AN INVESTIGATION OF HUNTINGTON'S CHOREA IN VICTORIA

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IN making an investigation of Huntington's Chorea in the State of Victoria, we have, so far, been able to collect 150 indisputable cases of this disease, and others. This figure includes 83 deceased, and 57 living. Out of a total of 74, who were certified to mental hospitals, 26 are still either in hospital or on trial leave. This represents ·34 per cent. of the total present mental hospital population. As regards sex distribution, our figures indicate that there were 68 females as compared with 72 males.

As far as can be ascertained, the first recorded case of Huntington's Chorea occurring in Victoria was at the Yarra Bend Mental Hospital in 1878. Although diagnosed then as Choreic Dementia, the clinical description of the patient and the subsequent family history leave no doubt that the case was one of Huntington's Chorea. Victoria, which was first settled in 1834, must be regarded as a new country and one which has been subject to the impact of a continual migration from other parts of the world, particularly Great Britain. There are three recognized peaks in this migratory flow; during the Gold Rush of the 1850–60s, and after each of the two World Wars. The changing population has created many difficulties in our investigations, and consequently in quite a number of instances it has been impossible to trace either the predecessors, or all of the descendants of known cases of Huntington's Chorea. However, it seems clear that there have been multiple sources responsible for the introduction of this family disease into Victoria, from older countries as well as from other Australian States. Restlessness and a wandering disposition, so frequently found in numbers of Huntington's Chorea families, have no doubt contributed appreciably to the relatively large number of the descendants of affected

TABLE I

Country of Origin	Number of Families	Number of Cases in Victoria
England:		
Cumberland	1	18
Northumberland	1	8
Cornwall	3	14
London	2	9
Scotland	3	8
Ireland	2	18
Germany	1	3
Tasmania (*ex* England)	1	9
Total	14	87

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families migrating to any country, and, on arrival, moving about from place to place. In those instances where it could be definitely established, the country of origin of various Victorian families, and the number of cases found among the descendants of these families, is given in Table I.

TABLE II

Isolated Huntington's Chorea Cases whose Families are Non-Resident in Victoria

Country of Origin	No. of Cases
Italy	1
Canada	1
New Zealand	1
Australia:	
New South Wales	5
South Australia	3
West Australia	3
Queensland	1
Tasmania	7*
Total	22

There were 39 cases of Huntington's Chorea in which the country of origin was unknown, and of these there were five cases who were adopted or illegitimate.

AGE AT ONSET OF FIRST SYMPTOMS

In the 73 cases where the age of onset was determined with a degree of accuracy, the average age was 38·3. The following table shows the age of distribution.

TABLE III

Age Range—Years	No.
0– 9	0
10–19	3
20–29	13
30–39	28
40–49	21
50–59	6
60–69	2
Total	73

The average age of onset was given by Davenport (1916) as 37·8; Julia Bell (1934) 35·5; Minski and Guttman (1938) 42·4.

EARLY SYMPTOMS

In the 65 cases in which detailed study was possible, 33 presented neurological symptoms as the mode of onset; in 16 cases mental preceded neurological symptoms, and in the remaining 16 cases both neurological and mental symptoms appeared within such a short interval of each other that it was impossible to state accurately from the history available which did appear first. The most commonly found early involuntary movements in order of frequency were:

* Six of the seven isolated cases from Tasmania are descended from one family which has 105 known cases of Huntington's Chorea. The original member of this family migrated from Somerset to Tasmania with her family of twelve in 1853 (Brothers (1950)).

Fidgetiness
Motor restlessness
Swaying and staggering gait
Jerkiness of the head
Shrugging of the shoulders
Clumsiness of hand movements
Twitching of facial muscles

In several instances, early involuntary movements appear to be confined to a relatively small group of muscles for several years before the occurrence of gradual involvement of other muscular groups. In the majority of cases, however, once any involuntary movement had become manifest, gradual extension of choreiform movement to other skeletal musculature was the rule, resulting eventually in the almost incessant severe generalized choreiform movements characteristic of this disorder.

The most frequently found psychological disturbances were irritability and quick temperedness. This was the case in nearly 70 per cent. of cases where the appearance of the mental symptoms preceded the chorea. Other early abnormalities which were responsible for bringing the patient under notice were as follows:

Moodiness or depression
Abnormal excitability
Emotional apathy
Paranoid trends
Outbursts of violence
Irresponsibility
Defective concentration and memory
Loss of moral control resulting in erotic behaviour, delinquency, or alcoholism
Aimless wandering and vagrancy, and, in one particular instance, compulsive behaviour

Mental States Associated with Huntington's Chorea

Although the mental picture of Huntington's Chorea is that of progressive organic dementia, it is often preceded by emotional disturbances and changes of personality, amounting, at times, to progressive psychotic states requiring certification.

Depression

In our series there were seven cases whose admission to hospital was due to severe depressive states. Four showed depression prior to the onset of chorea, and three developed, subsequent to chorea. Suicide had been attempted in one of the first group and two of the second group. Two cases showed remission of the depression following electropexy. Of the four cases where mental symptoms preceded the physical, three developed chorea one year later, and the other, eight years later.

Mania

Three cases were admitted to hospital on account of acute maniacal outbursts. One had had several attacks of mania during the twenty years preceding the onset of chorea. In each of the other two cases, chorea preceded the initial

attack of mania. In all three, the attacks were definitely episodic and occasionally alternated with periods of mild depression.

Paranoid Reaction

Of the seven cases with marked paranoidal features, five developed psychotic symptoms prior to chorea, and two subsequently. In the former the duration between onset of mental and physical symptoms varied between one and eight years. The progress of the initial dementia appeared to be somewhat slower in these cases than in the average cases of Huntington's Chorea. In no instance was there any amelioration of symptoms at any time.

Schizoid Reaction

Four cases showed very typical schizophrenic-like psychosis with the usual symptoms of withdrawal, emotional displacement and auditory hallucinations. Three occurred prior to the appearance of chorea. Of these, one developed early signs of chorea within one year, and two showed evidence of chorea three years after the onset of mental symptoms. In the fourth case, as both neurological and psychotic symptoms were present on admission and the early history somewhat obscure, it was not possible to venture any opinion as to which appeared first.

Epilepsy

Only two cases were admitted on account of mental symptoms associated with epilepsy, and both had been subject to seizures since late adolescence several years prior to the onset of chorea.

