However, a few patients in our series do show considerable differences between the degree of narrowing in their coronary, carotid, and iliac systems. We should perhaps remember that this is a necropsy survey; that although the disease may progress in parallel in different arterial systems, there will be individual variability; that the vessels studied are of differing size and importance; and that the crucial event in producing clinical symptoms and death in patients with diseased vessels is not the wall disease alone but the occurrence of thrombotic occlusion. Thus patients in whom the wall disease progresses steadily in all affected sites, and who do not develop thrombosis, are more likely to be found in the living population than in a necropsy survey, and in a necropsy study such as ours they will be most common in the older age-groups of the unselected sample. On the other hand, patients in the younger age-groups dying of cardiac infarction or strokes may be those in whom the disease has progressed more rapidly in some arterial sites than in others and in whom thrombosis has supervened.

Summary

The prevalence of coronary artery disease was studied in an unselected necropsy sample of 137 patients and in a selected group of 79 patients with myocardial infarction; arterial plaques in the aorta and carotid and iliac arteries were also studied in these patients and in a further series of 156 unselected necropsies and 37 patients with myocardial infarction (a total of 293 unselected patients and 116 patients with infarction).

In individual patients severe coronary narrowing is often associated with stenosis of the carotid and iliac arteries. A group of patients with myocardial infarction have more aortic disease than a group of unselected patients of comparable age and sex, the area affected by complicated plaques accounting for most of the difference.

A group of patients with myocardial infarction have more stenosis and more plaque ulceration in the carotid and iliac arteries than unselected patients of comparable age and sex. This study suggests that patients who have severe arterial disease in one site are likely to have severe disease elsewhere.

We are grateful to Sir George Pickering for advice and encouragement and to Dr. A. H. T. Robb-Smith and the staff of the Morbid Anatomy Department for access to the necropsy material. Miss Sheila Briers, Mr. Bruce Abrahams, and Mr. Paul Manners gave invaluable technical assistance throughout.

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HUNTINGTON'S CHOREA IN THE MORAY FIRTH AREA*

BY

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The syndrome of chronic adult chorea accompanied by insanity and with a hereditary basis has for many years been associated with the name of George Huntington following his description of it in America in 1872. In Britain the first recorded account was by West (1887), who described the relationships of an affected family at Stoke-on-Trent. As the syndrome became more widely recognized many individual cases were put on record, so that by the end of the nineteenth century the condition had been demonstrated from Bristol (Clarke, 1897) to Leith (Elder, 1899).

Macdonald Critchley (1934) gave an explanation of how at least some of the American families came of Suffolk stock; he also gave figures which illustrate a fairly wide incidence over Britain.

A comparatively rare disease which runs in families should be found more frequently in some areas than in others, and, in fact, localized collections of cases of Huntington's chorea have been described in Cornwall and Northampton by Bickford and Ellison (1953) and Pleydell (1954) respectively.

A further centre of Huntington's chorea is detailed here. It is hoped to illustrate how the disease can remain localized so long as it is confined to a closed community, but that the current social tendencies have now scattered potential carriers in fact from Ullapool to Southampton, from Canada to Australia.

It has been recognized for some years that Huntington's chorea is common in the Moray Firth area, especially among the fishing families. The point of focus which has the highest incidence and the place from where all the families originate is a small fishing village on the east coast of Ross-shire. Mentioning in particular the surnames of Patience and McLemmon, a note has already been made by McWilliam (1937) to point out the presence of the disease in this area and to ponder its source. The personal details and family connexions of the affected cases have had to wait until now to be disentangled.

Present Investigation

Source.—The origin of these peoples defies all clarification, and it is thus impossible to link them to any of these other known communities. Numerous theories have been advanced. It is clear that men skilled in herring fishing were settled here about 300 years ago, but it is not known if the original families brought Huntington's chorea or if the disease was introduced later by brides who may have come north to boost the weaving industry. For many years this fishing community refrained from marrying outside their own village or at least always married into fisher families. Thus the clans are even yet completely unaffected.

