

Multiple sclerosis in island populations: prevalence in the Bailiwicks of Guernsey and Jersey

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Abstract

The aim of this study was to establish for the first time the prevalence of multiple sclerosis in the Bailiwicks of Guernsey and Jersey, as representing the most southerly part of the British Isles. All patients with multiple sclerosis in the Channel Islands resident on prevalence day were identified by contacting all medical practices, Multiple Sclerosis, and Action Research for Multiple Sclerosis societies by letter and visits. The crude overall prevalence rates were 113/100 000 (95% confidence interval (95% CI) 90.3-135.7) and 86.7/100 000 (95% CI 63.3-110.0) for the Bailiwicks of Jersey and Guernsey respectively. When standardised to the age and sex structure of a previously reported Northern Ireland population the standardised prevalence ratios were 120.2/100 000 (95% CI 96.0-144.3) for Jersey and 95.6/100 000 (95% CI 69.9-121.3) for the Bailiwick of Guernsey. When compared with recent studies in the northern United Kingdom the prevalence rates for multiple sclerosis in the Channel Islands lend some support to the proposed latitudinal gradient in the British Isles although the standardised prevalence ratio in the Bailiwick of Jersey is similar to those found in recent studies of southern Britain. The standardised prevalence rates of probable and definite multiple sclerosis for the male populations were 37.3/100 000 (95% CI 17.9-56.7) for the Bailiwick of Guernsey and 45.5/100 000 (95% CI 26.3-64.7) for the Bailiwick of Jersey whereas the standardised prevalence rates for the female populations were 97.5/100 000 (95% CI 73.9-143.5) and 139.5/100 000 (95% CI 112.6-181.2) respectively. Thus there is a striking and unexplained 43% higher prevalence of probable and definite multiple sclerosis in the female population of Jersey compared with that of the Bailiwick of Guernsey. This seems to be due to an unusually low prevalence of the disease among the female population of the Bailiwick of Guernsey compared with that of the United Kingdom mainland.

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Multiple sclerosis is a common cause of seri-

ous neurological disability in young adults; it affects some 60 000 people in the United Kingdom and perhaps two million people worldwide.¹ The disease shows an unusual geographical distribution in becoming commoner with increasing distance from the Equator in both the northern and southern hemispheres.² There is evidence for a similar geographical gradient in the British Isles.² Different studies have reported multiple sclerosis to be 1.9 to 3.1 times commoner in women than men and to have a peak age of onset of about 30 years, being rare in childhood and after the age of 50.² The clinical features, sex ratio, and age specific incidence curves for multiple sclerosis are similar whatever the underlying frequency of disease, suggesting the same worldwide aetiology.²

Among the many epidemiological surveys of multiple sclerosis in the past 40 years, several intriguing reports have studied the prevalence of the disease in geographically defined island populations. The Orkney and Shetland islands have the highest prevalence of the disease ever recorded—namely, 309 and 184 per 100 000 respectively.³ The prevalence on Sardinia⁴ and Sicily⁵ has been reported to be over 45 per 100 000, whereas on nearby Malta the prevalence is tenfold lower.⁶ Clustering of cases of multiple sclerosis has been recorded on Orkney⁷ and on the small island of Key West off the coast of Florida.⁸ An "epidemic" of multiple sclerosis in the Faroes after occupation by British troops has been proposed,⁹ and a similar rise in prevalence after troop incursions has been claimed on Orkney,¹⁰ Iceland,¹¹ and Sardinia.¹² That the incursions caused the apparent increase in the prevalence of multiple sclerosis in these situations has in most cases been disputed.²

The Channel Islands lie 70-100 miles south of the coast of England between 49° and 50° latitude and about 10-30 miles west of the French coast, with the nearest island (Alderney) only eight miles from the Cotentin peninsula. The British Channel Islands are not part of the United Kingdom, but are crown dependencies allied to the English crown since the Norman invasion. They represent the most southerly part of the British Isles. These Islands were invaded by Germany in June and July 1940 and liberated in May 1945¹³ and were the only part of the British Isles occupied in the second world war. The islands are divided into the Bailiwicks of Guernsey and Jersey. The Bailiwick of Guernsey consists of Guernsey, Alderney, Sark, and other small islands, and

