Epidemiology of Idiopathic Dilated and Hypertrophic Cardiomyopathy

A Population-Based Study in Olmsted County, Minnesota, 1975–1984

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Using the records linkage system of the Mayo Clinic and of the Rochester Epidemiology Project, which accesses diagnostic data on the entire population of Olmsted County, Minnesota, we identified 45 new cases of idiopathic dilated cardiomyopathy (DCM) and 19 new cases of hypertrophic cardiomyopathy (HCM) among county residents for the years 1975–1984. Overall age- and sex-adjusted incidence rates were 6.0/100,000 and 2.5/100,000 person-years, respectively. The incidence of DCM doubled from 3.9/100,000 in the first 5 years to 7.9/100,000 person-years in the last 5 years of study. The corresponding change for HCM was from 1.4 to 3.6/100,000 person-years. Age- and sex-adjusted prevalence rates as of January 1, 1985, for DCM and HCM were 36.5/100,000 and 19.7/100,000 population, respectively. The prevalence of DCM in persons less than 55 years old was 17.9/100,000, over a third of whom were New York Heart Association functional Class III or IV at diagnosis. These estimates may be of value in determining the potential use of health care resources, particularly cardiac transplantation. (Circulation 1989;80:564–572)

Since the term "cardiomyopathy" was coined 30 years ago to describe a group of myocardial diseases of unknown cause,1 investigators have described many of the characteristic clinical features of the idiopathic cardiomyopathies.2 Attempts have also been made to define their epidemiologic features, specifically the frequency of occurrence in the general population.3–10 However, with the exception of a population-based study from Malmö, Sweden,3,4 incidence and prevalence estimates have been based on data from selected population subgroups, such as patients referred to cardiac units of general hospitals or major medical institutions,6,7 cases identified at autopsy,5–7 hospital discharge and mortality statistics,9 cross-sectional survey data,8 or cases identified through voluntary participation in an industrial health screening program.10 Thus, the epidemiology of the cardiomyopathies among unselected patients in the general population is still largely unknown. The purpose of this study was to document the incidence and prevalence of idiopathic dilated (DCM) and hypertrophic (HCM) cardiomyopathy in the population of Olmsted County, Minnesota, during the 10-year period from 1975 to 1984. This appears to be the first study of idiopathic cardiomyopathy in the United States to be based on review of the complete medical records of all patients diagnosed in a delineated community.

Methods

Case Ascertainment

Population-based epidemiologic research is possible in Olmsted County, Minnesota, because medical care is practically self-contained within the community and is delivered by a handful of health care providers. Most care is provided by the Mayo Clinic, which has maintained a unified medical record system with its two large affiliated hospitals during the past 80 years. This dossier-type record contains both inpatient and outpatient data and is easily retrievable for review.11 The diagnoses and surgical procedures entered into these records are indexed. The index includes the diagnoses made for outpatient office or clinic consultations, emergency

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room visits, nursing home care, hospital admissions, autopsy examinations, and death certification. The medical records of the other providers in the area who have served the local population are also indexed and can also be retrieved. These include the Olmsted Medical Group and Olmsted Community Hospital, the Rochester State Hospital, and the University of Minnesota and Veterans Administration Hospitals in Minneapolis. Data from several small hospitals in surrounding counties, from a few family practitioners in Rochester, and from local nursing homes are also coded into the central data bank at the Mayo Clinic. Thus, the details of the medical care provided to residents of the entire community are available for study. This ensures nearly complete case ascertainment for all major illnesses diagnosed in Olmsted County residents. The potential value of this data system (the Rochester Epidemiology Project) for population-based studies has been described previously.11,12

Using this unique data base, we identified all Olmsted County residents in whom cardiomyopathy was diagnosed during the 10-year period from 1975 to 1984. To ensure complete case ascertainment, we retrieved and reviewed the records of all patients assigned to the following rubrics of the hospital adaptation of the International Classification of Diseases, Eighth Revision (H-ICDA): Code 425, which specifically pertains to the cardiomyopathies; Code 427, which relates to heart failure; and Code 429, which denotes a category of “ill-defined heart disease,” including unspecified cardiomegaly. In addition, records in the Department of Pathology were checked for cases identified at autopsy, and a computerized diagnostic index maintained in the echocardiographic laboratory was reviewed for cases not listed elsewhere. From a total of 3,250 potential cases that were screened, we identified 69 individuals who fulfilled predetermined diagnostic and residency criteria for idiopathic cardiomyopathy.