Material.—Many difficulties were unsurmountable because details had to be checked from times too long ago, and other difficulties arose even in the assessment

*Abridged and adapted from a thesis accepted for the Doctorate of Medicine, Edinburgh University.

of individuals who can still be remembered. As degenerative cerebral changes appear in many of the older age-groups, and mental deficiency tends to be common with the in-breeding found here, it was practicable, in a large number of cases, to consider only the symptom of chorea. Seven families have been investigated in this series, and it is highly probable that they can all be linked together. They have been traced back for about 150 years in most instances, but there are so many uncertainties associated with information earlier than this that an origin common to these families has not become apparent.

The following family trees and personal details have been compiled.

Family C

C, II 1.—Wife of a fisherman. Admitted to a mental hospital in 1895, aged 63, with established chorea as well as delusions of persecution, and may have had alcoholic tendencies.

C, III 2.—Daughter of above. Became badly disabled by chorea about the age of 50 but retained her faculties well enough to manage at home. Died from exposure in a severe snowstorm (1923).

C, II 5.—It is suspected that this patient's mother may have been the one to pass on the disease. He was a hard-working fisherman most of his life, renowned for his skill and indifference to risk and danger. He never exhibited any psychiatric upset, but shortly after he retired at the age of 65 the chorea started and was confined to the head and arms.

C, III 7.—Son of above but of a very different disposition, tending to nervousness and unable to take responsibility. After six years of heavy drinking he was admitted to a mental hospital in 1931 mainly because of his violence, but settled well, presumably when the alcohol was withdrawn.

C, IV 13.—Daughter of above. A nodding of the head was first noticed at the age of 50. Gradually a shaking developed and spread, interfering with her dressing. Only a slight tendency to depression. Now, 15 years after the onset, she is still at home and is ambulant.

C, IV 14.—Sister of above. Moved to Glasgow on marriage. Onset of symptoms when aged 40. Survived 12 years, and latterly was severely demented.

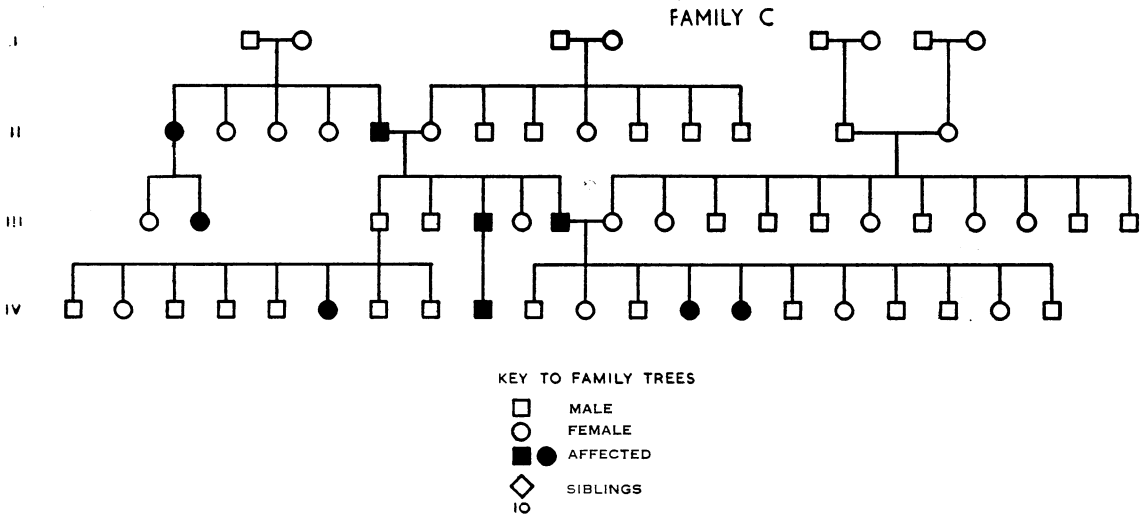
C, III 5.—Son of C, II 5. A robust hard-working fisherman until the last two years of his life. Said to be a very intelligent man but with a liking for drink. Chorea started at the age of 61, but advanced only a little in two years before he died with a cerebral haemorrhage.

C, IV 9.—Son of above, aged 42, has shown symptoms for eight years. Tendency to simplemindedness ran in mother's family and is shown by this patient as a complicating feature.

