BRITISH MEDICAL JOURNAL

LONDON SATURDAY NOVEMBER 13 1954

HUNTINGTON'S CHOREA IN NORTHAMPTONSHIRE

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Huntington's chorea is a fatal hereditary disease which is due to a dominant gene. The average age of onset is 35.5 years and the average duration of the disorder 13.7 years (Bell, 1934), but wide variations occur in both these factors. The involuntary athetoid movements are considered to be associated with the atrophy of the ganglion cells of the caudate nucleus and putamen; the mental deterioration with atrophy of the cerebral cortex; and rigidity, when it is present, with involvement of the large cells of the globus pallidus.

Although Huntington's chorea is described as a rare disease, it is probable that systematic search and accurate recording would reveal the incidence to be higher than is generally supposed. No attempt appears to have been made to obtain a true assessment of the number of cases in this country, and it was felt that a central Midland county such as Northamptonshire with a population of approximately 263,000 might provide an indication of the incidence of the disease. It was decided, therefore, to ascertain the number of cases in the county, and for this purpose the following procedure was adopted. (1) A questionary was sent to the senior partner of every practice in Northamptonshire asking whether or not there was a case of Huntington's chorea in the practice. The information thus obtained from each practice denoted the existence of six choreic families. (2) With the consent of the medical superintendent of the mental hospital which serves the town and county of Northampton, the records of two certified patients who were admitted from the county area were made available and their pedigrees traced back from information provided by relatives.

Inquiries of this nature can be undertaken only by medical observers, and this illustrates the value of medical rather than lay administration in a health service. The results of the survey indicated the existence of eight choreic families in the county; in one of these families the disease appears to have died out, but, as is frequently the case, marriages have been broken and relatives have separated, so that a complete history could not be obtained. Sixty-one cases of the disease were revealed by tracing back the pedigrees, and 13 patients are at present alive in the county. The details of the family histories are as follows.

Pedigree One (Fig. 1)

I 1:—Died in the Northamptonshire asylum; the cause of his insanity is not recorded.

II 1 (1834–87):—Suffered from Huntington's chorea and died in the asylum at the age of 53.

III 1:—Age of onset of disease not known; death aged 48 in the mental hospital. III 2:—Onset aged 40; death aged 48 in the mental hospital. III 3:—Onset aged 38; death aged 45. III 4:—Onset aged 40; death aged 57 in the mental hospital. He suffered also from pulmonary tuberculosis. III 5:—Onset aged 42; mania as well as chorea present; death aged 52. III 6, III 7, and III 8 did not inherit the disease, and no information is obtainable about the descendants of III 6 and III 7. III 9:—Onset aged 40, death aged 50. His wife committed suicide when she realized she had passed the disease on to her youngest child.

IV 1:—Onset aged 40; died aged 50 in the Poor Law Hospital. IV 2:—Onset aged 40; died aged 53 in the Poor Law Hospital. IV 4:—Onset aged 40; unable to concentrate or carry out simple functions; mental deterioration; speech indistinct; gait unsteady. Admitted to a mental hospital and diagnosed as Huntington's chorea. Died aged 53. IV 5:—Aged 46; onset aged 35 with involuntary movements of limbs and trunk; hesitant speech; her face is expressionless. Her gait is unsteady and she is unable to stand still; there is slight spasticity, and mental deterioration is present; she is unemployable. IV 6:—Aged 42; onset aged 34. Speech affected first, and at present his speech is of an explosive character; his face is expressionless. He has a festinating gait and there is spasticity of the limbs; mental deterioration is present and he quickly loses his temper; he is unemployable. IV 9:—Onset of disease aged 40 with characteristic involuntary movements. Died at 51. IV 11 was aware of the hereditary nature of the disease. He enlisted in the first world war, caring little whether he was killed or not. He was decorated twice, and after the war he married, but deliberately abstained from having children. IV 12:—Onset of disease aged 43 with difficult, slurred speech; later involuntary movements. Death aged 54. IV 13:—Onset aged 27, with difficulty in speech, followed by choreiform movements. Death aged 30 as a result of an accident. IV 14:—Onset at 27 with dyslalia; died aged 34. Her baby died a day after birth. IV 16:—Like his brother, IV 11, he was well aware of the hereditary nature of the disease. He also enlisted in the first world war with little regard whether he was killed or not, and was decorated three times. The onset of the disease was first noticed with choreiform movements at the age of 28 after demobilization, and he died aged 35. IV 20:—Onset aged 39 with involuntary movements and facial grimaces; slight spasticity present. There is mental deterioration, and she appears to be feeble-minded; she is unemployable. Present age, 42 years. IV 22:—Onset aged 18 with choreiform movements; died aged 26.

V 1–V 6:—No detailed information is available regarding this branch of the family, but the history that was obtained did not indicate that any member was suffering from Huntington's chorea. V 10:—Suffered from a nervous breakdown two years ago and was treated as an out-patient in the mental hospital. V 14:—Age of onset 18 years, while

she was in the A.T.S., with involuntary choreiform movements; mental deterioration. Admitted to a mental hospital in Northamptonshire, where a diagnosis of Huntington's chorea was made; later transferred to a mental hospital in Surrey. V 16:—Onset aged 19; now aged 26. He received a normal education and was employed before the war; he was accepted into the Army, but later was invalided out. Since his discharge from the Army he has not been employed. He has appeared before a magistrate's court on minor charges and for the past five years has been content to remain without any work in Part III accommodation. Here his mental deterioration became more and more marked, until at last his unkempt appearance and his curiosity in the opposite sex necessitated his removal to a mental hospital.

In 1953 his mental age assessed on the Stanford Binet intelligence scale Form L was 10 years 7 months and his intelligence quotient 71. Later he was examined with the Wechsler–Bellevue intelligence scale Form I, and on this he obtained an intelligence quotient of 95 on the verbal scale, 69 on the performance scale, and 82 on the full scale. Slight choreiform movements have developed over the past year, affecting the trunk and extremities. He repeats himself continuously, is unable to carry on a conversation, and, apart from occasional grimaces, his face is expressionless.

Nevertheless, despite the fact that both in his social and in his intellectual capacity he was a mental defective, he could not be dealt with as such on account of the age stipulation in the definition, which requires that the defectiveness shall be present before the age of 18 years. This definition, although introduced in order to deal with persons suffering from post-encephalitic defectiveness, associates disease with a sudden chronological event rather than a gradual pathological process, and for this reason is unsatisfactory.

