The public health burden of cardiomyopathies: A call for action

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Related article
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Cardiomyopathies (CMs) are defined as myocardial diseases in which structural or functional abnormalities of the myocardium occur in the absence of conditions that can explain them. In recent years, tremendous progress in understanding the molecular background, genetic architecture, and diagnostic strategies has dispelled the concept of CMs as rare entities. In fact, they are now recognized as a major health burden and an important cause of heart failure, arrhythmias, and sudden cardiac death [1]. However, the clinical course of patients with CMs is not always straightforward. Depending on phenotype, genotype and comorbidities, patients may remain asymptomatic or experience multiple exacerbations. The variable expression of CMs has influenced epidemiologic studies, underestimating the true prevalence. In addition, most information comes from studies conducted in tertiary centers and are affected by referral bias. Finally, studies describing the clinical course and contemporary management of CMs are lacking. In this issue of the *Polish Heart Journal*, Mizia-Stec and colleagues [2] publish a cross-sectional, nationwide, retrospective study providing new insights into the management of patients with CMs. The authors collected data from the national healthcare provider sample using International Classification of Diseases 10th Revision (ICD-10) codes, including information on hospitalizations, either elective or urgent due to exacerbation, and outpatient care, either at tertiary centers or as first-line care. 65383 CM patients accessing the public health system in Poland between 2016 and 2021. They were stratified into three subgroups according to Charlson Comorbidity Index (CCI) score. Across the CCI score spectrum, male sex was prevalent and, as expected, age increased in parallel with CCI score. Conversely, higher CCI scores were associated with fewer healthcare interventions. In addition, patients with more comorbidities were more likely to be hospitalized and had the lowest rate of third level outpatient care. Interestingly, the authors identified hospitalization as a critical step in the diagnostic work-up for the majority of patients. In fact, while only 3.3% and 3.0% of patients were diagnosed in tertiary centers and first-line care, respectively, 93.4% of patients were diagnosed with CM during hospitalization (mostly urgent). In addition, outpatient diagnosis rates were further reduced in patients with CCI >5. Subsequent interactions with the health care system were rare. In fact, one-third of hospitalized patients were not re-registered, while one-fifth died. Most patients who had a second contact with the health system were hospitalized, while referral to tertiary care had the lowest rate. Despite the novelty and importance of the study, there are some limitations that readers should be aware of. The diagnosis of CMs was based on ICD-10 codes, which may be misreported or otherwise not necessarily confirm the diagnosis. However, the authors correctly mitigated this limitation by excluding patients with codes related to ischemic heart disease. Second, because the period 2016-2021 includes the outbreak of the COVID-19 pandemic, its impact on hospitalizations and outpatient visits cannot be excluded. Finally, the interpretation of data derived from such a heterogeneous cohort including different CM phenotypes must

be done with caution. In this regard, we eagerly await the analysis focusing on hypertrophic (HCM) and dilated phenotypes. Furthermore, this work offers some points for reflection. As indicated by CCI, the patients included in this study had several comorbidities and a higher score accounted for worse outcomes. A recent analysis of the EURObservational Research Programme Cardiomyopathy registry, designed by the European Society of Cardiology, evaluated the association between common cardiovascular risk and severity of HCM phenotype [3]. Although the study was not designed to assess the impact of traditional risk factors on outcomes, the authors reported significant associations of obesity, hypertension, and diabetes with left ventricular hypertrophy, higher New York Heart Association class, higher left ventricular outflow tract gradients, and atrial fibrillation. Collectively, CMs represent a significant clinical burden to healthcare systems due to their increasing prevalence and poor prognosis. Similar data have been observed in cohorts including either different CM phenotypes or exclusively obstructive HCM [4, 5]. The increasing number of hospitalizations and surgical procedures results in increased costs. In addition, frequent delays in diagnosis add to the financial burden [6]. Furthermore, the heterogeneous course of CM necessitates lifelong follow-up and the implementation of a framework between primary care and third level centers [7, 8]. Indeed, a previous analysis from the Sarcomeric Human Cardiomyopathy Registry showed that most events, especially heart failure and atrial fibrillation, occur after the fifth decade, regardless of age at diagnosis [7]. The importance of continued follow-up and communication between different levels of care was recently highlighted by Garmany et al [8]. 2058 patients studied at the Mayo Clinic reported high rates of hospitalization and cardiac-related procedures outside of tertiary care, emphasizing the need for continuity of care, especially for patients considered at high risk. It is clear that CMs represent a major health burden that requires multiple interventions at different levels: 1) Management of CMs is no longer the prerogative of CMs specialists. Clinicians need to be aware of CMs as a possible underlying cause of using a "cardiomyopathy-mindset" as suggested by the recent comprehensive European Society of Cardiology guidelines [9]; 2) Patients and caregivers education is crucial for shared decision making; 3) Finally, implementation and consolidation of an infrastructure that incentivizes research and continuity of care is paramount to address the unmet needs of this complex population.

