Sudden Cardiac Death in Children

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Sudden cardiac death is a relatively infrequent occurrence in the pediatric population. In a review of death certificates in Olmstead County, Minnesota, Driscoll and Edwards found 2.3% of 515 deaths in the age range of 1–22 years to be sudden and unexpected, yielding an incidence of 1.3 cases per 100,000 patient years. Of these sudden deaths, only one third were definitely cardiac related, and an additional one fourth were probably cardiac in origin. A similar study reviewing the cardiovascular registry of the United Hospitals, St. Paul, Minnesota, from 1960 to 1983 identified only 50 cases of sudden cardiac death in the age range of 7–35 years. In a retrospective international study of sudden cardiac death in patients 1–21 years old encompassing 42 centers in Europe and North America, only 254 cases were collected. In contrast, the incidence of sudden cardiac death in the adult population in the United States is on the order of 300,000 cases per year. One representative study of acute myocardial infarction and sudden death from coronary heart disease in adults 35–74 years old in Nashville, Tennessee, identified 258 sudden cardiac deaths in a single year. Although the epidemiology, mechanisms, and prevention of sudden cardiac death have been extensively studied in the adult population, the significantly lower incidence of life-threatening cardiac events in the pediatric population has made comparable studies of children and adolescents relatively difficult. In children and young adults with ventricular tachycardia, poor outcome has been associated with preexisting clinical or subclinical heart disease, particularly cardiomyopathy. Cardiac diagnoses most frequently associated with sudden cardiac death in children include acute myocarditis, dilated cardiomyopathy, hypertrophic cardiomyopathy, tetralogy of Fallot, Ebstein’s anomaly, aortic stenosis, mitral valve prolapse, previous surgery for congenital heart disease, complete heart block, Wolff-Parkinson-White syndrome (WPW), long QT syndrome (LQTS), pulmonary hypertension, and coronary abnormalities.

In this issue of Circulation, Silka et al report the results of a rigorous hemodynamic and electrophysiological protocol in 15 pediatric survivors of sudden cardiac death. Each of the subjects in this investigation had been recognized to have some form of heart disease before the presenting episode of sudden death. The authors conclude that the suppression of inducibility of a tachyarrhythmia that is consistent with sudden death is associated with improved outcome. This finding is consistent with previous studies in adults. It must be remembered, however, that the conditions that predispose to sudden cardiac death in the pediatric population are quite different from those of adult counterparts. There is an extremely low incidence of clinical or occult atherosclerotic heart disease in the younger age group. Accordingly, valuable perspective may be provided by a more detailed examination of the relation between sudden cardiac death in children and the underlying cardiac diseases that have been associated with it. In the population studied by Silka et al, six basic cardiac diagnoses were identified: WPW, previous surgery for congenital heart disease, dilated cardiomyopathy, LQTS, hypertrophic cardiomyopathy, and congenital complete heart block. Two patients also had anomalies of the coronary arteries.

Perhaps the most interesting group of patients in the study were those diagnosed as having WPW. Ventricular fibrillation is a well-known complication in patients with WPW. Three of the 15 patients studied by Silka et al had been known to have WPW before sudden death, and of these, two were taking propranolol. These data raise two important questions: What is the potential role of β-adrenergic blocking agents in the episodes of sudden cardiac death experienced by patients taking these drugs? Should patients such as these undergo invasive electrophysiological evaluation regardless of whether they have experienced syncope or sudden cardiac death? A causal relation between taking β-blockers and sudden death has not been established; in the past, the use of propranolol has been advocated to treat WPW in children in whom administration of digoxin is not considered safe. Nonetheless, there are theoretical concerns regarding this therapy. The major electrophysiological effect of β-blocking agents is to increase atrioventricular nodal refractoriness and to slow atrioventricular nodal conduction. Two other drugs with similar actions, digitalis and verapamil, have been shown to promote rapid ventric-
ular responses to atrial fibrillation in patients with WPW by shortening the refractory period of a rapidly conducting accessory pathway. While β-blockers do not shorten the refractory period of the accessory connection, they generally do not increase it. Thus, taking β-blockers may not reduce the risk of rapid atrioventricular conduction of atrial fibrillation. Moreover, during atrial fibrillation, retrograde concealed penetration of the accessory connection by impulses reaching the ventricle via the atrioventricular node may interrupt atrioventricular conduction over the accessory pathway.19 β-Blockers, by decreasing conduction via the atrioventricular node, could in theory interfere with retrograde penetration of the accessory connection and enhance antegrade conduction. It therefore seems prudent to refrain from the use of β-blocking agents in children with WPW unless the accessory connection is known to be incapable of rapid conduction.

