolarization; v=ventricular depolarization.

Fig. 2B. Marked prolongation of sinus node recovery time detected from surface electrocardiogram. P=atrial depolarization; SNRT=sinus node recovery time; S=stimulus.

In Case 11 with drug refractory supraventricular tachycardia, EPS revealed paroxysmal atrial flutter-fibrillation, ectopic atrial rhythm and intraventricular conduction disturbances (Fig. 3). During the tachycardia, verapamil was administered intravenously and severe atrioventricular complete block developed which required temporary pacing. In 2 cases (Cases

Table II. Data of Drug-Electrophysiologic Study

Case	Arrhythmia	Previous drug(s)	Acute drug(s) given	Drug acutely successful?	Chronic drug(s)	Chronic drug successful?
7	PSVT	D, Prop	P	+	P	+
8	Sick sinus syndrome*	D	P	+	P**	+
9	Non-paroxysmal SVT	D, Prop, P	F, V	—	A	—
10	PSVT	D, P	V	+	V	+
11	PAFF	D, Prop	V	?***	A	+

D=digoxin; Prop=propranolol; P=propafenone; F=flecainide; V=verapamil; A=amiodarone; *=bradycardia-tachycardia syndrome; **=permanent pacemaker; ***=complete AV block due to verapamil; +=successful; —=unsuccessful. The other abbreviations are explained in Table I.

Fig. 3. Ectopic atrial rhythm. Atrial activation was recorded at proximal (PCS) and distal coronary sinus (DCS) first and followed by right atrium (HRA). The prolongation of the HV interval was noted.

7 and 10) with drug refractory supraventricular tachycardia, EPS was able to determine the tachycardia mechanism. During the tachycardia antiarrhythmia drugs were used to control the tachycardia successfully and stimulation protocols failed to provoke the arrhythmia. These patients have used the same drugs on a long term basis.

In 1 case with bradycardia-tachycardia syndrome in which the tachycardia attacks could not be controlled with digoxin, EPS showed normal sinus node functions and atrioventricular nodal reentry (Case 8) (Fig. 4). The tachycardia was stopped by repetitive extrastimuli and intravenous propafenone was administered. The stimulation attempts failed to induce

Fig. 4. Normal sinus node recovery time.

Fig. 5. 2° type II 2:1 AV block. Atrial activations were not followed by ventricular depolarizations at 2:1 rate. The site of block was infrahisian.

the tachyarrhythmia again. This case was treated with a permanent VVIR endocardial pacemaker and propafenone. The last case with dilated cardiomyopathy findings and non-paroxysmal supraventricular tachycardia (Case 9) underwent EPS. The study revealed atrioventricular nodal reentry which was stopped by programmed extrastimuli for brief periods. Various antiarrhythmia drugs (propafenone, verapamil, flecainide) failed to suppress the tachycardia. The case was treated with oral amiodarone for 6 months. The drug was unsuccessful and atrioventricular nodal ablation was planned.

In the cases with bradyarrhythmia (Cases 12–18), EPS revealed various

degrees of atrioventricular block and conduction disturbances (Table I). In 2 cases (Cases 14, 18) the surface electrocardiogram showed sinus bradycardia in contrast to EPS which determined atrioventricular block (Fig. 5). In the case with sinus arrest (Case 17) EPS showed extreme vagotony and atrioventricular conduction disturbance. In this group 5 cases were treated with permanent pacemaker. Two cases have been treated medically with no symptoms.

DISCUSSION

The cardiac electrophysiologic study is a very sensitive invasive method for precise diagnosis of cardiac arrhythmias and planning treatment. The method is used for various arrhythmias such as refractory supraventricular tachycardia, ventricular tachycardia, sinus node disease and atrioventricular block.⁵⁾ Patients with syncope and nondiagnostic noninvasive evaluation are also subject to this method.⁶⁾