C, IV 6.—The patient's father, a brother of C, III 7, was lost at sea aged 44 without having developed an obvious chorea. This patient moved to London on marriage. Symptoms started at the age of 40, and nine years later she had moderate incapacity.

Family K

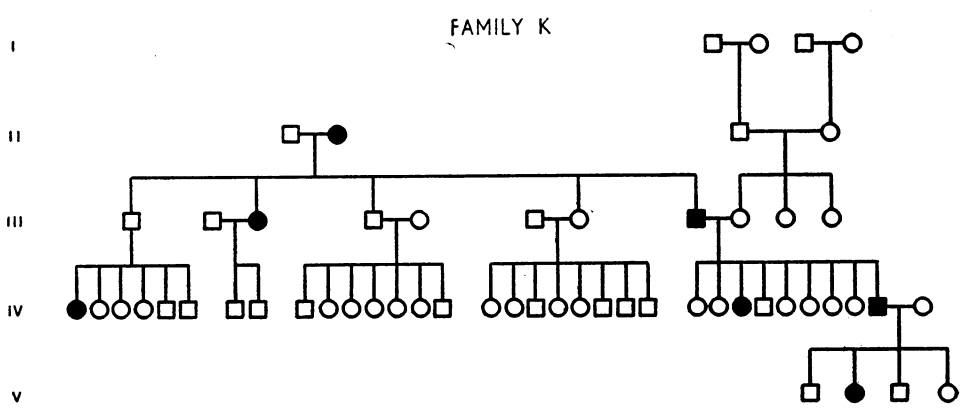
K, II 2.—No adequate details are present about the patient's parents, but an unconfirmed story suggests that she may have been a sister of C, I 2. She is still remembered with terror by the older villagers, as she was choreic and aggressive for the last 15 of her 80 years.



K, III 8.—Son of above. Was small as a child, but later grew tall and very strong. Employed as a fisherman. Lived to the age of 80 and developed mild choreiform movements during his last five years.

K, IV 26.—Daughter of above. A fisherman's wife. Developed athetosis at the age of 48. Maintained her mental composure up to the night she died. Laparotomy a year earlier had revealed a colonic neoplasm.

K, IV 32.—Son of K, III 8. From mother's side he inherited mental deficiency. Strong religious tendencies as a youth and for 20 years was a street-corner preacher. When aged 59 he received a head injury and after this a coarse tremor



developed in the hands. During the next year the chorea spread to involve the whole body. Liable to alternating depression and violence. Still living five years after start of chorea.

K, V 2.—Daughter of above. Symptoms started at the age of 31 during her second pregnancy nine years ago. Mood tends to fluctuate and chorea is more obvious when she is depressed.

K, III 3.—Fisherman's wife. Two sons were mentally defective. Died aged 80 after exhibiting chorea for her last 12 years.

K, IV 1.—Patient's mother came from another Moray Firth port, where Huntington's chorea is unknown. Father died aged 65 without showing disease, but he could have been the carrier. Died of cerebral haemorrhage at the age of 75 after five years of chorea. Marked dementia in the last few years.

Family L

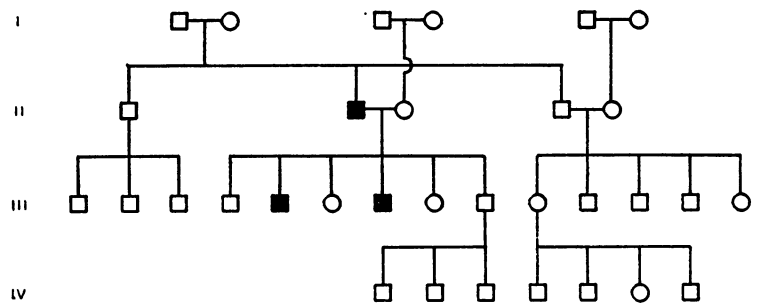
The name for L, I 2 appears on her children's birth certificate, but few memories of her remain. It is possible that she was a sister of P, II 12, and this could be a connexion between Families L and P.

