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Review

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Prevalence and Incidence of Dilated Cardiomyopathy in the United States and Western Europe: A Systematic Review

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Abstract

Background: Dilated cardiomyopathy (DCM) is a major contributing factor for heart failure and cardiac transplantation worldwide. Estimating the prevalence and incidence of DCM is critical for understanding the burden of illness in these patients and improving the landscape of preventative treatments. Previous reviews have shown substantial prevalence and incidence estimates for DCM within key regions such as the United States and several European countries. This review aimed to describe the published evidence on the prevalence and incidence of DCM within the United States, France, Germany, Italy, Spain, and the United Kingdom.

Methods: MEDLINE® and Embase were searched from database inception to May 9, 2023 for English-language studies reporting the prevalence or incidence of DCM within general populations of adults or children in countries of interest. Manual searches of relevant conferences and bibliographies of previous literature reviews were also conducted.

Results: Of 6,145 identified articles, 10 unique studies were included in the review. Six studies reported prevalence, and five studies reported incidence of DCM in various populations. Prevalence estimates of DCM, including idiopathic and non-idiopathic causes, within adults (≥ 18 years) and/or heterogeneous (all ages) populations ranged from 42.8 to 118.3 per 100,000 persons; idiopathic DCM estimates ranged from 8.3 to 59.2 per 100,000 persons. Prevalence of adolescent (about 11 - 18 years) DCM, including idiopathic and non-idiopathic causes, ranged from 2.6 to 212.8 per 100,000 persons. Annual incidence rates of idiopathic DCM in adult/heterogeneous populations ranged from 6.0 to 7.0 per 100,000 persons. Annual incidence of DCM due to idiopathic/non-idiopathic causes among pediatric populations was reported as 0.6 per 100,000 persons. Reported prevalence and incidence rates by sex showed male preponderance, and estimates were higher in Black persons compared with White and Hispanic persons; higher DCM prevalence estimates were observed in studies utilizing newer DCM definitions using ICD coding compared with older definitions.

Conclusion: This study highlights the varied prevalence and incidence rates of DCM reported across different geographic locations, time periods, sexes, races, and disease definitions. When comparing these rates, it is crucial to consider factors such as data sources, case definitions, case-finding methodologies, and study populations.

Keywords: Dilated cardiomyopathy; Prevalence; Incidence; Epidemiology; Systematic review

Introduction

Dilated cardiomyopathy (DCM) is a phenotype within the hierarchy of cardiomyopathies, defined as left ventricular dilatation and global or regional systolic dysfunction unexplained solely by abnormal loading conditions or significant coronary artery disease sufficient to explain the myocardial impairment [1]. DCM is also one of the main causes of heart failure [2]. Within DCM, heart failure with reduced ejection fraction has high prevalence and incidence and represents the most frequent cause of death, despite improvements in treatment. In addition, advanced heart failure in DCM is one of the leading indications for heart transplantation [3].

The DCM phenotype can be broadly subdivided into idiopathic (of unknown cause) and non-idiopathic (e.g., ischemic or acquired DCM) forms based on clinical or lifestyle-related factors. Idiopathic DCM may often present as familial disease; estimates obtained by screening relatives of patients with DCM identified probable familial disease in about 20-35% of cases [4]. Published studies have already identified more than 50 genes associated with DCM [5]. Factors associated with non-idiopathic DCM include drugs and toxins (most notably excess alcohol consumption), myocarditis, tachycardia, infectious or inflammatory causes, hypertension, and peripartum cardiomyopathy [4].

The etiological diversity of the DCM phenotype presents methodological difficulties for epidemiological studies on the prevalence and incidence of DCM. The most commonly cited sources have reported the prevalence of age-adjusted idiopathic DCM in the United States (US) to be 36.5/100,000

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persons within the general population, and incidence rates of 6.0/100,000 person-years [6]. Notably, the study reporting this prevalence estimate was conducted between the years 1975 and 1984, and is limited to individuals from Olmsted County, Minnesota. More recent estimates from reviews based on clinical trial data predict the prevalence of DCM to be approximately 400 per 100,000 people within North America [7]; the annual incidence in Europe and North America ranges between 5 and 7.9 cases per 100,000 people. Furthermore, this disorder accounts for around 60% of childhood cardiomyopathies, with infants younger than 12 months having the highest incidence [8].

The review by Hershberger et al 2013 [9] underscores that the true prevalence of DCM may be significantly underestimated, particularly in its early or subclinical stages. While traditional estimates suggest a prevalence of approximately 40 per 100,000 people, population-based genetic studies have revealed that pathogenic variants associated with DCM are present in as many as 400 individuals per 100,000 in the general population. This discrepancy suggests that many individuals with genetic predisposition to DCM may remain undiagnosed due to incomplete penetrance, lack of clinical screening, or mild/asymptomatic phenotypes.

Determining the prevalence and incidence of DCM has been challenging not only because of the inherent heterogeneity of the phenotype but also due to geographic variations, patient selection, and the evolution of diagnostic criteria over time [10]. However, prior reviews have shown substantial prevalence and incidence estimates for DCM within the US and select countries in Europe [11]. Various global burden studies have described higher prevalence estimates for cardiomyopathies in North America and Western Europe for both adults and children [12, 13]. Moreover, research and development initiatives have supported an increase in emerging treatments for DCM, particularly in US and European region. Moreover, these regions presented similar health system characteristics, data availability, and surveillance quality, thus enabling meaningful comparison. In light of this, the present systematic literature review aimed to characterize the published evidence on the prevalence and incidence of DCM within general populations of the US and select European countries, namely, France, Germany, Italy, Spain, and the United Kingdom (UK).

