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SOCIAL DETERMINANTS OF HEALTH IN THE SETTING OF HYPERTROPHIC CARDIOMYOPATHY

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ABSTRACT

Introduction: Social determinants of health play an important role in explaining poor health outcomes across many chronic disease states. The impact of the social gradient in the setting of an inherited heart disease, hypertrophic cardiomyopathy (HCM), has not been investigated. This study sought to profile the socioeconomic status of patients attending a specialized multidisciplinary clinic and to determine the impact on clinical factors, psychosocial wellbeing and adherence to medical advice.

Methods: Patients with HCM and at-risk relatives attending a specialized multidisciplinary clinic in Sydney Australia between 2011-2013 were included. Clinical, socioeconomic, geographic remoteness and adherence data were available. A broader clinic and registry-based group completed a survey including psychological wellbeing, health-related quality of life, Morisky Medication Adherence Scale and individual-level socioeconomic information.

Results: Over a 3-year period, 486 patients were seen in the specialized clinic. There was an over-representation of patients from socioeconomically advantaged and the least geographically remote areas. Socioeconomic disadvantage was associated with comorbidities, poor psychological wellbeing and health-related quality of life, lower understanding of HCM and more complex clinical management issues such as NYHA class, atrial fibrillation and left ventricular outflow tract obstruction. Approximately 10% of patients were non-adherent to medical advice, and poor medication adherence was seen in 30% of HCM patients with associated factors being younger age, minority ethnicity, anxiety and poor mental quality of life.

Conclusions: Of patients attending a specialized cardiac genetic clinic, there is an overrepresentation of patients from very advantaged and major metropolitan areas and suggests those most in need of a multidisciplinary approach to care are not accessing it.

Keywords: Hypertrophic cardiomyopathy, social determinants of health, adherence
INTRODUCTION

A universal healthcare system endeavors to provide equitable access to all avenues of health care and services for the population. In reality, social determinants, including social and economic circumstances, affect health throughout life. The World Health Organization (WHO) defines social determinants of health as the conditions into which people are born, live, work and age [1]. These include socioeconomic characteristics, which are potent determinants of health. Individuals further down the socioeconomic ladder have an increased risk of serious illness and premature mortality relative to those of higher status, and these effects are not restricted to the most disadvantaged, but rather operate as a social gradient across society [2]. Socioeconomic factors also influence how individuals interact with health care services. For example, low socioeconomic status has been consistently associated with lower utilization of general and preventive health checks [3], lower uptake of disease-specific screening programs such as breast cancer screening [4] and poorer access to disease-specific health care services such as cardiac services [5]. These social gradients in the use of health services may contribute to social inequities in health outcomes, and hence are important considerations in optimizing health care and services.

To date, no study has investigated the importance of socioeconomic characteristics as determinants of health service use or clinical outcomes in the setting of a genetic heart condition such as hypertrophic cardiomyopathy (HCM). HCM is the most common genetic heart condition, affecting at least 1 in 500 of the general population and is characterized by left ventricular hypertrophy in the absence of other loading conditions [6]. There is significant variability in the clinical presentation and natural history of disease, but undoubtedly the most tragic outcome is sudden cardiac death (SCD) [7]. Being a genetic condition, first-degree (at-risk) relatives are advised to undergo lifetime clinical surveillance due to a 1 in 2 risk of inheriting the gene mutation. HCM is an autosomal dominant genetic
disease, and despite rare founder mutations [8] affects the population equally. Disease guidelines exist, and while some controversies remain, the diagnosis, medical management and clinical/genetic surveillance of the family is established [9]. In Australia and worldwide, the multidisciplinary specialized cardiac genetic clinic is advocated as the ideal model of care for HCM families, offering expertise from genetic counselors, cardiologists, and others (Figure 1). The cost-effectiveness of a specialized multidisciplinary service incorporating genetic testing has recently been shown both in an Australian [10] and British [11] health care system highlighting the significant benefit of this type of service.

