study cohort. The endpoints were mortality at 30 days and 5 years, and in-hospital length of stay. 1684 patients (median age 83 [79 – 87] years; 32% female) were screened, of whom 121 (7.2%) had RBBB. 41 (33.9%) patients received a prophylactic PPM by clinical preference. Of the remaining 80, 45 (56%) patients received a PPM after TAVI. Baseline demographic and procedural characteristics were similar. Among those patients in sinus rhythm who had a prophylactic PPM, the PR interval was longer than in those who did not receive a prophylactic pacemaker (200 ms [170 – 228] vs. 175 ms [158 – 196]; p=0.05). Similarly, the QRS duration was longer (140 ms [126 – 156] vs. 131 ms [120 – 142]; p=0.01). There was numerically but not statistically more left anterior hemiblock (24 [62%] vs. 29 [45%]; p=0.10). All-cause mortality was similar at 5 years with death in 17 patients (41.4%) in the prophylactic PPM group vs. 27 (33.8%) in the no prophylactic PPM group; hazard ratio 1.27 (95% CI 0.69 to 2.33; p=0.44; Figure 1). In the no prophylactic PPM group 16 (35.6%) deaths occurred in those receiving PPM after TAVI and 11 (31.4%) in those not receiving PPM; hazard ratio 0.95 (95% CI 0.43 to 2.09; p=0.90). 30-day all-cause mortality was also similar with 2 deaths (5.3%) occurring in the prophylactic group vs. 5 deaths (6%) in those without a prophylactic PPM, 3 (8.6%) of which were in those not receiving PPM and 2 (4.4%) receiving PPM after TAVI. There was a trend to a reduced hospital length of stay in those receiving prophylactic PPM compared to those who either received a post TAVI PPM or who were discharged without PPM (2.5 [1.6] days vs. 4.0 [4.8] days, respectively; p = 0.08). When comparing the prophylactic PPM group to those who received PPM after TAVI, there was a statistically significant reduction in length of stay (2.5 [1.6] vs. 4.3 [4.5] days, respectively; p=0.02; Figure 2). Two patients in the prophylactic PPM group developed a pocket haematoma managed non-operatively.

Conclusions Over half of patients with RBBB undergoing TAVI require a pacemaker after their valve implant for high-grade AV block. A prophylactic pacing strategy in this high-risk cohort is safe and reduces length of hospital stay.

Conflict of Interest None

10 DISEASE PENETRANCE IN ASYMPTOMATIC CARRIERS OF FAMILIAL CARDIOMYOPATHY VARIANTS

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Introduction Predictive genetic testing allows the identification of at-risk first-degree relatives of patients with genetic cardiomyopathies. Data on the penetrance of genetic variants associated with cardiomyopathies is limited. The aim of this study was to investigate disease penetrance in asymptomatic carriers of familial cardiomyopathy variants associated with hypertrophic cardiomyopathy (HCM), dilated cardiomyopathy (DCM) and arrhythmogenic right ventricular cardiomyopathy (ARVC).