Methods

Standard methodologies for conducting and reporting systematic reviews as recommended by the Cochrane Handbook for Systematic Reviews of Interventions [14] were followed. Results from the review were reported according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines [15]. Relevant studies were identified by searching MEDLINE® and Embase via OvidSP from database inception to May 9, 2023, using predefined search strategies (Supplementary Materials 1 and 2, cr.elmerpub.com). Database search strategies were designed to be comprehensive and sensitive. Medical subject headings (MeSH) and Emtree terms, as well as "exploded" terms (e.g., exp incidence/), were used where possible to systematically capture all relevant publications, and reviewers manually screened through all identified records to determine their relevance to the research question. Additionally, we searched select conferences and the bibliographies of included literature reviews. Eligible studies for inclusion were observational studies reporting prevalence or incidence of DCM within general populations of adults or children in countries of interest (i.e., US, France, Germany, Italy, Spain, and the UK). Additionally, studies were excluded if they did not report on the incidence or prevalence of DCM within the general population. Publications not available in the English language were excluded. Observational studies of interest included cohort studies, case-control studies, and cross-sectional studies. A senior reviewer was responsible for reviewing abstracts and conference proceedings according to the pre-defined selection criteria.

All eligible studies identified during title/abstract screening proceeded to the full-text screening phase, where they were assessed for eligibility by two independent reviewers. During the full-text screening phase, reviewers reconciled differences between their inclusion decisions. A third reviewer intervened to reach consensus on any unresolved discrepancies. Studies that matched the inclusion criteria following the full-text screening were included for data extraction. Lists of records that were included and excluded after full-text screening are provided in Supplementary Materials 3 and 4 (cr.elmerpub.com), respectively. Investigators assessed the quality of the included studies using the Newcastle-Ottawa Scale [16] for case-control or cohort studies and the Joanna Briggs Institute checklist for cross-sectional studies [17].

A meta-analysis was not considered appropriate for this review after assessing heterogeneity in the disease definition, study design, and data collection methods. Thus, a descriptive summary of the evidence was carried out. Where necessary, data provided by included studies were used to calculate denominators or numerators. When studies offered both crude and adjusted rates, the adjusted rate (e.g., adjusted for sex and age) was selected for presentation. Reported rates were converted to rates per 100,000 persons as needed.

Results

Study selection and study characteristics

In total, 6,145 abstracts were identified from the searches including 6,132 records via MEDLINE® and Embase, and 13 records through manual searches (Fig. 1). Of these, 1,778 duplicate records (about 30% of records), captured in both Embase and MEDLINE®, were removed prior to screening. Common reasons for exclusion at the screening stage were: population (about 30% of exclusions), in which case the study was focused on another condition besides DCM, such as heart failure, heart transplant, arrythmia, or sudden cardiac death; and study design (about 35% of exclusions), where a captured study was not an epidemiological study and did not report prevalence or incidence data (e.g., clinical trials, case reports, studies on risk factors). Additionally, approximately 1% of excluded studies

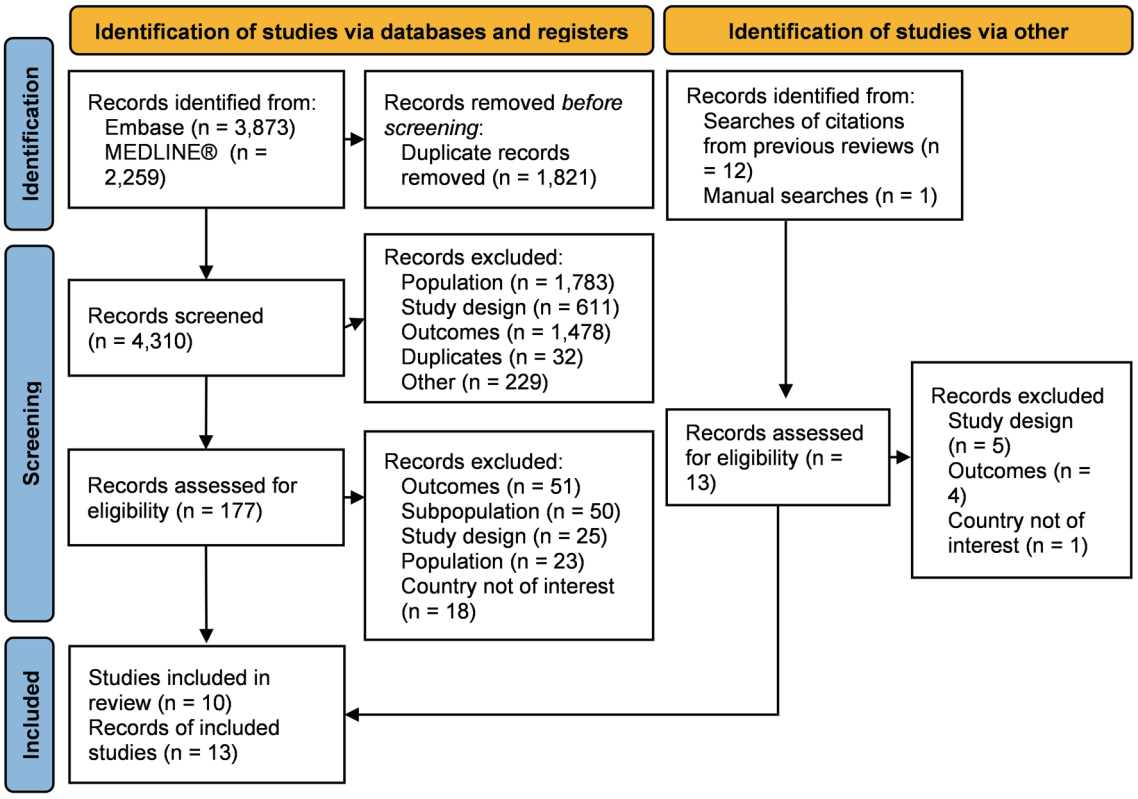

Figure 1. PRISMA flow diagram. PRISMA: Preferred Reporting Items for Systematic Reviews and Meta-Analyses.

were omitted because they reported the proportion of patients with DCM within specific subpopulations, such as individuals with diabetes, alcoholism, or those who had undergone heart transplantation, rather than reporting incidence or prevalence estimates in the general population. Following full-text screening, a total of 10 unique studies [6, 18-29] pertaining to 13 publications were included in the review.

