Abstracts

303 Predictors of a Coronary Artery Calcium Score of Zero in Patients with Familial Hypercholesterolaemia
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Background: A coronary artery calcium score (CACS) of zero is a hallmark of a very low risk of a coronary event over 15 years in asymptomatic subjects. Familial hypercholesterolaemia (FH) is a condition characterised by a markedly high risk of premature coronary artery disease (CAD). Therefore, it is useful to define the factors that predict a CACS of zero in this high-risk population.

Aim: To determine the predictors of a CACS of zero, as assessed by cardiac computed tomography (CT) scanning, in asymptomatic patients with FH.

Methods: Cross-sectional study of asymptomatic patients diagnosed with FH in a specialist clinic at Royal Perth Hospital in whom a cardiac CT for CACS was performed. Univariable and multivariable logistic regression were carried out to investigate the predictors of a CACS of zero.

Results: Data from 198 adult patients (40% males) were analysed. The mean age was 50.9 ± 10.0 years and 50% had a pathogenic mutation causative of FH. A CACS = 0 was found in 42% of the patients and the median CACS was 4.9 Agatston units (IQR 79.5). From a set of potential predictive variables, age, pre-statin LDL-C, hypertension, phenotypic Dutch Lipid Clinic Network Score (DLCNS), and the absence of a genetic mutation, were included in the multivariable analysis. Age, hypertension and DLCNS, particularly LDL-C concentrations, were independent predictors of a CACS of zero (p < 0.001, p = 0.014 and p = 0.011, respectively).

Conclusion: In asymptomatic, middle-age patients with FH, younger age, the absence of hypertension and a lower DLCNS are the major drivers of the absence of CAD, as assessed by CACS.

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304 Preload Dependence of Myocardial Work Parameters
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Background: Myocardial work (MW) is a novel technique which utilises speckle tracking strain derived global longitudinal strain (STGLS) in conjunction with the blood pressure to calculate a parameter of MW however preload should be considered when performing serial follow up measurements using MW.

Methods and results: In a double blind, cross over study of 6 healthy male subjects we compared the effects of IV administration of 30 ml/kg of 0.9% saline, Hartmann’s solution and 4% albumin, and 6 ml/kg of 20% albumin (albumin dose equivalent) on global work index (GWI), global constructive work (GCW), and global work efficiency (GWE) as measured by STGLS. Preload, as measured by 3D left ventricular end-diastolic volume (LVEDV) increased in all fluids as did GWI and GCW (Table 1) however there was no change in GWE with changes in preload. 0.9% saline induced the largest change in LVEDV, nonetheless Hartmann’s solution had the largest effect on both GWI and GCW although this change was not statistically different from the changes observed with the other fluids.

Conclusion: GWI and GCW are parameters that are preload dependent. GWE may be the most reproducible parameter of MW however preload should be considered when performing serial follow up measurements using MW.

Table 1.

<table>
<thead>
<tr>
<th>Fluid</th>
<th>ΔLVEDV (mL)</th>
<th>ΔGWI (mmHg)</th>
<th>ΔGCW (mmHg)</th>
</tr>
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<tbody>
<tr>
<td>0.9% Saline</td>
<td>10.9 ± 7.4</td>
<td>402.5 ± 227.8</td>
<td>372.0 ± 204.1</td>
</tr>
<tr>
<td>Hartmann’s</td>
<td>5.0 ± 5.3</td>
<td>497.0 ± 261.5</td>
<td>493.2 ± 256.0</td>
</tr>
<tr>
<td>4% albumin</td>
<td>5.7 ± 5.8</td>
<td>332.5 ± 120.3</td>
<td>282.7 ± 99.1</td>
</tr>
<tr>
<td>20% albumin</td>
<td>8.5 ± 8.9</td>
<td>247.5 ± 207.3</td>
<td>228.0 ± 139.2</td>
</tr>
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</table>

305 Prevalence of Fabry Disease in A Cohort with Unexplained Late Gadolinium Enhancement on Cardiac MRI
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Background: Fabry disease is a rare X-linked genetic disorder in which cardiac manifestations include LVH, contractile dysfunction, and fibrosis, visible on cardiac MRI (cMRI) as late gadolinium enhancement (LGE) of the myocardium. Fabry disease is an important diagnosis to make as treatment is available as lifelong replacement of the deficient enzyme.

Aim: To define the prevalence of Fabry disease in a cohort of patients with unexplained LGE on cMRI.

Methods: The study population was recruited from patients aged >16 years who had cMRI performed between 2010-2018 to investigate LVH, idiopathic LV dysfunction and/or idiopathic ventricular arrhythmia. Patients with ‘unexplained’ LGE (i.e. without a genetic diagnosis of an alternate cardiomyopathy such as HCM or biopsy-proven...