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the Bailiwick of Jersey contains several small uninhabited islands with only Jersey itself populated. Health care services in the two Bailiwicks are independent of the NHS and of each other. Also, as a popular holiday destination and an attractive residential choice due to favourable tax concessions, the Channel Islands are not as geographically isolated as some other islands—for example, Orkney and Shetland.⁷ No previous survey of the prevalence of multiple sclerosis in the Channel Islands has been carried out. The present study was undertaken to determine the prevalence of multiple sclerosis in the two Bailiwicks in the light of recent estimates of the prevalence of multiple sclerosis in southern Britain,¹⁴⁻¹⁸ and the proposed north-south gradient in the prevalence of multiple sclerosis in the British Isles.²

Methods

CASE ASCERTAINMENT

A list of potential patients with multiple sclerosis was compiled with information obtained from several sources, as the Southampton study¹⁶ had shown the value of this approach. The medical practitioners from both Bailiwicks were a major source of information. We also received help from the Multiple Sclerosis and Action Research for Multiple Sclerosis (ARMS) Societies in both sets of islands, and also from the hyperbaric oxygen unit on Guernsey. The nearest British major referral centre for neurology is the Wessex Neurological Centre at Southampton; this has become the predominant specialist referral centre for the Channel Islands but many patients have been seen elsewhere. Also, a consultant neurologist from Southampton visits Jersey regularly. Consultant neurologists at the Wessex Neurological Centre were therefore approached.

From these sources we compiled a provisional list of potential cases of multiple sclerosis covering the Bailiwicks of Guernsey and Jersey. This was then used for a more detailed study. Medical practitioners were contacted again by letter and requested to provide details (name, date of birth, sex, and whether the patient was aware of the diagnosis) of any patients under their care with multiple sclerosis who were alive and resident on the Channel Islands on prevalence day.

After this, consent was sought to approach the patient and obtain a venous blood sample for a separate genetic study. Prevalence days were 10 March 1991 for the Bailiwick of Jersey and 21 April 1991 for the Bailiwick of Guernsey. These days were chosen to coincide with local census days on each set of islands. (The difference between the two prevalence days is not considered relevant to the results of the study.) Further letters were sent to doctors not replying to initial enquiries and non-responders were contacted by phone, or personal visit, or both.

CLASSIFICATION OF PATIENTS

The provisional list of possible cases of multi-

ple sclerosis was then subjected to careful investigation. Sufficient clinical details to allow classification by both the criteria of Poser *et al*¹⁹ and of Allison and Millar²⁰ were sought. This was obtained either by three authors (GS, SEP, AL) being allowed access to the patients' notes or by the medical practitioners filling in a detailed questionnaire. The hospital notes of patients who had attended the Wessex Neurological Centre were also studied. Local Multiple Sclerosis Societies were requested to ask their members to supply details of place of birth and age of onset of the disease. Forty eight patients on Jersey and 36 on Guernsey and Alderney who volunteered had a venous blood sample taken by a member of the team.

STATISTICAL ANALYSIS

Population statistics were obtained from the census taken on prevalence day on Jersey,²¹ Guernsey,²² and Alderney.²³ No formal census data are gathered on Sark and it was estimated that 560 people were resident there: we did not find a single case of multiple sclerosis in this population and have excluded it from further analysis. To allow for differences between islands in age and sex structure and to allow comparison with published work, all crude prevalence data have been standardised against the 1961 Northern Ireland population²⁴ by two different methods. The indirect method was used to calculate standardised prevalence ratios with 95% confidence intervals (95% CIs) for each Bailiwick. To test the significance of differences in prevalence between the Islands and the 1961 Northern Ireland population for specific age groups, it was assumed that the observed number of cases follows a Poisson distribution with a mean equal to the expected number of cases under the standard prevalence rates. The direct method was used to calculate age standardised prevalence rates for each Bailiwick and the difference in standardised rates between islands, along with 95% CIs.²⁵