Little is known about the sociodemographic profile of patients attending a multidisciplinary cardiac genetic clinic. This study sought to profile the socioeconomic and geographic characteristics of consecutive patients attending a specialized clinic over a 3-year period, and in a broader patient population to identify whether social determinants such as education, income and geographic remoteness, impact on clinical factors, psychosocial wellbeing and adherence to medical advice.
METHODS

Consecutive clinic patient group

Consecutive patients attending a specialized cardiac genetic clinic in Sydney Australia, over a 3-year period from January 2011 through December 2013 were included in this study. Patients with a diagnosis of HCM, those investigated for possible HCM and all first-degree at-risk relatives who attended during the time period were included. Patients who failed to attend their appointment, but for whom enough clinical information could be gathered from the referral letter were included. Information available for this patient group included clinical data (i.e. disease status, clinical history, echocardiographic data, family history and genetic status), postcode, adherence and referring doctor’s details.

Patient adherence to guidelines

Adherence to medical advice and current guidelines was determined. For those with HCM, individuals were assessed based on their regularity of attendance for cardiology review, with those not being seen on at least a 2-yearly basis classified as non-adherent. Patients who declined to take a medication prescribed to them, who continued to participate in high-level or competitive sports, and who declined ICD therapy against recommendations were also considered non-adherent. Non-adherence was adjudicated by the multidisciplinary team and based on clinic encounters within the study period. At-risk relatives were considered non-adherent if they failed to attend for clinical screening as per the guidelines despite knowledge of this recommendation.

Socioeconomic status and geographic remoteness

Socioeconomic status was assessed using an area-level approach to categorize participant’s neighbourhood of residence using the Index of Relative Socioeconomic Advantage and Disadvantage (IRSAD) and the Index of Education and Occupation (IEO). Geographic remoteness of participant’s place of residence was determined by a
Remoteness Areas (RA) classification, developed by the Australian Bureau of Statistics and based on postcode.

The IRSAD is one of the Socioeconomic Indexes for Areas (SEIFA) 2011 developed by the Australian Bureau of Statistics [12], and ranks every region of Australia by relative socioeconomic advantage and disadvantage based on data collected in the 5-yearly Census. Decile 10 is the most advantaged and least disadvantaged area. The IEO was also developed as part of the SEIFA 2011, and is an index based around the skills of the people in an area, including both formal qualifications and skills required for different occupations. For this study, both indexes were grouped by top and bottom 50% of deciles to create a binary variable. The base unit of postal area code was used.

Geographic remoteness was determined by the ABS Australian Standard Geographical Classification (ASGC) Remoteness Areas classification, which correlates with the categories of the previously used Accessibility/Remoteness Index of Australia (ARIA+) [13]. Areas are defined by the distance a person must travel along a road network to reach goods, services and opportunities for social interaction. Remoteness area classifications include RA1, “Major cities of Australia”; RA2, “Inner regional”; RA3, “Outer regional”; RA4, “Remote” and RA5, “Extremely remote”.

**Psychosocial survey patient group**

A broader patient group was approached to complete a detailed psychosocial survey (Figure 2). This included all patients with a diagnosis of HCM and first-degree relatives who had previously attended the specialized cardiac genetic clinic, or those who were enrolled in the Australian Genetic Heart Disease Registry [14] (enrolled from the Sydney site or not associated with any major recruiting sites, i.e. self-enrolled participants). Participants were aged 18 years and above and were required to have sufficient English
skills to complete the survey. Participants were approached either in person while attending the specialized clinic, by phone call, post or by email between 2012 and 2014.

The survey comprised the Medical Outcomes Short-Form 36 version 2 (SF36v2) [15], Hospital Anxiety and Depression Scale (HADS) [16], HCM Patient Experience Scale,[17] Understanding and Satisfaction with Understanding Scale,[17] Morisky Medication Adherence Scale (MMAS-8) [18] and sociodemographic questions (further information in supplementary material).

Socioeconomic status was determined using the area-level measures, IRSAD and IEO, and in addition was based on individual-level data including self-reported highest education level and household income (selected from predefined categories), two commonly used proxies for socioeconomic status [19]. For education, a binary variable was created to identify individuals who had attained a university level qualification (high education) and those who had not (low education). Similarly, for income, responses were converted to a binary variable where those who indicated their household income was $AUD1300 ($US1220, €910) per week and below considered low income (based on approximate Australian median household income) [20].