Mental Confusion

In the cases of two brothers (Family F) the mental picture on admission was that of mental confusion, with marked clouding of consciousness. In these two cases choreiform movements had not been apparent prior to the onset of confusion, but were certainly present in a mild form shortly after admission. Both cases progressed to increasing dementia without any amelioration of symptoms.

Mental Deficiency

Apart from three instances of Huntington's Chorea occurring in cases of low grade feeble-minded members of known Huntington's Chorea families, two other familes have been discovered in which chorea was associated with mental deficiency. In each instance, there was no evidence of Huntington's Chorea having occurred in previous generations. Consequently, until further investigations can be carried out, it is premature at the moment to regard them as cases of Huntington's Chorea, although when seen as adults they are apparently indistinguishable from typical cases of this disease. In one family (Plate 1), the parents, who were of Cornish stock, were first cousins. There

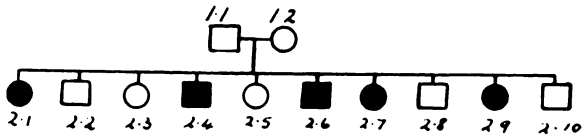


PLATE 1

were ten siblings in the generation investigated. Five of these were affected with chorea, and the other five were reported as being mentally and physically normal. Each of the five affected members was a mental defective of imbecile level and had had choreiform movements since infancy. The first affected member of this family at the age of eighteen was described by the Commonwealth Medical Referee as having been mentally defective since birth, and as suffering from chronic chorea. It was particularly stressed at the time that there was marked unsteadiness of the head and limbs. Thirty years later, at the age of forty-eight, this patient was admitted to Mont Park Mental Hospital physically helpless as a result of gross generalized choreiform movements. The fourth member of this family, an imbecile of low grade, is reported as having had muscular twitchings since infancy. At the age of twenty-five he was admitted to Sunbury Mental Hospital, grossly choreic. There was marked irritability and inability to sit still, facial twitchings, marked dysarthria, and choreiform movements. The sixth member of the family was admitted to Sunbury Mental Hospital at the age of twenty-four, when he was diagnosed as a high grade imbecile with chorea which had been present since the age of three. There was unsteadiness in gait and constant choreiform movements involving the whole voluntary musculature. Death occurred seven years later at the age of thirty. Information regarding post-mortem findings in this case is not available. Unfortunately, in the past, there does not appear to have been any systematic attempt to study in detail any of the post-mortem specimens of brains from choreic subjects. The seventh member has been reported by a social worker as being a mental defective with chorea. This patient is still living at home and no further details are available. The ninth member was admitted to Sunbury Mental Hospital at the age of sixteen, when he was described as an imbecilic youth with continuous choreiform movements of muscles, head, face, limbs and trunk. Information given by relatives suggested that the choreiform movements had been noticeable since at least the age of three.

In the second family (Plate 2), the parents, both of whom originally came from Devon, England, were second cousins. Neither the father, who died at the

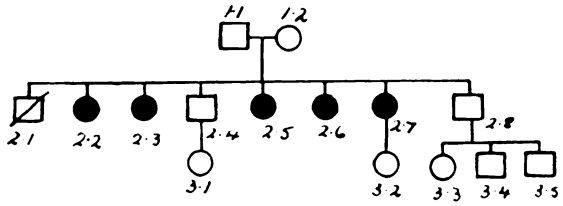


PLATE 2

age of eighty-two, nor the mother, who is still alive, aged eighty, apparently showed any signs of chorea. There were eight siblings in the family. The first died as a result of meningitis in infancy. Cases 2.2 and 2.3, who were identical twins, were mental defectives (feeble minded). The history relates that they had had an indistinct and slurred speech from childhood. In their early adult lives they were able to perform domestic duties of a routine nature, but at approximately the age of forty they began to show mental deterioration, and being unable to care for themselves were admitted to the Ballarat Mental Hospital. It was about this time that choreiform movements were first definitely noticed. At the time of death, several years later, both were completely bedridden on account of gross chorea. Case 2.4 is reported as being normal. Case 2.5, who

was an epileptic mental defective, died in the Kew Mental Hospital at the age of sixteen. Case 2.6, also an epileptic defective, died in the same hospital at the age of eighteen. Case 2.7 was a congenital mental defective (feeble minded) who was admitted to Royal Park at the age of forty-three, at which time she was showing evidence of being mildly choreic. Case 2.8 is reported as being of good intelligence and shows no signs of chorea.

Diagnosis and Prediction

In the absence of details of family history, mistaken diagnoses are most commonly found in early cases where the mental symptoms have appeared prior to the onset of choreiform movements, or, where the latter are so mild as to have been overlooked at the time of examination. A number of such cases in our series have been originally diagnosed as paranoid psychoses, schizophrenia, psychopathic states, neuroses, depressive states, dementia, and mental deficiency. Even in more advanced cases where chorea is manifest but family history obscure, occasional misdiagnoses are made, the most common instances being when they have been mistaken for post-encephalitis, general paralysis of the insane, disseminated sclerosis, senile and Sydenham's chorea.

On the other hand, in a few instances, certain cases originally diagnosed as Huntington's Chorea have subsequently proved to be cases of either hysteria, adult mental deficiency with chorea, or reactive depression. The depressive states may occur as symptoms of Huntington's Chorea, but, at the same time, it is realized that there is a marked tendency for reactive fears of family ill health and disease to occur in unaffected members of such families.

In well established cases where the choreiform movements are marked, and the family history is known, the diagnosis presents no difficulty. However, there still remains the problem of ascertaining which of the as yet unaffected members of a "Huntington's family" are likely to succumb to the disease. Since Huntington's Chorea has been shown to be a Mendelian dominant, it is clear that the probability is one in two that any descendant will be affected. It is obvious that years of apprehension and anxiety would be obviated for non-affected members if means could be found of predicting which individuals were not likely to become victims.

The use of EEG and psychological tests suggested themselves as possible methods of obtaining criteria for prediction.

