CLINICAL PROFILE OF TRANSTHYRETIN AMYLOID CARDIOMYOPATHY PATIENTS IN IRELAND

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Abstract 15 Table 1 Comparison of clinical characteristic

<table>
<thead>
<tr>
<th>Age (years)</th>
<th>hATTR-CM</th>
<th>wtATTR-CM</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>64-67.3</td>
<td>75.5</td>
<td>0.003</td>
<td></td>
</tr>
<tr>
<td>2-37</td>
<td>2 (20)</td>
<td>2 (20)</td>
<td>0.8</td>
</tr>
<tr>
<td>58</td>
<td>100</td>
<td>0.017</td>
<td></td>
</tr>
<tr>
<td>Mean NYHA Class</td>
<td>2</td>
<td>2</td>
<td>0.8</td>
</tr>
<tr>
<td>Permanent pacemaker in situ (%)</td>
<td>25</td>
<td>20</td>
<td>0.8</td>
</tr>
<tr>
<td>Mean LVEF (%)</td>
<td>51.13.6</td>
<td>45.4-10.8</td>
<td>0.3</td>
</tr>
<tr>
<td>Prevalence of atrial arrhythmia (%)</td>
<td>58</td>
<td>100</td>
<td>0.017</td>
</tr>
<tr>
<td>Prevalence of spinal stenosis (%)</td>
<td>16.7</td>
<td>60</td>
<td>0.04</td>
</tr>
<tr>
<td>Prevalence of peripheral neuropathy (%)</td>
<td>91.7</td>
<td>30</td>
<td>0.003</td>
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Background Transthyretin amyloid cardiomyopathy (ATTR-CM) is an under-recognised aetiology for heart failure with preserved ejection fraction. Early recognition and diagnosis of ATTR-CM should translate into better outcomes for these patients as targeted medications are licensed for use in ATTR-CM. European amyloid cardiomyopathy registries are establishing the clinical characteristics of this disease while creating diagnostic and prognostic tools with this data. Aim: To compare the clinical profile of patients with hereditary ATTR-CM (hATTR-CM) and wild-type ATTR-CM (wtATTR-CM) in Ireland.

Methods A retrospective review was performed on the electronic clinical documents of all ATTR-CM patients referred to our centre. The review period included dates from 1st January 2019 to 1st February 2021. The two groups were compared in terms of clinical characteristics (mean age, female prevalence, New York Heart Association (NYHA), left ventricular ejection fraction (LVEF)), and the prevalence of co-morbidities such as arrhythmias, permanent pacemaker insertion, carpal tunnel syndrome, spinal stenosis, and peripheral neuropathy. Statistical analysis was made using software package SPSS.

Results Twenty two patients in total were enrolled. 55% (n=12) had hATTR-CM compared to 45% (n=10) with wtATTR-CM. In the hATTR-CM group, 91.7% (n=11) of the patients had the T60A gene mutation and 8.3% (n=1) had the S50R gene mutation. There was a significant difference in the mean age between the two cohorts; hATTR-CM cohort mean age was 64.6±7.3 years compared to 76±8.4 years in the wtATTR-CM cohort (p-value = 0.003). (Table 1) There was a significant difference of the prevalence of atrial arrhythmia between the two groups; 58% (n=7) patients in the hATTR-CM group compared to all patients in the wtATTR-CM group (p-value = 0.02). In the hATTR-CM, 25% (n=3) were classified as NYHA 1, 50% (n=6) were NYHA 2, and 25% (n=3) were NYHA 3. In the wtATTR-CM group, 30% (n=3) were NYHA 1, 30% (n=3) were NYHA 2, and 40% (n=4) were NYHA 3. Pacemaker insertion rates were similar between groups 25% hATTR-CM and 20% wtATTR-CM respectively. The mean LVEF was 51±13.6% and 45.4±10.8% in the hATTR-CM and wtATTR-CM (p=0.3), respectively. The prevalence of carpal tunnel syndrome (33.3% vs 85.7%; p = 0.03) and spinal stenosis (16.7% vs 60%; p=0.04) was significantly lower in the hATTR-CM group compared to the wtATTR-CM group. Peripheral neuropathy was significantly more prevalent in the hATTR-CM group compared to the wtATTR-CM group (91.7% vs 30%, p=0.003).

Conclusion In this study, both sub-categories of ATTR-CM had similar prevalence. The T60A mutation is regarded as the most common hATTR gene mutation in Ireland which was reflected in this study. This study has limitations primarily due to the small size but demonstrates the emerging clinical profile of patients with ATTR-CM in Ireland. These findings will
have implications on the development of assessment and management pathways for this under-recognised patient cohort.