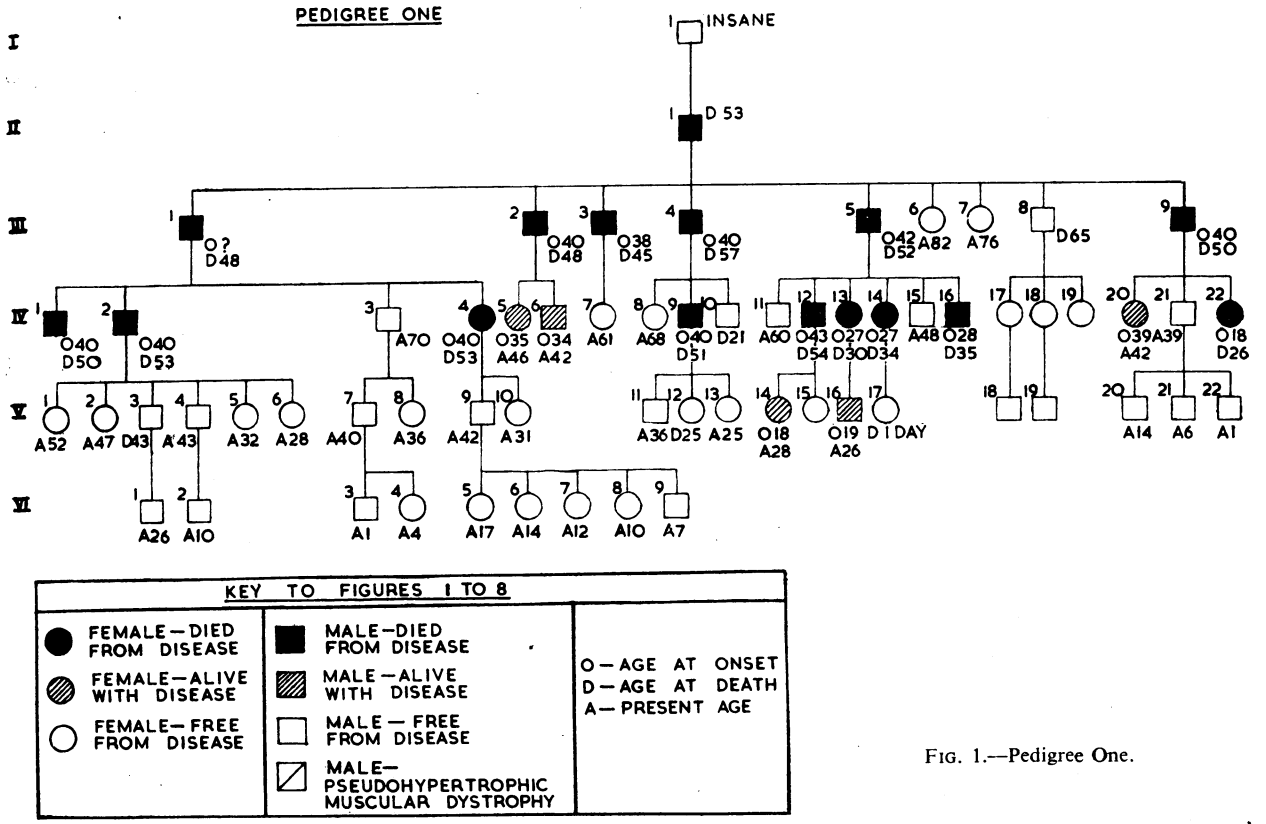

FIG. 1.—Pedigree One.

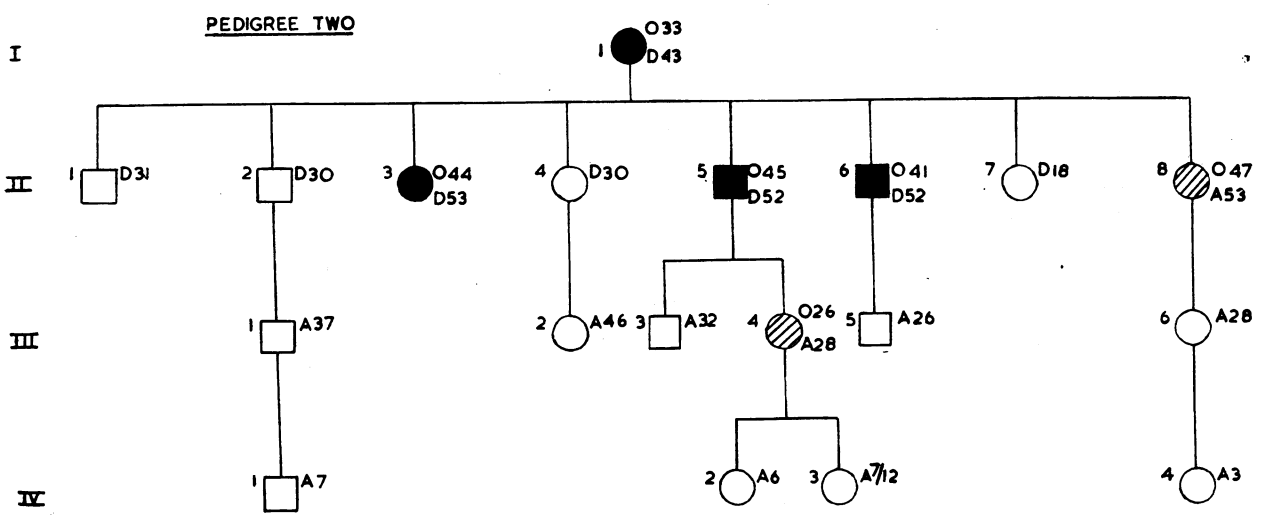

FIG. 2.—Pedigree Two.

