Social Perspectives in Huntington’s Chorea

M. R. HAYDEN, R. EHRLICH, H. PARKER, S. J. FERERA

SUMMARY

The social implications of Huntington’s chorea are serious and far-reaching, affecting all members of the family and the community as a whole. The relatively common occurrence of suicide and of major and minor crimes are cause for concern. The disease imposes a significant economic burden on both the family and society. The minimum direct cost to the state of a single affected person with Huntington’s chorea in South Africa is estimated at R23 000.

During the current national survey, in which data concerning nearly 500 persons with Huntington’s chorea have been analysed, it was soon apparent that the psychosocial implications of this disorder were extremely important and largely unexplored.

The prevalence of Huntington’s chorea in the different population groups of South Africa has recently been reported.1 It is important, however, that data concerning the number of affected persons be seen in the correct perspective, as Huntington’s chorea is a much more important disease than can be assessed from estimates of its prevalence. It is a prime example of a family disease; for every affected individual there are approximately 10 people in the immediate environment, including those at risk, the unaffected spouse, and other family members, who suffer from the far-reaching social consequences of the disorder. The cost of the disease to the community in economic terms is also cause for concern.

The purpose of this article is to provide a greater understanding of the social repercussions of Huntington’s chorea, in the hope that this may highlight the unmet social needs of the patient and his family.

PSYCHOSOCIAL CONSEQUENCES FOR THE AFFECTED PERSON

Many patients in the earliest phase of their illness expressed great apprehension at the possible social implications of being afflicted with ‘that dreadful hereditary disease, the slow killer, which slowly and inexorably disintegrates the mind and body’. They feared the physical and mental disability, economic impoverishment and social stigma of the ‘disease of inherited madness’. Many also felt guilty about not giving their children a stable home and about possibly transmitting the gene for Huntington’s chorea to their offspring.

The tragedy of this disorder is that it strikes in the prime of life, when social responsibilities and personal and financial possibilities are greatest. Patients ‘grow senile without growing old’.2 The affected persons dread losing personal dignity, exemplified by the failure of control of bladder and bowel function. At the time that they most need support, there is the unvoiced fear of anticipated abandonment and consequent loneliness. All these factors stimulate an attitude of extreme dependence and depression.

‘Insanity with a tendency to suicide’ was one of three features mentioned by Huntington3 in his definitive description of the illness (1872). We have had notification of 11 affected persons in the RSA who have died from self-inflicted wounds. All but 1 of these persons were male and this is equivalent to 3,35% of all deaths due to Huntington’s chorea in South Africa. Based on reported rates for 1976, the chance of an affected person with Huntington’s chorea in South Africa committing suicide is approximately 2 200 times greater than in the general population. While no definitive pattern has emerged, patients with this disorder are more likely to commit suicide in the early phases of the illness, when depression is severe and dementia is minimal.4,5

A further 24 persons made serious attempts on their lives which necessitated hospitalization. There were 16 females and 8 males in this group. In other words, males with Huntington’s chorea are more likely to succeed in their suicide attempts, in contrast to the greater frequency of attempted suicide in affected females.

It is clear that suicide is a very real danger for patients with Huntington’s chorea and it is important that this alternative be averted by swift and appropriate intervention.

CRIME AND HUNTINGTON’S CHOREA

It is on record that the earliest transmitters of the gene to the USA clashed with the law of their adopted country, as a result of repeated crimes and misdemeanours.6

In the current survey of Huntington’s chorea in South Africa, there have been repeated instances of antisocial behaviour, including suicide, assaults, stabbings, shooting, theft, two reports of murder, and other more minor crimes such as offences against property. Sexual aber-
rations have included indecent exposure, prostitution and rape. Similar offences have been reported by numerous other authors.  

The presence of antisocial behaviour in patients with Huntington's chorea is clearly established, but the precise determinants of such conduct are more difficult to ascertain. Whether this behaviour is the result of the underlying biochemical defect or rather the consequence of a disturbed social environment is uncertain.

Neurological syndromes have been described which are associated with specific patterns of conduct, such as the frequent presence of religiosity and a deepened interest in moral and ethical issues in some patients with temporal lobe epilepsy.  In other words, specific diseases may be associated with predictable modes of behaviour, which are directly related to the ongoing pathological process. Is this also true for Huntington's chorea?

A good group for comparison with Huntington's chorea patients are the unaffected members of the family, as they are exposed to a similar psychological, social and biological environment. Although no formal study has been performed, it was apparent in the current survey that antisocial behaviour is less common in this 'control' group. Oliver and Dewhurst have reported a large family where, even though the children of affected persons did not carry the gene, their disturbed upbringing resulted in their behaviour, their disturbed upbringing resulted in psychiatric and antisocial sequelae in adult life. However, these misdemeanours seemed less prevalent than in their affected progenitors.

It is likely, therefore, that the offences committed by affected persons are consequent to disinhibition or lack of control of aggressive impulses, which are in part the result of the degenerative process of Huntington's chorea. At the present time, further studies are needed to determine the extent to which the disease and the disturbed home environment contribute to the causation of antisocial behaviour in Huntington's chorea. This problem could be approached by using control groups in the general population, other chronic neurological patients and the unaffected family members.

It is important that the legal profession and prison authorities be aware of this disorder and its social implications. In this context it is relevant that, during the course of the current survey, it was brought to our attention that at least 2 persons had developed obvious signs of Huntington's chorea while in prison.

