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Increased rate of suicide among patients with Huntington's disease

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SUMMARY The proportion of deaths attributed to suicide was examined among 506 deceased individuals with diagnosed or suspected Huntington's Disease from New England USA. Comparison of this proportion with that of the general population indicated that the odds of a death being due to suicide in the Huntington's disease group is 8·2 times that of the Massachusetts population for persons aged 50 to 69 yr, but no difference appears in the 10 to 49 yr age group. Among the 157 Huntington's disease patients for whom cause of death was known, the corresponding odds estimates are 23·0 for the 50 to 69 yr age group and 2·7 for the 10 to 49 yr age group. More than half of the suicides occurred in individuals who showed early signs of the illness but who had not been diagnosed, suggesting that suicide may occur more frequently in the early stages of the illness.

Huntington's disease, an autosomal dominant neurological disorder with onset in mid life, results in progressive involuntary choreiform movements and cognitive impairment which may be preceded or accompanied by emotional disturbance.[1] Onset of symptoms usually occurs between 30 and 50 years of age but onset as early as 4 years and as late as 65 has been reported.[2]

Death by suicide among individuals affected by Huntington's disease is one of the serious consequences reported for this illness.[3–7] In fact, in his now classic 1872 description, George Huntington included the tendency to suicide as one of the major features of the illness which bears his name.[8] Although suicide is a leading cause of death in the United States, ranking ninth in 1976,[9] suicide prediction remains problematic.[10] However, the recognition of factors associated with suicide in Huntington's disease patients may help in anticipating this risk. This paper presents data collected by the New England Huntington's Disease Center Without Walls on patterns of suicide among patients with the disease.

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Methods

As part of an ongoing census of all persons known or reliably reported to have Huntington's disease in New England, histories of 149 apparently unrelated families with Huntington's disease involving 4919 individuals were collected. Families and individuals were identified through their physicians, other health professionals, the Massachusetts Chapter of the Committee to Combat Huntington's Disease and self-referrals to the research programme. These families comprise all those identified by the New England Huntington's Disease Center Without Walls between July 1980 and January 1983 but do not represent the entire Huntington's disease population because some new cases have been ascertained since then.

Information on the family histories was provided by individuals at risk for Huntington's disease, their spouses, the spouses of diagnosed persons, and diagnosed persons themselves. In all families with living Huntington's disease patients, the diagnosis was made or confirmed by a neurologist. All families had more than one affected member. When additional family members were reported to have had symptoms of Huntington's disease, we checked these reports as thoroughly as possible by correspondence with other relatives, physicians, hospitals,

nursing homes and review of death certificates. Individuals who did not seek medical attention but who were reliably reported by family members as showing signs of the illness are included in this study as "suspected Huntington's disease". Cause of death was established for as many people as possible by medical records, death certificates and non-contradictory lay reports.

In order to determine whether the number of deaths attributed to suicide among Huntington's disease patients was elevated, the percentage of deaths by suicide occurring among the deceased patients was compared with the percentage of the general Massachusetts population in 1976 who died of suicide.[11] We examined suicide trends in Massachusetts between 1920 and 1978 and found them to be similar,[12] and therefore we used 1976 as a representative year. Comparisons of the percentages and calculation of odds-ratios (OR, odds of a death being due to suicide) with adjustment for age and sex were done using the Mantel-Haenzel chi-square statistic and logistic regression analysis.[13]

Results

Within the 149 family histories, there were 403 deceased individuals diagnosed with Huntington's disease (198 males, 201 females, four unknown sex), and 103 deceased individuals suspected of having Huntington's disease (57 males, 45 females, one unknown sex). Cause of death was ascertained for 157 of these 506 individuals (table 1).

