CONTEMPORARY REVIEW

Sudden death in adult congenital heart disease: Risk stratification in 2014

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Arrhythmias and sudden death continue to plague a subset of adult patients with congenital heart disease. Despite investigative efforts spanning many decades, accurate identification of the high-risk patient remains challenging owing to a limited population size, relatively low event rate, and constantly evolving surgical approaches to the various malformations. Furthermore, until recently, most studies of the subject involved single-center formats with limited statistical power. The number of adult survivors has now reached a critical size where larger collaborative projects are beginning to generate more objective criteria for assessing risk. This review will provide an update on risk stratification for several of the major congenital cardiac lesions and outline the current recommendations for surveillance and management.

KEYWORDS Congenital heart disease; Sudden cardiac death; Ventricular tachycardia; Implantable cardioverter-defibrillator

ABBREVIATIONS CHD = congenital heart disease; **D-TGA** = D-looped ventricles and transposition of the great arteries; **EPS** = electrophysiology study; **ICD** = implantable cardioverter-defibrillator; **L-TGA** = L-looped ventricles and transposition of the great arteries; **LV** = left ventricle/ventricular; **PACES** = Pediatric and Congenital Electrophysiology Society; **SCD** = sudden cardiac death; **RV** = right ventricle/ventricular; **TGA** = transposition of the great arteries; **VT** = ventricular tachycardia

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Introduction

The late 1930s marked the beginning of a remarkable series of surgical advances for congenital heart disease (CHD) that permitted long-term survival for a unique group of patients who would otherwise have died during early childhood.[1] Improved longevity would eventually expose a number of unanticipated late complications, central among which were atrial and ventricular arrhythmias contributing to sudden cardiac death (SCD). Wolff et al[2] were the first to sound the alarm in 1972 when they published observations on disrupted conduction patterns and ventricular tachycardia (VT) associated with SCD in patients who have undergone repair of tetralogy of Fallot. Since then, the topic of late arrhythmias has received the attention of all cardiologists involved with the longitudinal care of this growing population.

Arrhythmias in CHD arise from the abnormal myocardial substrate caused by variable pressure/volume loads, cyanosis, and certain anatomic features specific to the individual structural lesion. The situation is further complicated by palliative or corrective surgery, creating myocardial scars that can function as conduction barriers and central obstacles for macroreentrant circuits. Of note, the most malignant arrhythmias in the CHD population typically do not become manifest until the third decade of life or beyond, suggesting that a period of degenerative remodeling also plays a role in their genesis. For this reason, SCD looms as a far greater concern once CHD patients reach adulthood than it did during childhood and adolescence.

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Magnitude of the problem

The number of adults with CHD living in North America is now estimated to exceed 1 million. Included in this group are some cases with relatively minor disease in whom arrhythmia risk is known to be low (eg, repaired atrial septal defect or ligated ductus arteriosus), but the majority can be classified as having moderate or severe malformations[3] with the potential for life-threatening rhythm disturbances. In several large series examining long-term outcomes after CHD surgery, sudden unexpected events (usually arrhythmic, but occasionally vascular or thrombotic) ultimately accounted for 20% or more of the total mortality in patients with complex lesions.[4–6]

Three important articles have provided detailed data on the overall SCD risk in large populations with CHD. Silka et al[7] examined outcomes for patients from the state of Oregon who underwent CHD surgery between 1958 and 1996 and identified sudden arrhythmic death in 30 individuals from a cohort of 3589. When event rate was adjusted

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according to specific lesion type and follow-up duration, the incidence appeared highest for adult patients with transposition of the great arteries (TGA) and a systemic right ventricle (RV) after atrial baffling operations, followed by those with left ventricular (LV) outflow obstruction (aortic stenosis or coarctation of the aorta), followed by those with tetralogy of Fallot. A different study format was used by Koyak et al[8] in their multinational case-control study of more than 25,000 adult CHD patients that included surgical follow-up as well as natural history of nonoperable cases. They identified 171 cases of SCD due to arrhythmias within this large group. Denominators were not reported for specific lesion type to allow calculation of incidence, but the conditions with the highest number of SCD events included Eisenmenger syndrome, TGA with a systemic RV, and tetralogy of Fallot. More recently, Gallego et al[9] reported their single center experience of more than 3000 adult patients with CHD, in which patients with TGA and a systemic RV were again the highest risk group, followed by those with a single ventricle. These studies are among the few that have looked at SCD among CHD patients in a global sense, and all 3 arrived at nearly identical conclusions: (1) malignant arrhythmias will occur in 1% of all patients with some form of CHD over a mean follow-up period of 10 years, (2) these events will be largely concentrated in adult aged patients with complicated hemodynamic lesions for whom the SCD risk may reach 10% per decade of follow-up, and (3) abnormal "systemic" ventricular function (whether this involves an anatomic RV or LV) is among the strongest predictors of malignant arrhythmias.