**Statistical analysis**

Data were analysed using SPSS version 22.0 software. Descriptive statistics were used to examine the socioeconomic and geographic profile of clinic patients. Associations between socioeconomic and geographic indicators and continuous and categorical outcome variables were analysed using student’s t-test and chi-squared analysis, respectively. Factors associated with medication adherence were assessed using bivariate logistic regression models allowing for adjustment by patient age, given the high likelihood of confounding.
RESULTS

Characteristics of the consecutive clinic patient group

Over a 3-year period from January 2011 to December 2013, 486 individuals meeting the inclusion criteria were seen in the specialized cardiac genetic clinic (Table 1). This included 342 (70%) individuals with HCM and 144 (30%) at-risk relatives. For those with disease, the mean age was 46 ± 17 years and 212 (62%) were males. Mean age at diagnosis was 39 ± 19 years and 147 (54%) presented symptomatically. There were 98 (35%) with an ICD, 142 (53%) with a family history of HCM including 45 (16%) with a relative dying suddenly. The mean maximum left ventricular (LV) wall thickness was 21 ± 6 mm and 32 (12%) had severe hypertrophy of 30mm or more. In addition, 67 (25%) had evidence of LV outflow tract obstruction at rest, 46 (17%) had atrial fibrillation, 101 (44%) had NYHA functional class of II-IV, and 16 (6%) had a previous SCD event (including resuscitated cardiac arrest, appropriate ICD therapy or SCD). At-risk relatives had a mean age of 29 ± 16 years and 69 (48%) were males.

Characteristics of the referring doctor for patients seen in the specialized clinic

There were 137 (41%) new patients seen over the study time period, and the remainder included those who were regularly followed up in the specialized clinics. In most cases, new patients had an existing diagnosis and this was their first presentation at a specialized clinic. Overall, 211 (64%) HCM patients were referred to the clinic by a specialist, which included adult and pediatric cardiologists, cardiac electrophysiologists and clinical geneticists. Of new patients seen in the clinic over the study period, 121 (88%) were referred by a specialist, compared to 90 (46%) specialist referrals for follow-up patients (p<0.0001). Only 16 (11%) at-risk relatives had a specialist referral to the clinic (p<0.0001) and 69 (49%) were seen as new patients during the study period.
Adherence to guidelines amongst clinic patients

Adherence to clinical guidelines was investigated across the consecutive clinic group, with 35 (10%) HCM patients and 10 (7%) at-risk relatives identified as non-adherent. Of those with HCM, 12 of 112 (10%) patients with clear risk factors for SCD declined ICD therapy despite medical advice. There were 13 (4%) HCM patients who did not see their cardiologist on a regular basis, 2 (0.6%) who continued to play high-level competitive sports and 8 (2%) who declined to take a prescribed medication, including beta-blockers, other anti-arrhythmics and anticoagulants.

Socioeconomic and geographic profile of patients attending the specialized clinic

The socioeconomic and geographic profile of clinic patients is summarized in Figure 3. Of the consecutive patient group, there were 241 (71%) HCM patients residing in the most advantaged 50% of IRSAD deciles (i.e. top 50% most advantaged areas) and 89 (26%) from the top most advantaged decile (decile 10; i.e. top 10% most advantaged areas). There were 105 (73%) at-risk relatives from the most advantaged 50% of IRSAD deciles and 27 (35%) from the top most advantaged IRSAD decile. There were 250 (76%) HCM patients residing in areas with an IEO in the top 50% of deciles (i.e. top 50% most educated areas) and 74 (22%) in the top most educated decile (decile 10; i.e. top 10% most educated areas). There were 106 (74%) at-risk relatives in the top 50% IEO deciles and 31 (22%) in the top decile.

There were 403 (85%) clinic patients who resided in the least remote category, “RA1 – Major cities of Australia”, including 278 (84%) HCM patients and 125 (87%) at-risk relatives. There were 40 (12%) HCM patients and 12 (8%) at-risk relatives from category “RA2 – Inner Regional”; and 9 (3%) HCM patients and 6 (4%) at-risk relatives from “RA3 – Outer Regional”. Only 2 (0.6%) HCM patients and no at-risk relatives resided in “RA4 – Remote” region of Australia. No patients represented the most extreme remoteness
category “RA5 – extremely remote”. The proportion of the Australian population that reside in these remoteness areas is RA1 60.7%, RA2 24.6%, RA3 11.7%, RA4 2.0% and RA5 1.0% (based on 1996 Australian Census data).

