Assessment and Follow-up of Pediatric Survivors of Sudden Cardiac Death

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In the young patient resuscitated from sudden cardiac arrest, the risks of recurrence are uncertain and so are the criteria defining therapeutic efficacy for the presumed cause of the initial event. In this study, we analyzed the outcome of 15 consecutive young patients, who were resuscitated from pulseless ventricular tachycardia or ventricular fibrillation and who were evaluated by comprehensive hemodynamic and electrophysiological testing. Patients were 11.2 ± 2.7 (mean ± SD) years old at the time of their event, and each was known to have some form of heart disease before sudden cardiac arrest. Ventricular tachycardia or fibrillation was inducible by programmed electrical stimulation in eight patients. Accessory atrioventricular connections, with antegrade effective refractory periods less than 220 msec, were identified in three patients. Sustained atrial flutter was the only arrhythmia inducible in two patients, and no arrhythmias were inducible in two other patients. Surgical or electrophysiological-guided medical therapy resulted in noninducibility of the ventricular arrhythmias in six patients. Surgical division of the accessory atrioventricular connections was performed in three patients, and arrhythmias were not inducible after operation. The four patients with atrial flutter or without defined arrhythmia were treated with an empiric therapy. During 37 ± 14 months of follow-up, the nine patients with documented noninducibility of a defined cause of sudden cardiac arrest were free of recurrent events. In contrast, during 18 ± 10 months of follow-up, two of the six patients with empiric therapy or persistent inducibility of ventricular tachycardia died suddenly, and three others had recurrence of ventricular tachycardia or fibrillation. Suppression of inducibility of a tachyarrhythmia consistent with sudden cardiac arrest was the only variable correlated with improved outcome (p = 0.03). Young survivors of sudden cardiac arrest are at high risk for recurrent life-threatening events, unless the efficacy of their therapy is proven by electrophysiological testing. Cardioverter-defibrillator therapy may improve the otherwise guarded prognosis of patients without proven therapy. (Circulation 1990;82:341–349)

Sudden cardiac death in the young patient is an uncommon event, and most studies of this problem have been limited to postmortem description of the unresuscitated victim.1–4 Although some form of heart disease is recognized in most young patients before sudden cardiac arrest,5,6 this event may be a primary manifestation of cardiovascular disease.7 Historically, there has been reluctance to attribute the death of a child to a sudden cardiac event, which may be reasonable, in part, for the infrequency of this diagnosis in the young.8

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Supported in part by the American Heart Association, Oregon Affiliate, grant 70172-1.

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Received July 27, 1989; revision accepted March 13, 1990.

In the past 10 years, we have witnessed the evolution of rapid response emergency care medical systems and an increase in the number of children surviving complex repairs of congenital heart defects.9–11 Thus, an increasing number of young survivors of sudden cardiac arrest may be identified. In this report, we present the results of hemodynamic and electrophysiological (EP) testing in 15 young survivors of sudden cardiac arrest in an attempt to define criteria predictive of a favorable outcome and to establish potential selection criteria for use of the cardioverter-defibrillator in this patient group.

See p 629

Methods

Study Patients

Between January 1984 and June 1988, 20 patients who ranged in age from 1.5 to 18 (9.6 ± 3.2) years were
referred to our institution after resuscitation from documented ventricular fibrillation (VF) or pulseless ventricular tachycardia (VT). Of the 20 patients, seven had congenital heart disease, each with prior surgical correction or palliation, seven had a primary electrical abnormality, and six had a cardiomyopathy. All patients required basic cardiopulmonary resuscitation in conjunction with direct-current defibrillation or cardioversion. Ultimately, fatal neurological or cardiovascular injuries were sustained by five patients; the 15 surviving patients, without irreversible injury, provide the basis of this study.

**Evaluation After Resuscitation**

After stabilization of cardiac rhythm and hemodynamics, patients were transferred to this institution. The initial evaluation consisted of a detailed history and physical examination and the establishment of continuous electrocardiographic (ECG) and hemodynamic monitoring. Serum electrolytes and levels of antiarrhythmic medications were determined on admission. An initial assessment of intracardiac anatomy and ventricular function was obtained with echocardiography.

