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Prevalence and incidence of dilated cardiomyopathy in the United States, France, Germany, Italy, Spain, and the United Kingdom: a systematic literature review

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Introduction

- Dilated cardiomyopathy (DCM) is a heart muscle condition characterized by ventricular dilation and depressed myocardial performance without hypertension, valvular, congenital, or ischemic heart disease[1]
  - DCM is a major cause of heart failure,[2] a leading indication for heart transplant[3]
  - It can be caused by genetics, viral infections, alcohol, pregnancy, and metabolic disorders; however, DCM with no known cause (idiopathic DCM) is common[2]

Objectives

- This review aimed to synthesize, from the published literature, the prevalence and incidence of DCM in the general population within the United States (US), France, Germany, Italy, Spain, and the United Kingdom (UK)

Methods

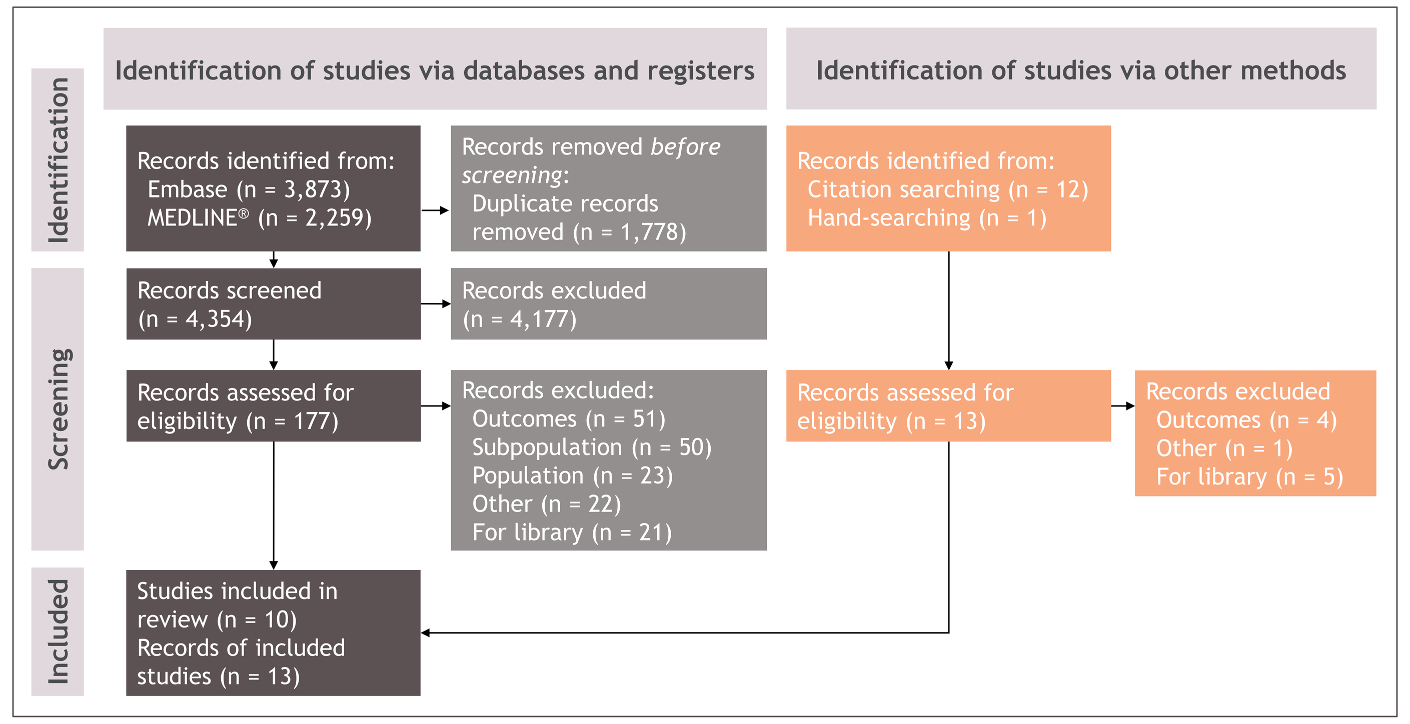
- A systematic literature review was conducted by searching MEDLINE® and Embase from database inception to May 9, 2023
  - Manual searches of relevant conferences within the last two years and bibliographies of previous literature reviews were also conducted
- English-language studies reporting prevalence or incidence of DCM within general populations of adults or children in countries of interest were included
- Investigators assessed the quality of the included studies using the Newcastle-Ottawa Scale[4] for case-control or cohort studies and the Joanna Briggs Institute checklist for cross-sectional studies[5]