Of the 10 studies included in review, seven were retrospective cohort studies [6, 18, 20-22, 24-28] and three were cross-sectional surveys [19, 23, 29]. Most studies were conducted in the US (number of unique studies (n) = 5) [6, 18, 19, 21, 24, 26-28], followed by the UK (n = 3) [20, 22, 29], Italy (n = 1) [25], and France (n = 1) [23]. Three studies reported relevant epidemiology outcomes for adult populations [20, 22, 25], three unique studies reported on pediatric populations [18, 19, 24, 26-28], and five reported heterogeneous populations (both adults and children) [6, 18, 21, 23, 29]. All studies focused on incidence or prevalence of DCM as the primary endpoints of analysis. A summary of the study and participant characteristics is presented in Table 1.

Of the nine studies reporting, the total sample sizes (denominators) of the population within which DCM cases were identified ranged from 5,169 [19] to over 74 million participants [24, 26-28], with a median of 589,579 participants. Only one study investigated a population of exclusively adults (≥ 18 years) [20]. Among two reporting studies, roughly half of their population consisted of male participants [18, 19]. Participants were majority White in two studies [18, 21], and majority Hispanic in a third study [19]; remaining studies did not report population demographic data.

Prevalence of DCM

Of the 10 studies included in the present review, six studies reported data for prevalence of DCM [6, 18-20, 23, 29].

Adult and heterogeneous populations

Five studies [6, 18, 20, 23, 29] reported data for the prevalence of DCM in the adult/heterogeneous population (Fig. 2). Prevalence estimates of DCM, including both idiopathic and non-idiopathic causes, ranged from 42.8 [23] to 118.3 [18] per 100,000 persons within any country. Prevalence estimates of idiopathic DCM within adults/heterogeneous populations ranged from 8.3 [29] to 59.2 [18] per 100,000 persons across countries. Case identification criteria varied across studies, and included ICD codes, World Health Organization/International Society and Federation of Cardiology (WHO/ISFC) Task Force criteria, as well as bespoke criteria.

Notably, the upper bound of the prevalence range for validated DCM including both idiopathic and non-idiopathic causes (118.3 per 100,000 individuals) was reported by Ababio

Table 1. Characteristics of Included Studies and Their Populations

Study	Study design	Data source; time period	DCM definition	N, total population	Age group	Age, total population	Males, total population	Race, total population
Ababio 2023 [18]	Retrospective cohort	National population-based electronic health records, US; 2017 - 2019	≥ 1 inpatient DCM ICD-10 code or ≥ 2 outpatient DCM ICD-10 codes ≥ 30 days apart (data stratified by idiopathic DCM, non-idiopathic DCM, and total DCM)	56,812,806	Heterogeneous; pediatric	≤ 11 years: 13.1%; 12 - 17 years: 6.0%; 18 - 29 years: 15.3%; 30 - 49 years: 25.6%; 50 - 64 years: 21.4%; ≥ 65 years: 18.4%; missing: 0.1%	44.6%	White: 66.9%; Black: 10.4%; Asian: 2.2%; missing: 20.5%
Angelini 2018 [19]	Cross-sectional	Schoolchildren of Harris County, Houston, Texas, US; 2010 - 2017	LVEF < 0.40 - 0.45 and size measurements >1 standard deviation from normal, including idiopathic and non-idiopathic DCM	5,169	Pediatric	11 - 14 years: 83.4%; 15 - 18 years: 16.6%	56%	Asian: 7%; Black: 18.5%; Hispanic: 41.8%; White: 31.4%; other: 1.3%
Codd 1989 [6]	Retrospective cohort	Population-based health records of Olmsted County, Minnesota, US; 1975 - 1984	Idiopathic DCM as per WHO/ISFC Task Force	98,423*	Heterogeneous	NR	NR	NR
Coughlin 1993 [21]	Retrospective cohort	Washington County Hospital, Maryland, US; 1975 - 1991	Idiopathic DCM as per ICD-9	121,393	Heterogeneous	NR	NR	White: ~93%
Lipshultz 2003 [24]; Wilkinson 2010 [27]; Towbin 2006 [26]; Wilkinson 2008 [28]	Retrospective cohort	Pediatric Cardiomyopathy Registry, New England & Central Southwest US; 1996 - 1999 [24, 27] and 1996 - 2002 [26, 28]	"Pure" DCM, including idiopathic and non-idiopathic, excluding additional overlapping cardiac phenotypes	74,212,292	Pediatric	NR	NR	NR
Brownrigg 2022 [20]	Retrospective cohort	Clinical Practice Research Datalink, England; 2000 - 2018	ICD-10 including idiopathic and non-idiopathic DCM	about 9,000,000	Adults	≥ 18 years: 100%	NR	NR
Herd 1991 [22]	Retrospective cohort	Bangour General Hospital, Dechmont, West Lothian, Scotland; 1982 - 1986	NR	145,000	Adults	NR	NR	NR
Williams 1985 [29]	Cross-sectional	General practitioner survey of East Anglia, Essex, Hertfordshire and Bedfordshire, England; 1983 - 1984	Idiopathic DCM as per WHO/ISFC Task Force	913,836	Heterogeneous	NR	NR	NR
Lannou 2020 [23]	Cross-sectional	National inpatient database, France; 2008 - 2015	ICD-10 including idiopathic and non-idiopathic DCM	97,300,000 hospitalizations	Heterogeneous	NR	NR	NR
Rakar 1997 [25]	Retrospective cohort	Cardiomyopathies registry and post-mortem database, Trieste, Italy; 1987 - 1989	Idiopathic DCM as per WHO/ISFC Task Force	265,321	Adults	NR	NR	NR

*Sample size reflects the population size of Olmsted County as of January 1, 1985, as reported by the United States Census Bureau. DCM: dilated cardiomyopathy; ICD: International Classification of Diseases; LVEF: left ventricular ejection fraction; NR: not reported; US: United States; WHO/ISFC: World Health Organization/International Society and Federation of Cardiology.

