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SHORT COMMUNICATION

Origin and Migration of Huntington's Chorea in Canada:

Preliminary Report

ANDRE BARBEAU, M.D., F.R.C.P.[C]* and
GABRIELLE FULLUM, R.N., *Montreal*

ABSTRACT

This preliminary report is part of a full-scale investigation of Huntington's chorea throughout the world. Data were obtained on some 820 possible cases of Huntington's chorea in Canada, and they were of sufficient quality in 633 cases to enable pedigrees to be drawn up of 104 families. The origin of 75 of these families was traced outside Canada. It was found that 55 of these kinships originally came from the British Isles, contrary to the prevalent feeling that incriminated United States sources. Only 57 of the 633 cases had moved from their first province of residence at the time of reporting. Thus, large and frequent migrations are not the rule, in the Canadian group, as had been previously reported by other authors.

HUNTINGTON'S chorea is a relatively rare disease of the central nervous system characterized mainly by dementia and choreiform movements. The main interest lies in the facts that it is inherited as an autosomal dominant and that its history can be traced back many hundreds of years.

In the course of a study of the world-wide distribution and migration patterns of this illness, we have gathered information on some 820 Canadian cases. The purpose of this preliminary report is to draw the attention of physicians and administrators to this problem in the hope that they will transmit their information to our Huntington's Chorea Registry in Montreal. It is planned to pursue this study in much greater detail and to pool the information accruing from many countries.

MATERIAL AND METHODS

Following announcements in various medical journals as well as letters to the superintendents of a number of mental hospitals, we received information on 820 possible cases of Huntington's chorea, living or dead, and from all 10 Canadian provinces. After preliminary examination of this material, it was considered necessary to exclude 187 of these cases for the following reasons: incomplete or uncertain diagnosis; no family history obtained; or only one generation of chorea known. It was realized, of course, that many of these cases are true examples of Huntington's chorea, but they cannot be properly classified and studied until more information is received. On the remaining 633 cases data were obtained of sufficient quality to enable us to prepare 104 pedigrees. Each pedigree was traced as far back as possible, all places of residence and the earliest known origin being noted. It was thus possible to trace the first ancestor who came to Canada in each of 75 families. Twenty-nine pedigrees could not be completed and their country of origin remains unknown. The present report concerns mainly the families comprising the first-noted group.

RESULTS

Table I shows the country of origin of 75 kinships now known to reside in Canada. It is readily apparent that the British Isles were the source of most of the cases; a similar pattern is also reported from the United States of America[1] and Australia.[2] Fifty-five of the 75 families (72%) in our study came from that area. The United States is the second most frequent source, but it is likely that some of the six kinships reported from that country also originated from Great Britain, Scotland or Ireland. The other countries are fairly evenly represented, most of them being from Northern or Central Europe. It is noteworthy that none of our 75 kinships originate from the so-called "Latin" countries: Italy, Spain or South America.

TABLE I.—HUNTINGTON'S CHOREA: ORIGIN OF 75 CANADIAN KINSHIPS

	No. of kinships
England	31
Scotland	15
Ireland	9
U.S.A.	6
Germany	4
Holland	3
France	3
Norway	2
Greece	1
Belgium	1

Of the 29 kinships whose first Canadian ancestor could not be traced, 19 are from Ontario, four from Quebec, three from New Brunswick, two from Saskatchewan and one from Prince Edward Island.

*Section de Neurologie, Université de Montréal.

TABLE II.—HUNTINGTON'S CHOREA: DISTRIBUTION OF REPORTED CASES AT TIME OF REPORT

Province	*No. of kinships*	*No. of cases*
Newfoundland	—	—
Prince Edward Island	1	1
Nova Scotia	5	55
New Brunswick	4	7
Quebec	17	99
Ontario	63	387
Manitoba	6	9
Saskatchewan	12	23
Alberta	14	31
British Columbia	15	21
Total	137	633

Table II indicates the distribution of the reported cases (living or dead) throughout Canada. Cases were reported from Newfoundland, but data on these were received too late to be included in this study. Again Ontario leads all provinces, both in total number of cases and in the number of kinships. Some kinships have members in more than one province, which explains the total of 137 instead of 104, to account for the 633 reported cases.