Continuous ECG monitoring was performed in each patient for an interval of 7–21 days after resuscitation. After normalization of neurological status, hemodynamic cardiac catheterization and angiography were performed. Additional evaluation included thallium scintigraphy and coronary arteriography in six patients who had ECG evidence of myocardial ischemia and endomyocardial biopsy in three patients who had an idiopathic dilated cardiomyopathy. Radionuclide ventriculography was performed in all patients who had an echocardiographic left ventricular shortening fraction less than 28% or d-transposition of the great arteries.

**Electrophysiological Testing**

After completion of the hemodynamic evaluation, we performed programmed electrical stimulation in patients 1–3 weeks after the episode of aborted sudden cardiac arrest. Antiarrhythmic medications were discontinued for a minimum of five half-lives before baseline EP study. Patients were studied in the postabsorptive state and were sedated as required with meperidine and chlorpromazine. Multiple electrode catheters were inserted percutaneously through the femoral, antecubital, or subclavian veins and were positioned at the high right atrium, right atrioventricular (AV) junction to record the His bundle potential, right or left ventricular apex, and coronary sinus. Programmed stimulation was performed with an impulse duration of 2 msec and current strength delivered at twice diastolic threshold. Three surface ECG leads were recorded together with three to five intracardiac electrogams. The intracardiac electrograms were filtered at 30–250 Hz and were recorded at paper speeds of 100–250 mm/sec.

The EP study initially evaluated baseline intracardiac intervals and characteristics of AV conduction. Programmed stimulation began with atrial pacing. Paced cycle lengths of 750 to 300 msec were tested for 30 seconds, with a progressive 50-msec decrement in the pacing interval. The presence of an accessory AV connection was evaluated by determining the characteristics of AV conduction during atrial pacing, by mapping the sequence of retrograde atrial activation during ventricular stimulation, and by atrial pacing from multiple sites in the right atrium and coronary sinus. Patients with accessory AV connections underwent detailed mapping studies during orthodromic reciprocating tachycardia to localize insertions of the pathway(s) and the induction of atrial flutter or fibrillation to determine the shortest preexcited RR intervals.

Regardless of the results of the atrial pacing, programmed ventricular stimulation was performed in each patient. Ventricular pacing was performed at two right ventricular sites in each patient, except for three patients who underwent left ventricular stimulation because of prior Mustard repair of d-transposition of the great arteries. The pacing protocol consisted of the sequential addition of one and two ventricular extrastimuli to the intrinsic rhythm, followed by one to four extrastimuli coupled to an 8-beat drive cycle length of 600 or 500 msec, then 400 msec. Each extrastimulus was scanned through diastole until refractory, at which point it was coupled 50 msec later to the preceding train of stimuli. The endpoints of testing were the induction of sustained VT or VF or completion of the protocol. The baseline ventricular stimulation protocol was repeated on the following day, with the addition of isoproterenol (0.05 μg/kg/min) in patients who had no arrhythmia inducible at baseline or who experienced sudden cardiac arrest during exercise. Patients with no arrhythmia inducible at baseline, who were receiving an antiarrhythmic drug at the time of their event, were restudied after four to six doses of that medication.

**Evaluation of Therapeutic Efficacy**

All patients with an inducible sustained ventricular arrhythmia, atrial flutter, or atrial fibrillation with capability of a rapid ventricular response underwent serial EP testing to evaluate responses to medical and surgical therapies. Patients with inducible VT underwent serial electropharmacological testing to assess both the suppression of inducibility and proarrhythmic effects. This testing was performed in two patients with VT after surgical procedures and in two patients after pacemaker implantation. The following schedule of antiarrhythmic agents was used: procainamide 15–20 mg/kg i.v. during 60 minutes; procainamide i.v. combined with mexiletine 3–4 mg/kg/dose p.o. at 6-hour intervals; propranolol 0.2 mg/kg i.v. during 20 minutes in four divided doses; and flecainide 1.5–3 mg/kg/dose p.o. at 12-hour intervals. Four to six doses of the oral medications were administered before serial EP testing. Serum drug concentrations were determined at the time of serial EP testing.