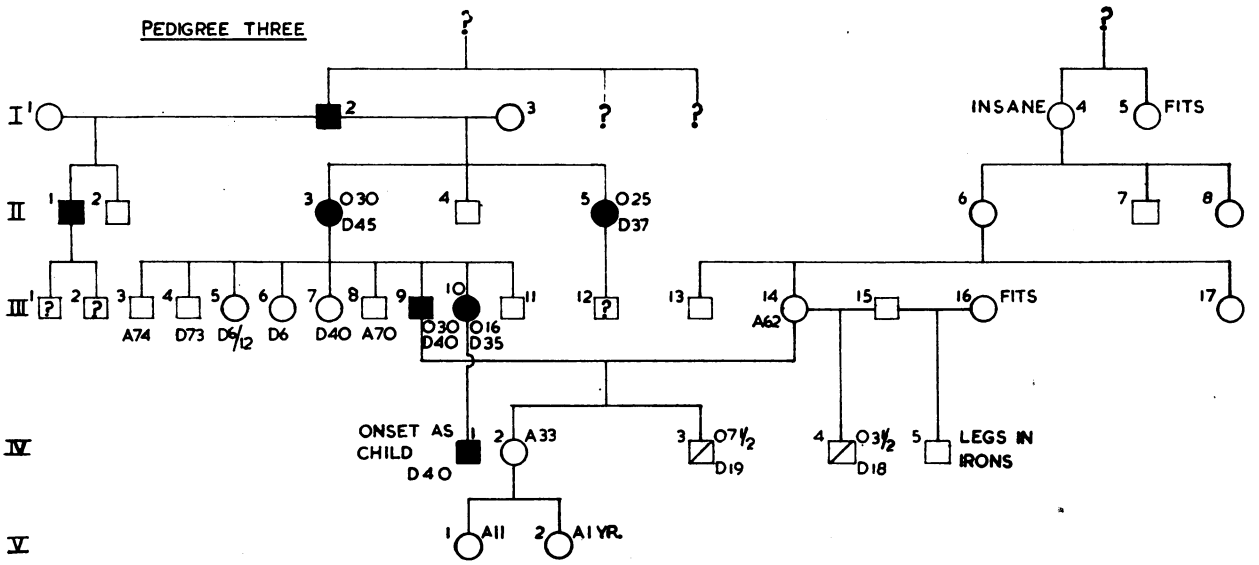

FIG. 3.—Pedigree Three.

Davenport stated that the way in which the disease affected a family might well be characteristic and that the symptoms could breed true in that family. This observation is supported by the information obtained in the present survey. Thus difficulty in speech was frequently the first sign of the disease amongst afflicted members of the family in this pedigree. In all the other families dyslalia occurred late, and Bell remarks on its rarity in the early stages as characteristic of the disease.

Pedigree Two (Fig. 2)

I 1:—Came to the county from Derbyshire; onset of disease at 33; death at 43.

II 1 was killed in the first world war aged 31. II 2 was killed in the first world war aged 30. II 3:—Onset of disease, aged 44; admitted to County Mental Hospital; died aged 53. II 4:—Died from influenza in 1919 aged 30. II 5:—Onset of disease aged 45; admitted to County Mental Hospital; died aged 52. II 6:—Onset aged 41; admitted to County Mental Hospital; died aged 52. II 7:—Died suddenly of heart trouble aged 18. II 8:—Onset of disease aged 47; first noticed when she started dropping things; later developed jerky movements of arms and legs, followed by facial grimaces: mental deterioration only slight. On examination she is unable to remain still; gross movements of limbs and trunk; involuntary grunting noises. Her face is expressionless. Present age 53.

III 3:—No signs of disease; has received hormone therapy on account of infertility. III 4:—Onset of disease aged 26; present age. 28. Physical characteristics: coarse, non-repetitive, involuntary movements affecting arms, legs, and trunk; occasional involuntary grunting noises; perioral twitchings. Mental characteristics: change of character; very excitable with men; nervous and talkative. III 6:—Stated to be nervous by relatives. No evidence of Huntington's chorea present.

Pedigree Three (Fig. 3)

The first member of this family known to be suffering from Huntington's chorea, I 2 married twice, and both wives passed on the disease to the next generation.

II 1:—No details of age of onset and death are available. II 3:—Age of onset 30 years with choreiform movements; death at 45. II 5:—Onset aged 25 with choreiform movements; death aged 37.

III 1:—Unable to walk. III 2:—It is thought that this child was not healthy, but no further details could be obtained about these two children. III 5:—Died aged 6 months. III 6:—Died aged 6 years from bronchitis. III 7:—Died aged 40. III 9:—Onset of disease with choreiform movements, aged 30; died aged 40. III 10:—Onset of disease with choreiform movements at 16 years; died aged 35. III 11:—Has emigrated to Canada and nothing is known of his health.

IV 1:—It is stated by III 3 that he suffered from the disease from birth and that he died at the age of 40. III 9 married a woman (III 14) who carried the gene for pseudohypertrophic muscular dystrophy. One of their two children, IV 3, suffered from the dystrophy at 7½ years and died at 19. III 14 had previously had a baby by another man (III 15), and the child of this union suffered from the dystrophy at 3½ years of age and died at 18. This man (III 15) had also had a child by another woman (III 16), who suffered from fits, and the child born as a result of this union had his legs permanently in irons.

There is a history of insanity and fits in the antecedents of III 14: thus I 4 suffered from insanity and I 5 suffered from fits.

Pedigree Four (Fig. 4)

I 1:—Lived in Scotland and suffered from the family chorea; age at death not known.

II 1:—Free from disease; died aged 60. II 2:—Onset of disease at about 27; death aged 42. II 3:—Onset of disease at about 27; death at about 42.

III 2:—Onset of disease aged 31. Admitted to mental hospital two years ago as she was unable to manage her domestic duties and was worried and emotionally disturbed. Admitted to mental hospital again for the same reason one year ago, when the diagnosis of Huntington's chorea was made. Physical characteristics—typical athetoid movements of arms, trunk, and legs. She is unable to walk steadily. Her face is expressionless. Present age 40 years.

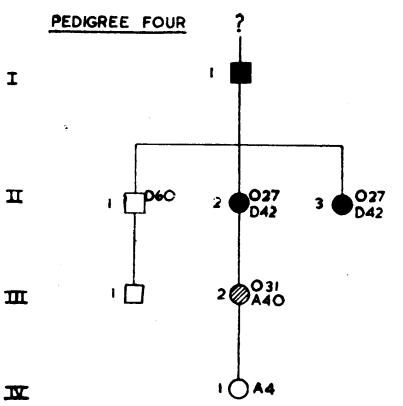

FIG. 4.—Pedigree Four.

Pedigree Five (Fig. 5)

No definite information is available about I 1, but it is known that I 2 was insane.