L, II 2.—A fisherman of great muscular strength. Died at the age of 40 after being choreic for 10 years. Suffered also from chronic alcoholism. His wife must have suffered greatly, as she made her children promise never to marry. Only one disobeyed, and his issue are free from the disease so far.

L, III 5.—Son of above. Trained as a fisherman but joined the Navy in the first world war. In 1917 he started to fidget and later developed chorea. Emigrated to Canada and was drowned while bathing. If the suggestion is true that he intentionally waded out into the water until he was overcome, then this is the only case of suicide in the series.

L, III 7.—Brother of above. Served with the Army during the first world war. Symptoms appeared in 1918 and he died at home two years later.

FAMILY L



Family N

Early details of this family are unknown because the marriage certificate of the parents cannot be traced.

N, II 15.—A fisherman; developed only mild chorea during his last two years. Died at the age of 74 of a cerebral thrombosis.

N, III 27.—Had a reputation for being intellectual as a youth. Later took to street preaching. Chorea started at the age of 64 and he died of bronchopneumonia 11 years later.

N, III 10.—State of mother unknown. Patient left village when young to be a policeman in the south. Died aged 75 after being choreic for one year.

N, III 18.—It has been impossible to verify the medical details of this patient's parents, but they are said to have been healthy. This patient worked in the village and then married a fisherman. Chorea started at the age of 71 and spread quickly. In spite of severe disability she was not mentally disturbed.

N, V 1–4.—The grandparents married in the village, then moved to a port further south. They were both said to be from affected families but their connexions are not clear. N, II 8 had chorea as an old woman. N, III 5 died young of pulmonary tuberculosis. All four children are in mental hospitals suffering from Huntington's chorea.

Family P

P II 12.—Patient's mother and her husband's mother were half-sisters. Her mother may have been afflicted. Patient was admitted to a mental asylum in 1892 as a case of chronic mania (a hereditary cause was suspected) with marked chorea. A diagnosis of Huntington's chorea was entered in the record the following year: the first proved diagnosis for this area. Died three years later from pulmonary tuberculosis.

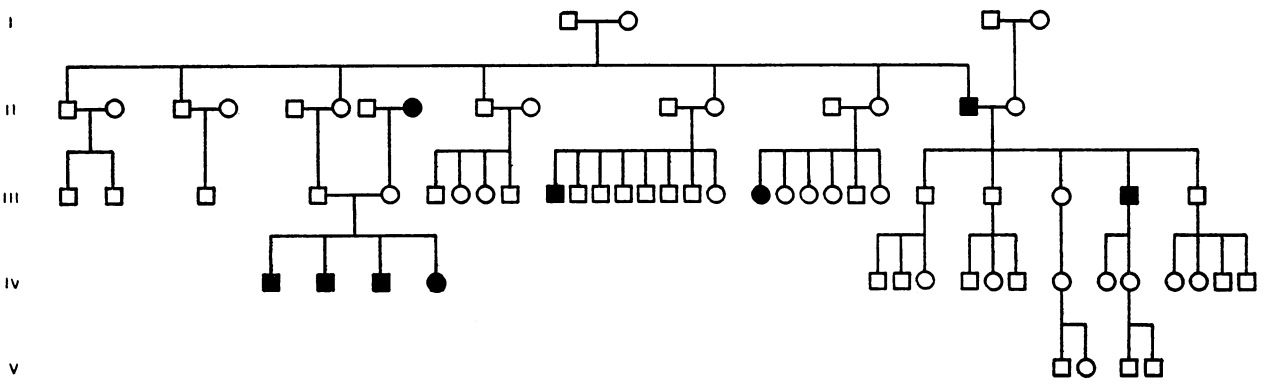
P, III 6.—Son of above. A fisherman. About the age of 44 began to be moody and later dull and depressed. Admitted to local asylum 10 years later as he threatened to harm himself and others. No chorea was noted on admission, but it had developed before his death three years later. Diagnosed as Huntington's chorea.

P, IV 5.—Daughter of above. Married a farm worker. Athetosis started at the age of 42, but it was eight more years before walking was impaired. Became very violent and was removed to hospital. Died 18 months later of a cerebral thrombosis.