The surgical division of accessory AV connections was evaluated by postoperative EP testing. The absence of preexcitation and noninducibility of orthodromic
reciprocating tachycardia was evaluated by programmed stimulation. Patients in whom an arrhythmia consistent with sudden cardiac arrest was not identified did not undergo repeat EP testing because of the lack of an objective standard by which to assess therapeutic efficacy. After the initiation of empiric antiarrhythmic therapy, persistent sinus rhythm was documented in each patient with no inducible tachyarrhythmia by continuous ambulatory monitoring.

Follow-up

All survivors have been evaluated at 6-month intervals since discharge. Any symptom of near syncope or palpitations has been examined in detail by 12-lead ECG, 24-hour Holter monitoring, exercise testing, and determination of serum drug levels. Pacemaker tachycardia detection algorithms and cardioverter-defibrillator event counters were interrogated when present.

Statistical Analysis

Continuous data are expressed as mean ± SD. The relation between the means of continuous variables was determined by the Student’s t test. Comparison between variables distributed into categories was based on Fisher’s exact test. Statistical significance was inferred at a probability value less than 0.05.

Definitions

Sudden cardiac arrest: the unexpected, abrupt cessation of breathing and circulation, caused by heart disease. Ventricular fibrillation: random, chaotic electrical activity of the ventricles, with no associated cardiac output. Sustained ventricular tachycardia: ventricular tachycardia more than 30 seconds in duration or requiring termination because of hemodynamic deterioration. Nonsustained ventricular tachycardia: ventricular tachycardia of 6 beats up to 30 seconds in duration. Suppression: noninducibility of a previously inducible tachyarrhythmia. Proarrhythmia: the development of a new arrhythmia during drug testing, neither spontaneous nor inducible at baseline EP testing.

Results

Clinical Features

The clinical features of the survivors of sudden cardiac arrest are listed in Table 1. Five patients had undergone correction of congenital heart defects with surgical repair 7.8 ± 2.1 years before their event. All patients had undergone postoperative cardiac catheterization from 2 to 6 years before their cardiac arrest, which documented normal pulmonary vascular resistance and no residual intracardiac shunt. Two patients with congenital heart defects had undergone pacemaker implantation before sudden cardiac arrest due to symptomatic bradycardia. Four patients had a cardiomyopathy: Three had a dilated cardiomyopathy, with an echocardiographic left ventricular shortening fraction of 12–22%, and one had a nonobstructive hypertrophic cardiomyopathy in association with Friedreich’s ataxia.

Six patients had a primary electrical abnormality. Three patients had overt preexcitation with up to two episodes of clinically recognized tachycardia per patient during the year before sudden cardiac arrest. Two patients had the long QT syndrome: One had a single episode of syncope 12 years before VF; the other had undergone a prior left cervicothoracic sympathectomy because of recurrent episodes of nonsustained VT. One patient had congenital com-

TABLE 1. Clinical Features

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yr)</th>
<th>Gender</th>
<th>Associated diagnosis</th>
<th>Prior symptoms</th>
<th>Medication at sudden cardiac arrest</th>
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<tr>
<td>1</td>
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<td>M</td>
<td>d-TGA</td>
<td>Syncope</td>
<td>Flecainide, digoxin</td>
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<td>d-TGA</td>
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</tr>
<tr>
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<td>Dose</td>
</tr>
<tr>
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<td>9</td>
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<td>Dose</td>
</tr>
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</tr>
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<td>F</td>
<td>DCM</td>
<td>Syncope</td>
<td>Dose</td>
</tr>
<tr>
<td>7</td>
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<td>F</td>
<td>DCM</td>
<td>Syncope</td>
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</tr>
<tr>
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<td>M</td>
<td>DCM</td>
<td>Syncope</td>
<td>Dose</td>
</tr>
<tr>
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</tr>
<tr>
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<td>Propranolol</td>
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<td>F</td>
<td>WPW</td>
<td>Palpitations</td>
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</tr>
<tr>
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<tr>
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<td>2</td>
<td>F</td>
<td>CCHB</td>
<td>Syncope</td>
<td>Propranolol</td>
</tr>
</tbody>
</table>

d-TGA, d-transposition of the great arteries; DORV, double outlet right ventricle; PAPVR, partial anomalous pulmonary venous return; DCM, dilated cardiomyopathy; HCM, hypertrophic cardiomyopathy; WPW, Wolff-Parkinson-White syndrome; LQTS, long QT syndrome; CCHB, congenital complete heart block.
plete AV block with a narrow QRS escape rhythm of 70 beats/min as an isolated cardiovascular abnormality. This patient had no arrhythmia or sudden prolonged pauses during prior Holter monitoring and a corrected QT interval of 0.45 seconds.