Methods We included asymptomatic individuals referred to the Royal Brompton Hospital for predictive testing after the finding of a pathogenic or likely pathogenic genetic variant in a first-degree relative with cardiomyopathy between January 2017 and December 2019. Cardiomyopathy diagnosis was defined by international guidelines. Those with a prior diagnosis or signs or symptoms of heart disease at the time of testing were excluded. Results: A total of 105 genotype-positive individuals from 80 families were evaluated (median age 24.9 years [interquartile range: 16.1 to 45.0 years], 51 [48.6%] males). Variants in genes associated with DCM included: TTN n = 16 (51.6%), MYH7 n = 5 (16.1%), LMNA n = 3 (9.7%), TNNI3 n = 3 (9.7%), RBM20 n = 2 (6.3%), BAG3 n = 1 (3.2%), DMD n = 1 (3.2%); variants in genes associated with HCM included: MYBPC3 n = 31 (47.7%), MYH7 n = 25 (38.5%), TPM1 n = 5 (7.7%), PLN n = 1 (1.5%), TNNC1 n = 1 (1.5%), TNNI3 n = 1 (1.5%), TNN2T n = 1 (1.5%); and variants in genes associated with ARVC included: PKP2 n = 4 (44.4%), DSP n = 4 (44.4%), FLNC n = 1 (11.1%). On first clinical evaluation 2 of 31 carriers of DCM variants were diagnosed with DCM, 5 with hypertrophic non-dilated cardiomyopathy (HNDc) and 4 with isolated left ventricular (LV) dilatation; 10 of 65 carriers of HCM variants were diagnosed with HCM; and 0 of 9 carriers of ARVC variants were diagnosed with ARVC. Over a median follow-up of 2.4 years (interquartile range: 1.0 to 4.3 years) an additional 13 of 84 carriers developed cardiomyopathy phenotypes (0 of 20 DCM, 3 of 20 HNDc, 3 of 20 isolated LV dilatation, 7 of 35 HCM, 0 of 9 ARVC). Furthermore, 2 individuals with DCM/HCM received implantable cardioverter defibrillators and 2 individuals with DCM/HCM received implantable loop recorders.

Conclusions Approximately one-third of asymptomatic carriers of familial cardiomyopathy variants were diagnosed with a cardiomyopathy phenotype at initial screen or during a short follow-up period. This confirms the importance of predictive testing and the need for follow-up of genotype-positive, phenotype-negative individuals.

Conflict of Interest None

11 IS PULMONARY ARTERY DILATATION PART OF THE ATHLETIC ADAPTATION TO EXERCISE?

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Introduction The ‘Athlete’s Heart’ is a well-established term used to describe the spectrum of structural and electrical cardiac adaptations to intensive exercise. Several studies have demonstrated the development of ventricular dilatation and hypertrophy in athletic individuals and cardiac magnetic resonance imaging (CMR) is well suited to assess these changes. However, there is a paucity of studies looking at pulmonary artery (PA) dimensions in athletic individuals. One could hypothesize that increased stroke volumes and pulmonary flow may cause dilatation of the PA. The aim of this study is to ascertain whether PA dilatation is part of the cardio-pulmonary adaptation to intensive exercise and establish normal parameters of PAs in athletes.

Methods The CMRs of male athletes performed at a single-centre between January 2017 and October 2021 were reviewed. Those with any evidence of a structural abnormality
Abstract 11 Figure 1 Study consort diagram. Abbreviations: ESE, exercise stress echocardiography; AVR, aortic valve replacement; AS, aortic stenosis; HCM; hypertrophic cardiomyopathy; TiA, transient ischaemic attack; CABG, coronary artery bypass graft; NSVT, non-sustained ventricular tachycardia.

Abstract 11 Figure 2 Risk of maternal cardiovascular event based on associated factors. Abbreviations: NYHA, New York heart association; BMI, body mass index; mWHO, modified world health organisation risk source; ESE, exercise stress echocardiography.

or cardiomyopathy were excluded. The volumes of all cardiac chambers were recorded, indexed to body surface area (BSA). The PA was measured in the transaxial stack at the level of the bifurcation of the main PA. This was repeated for a control group of sedentary individuals with otherwise structurally normal hearts. Sporting discipline of the athletes and sedentary controls was confirmed by retrospective review of the patients’ clinical records. Differences between the two groups were compared, with statistical significance for P-values set at <0.05.

