Prevalence of overt dilated cardiomyopathy in two regions of England

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SUMMARY The prevalence of dilated cardiomyopathy was assessed in two regions in England from 420 replies to 771 questionnaires sent to general practitioners. Overall point prevalence was 8.317 per 100 000 population in areas covering a total sample of 913 836 inhabitants. There was a statistically significant regional variation between East Anglia (Norfolk, Suffolk, and Cambridgeshire) and Essex, Hertfordshire, and Bedfordshire.

Though the natural history of dilated cardiomyopathy remains to be defined, this study has provided an assessment of point prevalence of overt cases of this condition.

Dilated cardiomyopathy belongs to the group of diseases defined as heart muscle diseases of unknown cause.¹ Studies of dilated cardiomyopathy have been beset by many problems in the past including the large number of synonyms under which this condition has masqueraded.² Clinical recognition and definition of the natural history are still not fully established.^{3,4} The reason for this is that myocardial hypertrophy and dilatation may be symptomless, and even when symptoms become manifest other diagnoses, which include that of ischaemic heart disease, may be ascribed to patients with this condition unless invasive and non-invasive investigations are undertaken.

Examination of endomyocardial biopsy tissue obtained by biopsies^{5,6} has contributed greatly to defining an infectious-immune mechanism for some patients with dilated cardiomyopathy⁷ so that a better understanding of the natural history is slowly emerging.

With the exception of two studies from the same centre in Sweden^{8,9} the prevalence of dilated cardiomyopathy is unknown. Numerous reports of often large numbers of cases from various centres have been published over the years,^{10,11} but these often act as reference centres, precluding therefore an assessment of prevalence. The purpose of this inquiry, which has hitherto not been undertaken in this country, was to establish an estimate of the prevalence of this condi-

tion. Furthermore, it was considered relevant to undertake this inquiry at this stage in view of the increasing number of patients with dilated cardiomyopathy being considered for transplantation.

Patients and methods

In the winter months of 1983-84 questionnaires were sent to general practitioners who were fellows, members, or associates of the Royal College of General Practitioners and resident in Norfolk, Suffolk, Cambridgeshire, Essex, Bedfordshire, and Hertfordshire. The questionnaire explained the need for information on the prevalence of dilated cardiomyopathy and Table 1 gives an example of the questions asked. An additional letter was sent stressing that only patients with dilated (congestive) cardiomyopathy according to the definition that excludes those patients who suffered from heart failure due to alcohol abuse or occurring in pregnancy or the puerperium or who had other known factors that could account for the symptoms were included. The total populations on the doctors' lists could be provided easily as they are regularly calculated by the local Family Practitioners Committee of the National Health Service. Furthermore we permitted respondents to reply on behalf of the group practice since in this way the scope of the inquiry could be considerably widened. In a few instances where the response was equivocal about where the diagnosis was made, a further letter was sent stressing the importance of the definitive diagnosis after reliable investigations. Only on receipt of a satisfactory reply were these cases included in the study.

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Accepted for publication 22 March 1985

Table 1 *Questionnaire sent to general practitioners*

How many patients do you have with dilated (congestive) cardiomyopathy on the date of receipt of this enquiry?		Please answer below
On my own list		
My list size is approximately		
On the practice list		
Practice list size		
How confident are you that a patient is not unknown to you?		
On your list		Fairly confident/Not confident
On practice list		Fairly confident/Not confident (please delete as applicable)
In <i>either case</i> do still return the form.		
If possible, where was the patient diagnosed?		
Approximately when?		

STATISTICAL ANALYSIS

The statistical test applied was the χ^2 test of independence (prevalence against area) significant at the 0.995 level.

Of the 771 questionnaires sent, 420 replies were received, which was considered acceptable. Some practitioners had retired or were engaged in other areas of primary care and did not possess a personal list of patients.

Results

Table 2 summarises the results.

Discussion

This study has provided an insight into the prevalence of symptomatic patients with this disease. The World Health Organisation (WHO)/International Society and Federation of Cardiology (ISFC) Task Force recommendations on definition and classification based on the original concepts of Goodwin and his colleagues were strictly followed. As has already been stated, cardiomyopathies are defined as "heart muscle diseases of unknown cause" and classified into dilated (congestive), hypertrophic, and restrictive forms.¹ To minimise confusion the term "specific heart muscle disease" was introduced and included all those cases with known secondary involvement of the heart due to metabolic diseases, including those due to deficiency, general systemic diseases, hereditary familial neuromuscular diseases, and sensitivity and toxic reactions, among which alcohol was listed. Heart fail-

ure in peripartur heart disease was excluded according to the definition.