All patients were in sinus rhythm on the most recent ECG preceding sudden cardiac arrest, with the exception of the two patients with pacemakers and the one with congenital complete AV block. Within a 24-month interval before sudden cardiac arrest, the following tachyarrhythmias were recognized in individual patients: atrial flutter (two patients), nonsustained VT (two), atrial ectopic tachycardia (one), and reciprocating tachycardia (three). Six patients had a history of syncope, which was temporally related to exertion in two. In the other four patients, the etiology of syncope was unknown, although the circumstances were not suggestive of vasodepressor syncope. Eight patients were receiving antiarrhythmic medications, and five were receiving digoxin.

During postresuscitation ECG monitoring, four patients developed spontaneous sustained VT or VT degenerating into VF. In addition, one patient had atrial ectopic tachycardia, and two patients had episodes of nonsustained VT. Spontaneous atrial flutter was not observed, and no patient with preexcitation developed an arrhythmia.

**Hemodynamic Evaluation**

By radionuclide ventriculography, four patients had a left ventricular ejection fraction less than 35%. Each of these patients had a mean pulmonary capillary wedge pressure greater than 18 mm Hg at cardiac catheterization. Right ventricular ejection fraction was normal (range, 45–55%) in the three patients with transposition of the great arteries. Abnormalities of myocardial perfusion were identified by coronary arteriography in two patients: anomalous origin of the left coronary artery from the pulmonary artery in one, and diffuse hypoplasia of the left coronary artery in the other. One patient had a fixed perfusion defect of the ventricular septum by thallium scintigraphy. Angiographic evidence of arrhythmogenic right ventricular dysplasia was not identified in any patient.

**Electrophysiological Testing**

Three classes of tachyarrhythmias were induced during baseline EP testing: sustained atrial flutter, orthodromic reciprocating tachycardia, and VT or VF. The following abnormalities were also defined: corrected sinus node recovery time greater than 525 msec (four patients), AV dissociation with block proximal to His (one), enhanced AV node conduction (one), and HV interval greater than 55 msec (two).

Atrial flutter occurred in four patients during EP testing. In two patients with preexcitation, atrial flutter developed spontaneously after the induction of orthodromic reciprocating tachycardia. In two other patients with congenital heart defects, sustained atrial flutter, induced by rapid atrial pacing, was the only tachyarrhythmia identified. The induction of atrial flutter was accompanied by a hypotensive 1:1 ventricular response in one patient with enhanced AV node conduction. Atrial flutter was not inducible during EP testing in one other patient with a congenital heart defect who did have clinically documented episodes of atrial flutter.

Orthodromic reciprocating tachycardia was induced in the three patients with preexcitation. Mapping of the retrograde atrial activation sequence during tachycardia identified a posterior septal pathway in two patients and multiple pathways in one patient. Preexcited RR intervals less than 220 msec occurred after spontaneous atrial flutter in two patients and induced atrial fibrillation in one patient. Sustained ventricular arrhythmias were not inducible in these patients, and no patient in this series had a concealed accessory AV connection.

Sustained VT was induced in six patients at baseline EP testing. VT was induced with two extrastimuli in one patient, and with three extrastimuli in five patients. No arrhythmias were inducible at baseline EP testing in the two patients receiving quinidine at the time of sudden cardiac arrest; however, polymorphic VT was induced in both patients after the reintroduction of quinidine therapy. The cycle length and QRS configuration of induced VT corresponded to those of the spontaneous VT observed in four patients after resuscitation. The mean cycle length of the induced VT was 210±15 msec, and VT degenerated into VF in four patients. Characteristics of the clinical and inducible tachyarrhythmias are summarized in Table 2.