Regarding the EEG, we are most grateful for a personal communication from Dr. J. Daniel Palm, of the Dight Institute for Human Genetics, University of Minnesota, in which he states:

> "The information we have of test results of affected persons is indeed scanty and in need of further verification. An antithesis apparently exists in the EEG tests. In the very early stages of the disorder the persons show an abnormal EEG of the low voltage fast type which is also present in half of the adult offspring of affected persons. We believe that destruction of the connection of the caudate to the thalamus by the ravages of the disease may interrupt the feed back system to area IV and thus give a normal EEG. This hypothesis is certainly open to criticism, but is all that we have now.
>
> "We have administered various psychological tests to the offspring of choreics but only one test seems to show promise. The Rorschach Ink Blot tests show a significant number of abnormal personality traits present in the group tested but are not correlated with the EEG."

In this department it was thought that, in addition to assisting diagnosis and indicating rate of deterioration, the comparison between the psychological tests of affected and non-affected members of Huntington's descendants might yield some clue as to which unaffected members were likely to be affected.

With early affected cases it was possible to apply the Wechsler (1939),

Rorschach (1937), Goldstein Colour Form (1941), Cattell 16 P.F. (1946), Cornell Index (1945), and standardized tests of educational attainment and writing. With advanced cases of the disease it was sometimes only possible to complete one or two of the sub-tests of the Wechsler, generally repeating digits, arithmetic, and information, and to obtain a word recognition index (Burt's Reading Scale) (1922).

An examination of the Wechsler results from thirteen of the earliest cases of the disease indicated that at first the Huntington's Chorea patient retains his ability to recount general information, and he understands and makes use of his everyday vocabulary. On the other hand his ability to manipulate numbers and his immediate rote memory show considerable deterioration, with the earliest onset of physical or mental symptoms. On the whole the patient does very much worse on performance tests. The greatest reduction is shown in (1) the ability to analyse and synthetize perceptions (Block Design test), and (2) learning association flexibility and motor co-ordination (Digit Symbol test).

Use of the Wechsler formula indicated with this group that deterioration ranged from 14 per cent. to 80 per cent., mean 43 per cent. (S.D. 19 per cent.). There was also a small positive correlation between the amount of deterioration and the duration of the disease.

In two cases of the disease it was possible to retest the patients with a two years' interval. The results suggest that there is an irregular decline of one to two points of weighted score per year in the "Don't Hold" Wechsler tests at the early stages of the disease.

The Rorschach test, although applied to only a few of the earliest affected cases, invariably gave a record characteristic of organic lesion, namely poor output, percentage of "F" higher than normal, absence of "FK", slow reaction times, little or no "M", colour naming, impotence, perplexity, and use of automatic phrasing.

The Inventories, Cattell 16 P.F. and Cornell N.2, revealed in detail personality dysfunction of the affected compared with the unaffected, and supported the general picture given by the psychiatric examination.

The results of educational tests indicated a definite falling off in the affected in the ability to make arithmetical calculation. In the same group there was also a decline in word recognition, which was equal on the average to the difference between an Eighth Grade, compared with a Sixth Grade educational level.

In all cases where samples of handwriting were taken of the affected group it was obvious that the choreiform movements interfered with the writing to a greater or lesser degree, producing in many cases a characteristic "saw tooth" effect.

The testing of a small number of children from Huntington's families yielded insufficient data on which to base any positive hypothesis. However, the test results of an unaffected descendant, aged 37, were remarkably similar to those of undoubted cases, for example, three of the Wechsler tests associated with deterioration were significantly lower than expected. This person showed physical symptoms of the disease two years later.

PROGNOSIS

Once established, the chorea progressively becomes worse. There are no remissions, although in certain families its progress is much slower than in others. In the terminal stages there is usually gross emaciation and weakness,

and unless some inter-current illness intervenes they become chronically bed-ridden and eventually die as a result of exhaustion. Although in the large majority progressive organic dementia is usual, nine cases in our series showed no apparent personality change throughout the course of their illness. One (H. 2.1) who developed chorea at the age of sixty is still at the age of seventy-five very well preserved mentally, in marked contrast to his son who at the age of forty-four, five years after the onset of illness, is already showing marked mental deterioration. Six cases showed remission of the acute early mental symptoms necessitating their original admission to hospital, but in each case there was probably some slight residual deterioration—one was able to remain at home for nearly twenty years, and another twelve years, before being re-admitted in an advanced state of dementia.

Based on thirty-six cases in which the ages of onset and of death were both ascertained, the average duration of the disease was thirteen years. The shortest duration was one instance in which death was recorded after only two years of symptoms; the longest fifty years. The duration of the disease did not appear to be influenced by the age of onset. The average age at death which was established in fifty-seven cases was 51·3 years. Table IV shows the age distribution of death.

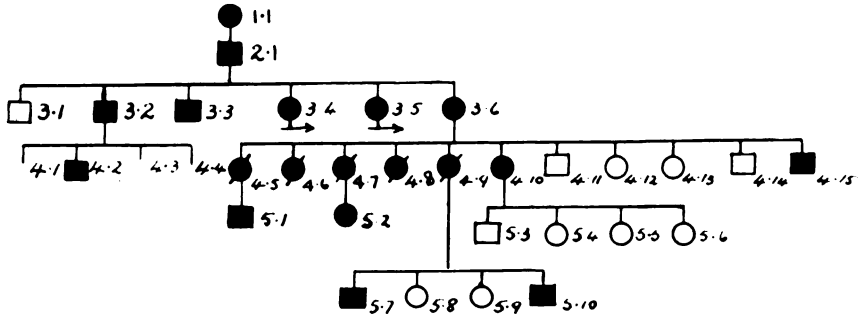
TABLE IV

Age Range—Years	No.
0–10	—
11–20	—
21–30	1
31–40	6
41–50	18
51–60	20
61–70	7
71–80	5
Total	57

EXAMPLES OF FAMILY HISTORIES

Family "A"

The first recorded case of Huntington's Chorea occurring in the State of Victoria was a male patient (Plate 1) (Case 2.1) who, at the age of 41, was admitted in the year 1878 and died 12 years later at the Ararat Mental Hospital. He was stated to have migrated from the County of Cumberland, England, where his mother was supposed to have died from a similar illness some years previously. As far as is known, there were no siblings. In the following generation, there were six siblings and interestingly enough all but one were apparently afflicted with Huntington's Chorea. There is no record, however, of any of them having been admitted to a mental hospital. There are no known descendants of Cases 3.1 and 3.3. As regards Cases 3.4 and 3.5 there were descendants, but no information concerning them has been obtained. Case 3.2 had four children, two of whom were diagnosed at the Sunbury Mental Hospital as cases of Huntington's Chorea, one of whom (Case 4.2) was originally an epileptic mental

