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SUDDEN CARDIAC DEATH AND CONGENITAL HEART DISEASE (CHD)

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CHD remains an important cause of sudden cardiac death to be recognized at adult autopsy. Bicuspid aortic valve and anomalous coronary anomalies are the most common malformations, comprising 36.9% and 26.2% of cases, respectively [1]. However, a wide spectrum of simple to complex malformations can be seen, with or without prior surgery, and over a wide age spectrum. CHD is now "grown-up" and will likely be diagnosed by forensic pathologists with increased frequency in the future [2].

The risk of sudden cardiac death (SCD) rises with the complexity of the cardiac abnormality and the surgery undertaken. In infancy mortality used to be high but now there is prolonged survival to adulthood and less operative mortality. Obviously complex lesions have a higher mortality. In a recent review forty-six patients had 2 functional ventricles and 23 had received palliation for a single-functional ventricle. Patients with a single ventricle died at a younger age than patients in the biventricular group. Thrombosis was the most common cause (61%) of death in the single-ventricle group. Arrhythmia or presumed arrhythmia was the most common cause (46%) of death in the biventricular group. Fifty-one patients had undergone surgery. Six patients had primary electrophysiological disease, and 5 had cardiomyopathy. Eight deaths occurred in patients with pulmonary vascular disease. Sudden unexpected death occurred at a frequency of at least 10 patients per year over an 8-year period with 55,730 patient encounters. Arrhythmias (30%) and pulmonary vascular disease (13%) are important causes of sudden death. Infants and young children with surgical shunts comprise 23% of sudden unexpected deaths that occur within a month of the last evaluation [3].

Simple aortic valve disease and hypertrophic cardiomyopathy are rare (4%) causes of sudden death in childhood.

Many patients with ventriculoarterial discordance survive to adulthood. Those with complete transposition of the great arteries have often had an atrial switch procedure (Mustard or Senning operation) performed, which leaves the morphological right ventricle (RV) supporting the systemic circulation. RV failure and tricuspid regurgitation are common. Atrial arrhythmias are frequent, and atrial flutter may be a marker for sudden death. Patients with an arterial switch procedure also have long-term problems include coronary stenoses, distortion of the pulmonary arteries, dilatation of the neoaortic root, and aortic regurgitation.

Patients with congenitally corrected transposition have both atrioventricular and ventriculoarterial discordance and therefore also have a morphological RV and delicate tricuspid valve in the systemic circulation. Associated defects, such as abnormalities of the tricuspid valve, ventricular septal defect, and pulmonary stenosis, occur in the majority of patients. Heart block occurs with increasing age. Atrial arrhythmias occur frequently. Progressive tricuspid regurgitation occurs with age and is associated with deterioration of RV function and sudden death [4].

Tetralogy of Fallot is the most common cyanotic heart disease. Its operative mortality and long-term result are good. At the late phase after the correction, pulmonary valve regurgitation associated with right side heart failure, aortic valve regurgitation, arrhythmia and sudden death become major adverse outcomes. The electrophysiological and haemodynamic substrate of sudden death resembled that of sustained ventricular tachycardia, with pulmonary regurgitation being the predominant haemodynamic lesion [5]. Pulmonary regurgitation following repair of Tetralogy of Fallot is a common postoperative sequela associated with progressive right ventricular enlargement dysfunction. Although pulmonary regurgitation may be well tolerated for many years following surgery, it can be associated with progressive exercise intolerance, heart failure, tachyarrhythmia, and late sudden death. It also often necessitates re-intervention. Identifying the appropriate timing of such intervention could be very challenging given the risk of prosthetic valve degeneration and the increased risk of reoperation. Pulmonary valve replacement performed in an experienced tertiary referral centre is associated with low operative morbidity and mortality and very good long-term results [6]. Double-outlet right ventricle is a cyanotic heart disease with a wide spectrum of morphology and is divided according to the site of ventricular septal defect: subaortic, subpulmonary, doubly committed and remote type. Its operative methods are completely dependent on its morphology, and vary such as intracardiac tunnel repair, Rastelli type repair, arterial switch procedure and Fontan type repair. Left ventricular outflow tract obstruction is one of the most important problems after the correction. Recent operative strategy for the treatment of tricuspid atresia and single ventricle is the completion of right heart bypass operation using total cavo-pulmonary connection with staging strategy. Pleural effusion, ascites, protein loosing enteropathy and supraventricular arrhythmia are major adverse outcomes after Fontan type repair, while extracardiac total cavopulmonary connection is expected to reduce the incidence of supraventricular arrhythmia [7].

Adults with congenital heart disease

Adults with repaired congenital heart disease represent a complex and heterogeneous group of patients that are increasingly surviving beyond childhood. Patients have a variety of diagnoses that include specific structural anomalies, assorted physiologic derangements, and unique techniques for surgical repair. During long-term follow-up, even the most excellent surgical outcome may result in anatomic stenosis and insufficiency, cardiac rhythm disturbance, and myocardial dysfunction. Any of these abnormalities, alone or in combination, may result in significant morbidity and mortality. Sudden death is common and arrhythmias are frequently suspected to be the cause. Unfortunately, arrhythmias are difficult to predict and may be lethal at their initial presentation. In addition, a wide spectrum of arrhythmias, both supraventricular and ventricular, are possible, depending on the specific diagnosis and type of repair performed. Supraventricular arrhythmias may be as lethal as ventricular arrhythmias, arrhythmia substrates develop in a unique manner when compared with other adult cohorts [8]. Sudden death may also occur in Turners syndrome [9].

In our database of sudden cardiac death 77/2000 (3.9%) cases died of a sudden cardiac death with congenital cardiac lesions. Simple CHD cases were atrial/ventricular septal defect (ASD/VSD, n=23), aortic stenosis (AS, n=3), Noonan syndrome (n=3) and Williams syndrome (n=2). Complex CHD cases were double outlet right ventricle (n=10), Tetralogy of Fallot (n=6), transposition of the great vessels (TGV, n=4), congenitally corrected TGV (n=2), atrioventricular septal defect (AVSD, n=4), anomalous pulmonary venous drainage (n=2), Ebstein anomaly (n=1) and hypoplastic left heart syndrome (n=1). There were cases of morphologically normal hearts with a functional bicuspid aortic valve (BAV) and no complications (n=3), healed VSD (n=2) and small ASD (n=1). Aortic CHD cases included coarctation of the aorta (n=4), sinus of Valsalva aneurysm (n=1) and aortic dissection with BAV (n=1). Complications of bacterial endocarditis occurred with AS (n=1) and AVSD (n=1). Finally there were cases of sudden death with pulmonary hypertension (n=2) secondary to CHD and ischaemic heart disease with CHD (n=2) [10].

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