**Therapy**

**Patients with inducible VT.** Two of the eight patients with inducible VT underwent surgical procedures: a left internal mammary artery graft to an anomalous left coronary artery from the pulmonary artery in one patient and a left cervicothoracic sympathectomy in a patient with the long QT syndrome. Baseline preoperative and postoperative EP testing was performed in both patients. Pacemaker implantation was performed in two patients with VT before serial drug testing: one with congenital complete AV block and the other with sinus bradycardia after the Mustard procedure.

Four of six patients with VT inducible at baseline became noninducible during subsequent testing. The inducibility of VT in two patients only after the reintroduction of quinidine was interpreted to represent a proarhythmic effect. The two patients with persistent inducibility of VT were discharged on the antiarrhythmic regimen requiring the most aggressive stimulation protocol to induce VT.

**Patients without inducible VT.** In the three patients with preexcitation, surgical division of the accessory AV connections was performed in preference to serial drug testing. Postoperative EP testing in each patient
documented the division of the accessory AV connection and no other inducible tachyarrhythmia. The two patients with only inducible atrial flutter were treated with digoxin and verapamil and had sinus rhythm documented at the time of discharge. The suppression of inducibility of atrial flutter was not tested. The two survivors with no defined cause of sudden cardiac arrest were treated with β-adrenergic blockers; a cardioverter-defibrillator was also implanted in one of these patients. The inducible arrhythmias and initial therapies are summarized in Figure 1.

Patient Outcome

After resuscitation, patient follow-up has averaged 33.7±17 (range, 5–68) months. During this interval, two patients died suddenly, and three others had recurrence of pulseless VT or VF. These five events occurred at an average of 18.8 months after the first episode of sudden cardiac arrest. One patient with persistent inducibility of VT died suddenly 13 months after initial resuscitation, and the other had a documented recurrence of VF during a febrile illness. VF recurred in one patient with inducible atrial flutter during therapy with digoxin and verapamil. This patient had significant left ventricular dysfunction, and it is uncertain whether VF was precipitated by a ventricular arrhythmia not inducible during EP testing or atrial flutter with a rapid ventricular response. This patient received a cardioverter-defibrillator implant after the second episode of VF, which has since discharged in response to a syncopal arrhythmia (Figure 2).

Recurrent events have occurred in both patients with an undefined etiology of sudden cardiac arrest. One patient, with no arrhythmia identified by EP testing or Holter monitoring, died suddenly 5 months after resuscitation. The other patient, with a prior cardioverter-defibrillator implant, had tachycardia detection during near syncope and defibrillator discharge. Whether this was an appropriate response to a pathological tachycardia is speculative; however, this patient has remained on a β-blocker regimen and has been unable to attain a heart rate greater than 125 beats/min during exercise.

The six patients with VT inducible at baseline, who had arrhythmia noninducibility before discharge, have remained free of recurrent events. No arrhythmias have occurred in the three patients who had division

<table>
<thead>
<tr>
<th>Patient</th>
<th>Prior arrhythmia</th>
<th>Postresuscitation ECG</th>
<th>Arrhythmia</th>
<th>Inducible arrhythmia</th>
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<tr>
<td>1</td>
<td></td>
<td>Sinus B</td>
<td>NSVT</td>
<td>VT/VF</td>
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<td>AF</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>AF/sinus B</td>
<td>VVI paced</td>
<td>VT</td>
<td>VT*</td>
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<td>CCHB</td>
<td>VT</td>
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VT, ventricular tachycardia; B, bradycardia; NSVT, nonsustained ventricular tachycardia; AET, atrial ectopic tachycardia; AFib, atrial fibrillation; RT, orthodromic reciprocating tachycardia; WPW, Wolff-Parkinson-White; CCHB, congenital complete heart block; VF, ventricular fibrillation; AF, atrial flutter.

*Quinidine.