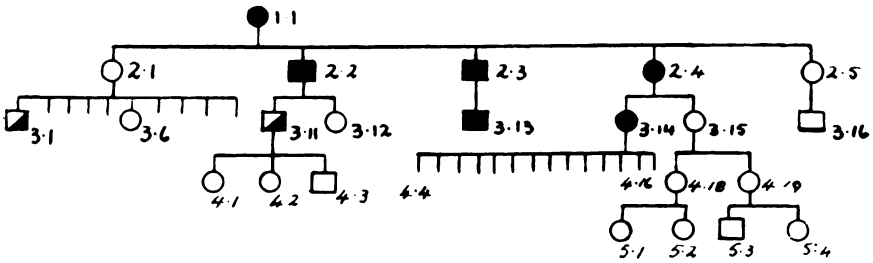


defective. By the time he had reached the age of 17, he was grossly choreic. Case 3.6 had eleven children, the six eldest (4.5 to 4.10) and the youngest (4.15) all of whom eventually developed Huntington's Chorea. The age of onset of these seven cases was uniformly about thirty-five and the age of death approximately forty-five, the deaths being manifest by chorea followed by simple dementia. Cases 4.5, 4.6 and 4.7 died in benevolent homes. Cases 4.8, 4.9 died in mental hospitals, while Case 4.10 is still alive in a mental hospital.

In the present generation, Cases 5.1, 5.2, 5.7, 5.10, have already been admitted to mental hospitals with the diagnoses of Huntington's Chorea. In three of these cases the early mental symptoms were somewhat schizoid in nature, and in each case there was marked violent behaviour immediately prior to admission to hospital. Cases 5.8, and 5.9, as yet show no apparent mental change or choreiform movements. The age of onset in this generation was 31, 23, 21 and 18 respectively. Cases 5.3, 5.4, 5.5 and 5.6 whose present ages range from seven to twenty-five years, are as yet still unaffected.

Family "B"

Case 1.1 arrived in Victoria from Northumberland some time during the middle of the second half of the nineteenth century. She was the only member of her family to migrate to Australia. Although showing definite evidence of chorea before she died, there were apparently no gross mental changes. Her eldest daughter (Case 2.1) together with her family of ten children, lived in New South Wales. She died at the age of seventy, somewhat mentally enfeebled, but apparently not choreic. This is not conclusive as no really reliable information could be obtained. Her eldest son (Case 3.1) died in a New South Wales mental hospital before the age of forty. No data is available about the other siblings, except in Case 3.6, who is known to be free from chorea at the present time. Case 2.2 died of Huntington's Chorea in a benevolent home. There were two children, the elder of whom was showing early signs of

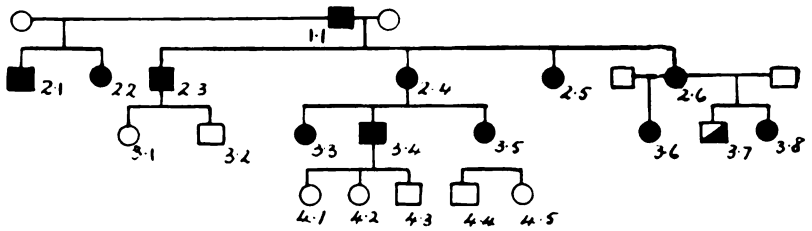


chorea four years ago. Case 2.3 died as the result of Huntington's Chorea in a benevolent home. His only son (Case 3.13) was admitted to Mont Park Mental Hospital at the age of forty-two in an advanced stage of Huntington's Chorea and died five years later. Case 2.4, who died at the age of fifty from Huntington's Chorea, showed no gross mental abnormality. Her eldest daughter (Case 3.14) at the age of thirty-nine developed marked irritability and became subject to outbursts of uncontrollable temper. Shortly afterwards, she developed choreiform movements of arm, head and face. A year later she was admitted to Mont Park Mental Hospital, where she rapidly deteriorated mentally and physically and died three years later at the age of forty-three. There were fourteen children, whose present whereabouts are unknown. Case 3.15, whose present age is fifty-two, is very tense and over-anxious but appears to be free from any suggestion of choreiform movements, although two years ago was regarded as an early case of Huntington's Chorea. Her two children, 4.18 and 4.19, are reported to be neurotic. The youngest sibling of the second generation (Case 2.5) died as a result of cerebral haemorrhage.

Family "C"

Little is known concerning the original affected member of this family beyond the fact that he was described as being nervy and probably of Irish extraction. It is presumed that he had chorea, and this is supported by the fact that all of his six immediate descendants (two by his first marriage and four by his second) have proved to be cases of Huntington's Chorea. In the first family (Case 2.1) an invalid pensioner, has marked choreiform movements. Case 2.2 is alleged to be very nervy and mildly choreic. Case 2.3 (second family) was admitted to the Kew Mental Hospital at the age of fifty-one, when he was markedly mentally enfeebled and practically helpless on account of advanced chorea. He died several months later. Case 2.4, according to her husband, began to exhibit mental changes at the age of twenty-eight, when she was hallucinated and developed delusions of poisoning. Later she began to show fidgety movements. On account of suicidal threats and violence towards her husband she was admitted to a mental hospital four years later. She remained in hospital for seventeen years until she died at the age of forty-nine. Case 2.4 was admitted to Kew Mental Hospital in her late twenties as a case of Huntington's Chorea. Case 2.6 is reported as having had a nervous breakdown at the age of eighteen, and apparently recovered. She attempted suicide twelve years

later, after which she began to show signs of irritability, mental deterioration, and signs of chorea in the nature of facial twitchings, dysarthria, and unsteadiness of gait. On admission to hospital at the age of thirty-five she showed mild right hemiparesis. Death occurred at the age of forty-five. In the succeeding generation, nothing is known concerning the medical history of the children, 3.1 and 3.2. Case 2.4 had three children, still alive, but all affected by Huntington's Chorea (3.3 to 3.5). The eldest, 3.3, was admitted to Sunbury Mental Hospital at the age of thirty-nine, but the onset of the condition had been noticed approximately three years previously after an attempted suicide, shortly after which she began to develop choreo-athetoid movements. She became extremely irritable, abusive, destructive, and had attacks of uncontrollable temper. At the time of admission she had developed visceral delusions. She is