Since the natural history of dilated cardiomyopathy has not yet been fully established,⁴ and the fact that symptoms may not be manifest despite severely depressed left ventricular function, the cases included in this study represent only the tip of the iceberg of the true incidence of dilated cardiomyopathy.¹² Studies of patients with cardiomegaly or left bundle branch block in asymptomatic individuals undergoing medical check ups were also considered unhelpful for assessing the incidence of this condition. There is no evidence that asymptomatic subjects with a fascicular block develop dilated cardiomyopathy in later life. When cardiomegaly is evident on chest radiographs the condition is usually well advanced. Alternatively, despite evidence of left ventricular dilatation or malfunction by other investigations, cardiomegaly may not be evident on chest radiographs.¹² When these various factors are considered together with the fact that patients may be asymptomatic despite depressed left ventricular function being accidentally discovered it is evident that the true prevalence of dilated cardiomyopathy may be far greater. Our results therefore represent overt disease in patients who, as a result of their symptoms, presented to their doctor.

The only other study attempting to give an assessment of the incidence of dilated cardiomyopathy was undertaken by Torp in Malmö, Sweden, who during a period of eight years reported an incidence of 3/100 000 per year of the hospital population for a city of 250 000 inhabitants. If postmortem studies were included during the same period, however, the

Table 2 *Details of results from 420 replies to 771 questionnaires sent to general practitioners*

	No of cases of dilated cardiomyopathy	Patient population	Point prevalence (per 100 000)*
East Anglia (Norfolk, Suffolk, Cambridgeshire)	52	384 220	13.534
Essex, Hertfordshire, and Bedfordshire	24	529 616	4.532
Total	76	913 836	8.317

* χ^2 test of independence (prevalence against area): $p < 0.05$.

incidence rose to 5/100 000 patients per year.⁸ In a subsequent extended study an incidence of 7.5 cases was found with an estimate that 10/100 000 patients might be affected.⁹

Our overall figures of 8.317 cases per 100 000 population is in keeping with these results. Existing evidence points to a poor overall prognosis in symptomatic patients,¹⁰ so that prevalence figures would not be many times greater than those for incidence. It must also be remembered that Torp's study represented a sustained effort in one centre to remain vigilant for this particular diagnosis.

The variation in the prevalence between East Anglia (Norfolk, Suffolk, and Cambridgeshire) and Essex, Hertfordshire, and Bedfordshire is statistically significant ($p < 0.05$) and represents the first indication of an uneven geographical distribution of cases.

The reason for limiting the mailing of questionnaires to fellows, members, or associates of the Royal College of General Practitioners was the belief that these practitioners would co-operate in this study and would be more likely to be in possession of accurate medical records. In addition, a disease-index register was also more likely to have been maintained.

General practitioners are in the favourable position in that after initial appraisal and subsequent specialist investigation and diagnosis they have continued supervision of the patient. Even when the patient presented initially in hospital the practitioner would subsequently be fully informed by the specialist. It could be argued that a patient normally on the doctor's list might have presented to or continued under the care of a partner and therefore would be unknown to the respondent. This factor would of course also operate in the reverse direction and therefore should not materially affect the overall number of cases. Furthermore, many practitioners replied as units, and some were in the possession of detailed diagnostic registers.

Positive replies gave the name of the hospital where the diagnosis had been established after investigation. Often additional information was volunteered and respondents were well aware, in the additional information provided, of the distinction made between idiopathic dilated cardiomyopathy and cases secondary to pregnancy, alcoholic excess, or other factors contributing to heart failure. If in the initial reply any queries were raised about the accuracy of the diagnosis of dilated cardiomyopathy, or whether a history of alcohol intake or pregnancy could possibly be pathogenetically linked to the heart failure, the practitioner was asked for additional information to eliminate any mistaken inclusion of these patients. Only patients with dilated cardiomyopathy, as defined,¹

were included in the study.

This preliminary study has therefore provided an assessment of the prevalence of overt dilated cardiomyopathy. Clearly, other regions and a greater number of the population must now be investigated. We hope that this report will act as a catalyst for others to undertake similar studies.

We thank Dr G Janacek, University of East Anglia, for statistical advice, and the Royal College of General Practitioners for their help.

This study was undertaken as part of the Multicentre Research study on dilated cardiomyopathy of the Scientific Council on Cardiomyopathies of the International Society and Federation of Cardiology.

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