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ORIGINAL ARTICLE

The Portuguese Registry of Hypertrophic Cardiomyopathy: Overall results

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KEYWORDS
Hypertrophic cardiomyopathy;
Registry;
Left ventricular hypertrophy;
Outcome

Abstract
Introduction: We report the results of the Portuguese Registry of Hypertrophic Cardiomyopathy, an initiative that reflects the current spectrum of cardiology centers throughout the territory of Portugal.
Methods: A direct invitation to participate was sent to cardiology departments. Baseline and outcome data were collected.
Results: A total of 29 centers participated and 1042 patients were recruited. Four centers recruited 49% of the patients, of whom 59% were male, and mean age at diagnosis was 53±16 years. Hypertrophic cardiomyopathy (HCM) was identified as familial in 33%. The major reason for diagnosis was symptoms (53%). HCM was obstructive in 35% of cases and genetic testing

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[1] List of participating centers and principal investigators are provided in Appendix A.

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was performed in 51%. Invasive septal reduction therapy was offered to 8% (23% of obstructive patients). Most patients (84%) had an estimated five-year risk of sudden death of <6%. Thirteen percent received an implantable cardioverter-defibrillator. After a median follow-up of 3.3 years (interquartile range [P25-P75] 1.3-6.5 years), 31% were asymptomatic. All-cause mortality was 1.19%/year and cardiovascular mortality 0.65%/year. The incidence of heart failure-related death was 0.25%/year, of sudden cardiac death 0.22%/year and of stroke-related death 0.04%/year. Heart failure-related death plus heart transplantation occurred in 0.27%/year and sudden cardiac death plus equivalents occurred in 0.53%/year.

Conclusions: Contemporary HCM in Portugal is characterized by relatively advanced age at diagnosis, and a high proportion of invasive treatment of obstructive forms. Long-term mortality is low; heart failure is the most common cause of death followed by sudden cardiac death. However, the burden of morbidity remains considerable, emphasizing the need for disease-specific treatments that impact the natural history of the disease.

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PALAVRAS-CHAVE
Miocardiopatia hipertrófica;
Registo;
Hipertrofia ventricular esquerda;
Prognóstico

Registo Português de Miocardiopatia Hipertrófica: resultados globais

Resumo

Objetivo: Apresentação dos resultados do Registo Português de Miocardiopatia Hipertrófica.

Metodologia: Convite direto aos diferentes centros de cardiologia de Portugal, com análise de dados basais e de seguimento.

Resultados: Foram 29 os centros participantes e 1042 doentes incluídos. Quatro centros incluíram 49% dos doentes, 59% do sexo masculino, idade média de diagnóstico 53 ± 16 anos. A doença foi considerada familiar em 33% e a presença de sintomas foi a principal causa de diagnóstico (53%). A miocardiopatia hipertrófica foi obstrutiva em 35%. O estudo genético foi efetuado em 51%. Oito por cento dos doentes fizeram terapêutica invasiva de redução septal (23% dos doentes com obstrução). A maioria dos doentes (84%) apresentava um risco estimado de morte súbita aos 5 anos < 6%. Em 13% foi colocado desfibrilhador cardioversor implantável. Após um seguimento de 3,3 anos, intervalo interquartil (P25-P75) 1,3–6,5 anos, 31% estavam assintomáticos. A mortalidade total foi de 1,19%/ano e a cardiovascular de 0,65%/ano. A incidência de morte por insuficiência cardiaca foi de 0,25%/ano, a de morte súbita de 0,22%/ano e a de morte por acidente vascular cerebal de 0,04%/ano. A mortalidade por insuficiência cardíaca e transplante cardíaco foi de 0,27%/ano e a de morte súbita e equivalentes de 0,53%/ano.

Conclusões: A miocardiopatia hipertrófica em Portugal apresenta idade de diagnóstico elevada e é frequente o tratamento invasivo de formas obstrutivas. A mortalidade é baixa, a insuficiência cardíaca é a principal causa de morte, seguida pela morte súbita. A doença apresenta elevada morbilidade, realça a necessidade do desenvolvimento de tratamentos específicos com impacto na sua história natural.

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List of abbreviations

AF	atrial fibrillation
ASA	alcohol septal ablation
CMR	cardiac magnetic resonance
CRF	case report form
HCM	hypertrophic cardiomyopathy
HF	heart failure
ICD	implantable cardioverter-defibrillator
IVS	interventricular septum
LVH	left ventricular hypertrophy
PRo-HCM	Portuguese Registry of Hypertrophic Cardiomyopathy
SCD	sudden cardiac death
TIA	transient ischemic attack

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