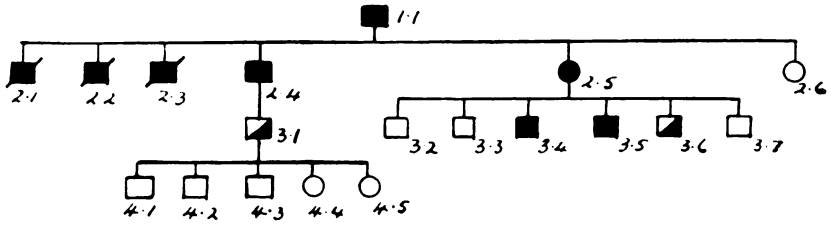


now showing evidence of progressive mental enfeeblement. The second child (Case 3.4), is at present attending a general hospital on account of staggering gait and severe headaches since the age of thirty-six. The third child (Case 3.5) is described by her parents as being very irritable, and recently having developed a peculiar staggering gait at the age of thirty-six. Case 3.6 is the only issue of the first marriage of Case 2.6. She was admitted to the Ararat Mental Hospital at the age of twenty-four presenting schizoid features and early signs of chorea. Nine years later, just prior to death, she was extremely demented and physically helpless on account of advanced chorea. Case 3.7 (issue of second marriage of Case 2.6) is reported as being an invalid pensioner, who "sways about all over the place". Case 3.8 was admitted to the Ballarat Mental Hospital at the age of twenty-six when the clinic records reveal her as being unco-operative, refractory, regressive, delusional and hallucinated. She was originally regarded as a case of schizophrenia, but several months later it was noted that she had developed mild twitchings of the shoulders and fidgetiness of the hands. One year later the florid mental symptoms had somewhat abated, but there was some mental enfeeblement. The choreiform movements, on the other hand, had become much more pronounced.

It is interesting to note that, apart from the members of the last generation, whose ages are still below twenty, and Cases 3.1 and 3.2 about whom no information is available, Huntington's Chorea has been diagnosed in every member of this family.

Family "D"

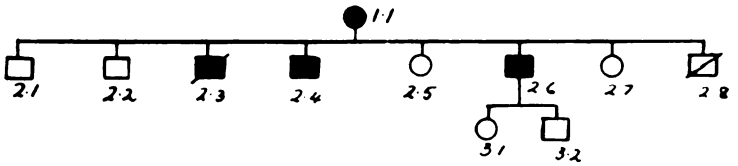
The earliest member of this family known to us to have suffered from Huntington's Chorea was an English migrant whose family formerly resided in the County of Cornwall. Clinically, little is known of this particular case beyond the fact that he died in early middle age from Huntington's Chorea. Of his six children, all but the youngest, who is still alive, developed signs of chorea at about the age of forty. A son, 3.1, is said to have been nervous and committed suicide. Case 2.5 lived for over twenty years after the onset of the symptoms. Apparently



none of them showed any gross abnormality. In the next generation, information is available in the case of the descendants of Case 2.5. Two of her six sons (Cases 3.4 and 3.5) are at present suffering from advanced chorea, and another son (Case 3.6) is now showing early mild symptoms of this condition. Case 3.4 developed choreiform movements four years prior to his admission to Royal Park on account of repeated maniacal outbursts. On examination, there was marked dysarthria, twitchings of the face, and gross purposeless movement of the neck and arms. Mentally, he is euphoric, distractible, and exhibiting flight ideation, lack of attention, and memory defect. Whilst knowing all about the hereditary nature of his complaint and his own family history, he nevertheless appears to be quite lacking in any insight into the severity and seriousness of his own condition. Case 3.5, although markedly choreic, is still able to carry on his usual occupation as a wharf labourer.

Family "E"

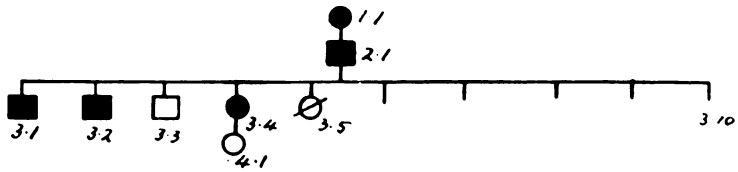
The Case 2.6 in family "E" was formerly a resident of New South Wales where his mother (Case 1.1) and two brothers (Case 2.3 and Case 2.4) were known to be definite cases of chorea.



He himself was admitted to the Kew Mental Hospital at the age of forty-one, grossly choreic and mentally enfeebled. The outstanding early mental symptoms described in this case were uncontrollable fits of temper and aimless wandering. Death occurred twelve years later at the age of fifty-three. He was the father of two children whose present whereabouts and clinical histories are unknown.

Family "F"

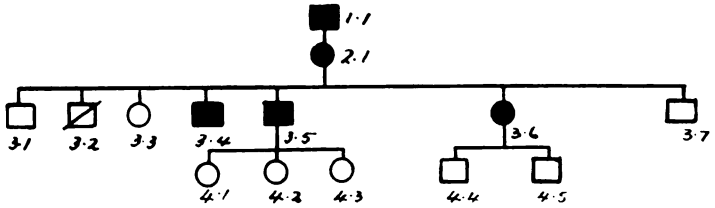
Case 1.1 died in an English Mental Hospital apparently from Huntington's Chorea. The son (Case 2.1), who migrated to Victoria, was admitted to the Sunbury Mental Hospital at the age of sixty-six and died three years later grossly demented and choreic. Information regarding duration of illness, early signs and symptoms, is not available. In the succeeding generation there were ten siblings. Of these, three were afflicted with Huntington's Chorea and one died



from rheumatic fever at the age of eighteen. The others are reported as being in good health. Case 3.1 was admitted to Ararat Mental Hospital in an advanced stage of Huntington's Chorea when aged forty-seven, and he died at the age of fifty-four. Case 3.2 was admitted to the same hospital at forty-three and died four years later. Case 3.4 was admitted to Mont Park at forty-one, five years ago. All these cases were grossly demented on admission. The onset in each case was that of simple mental deterioration associated with mental confusion and early choreiform movements.