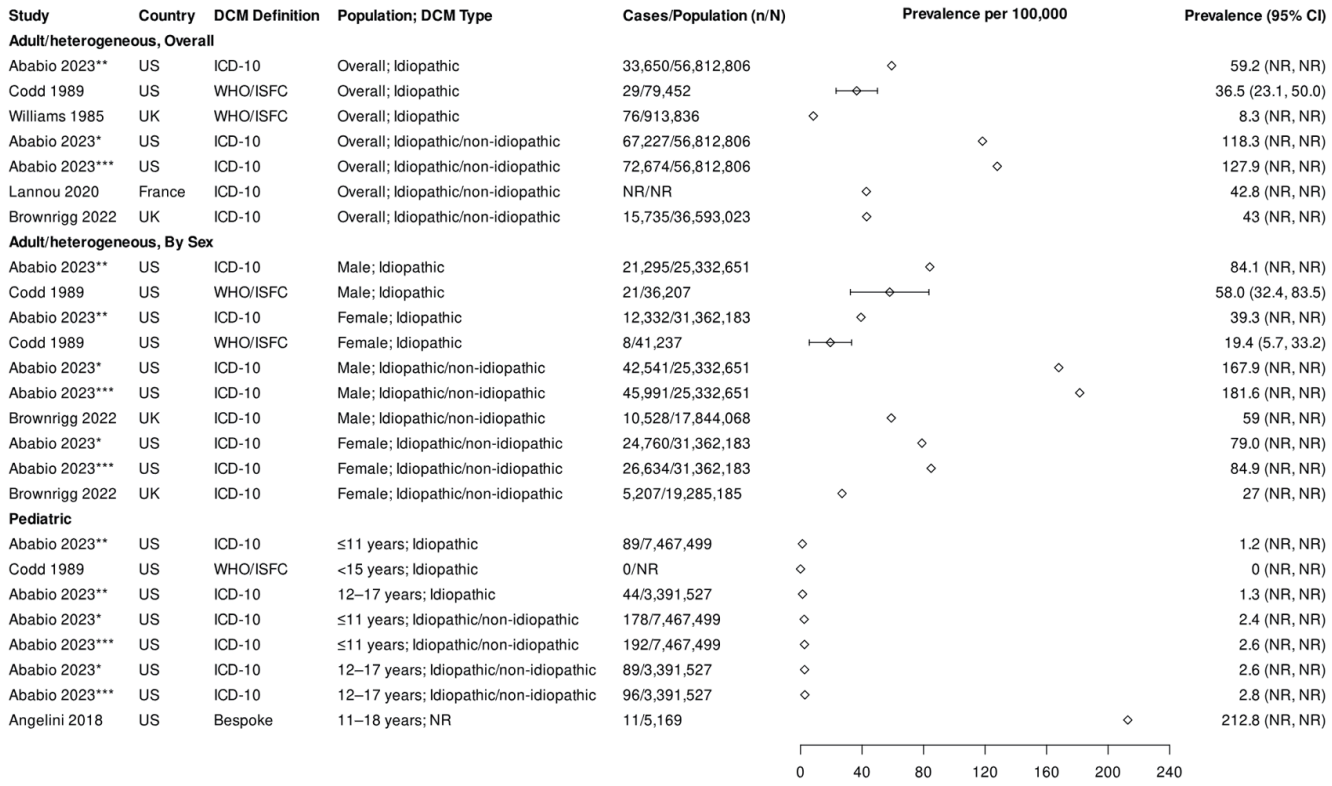

Figure 2. Prevalence of dilated cardiomyopathy, sorted by age group and sex. *Validated dilated cardiomyopathy. Investigators performed clinical validation of cases of dilated cardiomyopathy by choosing a random sample of 1,000 suspected cases in the database, and verifying the diagnosis based on clinical notes. **Idiopathic dilated cardiomyopathy. ***Total dilated cardiomyopathy (idiopathic and non-idiopathic). Please note all prevalence values are expressed per 100,000. CI: confidence interval; DCM: dilated cardiomyopathy; ICD: International Classification of Diseases; NR: not reported; UK: United Kingdom; US: United States; WHO/ISFC: World Health Organization/International Society and Federation of Cardiology.

et al (2023) [18], who reported DCM prevalence using various definitions. This study analyzed de-identified retrospectively collected data from the Optum® Electronic Health Records database, a large, racially diverse population with clinical encounter data for over 101 million patients from a network of 700 hospitals and 7,000 clinics across all 50 states in the US as of September 30, 2019. Investigators performed a clinical validation of DCM cases by choosing a random sample of 1,000 suspected cases of DCM in the database, and verifying the diagnosis based on clinical notes. Validated DCM accounted for 118.3 DCM cases per 100,000 persons, while idiopathic DCM accounted for 59.2 cases per 100,000 (total DCM including idiopathic and non-idiopathic DCM was 127.9 per 100,000 persons).