**16 SINGLE-CELL RNA SEQUENCING REVEALS CARDIAC CELL-SPECIFIC TRANSCRIPTOMIC CHANGES IN DILATED CARDIOMYOPATHY**

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NOT AVAILABLE FOR PUBLICATION

**17 THE NATURAL HISTORY AND STAGING OF PRE-CLINICAL HEART FAILURE**

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Introduction Heart failure (HF) is growing global epidemic. Given the high morbidity and mortality associated with HF, and the impact it has on an already stretched health care system, its effective management is a public health priority. Prevention is a critical component of this strategy and this is dependent on a more complete understanding of the natural history of the condition, especially its preclinical precursors.

Methods St Vincent’s University Hospital offers a HF Prevention service (STOP-HF) targeted towards patients in its catchment area that have risk factors for HF. We retrospectively analysed the serial echocardiographic and natriuretic peptide data of 1,425 of these patients who have had at least two visits. Stage A was defined as no previous diagnosis of HF and a normal echocardiogram with one or more of the following: hypertension, hypercholesterolaemia, obesity, vascular disease, diabetes mellitus, arrhythmia requiring therapy or significant valvular disease. Stage B, a cohort at higher risk of development of heart failure, was defined as no previous diagnosis of HF and evidence of a structural or functional abnormality on echocardiography including: ejection fraction <50%, diastolic dysfunction, left ventricular hypertrophy, valvular disease of at least moderate severity, an enlarged left ventricle or an abnormal LAVI. Demographics, comorbidities, medication use and biochemical data were also analysed from each visit.

Results A population of 1,425 was analysed. On average, visit 2 was completed 4.8 years after visit 1. The average age at visit 1 was 64yrs with females accounting for 46% (n=649). At visit 1, 72% (n=1022) of the population were classified as Stage A and 28% Stage B. The average BNP of a Stage A patient was 28pg/ml while the average BNP of a Stage B patient was 64pg/ml. At visit 2, 60% (n=858) of patients were classified as Stage A with 40% Stage B. At visit 2, the average BNP of a Stage A patient was 35pg/ml while the average BNP of a Stage B patient was 141pg/ml. In terms of progression, 53% of patients remained in Stage A, 18% progressed to Stage B, 21% remained in Stage B and 7% had regressed from Stage B to Stage A. The most prevalent manifestation of progression was an increase in LAVI while the notable change in patients demonstrating regression from Stage B to Stage A was a reduction in LVH. In terms of changes in BNP between the two visits, those who remained in Stage A had an average BNP increase of 10pg/ml from visit 1 to visit 2. Similarly, those who regressed from Stage B to Stage A had an average increase in BNP of 22pg/ml whereas those that progressed from Stage A to Stage B had an average increase in BNP of 68pg/ml.

Summary This natural history study of a large sample of patients at risk for the development of heart failure demonstrates a significant proportion of patient with Stage B and furthermore a concerning progression rate of progression from Stage A to Stage B. Our data also identify a change in NP as a useful clinical biomarker of Stage B and risk of progression. It is hoped that this initial study will form as a baseline for further analysis and help guide screening and prevention strategies in the future.

**18 MANAGEMENT AND OUTCOMES OF PATIENTS WITH LEFT ATRIAL APPENDAGE THROMBUS PRIOR TO PERCUTANEOUS CLOSURE. LAPTOP REGISTRY**

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**19 4 YEAR RESULTS OF THE CARDIOLOGY TRAINEE FEEDBACK PROJECT**

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Background The Higher Specialist Training (HST) Scheme is run by the Royal College of Physicians (RCPI) overseen by the Irish Board for Training in Cardiovascular Medicine (IBTCM). The stated aim of the scheme is to provide doctors upon completion, with the necessary skills and knowledge to practice competently, professionally and independently as cardiology specialists. In order to inform dialogue surrounding improvements to the training scheme, those enrolled have been surveyed annually over the past 4 years to gage their views regarding the following domains – trainer/trainee interaction, access to training, procedural training, service commitments and training, research and audit, training program structure, gender balance, future career planning and location, quality of life and in the past year the impact of the Covid-19 pandemic on training.

Methods For the past 4 years, the second author surveyed trainees in training years 1-4 through the medium of an online questionnaire. Trainees were asked to respond to a range of questions pertaining to the domains of interest listed above, predominantly indicating their level of agreement with a statement in the format of a 5 point Likert scale.

Results The response rate to the survey was excellent and averaged at 65% over the four years. Individual years ranged from 52% to a high of 82%. Even in the year with the lowest response (52%) a significant proportion of the trainees were represented. Overall trainees are happy with the training scheme with 88% saying they would recommend the