Diagnostic Criteria
Diagnostic categories were as outlined by the World Health Organization/International Society and Federation of Cardiology Task Force on Cardiomyopathies.13

Dilated cardiomyopathy. DCM involves dilatation of the left, right, or both ventricles, with impaired systolic function; congestive heart failure is a frequent but not invariable finding.13 The diagnosis of DCM is essentially one of exclusion2 and is based on symptoms, physical signs, electrocardiographic, and chest roentgenographic findings of ventricular dysfunction, supported by echocardiographic or angiographic confirmation of global ventricular dilatation. In this study, DCM was deemed present in 46 patients. In four individuals, the diagnosis was confirmed only at autopsy, whereas the other 42 had the diagnosis confirmed by echocardiography, angiography, or both (Figure 1). Of those who had echocardiography (n=41), 10 were diagnosed before the availability of two-dimensional scanning, although eight subsequently had two-dimensional scans. All 42 patients fulfilled the following criteria: left ventricular end-diastolic (LVEDV) and end-systolic (LVESV) volumes two standard deviations or more greater than volumes established for normal subjects (LVEDV≤110 cc/m²; LVESV≥44 cc/m²) and left ventricular ejection fraction (LVEF) two standard deviations or more below that in normal subjects (LVEF≤51%).14

In screening the clinical records for potential DCM cases, those in whom ventricular dilatation or dysfunction was most likely due to significant coronary artery disease were excluded. Members of the final DCM cohort who had a history of angina or had electrocardiographic findings of ischemia (n=14) were shown to have normal coronary arteries or minimal atherosclerosis by coronary angiography (n=12), treadmill exercise testing (n=1), or autopsy (n=1).

There were 15 individuals (33%) from whom cardiac tissue was available for pathologic examination (nine patients had endomyocardial biopsies, seven had autopsies, one had both examinations). Because there are no specific morphologic features that characterize DCM, these specimens were examined primarily for the exclusion of other conditions. All showed interstitial or perivascular fibrosis (n=11), myohypertrophy (n=10), or both in the absence of stainable iron, amyloid deposition, or lymphocytic infiltration.
Hypertrophic cardiomyopathy. HCM is characterized by disproportionate hypertrophy of the left ventricle, and occasionally also of the right ventricle, which typically involves the septum more than the free wall but sometimes is concentric. This occurs in the absence of a recognizable stimulus to hypertrophy. In this study, HCM was diagnosed during life in 18 persons; another three cases were first diagnosed at autopsy (Figure 1). The diagnosis of HCM is based on specific echocardiographic criteria. Initial echocardiographic evaluation was by M-mode alone in seven patients. The findings were subsequently confirmed in five of these by two-dimensional echocardiography, in one by contrast ventriculography, and in one at autopsy. All patients had a thickened interventricular septum (median, 17 mm; range, 13–23 mm). Sixteen patients had an increased septal to posterior wall thickness ratio (≥1.5); in the remaining two individuals diagnosed during life, this ratio was 1.4. Systolic anterior motion of the mitral valve was present in 11 patients, and midsystolic closure of the aortic valve was present in three.

Specifically excluded from this cohort were patients with left ventricular hypertrophy secondary to known causes, such as systemic hypertension or aortic stenosis. As with DCM, there are no pathognomonic morphologic features of HCM. Pathologic examination (four autopsy specimens) revealed nonspecific interstitial fibrosis and myohypertrophy in all patients, without stainable iron, amyloid deposits, or lymphocytic infiltration.

Restrictive cardiomyopathy. RCM is the end result of endomyocardial scarring that may affect one or both ventricles, restricting ventricular filling. The diagnosis of RCM, based on characteristic hemodynamic features at cardiac catheterization, was established in one individual in our study.

Unclassified cardiomyopathy. A few cases of idiopathic cardiomyopathy do not fit readily into any group defined above. These have been termed “unclassified” cardiomyopathies, and best describe one patient in our cohort. A 6-year-old girl, who died suddenly and unexpectedly, was found at autopsy to have mild concentric left ventricular hypertrophy with lymphocytic infiltration of the myocardium; she did not fulfill diagnostic criteria for myocarditis.