P, IV 7.—Daughter of P, III 6. Settled in Newcastle after her marriage and was lost to local knowledge until her daughter started inquiring about the advisability of her own marriage.

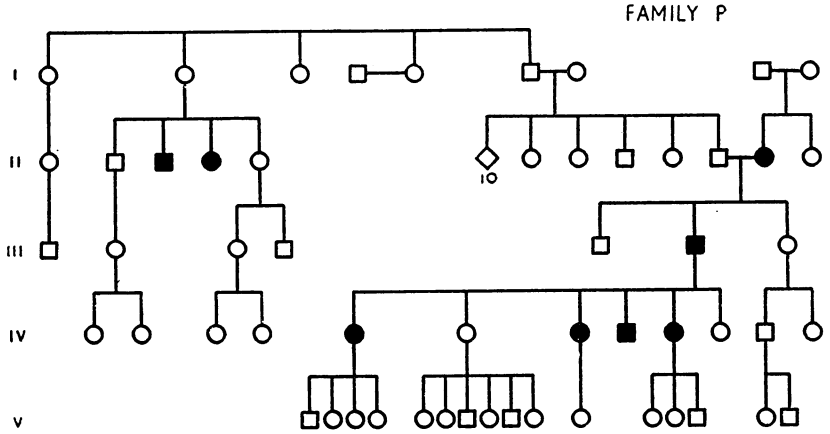
P, IV 8.—Son of P, III 6. Joined the Merchant Navy. Two head injuries. Invalided home in 1943. After examination two years later, diagnosis of pseudodementia. Involved in a charge of indecent exposure in 1948 and Huntington's chorea was diagnosed. Died of bronchopneumonia aged 44.

FAMILY N



P, IV 9.—Daughter of P, III 6. Married a farm worker. Shake of hands started at the age of 32 (14 years ago). Still able to dress herself. Marked dementia.

FAMILY P



P, II 3.—No details available about his parents. Admitted to asylum in 1904 with advanced mental degeneration and marked chorea. Huntington's chorea appears on record as the diagnosis. Died a year later.

P, II 4.—Sister of above. Admitted to asylum in 1894 on account of violence and "hereditary chronic chorea." Died of pulmonary tuberculosis five years later.

Family R

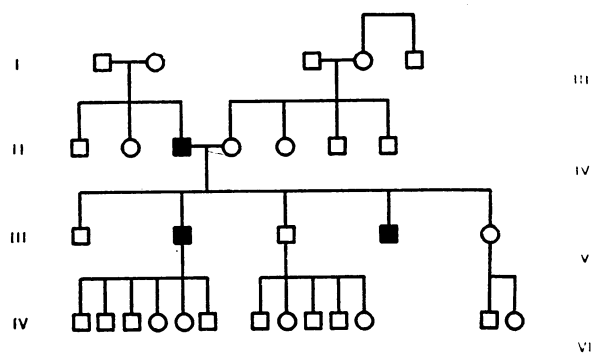
R, I 2.—Was born in 1825 and died aged 25 without showing any disorder. Her husband's second family are clear. R, II 2 died at age of 12. R, II 1 emigrated to Australia and is lost to this review.

R, II 3.—A strong and courageous skipper. Retired from the sea when aged 68, then developed chorea. Deteriorated quickly and died of cerebral haemorrhage two years later.

R, III 2.—A fisherman. Chorea became obvious 10 years ago at the age of 67. At present very frail, with chronic asthma. He exhibits that peculiar catch in his breath which has been described with the disease.

R, III 4.—A fisherman. Admitted to mental hospital in 1952 with athetosis; quarrelsome and degraded. Died of bronchopneumonia in 1959 aged 71.

FAMILY R



Family S

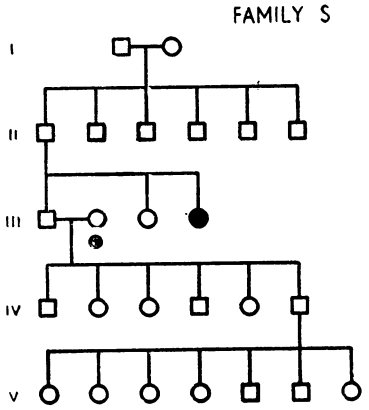
Most of the early details of this family cannot be verified as they have been handed down through the generations. As these may form a link between families they are included for interest.

