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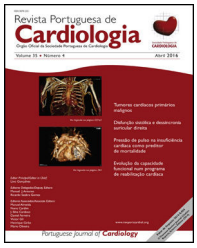

SHORT COMMUNICATION

Impact of the creation of specialized units for patients with hypertrophic cardiomyopathy

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Abstract
Introduction and objectives: According to current international guidelines, hypertrophic cardiomyopathy (HCM) patients should be managed in specialized units. However, there is lack of data on the impact of the creation of these units in the management of HCM patients. Our goal was to assess the impact of the creation of an Inherited Inherited Diseases Cardiac Unit (ICDU) in the current management of patients with HCM.
Methods: We analyzed 114 consecutive patients (62.6±8 years old, 70.2% males) with HCM. Variables related to optimal management of HCM patients and their family study were recorded, as well as guidance on the risk of sudden death. We analyzed whether patients were assessed by the ICDU or at a general cardiology consultation (GCC).
Results: 50 patients were assessed in the IDCU and 64 in the GCC. Familial screening was more frequent in patients assessed by the IDCU (45.3% vs. 4%; p<0.01), requesting more genetic studies of the index case (70.3% vs. 14%; p<0.01) and cardiac magnetic resonance (53.1% vs. 18%; p<0.01). Sudden death risk score was performed more frequently in patients after the creation of an IDCU (67.2% vs. 28%; p<0.01). Treatment with beta-blockers was similar in both groups (72% vs. 78.1%; p=0.24). An implantable cardiac defibrillator was indicated similarly in both groups (12.5% in ICDU and 6% in GC; p=0.24).
Conclusions: The implementation of an IDCU improved the quality of the medical care for HCM patients by performing a better study of the patients and their families.

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PALAVRAS-CHAVE
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Impacto da criação de uma unidade especializada no atendimento de doentes com miocardiopatia hipertrófica

Resumo
Introdução e objetivos: De acordo com as recomendações internacionais atuais, o tratamento de doentes com miocardiopatia hipertrófica (MCH) deve ser efetuado em unidades especializadas. No entanto, há escassez de dados sobre o impacto da criação de uma unidade de doenças cardíacas hereditárias (UDCH) no tratamento dos doentes com MCH.
Métodos: Analisámos 114 doentes consecutivos (62,6±8 anos, 70,2% homens) com MCH. Foram registadas as variáveis relacionadas com o tratamento aprimorado destes doentes e o respetivo estudo familiar, assim como o aconselhamento sobre o risco de morte súbita. Verificámos se os doentes foram avaliados em UDCH em consulta geral de cardiologia (GC).
Resultados: Foram avaliados respetivamente 50 e 64 doentes na UDCH e na CG. O rastreio familiar foi mais frequente em doentes avaliados na UDCH (45,3% *versus* 4%, p<0,01) requerendo a UDCH mais estudos genéticos no caso índice (70,3% *versus* 14%, p<0,01) e mais ressonância magnética cardíaca (53,1% *versus* 18%, p<0,01). O *score* de risco de morte súbita foi realizado com mais frequência em doentes após a criação da UDCH (67,2% *versus* 28%, p<0,001). O tratamento com betabloqueantes foi semelhante em ambos os grupos (72% *versus* 78,1%, p=0,24). Foi indicado desfibrilhador cardíaco implantável de forma semelhante em ambos os grupos (12,5% na UDCH e 6% na CG, p=0,24).
Conclusões: A implantação de uma UDCH melhorou a qualidade do atendimento médico aos doentes com MCH através de um estudo mais completo dos doentes e das suas famílias.

Hypertrophic cardiomyopathy (HCM) is the most frequent cardiomyopathy and represents a major health burden, causing heart failure and sudden death (SD) among young patients.[1] In its management, a variety of areas of cardiology work in partnership as the assessment of these patients is a complex process that should be developed into specialized units.[2]

The impact of these units is also complex and hard to measure. Before our study, there was no data that we are aware of on the impact of a specialized unit in the clinical assessment of patients with HCM. We present our initial experience in the assessment of these patients in an Inherited Cardiac Diseases Unit (ICDU).

We analyzed 114 consecutive HCM patients diagnosed by imaging testing according to current guidelines.[3] Fifty patients were assessed in a general cardiology consultation (GCC) and sixty-four in an ICDCU. The main clinical courses of action related to HCM clinical assessment were analyzed.

70.2% of patients were male and the mean age was 62.6±8 years. Prevalence of cardiovascular risk factors was higher in the GCC group, where more patients had hypertension (85% vs. 39.1%; p<0.01) and dyslipidemia (23.9% vs. 19.5%; p=0.03). Atrial fibrillation was also more prevalent in the GCC group (42.9% vs. 21.9%; p=0.017). Family history of sudden cardiac death (SCD) was found in 28.1% of patients assessed in the ICDU and in 4% of those assessed assessed in the GCCs (p<0.01).

The most frequent phenotype was septal hypertrophy (70.2%), followed by apical (14.9%). 38.6% of patients had left ventricle outflow tract obstruction, which was similar in both groups (p=0.51), with a mean peak gradient of 52.3± 28 mmHg in GCC and 67.9±42 mmHg in ICDU (p=0.19).

Table 1 shows the main characteristic related to the clinical management of the patients. In patients assessed in the IDCU, family screening occurred more frequently, as well as recording the family history and family tree.

The main tests required for a complete assessment of HCM[2,4] (including cardiac magnetic resonance, exercise test and genetic test) were performed more frequently in patients assessed in the IDCU. SCD risk assessment, using the HCM risk-SCD Score was also performed more often in the IDCU. Beta-blockers were administered in high numbers in both groups. Implantable cardioverter defibrillators were indicated much more frequently by the IDCU, although statistical significance was not reached.

In spite of the limited conclusions that can be drawn from a single experience, our results endorse the benefits of specialized units for the clinical assessment of HCM patients. They fulfil all the care needs of patients and their families, providing a comprehensive approach that is usually impossible at general consultations. The differences observed could be derived from the higher grade of staff training and implementation of specific protocols for each specific disease.

Table 1 Main actions related to the assessment of patients with hypertrophic cardiomyopathy in clinical practice.

	General Consultation (n=50)	Inherited Cardiac Diseases Unit (n=64)	p
Registry of family history/family tree	4 (8%)	64 (100%)	<0.01
Assessment of relatives	2 (4%)	29 (45.3%)	<0.01
Genetic test performance	7 (14%)	45 (70.3%)	<0.01
HCM Risk-SCD Score	14 (28%)	43 (67.2%)	<0.01
Exercise test	16 (32%)	42 (65.6%)	<0.01
Cardiac magnetic resonance	9 (18%)	34 (53.1%)	<0.01
Beta-blocker treatment	36 (72%)	50 (78.1%)	0.32
Defibrillator indication	3 (6%)	8 (12.5%)	0.24

HCM: hypertrophic cardiomyopathy; SCD: sudden cardiac death.

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Conflicts of interest

The authors have no conflicts of interest to declare.

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