Results The CMRs of 169 male athletes were compared with 95 male sedentary controls. There was no difference in the mean ages of the groups (athletes 33±11 years (range 17–59) versus controls 30±13 years (range 17–60); p=0.10) The main indications for CMR in the athletic cohort included cardiac symptoms 25% (n = 44), ECG abnormalities 24% (n = 41) and screening due to a family history of cardiac disease 22% (n = 37). The majority of athletes engaged in endurance sport (60%; n = 101) and football (18%, n = 30).The differences between the groups is shown in table 1. The volumes of all cardiac chambers, were significantly larger in the athletes. LV ejection fraction (LVEF) was significantly lower in athletes. The average heart weight was 507±152 grams and left ventricular (LV) fibrosis was found in 28 (42%) cases (Figure 1A). Death was more common between 16 and 20 years of age (n=24) (Figure 1B). Death occurred during exertion in 25 (38%) individuals and at rest or during daily activities in the remaining 41 (62%), including 5 individuals who died during sleep. Male sex was more represented among decedents who died during exertion (88% compared with 68% in the group that died at rest, p=0.07); LV fibrosis was significantly larger than in sedentary controls (athletes 25.1 (±3.7)mm vs controls 22.4(± 3.0)mm). The distribution of PA diameters is shown in figure 1. There was no significant correlation between PA diameter and either RV end-diastolic volume (r=0.10) or RV stroke volume (r=0.04). Incidentally, a significantly higher proportion of athletes had fibrosis at the RV insertion point.

Conclusion Male athletes have a significantly larger PA than sedentary controls with 22% athletes having a PA≥27 mm compared to just 7% of controls. This suggests that PA dilatation may be part of the cardiopulmonary adaptation to exercise. These findings need corroboration in larger-scale, prospective studies, including pulmonary pressures and a wider array of sporting disciplines. The long-term implications of pulmonary artery dilatation in athletes is unknown but it may be used as an additional parameter to indicate athletic adaptation.

Conflict of Interest Nil

Background Sudden cardiac death (SCD) in young individuals and athletes is generally caused by hereditary cardiac conditions, including cardiomyopathies such as hypertrophic cardiomyopathy (HCM). Although historically HCM has been reported as the predominant cause of SCD in young athletes, it is unclear as to what degree exercise is a trigger for possible fatal arrhythmias.

Aim We aimed to report on the circumstances of SCD in a cohort of young individuals aged ≥ 10 and < 30 whose autopsy was consistent with HCM.

Methods We reviewed 6860 consecutive cases of SCD referred to our specialist cardiac pathology centre 1994 and 2020. SCD was defined as death from a cardiovascular cause within 12 hours of apparent well-being. All cases underwent detailed autopsy evaluation of the heart, including histological analysis, by expert cardiac pathologists. A minimum of 10 blocks of tissue were taken for histological analysis. HCM was defined by the presence of increased heart weight or increased wall thickness and significant myocyte disarray at histological examination.

Results Of the total cases of SCD, 264 (4%) were due to HCM. Our cohort of young decedents comprised of 66 individuals (average age 21±5 years, males 76%). For the majority (n=52, 79%) SCD was the first manifestation of HCM. Our cohort of young decedents comprised of 66 individuals (average age 21±5 years, males 76%). For the majority (n=52, 79%) SCD was the first manifestation of HCM. The average heart weight was 507±152 grams and left ventricular (LV) fibrosis was found in 28 (42%) cases (Figure 1A). Death was more common between 16 and 20 years of age (n=24) (Figure 1B). Death occurred during exertion in 25 (38%) individuals and at rest or during daily activities in the remaining 41 (62%), including 5 individuals who died during sleep. Male sex was more represented among decedents who died during exertion (88% compared with 68% in the group that died at rest, p=0.07); LV fibrosis was significantly larger than in sedentary controls (athletes 25.1 (±3.7)mm vs controls 22.4(± 3.0)mm). The distribution of PA diameters is shown in figure 1. There was no significant correlation between PA diameter and either RV end-diastolic volume (r=0.10) or RV stroke volume (r=0.04). Incidentally, a significantly higher proportion of athletes had fibrosis at the RV insertion point.

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Conflict of Interest Nil

Abstracts

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