Patients with specific heart muscle diseases caused by known infective agents, metabolic excesses or deficiencies, sensitivity or toxic reactions, or specific heart muscle diseases associated with systemic diseases, storage diseases, or hereditary or familial disorders were not included in the study. However, family members were not contacted or investigated to detect cardiac disease. Two patients with HCM and three with DCM had a documented family history possibly compatible with cardiomyopathy, but such history is likely to be incomplete.

Residency Criteria

In addition to fulfilling diagnostic criteria, patients for the incidence study must have established residence in Olmsted County for at least 1 year before diagnosis, thus excluding those who might have moved into the area for diagnosis or treatment of their cardiomyopathy. Patients for the prevalence study were required to be residing in the county on January 1, 1985, and to have had established residence in Olmsted County for at least 1 year before that date.

Data Analysis

The entire population of Olmsted County was considered to be at risk of developing idiopathic cardiomyopathy. Thus, incidence rates were calculated with the observed number of cases divided by the age- and sex-specific person-years of observation estimated from decennial census data for Olmsted County. Rates were also calculated separately for the city of Rochester (urban) and the balance of Olmsted County (rural). Rates were directly age- and sex-adjusted to the population structure of US whites in 1980; for comparisons between sexes, rates were age-adjusted to the same standard population. The denominator for the calculation of prevalence rates was the estimated county population on January 1, 1985. Ninety-five percent confidence intervals (95% CI) were estimated from the cumulative Poisson distribution. Statistical significance indicates p less than 0.05 unless otherwise noted.

Results

During the 10-year study period, 69 Olmsted County, Minnesota, residents were diagnosed as having idiopathic cardiomyopathy. All of these individuals were white, reflecting the racial composition of the community. Of the total, 46 (67%) had dilated cardiomyopathy, and 21 (30%) had hypertrophic cardiomyopathy. One 67-year-old woman had typical hemodynamic features of restrictive cardiomyopathy, and the remaining patient's cardiomyopathy could not be classified. Sixty-six of these patients represented new diagnoses of cardiomyopathy, whereas the remaining three individuals moved to Olmsted County after the diagnosis was established and were only counted among the prevalence cases. Altogether, there were 45 patients with idiopathic cardiomyopathy residing in the community on the prevalence day of January 1, 1985.

Based on 45 new cases of DCM and 19 new cases of HCM, the age- and sex-adjusted incidence rates were 6.0/100,000 person-years (95% CI, 4.2–7.7) and 2.5/100,000 person-years (95% CI, 1.4–3.7), respectively. With a single case, the crude incidence of restrictive cardiomyopathy was estimated at 0.1/100,000 person-years. The overall age- and sex-adjusted incidence of all cardiomyopathies combined was 8.7/100,000 person-years (95% CI, 6.6–10.9) with the 66 new cases observed. There was a marked preponderance of men among the DCM and
HCM incident cases: 33 men vs. 12 women with DCM and 11 men vs. eight women with HCM. The male:female ratio of age-adjusted incidence rates was 3.4 and 1.7, respectively (Table 1). Table 1 also shows the age-specific incidence rates for each sex. With the exception of HCM in elderly women, incidence rates for DCM and HCM were greater for men than for women in all age groups. Rates generally increased with advancing age, although the small number of cases in certain age groups makes the pattern unstable. Median age at diagnosis of DCM was 54 years (range, 24–88 years), whereas that for HCM was 59 years (range, 16–94 years).

The manner in which these incident cases presented is given in Table 2. Forty-one (91%) of the 45 DCM cases were diagnosed during life. Of these 41 cases, 36 (88%) were symptomatic before diagnosis, most commonly with dyspnea that occurred in 33 patients by New York Heart Association (NYHA) guidelines. In 27 of these 33 patients, the dyspnea was severe (NYHA functional Class III or IV) and was accompanied by signs of heart failure. Nine patients had prior anginal symptoms, all of whom were shown to have insignificant or no coronary artery disease. Five persons had experienced a syncopal episode (defined as sudden loss of consciousness) before diagnosis. Four of the 45 DCM cases (9%) were first correctly diagnosed at autopsy. All four had previous symptoms: three with dyspnea, 2 with heart failure, and one with recurrent preexcitation syndrome.