II 1:—Suffered from insanity and died in the mental hospital, but there is no evidence to indicate that her mental condition was due to Huntington's chorea. II 2 :—Onset aged 46. Died in the mental hospital and is thought to have suffered from Huntington's chorea. There is no record of a definite diagnosis having been made, but the description of the signs indicate that almost certainly this was the condition from which he was suffering.

III 1 is aged 61. She has been in a mental hospital and is still suffering from depression, but there is no evidence that she has contracted Huntington's chorea. III 2 developed the disease at the age of 40 and died in the mental hospital at the age of 57.

IV 1 :—Onset of the disease at the age of 39 with dyslalia followed by jerky movements; mental deterioration occurred late in the disease. She died in the mental hospital aged 51. IV 2 was smothered at the age of 1 year. IV 6 :—Onset at age of 31 with mental peculiarity, dropping things, jerking movements; speech affected five years after onset of chorea. Died aged 43. IV 7 died aged 11 years and is known to have suffered from pulmonary tuberculosis. IV 9 died of pneumonia aged 15 months. IV 11 was killed aged 21. IV 12 died aged 12 of diphtheria. The history indicates that the remaining members of the family do not show any signs of the disease.

The apparently low incidence of the transmission of the disease-bearing gene in this family is probably explained by the fact that 5 of the 13 children in the fourth generation died before the age of 22—that is, before the age at which the disease is generally manifest. It is too early yet to say that the remaining members of this generation have not inherited the disease.

Pedigree Six (Fig. 6)

Of the eight Northamptonshire families suffering from Huntington's chorea, this is the only one in which the origin of the disease can be demonstrated. The first person to suffer from the disease was III 6. His father (II 4) worked in a factory until his death at 70; his mother (II 5) worked at making lace until her death at 71. The history was obtained from III 7, aged 78, who is emphatic that none of her mother's or father's siblings—all of whom lived to between 60 and 70 years—suffered from the disease; she is equally emphatic that III 6 was the only member of her siblings to be affected. In this patient, who died at 73, the disease originated at the age of 55, which appears to be in

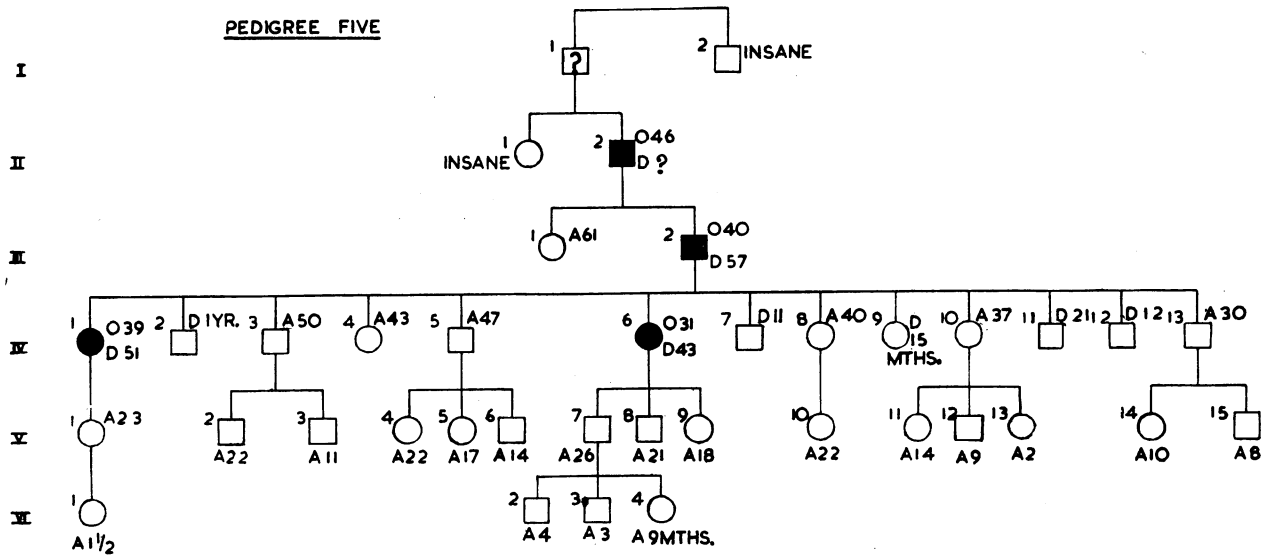

FIG. 5.—Pedigree Five.

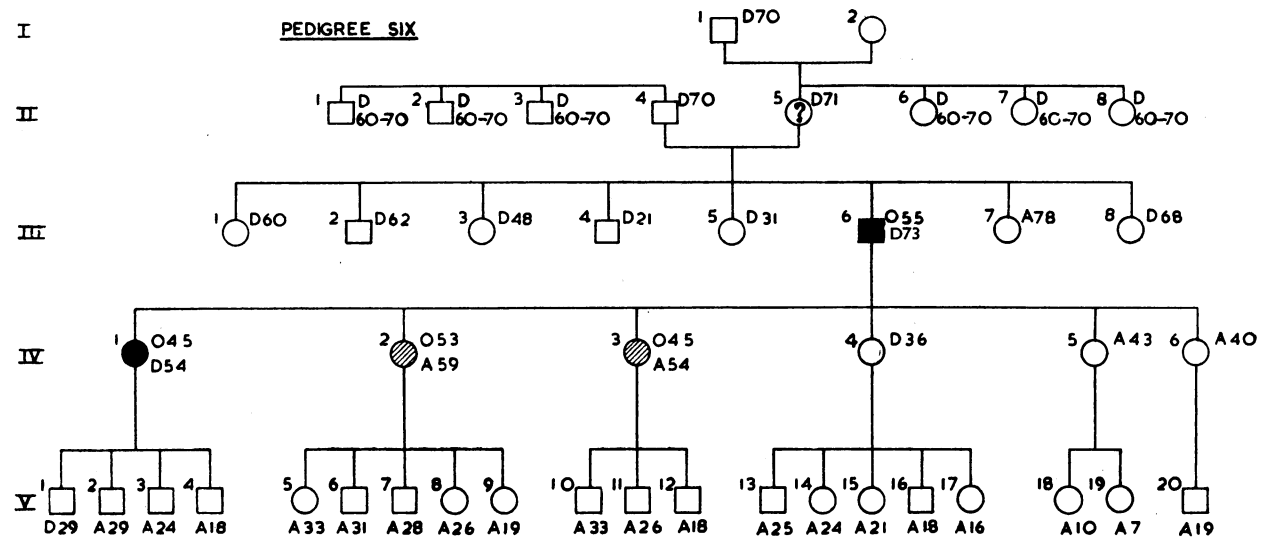

FIG. 6.—Pedigree Six.

conformity with Bell's records of late onset in initial manifestation of the chorea. He had six children, three of whom have so far inherited the disease.