Results

Study selection

- Of 6,144 identified articles, 10 unique studies (13 publications) were included in the review (**Figure 1**)

Figure 1. PRISMA diagram of study selection



n - number of records

Study characteristics and study quality

- Seven studies were retrospective cohorts,[6-15] three were cross-sectional surveys[16-18]
- Most studies were conducted in the US (number of studies [n] = 5),[6,8,9,11,13-16]
  - Other countries included the UK (n = 3),[7,10,18] Italy (n = 1),[12] and France (n = 1)[17]
- Four studies reported prevalence or incidence for adults[7,9,10,12]
- Three studies reported on pediatric populations[6,11,13-16]
- Four studies reported heterogeneous populations (both adults and children)[6,8,17,18]
- Sample sizes ranged from 5,169[16] - 74,212,292[11,13-15] participants (median: 326,441)

Prevalence of idiopathic DCM ranged from 8.3 to 59.2 per 100,000 persons within the US and key European countries

Annual incidence of idiopathic DCM ranged from 1.1 to 6.95 per 100,000 persons

Table 1. Prevalence of dilated cardiomyopathy within the general population, sorted by region. Purple **indicates idiopathic DCM**

Study	Study Features	Case Identification Criteria	Age Group	Prevalence per 100,000 (95% CI)
Ababio 2023[6]	National population-based electronic health records, **United States**; 2017-2019	≥1 inpatient DCM ICD-10 code or ≥2 outpatient DCM ICD-10 codes ≥30 days apart (data stratified by validated non-idiopathic DCM, idiopathic DCM, and total DCM)	All ages	118.33[a] 59.23[b] 127.92[c]
			Children ≤11 years	2.38[a] 1.19[b] 2.57[c]
			Children 12-17 years	2.62[a] 1.31[b] 2.83[c]
Codd 1989[8]	Population-based health records of Olmsted County, Minnesota, **United States**; 1975-1984	Idiopathic DCM as per WHO/ISFC Task Force	All ages	36.5 (23.1-50.0)
Angelini 2018[16]	Schoolchildren of Harris County, Houston, Texas, **United States**; 2010-2017	LVEF <0.40-0.45 and size measurements >1 standard deviation from normal (idiopathic and non-idiopathic DCM)	Children 11-18 years	210
Lannou 2020[17]	National inpatient database, **France**; 2008-2015	ICD-10 including idiopathic and non-idiopathic DCM	All ages	42.84
Brown-rigg 2022[7]	Clinical Practice Research Datalink, **England**; 2010-2018	ICD-10 including idiopathic and non-idiopathic DCM	Adults ≥18 years	2010: 57 2018: 43
Williams 1985[18]	General practitioner survey of East Anglia, Essex, Hertfordshire & Bedfordshire, **England**; 1983-1984	Idiopathic DCM as per WHO/ISFC	All ages	8.32

[a] Validated DCM; [b] Idiopathic DCM; [c] Total DCM; CI - Confidence interval; DCM - Dilated cardiomyopathy; ICD - International Classification of Diseases; ISFC - International Society and Federation of Cardiology; LVEF - Left ventricular ejection fraction; WHO - World Health Organization

Table 2: Incidence of dilated cardiomyopathy within the general population, sorted by region. Purple **indicates idiopathic DCM**