Sixteen of the 19 HCM incident cases (84%) were diagnosed during life, and 10 of these 16 (62%) were symptomatic before diagnosis. Angina pectoris, occurring in eight of the 10, was the most common symptom, with dyspnea (predominantly NYHA Class I or II), heart failure, syncope, presyncope, and paroxysmal supraventricular tachycardia occurring less commonly (Table 2). Three of the 19 patients with HCM were first correctly diagnosed at autopsy. Two of these were young men (aged 28 and 34 years) who died suddenly and unexpectedly. One was an elderly woman known to have heart disease.

There were five persons with DCM and six with HCM in whom the diagnosis was made in the course of preemployment or other general medical examination or during inpatient work-up for an unrelated condition. None of these had a history of cardiac symptoms. Abnormalities that focused attention on the cardiovascular system included a cardiac murmur in six patients, an increased cardiothoracic ratio on chest roentgenogram in two and electrocardiographic abnormalities in three patients. Resultant echocardiographic studies confirmed a diagnosis of DCM or HCM. Nine of these 11 asymptomatic individuals came to attention in the last 5 years of the study, 1980–1984, compared with only two in the first 5 years. This may have influenced the incidence rate, which rose significantly between 1975 and 1979 and 1980 and 1984 (Table 3). The overall age- and sex-adjusted incidence of DCM increased from 3.9/100,000 person-years in the first 5-year period to 7.9/100,000 person-years in the second, whereas the corresponding change for HCM was from 1.4 to 3.6/100,000 person-years. Given the small numbers, incidence rates in the urban and rural areas of Olmsted County cannot be rigorously compared. The rates for 1980–1984 appear quite similar in the two areas, however, which raises the possibility that the somewhat lower rates for 1975–1979 may be due to underascertainment among rural patients (Table 3).

The age- and sex-adjusted prevalence of the idiopathic cardiomyopathies (all types) on January 1, 1985, was 57.8/100,000 population (95% CI, 40.7–
74.9. The prevalence of DCM, 36.5/100,000 (95% CI, 23.1–50.0), was almost twice that of HCM, 19.7/100,000 (95% CI, 9.6–29.8). This mainly reflects the underlying difference in incidence of these two conditions. Prevalence rates were greater for men, with male:female ratios of age-adjusted rates of 3.0:1 for DCM, 2.0:1 for HCM, and 2.4:1 overall. Because of the small number of cases in most age groups, no clear pattern by age was discernible, although there was a suggestion of higher rates among older individuals (Table 4). Crude prevalence rates in those less than 55 years of age were 17.9/100,000 for DCM, 5.1/100,000 for HCM, and 23.0/100,000 for all idiopathic cardiomyopathies together. For patients with DCM in the prevalence study, the median age on January 1, 1985, was 61 years (range, 32–81 years); median LVEF at diagnosis was 26% (range, 12–40%).

**Discussion**

Previous attempts to describe the epidemiologic characteristics of the idiopathic cardiomyopathies

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**Table 2.** Clinical Spectrum at Presentation of Idiopathic Dilated Cardiomyopathy and Hypertrophic Cardiomyopathy Among Olmsted County, Minnesota, Residents, 1975–1984

<table>
<thead>
<tr>
<th>Diagnosis before death</th>
<th>Idiopathic dilated cardiomyopathy</th>
<th>Hypertrophic cardiomyopathy</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>( n )</td>
<td>%*</td>
</tr>
<tr>
<td>Symptomatic</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dyspnea (NYHA)§</td>
<td></td>
<td></td>
</tr>
<tr>
<td>I/II</td>
<td>41</td>
<td>91</td>
</tr>
<tr>
<td>III/IV</td>
<td>36</td>
<td>88</td>
</tr>
<tr>
<td>Clinical heart failure</td>
<td>27</td>
<td>75</td>
</tr>
<tr>
<td>Angina pectoris</td>
<td>9</td>
<td>25</td>
</tr>
<tr>
<td>Syncope</td>
<td>5</td>
<td>14</td>
</tr>
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<td>Near syncope</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Paroxysmal supraventricular tachycardia</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Asymptomatic</td>
<td>5</td>
<td>12</td>
</tr>
<tr>
<td>First diagnosis at autopsy</td>
<td>4</td>
<td>9</td>
</tr>
<tr>
<td>Symptomatic</td>
<td>4</td>
<td>100</td>
</tr>
<tr>
<td>Dyspnea (NYHA)§</td>
<td></td>
<td></td>
</tr>
<tr>
<td>I/II</td>
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<td>25</td>
</tr>
<tr>
<td>III/IV</td>
<td>2</td>
<td>50</td>
</tr>
<tr>
<td>Clinical heart failure</td>
<td>2</td>
<td>50</td>
</tr>
<tr>
<td>Paroxysmal supraventricular tachycardia</td>
<td>1</td>
<td>25</td>
</tr>
<tr>
<td>Asymptomatic</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>45</td>
<td>100</td>
</tr>
</tbody>
</table>

