PREVALENCE STUDY

The prevalence of Huntington's chorea in an area of East Anglia

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SUMMARY. The prevalence of Huntington's chorea in East Anglia was sought by following up a series of different sources, both in hospitals and in the community. The results show that the prevalence of the disease in this area is very much higher than has previously been reported and is comparable to the highest rates known anywhere in the world.

Introduction

A GENERAL study of Huntington's chorea in East Anglia was begun in 1972. One of the early findings was a much higher prevalence of the disease in the area than suggested by previously published work (Critchley, 1934). In 1975 it was higher than that quoted for most studies which had a range of 4-7/100,000 (Myrianthopoulos, 1966). An attempt has been made to analyze the reasons for the change in apparent prevalence and also to explain local concentrations of patients. No attempt has been made to analyze characteristics of the disease or the psychiatric problems it causes in the way that Bolt (1970), Pleydell (1954), and Oliver (1970) and many others have done, although these data are available, as similar findings would almost certainly emerge.

Aim

To assess as completely as possible the prevalence of Huntington's chorea in the northern part of East Anglia and compare the success of the various methods used.

Method

The region originally studied was Norfolk and the Norfolk/Suffolk border, but it soon became clear that fuller information was obtainable within the area covered by the two central Norfolk mental hospitals. For the purposes of this paper this area is conveniently described by an arc of radius approximately 40 miles centred on Cromer, Norfolk (Figure 1).

The number of patients with a diagnosis of Huntington's chorea in the area described was ascertained by various methods listed below. To these were added new cases, as yet undiagnosed, in whom a clear diagnosis was possible owing to their identification during the family studies that were carried out after the initial ascertainment of patients. These patients were identified from several sources, some of which overlapped, thus some patients were identified more than once. It would be foolish to rely too much on numerical differences between the methods of ascertainment, as most of the cases would eventually have come to light using the methods available to the worker in this field (Table 1). Similar diseases such as that described by Cameron in Ampthill, Bedfordshire (Cameron and Crawford, 1974), and Joseph's disease were not included in this study.

Case finding procedures

1. Handsearching all the death and discharge records at the mental hospitals from the beginning of the NHS in 1947. All patients in whom the diagnosis of Huntington's chorea had been made were noted, their medical records borrowed from the relevant hospitals and studied with particular reference to next of kin and family history, both past, present and future (children). The date and place of birth were noted in particular.
2. The indexes of the general hospitals were searched for numbers in the *International Classification of Disease* covering Huntington's chorea, and those bearing 331.0 (or 355 before 1968) were found and the relevant notes borrowed.
3. A letter was sent to each general practitioner in the area requesting any information about known cases of Huntington's chorea, or information about families in whom a family history was known.
4. Permission was granted by the Registrar General to search manually the death registers of the area covered