The UK population-based cohort study by Brownrigg et al (2018) [20] studied prevalence of DCM from 2000 to 2018 utilizing the UK Clinical Practice Research Datalink research database. Authors reported an overall increase in the prevalence estimates of DCM through these years, with a peak in 2010 (57 per 100,000 individuals) and decreasing in 2018 (43 per 100,000 individuals). Of note, authors observed a decline in the prevalence of DCM cases during this period (2010 - 2018) and an increase in the prevalence of arrhythmogenic right ventricular cardiomyopathy cases. A potential reason for this increase, as reported by the authors, is due to specific diagnostic imaging parameters introduced by the Task Force Criteria in 2010 as well as the expanded use of cardiac magnetic resonance, all of which may have increased overall detection.

A substantial shift in prevalence estimates was observed across studies with the use of different diagnostic criteria for DCM. For idiopathic DCM, higher prevalence estimates using newer ICD codes (59.2 [18] per 100,000 persons) were observed in comparison with those using older WHO/ISFC Task Force diagnostic criteria (8.3 [29] to 36.5 [6] per 100,000 persons). In addition, three studies [6, 18, 20] reported higher DCM prevalence estimates in males compared with females (Fig. 2). Two studies reported age-specific prevalence estimates, with older populations showing significantly higher prevalence compared with younger ones [6, 18]. Moreover, Ababio et al (2023) [18] reported prevalence categorized by race, with higher estimates observed in Black persons followed by White and Asian persons (106.9, 66.0, and 22.4 per 100,000 persons for idiopathic DCM, respectively). Trends were similar for validated DCM cases to total DCM cases (idiopathic and non-idiopathic; data not shown). Furthermore, Lannou et al (2020) [23] reported an increase in the prevalence of DCM from 419.9 (2008) to 472.8 (2015) per 100,000 individuals.

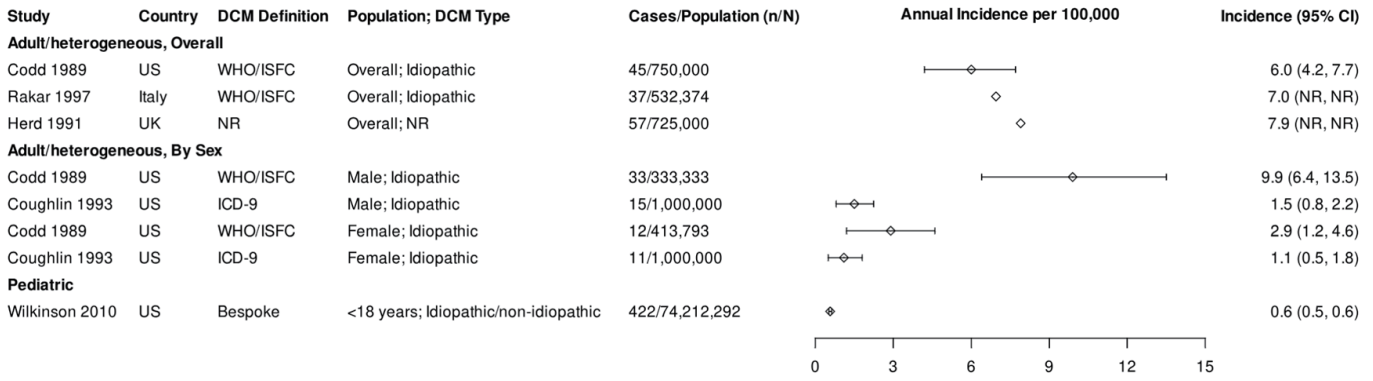

Figure 3. Annual incidence of dilated cardiomyopathy, sorted by age group and sex. Please note all incidence values are expressed per 100,000. CI: confidence interval; DCM: dilated cardiomyopathy; ICD: International Classification of Diseases; NR: not reported; UK: United Kingdom; US: United States; WHO/ISFC: World Health Organization/International Society and Federation of Cardiology.

Pediatric populations

Three studies reported data for the prevalence of DCM in the pediatric population of the US (Fig. 2) [6, 18, 19]. All studies utilized different criteria for diagnosis of DCM patients. Ababio et al (2023) [18] utilized ICD-10 code I42.0 while Angelini et al (2018) [19] used left ventricular ejection fraction < 0.40 - 0.45 and size measurements > 1 standard deviation from normal as the basis for DCM diagnosis, and Codd et al (1989) [6] utilized WHO/ISFC diagnostic criteria.

In Ababio et al (2023), pediatric DCM prevalence estimates showed variation across types of DCM: validated DCM had a prevalence of 2.4 and 2.6 per 100,000 persons, idiopathic DCM showed 1.2 and 1.3 cases per 100,000 persons, and total DCM showed 2.6 and 2.8 cases per 100,000 persons for children aged ≤ 11 years and 12 - 17 years, respectively [18]. Older children showed higher prevalence estimates in comparison with younger ones. Angelini et al (2018) estimated the prevalence of DCM as 212.8 per 100,000 persons aged 11 - 18 years [19]. Notably, their sample was drawn from a small population of schoolchildren of Harris County, Houston, Texas, and authors did not report whether cases included idiopathic or mixed idiopathic/non-idiopathic DCM. Codd et al (1989) reported no idiopathic DCM cases in those younger than 15 years of age [6].

Incidence of DCM

Five studies reported data for incidence of DCM in the general population [6, 21, 22, 24, 25], four of which reported for the adult/heterogeneous population (Fig. 3) [6, 21, 22, 25]. Few recent studies were identified, with all adult data originating from publications dated 1997 or earlier.