FIGURE 1. Flow chart of inducible tachyarrhythmias and initial therapies in the 15 survivors of sudden cardiac death (SCD). Patients with therapy proven to result in noninducibility by programmed stimulation are indicated by the boxes with shadows (six patients with ventricular tachycardia and three patients with reciprocating tachycardia). In two patients with ventricular tachycardia, inducibility was not suppressed, and in four patients, therapy was empiric, because of lack of definition of a specific etiology of sudden cardiac death. Pace, pacemaker; AICD, automatic implantable cardioverter-defibrillator.
of accessory AV connections. When compared with the outcome of patients with persistent inducibility or an undefined cause of sudden cardiac arrest, the outcome of these patients with documented noninducibility of an arrhythmia consistent with sudden cardiac arrest was significantly improved ($p=0.03$).

Holter monitoring has been obtained at 6–12-month intervals in all survivors. No significant arrhythmias have been detected by this method, except for nonsustained VT in the patient with persistent inducibility of VT who died suddenly. Exercise testing has also been performed in most survivors, and no arrhythmias have been determined by this method. The relation between arrhythmia inducibility at discharge and patient outcome is summarized in Figure 3.

Analysis of Factors Other Than Electrophysiological Testing

Multiple variables were evaluated in determining the risk of recurrent sudden cardiac arrest and thus for consideration for cardioverter-defibrillator implant. By univariate nonparametric analysis (Fisher's exact test), the ability to suppress inducibility of a defined tachyarrhythmia was the only factor associated with freedom from recurrence ($p=0.03$). Definition of an arrhythmia, without proven therapeutic efficacy, did not confer an improved prognosis. Sudden cardiac arrest, VT, or VF recurred in three of four patients with an ejection fraction less than 35% compared with two of 11 patients with normal systolic function ($p=0.2$). The variables of congenital heart disease, ischemia, age at the first episode of sudden cardiac arrest, and sinus or paced rhythm were not predictive of a recurrent event.

Discussion

The results of this study suggest that young patients resuscitated from sudden cardiac arrest remain at risk for recurrent events, except for patients who undergo division of an accessory AV connection. For the
The remainder of these patients, the identification and EP-guided treatment of a specific etiology of sudden cardiac arrest offer an improved prognosis. In the absence of this standard, cardioverter-defibrillator therapy appears appropriate. The risk of a recurrent event is particularly high for patients with persistent inducibility of VT or those in whom the mechanism of sudden cardiac arrest is undefined.

Evaluation of the Young Survivor of Sudden Cardiac Arrest

One objective of this study was to determine whether EP testing and guided therapy improved the long-term prognosis of young survivors of sudden cardiac arrest. Although serial EP testing has proven to be a superior method of prospective treatment selection for most adult survivors of sudden cardiac arrest,19,20 the validity of this testing has been questioned in patients with nonischemic substrates of heart disease, such as the long QT syndrome or idiopathic dilated cardiomyopathy.21,22 It is also uncertain whether arrhythmias constitute an independent risk factor for sudden cardiac arrest in the absence of ventricular dysfunction23 and whether atrial tachyarrhythmias can be implicated in the genesis of VF or sudden cardiac arrest.24

In this study, a tachyarrhythmia was inducible by programmed stimulation in 13 of 15 patients, and suppression of inducibility was correlated with an improved outcome. This finding suggests that in the absence of a defined reversible cause of sudden cardiac arrest, the induction of any sustained tachyarrhythmia represents a significant finding and provides an objective standard by which to assess therapeutic efficacy. Programmed stimulation protocols must evaluate atrial and ventricular tachyarrhythmias as potential etiologies of sudden cardiac arrest in young patients, because a rapid ventricular response to a supraventricular tachyarrhythmia may result in as marked a detrimental effect on the hemodynamic status of a patient as a primary ventricular arrhythmia. The possibility of atrial induction of ventricular arrhythmias also deserves consideration.25

The validity of arrhythmia suppression as a factor resulting in improved patient prognosis is based on the presumed significance of inducible tachyarrhythmias. Each patient in this study with clinically documented VT had the arrhythmia induced during programmed stimulation. The rapid cycle lengths of VT (210±15 msec) in these patients contrasts with the slower, sustained forms of VT that may not degenerate into VF.26 The four cases of VF during programmed stimulation in this series were each preceded by VT. Although the specificity of VF induction by programmed stimulation is uncertain, VF has not been induced in any other patients less than 18 years old studied in our laboratory.27

The inducibility of VT in two patients only after the reinitiation of quinidine is of concern. Similar results have been reported by Ruskin et al.28 Both patients in this series were receiving quinidine for primary atrial arrhythmias (atrial ectopic tachycardia and atrial flutter) at the time of sudden cardiac arrest. These findings emphasize the proarrhythmic potential of antiarrhythmic drugs and the need for caution and objective measurement in their use.