**Characteristics of the survey patient group**

The broader group approached to complete the survey included 610 patients from the Australian Genetic Heart Disease Registry and those seen in the specialized clinics from 2002-2013, with 275 included in the final analysis (response rate 45%). This included 209 (76%) patients with HCM and 66 (24%) at-risk relatives (Figure 2).

Of the 209 HCM patients, 156 (78%) had previously been seen in the specialized clinic, while the remainder had self-enrolled in the Australian Genetic Heart Disease Registry or were family members of clinic patients (Supplementary Table). Mean age was 53 ± 15 years and 120 (60%) were male. Clinically, the mean maximum LV wall thickness was 18 ± 8 mm, 37 (19%) had atrial fibrillation, 44 (23%) had LV outflow tract obstruction and 56 (32%) reported NYHA functional class II-IV symptoms. Eighty-four (41%) patients had an ICD.

**Psychosocial and clinical factors associated with education level, household income and geographic remoteness in HCM patients**

Compared with HCM patients with a high household income, those patients with a low household income (Supplementary Table) were on average older (57 ± 16 versus 49 ± 13 years, p<0.0001). Furthermore, HCM patients with a lower income were less likely to have been seen in the specialized clinic (71% versus 86%, p=0.01) and more likely to express a more complex disease phenotype including atrial fibrillation (24% versus 12%, p=0.03) and LV outflow tract obstruction (29% versus 17%, p=0.05). They were also more likely
than higher income patients to present with comorbidities (51% versus 33%, p=0.009). In terms of psychosocial differences, HCM patients with low income were more likely to be depressed (HADS-Depression scores ≥ 8, 28% versus 11%, p=0.004) and reported poorer physical quality of life (SF36v2 PCS, 44 ± 10 versus 47 ± 11, p=0.03). There was also a worse level of understanding of HCM in those with low incomes (14 ± 5 versus 16 ± 6, p=0.002).

Compared with HCM patients with a higher education level, those with a lower level of education (Supplementary Table) reported a poorer understanding of HCM (14 ± 6 versus 16 ± 6, p=0.03) and less satisfaction with their understanding (10 ± 3 versus 12 ± 3, p=0.001). They also had lower mental quality of life scores (SF36v2 MCS, 42 ± 17 versus 47 ± 13, p=0.002) and less involvement in their medical management (11 ± 2 versus 12 ± 2, p=0.01). In terms of symptoms, those from the lower educated group were more likely to have NYHA symptoms II-IV, though this only just reached statistical significance (41% versus 26%, p=0.05).

Patients who resided in more remote areas of Australia (i.e. RA2 and RA3) were less likely to have been seen in the specialized clinic (60% versus 86%, p<0.0001), were less satisfied with their understanding of HCM (11 ± 3 versus 10 ± 3, p=0.04) and reported worse physical quality of life (SF36v2 PCS, 46 ± 11 versus 43 ± 10, p=0.05) (Supplementary Table). The sub-domains of the SF36v2 were assessed (Figure 4) to provide further information about the quality of life impairments seen between the different socioeconomic groups.

**Medication non-adherence amongst HCM patients**
Poor self-reported medication adherence was identified in 45 (30%) HCM patients who completed the survey. Factors associated with poor medication adherence compared to moderate-good adherence included age (52 ± 14 versus 59 ± 13 years, p=0.004), mental quality of life (SF36v2 MCS, 40 ± 17 versus 46 ± 14, p=0.03), non-white/Caucasian ethnicity (24% versus 8%, p=0.005) and HADS-Anxiety (44% versus 24%, p=0.01). After adjusting for age, non-white/Caucasian ethnicity remained significantly associated with poor medication adherence, despite wide confidence intervals (OR 3.43, 95%CI 1.24-9.52, p=0.018), while HADS-Anxiety showed only borderline significance (OR 2.12, 95% CI 0.99-4.56, p=0.05) and mental quality of life remained significantly associated (SF36v2 MCS, OR 0.98, 95% CI 0.95-0.99, p=0.045). None of the indicators of socioeconomic status or geographic remoteness were associated with poor medication adherence.