S, I 1.—Must have been born about 1790 and was of Highland stock and with a clan name. He was said to have married into the fisher village, but was drowned young.

S, III 4.—Moved to another port to the east of the Moray Firth, where she raised a family and has handed on Huntington's chorea after exhibiting it herself in later years.

S, III 2.—Can probably be identified as T, I 2 (married November 1, 1821). Her fate is uncertain, but it is likely that she introduced Huntington's chorea into Family T.

FAMILY S



Family T

This family forsook the sea and for several generations were employed as estate gardeners, moving first to Morayshire then to Easter Ross.

T, II 1.—Can just be remembered as a very old man with a pronounced shake.

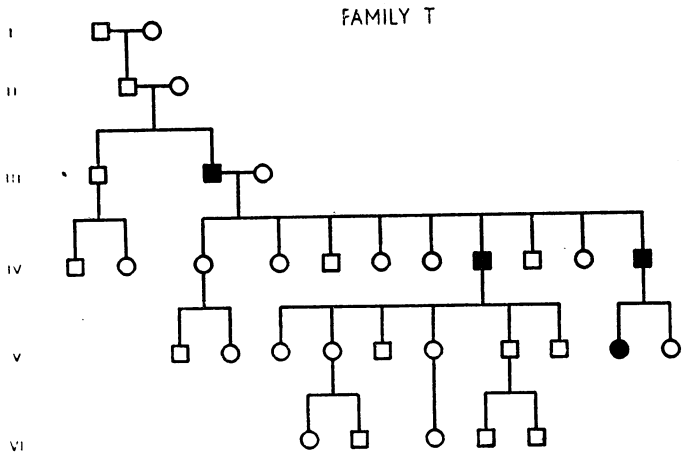
T, III 2.—Patient had a good memory up to the time of his death at the age of 78, but had well-marked chorea for 14 years.

T, IV 8.—Became moody and irritable at the age of 60. Chorea soon developed and he died two years later.

T, IV 11.—Brother of above. Liable to drunken outbursts for many years, and this disguised the true diagnosis. Chorea severe for the last seven years of his life.

T, V 9.—Daughter of above. Married a Londoner and later divorced. Husband and four boys last heard of in London. When she was 43 she became tired, clumsy, unemotional, and showed jerky movements. Now, seven years later, she has gross dementia but very little remaining chorea.

FAMILY T



Analysis of Clinical Details

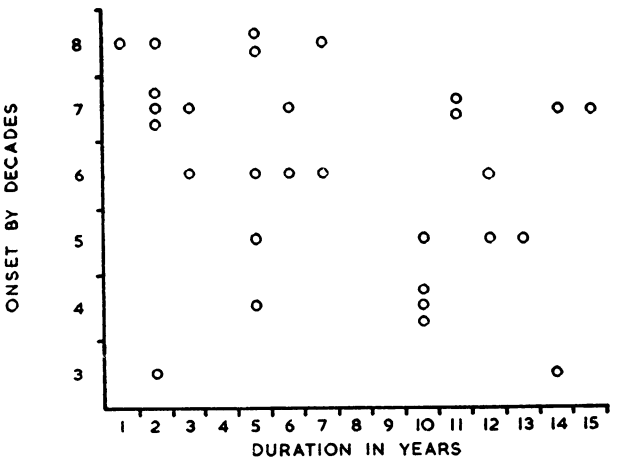
Many difficulties have been encountered which reduce the value of statistical examination of this material. In particular, the number of cases is small, but it seems more important to keep them small and accurate rather than swell the numbers by allowing in doubtful cases.

Incidence.—At present there are 896 names on the village voters roll, and although 12 of the above patients are alive only five were born in the village, representing

an incidence of 0.56%, which is 100 times the incidence of Cornwall or Northampton. No useful conclusions can be drawn in a disease so irregularly spread.

Age of Onset.—In 41 patients the average age when symptoms started is 51.59 years (S.D. ± 15.6). The earliest was at 27 years (P, II 3), the latest at 75 years (K, III 8). These figures show a much later onset of symptoms than the usually quoted figure of 35 years.

