Sudden Cardiac Death in Adult Congenital Heart Disease

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KEYWORDS

• Congenital heart disease • Implantable cardioverter-defibrillator • Risk stratification
• Sudden cardiac death • Ventricular tachycardia

KEY POINTS

- Sudden cardiac death (SCD) is a leading cause of mortality in adults with congenital heart disease.
- SCD is predominantly due to malignant arrhythmias but other causes include myocardial infarction, heart failure, thromboemboli, and aneurysm rupture.
- Cardiac arrest survivors and patients with hemodynamically unstable ventricular tachycardia with no clearly identified reversible cause generally benefit from implantable cardioverter-defibrillator (ICD) therapy.
- Risk stratification for primary prevention ICDs remains challenging in this population of patients.
- Factors associated with SCD are relatively well defined in patients with tetralogy of Fallot, in contrast to those with systemic right ventricles or univentricular hearts.

INTRODUCTION

Remarkable medical and surgical breakthroughs in the care of children born with heart defects have resulted in a striking improvement in survival, particularly in younger age strata and in those with severe forms of congenital heart disease (CHD).[1] Consequently, most children now survive to adulthood, leading to a shift in population demographics with adults making up two-thirds of the entire CHD population.[2] Epidemiologic studies estimate that there are at least 3 million adults with CHD in North America and Europe.[3] Moreover, the prevalence of complex CHD in adults is steadily increasing.[2] Despite these medical achievements, patients with CHD cannot be considered cured even with successful repairs. They face a variety of long-term complications, including a broad spectrum of cardiac arrhythmias, and higher mortality compared with the general population.[4]

In this context, sudden cardiac death (SCD) is a leading cause of mortality. These catastrophic events often occur in otherwise relatively stable patients, typically in the third or fourth decade of life.[4–12] Identification of subjects deemed at high risk for SCD is a real challenge for the treating physician. These difficulties are reflected in a

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consensus document developed in partnership between the Pediatric and Congenital Electrophysiology Society and the Heart Rhythm Society, in which evidence-based recommendations were proposed.[13] This article focuses on mechanisms and risk factors for SCD in the adult with CHD and summarizes current recommendations regarding preventive strategies.

SCOPE OF THE PROBLEM

SCD is defined as death due to a cardiovascular cause that occurs within 1 hour of the onset of symptoms, or unwitnessed death in the absence of an obvious extracardiac condition as the proximate cause of death. The first reports that raised concerns about SCD following surgical repair of CHD were published more than 35 years ago.[14] The topic has since grown in interest, as reflected by numerous subsequent studies.

Contemporary studies on causes of late mortality in adults with CHD suggest that, after heart failure, SCD is the second most common cause of mortality, accounting for approximately 20% of all deaths (**Table 1**).[4–12] Trends indicate that the proportion of SCDs is on the decline.[11,12] Although the incidence of SCD in the CHD population at large is relatively low (<0.1% per year), identified subgroups at higher risk include tetralogy of Fallot, systemic right ventricle (ie, complete transposition of the great arteries [TGAs] with atrial switch or congenitally corrected TGA), left-sided outflow obstructive lesions (ie, aortic or subaortic stenosis, aortic coarctation), and Eisenmenger syndrome.[5,7,9,10] Furthermore, factors associated with SCD seem to differ among specific defects, suggesting that risk stratification is best performed on a lesion-by-lesion basis.[15]

MECHANISMS OF SUDDEN CARDIAC DEATH

Arrhythmias account for approximately 80% of all SCDs in the CHD population. Ventricular arrhythmias, both monomorphic and polymorphic (**Figs. 1** and **2**), are the most common events. However, atrial tachyarrhythmias with rapid (eg, 1:1) atrioventricular (AV) conduction degenerating into ventricular fibrillation (VF) have also been described, as have bradyarrhythmic deaths due to AV block. It is also important to bear in mind that other pathologic conditions, such as thromboembolism, myocardial infarction, aortic dissection, and aneurysm rupture, account for up to 20% of SCDs.[13] In contrast to other forms of heart disease that afflict young adults (eg, hypertrophic cardiomyopathy and arrhythmogenic right ventricular cardiomyopathy), exercise and physical activity have not been associated with SCD in adults with CHD,[16] with few exceptions such as TGA with atrial switch surgery.[17] Overall, less than 10% of SCDs in adults with CHD occur during exercise.[8,18] Most adults with CHD should be encouraged to exercise regularly, although the nature and intensity of exercise training should be tailored following a comprehensive evaluation.[16,19]

SECONDARY PREVENTION OF SUDDEN CARDIAC DEATH

There are no randomized trials for secondary prevention implantable cardioverter-defibrillators (ICDs) in subjects with CHD. In general, ICDs are

Table 1
Sudden cardiac death in adults with congenital heart disease

Authors	Years	Subjects	Deaths	SCD
Oeschlin et al,[6] 2000	1981–1996	2609	197	26%
Silka et al,[5] 1998	1958–1996	3589	176	23%
Verheugt et al,[7] 2010	2001–2009	6933	197	19%
Zomer et al,[8] 2012	2001–2010	8595	231	22%
Gallego et al,[9] 2012	1990–2010	936	50	44%
Koyak et al,[10] 2012	1970–2011	25,790	1189	19%
Diller et al,[4] 2015	1991–2013	6969	524	7%
Engelings et al,[11] 2016	2001–2015	2596	239	23%
Raissadati et al,[12] 2016	1953–2009	10,964	721	21%
Total or average	—	63,936	3524	19%

Adapted from Khairy P, Van Hare GF, Balaji S, et al. PACES/HRS expert consensus statement on the recognition and management of arrhythmias in adult congenital heart disease: Developed in partnership between the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Endorsed by the governing bodies of PACES, HRS, the American College of Cardiology (ACC), the American Heart Association (AHA), the European Heart Rhythm Association (EHRA), the Canadian Heart Rhythm Society (CHRS), and the International Society for Adult Congenital Heart Disease (ISACHD). Heart Rhythm 2014;11(10):e132; with permission.