In IV 1 the onset was manifest at the age of 45 and was characterized by scanning speech and choreic movements of the face, mouth, and neck. She also developed writhing movements of the shoulders, arms, and legs. Slight spasticity was present; her gait was unsteady and she used to fall down. She died at the age of 54 in accommodation provided under Part III of the National Assistance Act. IV 2 :—Onset aged 53 with choreiform movements of the fingers, arms, and legs. The progress of the disease has been slow and she is still at work. Present age, 59. She states that she can remember being taken as a child to visit her grandmother, who was in bed and who "shook," but she does not think that any of her other relatives "shook." This statement throws some doubt on the evidence of III 7 about earlier generations. Presumably a person who suffers from the disease is more likely to describe other people as suffering from a similar affliction than a person who is free from the disease, since the former does not wish to consider that she is the only member of the family suffering from "nerves," whereas the latter unconsciously opposes any suggestion that there is a family taint. IV 3, aged 54, is in an advanced state of dementia. There are ceaseless writhing movements and the face is expressionless. The age of onset was 45. IV 4 died aged 36 from ? cancer. Another female sibling in the fourth generation died at the age of 2½ years from "inflammation of the brain." She has not been included in the pedigree since there was uncertainty regarding her date of birth.

It is still too early to say that the remaining two sisters in the fourth generation are not carrying the disease-bearing gene. The three affected sisters have given birth to 12 children, so that the incidence of the chorea may well be doubled in the fifth generation. The history does not indicate that any of these children have been affected as yet.

Pedigree One illustrates how it is possible for the incidence of the disease to diminish in a family when the members become aware of the hereditary nature of the affliction; this pedigree illustrates how the disease can spread when members of a family have no inkling of its hereditary nature.

Pedigree Seven (Fig. 7)

This family came to the county from Wiltshire, and only a relatively small pedigree can be traced.

In I 1 the disease was first noted at the age of 53, and death occurred at the age of 68.

II 1 suffered from Huntington's chorea and died aged 64. II 2 also developed the disease, and died aged 51. II 3 died from pulmonary tuberculosis. No information can be obtained about II 4 or II 5. II 6 died from the disease aged 51. II 7 is in a mental hospital suffering from Huntington's chorea; she is aged 55. In her case she was admitted at the age of 46 in a state of agitated melancholia. This state progressed into dementia, and choreiform movements did not become apparent until she was 51.

The history does not indicate that any of the children of II 1 have been affected as yet.

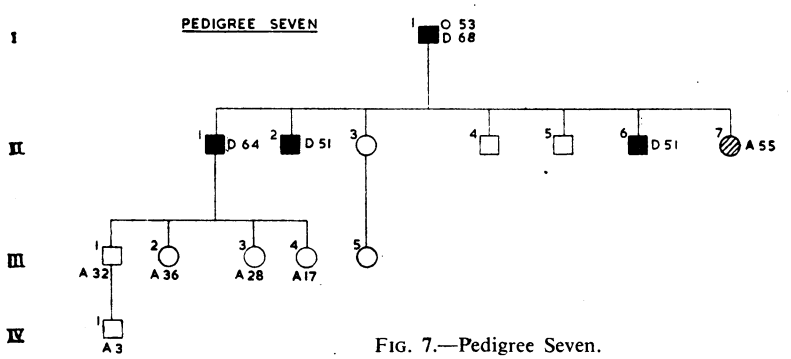

FIG. 7.—Pedigree Seven.

Pedigree Eight (Fig. 8)

I 1:—No cause for the insanity of this person can be established, but it seems probable that she suffered from familial chorea. Of her six children, only II 3 is definitely known to have been affected. The onset in his case was at 43 and death at 51 years. The ages at death of II 2, II 4, II 5, and II 6 are not known.

II 3 had six children, of whom four developed the disease. In III 1 the onset was at 41 years and death at 58. In III 2 the onset was at 45 years and death at 55. In III 3 the onset was at 16 years and death at 36; when she was 19 she had an illegitimate baby. In III 4 the onset was at 17 years and death at 40. III 5 is living, aged 70. III 6 died aged 14—? typhoid fever.

IV 1 is aged 58; onset of disease aged 53; choreiform movements slight, affecting trunk and limbs. IV 2 was killed in 1914 aged 19. IV 3 is aged 53; onset of disease was at 46, with involuntary movements of the shoulders; arms and legs affected later; at present there are jerky movements of the whole body. She appears normal mentally, but her husband states that her behaviour is irrational at times. Her face is expressionless. IV 4:—Aged 47, onset of disease aged 39, with involuntary movements of arms and legs. Unsteady in her gait; speech and mentality appear normal; her face is expressionless. IV 5 died aged 3 years as a result of an accident. IV 7 :—Aged 41; onset of disease aged 34, with involuntary movements of the hands; later the arms and legs were affected, then the speech was involved; and finally mental instability was noticed. At present her athetoid movements are general and almost continuous. She has an ataxic, high-stepping gait, walks on a wide base and not infrequently falls down. She is mentally peculiar, readily becomes angry, and is abusive and violent towards her older son. The younger son is in a children's nursery. IV 8 :—Aged 55; very nervous and emotional; would not be seen. She has been in the mental hospital for anxiety neurosis. IV 9 :—Onset of disease aged 3 years; death aged 12. This information was obtained from III 5 and her husband, who have seen the disease in three generations and are emphatic that the child suffered from the hereditary chorea and not from any other disease.

V 2, aged 34 years, is very nervous, hates crowds, and is of an excitable disposition; does not remain still. He is not classed as a case of Huntington's chorea. V 5 and V 6 were stillborn. V 7 died aged 18 months—"paralysed."

Diagnosis

In a survey of this nature it is necessary that the diagnosis shall be established with certainty. Of the 13 patients in the series who are alive, 12 exhibit the involuntary athetoid movements which are pathognomonic of the disease, while in the thirteenth the criteria for diagnosis are: serious mental deterioration, spasticity, an expressionless face, a stooped posture with a festinating gait, and the gross difficulty in speech which has characterized the disease in his family.