**Factors associated with clinical screening adherence amongst at-risk relatives**

Sixty-six at-risk relatives completed the survey, with a mean age of 47 ± 17 years and 22 were males (33%). Of these, 17 were identified as being non-adherent to clinical screening advice (including 4 patients identified as non-adherent in the clinic patient group). Twenty-three (35%) at-risk relatives had never been seen in the specialized clinic, but were recruited from the Australian Genetic Heart Disease Registry. There were 51 (77%) at-risk relatives residing in ‘RA1 – Major cities of Australia’, 12 (18%) in ‘RA2 – Inner regional’ and 3 (5%) in ‘RA3 – Outer regional’ areas. There was a statistically significant association between geographic location and non-adherence amongst at-risk relatives [RA1 (n=9, 18%) and RA2/3 (n=8, 53%), p=0.015]. The self-reported level of understanding of HCM was found to be significantly worse in the non-adherent relatives compared to the adherent relatives (15.9 ± 5.5 versus 11.9 ± 5.5, p=0.014). No differences in IRSAD, IEO, psychological wellbeing and health-related quality of life were found between adherent and non-adherent relatives.
DISCUSSION

Globally, health outcomes are socially determined, but to date no study has evaluated the impact of social determinants on health and clinical characteristics in the setting of genetic heart diseases such as HCM. The current study supports the notion that it is primarily the advantaged and well educated who comprise the greatest proportion of patients who attend a specialized HCM clinic. Lower socioeconomic groups were under-represented in the clinic setting. Further, those patients from lower socioeconomic groups were characterized by worse psychological wellbeing and health-related quality of life, lower understanding of HCM and had more complex disease presentations (including atrial fibrillation, LV outflow tract obstruction and NYHA class II-IV symptoms and comorbidities). Overall the study suggests that the patient groups with the most significant and complex health issues are those from a lower socioeconomic class, and that specialized multidisciplinary clinics that are advocated as an ideal model of care for HCM are not serving this patient group in an equitable way.

Socioeconomic status, that is the individual’s social position relative to others, is known to be an important factor in the onset and progression of a number of health conditions including cardiovascular disease, with a low socioeconomic status associated with greater mortality and acting as a barrier to accessing high-quality health care [21, 22]. Inequities are considered avoidable disparities in delivery and access to health care and are driven by social, political and economic factors, and addressing these gaps is a major focus globally [22]. In Australia, whilst a universal health care system is in place, the provision of health care outside of hospitals is largely accessed via a combination of both private and public services, often resulting in a “user-pays” scenario [23]. A Medicare system exists, providing subsidies for most of the cost of doctor visits, incentives to provide bulk billing to concession card holders and a ‘Safety Net’ to cap out-of-pocket medical costs over a certain annual threshold [24]. However it is known that advantaged groups
disproportionally access and gain benefit from primary health care and specialist medical services, both in Australia [25] and elsewhere [26]. The confounding issue is that low socioeconomic status is frequently associated with a greater prevalence of chronic diseases and disease risk factors which taken together often give rise to more complex clinical management needs [27]. Our study highlights an overrepresentation of patients from very advantaged and major metropolitan areas, suggesting that the groups most in need of a multidisciplinary approach to care are not accessing it.

The clinical profile of patients referred to and seen in a specialized clinic present more challenging management issues and often more complex patients. The HCM clinic in Sydney is a tertiary referral center, with 64% of the HCM patients referred by another specialist, including general cardiologists, pediatric cardiologists and cardiac electrophysiologists. Typically in this clinical setting, patients may be either (1) seen for a particular management issue then will continue their ongoing cardiac follow-up with their own cardiologist, or (2) choose to be followed-up in the clinic (Figure 1). Ideally, all HCM patients should be referred at least once to the specialized clinic where they can be reviewed by the cardiology team and engage with the genetic counselor to discuss familial aspects of disease and any genetic testing options available to them. More realistically however, the specialized clinic serves to support the most complex clinical and genetic management scenarios where expertise is critical to ensure the best outcomes for the patient. Based on what we understand about increased morbidity and mortality in more socioeconomic disadvantaged groups, there may be a greater need for multidisciplinary specialized clinic care in this population.