Results

The provisional register of potential patients with multiple sclerosis in the Channel Islands contained 193 names; of these 45 were eventually excluded because they were not resident on prevalence day ($n = 8$), or because the diagnosis of multiple sclerosis could not be confirmed ($n = 9$), or the person was not traced ($n = 15$). Additionally, one patient was registered with more than one practice and six were registered under both married and maiden names. Of the 45 patients 21 came from Jersey and 24 from Guernsey. A diagnosis of multiple sclerosis was accepted if it was confirmed by information from the patient's medical practitioner or from the medical notes. Some excluded patients had other diagnoses—for example, hereditary spastic paraparesis (which can mimic multiple sclerosis) ($n = 4$), Parkinson's disease ($n = 1$), or benign intracranial hypertension ($n = 1$). Others had suspicious symptoms but no signs

(n = 4). Every medical practice in the Bailiwicks of Guernsey and Jersey responded to this survey. Table 1 shows the number of resident cases found in each Bailiwick on prevalence day together with the recorded populations from the official census returns. It also shows the 53 patients from the Bailiwick of Guernsey and the 95 patients from Jersey classified according to the criteria of Allison and Millar²⁰ and those of Poser *et al.*¹⁹ Ninety four per cent of the patients of the Bailiwick of Guernsey and 88% of the patients on Jersey came under the "probable and possible" Allison and Millar classification²⁰; 85% of the patients of the Bailiwick of Guernsey population and 88% of the Jersey population were fully classifiable under the system of Poser *et al.*¹⁹ Of those who could not be readily classified under the Poser system 10 could not be categorised even with full clinical details (mainly those with progressive spinal cord disease) and with the remainder (n = 9) we were unable to attain sufficient clinical details to make classification possible. The four patients from Alderney in the Bailiwick of Guernsey (one male and three female) were all in the clinically definite category. Of the total identified cases of multiple sclerosis, 19 (13%) had a positive family history but in only 5% of cases was a first degree relative also affected. Only one affected sib pair was found on Guernsey, and one patient with a normal twin was resident on Jersey, as was one male patient with a sister not resident in the Channel Islands but being investigated for the disease elsewhere.

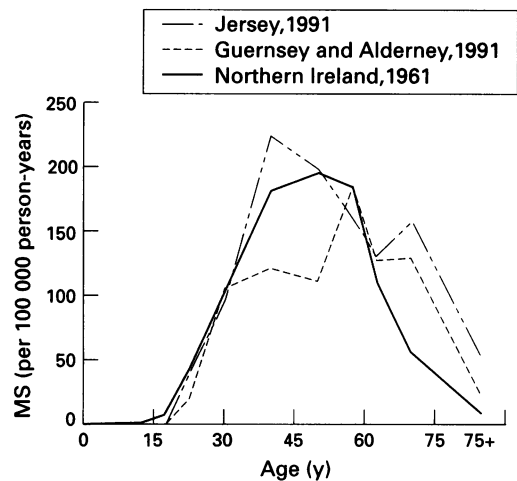
Table 2 shows the overall crude preva-

lences and the standardised prevalence ratios with the populations of the two Bailiwicks age and sex standardised against the 1961 Northern Ireland population.²⁴ Also shown are the figures from other recent studies in the south of the United Kingdom.¹⁴⁻¹⁸ The census reports showed that the age and sex structures of the populations of the two Bailiwicks were almost identical (results not presented). Further analysis was confined to patients with multiple sclerosis who came into the "definite and probable" categories of the criteria of Poser *et al.*¹⁹ These comprised 85% of the patients from Guernsey and Alderney and 88% of the patients from Jersey. The figure shows the age specific prevalence of probable and definite cases of multiple sclerosis for both sets of islands compared with that of the Northern Ireland population.²⁴ Whereas the overall shape of the distribution is similar in all three cases (especially at earlier ages), both Bailiwicks have higher prevalences in older age groups than the original Northern Ireland study.²⁰ At ages 60 and older, 8.35 cases would have been expected in Jersey by applying the prevalence of the standard populations, but 18 cases were found. This excess was highly significant (p = 0.003). At these age groups in Guernsey and Alderney, 12 cases were observed compared with 6.95 expected (test for excess p = 0.5). Table 3 shows the crude prevalence rates and the age adjusted rates for male and female cases in the Channel Islands. Surprisingly, whereas the male populations only showed a difference of 8.2 cases per 100 000 males between the two