It will be seen that the pedigree is of great value in establishing the diagnosis, particularly in those cases in which insanity precedes the chorea. Jelliffe (1910) stated that some English psychiatrists did not seem to recognize Huntington's chorea and that cases in English asylums were diagnosed as catatonia, dementia, or chronic mania. This observation was reiterated only last year by Bickford and Ellison, who wrote that they had seen cases in Cornwall diagnosed as Parkinson's disease, disseminated sclerosis, neurasthenia with ataxia, and dementia with creeping paralysis. Further evidence is provided by Spillane and Phillips, who, in their inquiry in South Wales in 1937, reported that *only two of the fifteen cases they observed had previously been*

diagnosed; they considered that other cases were hidden away among the population. From this it would seem that accurate collation of family pedigrees by the medical officers of the local health authority would render a useful service both to practitioners and to hospital clinicians in all cases where Huntington's chorea should be excluded from the differential diagnosis.

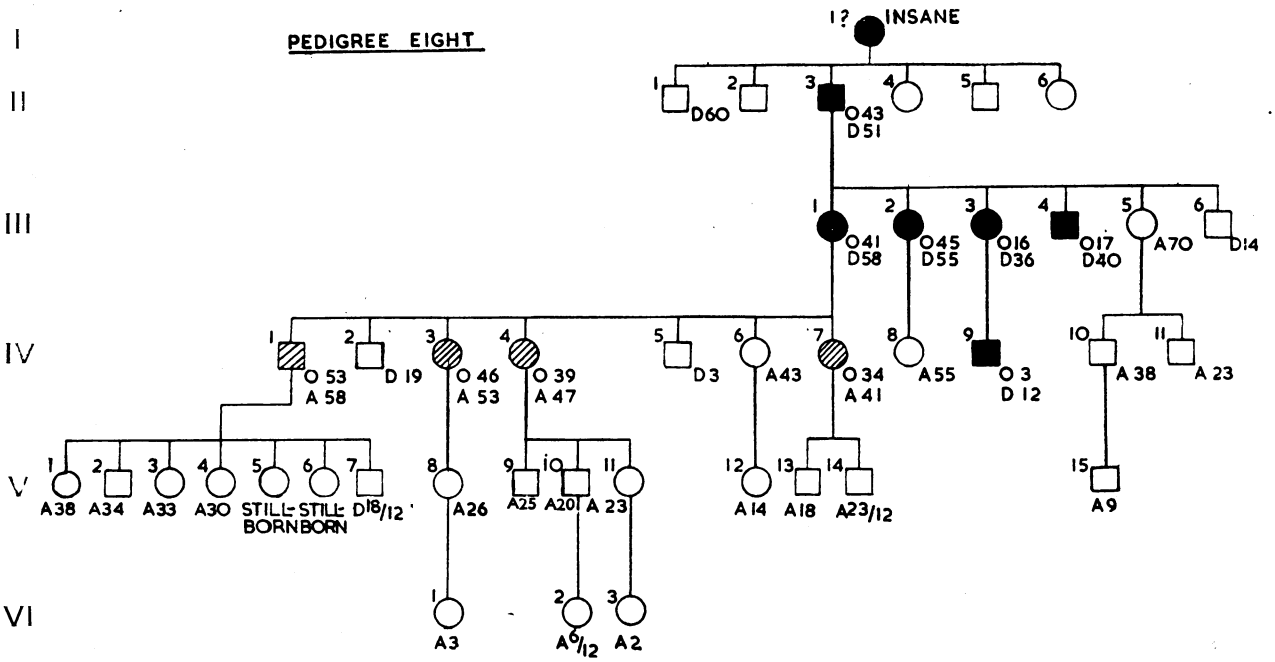

FIG. 8.—Pedigree Eight.

Incidence

On first consideration it might seem surprising that a fatal hereditary disease such as Huntington's chorea still persists in the community and that so deleterious a gene has survived the process of evolution. Haldane suggests that the age of onset in middle life may merely mean that primitive men and women seldom lived much beyond 40, so that postponement of onset beyond that age had no selective advantage. In addition, heightened sexual interest, possibly due to cortical degeneration, is not infrequently one of the early features of the disorder, and it may be that in this peculiar manner afflicted families have propagated the disease and so avoided extinction. Thus Reed and Palm (1951) found that in the pedigrees they investigated the average number of children from affected individuals was 6.07 ± 0.9 and from their unaffected sibs 3.33 ± 0.5, the difference between the two means of 2.74 ± 1.03 being statistically significant. In this country, Bell records an average number of 4.4 children where the mother is affected and 5.0 children where the father is affected.

The findings of the present survey denote an incidence of approximately 5 patients suffering from Huntington's chorea per 100,000 of the population. Bickford and Ellison (1953) obtained a higher figure amongst only those Cornish families who had a member in the mental hospital, so that the true incidence of the disease in that county is likely to be considerably greater. This, however, is probably related to the limited movements of the population in Cornwall on account of its isolated geographical position. In the London area, the 80 patients described by Minski and Guttmann in London County Council hospitals in 1938, if expressed as a proportion of the London County Council population at that time, give an incidence of 1.8 cases per 100,000; but here, too, reference was made only to those families which had a member in hospital. Critchley's survey in 1934 indicated a relatively high incidence of the disease in Suffolk, but again he was dealing only with certified patients, and the majority of choreics do not enter a mental hospital until the disease has reached an advanced stage. In this connexion Hughes (1925) found that in the state of Michigan there were, on an average, four domiciled choreics for each hospital case, but this observation may not be applicable here. Curran (1930) states that the incidence is higher in rural than in urban communities, but this does not hold good in Northamptonshire, where most of the families live in the industrial districts.

In a disease transmitted by a dominant gene the number of children who will inherit the disorder from an affected parent must vary greatly. It follows, therefore, that in a survey of this nature, unless the population sampled is very large, the number of persons found to be suffering from Huntington's chorea will show consequent variations. Nevertheless, it can be said that there is no evidence to indicate that the incidence of the disease is higher in this county than elsewhere; and if these figures are representative for the country as a whole then there are at present more than 2,000 people suffering from Huntington's chorea in England and Wales.

Care of the Patient

Treatment.—Goldman (1952) reported that oral procaine amide had given beneficial results in the control of choreiform movements. In this survey six patients with gross movements were treated with increasing doses of procaine amide over periods of more than six weeks, but no beneficial effect was noted which could be attributed to the use of the drug.