Article information

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REFERENCES

1. Masarone D, Kaski JP, Pacileo G, et al. Epidemiology and clinical aspects of genetic cardiomyopathies. Heart Fail Clin. 2018; 14(2): 119–128, doi: 10.1016/j.hfc.2017.12.007, indexed in Pubmed: 29525641.
2. Mizia-Stec K, Grzybowski J, Cegłowska U, et al. Treatment pathways defined as the sequence of visits to the public health system of patients with cardiomyopathies in Poland in the period 2016–2021. Pol Heart J. 2024; 82(5): 500–506, doi: 10.33963/v.phj.100178, indexed in Pubmed: 38606740.
3. Lopes LR, Losi MA, Sheikh N, et al. Association between common cardiovascular risk factors and clinical phenotype in patients with hypertrophic cardiomyopathy from the European Society of Cardiology (ESC) EurObservational Research Programme (EORP) Cardiomyopathy/Myocarditis registry. Eur Heart J Qual Care Clin Outcomes. 2022; 9(1): 42–53, doi: 10.1093/ehjqcco/qcac006, indexed in Pubmed: 35138368.
4. Lannou S, Mansencal N, Couchoud C, et al. The public health burden of cardiomyopathies: Insights from a nationwide inpatient study. J Clin Med. 2020; 9(4): 920, doi: 10.3390/jcm9040920, indexed in Pubmed: 32230881.
5. Butzner M, Maron M, Sarocco P, et al. Healthcare resource utilization and cost of obstructive hypertrophic cardiomyopathy in a US population. Am Heart J Plus. 2022; 13: 100089, doi: 10.1016/j.ahjo.2022.100089, indexed in Pubmed: 38560082.
6. Naidu SS, Sutton MB, Gao W, et al. Frequency and clinicoeconomic impact of delays to definitive diagnosis of obstructive hypertrophic cardiomyopathy in the United States. J Med Econ. 2023; 26(1): 682–690, doi: 10.1080/13696998.2023.2208966, indexed in Pubmed: 37170479.
7. Ho CY, Day SM, Ashley EA, et al. Genotype and lifetime burden of disease in hypertrophic cardiomyopathy: Insights from the Sarcomeric Human Cardiomyopathy Registry (share). Circulation. 2018; 138(14): 1387–1398, doi: 10.1161/CIRCULATIONAHA.117.033200, indexed in Pubmed: 30297972.
8. Garmany R, Bos JM, Ommen SR, et al. Clinical course of patients with hypertrophic cardiomyopathy away from tertiary referral care. ESC Heart Fail. 2023; 10(3): 1919–1927, doi: 10.1002/ehf2.14345, indexed in Pubmed: 36987533.
9. Arbelo E, Protonotarios A, Gimeno JR, et al. 2023 ESC Guidelines for the management of cardiomyopathies. Eur Heart J. 2023; 44(37): 3503–3626, doi: 10.1093/eurheartj/ehad194, indexed in Pubmed: 37622657.