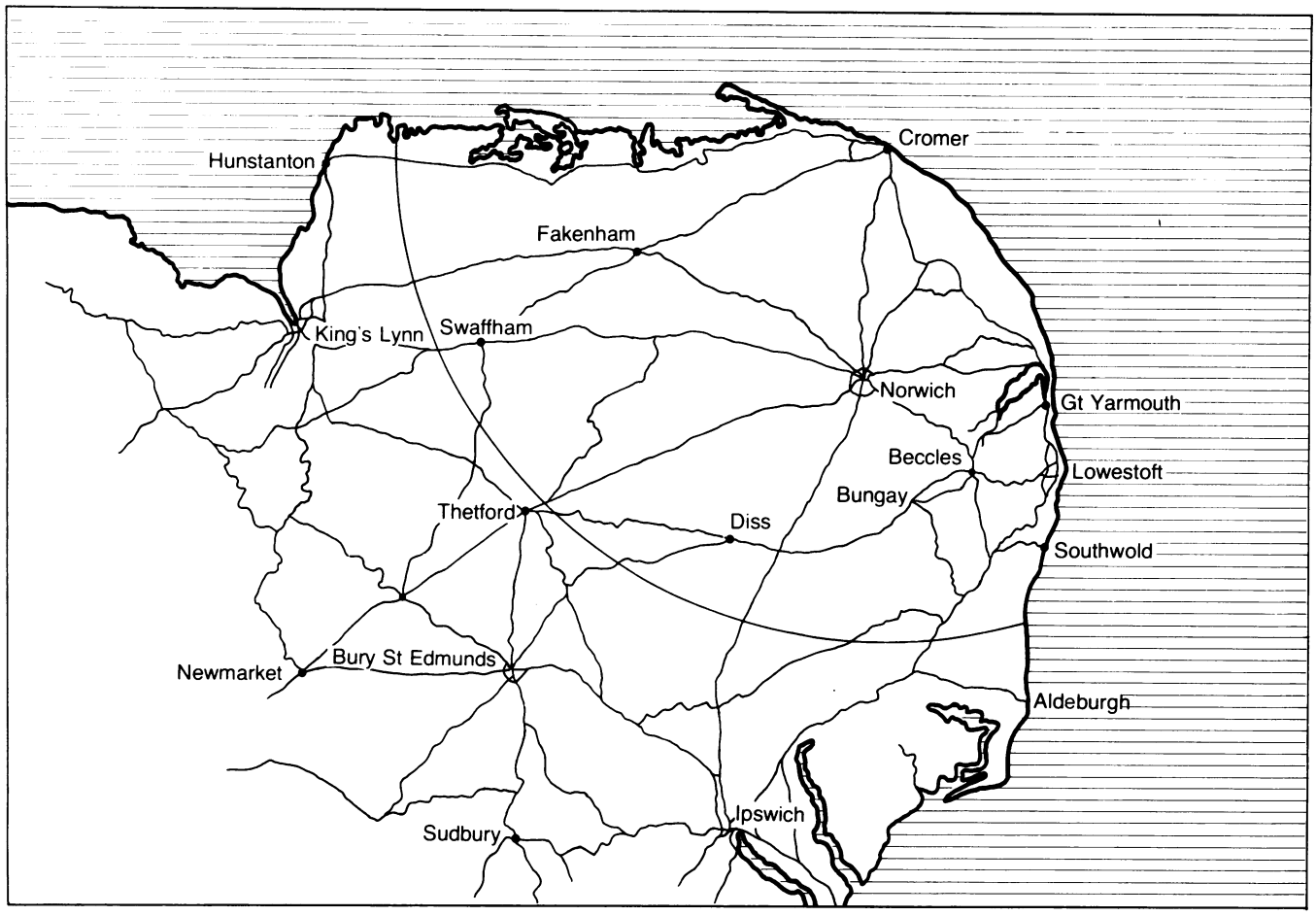

Figure 1. *Map of East Anglia showing study area.*

Table 1. Cases found by different methods of searching.

Method	Dead	Alive	Total
General hospital notes searched under heading *International Classification of Disease*	4	0	4
Mental hospital notes searched under diagnosis	22	5	27
General-practitioner letter	10	14	24
Total	36	19	55

by the two mental hospitals from when they began, in 1837, to the present day. Some 100,000 entries were examined, a note being made of any deaths with a diagnosis that would fit in with Huntington's chorea, or of a person who was a known part or suspected part of one of the kindreds constructed up to that time.

5. Permission was then sought from the general practitioners to visit the families thus identified. The next of kin, children, or sufferer were then asked to provide as much additional information about the family as possible to enable the family tree to be extended backwards into history and laterally in an attempt to join up small family units into large kindreds.

6. Further family tracing was carried out using parish registers, the search being based on the information gathered above.

7. Various other methods of detection arose during the study such as interviewing retired psychiatrists, hearsay, and comments from social workers and allied members of the community services. None of these sources was numerically large nor were the identified people included unless the diagnosis was substantiated by other means.

Results

Using progressively more stringent methods of ascertainment the prevalence of the disease was found to rise to 9.24/100,000, much higher than the figure suggested in the only other East Anglian study (Critchley, 1934). It was found that the prevalence was unevenly distributed in the area and that a considerable pocket existed in Lowestoft. This prevalence is higher than any other figure given for a comparably sized population in the

UK, being exceeded internationally only by the level in Tasmania (Brothers, 1949), but being higher than all other areas studied throughout the world (Shokeir, 1975; Myrianthopoulos, 1966; Pratt, 1967).

Discussion

The previous study in East Anglia (Critchley, 1934) used methods of ascertainment which produced a zero prevalence rate of Huntington's chorea in Norfolk. However, retrospective family studies enabled 20 cases to be identified at the time when the above study was carried out. Critchley's findings did not support the common belief that there was an unusually high prevalence of Huntington's chorea in East Anglia due to its historical connections with the New England group of choreics. However, his conclusions drawn from the above historical connections (Critchley, 1934, 1964, 1973) are substantially incorrect (Hans and Gilmore, 1969; Caro and Haines, 1975).

I have tried to show the effects of using other studies' criteria on the data from our own catchment area to demonstrate how these criteria will change the prevalence rate in a given area (Table 2). This would agree with the statement that prevalence rates are not strictly comparable owing to differing methods of ascertainment (Myrianthopoulos, 1973).

Using the information collected about these families it is possible to estimate the number of people within stem families suffering from Huntington's chorea during the last century. These figures are, of course, crude, but are unlikely to exaggerate the numbers. Table 3 shows the prevalence due to studied families and absolute numbers in Norfolk using this technique from 1851 to 1971 by decade.

The East Anglian rise from 1851 to 1971 probably contains a real increase, but is considerably distorted and exaggerated by family errors in detection, and loss of patients with the passage of time, the incredible rise being largely artefactual. However, it does demonstrate that a small number of sufferers in known families could be the forefathers of a large number of present-day cases. There is no evidence from this study to suggest that a similar number of families have died out (none of the kindreds end), although equally there is no proof that they have not. It has been shown (Reed and

Table 2. Prevalence rates reported by different authors.