Supervision.—Domiciliary care obviously is preferable to institutional supervision, but unfortunately the antisocial behaviour of the patient frequently means that control in the home is inadequate. Mental instability, character changes, and sexual precocity not infrequently precede the chorea and are accompanied by a fall in the standard of living and the social class. It is probable that patients from the professional classes can be cared for longer in their homes, and Curran states that the majority of patients come from these classes. In this county, however, the families were found mainly in that section of the community classed by the Registrar-General as Group IV (namely, partly skilled occupations) where domiciliary supervision presents considerable difficulties.

Serological Table

Pedigree No.	Blood											Taste
Pedigree Two												
II 8*	O	MN	R_1r	PP	Le^{a-}	Le^{b+}	ss		$Lu^b\ Lu^b$	kk	Fy^{a+}	N
III 1	A_2	MN	R_2r	Pp	Le^{a-}	Le^{b+}	S		$Lu^b\ Lu^b$	kk	$Fy^{b+}Fy^{b+}$	N
III 2	B	MN	R_1r	Pp	$Le^{a+}Le^{a+}$		ss		$Lu^b\ Lu^b$	kk	Fy^{a+}	N
III 3	A_2	MN	R_1r	Pp	Le^{a-}	Le^{b-}	S		$Lu^b\ Lu^b$	kk	Fy^{a+}	N
III 4*	O	N	R_1R_2	Pp	Le^{a-}	Le^{b+}	S		$Lu^b\ Lu^b$	kk	Fy^{a+}	N
III 5	O	N	R_2r	Pp	Le^{a-}	Le^{b+}	ss		$Lu^b\ Lu^b$	kk	Fy^{a+}	N
III 6	A_1	MN	R_1r	Pp	$Le^{a+}Le^{a+}$	Le^{b-}	ss		$Lu^b\ Lu^b$	kk	Fy^{a+}	N
Pedigree Eight												
III 5	O	MN	R_1R_2	PP	Le^{a-}	Le^{b+}	ss		$Lu^b\ Lu^b$	kk	$Fy^{b+}Fy^{b+}$	N
IV 1*	A_1	MN	R_1R_2	pp	Le^{a-}	Le^{b+}	ss		$Lu^b\ Lu^b$	kk	$Fy^{b+}Fy^{b+}$	N
IV 3*	O	N	R_1R_1	Pp	Le^{a-}	Le^{b+}	ss		$Lu^b\ Lu^b$	kk	$Fy^{b+}Fy^{b+}$	N
IV 4*	A_1	N	R_1R_2	Pp	Le^{a-}	Le^{b+}	ss		$Lu^b\ Lu^b$	kk	Fy^{a+}	N
IV 6	A_1	N	R_1r	Pp	Le^{a-}	Le^{b+}	ss		$Lu^b\ Lu^b$	kk	$Fy^{b+}Fy^{b+}$	N
IV 7*	A_1	MN	R_1R_2	Pp	Le^{a-}	Le^{b+}	ss		$Lu^b\ Lu^b$	kk	$Fy^{b+}Fy^{b+}$	N
IV 10	O	MN	R_1r	Pp	Le^{a-}	Le^{b+}	S	Lu^{a+}		kk	Fy^{a+}	N
IV 11	O	MN	R_2r	Pp	Le^{a-}	Le^{b+}	S	Lu^{a+}		kk	Fy^{a+}	N
V 1	A_1	M	R_2r	Pp	$Le^{a+}Le^{a+}$	Le^{b-}	ss		$Lu^{b+}Lu^{b+}$	kk	$Fy^{b+}Fy^{b+}$	N
V 2	A_1	MN	R_1r	Pp	$Le^{a+}Le^{a+}$	Le^{b-}	ss		$Lu^b\ Lu^b$	kk	$Fy^{b+}Fy^{b+}$	No record
V 3	O	MN	R_2r	Pp	Le^{a-}	Le^{b+}	ss		$Lu^{b+}Lu^{b+}$	kk	$Fy^{b+}Fy^{b+}$	N
V 8	O	N	R_1R_1	Pp	Le^{a-}	Le^{b+}	ss	Lu^{a+}		kk	Fy^{a+}	N
V 10	A_1	N	R_1R_2	Pp	Le^{a-}	Le^{b+}	ss		$Lu^{b+}Lu^{b+}$	kk	Fy^{a+}	N
V 11	A_1	MN	R_1R_2	Pp	Le^{a-}	Le^{b+}	ss		$Lu^{b+}Lu^{b+}$	kk	Fy^{a+}	N
V 12	A_2	N	R_1r	PP	Le^{a-}	Le^{b+}	ss		$Lu^{b+}Lu^{b+}$	kk	$Fy^{b+}Fy^{b+}$	NT
V 13	O	N	R_2r	PP	Le^{a-}	Le^{b+}	ss		$Lu^{b+}Lu^{b+}$	kk	Fy^{a+}	N
V 14	A_1	M	R_1r	Pp	Le^{a-}	Le^{b+}	ss		$Lu^{b+}Lu^{b+}$	kk	$Fy^{b+}Fy^{b+}$	No record
V 15	O	N	R_1R_1	PP	Le^{a-}	Le^{b+}	S	Lu^{a+}		kk	Fy^{a+}	N
VI 1	O	N	R_1R_1	Pp	Le^{a-}	Le^{b+}	S	Lu^{a+}		kk	Fy^{a+}	N

* Suffers from Huntington's chorea. N = Normal perception of taste. NT = Non-taster.

Prevention of the Disease

(a) Prediction

Serological and Hereditary Factors.—In this present survey samples of blood were taken from members of two of the families (Pedigrees Two and Eight) and subgrouped at the Radcliffe Infirmary, Oxford, as it was hoped that the results obtained might have some bearing on genetic linkage. These persons were tested also for taste factor (phenylthiocarbamide—50 parts per million). In the event, no significant data were obtained, but the results are placed on record (see Serological Table).

Electroencephalography.—Attempts at accurate prediction by means of electroencephalography have been made, but with conflicting results, and the original claims of Patterson *et al.* (1948) have not been substantiated by Harvald (1951) or Leese *et al.* (1952). In this inquiry electroencephalographic records were obtained in six persons, four of whom were suffering from the disease in an advanced stage. These records, which were made by the electroencephalographic department of the Radcliffe Infirmary, Oxford, did not reveal any definite abnormalities which might be of value in the diagnosis of the disease, so it was felt that no useful purpose would be served by obtaining tracings from other patients.