Adult and heterogeneous populations

Annual incidence estimates of idiopathic DCM were 6.0 [6] and 7.0 [25] per 100,000 persons in adult/heterogeneous populations within any country (Fig. 3). The lower estimate was reported by Codd et al (1989) [6], a population-based study of Olmsted County, Minnesota; authors utilized a central data bank of Mayo Clinic and the Rochester Epidemiology Project. The higher estimate was reported by Rakar et al (1997) [25], who utilized a registry for cardiomyopathies and a post-mortem database of a cardiology department in Trieste University, Italy from November 1987 to November 1989. Both studies used WHO/ISFC diagnostic criteria to ascertain DCM cases. For unspecified DCM (not reported as idiopathic or non-idiopathic), Herd et al (1991) conducted a retrospective review of all patients attending Bangour General Hospital, West Lothian, Scotland during the 5-year period from 1982 to 1986 [22]. By identifying DCM patients from clinic letters, discharge summaries, and post-mortem results, incidence was estimated as 7.9 per 100,000 persons.

In addition, two studies reported annual incidence rates of idiopathic DCM by sex (Fig. 3), with higher estimates in males (ranging from 1.5 [21] to 9.9 [6] per 100,000) compared with females (1.1 [21] to 2.9 [6] per 100,000). The greater variation in annual incidence estimates for both males and female could be attributed to the varying diagnostic criteria used. Lower estimates were reported by Coughlin et al (1993) [21] using ICD-9 criteria, while higher estimates were reported by Codd et al (1989) [6] using WHO/ISFC criteria.

Furthermore, Codd et al (1989) [6] reported a temporal increase in the incidence of DCM over the study period. The incidence rate rose from 3.9 per 100,000 person-years during the first 5 years (1975 - 1979) to 7.9 per 100,000 person-years in the final 5 years (1980 - 1984), effectively demonstrating a twofold increase over time.

Pediatric populations

Incidence of pediatric DCM was reported by four publications [24, 26-28] reporting on the same cohort from the Pediatric Cardiomyopathy Registry sponsored by the National Heart,

Lung, and Blood Institute (Fig. 3), albeit at two different time points (1996 - 1999 [24, 27] and 1996 - 2002 [26, 28]). This registry has collected data on all children with newly diagnosed cardiomyopathy in New England and the Central Southwest region of the US. Of the two publications which reported on all children in these regions who received this diagnosis between 1996 and 1999 [24, 27], the reported annual incidence of DCM (including both idiopathic and non-idiopathic causes) was 0.58 per 100,000 persons (95% confidence interval (CI): 0.51 - 0.65). Two subsequent publications [26, 28] followed the reporting from this registry, extending the reporting period until 2002. The updated value of the annual incidence of pediatric DCM was 0.57 per 100,000 (95% CI: 0.52 - 0.63). For this cohort, Towbin et al (2006) also reported higher incidence rates in males than females (male: 0.66 (95% CI: 0.58 - 0.75) vs. female: 0.47 (95% CI: 0.40 - 0.55)), and in Black persons (0.98 (95% CI: 0.78 - 1.21)) compared with Hispanic (0.58 (95% CI: 0.48 - 0.70)) and White persons (0.33 (95% CI: 0.29 - 0.38)) [26]. Lipshultz et al (2003) [24] also reported varying annual DCM incidence rates of 0.61, 0.71, 0.58, and 0.48 per 100,000 for the years 1996, 1997, 1998, and 1999, respectively.

Quality assessment

The risk of bias across the seven included cohort studies was assessed using the Newcastle-Ottawa Scale. It should be noted that incidence or prevalence of DCM was captured irrespective of follow-up time; thus, the domain assessing sufficiency of follow-up length was considered as nonapplicable in all studies reviewed. All studies reached the maximum score of 6/6 or 4/4. The high quality of these cohort studies was attributable to the adoption of multi-center hospital data or population-based data retrieved from national electronic health records or large-scale registries, thus having good representativeness of the exposed cohort in the community.

The risk of bias across the three cross-sectional studies was assessed using the Joanna Briggs Institute quality assessment tool. Studies were generally at low risk of bias, since all studies clearly defined their inclusion criteria, as well as provided detailed descriptions of their participants and study settings.

Discussion

To our knowledge, this is the first systematic review to investigate the prevalence and incidence of DCM within adults and children of general populations across the US and select European countries.

The most striking result of the reported prevalence and incidence of DCM is the large variability of the estimates across the studies. In the five studies reporting adult/heterogeneous DCM prevalence, we saw more than a six-fold increase in idiopathic DCM prevalence from the lowest [29] (8.3 per 100,000) to the highest [18] estimate (59.2 per 100,000) and three-fold increase in idiopathic/non-idiopathic DCM prevalence (42.8 [23] to 127.9 [18] per 100,000). Similar variability can be seen in the pediatric prevalence estimates, and to a lesser degree in the incidence estimates. There may be multiple reasons for these observed discrepancies. First, the clinical course of DCM can vary substantially across patients, mostly due to the variety of causes and triggers affecting the natural history of the disease [30]. Moreover, some symptoms of DCM overlap with other non-ischemic cardiomyopathies [10]; thus, having a correct diagnosis of DCM may be difficult to achieve.