Family "G"

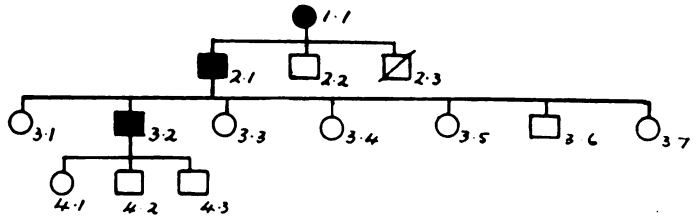
Apart from the fact that Case 1.1 died as a result of Huntington's Chorea, no further information is available regarding him or his antecedents. Case 2.1 died at the Royal Park Mental Hospital. The family consisted of seven siblings, of whom Case 3.1 is living in Canada, Case 3.2 was killed whilst a young man under the age of thirty, Case 3.3 and Case 3.7 are reported as being normal. Case 3.4 originally diagnosed as a case of multiple sclerosis, eventually died at the age of sixty-six from Huntington's Chorea. The fifth member, Case 3.5, was



admitted to the Bundoora Repatriation Mental Hospital, at forty-three, eight years ago. The onset of his condition was characterized by marked irritability and paranoid features which preceded the appearance of clumsiness and other features of chorea. Of his three children, whose present ages range between four and seventeen years, the only point of interest is the psychopathic behaviour manifested by the eldest. Case 3.6 died at Mont Park at the age of forty-three, where she was originally diagnosed as suffering from schizophrenia. Her subsequent history revealed that she was a case of Huntington's Chorea. Her two children, the eldest of whom is now twenty-six, as yet have not manifested any demonstrable abnormality.

Family "H"

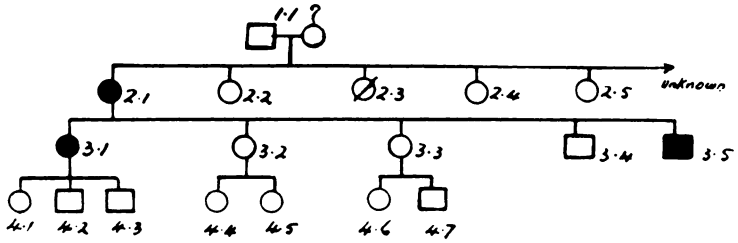
Family "H" originally came from Glasgow, Scotland. Case 1.1 spent a short period in a mental hospital in Scotland, but beyond the fact that she had chorea, no details of her condition are available. Of her three children, the eldest, Case 2.1, who migrated to Victoria with his seven children, developed signs of chorea at the age of sixty, when he was "paid off" from work on account of his physical inability to carry on the skilled operations of a glass cutter.



His present age is seventy-five, but there are no indications of any appreciable mental deterioration. The second member, Case 2.2, migrated to America, and the third, Case 2.3, died in adolescence. Of the seven children of Case 2.1, only one, Case 3.2, is known to have so far developed signs of Huntington's Chorea, which became manifest at the age of thirty-nine. Five years later he was admitted to Royal Park Receiving House grossly choreic. Mentally, he was described as being paranoidal and abnormally excited. Although no longer in hospital, he is now showing marked signs of mental enfeeblement. His three children, who are below the age of twenty, so far give no indication of the onset of Huntington's Chorea.

Family "I"

The parents of Case 1.1 came from Maitland, New South Wales. Unfortunately, there is no evidence to indicate that either of them suffered from Huntington's Chorea. It is known that they had a large family. We have been able to trace only three cases with Huntington's Chorea, and in each instance mental symptoms preceded chorea. Case 2.1 was described as being exceedingly jealous, violent tempered, and neglectful of her five children, and for some years prior to her admission to hospital, at the age of thirty-four, she spent most of the



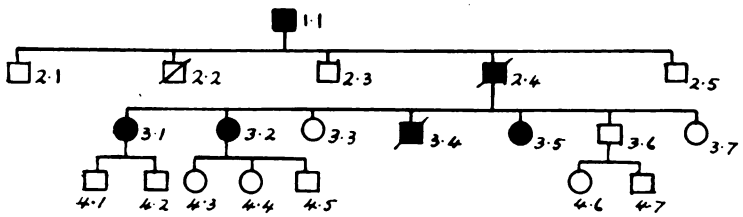
day in bed and completely neglected her household duties. She died at the age of forty-three, grossly choreic. Her eldest daughter, Case 3.1, under the age of twenty, was apparently a placid, even tempered, likeable, but somewhat gullible person. At about the age of twenty she became very erotic, neglectful of her appearance and household duties, threatened to commit suicide and injure the children. At the age of twenty-three, just prior to her admission to a mental hospital, she began to lose control of her legs. Dementia was rapid and profound, death taking place at the early age of twenty-nine. Her three children are below the age of ten years.

Case 3.2 is described as being a silly, giggling, "feather brained" girl, subject to violent outbursts of temper. There are as yet no signs of chorea.

Case 3.3 is known to be a prostitute. Case 3.4 is a stammerer who is very tense and over anxious. Case 3.5, the youngest member of this generation, is below average intelligence and sits for hours without speaking. He has definite twitchings of the face, and so it would appear that he is an early case of Huntington's Chorea. In those members of this family not yet diagnosed as suffering from this disease, the abnormal personalities could obviously be attributed to the deplorable environmental conditions—broken home—alcoholic father—years of poverty and neglect—as was the case in this particular family, but despite this they may still possibly be found to be also associated with Huntington's Chorea eventually.

Family "J"

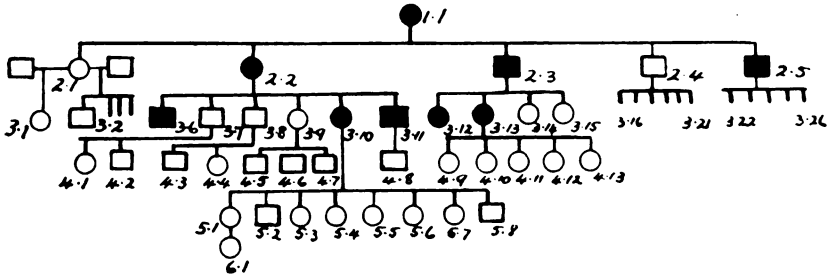
The original member of this family (Case 1.1) was born in County Clare, Ireland, and arrived in this country during the latter half of the last century. He is reported by his grandchildren as having suffered from "St. Vitus Dance". Of the first generation born in Australia,



Case 2.2 died in a mental hospital with the diagnosis of senile dementia. The only member of this generation known to have definitely suffered from Huntington's Chorea was Case 2.4. Of his seven children Cases 3.1, 3.2, 3.4, and 3.5 are all known to be suffering from Huntington's Chorea at the present time.

