Anomalous coronary arteries and sudden death: time for action

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Coronary artery anomalies are rare congenital disorders of epicardial coronary arteries that comprise diverse malformations involving their number (e.g. single or duplicated artery), origin (e.g. artery arising from opposite coronary sinus of Valsalva or pulmonary artery), course (e.g. intramural course, course in relation to the aorta and pulmonary trunk), termination (e.g. fistula) and structure (e.g. stenosis, atresia). Most of these anomalies are clinically silent and prognostically insignificant. However, some specific malignant forms induce myocardial ischaemia, potentially leading to lethal complications. Anomalous origin of left coronary artery from the pulmonary artery causes myocardial infarction in infancy that, if left untreated, results in 90% mortality in the first year of life.[1] Anomalous origin of the left main coronary artery (ALMCA) from the right sinus of Valsalva or anomalous origin of the right coronary artery (ARCA) from the left sinus, combined with a coronary artery course between the aorta and pulmonary trunk, are associated with sudden death in young athletes, especially during or after exercise.[2] Death has been attributed to various mechanisms: presence of a narrowed slit-like coronary orifice, acute angle vessel takeoff, proximal intramural arterial course and exercise-induced coronary artery compression between the aorta and pulmonary trunk. However, these factors did not appear to be predictive for sudden death in a study that compared the anatomical features of ALMCA or ARCA in 12 patients who died by these anomalies versus 18 others who died of unrelated causes.[3] Other non-anatomical mechanisms have therefore been invoked, including spasms from endothelial injury and ventricular tachyarrhythmia.[4]

In this journal issue, Türkmen et al reported the postmortem findings in two cases of sudden death due to ARCA (associated with a noncontributory fenestrated membrane of the sinus coronarius),[5] and single coronary artery.[6] In each, the culprit artery traversed a course between the aorta and pulmonary trunk. Both subjects experienced mild premonitory symptoms, the significance of which were not appreciated in life. Notably, sudden death occurred at rest, and not during exercise. This is rare, but by no means unknown.[7] In one case, the authors found an area of subendocardial bleeding at the ventricular septum without histological evidence of acute or chronic ischaemia.[5] However, they did not expressly document autopsy evidence of ventricular myocardial infarction in the other case,[6] which might have shed light on the pathogenesis of sudden death.

Antemortem diagnosis of ALMCA or ARCA with vulnerable interarterial course is notoriously difficult. Sudden death is the first presentation in over half of the cases. Patients with ALMCA are more likely to report ischaemic symptoms such as chest pain and/or syncope, but are unlikely to raise the diagnostic alarm, given their youth and apparent good health. Rest and even exercise stress electrocardiography are rarely abnormal.[2] Cardiac magnetic resonance imaging and multi-slice computed tomography depict the proximal coronary artery course accurately, and are excellent for assessment of the course of the anomalous artery in relation to the aorta and pulmonary trunk.[8,9] However, their high cost and the risk of ionising radiation, and nephrotoxic contrast exposure in the latter, prohibit use of these modalities for routine screening.

For a diagnostic screening tool to be useful, it must be safe, noninvasive, inexpensive and widely available. Prior studies report favourable results with the use of echocardiography to screen for anomalous coronary arteries. In 1,360 young athletes prospectively evaluated by echocardiography, Pelliccia et al were able to visualise the ostium and proximal epicardial course of the left main coronary artery in 97% and the right coronary artery in 80% of subjects.[10] Similarly, Zeppelli et al reported clear echocardiographic visualisation of the ostia and proximal tracts of coronary arteries in 3,150 out of 3,650 competitive athletes.[11] More recently, Davis et al successfully employed echocardiography to screen 2,388 asymptomatic children and adolescents from the general population, and discovered four cases of anomalous coronary arteries (0.17%).[12] These studies underscore the feasibility and usefulness of echocardiographic screening for ALMCA or ARCA. Cardiologists and ultrasonographers must be taught to specifically look out for, and report, the origins and proximal courses of coronary arteries, especially in young patients. This is especially relevant in countries, like Singapore, which practise compulsory military conscription of young men, who will have to undergo strenuous physical training. Routine echocardiographic screening for anomalous coronary artery origins in these subjects should identify patients with malignant anomalous coronary arteries for curative surgery, thereby averting sudden death from

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exercise training. At the very least, echocardiographic assessment of coronary origins must be obligatory for young adults presenting with atypical cardiac symptoms. In cases where these cannot be visualised with certainty, cardiac magnetic resonance imaging or computed tomography should be strongly considered.

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