(b) Health Education

There can be few diseases more distressing than Huntington's chorea either to the patient or to the relatives. The former, after suffering from increasingly uncontrollable movements for many years, inevitably die in a demented state. Often they have to be nursed in cot beds in order to prevent them from injuring themselves. The relatives spend their lives in fear and apprehension lest they and their children fall victim to the disease.

The real gravity of the chorea, however, lies in its hereditary nature. The child of an affected parent stands a 50% chance of contracting the disease; if he marries without waiting to see if he has the disorder, then he can assume that his children stand a 25% chance of inheriting it. The hereditary pattern is so definite that every effort should be made to prevent its spread, and eugenic advice should be given to members of afflicted families. The experience of this survey indicates that such advice is welcomed. Although many members of a family may be afflicted, the true hereditary character of the disease is not understood by them, and their lack of knowledge makes them the more fearful. Rational explanation can do much to relieve them of the wretchedness of unnecessary anticipation.

Such health education, to be effective, should be given to all members of the family, but as a rule only a few members are under the care of any one practitioner. It may well be that a doctor does not know that his patient comes from a choreic family. As an illustration of this, III 4 of Pedigree Two was advised not to have any more children, but her brother and his consort were receiving treatment for infertility.

The local health authority is in a fortunate position in being able to work in close liaison with the general practitioners and the mental hospital. Moreover, under Section 51 of the National Health Service Act they are responsible for the prevention and aftercare of mental illness. In undertaking home visiting the medical officer can obtain details of family trees which may assist both the practitioner and

the hospital clinician regarding the diagnosis, and at the same time he can give the necessary advice to prevent the spread of the disease.

Mutation

It might be argued with justification that if the mutation rate of a hereditary disease is high there is little object in trying to prevent its spread. In Huntington's chorea, however, it is clear that the great majority of cases are inherited, and the proportion of persons who owe their disease to a mutation in one or other parent is very small indeed. Hence, if it were possible to prevent the transmission of the gene the incidence of the disease would be very greatly reduced. Although one possible instance of mutation in an earlier generation is demonstrated in the present series of eight pedigrees, Bell quotes only 6 out of 151 pedigrees.

These observations indicate that the following measures would be of value: (1) Practitioners should be encouraged to notify all cases of Huntington's chorea to the local health authority so that the family pedigrees can be investigated. (2) (i) Offspring of an affected parent who are of Roman Catholic faith should be advised against marriage. (ii) Offspring of an affected parent who are of Protestant faith should be advised not to have children, and should be encouraged to attend a birth-control clinic. The help of all churches would be of great value in undertaking these measures. It is noteworthy that the Minister of Health has sanctioned contraceptive advice at birth-control clinics " only in cases where further pregnancy would be detrimental to health." It would, however, appear only logical that the health of the offspring should be protected in the same way as the health of the parent. (3) Termination of pregnancy should be considered if either parent is suffering from the disease or may be carrying it. (4) Friendly supervision of members of choreic families should be maintained by whoever is in the best position to do so effectively. In some cases this could be the responsibility of the practitioner, in others of the district nurse, the health visitor, or the mental health worker. (5) As soon as domiciliary care is found to be inadequate, the patient should be admitted to an institution or mental hospital.

Summary

Although Huntington's chorea is described as a rare disease, no attempt has been made to assess the incidence in this country. In the present survey a questionary was sent to the senior partner of every practice in Northamptonshire, and with the consent of the medical superintendent of the county mental hospital the records of certified patients were made available. The results indicated that the incidence of the disease in Northamptonshire is approximately 5 diagnosed cases per 100,000 of the population. No conclusions can be drawn from such small figures in a population of about 263,000, but if this incidence is representative for the country, then there are at present more than 2,000 people suffering from Huntington's chorea in England and Wales.

It is noted that choreic families appear to be drawn mainly from Group IV of the Registrar-General's social classification.

In this survey six patients were treated with procaine amide, but no beneficial results were observed which could be attributed to the drug.

Electroencephalographic and serological investigations were carried out on members of two choreic families. The results were not found to be of any significance in prediction or diagnosis, but the serological findings may be of value in future work on this subject.

Adequate supervision of choreic individuals is very desirable on account of the heightened sexual interest and fecundity associated with cortical degeneration in the disease. As soon as domiciliary control is insufficient patients should be admitted to hospital.

Since there is no treatment for this hereditary disease, attention is drawn to the urgent need for its prevention through the health education of afflicted families. If medical practitioners reported the disease voluntarily, the local health authority could help by investigating family pedigrees and giving the necessary advice. This work would be both of preventive and of diagnostic value. It is felt that prevention is by far the most important aspect of Huntington's chorea, and that every effort which is possible in a voluntary capacity should be made in an attempt to reduce the incidence of the disease.

I should like to express my thanks to Dr. C. M. Smith, County Medical Officer of Health, Northamptonshire, for his help and suggestions in undertaking the survey; to Dr. J. A. Fraser Roberts for his advice in the preparation of this paper; to Dr. A. N. Graham for granting me facilities to examine patients at St. Crispin Hospital; and to Dr. J. C. Goodchild for undertaking the treatment of patients with procaine amide. I should also like to thank Dr. M. M. Pickles for undertaking the serological investigations; and, finally, to express my appreciation of the co-operation received from the medical practitioners in the county, without whose help this survey would not have been possible.

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PARTIAL GASTRECTOMY: TEN YEARS LATER

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The operation of partial gastrectomy has received perhaps more study and investigation than almost any other operation in surgery. The value of the procedure in the treatment of peptic ulcer is now established, and the immediate results are well known. The operative mortality rate in good hands has been established at 1–2%. The post-gastrectomy syndromes have been studied in detail by Adlersberg and Hammerschlag (1947), Irvine (1948), Mimpriss and Birt (1948), Muir (1949), Wells and Welbourn (1951), and many others. Very little is known, however, about the late results of the operation and the prospects for the patient ten or more years after surgery.

Of 119 cases of partial gastrectomy performed for the treatment of peptic ulcer before the early months of 1944, 28 were traced to 1946 only, a further 16 maintained contact by letter to 1950, and the remaining 75 have been examined at frequent intervals to the present day (1954). It is the history of these 75 patients which