Family "K"

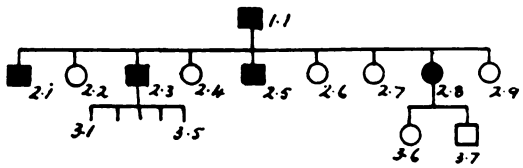
Case 1.1 arrived in Victoria from Tasmania, but it is reasonably supposed that she had originally migrated from England. At the age of fifty, she developed restlessness and choreiform movements. Death occurred twenty years later at the age of seventy. There is no record of any siblings being in this country. As a result of her first marriage, there were five children, three of whom developed Huntington's Chorea at approximately the age of forty. One of these (Case 2.2) spent a short period in a mental hospital. There was no issue of the second marriage. Of the six immediate descendants of Case 2.2, three developed chorea. Case 3.6 first showed typical choreiform movements at the age of forty-four and five years later began to show signs



of mental enfeeblement and confusion. Cases 3.10 and 3.11 are as yet presenting early signs only of chorea without any apparent mental change. Of the descendants of Case 2.3, Case 3.12 was admitted to Royal Park Mental Hospital at the age of thirty-eight in an advanced state of dementia. The choreiform movements were gross and generalized, death occurring one year after admission. There was no issue of her marriage. Case 3.13 first showed signs of the disease at the age of thirty-two. Fourteen years later, at the time of examination, she was moderately demented and completely bed-ridden. There were five children of Case 2.5, but no information is available. The children of all affected members of the third generation are still below the age of twenty-two, and as yet there are no signs of the disease in any one of them.

Family "L"

The original members of this family migrated from Germany to Victoria approximately ninety years ago. Nothing is know of their immediate descendants, except that a son (Case 1.1) and his family moved from Victoria to Tasmania early this century. He was admitted to the New Norfolk Mental Hospital at the age of fifty-five with a history of choreiform movements of the upper limbs and mental deterioration. Two sons (Cases 2.1 and 2.3) developed chorea and showed personality changes at about the age of forty, and both were subsequently admitted to the same hospital. Case 2.5 was admitted to Mont Park Mental Hospital at the age of 31, the provisional diagnosis being general paralysis of the insane. It was later revealed that his Wasserman reaction was negative and that he had been subject to epilepsy for a number of years. On examination, there was a stumbling gait, slurred speech and at



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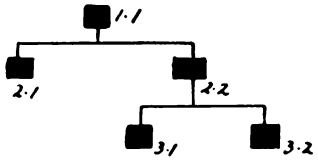
times a marked restlessness and confusion. He died two years later as a case of Huntington's Chorea. Case 2.8 presented herself for treatment at the age of thirty-nine, when the history given was that she had for the past three years suffered from choreiform movements, insomnia, and inability to manage her household duties to the satisfaction of her husband. On examination, there was marked chorea of the upper limbs and head and explosive articulation. She has since been admitted to Royal Park Mental Hospital.

Family "M"

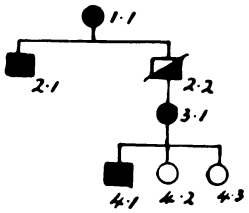
Detailed information concerning Cases 1.1, 2.1 and 2.2 of Family "M" is unavailable, except for the fact that all three had suffered from chorea and were insane. Case 3.1 was admitted to the Ballarat Mental Hospital at the age of thirty-seven. It is recorded that the earliest mental symptoms shown consisted of abnormal aggression and maniacal excitement



just prior to his admission to hospital, where he died two years later, twelve years after the onset of choreiform movements. His brother (Case 3.2) was admitted to the Bundoora Repatriation Mental Hospital at the age of forty, ten years after the onset of chorea. Mentally he is described as being restless, irritable, aggressive and very depraved in his habits. Death occurred five years later at the age of forty-five.

Family "N"

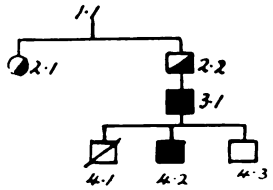
Case 1.1 was reported to have had St. Vitus Dance for some years prior to her death in the Kew Mental Hospital some seventy years ago. Details of her clinical history are unavailable, but it is recorded that she had arrived in this country from England some twenty years previously. Her elder son, Case 2.1, as a result of Huntington's Chorea, was admitted at the age of forty to a mental hospital in New South Wales. The younger son was suspected of having committed suicide at an early age. Although no actual chorea was observable prior to his death he was known to be subject to violent outbursts of temper. In the case of his daughter, Case 3.1, who was admitted to the Sunbury Mental Hospital at the age of thirty-five, the clinical notes state that since the age of twenty-eight she had been showing signs of becoming



mentally unbalanced. The earliest symptoms shown were paranoid trends and extreme irritability. She eventually became quite incapable of managing her home. Choreiform movements did not become particularly noticeable until several months prior to her admission to hospital. Death occurred at the age of thirty-nine. In the next generation, Case 4.1 began to show signs of chorea at the age of twenty-six whilst he was still in the army. The first symptom was his inability to roll a cigarette. Later, it was noticed that he began to make grunting noises and a year after this began to show unsteadiness in his gait. In spite of the fact that he was drinking heavily, he continued working as a labourer for three years after the onset of his illness before he was eventually admitted to the Repatriation Hospital and later transferred to the Bundoora Repatriation Mental Hospital on account of his mental deterioration. When last heard of his two siblings, Cases 4.2 and 4.3, were in good health and showing no signs of chorea.

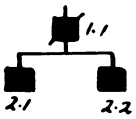
Family "O"

The records of this family are still incomplete. Cases 2.1 and 2.2 had both been rather loosely described as being nervy and it is thought that they both possibly suffered from Huntington's Chorea. Case 3.1 developed choreiform movements and irritability at the age of forty and died twelve years later. Case 4.1 was killed in action in World War II. Case 4.2 was discharged from the Army after five years' service, on account of nerves. The earliest symptoms were irritability, irresponsibility, and erotic behaviour, for at least three years prior to the appearance of choreiform movements. He was admitted to Sunbury Mental Hospital at twenty-nine and died ten years later. Case 4.3, who has three children, is apparently normal.