Table 1 Clinical classification of patients with multiple sclerosis on the Channel Islands

	Bailiwick	
	Guernsey (Population 61 624)	Jersey (Population 84 082)
Total cases recorded	53	95
Female:male sex ratio	2.6:1	3:1
Alison and Millar criteria ²⁰ :		
Probable	42	78
Possible	8	6
Early	3	11
Poser <i>et al.</i> criteria ¹⁹ :		
CDMDS	40	80
LSDMS	1	1
CPMS	2	1
LSPMS	2	2
Unclassifiable	8	11

CDMDS = Clinically definite multiple sclerosis; LSDMS = laboratory supported definite multiple sclerosis; CPMS = Clinically probable multiple sclerosis; LSPMS = Laboratory supported probable multiple sclerosis; Unclassifiable = Unclassifiable with Poser *et al.* criteria.



Age specific prevalence of probable and definite cases of multiple sclerosis (MS).

Table 2 Comparison of Channel Islands study with other recent south British studies

	Jersey 1991	Guernsey and Alderney† 1991	Cambridge 1990	Southampton and SWHHA 1987	SE London (Sutton) 1985	SE Wales 1985
No of cases	95	53	374	411	195	441
Population	84 082	61 164	288 410	411 000	169 600	376 710
Crude prevalence rates/100 000	113 (90.3-135.7)	86.7 (63.3-110.0)	130 (117-143)	99 (89-109)	115 (99-131)	117 (106-128)
*SPR	120.2 (96.0-144.3)	95.6 (69.9-121.3)	121 (108-133)	115 (108-133)	129 (111-127)	139 (126-150)

*Standardised prevalence ratio. The figure shown is the one that would have been obtained if these populations had the same age and sex structure as the population surveyed in Northern Ireland 1961²⁰ and taking the prevalence found in that study as = 100.¹⁴

†The Bailiwick of Guernsey consists of Guernsey, Alderney, and Sark. The population of Alderney was 2297 with four recorded cases (one male, three female). No cases were recorded on Sark (population estimate 560) which was excluded from further analysis. SWHHA = South West Hampshire Health Authority.

Table 3 Age standardised prevalence rates of probable and definite multiple sclerosis in male and female populations in the Channel Islands

	Crude prevalence	Age standardised* prevalence
Males:		
Bailiwick of Guernsey	47.6/100 000	37.3/100 000 (95% CI 17.9–56.7)
Bailiwick of Jersey	53.8/100 000	45.5/100 000 (95% CI 26.3–64.7)
Females:		
Bailiwick of Guernsey	110.2/100 000	97.5/100 000 (95% CI 73.9–143.5)
Bailiwick of Jersey	162.0/100 000	139.5/100 000 (95% CI 112.6–181.2)

*To Northern Ireland 1961 crude prevalence = 65.8/100 000 (male) 94.6/100 000 (female). Difference in age standardised prevalence between two Bailiwicks: male = 8.2 cases/100 000 (19.8 to 36.1), *p* value for difference = 0.6; female = 42 cases/100 000 – 6.4 to 90.3) *p* = 0.09.