The importance of knowing the electrophysiological characteristics of the accessory pathway has prompted some authors to advocate invasive electrophysiological testing in all children with WPW and frequent tachycardia or palpitations.20 The appropriate method of evaluation of the asymptomatic patient with WPW or the patient with infrequent episodes of tachycardia as their only manifestation of WPW is, however, controversial. Rinne et al21 have presented evidence favoring invasive electrophysiological testing in all patients with WPW and palpitations. Others argue that invasive electrophysiological testing of all symptomatic patients may not be warranted.22 To date, the small numbers of pediatric patients with WPW do not allow a definitive answer to this question in children. However, invasive electrophysiological testing in children with WPW furthers the collection of important data in the pediatric age group and has the potential to avert episodes of sudden cardiac death such as those reported by Silka et al.9

The diagnosis determined for the largest single group of patients in the study by Silka et al9 is postoperative congenital heart disease. In this area of research, perhaps the greatest body of information has been collected in patients who have undergone corrective surgery for tetralogy of Fallot, because of its well-recognized association with ventricular ectopy and sudden death.23,24 However, the advent of complete correction of this lesion in infancy may prove to be a factor in decreasing both ventricular ectopy and late mortality.25 The repair of two other lesions, d-transposition of the great arteries (d-TGA) and double-outlet right ventricle (DORV), has gained increasing recognition as a possible precursor to arrhythmia and sudden death in the pediatric population. A recent study of 89 patients late after repair of DORV26 found a very high incidence of sudden cardiac death (18% during a mean follow-up period of 82 months). Because greater age at the time of operation was a significant risk factor for sudden cardiac death in this study, it is possible that surgical advances allowing correction at earlier ages will cause a decrease in late mortality.

Of the various surgical corrections of d-TGA, the Mustard operation and its concomitant incidence of sudden cardiac death is best known. In two studies with follow-up of reasonably long duration (mean, >4 years), the incidence of sudden cardiac death was 4.4%27 and 2.9%.28 Holter analysis indicates that loss of sinus rhythm is frequent in patients who have undergone the Mustard operation.27–29 Postoperative electrophysiological testing reveals atrial flutter to be the most frequently induced arrhythmia.30 Although the correlation is not strong, atrial tachyarrhythmias have been the only predictor of sudden cardiac death in this group.27 There is evidence that the arterial switch procedure31 for the repair of d-TGA will reduce the incidence of electrophysiological abnormalities.32

The other large group of patients in the study by Silka et al9 had dilated cardiomyopathy. The prognostic significance of ventricular ectopy in dilated cardiomyopathy in adults remains controversial.33–35 Several investigators have found no correlation between inducible ventricular arrhythmias and subsequent sudden cardiac death in patients with this disease.36,37 In an interesting study of sudden cardiac death in hospitalized adults with severe heart failure, Luu et al38 found that 13 of 21 monitored deaths resulted from bradyarrhythmia or electromechanical dissociation, and only eight were the results of ventricular tachycardia or fibrillation. It is of interest that two of the patients with bradycardic death had a previous history of ventricular tachycardia. While an association between complex ventricular ectopy and sudden cardiac death has been suggested in children with dilated cardiomyopathy,39 no strong correlation has been identified. In addition, the value of electrophysiological testing in this younger patient population has not been investigated. Thus, it is not surprising that electrophysiological testing was not helpful in guiding therapy in the three patients with dilated cardiomyopathy in the study by Silka et al.

Similarly, the usefulness of invasive electrophysiological testing in survivors of sudden cardiac death with LQTS has not been determined.40 It has been recommended that patients with LQTS who are at high risk of sudden cardiac death because of factors such as history of syncope, congenital deafness, or a family history of sudden cardiac death be treated with a β-blocker at the maximum tolerated dosage. Those who experience syncope despite this therapy are candidates for left stellate ganglionectomy.41 One final diagnosis that must be considered in a discussion of sudden cardiac death in children is an anomaly of the coronary arteries. Anomalies of the coronary arteries have been identified in as many as 35% of children and young adults who die suddenly.1,42

In conclusion, a single solution to the problem of sudden cardiac death in children is likely to remain elusive. Due to the diversity of causes and the small numbers of patients who have been studied, no
definitive empirical approach to the treatment of the pediatric patient who has experienced or is at risk of experiencing sudden death from a cardiac cause is possible. It would seem prudent to carefully search for and treat underlying heart diseases such as WPW, LQTS, and coronary anomalies; to pursue newer and better surgical techniques for correcting congenital heart disease such as DORV, d-TGA, and tetralogy of Fallot; and to continue to search for the underlying mechanisms of sudden cardiac death in diseases such as dilated cardiomyopathy.

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