Family "P"

Case 1.1 died at a Mental Hospital from Huntington's Chorea. No other details of this case are available. Of the two children, Case 2.1 began to show signs of chorea at fifty-four, symptoms being clumsiness, fidgety movements, jerkiness in writing, and unsteadiness in gait.

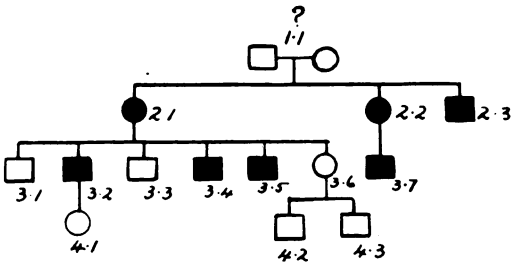


The first mental symptoms, ante-dating chorea for at least two years, were irritability, abusiveness, lack of concentration, and gradual deterioration generally of the higher mental faculties. His brother (Case 2.2) developed chorea at the age of fifty-three. Apart from having no insight into his physical disability, there are as yet no other apparent mental symptoms, and he is still able to carry on his business as a hotel proprietor.

Family "Q"

This family includes seven known cases of Huntington's Chorea, but details are only available in the following three cases:

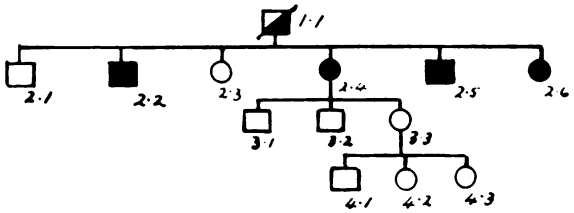
Case 2.1 was admitted to the Ballarat Mental Hospital at the age of forty-six when both dementia and chorea were well advanced. She died five years later. Case 3.4 first showed signs of chorea at forty-five and was admitted to Royal Park eleven years later on account of



depression. He died in a benevolent home at the age of fifty-three. Case 3.5 was admitted to Kew Mental Hospital at the age of thirty-seven and was diagnosed originally as a mental defective, no doubt due to his simple dementia associated with infantilism and absence of secondary sexual characteristics. Choreiform movements, which were relatively slight on admission, gradually became more marked, and, at the age of forty-three, when he died, he was grossly choreic.

Family "R"

Case 1.1 committed suicide at the age of thirty, and although no factual information is available it is reported that he was probably a case of Huntington's Chorea. Four children



are known to have been affected by Huntington's Chorea, although Case 2.4 was the only one admitted to a mental hospital, which was at the age of sixty-two. Signs of chorea had been present since the age of fifty. She was paranoid and mentally enfeebled.

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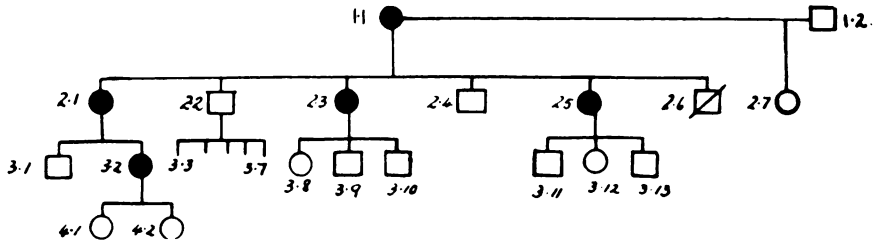
Family "S"

Case 1.1, who lived in Scotland and was known to have died of Huntington's Chorea, had six children, the eldest of whom (Case 2.1) migrated to Victoria. Case 1.2, the only known sibling of this generation, is still quite well and is living near Edinburgh. Case 2.1 first showed symptoms of Huntington's Chorea at the age of forty and died fourteen years later. Case 2.2, at present living in Edinburgh, is reported as being quite well. Case 2.3 died of Huntington's Chorea five years ago. Case 2.4, who is living in Bathgate, is reported as being normal. Case 2.5 is at present in the Edinburgh Infirmary suffering from Huntington's Chorea. Case 2.6 died at the age of fourteen.



In the third generation, Case 3.1, at present aged thirty-three years, is reported as being quite normal. Case 3.2, who was recently admitted to Royal Park Receiving House at the age of twenty-seven, suffered from Sydenham's Chorea at the age of seven. On admission to hospital she appeared to be emotionally impoverished and rather indifferent in manner. She had apparently been anergic and neglectful of herself, her children, and her home, and is said to have been extremely irritable and violent towards her family. She was provisionally diagnosed as a schizophrenic, and, although there are as yet no signs of choreiform movements, the probability is that she is an early case of Huntington's Chorea in which mental symptoms have preceded the appearance of chorea.

References

Babcock, H., *Arch. Psychol.*, 1930. No. 117.

Idem, *J. Abnorm. (soc.) Psychol.*, 1932, 27–63.

Bell, J., "Nervous Diseases and Muscular Dystrophies, Part 1. Huntington's Chorea," *The Treasury of Human Inheritance*, 1934, **4**. London: Cambridge Univ.

Brothers, C. R. D., *The History and Incidence of Huntington's Chorea in Tasmania*, 1950. A'sian Med. Pub. Co.

Burt, C., *Mental and Scholastic Tests*, 1922. London: King and Son, Ltd.

Cattell, R. B., *The Description and Measurement of Personality*, 1946. New York: World Book Co.

Idem, "The Confirmation and Clarification of Primary Personality Factors", *Psychometrica*, 1947, 197–220.

Davenport, D., *Huntington's Chorea in Relation to Hereditary and Eugenics*, 1916. Cold Spring Harbour, Long Island, N.J.

Goldstein, K., and Scheerer, "Abstract and Concrete Behaviour. An Experimental Study with Special Tests", *Psych. Monog.*, 1941. Illinois: North Western University.

Klopfer, B., and Kelly, D. M., *The Rorschach Technique*, 1946. London: G. G. Harrap and Co.

Minski, L., and Guttmann, E., "Huntington's Chorea", *J. Ment. Sci.*, 1938, 348, 21–96.

Office of Scientific Research and Development and Cornell University, "The Use of the Cornell Service Index", *War Medicine*, 1945, **7**, 214–217.

Idem, "The Cornell Index", *Psycho. Som. Med.*, 1946, **8**, 411–413.

Rorschach, H., *Psychodiagnostics*, 1937. 3rd ed. Bern: Huber.