Understanding the reasons for the disparity in the patient group accessing the specialized multidisciplinary clinic is challenging. Other groups have investigated inequities in access to specialist medical services and suggest it may be shaped by the health care system
itself, where despite a public system that can provide subsidies for many services, there still may be an out-of-pocket and/or private aspect [28]. Korda and colleagues suggest the reasons go beyond just cost, and shine a spotlight on the primary care physician/general practitioner as the gatekeeper for specialist referral [24]. Others suggest patient referral may be moderated by the perceived socioeconomic status of the patient, for example more disadvantaged patients may be expected to be non-compliant, and so the health practitioner does not make the referral [28]. In the setting of genetic heart disease management in a specialized clinic, it is unclear what the barriers are preventing referral of disadvantaged patients, though are likely a complex interaction of many different factors.

The influence of socioeconomic status on patient adherence to medical advice and therapies has been previously described [3, 29]. Our study reports the overall prevalence of non-adherence in a consecutive patient series of 10%. In all cases after lengthy discussion it was ultimately the patient who decided whether they follow medical advice. In one instance, a young man with 3 well-established risk factors for SCD declined ICD therapy due to a belief that a change in his lifestyle to incorporate better diet and exercise would be protective. The patient still remains without an ICD despite his very high risk of an event. An analysis of socioeconomic factors that may explain non-adherent behaviour in this patient group found no association in the present study. Non-adherence is likely the result of a number of factors including experience with the disease (i.e. family history of SCD), symptomatic status and the various coping strategies people use to negotiate difficult situations. Further, poor medication adherence was reported in 30% of the surveyed patients who were taking a regular medication for their heart condition (n=150). Factors associated with poor medication adherence were younger age, non-white/Caucasian ethnicity, anxiety and mental quality of life. Due to the high correlation between age and adherence, subsequent analysis adjusting for this was performed, and ethnicity, anxiety and mental quality of life remained significant suggesting greater
attention and a higher clinical suspicion of poor medication adherence for younger patients, those from minority ethnic groups and those indicating some anxiety or poor mental quality of life (e.g. lowered social functioning, lower perceived general health).
Conclusions

The effect of socioeconomic status on clinic attendance, and on clinical and psychosocial outcomes is evident in the HCM patient population, and determining ways to better serve those from more disadvantaged groups who likely present more complex clinical management issues is critical. The underlying reasons for the disproportionate number of advantaged and highly educated individuals attending a specialized clinic are likely to be multifactorial. Further work to better understand these inequities is imperative if we are to provide equitable care to all HCM patients.
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TABLE 1: Characteristics of consecutive patients seen in a specialized HCM clinic over a 3-year period

<table>
<thead>
<tr>
<th></th>
<th>HCM patients</th>
<th>At-risk relatives</th>
<th>All patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>N (%)</td>
<td>342 (70)</td>
<td>144 (30)</td>
<td>486</td>
</tr>
<tr>
<td>Mean age, years (SD)</td>
<td>46 ± 17</td>
<td>29 ± 16</td>
<td>41 ± 18</td>
</tr>
<tr>
<td>Male gender (%)</td>
<td>212 (62)</td>
<td>69 (48)</td>
<td>281 (58)</td>
</tr>
<tr>
<td>White/Caucasian ethnicity</td>
<td>229 (67)</td>
<td>102 (71)</td>
<td>331 (68)</td>
</tr>
<tr>
<td>Specialist referral</td>
<td>211 (64)</td>
<td>16 (11)</td>
<td>227 (48)</td>
</tr>
<tr>
<td>Top 50% IRSAD areas*</td>
<td>241 (71)</td>
<td>105 (73)</td>
<td>346 (71)</td>
</tr>
<tr>
<td>Top 50% IEO areas*</td>
<td>250 (76)</td>
<td>106 (74)</td>
<td>356 (73)</td>
</tr>
<tr>
<td>Geographic remoteness†</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Major Cities of Australia (RA1)</td>
<td>278 (84)</td>
<td>125 (87)</td>
<td>403 (85)</td>
</tr>
<tr>
<td>Inner regional (RA2)</td>
<td>40 (12)</td>
<td>12 (8)</td>
<td>52 (11)</td>
</tr>
<tr>
<td>Outer regional (RA3)</td>
<td>9 (3)</td>
<td>6 (4)</td>
<td>15 (3)</td>
</tr>
<tr>
<td>Remote (RA4)</td>
<td>2 (0.6)</td>
<td>0 (0)</td>
<td>2 (0.4)</td>
</tr>
<tr>
<td>Non-adherence</td>
<td>35 (10)</td>
<td>10/144</td>
<td>45/484</td>
</tr>
<tr>
<td>Declined ICD‡</td>
<td>12 (10)</td>
<td>-</td>
<td>12 (10)</td>
</tr>
<tr>
<td>Cardiology review</td>
<td>13 (4)</td>
<td>-</td>
<td>13 (4)</td>
</tr>
<tr>
<td>Competitive sports</td>
<td>2 (0.6)</td>
<td>-</td>
<td>2 (0.6)</td>
</tr>
<tr>
<td>Decline medication</td>
<td>8 (2)</td>
<td>-</td>
<td>8 (2)</td>
</tr>
<tr>
<td>Clinical screening</td>
<td>-</td>
<td>10/144</td>
<td>-</td>
</tr>
</tbody>
</table>