Method	Names of users	Number produced	Prevalence/10^5
Mental hospital inpatient medical certificates	Critchley (1934)	7	1.2
Circular letter and hospital notes and nuclear family studies	Pleydell (1954) Heathfield and Mackenzie (1971) Bolt (1970)	35	5.1
Extended kindred tracing	This paper	54	9.24

Table 3. Prevalence rate.

Date	Population × 10^3	Number of patients with Huntington's chorea	Prevalence/10^5
1851	385	3	0.78
1861	360	2	0.56
1871	396	2	0.51
1881	404	4	0.99
1891	415	7	1.68
1901	441	10	2.27
1911	465	8	1.72
1921	474	6	1.26
1931	475	20	4.21
1941	495	22	4.45
1951	513	27	5.27
1961	528	39	7.39
1971	583	54	9.24

Palm, 1951) that inheriting the gene for Huntington's chorea increases the reproductive fitness as compared to the unaffected members of a family and this may be one of the causes of the increased prevalence.

Lowestoft, which is in the catchment area, has a population of 53,000 and a prevalence rate of over 30 per 100,000. This is six or seven times higher than the average for the country as assessed by various other studies (Critchley, 1934; Pleydell, 1954; Oliver, 1970; Heathfield and Mackenzie, 1971).

In the Moray Firth area the effects of localized populations shows an unusually high concentration of sufferers (Lyon, 1962), but the population he drew from was very small, and was calculated, it appears, from the electoral roll, thus missing out many people who should otherwise have been included in the population figures. However, he did recognize some of these shortcomings. Using an area with the same population as that used in this study we can produce even higher prevalence rates depending on which area of a town we include in the study (Table 4). In this table it can easily be seen that the prevalence rate falls to the average for the town as one leaves a fixed spot where there are sufferers in one household.

Heathfield (1973) suggests that localized raised incidences may be due to heterozygotes marrying, although I fail to see that this will make any difference to the gene frequency. In fact, it will reduce it if homozygotes do not survive through to breeding age. However, in Lowestoft these marriages do not appear to have occurred. Unlike the Moray Firth study (Lyon, 1962) where cases appear to be in related families, as far as it can be discovered the Lowestoft cases are from 11 separate families, very nearly half from within East Anglia and half immigrants to the area. This finding is very different from Great Yarmouth, another coastal town a mere nine miles away, where the prevalence is apparently no higher than the UK average. Both towns now have a population of just over 50,000, but in 1841 Great Yarmouth had a population of just half this (24,250), whereas Lowestoft contained less than a tenth of its current population (4,837). It was probably the vast influx of people into Lowestoft, which was a rapidly developing town, that caused this large difference in prevalence—less stable families being more mobile and perhaps moving to avoid the stigmas of the past or being less able to keep employment in their home areas owing to mental and social instability.

The Registrar General's figures show an increase of over 100 per cent in the deaths from Huntington's chorea since 1959 (Table 5). The Registrar's figures shown are an underestimate (Caro, 1976), but nevertheless a considerable rise is shown. This may be in part due to better diagnosis, but probably contains a real increase.

The usually accepted prevalence of the disease is 4-7/100,000. In 1972 48.8 million people were recorded living in England and Wales (Office of Population Censuses and Surveys, 1974). Using the maximum figure, if 7/100,000 have Huntington's chorea this gives a suffering population of 3,416.

The disease runs a 15-year course and so about 1/15

Table 4. Prevalence in a town from a fixed point.

Area in square miles	Number of patients	Population	Prevalence/10^5
0	2	5	40,000
0.05	3	250	12,000
1.76	4	8,800	45.45
4.31	6	21,550	27.80
8.70	11	43,500	25.28
10.00	16	54,000	30.18

Table 5. Registrar General's figures.

Years	*1959*	*1960*	*1961*	*1962*	*1963*	*1964*	*1965*
Death from Huntington's chorea	46	49	59	60	66	70	70
Total deaths × 10^3	605	526	550	557	572	535	549
Years	*1966*	*1967*	*1968*	*1969*	*1970*	*1971*	*1972*
Deaths from Huntington's chorea	77	90	74	72	80	90	100
Total deaths × 10^3	564	542	577	580	575	567	592

of these (227) will be dying each year. The mean age of death for Huntington's chorea patients is 56 years (Bell, 1934; Bird *et al.*, 1974), the average death rate for the two decades surrounding this age is about 10 per 1,000, so a small proportion will be dying of other conditions (about 34 people). The expected number of deaths from Huntington's chorea is 193, considerably more than shown by the Registrar's data.

Conclusion

The prevalence of Huntington's chorea in East Anglia is seen to be considerably higher than in most studied areas of the UK. Using the usually accepted criteria for making the diagnosis, in some areas of East Anglia the rates were very much higher than other local areas of England. This conflicts with work that has been done in East Anglia in the past and reasons for this have been discussed.

The information obtained from this study points to some important approaches which must be considered in order to gather anything like accurate information about the prevalence of a familial disease. If positive action is envisaged in the future for limiting the propagation of such disease, the information revealed in this study may prove to be valuable.

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