Syncope is a frequent symptom in children. The etiology is related to extracardiac rather than cardiac disturbances. Cardiac syncope is a very serious symptom requiring prompt diagnosis and treatment. The evaluation of syncope is made noninvasively by surface electrocardiography, exercise testing, Holter-monitoring and neurologic examination. Invasive testing must also be done in some patients where noninvasive methods have failed to determine the etiology. Some reports have proven the accuracy of the cardiac electrophysiologic studies in syncope evaluation and treatment.^{6),7)} In contrast to this opinion some authors have suggested that a negative study was not a good prognostic sign.⁸⁾ Also there have been controversial ideas about positive electrophysiologic findings.⁹⁾ In the study which was reported by Bass et al, the patients with positive electrophysiologic findings were more prone to sudden death than patients with negative findings.⁹⁾ Kushner et al reported that the patients with negative electrophysiologic findings had a lower risk of sudden death, but had repetitive syncopal attacks and follow-up revealed new rhythm disturbances, formerly undetected.⁸⁾ It has been proposed that close follow-up is essential in cases with negative findings.⁹⁾ Beder et al reported that arrhythmias are an important cause of syncope in some children with an otherwise normal heart, when other causes have been excluded by noninvasive methods.¹⁰⁾

Our study revealed the cause of the syncope in 2 patients, 1 with idiopathic long QT syndrome and 1 with sick sinus syndrome. Three patients with syncope had normal electrophysiologic findings. In 1 patient with exercise-induced ventricular tachycardia the electrophysiologic study showed

vagotony but an aggressive stimulation protocol failed to induce the ventricular tachycardia. Our experience shows the usefulness of the cardiac electrophysiologic study in childhood syncope. The study must be done on every patient with negative noninvasive evaluation and recurrent attacks.

In determining the mechanism of supraventricular tachycardia the cardiac electrophysiologic study is very efficient.^{3),4)} This arrhythmia can be controlled with empiric antiarrhythmic therapy. Some patients may be refractory and show various dangerous symptoms during the tachycardia and must be evaluated by cardiac electrophysiologic study. Garson et al reported the clinical features and response to treatment of the childhood supraventricular tachycardias.¹¹⁾ In Gillette's study the mechanisms of supraventricular tachyarrhythmias were determined by cardiac electrophysiologic study.⁴⁾ In addition to this evaluation the drug-electrophysiology study must be done in these arrhythmias. In adults, drug-electrophysiology studies have proven valuable in predicting the long term efficacy of oral antiarrhythmia therapy. This is a very successful method for prevention of the proarrhythmia effects of antiarrhythmia drugs that cannot be managed safely in clinical conditions. Kugler et al¹²⁾ reported the results of drug-electrophysiologic studies in 40 children and chronic arrhythmia suppression was achieved in 37 of them. In our study group with tachycardia the mechanism of supraventricular tachycardia was determined and drug-electrophysiology studies were done in all patients (Tables I and II). Three patients have been controlled with various antiarrhythmia drugs which had been found to be effective in drug-electrophysiology studies (Cases 7, 8, 10). One case had life threatening complete AV block during the trial (Case 11).

The non-paroxysmal atrioventricular reentry tachycardia in 1 case (Case 9) could not be controlled during the study and follow-up treatment with amiodarone has not been effective either. The results of our study show that cardiac electrophysiologic study in supraventricular tachycardia is feasible, safe and effective. We also recommend that the study should be done in all children with drug refractory supraventricular tachycardia. Also proarrhythmic events can be controlled by this procedure.

Complete AV block is very common in children. It may be congenital or acquired. The treatment of this conduction defect can be planned by noninvasive methods.¹³⁾ However indications for permanent pacemaker implantation are changing and some reports show the usefulness of the cardiac electrophysiologic study.^{2),14)} The site of block, ventriculoatrial conduction and secondary pacemaker recovery time can be determined during the study.^{2),13)-15)} Also secondary ventricular arrhythmias are evaluated by this method.^{13),14)} In our study group the site of block was determined in

all patients (Table I). Secondary pacemaker recovery times were evaluated in some patients. Five patients were treated by permanent pacemaker. Two patients have been symptom-free during follow-up. The study shows the effectiveness of this method in diagnosing and treating conduction disturbances.

In conclusion, various childhood arrhythmias can be successfully evaluated by cardiac electrophysiologic study. The indications and results of the study must be carefully determined and noninvasive methods must be the first step.

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