NYHA, New York Heart Association functional classification.
*Percentage by diagnosis during life or at autopsy.
†Percentage of symptomatic versus asymptomatic within each diagnosis category.
§Percentage with each symptom among those who were symptomatic. Percentages may total more than 100% because individuals may have more than one symptom.
§New York Heart Association (NYHA) functional classification for severity of dyspnea.23

**Table 3.** Secular Trends in Incidence by Type of Cardiomyopathy and Area of Residence Among Olmsted County, Minnesota, Residents 1975–1984

<table>
<thead>
<tr>
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</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>( n )</td>
<td>Rate*</td>
<td>( n )</td>
<td>Rate*</td>
<td>( n )</td>
<td>Rate*</td>
</tr>
<tr>
<td>Dilated idiopathic cardiomyopathy</td>
<td></td>
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</tr>
<tr>
<td>Urban</td>
<td>11</td>
<td>5.0</td>
<td>20</td>
<td>8.2</td>
<td>31</td>
<td>6.6</td>
</tr>
<tr>
<td>Rural</td>
<td>3</td>
<td>1.9</td>
<td>11</td>
<td>7.1</td>
<td>14</td>
<td>4.6</td>
</tr>
<tr>
<td>Total</td>
<td>14</td>
<td>3.9</td>
<td>31</td>
<td>7.9</td>
<td>45</td>
<td>6.0</td>
</tr>
<tr>
<td>Hypertrophic cardiomyopathy</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Urban</td>
<td>4</td>
<td>1.7</td>
<td>8</td>
<td>3.0</td>
<td>12</td>
<td>2.4</td>
</tr>
<tr>
<td>Rural</td>
<td>1</td>
<td>0.7</td>
<td>6</td>
<td>3.9</td>
<td>7</td>
<td>2.3</td>
</tr>
<tr>
<td>Total</td>
<td>5</td>
<td>1.4</td>
<td>14</td>
<td>3.6</td>
<td>19</td>
<td>2.5</td>
</tr>
</tbody>
</table>

*Incidence per 100,000 person-years, directly age- and sex-adjusted to the US white population in 1980.
have been limited by a number of factors. Patients with cardiomyopathies referred to tertiary care centers are unlikely to reflect the spectrum of disease as it occurs in the general population; studies based on autopsy series are limited by the selection of deaths that come to autopsy; and hospital discharge statistics are, by definition, based on inpatient cases of disease only. The strength of the present study lies in the fact that ascertainment of diagnosed cases of idiopathic cardiomyopathy is almost certainly complete for the delineated population of Olmsted County during the 10-year period of study. This is assured by 1) comprehensive and high-quality medical care provided to the local population by the Mayo Clinic and the other health care providers in the area; 2) the high use of medical care by the local community—it is estimated that in any 3-year period, about 95% of the population of Olmsted County is seen for routine examination or for major or minor illness at Mayo Clinic or its affiliates (L.J. Melton, unpublished data); 3) the unique nature of the centralized diagnostic index at the Mayo Clinic (the Rochester Epidemiology Project), which includes inpatient, outpatient, and autopsy diagnoses from almost all providers of care to the local population; 4) the extensive screening procedure used in this study that covers several diagnostic rubrics to which cardiomyopathy could be coded; and 5) the availability for review of the original and complete inpatient and outpatient clinical records on every case. Undiagnosed cases would, of course, have been missed.