Most other studies of the SCD issue in CHD tend to focus on a specific lesion, with an understandable predilection to choose a common malformation with an effective surgical solution and a large number of patients surviving into middle age. Hence, tetralogy of Fallot has been studied more extensively than any other condition, and so the mechanisms for SCD and its risk factors have been worked out reasonably well. Knowledge is less developed for other forms of CHD, such as TGA or single ventricle, either because the malformation is less common or because survival into adulthood is limited. The best available data suggest that it is hazardous to assume that lessons learned from tetralogy of Fallot are applicable to other forms of CHD. Only recently has multicenter attention been directed to SCD in alternate lesions, and risk stratification for such patients appears to differ in some important ways. It is useful, therefore, to evaluate arrhythmia risk in adult CHD on a lesion-by-lesion basis.

Risk assessment in specific CHD lesions

Tetralogy of Fallot

The mechanism of SCD in tetralogy of Fallot has been under investigation for more than 4 decades. Early on there was concern that atrioventricular block accounted for these events,[2] but it quickly became evident that VT was the culprit in most cases.[10] It is now understood that the intrinsic anatomy of the RV in tetralogy involves structural features that can potentially support macroreentry circuits near the outflow tract[11–13] and that traditional surgical repair might reinforce this potential (Figure 1). Patients with tetralogy can also develop RV and LV dysfunction from hemodynamic stress, putting them at additional risk of more disorganized polymorphic VT and ventricular fibrillation, similar to arrhythmias seen in any other form of dilated cardiomyopathy. Beyond the VT risk, these patients also carry a heavy burden of atrial macroreentrant tachycardias[14,15] involving the cavotricuspid isthmus and/or atriotomy scars (Figure 2). Atrial tachycardia, when conducted rapidly in a patient with tetralogy and depressed ventricular function, can contribute to the SCD risk in some individuals.

More than 100 studies have been published examining SCD in repaired tetralogy since the mid-1970s. The largest of these permit estimation of the SCD risk in a given cohort with tetralogy of approximately 2% per decade of follow-up.[7,16–19] This figure, however, is based on study groups that include both pediatric and adult subjects with variable follow-up duration. If attention is directed exclusively to adult patients 25 years or more after surgical repair, the SCD risk rises to the range of 6%–10% per decade of follow-up.[17,19]

The search for risk factors predicting VT and SCD in patients with tetralogy has generated an extensive catalog that includes patient age, surgical timing/technique, measures of hemodynamic status, electrocardiographic findings, noninvasive rhythm monitoring, and invasive electrophysiologic evaluation. The sheer length of the list reflects the challenge of working with a limited population size and low event rate, but perhaps the biggest obstacle to a strong

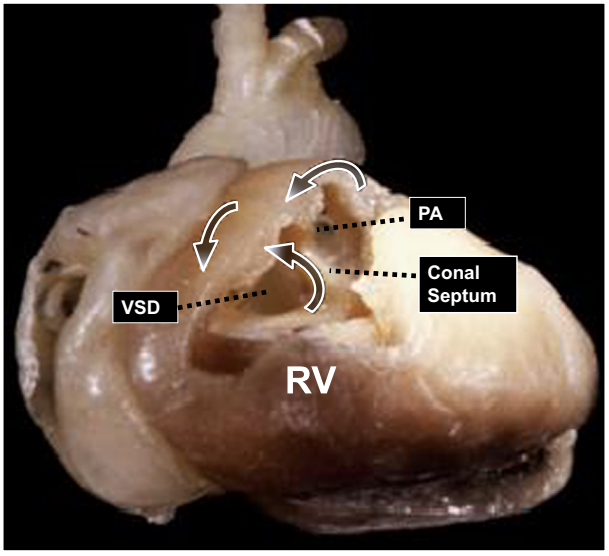

Figure 1 Pathologic specimen from a young patient with unrepaired tetralogy of Fallot demonstrating the intrinsic anatomic features that can contribute to macroreentrant ventricular tachycardia. A portion of the anterior right ventricle (RV) has been removed to expose the ventricular septal defect (VSD), the stenotic pulmonary outflow tract (PA), and the narrow band of muscle (Conal Septum) running between the VSD and the PA. The 3 curved arrows mark locations that could function as protected corridors supporting macroreentry after surgical repair.

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