Among the 403 deceased Huntington's disease patients, there were nine documented suicides. All were male. Of the 103 suspected Huntington's disease patients, there were eleven documented suicides (seven males, four females) (table 2). Two of these eleven were in psychiatric hospitals. With the exception of one suicide occurring in 1934, all suicides reported herein occurred between 1956 and 1982. Thirteen of the 20 persons committing suicide were known to be married at the time of death and 13 had children. Two of the female suicides were sisters; two of the males were uncle and nephew. No other individuals were related. Method of suicide was ascertained for 14 of these individuals and included drowning (four), asphyxiation by poisonous gas (two), gunshot (one), hanging (two), jumping from a high place (three), drug overdose (one), and fire (one). As shown in table 1, suicide is the third leading cause of death among the 157 individuals for whom cause of death was established. Age of death was ascertained for 342 of the 506 deceased Huntington's disease patients and the mean was 54·8 years. The age of death is known for 19 of the 20 individuals who committed suicide and who were diagnosed or suspected of having Huntington's disease. The mean was 47·7 years. The youngest suicide was at 24 and the oldest at 69 years.

Table 1 *Primary* cause of death among individuals with suspected or diagnosed Huntington's disease*

Cause of death	*Number of deaths*	*Percent*
Unknown	349	—
bronchopneumonia	50	31·8
heart disease	24	15·3
suicide	20	12·7
cerebrovascular disease	11	7·0
Huntington's chorea (only cause listed)	10	6·4
accidents	10	6·4
other neurological disease	9	5·7
cancer	7	4·5
psychiatric illness	4	2·5
tuberculosis	4	2·5
generalised arteriosclerosis	3	1·9
all other	5	3·2
Total	506	99·9

*Primary cause only, excluding Huntington's Disease, as listed on death certificates, in medical records or as reported by relatives.

Table 2 *Method of ascertainment of cause of death*

	Huntington's disease diagnosed N = 403	*Huntington's disease suspected N = 103*
Suicide confirmed by record	7	5
Suicide reported by family	2	6
Total	9	11

Suicide accounts for 2·3% of all deaths of the 403 diagnosed Huntington's disease patients, although no female diagnosed Huntington's disease patients committed suicide. Among the 506 deaths with suspected or diagnosed Huntington's disease, 20 committed suicide accounting for 4% of all deaths. This is significantly greater than the 1% occurrence of suicide reported for the general population ($p < 0{\cdot}00001$).[12] However, these percentages are not adjusted for age or sex differences. Examination of differences between males and females (using the combined group of patients with definite and suspected Huntington's disease), in the percentage of deaths due to suicide revealed that approximately four times as many male Huntington's disease patients committed suicide as females. However, this number is not significantly different from the Massachusetts population: in a pooled estimate of both the Huntington's disease patient group and the Massachusetts population, approximately twice as many male deaths are attributed to suicide as female deaths ($p < 0{\cdot}0001$). Because the sexes were similar in this respect for the two populations and because of small numbers of female suicides we combined males and females in table 3.

Table 3 *Suicide occurrence by age among 506 deceased Huntington's disease patients and the Massachusetts general population*

Age group (yr)	*Huntington's disease*			*Massachusetts general population*			*Odds-ratio*	*Overall Odds-ratio*
	Total deaths	*Observed suicide deaths*	*Percent suicide deaths*	*Total deaths*	*Observed suicide deaths*	*Percent suicide deaths*		
0–9	2	0	0	1029	0	0	—	
10–19	6	0	0	523	26	5·0	0	
20–29	10	2	20·0	979	158	16·1	1·3	
30–39	28	3	10·7	870	92	10·8	1·0	1·06
40–49	69	4	5·8	2115	82	3·9	1·5	
50–59	97	6	6·2	5612	74	1·3	4·9*	8·19
60–69	73	4	5·5	10118	52	0·5	11·2*	
>70	57	0	0	32792	37	0·1	—	
unknown	164	1	0·6	11	0	0	—	
Total	506	20	4·0	54049	521	1·0		

*p < 0·00005

Comparisons with the Massachusetts population using age-specific rates of suicide show definite age group differences. The results in table 3 suggest that the proportion of deaths due to suicide in Huntington's disease patients and the Massachusetts general population is not significantly different among the individuals dying between ages 10 and 49 yr. However, in the 50 to 69 yr age group, a significantly greater percentage of Huntington's disease deaths are attributed to suicide.

Logistic regression analysis confirmed this interpretation of the data. No difference between Huntington's disease patients and the Massachusetts population in the occurrence of suicide is observed for individuals dying between the ages of 10 and 49 (OR = 1·06 with 95% confidence interval 0·53, 2·11). However, in the 50 to 69 yr age group, the observed and expected frequencies are significantly different (OR = 8·19 with 95% confidence interval 4·21, 15·91). The odds-ratios of the two age groups of Huntington's disease patients are significantly difference from one another (p < 0·0001).