TABLE III.—HUNTINGTON'S CHOREA: INTERPROVINCIAL MIGRATION PATTERN

		No. of cases									
From		*1*	*2*	*3*	*4*	*5*	*6*	*7*	*8*	*9*	*10*
To	*1*	—	2	1	1	4	1	2	0	0	0
	2	0	—	0	0	16	0	1	0	0	0
	3	0	1	—	0	3	0	0	0	0	0
	4	0	0	2	—	2	0	0	0	0	0
	5	0	0	0	1	—	2	0	1	0	0
	6	0	0	0	0	0	—	0	0	0	0
	7	0	0	0	0	0	0	—	0	1	0
	8	0	0	0	0	1	0	0	—	0	0
	9	0	0	0	0	0	0	0	0	—	0
	10	0	0	0	0	0	0	0	0	0	—
	U.S.A.	0	0	1	0	11	3	0	0	0	0

1. British Columbia
2. Alberta
3. Saskatchewan
4. Manitoba
5. Ontario
6. Quebec
7. New Brunswick
8. Nova Scotia
9. Prince Edward Island
10. Newfoundland

A study was also made of the interprovincial migration pattern. Table III shows that only 57 cases had moved from their place of origin at the time of the report, which is considerably less than was expected from the impressions of many other authors and from Archibald's previous study in Canada.[3] Of these 57 cases, 15 moved to the United States of America. Such migration as was evident was mainly westward.

DISCUSSION

It is much too early to draw definite conclusions from a preliminary study of this nature. The only purpose of reporting these data is to draw attention again to the problem and to outline appropriate approaches to it. Even though Huntington's chorea is a rare disease, the social and economic difficulties resulting from it are such that its annual cost to the nation (in loss of income as well as in pensions, welfare costs, etc.) is over one million dollars. The personal and social problems involved cannot be measured.

Because of the dementia almost always present in its victims, Huntington's chorea is a good model to study both biochemically and genetically for a possible better understanding of mental illness. This study also serves the purpose of making such material available for other interested investigators.

Two general conclusions can already be drawn. Firstly, most Canadian kinships with Huntington's chorea originate from the British Isles. Secondly, there is a very limited amount of migration of affected families across the country. Once established in an area, these families tend to remain stationary, unless they live in large cities. This study is continuing, and we would again solicit further information from Canadian doctors concerning patients and families affected by this illness.

SUMMARY

A study of 633 Canadian cases of Huntington's chorea, arising from 104 independent pedigrees, shows that the majority of patients are of British origin (England, Ireland, Scotland) and that the families once established reveal little tendency to move.

We would appreciate the collaboration of Canadian doctors to enable us to continue this study which is at present in its earliest stages.

RÉSUMÉ

Une étude de 633 cas canadiens de chorée de Huntington provenant de 104 souches indépendantes montre que la majorité des patients sont de descendance britannique (Angleterre, Irlande, Ecosse) et que les familles, une fois établies, ont peu tendance à se déplacer. Nous aimerions recevoir la collaboration des médecins canadiens pour continuer cette étude qui n'est qu'au stade préliminaire.

REFERENCES

1. DAVENPORT, C. B. AND MUNCEY, E. B.: *Amer. J. Insan.*, **73**: 195, 1916.
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3. ARCHIBALD, C. H.: *Bull. Off. Int. Hyg. Publ.*, **30**: 2286, 1938.

PAGES OUT OF THE PAST: FROM THE JOURNAL OF FIFTY YEARS AGO

XIV. KELLY'S AMERICAN MEDICAL BIOGRAPHY

What more delightful in literature than biography? And yet, how uncertain and treacherous is the account which any man can give of another's life! And who is to be trusted to give a correct account of his own? Montaigne is the only great autobiographer; the only man whose spirit and pen make us feel that we know as much of him as any one of us could tell of himself; the only man we believe when he says, "I have either told all, or designed to tell all . . . I leave nothing to be desired or guessed at concerning me." However imperfectly told, the story of any life has an interest which appeals to us in direct proportion as we feel that brotherly sympathy with human effort, careless of the result, whether success or failure.—Sir William Osler, *Canad. Med. Ass. J.*, 2: 938, 1912.