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Editorial

Prediction and prevention of sudden cardiac death in transposition of the great arteries: A step closer

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Recently, Khairy et al. investigated the pathophysiology of sudden cardiac death (SCD) in patients with dextro-transposition of the great arteries (d-TGA) and Mustard or Senning baffles [1]. Based on his findings he postulated that myocardial oxygen supply/demand mismatch may provoke ischemia related tachyarrhythmias leading to SCD. In the current issue of this journal, Khairy et al. provide for the first time histopathological evidence in favor of this theory [2]. In the last decades, drastic changes in management of patients with d-TGA have resulted in excellent survival rates and improvements in quality of life. The Mustard and Senning procedures, also known as the intra-atrial baffle repairs, were introduced in the early 60s as the first corrective surgery available for d-TGA [3,4]. However, patients often had post-surgical complications such as baffle leakage and tachyarrhythmias [5]. The arterial switch procedure, introduced by Jatene et al. in 1975, led to superior outcomes with respect to incidences of ventricular tachyarrhythmias and subsequently long-term survival [6,7].

Unfortunately, delay in diagnosis and/or lack of resources in developing countries have resulted in a considerable number of d-TGA patients presenting after the neonatal period. The Mustard and Senning procedures are therefore still performed. This yields in a challenging population with a systemic right ventricle (SRV) at high-risk for SCD [8]. Data on the pathophysiology of SCD in these patients is scarce, thereby hampering identification of high risk d-TGA patients requiring implantable cardioverter-defibrillator (ICD) implantation.

As the majority of SCD cases in this population occur during exercise, it is plausible to assume that a misbalance in oxygen demand and supply may provoke ischemia and subsequent ventricular and/or atrial tachyarrhythmias [1]. In the current issue of this journal, Khairy et al. retrospectively analyzed 140 adults with d-TGA and atrial switch surgery for the incidence of cardiac arrest of presumed arrhythmic etiology [2]. Cardiac arrest occurred in 8 patients (6%, 5 men, 30.5 ± 8.6 years) and none were prior diagnosed with coronary artery disease or sustained ventricular tachyarrhythmias. Half of these patients had atrial tachyarrhythmias and only one patient had a history of non-sustained ventricular tachyarrhythmias. The majority of the patients (n = 6) had at least moderate SRV dysfunction and only 1 patient received beta-blocking agents prior to arrest. Cardiac arrest occurred during exercise (n = 3), after consuming methamphetamine (n = 1) and in rest in the presence of an atrial tachyarrhythmia (n = 1).

Interestingly, autopsies in 2 out of 5 patients revealed acute myocardial infarction and chronic subendocardial ischemic lesions of the hypertrophied SRV in absence of coronary artery disease. One patient died in the early post-operative period after tricuspid valve replacement when post-operative complications, such as cardiac tamponade or tachyarrhythmias might have played a role. The other patient who died had a history of atypical chest pain during exertion since the age of 29 years. Serial exercise testing by myocardial perfusion scintigraphy revealed no electrocardiographic or ischemic changes. Yet, ventricular fibrillation occurred 9 years later while walking. Hence, these observations by Khairy et al. help us to understand the pathophysiological changes underlying SCD in d-TGA.

Optimal medical management is important in this patient population in order to achieve efficient oxygen consumption. Therefore, beta-blocker therapy to reduce the oxygen consumption in order to prevent atrial tachyarrhythmias and improve stroke volume should be considered. Surprisingly, only one patient received beta-blocker therapy in this high risk SCD population.

Adult patients may have different risk profiles compared to pediatric patients. Previous data showed that SRV dysfunction and ventricular tachyarrhythmias are mainly encountered in adults [9,10]. Focusing on adults with d-TGA, age, SRV dysfunction and a prolonged QRS (≥140 ms) duration are associated with SCD [10].

Khairy et al. should be complimented for providing this first proof in favor of the myocardial ischemia hypothesis which is a step-closer in identifying high-risk SCD patients [2]. Yet, further research in a larger population is warranted to obtain more histopathological evidence supporting this theory. As suggested by Khairy et al., aggressive control

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of tachyarrhythmias and counseling in order to avoid stimulants and high intensity exercise should be considered in clinical practice [2]. However, it would also be interesting to implant ICDs for primary prevention in this high-risk population. Special attention should be paid to patients with severe ventricular dysfunction and QRS prolongation exposed to triggers, such as intensive exercise, which might provoke myocardial ischemia and subsequently lethal tachyarrhythmias resulting in SCD.

Disclosures

None.

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