The differences are even more pronounced if we only consider Huntington's disease patients for whom a cause of death was established. As shown in table 4, logistic regression analysis of the sub-sample of 157 individuals for whom cause of death was established shows significant differences in the occurrence of suicide in both the 10–49 yr age group (OR = 2·7 with 95% confidence interval 3, 5·6) and 50–69 yr age group (OR = 23, with 95% confidence interval 11·1, 46·1). The difference between these two groups of Huntington's disease patients is also significant (p < 0·0001). If those 157 patients are representative of the entire sample, the higher proportion of suicide which occurs in this smaller group may be a more accurate reflection of the occurrence of suicide among Huntington's disease patients.

Discussion

Over the past 50 years, suicide has accounted for approximately 1% of all deaths in Massachusetts.[12] Therefore, in this sample of 403 individuals with Huntington's disease, suicide occurred at least twice as often as in the general population, and among the 506 individuals with either suspected or diagnosed Huntington's disease, at least four times as frequently. The occurrence of suicide in our sample is comparable to the proportion of Huntington's disease patients committing suicide reported elsewhere.[5 7]

The above comparisons with the general popula-

Table 4 *Suicide occurrence by age among 157 Huntington's disease patients with an established cause of death and the Massachusetts general population*

Age group (yr)	*Huntington's disease*			*General population*			*Odds-ratio*
	Total deaths	*Observed suicide deaths*	*Percent suicide deaths*	*Total deaths*	*Observed suicide deaths*	*Percent suicide deaths*	
0–9	1	0	0	1029	0	0	—
10–49	49	9	18·4	4487	358	8·0	2·7*
50–69	67	10	14·9	15730	126	0·8	23·0*
>70	23	0	0	32792	37	0·1	—
unknown	17	1	6·0	11	0	0	—
Total	157	20	12·7	54049	521	1·0	—

*p < 0·00001

tion are based on crude rates which do not account for differences among age groups and between men and women. Age and sex adjusted rates show that suicide in the New England Huntington's disease population is eight times greater than in the general population among individuals who die between ages 50 and 70 yr but that there is no difference among people who die before age 50 yr. However, cause of death was established for only 157 affected individuals; therefore suicide cannot be excluded as having been the cause of death for some of the remaining 349. Because some of those patients with an unknown cause of death may have been suicides, these data may underestimate the occurrence of suicide among Huntington's disease patients in this sample.

Examination of the occurrence of suicide among those patients for whom cause of death has been established may yield a more accurate appraisal of risk of suicide. This analysis shows significant differences between Huntington's disease patients and the general population in both age groups. Between the ages of 10 and 49 yr, Huntington's disease patients are almost three times as likely to commit suicide and in the 50–69 yr age group, 23 times more likely.

The occurrence of suicide among Huntington's disease patients in this study may be an underestimate for several reasons. Only those patients for whom suicide was definitely reported as the cause of death (by close relatives, death certificates or medical records) were classified as such. While suicide may be suspected in the reported instances of accidental death, such cases were excluded from the suicide categorisation because of lack of verification.

It should be pointed out, however, that in the general population, officially reported suicide statistics may underestimate the actual occurrence of suicide by as much as 22%.[14 15] If the data in table 2 are reanalysed assuming this underestimate among the general population, Huntington's disease patients in the 50 to 70 yr age group still have a significantly increased rate of suicide (OR = 5·0 with 95% confidence interval 2·74 8·95, $p < 0.0001$).

Reports which indicate that suicide is not a major risk among Huntington's disease patients may be overlooking undiagnosed persons with early signs as recognised by the family.[16–18] It is striking that in our sample, the prevalence of suicide appears four times higher among suspected Huntington's disease patients than among those diagnosed. If many of the suspected cases were in the early stages of the illness, as seems likely, this implies that patients early in their disease are particularly prone to suicide. This possibility warrants further investigation with a larger group of patients and has clinical importance. If the diagnosis is made when subtle signs of Huntington's disease are apparent, the affected individual is given the opportunity to discuss feelings about Huntington's disease, and the physician or genetic counsellor can then provide necessary psychological support.