*Index for relative socioeconomic advantage and disadvantage (IRSAD), Index for Education and Occupation, high scores indicate those in top 50% of Australian population
†Australian Statistical Geographic Standard (ASGS)
‡Percentage calculated from the number of patients where an ICD was indicated
Abbreviations; ICD, implantable cardioverter defibrillator; RA, remoteness area
FIGURE LEGENDS

FIGURE 1: Participant recruitment to the study
The consecutive clinic group included those seen between 2011-2013. A broader group of participants from the specialized cardiac genetic clinic (2002-2013) and Australian Genetic Heart Disease Registry were invited to complete the survey.

FIGURE 2: Flowchart of the specialized cardiac genetic clinic
The specialized cardiac genetic clinic incorporates cardiology opinion and management, clinical surveillance of relatives, genetic counseling and testing options and education.

FIGURE 3: Distribution of (A) Index of Socioeconomic Advantage and Disadvantage (IRSAD), (B) Index of Education and Occupation (IEO) and (C) Geographic remoteness areas, for consecutive patients attending a specialized clinic
The majority of patients seen during the study time period resided in (A) socioeconomically advantaged (B) highly educated and (C) major metropolitan areas.
Abbreviations: RA1, major cities of Australia; RA2, inner regional; RA3, outer regional; RA4, remote; and RA5, extremely remote.

FIGURE 4: Spider-plots of the SF-36v2 domain scores by (A) Education level, (B) Household income and (C) Geographic remoteness amongst patients with hypertrophic cardiomyopathy
Health-related quality of life (HR-QoL) was impaired across a number of domains for patients with lower education level, lower household income and who were more geographically remote.
Abbreviations: PF, physical function; RP, role – physical; BP, bodily pain; GH, general health; VT, vitality; SF, social functioning; RE, role – emotional; MH, mental health; SES, socioeconomic status; RA1, metropolitan cities of Australia; RA2/RA3, inner and outer regional areas.

*Indicates a significant difference
FIGURE 1: Flowchart of the specialized cardiac genetic clinic

HCM patients

Cardiologist or other specialist referral
Direct contact from patient (primary care physician referral)

Opinion
Management
Genetics

Complex patients
Opinion regarding diagnosis, ICD etc.
Not seen long-term

Patients managed long-term (primary care physician referral)

Gene*c(counseling
Gene*c(test*ng
Inheritance & screening advice

At-risk relatives

Clinical screening

Relatives will return periodically long-term

First-degree relatives may attend the specialized clinic (primary care physician referral)
FIGURE 2: Participant recruitment to the study

Australian Genetic Heart Disease Registry
(NSW site or not linked to any recruiting sites, i.e. self enrolment)

HCM and Genetic Heart Disease clinics, Sydney (2002-2013)

(1) Clinic group:
All consecutive patients seen 2011-2013
N=486

(2) Survey group:
All HCM and at-risk relatives
Aged 18 years and over
Sufficient English skills
N=275
FIGURE 3: Distribution of socioeconomic indexes and geographic remoteness of consecutive patients attending a specialized clinic.

A

B

C

At-risk relatives

HCM

IRSAD Deciles

1

2

IEO

1

2

ASGS Remoteness Area

RA1
FIGURE 4: Spider-plots of the SF-36v2 domain scores by (A) Education level (B) Household income and (C) Geographic remoteness amongst patients with hypertrophic cardiomyopathy.