The exact implications of atrial flutter in relation to sudden cardiac arrest are uncertain. Atrial flutter with a rapid ventricular response represents one sequence that potentially could result in VF. One patient in this series, with recurrent atrial flutter and enhanced AV conduction, is known to have had the onset of this arrhythmia immediately preceding two episodes of sudden cardiac arrest. The Collaborative Study of Atrial Flutter in the Young has emphasized the increased mortality associated with this arrhythmia, although the mechanism of sudden cardiac arrest was not defined by this study.29 In the absence of other defined causes of sudden cardiac arrest, we now consider atrial flutter a plausible etiology, particularly in patients with congenital heart disease.

Of emphasis, evaluation of the young survivor of sudden cardiac arrest extends far beyond the EP study.30 Any potentially correctable factors, either anatomic, physiological, or hemodynamic, must be addressed before the initiation of programmed stimulation and arrhythmia suppression trials. However, after the correction of a presumed reversible factor, the noninducibility of VT or VF must be proven by EP testing. In the absence of a defined reversible cause of sudden cardiac arrest, therapy is guided by the ability to suppress inducible tachyarrhythmias.

Management of the Young Survivor of Sudden Cardiac Arrest

Five of the 15 survivors of sudden cardiac arrest in this series had a recurrent event after their initial resuscitation. However, these events were limited to patients with persistent inducibility of VT or those in whom empiric therapy was used, because of the lack of definition of a specific etiology of sudden cardiac arrest. Patients with a defined tachyarrhythmia and
documented noninducible status on specific therapy have been free of recurrence. These results suggest a positive predictive value of EP-guided therapy, when suppression of previously inducible tachyarrhythmias is documented (Figure 4). This conclusion is consistent with previous series of adult survivors of sudden cardiac arrest.31,32

Management of the young survivor of sudden cardiac arrest without a defined tachyarrhythmia remains uncertain. Potential transient abnormalities, such as ischemia, which produce an unstable electrophysiological substrate, require exclusion. Sudden cardiac arrest may also be the consequence of bradycardia or of nonreentrant tachyarrhythmia. Although the role of bradycardia in sudden cardiac arrest has been questioned,33 cardiac pacing is mandated in young survivors of sudden cardiac arrest with significant bradycardia or severe conduction system abnormalities.

Based on our results, we currently would recommend placement of a cardioverter-defibrillator in young survivors of sudden cardiac arrest with non-suppressible VT or an unknown cause of VF. These recommendations are based on 1) the apparent risk of recurrent sudden cardiac arrest, 2) the marked improvement in adult survival rates associated with use of this therapy,34,35 and 3) the apparent low risk of procedural complication associated with implant of this device in young patients weighing more than 25 kg.36 Definitive treatment of all tachyarrhythmias, including atrial flutter, is required. Recurrent atrial flutter in the young survivor of sudden cardiac arrest may be an indication for either AV node ablation or cardioverter-defibrillator implantation.

The suppression of inducible tachyarrhythmias must be proven in the selection of therapy for young survivors of sudden cardiac arrest. In the absence of this standard, implantation of the cardioverter-defibrillator is recommended, given the high risk of recurrent sudden cardiac arrest. Prospective comparative trials of the two therapies (drug versus cardioverter-defibrillator) and longer patient follow-up will be required for the validation of these recommendations.37–39

References


**KEY WORDS**  *arrhythmias* • electrophysiology • resuscitation • children
Assessment and follow-up of pediatric survivors of sudden cardiac death.

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_Circulation._ 1990;82:341-349
doi: 10.1161/01.CIR.82.2.341

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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