Despite these advantages, both this study and previous work in this area are limited by the lack of specific diagnostic criteria. Since the report of the World Health Organization/International Society and Federation of Cardiology Task Force, the term "cardiomyopathy" has been reserved for heart muscle disorders of unknown cause, which may be classified as dilated, hypertrophic, or restrictive, according to their anatomic and functional features. Heart muscle disease of known cause; that is, that due to systemic or pulmonary hypertension, coronary artery disease, valvular or congenital heart disease, infective or metabolic causes, and those associated with systemic disorders are specifically excluded. Thus, the diagnosis of idiopathic cardiomyopathy is primarily one of exclusion supported by a combination of clinical features, many of which are nonspecific. In the absence of a specific clinical finding or diagnostic test, there are, inevitably, problems in diagnosis and classification.

A particular question arising in relation to DCM concerns the possible coexistence of coronary artery disease. Ideally, all patients with DCM should be shown to be free of coronary artery disease. In practice, however, coronary arteriography is not routinely performed in all patients with congestive heart failure. Because retrospective studies cannot control the conditions under which patients are recruited or investigated, it follows that most epidemiologic studies of DCM have had to compromise on the completeness of data on coronary artery disease in their subjects. In the Icelandic study, three fourths of the patients were shown to have normal coronary arteries; the proportion of the Malmö series that underwent coronary arteriography is not specified, and there are no details of coronary imaging in the United Kingdom questionnaire study. A previous retrospective study based on the entire Mayo Clinic practice, including referral patients, found angiographic evidence of normal coronary arteries for 58% of DCM patients. In this population-based study, 47% of patients were shown

### TABLE 4. Prevalence of Idiopathic Dilated Cardiomyopathy, Hypertrophic Cardiomyopathy, and All Idiopathic Cardiomyopathies Among Olmsted County, Minnesota, Residents on January 1, 1985

<table>
<thead>
<tr>
<th>Age (yr)</th>
<th>Idiopathic dilated cardiomyopathy</th>
<th>Hypertrophic cardiomyopathy</th>
<th>All cardiomyopathies*</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Male n Rate†</td>
<td>Female n Rate†</td>
<td>Male n Rate†</td>
</tr>
<tr>
<td>&lt;15</td>
<td>0 0.0 0.0</td>
<td>0 0.0 0.0</td>
<td>0 0.0 0.0</td>
</tr>
<tr>
<td>15–24</td>
<td>0 0.0 0.0</td>
<td>0 0.0 0.0</td>
<td>2 25.9 0.0</td>
</tr>
<tr>
<td>25–34</td>
<td>3 36.0 10.2</td>
<td>0 0.0 0.0</td>
<td>3 36.0 20.4</td>
</tr>
<tr>
<td>35–44</td>
<td>0 0.0 0.0</td>
<td>0 0.0 0.0</td>
<td>0 0.0 0.0</td>
</tr>
<tr>
<td>45–54</td>
<td>9 198.3 22.2</td>
<td>1 22.0 0.0</td>
<td>10 220.3 22.2</td>
</tr>
<tr>
<td>55–64</td>
<td>3 89.1 109.2</td>
<td>5 148.5 0.0</td>
<td>8 237.6 109.2</td>
</tr>
<tr>
<td>65–74</td>
<td>3 148.5 35.9</td>
<td>1 49.5 107.7</td>
<td>4 198.0 179.5</td>
</tr>
<tr>
<td>≥75</td>
<td>3 237.5 35.0</td>
<td>0 0.0 70.1</td>
<td>3 237.5 105.1</td>
</tr>
<tr>
<td>Total</td>
<td>21 47.0 16.2</td>
<td>9 20.1 12.1</td>
<td>30 67.1 15.0</td>
</tr>
<tr>
<td>Adjusted‡</td>
<td>58.0 19.4</td>
<td>25.8 13.0</td>
<td>83.8 35.1</td>
</tr>
<tr>
<td>95% confidence interval§</td>
<td>32.4–83.5</td>
<td>5.7–33.2</td>
<td>8.8–42.8</td>
</tr>
</tbody>
</table>

*Includes one patient with restrictive cardiomyopathy.
†Prevalence per 100,000 population.
‡Prevalence per 100,000 population, directly age-adjusted to the U.S. white population in 1980.
§95% confidence interval around adjusted rate.
to have normal coronary arteries or minimal atherosclerosis. However, these did include all patients who complained of angina or had electrocardiographic findings of ischemia. Moreover, the median duration of documentation in Mayo records before diagnosis was 23 years in this study, indicating that most patients were under continuous long-term medical surveillance before the diagnosis of cardiomyopathy.