THE EXPERIENCE OF BEING AT RISK

Every child of an affected patient has an even chance of inheriting the gene for Huntington's chorea. The inability to escape from the unacceptable reality, that they may be 'passive victims of a totally random genetic accident', was devastating for all concerned. 'It's like living under a cliff, waiting for a landslide', was the way a 28-year-old woman described her feelings.

Others have portrayed the experience of being at risk as 'living with a time bomb', and 'playing Russian roulette with a two-barreled gun and somebody else's hand on the trigger'. All at-risk persons interviewed admitted having experienced anxiety at some time. In some the anguish was continuous and incapacitating, and necessitated therapy. For others, denial was often the only way they could continue enjoying constructive, creative lives. Some of these persons married and produced children without informing their spouses of the disease in their family, even though they were aware of its genetic implications. In this way, denial propagated the disease.

A smaller proportion of those at risk either refused to marry, or chose to have childless marriages. At present, this is the only way to decrease the incidence of this disorder in the community. For those who choose to refrain from procreation, there are no other viable prospects for parenthood. By virtue of their at risk status, such persons are not usually accepted as adoptive or foster parents. A theoretical alternative to adoption does arise if the potential carrier is a male. In such a case, artificial insemination of his wife would ensure that the gene would not be transmitted to the next generation and at the same time allow her the fulfilment of pregnancy. This method is, however, fraught with numerous ethical and legal dilemmas.

Individuals at risk of carrying the gene for Huntington's chorea confront numerous serious problems at different stages of their lives, which necessitate urgent and supportive intervention.

THE BURDEN TO THE UNAFFECTED SPOUSE

The unaffected spouse has unique concerns and needs. He or she is often alone, having to cope with the patient at home. The change of marital role, with the husband having to deal with increasing domestic chores or the wife being forced to become the breadwinner for the family, is most difficult for the respective spouses. This difficulty is compounded by the inevitable isolation that ensues following repeated social embarrassments and rejection by old friends.

The hardest fact for the unaffected partners to accept is that they have unwittingly been involved in the transmission of the disease to their children; many described this as their heaviest burden. Furthermore, they were often left with the invidious task of having to inform the children of the implications of their at-risk status. They often resented that they had not been told of the family illness in their partner before marriage. Almost all persons (80%) in the current survey said they would still have entered the marriage if they had known of the illness, but could then have planned for responsible parenthood.

The unaffected spouse often became the target for the patient's delusions, abuse and sometimes violent attacks. 'He used to hit me with his fist and on occasion threw his teacup at me.' While divorce ensued in some families, in
many others the spouse chose to stay with his or her partner. A feeling of acceptance and strength was expressed. 'I married him, and it is my duty to look after him.' Within these families, the spouse became a source of great comfort to all around.

Many partners complained of the indifference of the medical profession to their problems. One woman remarked, 'Doctors just seem to be not interested. The doctor at the hospital told me to take her (affected daughter) home as I probably knew more about this Huntington's chorea than he did. What must I do? I go to the doctors for help and they can do nothing for me.'

While the afflicted patient is the person who first draws medical attention to these families, it is often the unaffected spouse who needs the most attention and help to cope with his or her problems.

**THE ECONOMIC BURDEN**

Huntington's chorea imposes a significant economic burden on both the family and the community.

The largest single economic cost borne by the family is usually due to loss of earnings. This can be offset only partly by state grants, so that affliction entails a constant financial struggle. Furthermore, in contrast to non-genetic chronic diseases, expense in Huntington's chorea recurs in each generation.

The minimum direct cost to society, over the lifetime of a single sufferer in South Africa, is estimated at R23 000. This has been calculated for the father of a Cape Coloured family, over the average 14-year course of the disease, using 1979 values. It includes a 2-week acute hospitalization for initial assessment; attendance at a special clinic every 6 weeks; a daily regimen of pharmacotherapy; a monthly disability grant; a monthly maintenance grant (for two children over a 5-year period) and the price of hospitalization for the last 4 years of his life. Notably, it excludes loss of economic productivity as well as the cost of social work, transport and home nursing services.

With a minimum of 160 persons at present known to have the disease in the RSA, the lowest estimated total lifetime cost to society is thus R3,7 million. Although expenses per patient vary with socio-economic and family circumstances, the above total, based on conservative assumptions and minimum figures, can be regarded as a gross understatement of the real figure.

Although computed differently, it is of interest to compare our figure of R23 000 to a British estimate of £20 000 and an American figure of between $65 000 and $234 000 for the direct lifetime cost per patient.

**CONCLUSION**

The constellation of the mental and physical symptoms, together with the social and genetic implications for affected families, makes Huntington's chorea 'one of the most dreadful diseases that man is liable to.'

While there is no available medical cure, the major task confronting persons involved in the management of affected patients and their families is to provide improved care. Appropriate and adequate pharmacotherapy can alleviate many of the patient's symptoms. With greater understanding of the disease, the social stigma will diminish and families will be free to use the available community resources.

In an attempt to provide co-ordinated care, a special Huntington's chorea clinic has been established at Groote Schuur Hospital, to attend to the needs of affected persons and their families. It was felt that patients' interests would best be served by the simultaneous attendance of all necessary experts, and the staff of the clinic comprises a neurologist, a medical geneticist, a social worker, and a genetic nursing sister. In addition, a psychiatrist is on call. In the 12 months since its inception in January 1979, 98 consultations have been conducted and the clinic has proved to be a most important aid in the management of affected families.

The social concerns and needs of patients with Huntington's chorea are shared by persons suffering from numerous other unrelated genetic and chronic diseases. Awareness of the need for comprehensive care for the whole family in Huntington's chorea highlights the unmet needs and socioeconomic implications of other chronic disorders.

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**REFERENCES**