Study	Data Source; Time Period	Case Identification Criteria	Age Group	Annual Incidence per 100,000 (95% CI)
Codd 1989[8]	Population-based health records of Olmsted County, Minnesota, **United States**; 1975-1984	Idiopathic DCM as per WHO/ISFC Task Force	All ages	6.0 (4.2-7.7)[a] 2.5 (1.4-3.7)[b]
Coughlin 1993[9]	Washington County Hospital, **United States**; 1975-1991	Idiopathic DCM as per ICD-9	Men	1.5 (0.8-2.24)
			Women	1.1 (0.5-1.8)
Lipshultz 2003[11]; Wilkinson 2010[14]; Towbin 2006[13]; Wilkinson 2008[15]	Pediatric Cardiomyopathy Registry, New England & Central Southwest **United States**; 1996-1999[11,14] and 1996-2002[13,15]	"Pure" DCM, including idiopathic and non-idiopathic, excluding additional overlapping cardiac phenotypes	Children <18 years	1996-1999: 0.58 (0.51-0.65) 1996-2002: 0.57 (0.52-0.63)
Rakar 1997[12]	Cardiomyopathies registry and post-mortem database, Trieste, **Italy**; 1987-1989	Idiopathic DCM as per WHO/ISFC	Adults	6.95
Herd 1991[10]	Bangour General Hospital, Dechmont, West Lothian, **Scotland**; 1982-1986	Not reported	Adults	7.9

[a] Age-adjusted incidence; [b] Sex-adjusted incidence; CI - Confidence interval; DCM - Dilated cardiomyopathy; ICD - International Classification of Diseases; ISFC - International Society and Federation of Cardiology; WHO - World Health Organization

Participant characteristics

- Mean/median age of DCM onset ranged from 54[8] - 70 years[17] (n = 4; median: 58)
- Males among DCM patients ranged from 57.7[9] - 73.3%[8] (median: 65.0%)
- Studies were generally considered high quality, attributable to the use of clearly described multi-center hospital data or population-based data retrieved from national electronic health records or large-scale registries
- Case identification criteria varied across studies, and included International Classification of Diseases (ICD) codes, World Health Organization/International Society and Federation of Cardiology (WHO/ISFC) criteria, and bespoke criteria
  - Criteria also varied with regards to idiopathic versus non-idiopathic DCM

Prevalence of dilated cardiomyopathy

- Six studies reported prevalence of DCM,[6-8,16-18] (**Table 1**)
  - Prevalence of DCM due to any cause (idiopathic or non-idiopathic) ranged from 42.8[17] - 118.3[6] per 100,000 persons in adult/heterogeneous populations across countries
  - Prevalence estimates of idiopathic DCM ranged from 8.3[18] - 59.2[8] per 100,000 persons in adult/heterogeneous populations across countries
- Prevalence data for pediatric patients were scarce (n = 2):
  - DCM prevalence was between 2.6[6] and 210[16] cases per 100,000 persons within the US for children aged ~11-18 years

Incidence of dilated cardiomyopathy

- Five studies reported on incidence of DCM[8-15] (**Table 2**)
  - Annual incidence of idiopathic DCM ranged from 1.1[9] - 7.0[12] per 100,000 persons in adult/heterogenous populations across countries
- Incidence of pediatric DCM was reported by four articles[11,13-15] all using a cohort from the Pediatric Cardiomyopathy Registry sponsored by the National Heart, Lung, and Blood Institute at two different time points (1996-1999[11,14] and 1996-2002[13,15])

Discussion

- Large variability was observed across reported prevalence and incidence estimates
- Reasons for these observed discrepancies are numerous, and include:
  - Differences in the definitions of DCM (e.g., idiopathic vs. non-idiopathic DCM)
  - Varying methodologies (e.g., study design, sample size)
  - Varying data sources (e.g., population-based data sources vs. hospital databases)
  - Differing demographics of the patient populations (e.g., age, sex, race)
  - Differences in methods of DCM case ascertainment

Conclusions

- This review identified varied prevalence and incidence rates of DCM across different geographic areas and time periods
- Contributing factors to this variation include the sources of population data, the definition of DCM, methodologies used to identify and ascertain cases of DCM, and study populations when comparing prevalence or incidence rates across regions
- Lack of reliable data on pediatric DCM and separation of data by idiopathic vs. non-idiopathic DCM are unmet needs
- Future studies should standardize the selection criteria based on the established definitions of DCM
- As well, since prevalence estimates may vary across different population demographics, more granular reporting based on patient characteristics is needed

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