Furthermore, differences in the definition and diagnostic criteria for DCM across studies may have contributed to the heterogeneity in DCM estimates. More recent studies analyzed data from national databases of the US, the UK, and France, which relied on the utilization of a diagnostic code (ICD-10-I42.0) for ascertainment of DCM cases [18, 20, 23]. However, older studies [6, 29] tended to ascertain DCM cases based on the WHO/ISFC Task Force recommendations on the definition and classification of cardiomyopathies and were carried out in specific regions of the country. Indeed, the definition of DCM in the literature has evolved over time [31], and a growing number of new diagnostic markers are being identified [10, 32]. The early definition of DCM as per the 1980 WHO/ISFC was merely a broad classification of phenotype or structural myocardial changes with unknown origins [33]; this definition was then updated in 1995 to include a more detailed understanding of genetic, infectious, and other specific causes [34]. In 2006, the American Heart Association (AHA) published a framework for grouping cardiomyopathies that presented a new perspective on DCM classification [35]. Their model considered DCM as a heterogeneous cardiomyopathy that can have both genetic and acquired etiology. Two years later, in 2008, an influential paper by the European Society of Cardiology (ESC) Working Group was published on the classification of cardiomyopathies [36]. Their position statement provided a clinically oriented classification system in which disorders of the heart muscle could be categorized based on ventricular morphology and function instead of genetics or etiology. Subsequent updates from both AHA and ESC have followed in recent years, which have aimed to enhance our current and developing understanding of the genetics, physiology, and natural history underlying the disease [1, 5, 10]. In parallel to updates to the definition of DCM, diagnostics of the disease have been transforming. Diagnostic methods for DCM were historically based primarily on clinical characteristics, X-ray, and electrocardiogram (ECG) tools. However, more modern cardiac magnetic resonance (CMR) imaging and cardiac ultrasound devices can provide high-resolution images, thereby enabling the identification of additional characteristics such as myocardial scarring and inflammation. In addition, routine genetic testing is now recommended for familial cases of DCM, which can aid in identifying specific mutations that may contribute to the disease [37]. Improved understanding of its etiology and advancements in diagnostics over the years have helped classify different phenotypes of cardiomyopathies and distinguish DCM from other overlapping etiologies that previously led to misclassification of DCM patients [5, 37, 38]. In light of the adapting landscape of definitions and diagnostics for DCM, it is unsurprising that reported estimates of DCM prevalence and incidence have differed across studies con-

ducted at various times and across regions. Nevertheless, more recent studies using contemporary definitions of DCM should generally be considered more accurate in reflecting the true prevalence and incidence of DCM. Modern definitions typically encompass appropriate diagnostics and the most recent understanding of the etiology and natural history of the disease according to experts [31].

Finally, the reported discrepancies in prevalence and incidence estimates across studies may be attributable to different methodologies, data sources, and population characteristics. Specifically, differences in demographics of the patient population (e.g., age, sex, race/ethnicity) may explain some of the variation in observed figures. Although, within the studies included, important demographic information describing the sample population was often not reported. This omission is significant, as a substantial proportion of DCM cases are linked to genetic factors, and racial differences in genetic predisposition, such as variants in *TTN* and *LMNA* genes, as well as specific polymorphisms more common in individuals of African descent, may influence disease prevalence and severity. Moreover, disparities in healthcare access and socio-environmental exposures likely contribute to higher DCM burden in certain racial and ethnic groups. Difference in study design (e.g., cross-sectional vs. longitudinal) and data sources (e.g., hospital databases vs. population-based registries) further compound the variability in reported estimates.

It is especially important to consider the data source when interpreting estimates reported by different studies; for instance, studies reporting on specific regions or single-center studies may not be generalizable to the general population. Williams et al (1985) [29] conducted a questionnaire survey of family practitioners in two regions of England - East Anglia (Norfolk, Suffolk, and Cambridgeshire) and Essex, Hertfordshire, and Bedfordshire. Authors reported the prevalence of DCM as 8.3 per 100,000 individuals based on a response rate of only 54% and without objective confirmation of findings by echocardiographic or angiographic means. Thus, their findings served as only a preliminary study for assessing the prevalence of DCM in specific regions of the UK. Conversely, Codd et al (1989) [6] conducted a population-based study of Olmsted County, Minnesota, by utilizing a central data bank of Mayo Clinic and the Rochester Epidemiology Project database to report an idiopathic DCM prevalence of 36.5 per 100,000 individuals. This database, although is restricted to a specific county in the US, contains medical records of nearly the entire population of this county, irrespective of demographic or socioeconomic characteristics; only about approximately 5% of the target population do not consent for their records to be used in research [39]. In this way, this database can be considered an exceptionally comprehensive and well-designed data source for various research activities, including estimating prevalence and incidence. Nevertheless, extrapolating this dataset to the entire US population may still be considered biased. Previous research has demonstrated that although the county is typically well-matched to other regions of the US in terms of age and sex, Olmsted County was determined to be less ethnically diverse than the entire US population, wealthier, and more educated. Thus, the study conducted within Olmsted County may not be generalizable to the current US population.

Data on pediatric DCM prevalence were sparse, with only three studies reporting. The large discrepancy between the reported results is mostly due to different diagnostic criteria utilized by the respective studies. The prevalence estimates reported by Ababio et al (2023) [18] may be considered more precise since Codd et al (1989) [6] used dated diagnostic criteria and Angelini et al (2018) [19] was not a population-based study and cases were ascertained based on voluntary participation in the questionnaire followed by ECG and CMR evaluations which were subject to psychological, social, and economic pre-testing biases. As seen in the adult/heterogeneous population, pediatric DCM prevalence estimates also showed variation according to the DCM diagnostic criteria used, as reported by Ababio et al (2023) [18]. The prevalences of children with validated, idiopathic, or total (idiopathic and non-idiopathic) DCM were 2.4, 1.2, and 2.6 cases per 100,000 persons for children aged ≤ 11 years, and estimates were 2.6, 1.3, and 2.8 cases per 100,000 for those aged 12 - 17 years, respectively. Moreover, older children tended to show higher prevalence estimates in comparison with younger ones. This highlights the lack of reliable data on prevalence and incidence of pediatric DCM. More studies should be conducted, especially those stressing upon idiopathic and genetic forms of DCM.

