Clinical profiles and incident heart failure in cardiomyopathies: a population-based linked electronic health record cohort study

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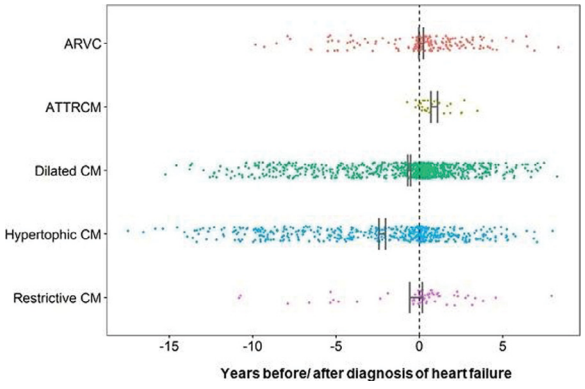
Background: Cardiomyopathies frequently cause heart failure (HF), however their prevalence in the general population and the natural history of incident HF across the spectrum of cardiomyopathy phenotypes is poorly understood. Improved understanding will help guide rational selection of diagnostic tests and accelerate the recognition of underlying causes of HF.

Purpose: To estimate the prevalence of cardiomyopathies using electronic health records; to compare clinical characteristics between patients with cardiomyopathy phenotypes; and to describe the temporal relationship between diagnosis of cardiomyopathy and incident HF.

Methods: A population-based cohort of patients with cardiomyopathy (n=4058) was provided by the UK Clinical Practice Research Datalink (CPRD) from a denominator sample of ~9 million individuals. Patients were phenotyped into groups according to ESC criteria: hypertrophic cardiomyopathy (HCM), dilated cardiomyopathy (DCM), arrhythmogenic right ventricular cardiomyopathy (ARVC) and restrictive cardiomyopathy (RCM). An additional group of transthyretin amyloid cardiomyopathy (ATTR-CM) was reported separately. Point prevalence was estimated for each cardiomyopathy subtype and clinical characteristics defined. An index date at first diagnosis of HF was determined for each patient and the time from/to first diagnosis of cardiomyopathy calculated relative to the index date and presented graphically.

Results: DCM was the most common cardiomyopathy phenotype among women and men with 3.4 and 7.7 cases per 10,000 population, respectively. The 2-fold increase in prevalence among men was consistent across DCM, HCM and RCM; the reverse trend was observed for ARVC which was found in 2.3 per 10,000 women and 1.1 per 10,000 men. At the time of first diagnosis of cardiomyopathy, most patients with ATTR-CM (73.5%), DCM (71.0%) and RCM (71.3%) had pre-existing HF though this proportion fell to 41.0% in ARVC and 31.0% in HCM. In relation to incident HF, a diagnosis of HCM and DCM were recorded earliest at a mean –2.2 years (SE 0.2) and –0.6 years (SE 0.1), respectively. We observed a clustering of diagnoses of RCM (mean –0.2 years, SE 0.4) and ARVC (mean 0.1 years, SE 0.1) around the time of onset of heart failure, whereas a diagnosis of ATTR-CM was first recorded at a mean of 0.9 years (SE 0.2) following the onset of heart failure.

Conclusions: Most diagnoses of ATTR-CM, DCM and RCM were preceded by clinical expression of HF whereas most people with ARVC or HCM developed HF after their cardiomyopathy diagnosis. Our findings in ARVC and HCM suggest a more indolent course with respect to cardiac function or better recognition in an asymptomatic phase. The clustering of a diagnosis of heart failure around the time of diagnosis of cardiomyopathy highlights a need for greater awareness of specific aetiologies of heart failure in routine practice and suggests opportunities for presymptomatic or earlier diagnosis.



Temporality of HF in cardiomyopathies