Bailiwicks (95% CI 19.8–36.1, *p* = 0.6), the female populations showed a profound difference, with a large excess of 42 cases per 100 000 of the female population seen on Jersey (95% CI – 6.4 to 90.3, *p* = 0.09). The mean age of male patients with multiple sclerosis was 49.6 for the Bailiwick of Guernsey and 53.6 for Jersey and the corresponding mean ages for female patients were 46.1 and 43.8 respectively. We were able to establish the year of onset of the disease in 46 (87%) of the patients on Guernsey and Alderney and in 69 (73%) of the patients on Jersey. There was no evidence of clustering by year of onset among these patients, as claimed in a previous Orkney study.⁷ We were unable to find reliable information on the prevalence of multiple sclerosis in the Channel Islands before the second world war and in only one patient in this study was the onset of the disease before 1940. Among the patients in whom age of onset was established there was a tendency for it to be earlier in the female (but not male) patients on Guernsey than in those on Jersey. We were able to establish the place of birth in 40 (75%) of the 53 patients in the Bailiwick of Guernsey and in 57 (60%) of the 95 patients on Jersey. Of these, 70% of the patients on Guernsey and Alderney and 58% of the patients on Jersey were native to their respective islands. From the census returns^{21–23} 65% of the total population of Guernsey and Alderney were born on the islands with 77% of the remaining 35% of population originating elsewhere in the British Isles. On Jersey 52% of the total population are native with 76% of the remaining 48% coming from elsewhere in the British Isles.

Discussion

The present study is the first survey of the prevalence of multiple sclerosis in the Channel Islands. The methods used were similar to other recent studies in southern England.^{16,18} Because of the geographically circumscribed island nature of the populations studied, the many sources of case identification, and the high response rate to inquiries, we believe that the estimate of numbers of patients in the Bailiwicks of Guernsey and Jersey is accurate. As the Channel Islands lie 70–100 miles south of the United Kingdom mainland, information on the

prevalence of multiple sclerosis there is of value in deciding whether a latitudinal gradient in the prevalence of the disease exists in the British Isles as seems to occur on a worldwide basis.² Whereas the current age and sex standardised prevalence ratio of 120.2 (if 100 = Northern Ireland 1961) in the Bailiwick of Jersey is little different from the standard prevalence ratios in Southampton (115),¹⁶ Cambridge (121),¹⁸ and Sutton (129),¹⁴ it is considerably lower than the most recent survey of north east Scotland (221).²⁶ The Bailiwick of Guernsey has an even lower prevalence ratio of 95.6. These figures overall would therefore lend at least some support to the proposed latitudinal gradient in prevalence of multiple sclerosis in the British Isles,² at least between northern and southern Britain. Between the two Bailiwicks the age specific prevalence profiles of multiple sclerosis for probable or definite cases show significantly higher prevalence in older age groups (60+) on both sets of islands (Jersey, *p* = 0.003; Guernsey, *p* = 0.5) than in the Northern Ireland population, which presumably reflects improved longevity of patients in the 30 years since that study was performed.²⁴ The familial incidence of multiple sclerosis in the Channel Islands also seems to be lower than in some surveys² and only seven (5%) patients have a first degree relative also affected.

The striking finding in the present study is the unexpectedly high prevalence of female patients with probable or definite multiple sclerosis on Jersey compared with the Bailiwick of Guernsey (139.5/100 000 *v* 97.5/100 000), which largely accounts for the difference in overall prevalence between the two populations (table 3). The female:male ratios of the general population of the two Bailiwicks are almost identical (1:1.07 and 1:1.05). Because the methods of ascertainment were the same on both sets of islands and because only probable or definite cases were included, this difference seems to be real. The observed prevalence per 100 000 females on Jersey is in line with that seen in recent surveys of southern Britain.^{14–18} Thus the female prevalence rate in the Bailiwick of Guernsey should be regarded as unusually low compared with mainland Britain. There is a dearth of prevalence data for nearby France, although a recent paper has quoted a figure of 40/100 000.²⁷ We are unaware of any established difference in the genetic make up of the populations of the two Bailiwicks and it is difficult to see why such factors should only apply to the female and not the male population.

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