A second unresolved issue in DCM is the etiologic contribution of ethanol. Several investigators have attempted to define the nature of the association between "excessive" alcohol consumption and myocardial disease, but there is still uncertainty about the degree and duration of alcohol abuse required to effect myocardial damage. There is the possibility that persons with ventricular dilatation and impaired systolic function are questioned more intensively about their drinking habits than persons with other conditions, and there is the recognized difficulty of obtaining an accurate account of alcohol intake from habitual drinkers. In this study, it would appear from entries in the medical records that 12 of the 46 DCM patients had a significant history of alcohol consumption. To have excluded these from the cohort would likely have resulted in underestimation of the incidence and prevalence of DCM. It is not clear whether or not persons with a history of alcohol abuse were excluded from other epidemiologic studies of DCM.

Other estimates of the incidence and prevalence of idiopathic dilated or hypertrophic cardiomyopathy are summarized in Table 5. These estimates vary considerably, reflecting variation in case ascertainment and diagnostic criteria, although the possibility of some geographic variation cannot be ruled out. In Malmö, Sweden, 550 individuals with myocardial disease of diverse causes were identified from 1970 to 1979. Extensive clinical investigation established the diagnosis of DCM "firmly" in 89 patients, giving a crude annual incidence of 3.9/100,000.

Another 43 persons were classified as "probably" cases, increasing the crude rate to 5.3/100,000/year, but specific criteria by which these categories were defined are not provided. The comparable group in this study (i.e., those with DCM diagnosed during life) totals 41 cases and yields a crude rate of 4.5/100,000 person-years. Although these rates appear quite similar, the Olmsted population is younger on average than that of Malmö. The Olmsted County age- and sex-specific rates applied to the Malmö population yield an adjusted incidence rate of 7/100,000 person-years, which can be directly compared with the Malmö rate of 5.3/100,000.

Both studies recognized additional patients in whom DCM was diagnosed at autopsy. These made up 30% of the Malmö series and 9% of the Olmsted County cohort series. This discrepancy is due to the higher proportion of elderly in the Malmö population and, more importantly, to quite different autopsy rates (90% among Malmö residents of all ages vs. about 40% of all Olmsted County deaths and 30% among older residents). Had all Olmsted County patients undergone autopsy, we could conceivably have found up to eight additional cases of DCM (and six more cases of HCM). Nonetheless, after the differences in age and sex distribution are accounted for by adjusting the Olmsted County rate to the Malmö population, the overall incidence rates inclusive of autopsy cases are 7.8 and 7.9/
100,000/year, respectively, in Olmsted County and Malmö. Of interest, the population of Olmsted County is ethnically quite similar to that of Malmö, with a high proportion of residents having Northern European ancestry.

A recent report from Western Denmark noted a crude annual incidence rate of DCM of 0.7/100,000/year. That study, conducted retrospectively during a 2-year period (1980–1981), used clinical notes from three cardiology referral centers and autopsy reports from 14 pathology departments. The low rate almost certainly reflects incomplete case ascertainment for the population under study.

To our knowledge, only one previous attempt has been made to define the prevalence of DCM. This was a questionnaire survey of family practitioners in two regions of England. The resulting prevalence rate of 8.3/100,000 was based on a response rate of only 54% and without objective confirmation of findings by echocardiographic or angiographic means. The overall age- and sex-adjusted prevalence of DCM in our study (36.5/100,000) is over four times that figure and seems likely to be a more accurate representation of the true prevalence.

The crude annual incidence of HCM in Western Denmark has been estimated at 0.4/100,000. This is in sharp contrast to the age- and sex-adjusted incidence from this study of 2.5/100,000 person-years. Even the latter figure may be an underestimate, however, because one recent report drew attention to the coexistence of HCM and hypertension, especially in the elderly, and patients with significant hypertension were excluded from this cohort. A study in Birmingham, England, identified 39 patients with HCM who presented to a district general hospital over a 6-year period. However, it is not possible to construct incidence rates from that study because of incomplete denominator data.