An association between organic disease and suicidal potential has been suggested among patients with cancer,[19] diabetes,[20] renal failure[21] and psychiatric disorders.[22] Necropsies of suicide victims reveal them to be on the whole, a group with serious physical illnesses.[23] Thus, the increased occurrence of suicide among patients with Huntington's disease may not represent a disproportionate increase over patients with other disorders.

The proportional mortality rates calculated in this analysis do not express absolute risk of suicide for living Huntington's disease patients. The higher proportion of Huntington's disease patients dying of suicide is suggestive of a difference with the general population and further studies are needed to clarify these data. However, these preliminary data show that the relative risk of suicide occurrence among Huntington's disease patients ranges from two to 23 times that of the general population. This higher proportion of deaths by suicide among Huntington's disease patients necessitates appropriate exploration of suicidal potential at the time of initial evaluation.

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References

[1] Myers RH, Schoenfeld M, Bird ED. Huntington's disease: Genetics, Chemical Pathology and Management. *Progress in Medical Genetics* (in press).
[2] Myers RH, Goldman D, Bird ED *et al.* Maternal transmission in Huntington's disease. *Lancet* 1983;**1**:208–10.
[3] Ladame P-L. Suicide et chorée de Huntington. *Encephale* 1911;**6**:422–7.
[4] Minski L, Guttman E. Huntington's chorea: a study of 34 families. *J Ment Sci* 1938;**84**:21–96.
[5] Reed TE, Chandler JH. Huntington's chorea in Michigan. I. Demography and Genetics. *Am J Hum Gen* 1958;**10**:201–25.
[6] Dewhurst K, Oliver JE, McKnight AL. Socio-psychiatric consequences of Huntington's disease, *Br J Psychiatry* 1970;**116**:255–8.
[7] Hayden MR, Ehrlich R, Parker H, Ferera SJ. Social

Perspectives in Huntington's Chorea. *S Afr Med J* 1980;**58**:201–3.
[8] Huntington G. On chorea. *Medical and Surgical Reporter* 1982;**26**:317–21.
[9] United States Department of Health and Human Services. *Vital Statistics of the United States* 1976;1–246, Vol II-A, Mortality.
[10] Kramer M, Pollack ES, Redick R, Locke BZ. *Mental Disorders/Suicide* 1972;175. Cambridge:Harvard University Press.
[11] United States Department of Health and Human Services. *Vital Statistics of the United States* 1976;7–283, Vol II-B, Mortality.
[12] Commonwealth of Massachusetts. Annual Report of Vital Statistics, Department of Public Health, Public Document #1, 1920–1978;15.
[13] Schlesselman JJ. *Case Control Studies,* Oxford University Press, 1982; 227–90.
[14] Sainbury P, Jenkins JS. The accuracy of officially reported suicide statistics for purposes of epidemiological research. *J Epidemiol Community Health* 1982;**36**:43–48.
[15] Brugha T, Walsh D. Suicide past and present. The temporal consistency of under-reporting. *Br J Psychiatry* 1978;**132**:177–9.
[16] Brothers CRD. Huntington's Chorea in Victoria and Tasmania. *J Neurol Sci* 1964;**1**:405–20.
[17] Mattson B. Huntington's Chorea in Sweden II. Social and Clinical Data. *Acta Psychiatr Scand* 1974;**Supp. 225**,221–35.
[18] Chiu E. Causes of death in Huntington's Disease. *Med J Austr* 1982;**1**:153.
[19] Louhivuori KA, Hakama M. Risk of Suicide Among Cancer Patients. *Am J Epidemiol* 1979;**109**:59–65.
[20] MacGregor M. Juvenile diabetes growing up. *Lancet,* **1** 1977;944–5.
[21] Washer GF, Schroter GP, Starzl TE, Weil R. Causes of death after kidney transplantation. *JAMA* 1983;**250**:49–54.
[22] Tsuang MT. Suicide in schizophrenia, manics, depressive, and surgical controls. *Arch Gen Psychiatry* 1978;**35**:153–5.
[23] Stewart I. Suicide: the influence of organic disease. *Lancet* 1960;**2**:919–20.