Four studies reported data for incidence of DCM [6, 21, 22, 25] in the adult/heterogeneous general population, ranging from 6 to 7.9 per 100,000 individuals. Coughlin et al (1993) [21] reported incidence rates of DCM based on ICD-9 diagnostic codes. Conversely, Codd et al (1989) [6] and Rakar et al (1997) [25] based their definitions on WHO/ISFC Task Force recommendations, while Herd et al (1991) [22] did not report any specific DCM diagnosis criteria. Incidence rates tended to be higher in studies utilizing non-ICD diagnostic codes. Of all the studies reporting incidence rates, only Codd et al (1989) [6] utilized a population-based database for ascertainment of DCM cases. However, this may not be generalizable to the current US population due to its limited sample size, and changes in disease epidemiology and clinical care since its publication. Rakar et al (1997) [25] utilized a cardiomyopathies registry and post-mortem database of a cardiology department in Trieste University for DCM case ascertainments, and thus cannot be generalized to the overall Italian population. Herd et al (1991) [22] ascertained DCM cases based on a retrospective review of data from hospitalizations, outpatient visits, and autopsy records in a single center. Likewise, their study results cannot be generalized to the overall UK population. Moreover, their study also considered cases of alcoholic cardiomyopathies. Overall, none of these studies presented incidence rates that were truly representative of a national population, as they were all carried out in specific regions within a country. Lastly, only one study reported data for incidence of DCM in the pediatric population (0.6 per 100,000 individuals) [27]. This highlights a knowledge gap in the existing literature in this population.

Strengths and limitations

Our review has several key strengths. Firstly, the review methodology was rigorous, and literature searches were compre-

hensive and thorough, encompassing two major electronic databases and gray literature searches. Our review compiles all published evidence on the topic, providing estimates of prevalence and incidence for DCM in the general population of select pre-specified countries. Further, this study identified various challenges and gaps in the reported studies, including issues related to inconsistent diagnostic criteria utilized to identify DCM patients, technological advancements in techniques over time causing significant variations in estimates, and the evolution of the definition of DCM which may lead to either under- or over-representation of DCM cases.

However, our review is not without limitations, which should be considered when interpreting the findings. First, we restricted inclusion to studies published in English; therefore, non-English but otherwise relevant studies would have been missed. Additionally, a key limitation of our research was that we focused on including data from select countries; namely, the US, France, Germany, Italy, Spain, and the UK. This geographical limitation may reduce generalizability of the results, and the global prevalence and incidence of DCM could not be estimated. Furthermore, as approximately 35% of DCM cases arise due to genetic factors, including data from Middle Eastern, South American, and Asian populations would provide more insight into the epidemiology of DCM and any variations across geographic regions. Further up-to-date research into the prevalence and incidence of DCM outside of our selected countries is warranted to understand the complete prevalence and incidence of the disease. Despite employing a rigorous methodology for the selection of studies from published literature, the possibility of having excluded potentially relevant studies on the topic cannot be completely ruled out due to the highly heterogeneous nature of the disease and overlapping etiologies of DCM with other cardiac-related issues. Furthermore, limitations associated with the studies included in our review should be noted. Inherent limitations are present due to the nature of the study designs and use of retrospective observational data. Also, subclinical DCM cases may not be adequately represented in these study populations which may result in underestimation of DCM prevalence or incidence rates. Another limitation is the possibility that the patients included in these databases may also be more likely to have insurance coverage when compared with the general population. This bias can result in underestimating the prevalence and impact of conditions like DCM in uninsured or underinsured groups, who may have different risk factors and healthcare access. Addressing this bias is crucial for more accurate and inclusive health research. Lastly, some prevalent data originated from small regions or single-center studies, which may not be representative of the general population. However, this was not considered to have strongly affected our results, which rely primarily on figures from large representative studies.

It is acknowledged that the described variations across studies have significant impact on the precise prevalence and incidence estimates of DCM and thus underscore the need for more carefully designed future studies. Moreover, guidelines providing diagnostic criteria can continue to develop to take into consideration evolving diagnostic and genetic markers, new disease presentations and reclassifications which will help improve the accuracy of case estimations. These developments will help tailor specific therapies for patients by imparting better clinical care and help improve prognosis as well.

Conclusions

Our review identified varied prevalence and incidence rates of DCM as reported across different geographic areas, time periods, sexes, races, and definitions of DCM. 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. The lack of reliable data on pediatric DCM and clear reporting of data on idiopathic and genetically associated DCM is an unmet need in this field. Moving forward, to obtain more accurate estimates of DCM prevalence and incidence, future studies should standardize selection criteria according to established definitions of DCM.

Supplementary Material

Suppl 1. Search Strategy for Embase (via OvidSP).

Suppl 2. Search Strategy for MEDLINE® (via OvidSP).

Suppl 3. List of Studies Included in the Review.

Suppl 4. List of Excluded Studies After Full-Text Screening.

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Conflict of Interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Author Contributions

MM: conceptualization, methodology, writing - review and editing. AB, YZ, SM, CB, and AB: methodology, writing - review and editing. KH: data curation, formal analysis, investigation, methodology, project administration, writing - review and editing. RK: data curation, formal analysis, investigation, methodology, writing - original draft, writing - review and ed-

iting. MSF: conceptualization, formal analysis, investigation, methodology, supervision, writing - review and editing. NG: conceptualization, methodology, supervision, writing - review and editing.

Data Availability

The authors declare that data supporting the findings of this study are available within the article and its supplementary files, and further inquiries can be directed to the corresponding author.

Abbreviations

AHA: American Heart Association; CI: confidence interval; CMR: cardiac magnetic resonance; DCM: dilated cardiomyopathy; ICD: International Classification of Diseases; LVEF: left ventricular ejection fraction; PRISMA: Preferred Reporting Items for Systematic Reviews and Meta-Analyses; NR: not reported; UK: United Kingdom; US: United States; WHO/ISFC: World Health Organization/International Society and Federation of Cardiology

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