The reported prevalence of HCM also shows considerable variation between studies. Based on 11 cases identified at autopsy during a 12-year period, Bjarnason et al estimated that HCM can be identified in 0.17% of all decedents in Iceland. This was extrapolated to provide a population prevalence for HCM of 33/100,000. Two studies of HCM that used echocardiography as a screening tool suggested prevalence rates of 830/100,000 and 170/100,000, respectively. Although there may be geographic differences in rates of HCM, the most likely reason for such a discrepancy is use of different echocardiographic criteria for HCM. The study based on the Framingham cohort involved echocardiographic markers of HCM, rather than HCM per se. The Japanese study was confined to adult male participants in an industrial heart screening program. Neither of these estimates is comparable to our prevalence rate of 19.7/100,000, based predominantly on symptomatic patients coming to medical attention.

Presentation of a single incidence rate for DCM or HCM for the 10 years covered by this study may be misleading because the incidence of both conditions appears to be rising. Although this may reflect a true increase in incidence, it is more likely due to better diagnosis of the condition because of a higher index of suspicion among clinicians and increased use of echocardiography. For example, a recent study of echocardiography in the Olmsted County population revealed that use of the procedure had increased from 552/100,000 population in 1979 to 1,435/100,000 population in 1985. Nonetheless, the incidence rates presented here may still be underestimates of the true rates, even for the later time period. With more thorough and definitive evaluation of cardiac symptoms and early detection of mild and asymptomatic cases, we may expect further increases in the reported incidence and prevalence of DCM and HCM. Currently, the only way to determine these rates more accurately would be to devise an ongoing screening program in the general population, which is a costly and time-consuming venture given the relative infrequency of the condition.

The impact of the idiopathic cardiomyopathies in terms of morbidity, years of productive life lost, and health care costs is largely unknown. A recent study based on hospital discharge statistics estimated that in the United States there are about 126,000 hospital discharges annually (55/100,000 population/year) that bear a diagnosis of cardiomyopathy. In 46,000 of these (20/100,000/year), cardiomyopathy is the principal diagnosis. The latter group alone accounts for 410,000 hospital-stay days. Interpretation of these data is difficult, however, because hospital statistics count discharges rather than individuals, thus taking no account of readmissions. Moreover, the diagnostic rubric used in most studies, ICD-425, includes specific as well as idiopathic forms of cardiomyopathy, and the diagnoses coded in hospital discharge data are not confirmed. This could represent a major source of error in a study of idiopathic cardiomyopathy. In the present study, for example, there were 517 individuals with diagnoses coded to ICD-425. A detailed review revealed that only 67 of the 517 (13%) could be considered bona fide cases of idiopathic cardiomyopathy. In addition, hospitalized patients represent only a fraction of all patients with cardiomyopathy. In the Olmsted County study, most patients were diagnosed and observed exclusively on an outpatient basis.

Projection of age- and sex-specific incidence and prevalence rates from the present study to the entire estimated population of the United States in 1989 suggests that there may be almost 140,000 patients with idiopathic cardiomyopathy living in the United States, with approximately 20,000 new patients coming to attention annually. These extrapolations do not take into account any regional variation in occurrence, nor do they allow for variation by race because 99% of the Olmsted County population is white. In light of recent data
suggesting that black race is an independent risk factor for DCM, our data may best be regarded as conservative estimates. There are, however, no epidemiologic studies based on multiracial urban populations with which to compare our results.

Treatment of DCM is aimed primarily at alleviating symptoms of congestive heart failure and preventing sudden death. Although many patients benefit from it, drug therapy does not treat the underlying cause of the cardiac dysfunction nor necessarily arrest progression of the disease. Cardiac transplantation may offer the only hope of improved quality of life and increased survival in patients with end-stage heart muscle disease due to DCM. In 1985, 57% of transplant patients had DCM as the underlying pathologic process. In this study, the prevalence of DCM in persons less than 55 years, who were NYHA functional Class III or IV at diagnosis, was 7.7/100,000 population. This estimate may be of value in assessing the potential contribution of cardiac transplantation to the management of DCM.

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References


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