Duration.—The average time that patients have survived after first exhibiting chorea has been calculated as 7.13 years (S.D. ± 4.5) in the 29 who have died. The longest is 15 years. Twelve patients still survive, having shown symptoms for 5 to 20 years. When the results are graphed some conclusions can be drawn (see Chart). Conditions in the village have changed greatly over the years this survey covers. A high infant mortality rate, the prevalence of tuberculosis, the hazards of the sea,



Scattergraph plotting survival time and age of onset of chorea in 29 patients.

the state of nutrition, and the sanitation have all improved greatly and affected survival and longevity. The Chart shows a fairly wide scatter. The increased density of the upper left quadrant is in keeping with the extra hazards of old age irrespective of Huntington's chorea. There is no suggestion that people affected early in life survive only a short time. No double population pattern appears to support the theory of anticipation which seemed likely during the clinical inquiries.

Heredity.—Huntington's chorea is a hereditary disease due to a dominant gene and therefore with a 50% chance of inheritance. These pedigrees show that 20 affected parents had 108 children, of whom 27 were affected, which is 25.93%. The data can be broken down further (Table I). Social customs may have influenced these results. The fishermen occasionally brought brides from other ports but seldom wandered off themselves. In this community unromantic courting customs are still remembered. Thus a girl from an affected family would have a poorer chance of matrimony.

TABLE I.—*Numbers and Condition of Children from Parents Affected with Huntington's Chorea*

	Affected		Clear		Total
	Male	Female	Male	Female	
Mother, 6 ..	3	4	15	10	32
Father, 14 ..	12	8	29	27	76
Either, 20 ..	15	12	44	37	108

Anticipation.—By analysing large series of cases both Davenport (1916) and Bell (1934) have discounted the importance of anticipation in Huntington's chorea. While some suggestion of it is given clinically, too few generations are presented to show a true pattern, but any such impression is destroyed by setting out the overall picture of some of the likeliest families.

TABLE II.—*Age of Onset of Chorea in Some Patients, Arranged According to Their Generation in Their Families*

Generation	Family C	Family K	Family P	Family T
I				
II	65, 59	65	44	
III	50, 57, 61	75, 69	55	64
IV	40, 50, 40, 42	48, 59, 70	42, 43, 32	60, 56
V		31		42

Fertility.—Even a casual survey of the pedigrees shows no evidence of infertility among these patients with Huntington's chorea. Celibacy has been demonstrated in only one family (L, II 2), but may well have been practised silently by other individuals. The strict morality of these fishermen who have been under scrutiny is noteworthy. Liaison might have been expected from the men when visiting other ports of the North-East of Scotland, but all the cases of Huntington's chorea which have been discovered are the issue of legal unions. This is in strong contrast with the American writings but in agreement with the statement by Russell Brain (1952).

Blood Groups.—Pleydell (1954) has already shown that the allelomorph is unconnected with the blood groups. While there are only seven living cases in this locality they fall into groups AB, A, and O with rhesus-positive and rhesus-negative.

Electroencephalogram.—It has been practicable to arrange E.E.G. recordings in only five cases of this series. Only non-specific changes are reported in each of these.

Developmental Defects.—Careful search and inquiries have failed to discover any congenital deformity or disease common to this community beyond the mental deficiency, already mentioned, which is not particularly associated with the families affected with Huntington's chorea.

Psychiatric Aspects.—It has already been explained that analysis of psychiatric details is impracticable in this series because of the confusion with the mental deficiency of the present as well as the lack of details in patients long since dead and the alcoholism of the past. Some impressions have been gained by those looking after this group and are worth recording.

The real psychosis of Huntington's chorea occurs a few years after the chorea is established, and yet many people who have practical dealings with the early stages of the disease prefer to base their diagnosis on behaviour changes rather than on neurological features. Moodiness, irritability, and neglect of home and person are all features which have been shown by these patients. With both K, V 2 and C, IV 13 the house is the best index of the patient's condition, and the appearance of neglect or disorder will herald a spell of worsening of chorea.

Some of the external influences affecting these patients are easily revealed on admitting them to hospital. Excitement tends to make the chorea more pronounced

and these patients tend to be more excitable than normal. They may be made worse by the excitement of attending a clinic or being admitted to a ward, while some are made worse even by a visit from a friend. From hospital records it is apparent that many patients have deteriorated on arrival in a ward and gradually improved spontaneously over two to three days.

Neglected nutrition and personal tidiness are often found in affected patients, who will improve greatly after these are corrected in hospital or when the patient is induced to live with relatives. Incidental febrile illness such as influenza has often been found to accentuate the choreic symptoms in our patients. The influence of alcohol and the improvement following abstinence has frequently been demonstrated in this series.

Religious tendencies were noted in some patients but assumed serious proportions only in the cases of K, IV 32 and N, III 27. Superstition and religion are commonly intertwined with the life of any fishing community and are well in evidence here.

Features of persecution, aggression, and grandiose behaviour are occasionally found in the hospital records.

"That form of insanity which leads to suicide" is part of Dr. George Huntington's original description of the disease. Bickford and Ellison (1953) have observed a high incidence of suicide in their series in Cornwall and give this as a reason for some of their cases receiving awards for gallantry during the war. The fishermen of North-East Scotland have long had a reputation for skill, endurance, and courage. This is probably a feature of the breed, whether affected or not, and as the patients here reviewed have a complete lack of insight it is unlikely that self-destruction need be feared here. Suicide has been suggested for only one patient (L, III 5), but of course cannot be verified.

Sexual forms of mental disturbance have been observed by American writers, who claim that it is commonly part of Huntington's chorea. This tendency does not occupy a prominent place in the accounts in this country and was encountered only once (P, IV 8) in this series.

Neurology.—Accurate information about the neurological changes is available for a very few of the cases in this series. The majority appear to have presented the typical picture, and the spread of the disease has followed the usual pattern, with individual variation covering a wide range. One case (K, IV 26) showed an increase in muscle tone suggesting an extension of the disease process to neighbouring areas. The involvement of the respiratory muscles by chorea is reported by Julia Bell (1934) as a great rarity. An example of this complication has been noted in R, III 2.

Summary

Huntington's chorea appears to have been first recognized in the Moray Firth area in 1893.

The disease is not found in the families of Highland stock, and some of the reasons for this are given. The affected patients can all claim their origins from one small fishing village in Ross-shire which was probably settled about 300 years ago. The place whence those peoples came is uncertain.

The earliest case of Huntington's chorea diagnosed by implication is S, II 1, who was probably born about 1810.

The pedigrees of seven affected families have for the first time been disentangled and are presented with some suggested links between them.

Some psychiatric aspects are discussed and mention is made of the clinical features employed locally for early diagnosis.

For the great interest and encouragement shown in this work I thank Dr. J. Ronald. Both he and Dr. Martin Whittet have kindly made all their hospital records available. Without the very willing help of the physicians, psychiatrists, local practitioners, and the district registrars, this survey could never have been completed.

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NON-HEREDITARY CHRONIC ADULT CHOREA AS A CLINICAL ENTITY

BY

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When Dr. George Huntington (1872) first described the form of chronic adult chorea which now bears his name he emphasized "its hereditary nature," stating that "it is confined to certain and fortunately a few families, and has been transmitted to them, an heirloom from generations away back in the dim past."

The appearance of Huntington's chorea in patients whose parents have been free from the disease has long been the subject of debate, and family circumstances are seldom clear enough to be acceptable as proof. In the Moray Firth area we find a sufficiently regular pattern of spread to permit conclusions being drawn from the breeding habits of patients' ancestors in assessing the likelihood of hereditary disorders.

During a recent survey of the disease in this area it has been shown that all the choreic patients can trace the family origin to a small fishing village in Ross-shire. These fishermen appear to have settled here about 300 years ago and to have kept themselves quite apart, marrying within their own community or occasionally bringing brides from other ports. In contrast to this nest of Huntington's chorea, two patients were discovered who appear to resemble true cases of Huntington's chorea so closely that they were in fact given this diagnosis, but on scrutiny they lack the necessary hereditary background.