

Huntington's disease – the experiences of voluntary organisations

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Huntington's disease is associated with a considerable psychiatric morbidity and mortality. These risks are not confined to the sufferer. The relations of victims, both biological and legal, are also at high risk for significant psychosocial dysfunction (O'Shea & Falvey, 1988). Despite recent advances in genetic research, there is still no cure for this chronic choreo-dementia syndrome. Medical management is confined to palliation of symptoms. The doctor and the patient's family, in the face of this unpleasant disease, are wont to exercise a variety of psychological defences which may hinder the delivery and receipt of adequate support (Martindale, 1987).

Cooperation between professionals and self-help groups in general is not always as good as it might be (Black, 1988). This, and my experiences with the Huntington's Disease Association of Ireland, led me to enquire more formally into the experiences of a cross-section of national Huntington societies.

The study

The Huntington societies of Britain, Holland, Ireland, New Zealand and the United States of America were each sent a questionnaire and a covering letter. Enquiries were made as to how helpful the associations had found the medical profession as a whole/general practitioners/psychiatrists in assisting them/showing an awareness of their national or local association/and in helping patients and their families. They were asked various historical details about individual groups and about efforts made to advertise themselves politically and medically.

Various questions concerned the settings in which Huntington patients were treated or nursed. The groups' feelings about genetic counselling, including who should deliver it, were sought, and specific enquiry was made into the exact composition of meetings.

The secretariats of all five national organisations returned detailed replies. A synopsis of each reply is now given:

Britain Association to COMBAT Huntington's Chorea (Head Office: 34a Station Road, Hinckley, Leicestershire LE10 1AP. Telephone 0455 615558).

A minority of "senior members of the medical profession go out of their way to assist us, but the

majority requires assistance from us". Doctors are aware of COMBAT and "many refer families to us". Some doctors "simply 'don't want to know' (and don't know – because they give inaccurate information to families)". Each category of doctor contains the "excellent" and "the frankly awful". Psychiatrists are increasingly reluctant "to give long term accommodation" in their wards.

COMBAT was founded in 1971 with the assistance of the American Marjorie Guthrie, and it was closely modelled on the US organisation. The first secretary was in London; the first branch was in Sheffield. Contacts with doctors are promoted in a wide variety of ways (e.g. advertising, professional seminars, patient support in hospitals) whereas political association tends to be with a few "friendly MPs".

For the greater part of their illness patients are managed at home. Between 12–17% of sufferers (700–1,000 at any one time) spend their last days in institutions. Apart from some places with special expertise (e.g. Sue Ryder Home, Stagenhoe) there are a variety of homes available, e.g. Leonard Cheshire Foundation homes. However, half are placed in psychiatric or psychogeriatric wards. This is rendered necessary by the nursing requirements rather than the psychiatric needs of victims. The closure of psychiatric hospitals is making matters worse for Huntington families. "Social Services Departments should provide residential care for the disabled, but not all do, and they are wary about taking Huntington's disease cases because of the behavioural problems and because the uncontrollable movements alarm other residents."

Genetic counselling is "imperative" and should be given by specialists at genetic clinics. It has, unfortunately, "frequently happened that people at risk are told they must not have children".

Holland *Vereniging van Huntington* (PA Verkuylaan 30, 1171 EE Badhoevedorp, Netherlands).

Some leading academic centres, e.g. Leiden, have been helpful and, if a contact has been established, this network can be expanded. An increasing number of young family doctors have become actively involved in the group's activities following the publication of newspaper articles and appearances in

television shows. Specialists, such as neurologists or psychiatrists, are either unable or unwilling to become involved, preferring to liaise instead with fellow professionals. However, a few psychiatrists in certain centres have shown an active interest.

The Dutch group was funded in 1976 on the initiative of the late Marjorie Guthrie and a number of professors of the Academic Hospital at Leiden. They had perceived a great need to provide support and information for patients and families. Information leaflets through the post and extensive media usage brought it in touch with doctors and legislators. The belief of the organisation that Huntington's disease is primarily a physical affliction has, over the last 12 years, progressively moved the domicile of sufferers away from the psychiatric institution and into nursing homes, two of which provide a special Huntington unit. Almost all cases spend their final days away from home. Short-term in-patient psychiatric care is acceptable only if the treatment offered there is likely to help; longer-term admission to a psychiatric hospital is advocated only in exceptional cases. Every academic hospital in the Netherlands provides skilled genetic counselling. About 2–5% of attenders at meetings are actual patients; the remainder consists of equal proportions of at-risk people and other relatives.

Ireland *Huntington's Disease Association of Ireland (HDAI)* (279 Sutton Park, Dublin 13. Telephone 01 323752)

The medical profession has been helpful when approached. However, the HDAI tends to adopt a low profile. Possibly as a result of this, very few medical referrals to the Association have been made. "There is very little evidence that the medical profession provide(s) meaningful assistance to patients and their relatives."

The HDAI was founded in Dublin in 1985 for similar reasons to COMBAT, on which the group was modelled. The HDAI tried to come in under the British umbrella but, unfortunately, this proved impossible. Contact with doctors was initiated via circulars and, from time to time, contact was established locally with politicians.

Again, most of the duration of the illness is spent at home. About 10–15% (although no hard figures are available) are thought to spend their final days in psychiatric institutions. Also, while various types of in-patient care are seen as necessary, psychiatric care should be specifically indicated. Trained genetic counsellors should provide family planning services, such specialists are practically nonexistent in Ireland, and it is not unknown for the familial diathesis to be hidden from potential marriage partners.

"Patients have attended our meetings. They are mainly supported by well relatives. In one instance

husband and wife attended. Wife had disease but husband said that wife did not know it."

New Zealand *NZ Huntington's Disease Association* (23 Konene Street, Rotorua, North Island).

Doctors, unless they have a specific interest in Huntington's disease, are unlikely to "support either the families or the Organisation". This may be because of the chronic, refractory course of the illness. The Association was founded in 1978 by Mrs Lorree Adam who had nursed her late husband, a victim of the disease. Mrs Adam had received advice and information from American and English sources. Today there are three groups: Auckland, Wellington, Christchurch.

Student nurses are lectured to in mental hospitals by the Association, and doctors are kept up to date with literature. Politicians tend to show transient interest. Most families cope with sufferers for as long as they are able but many, "unfortunately", are placed in mental institutions. The shift of psychiatric in-patients to private hospitals in New Zealand is welcomed by the Association: "... it means the patients are receiving better accommodation and more individual care". This move is a result of the cheaper rates offered by private enterprise over the State system. Very heavy costs are involved in this move if the patient is "not a certified patient under the medical act". Short-term stay in psychiatric hospitals has positive points: education for doctors and a rest for relatives. The Arthur Preston Centre in Melbourne (27 Yarrbat Avenue, Balwyn, Melbourne, Australia) "is truly fantastic, providing for the needs of these people with great understanding".

Attempts are currently being made to set up a genetic counselling service for at-risk persons. Following the recent advances in genetic predictive research everyone initially wanted to be told if they were carrying the gene. However, after some debate and thought, this enthusiasm for knowledge of genetic tests has died down considerably: "... they ... would rather take the 50% chance (that) they won't develop HD".

Actual patients are encouraged to attend meetings.

USA *Huntington's Disease Society of America* (140 W. 22nd Street, New York, NY 10011. Telephone 212 242 1968).

This group has found "the medical profession generally to be unaware of the facts about HD and of the information we provide". This applies to both general practitioners and psychiatrists.

In 1976, Marjorie Guthrie, the wife of folk-singer Woody Guthrie, founded the *Committee to Combat Huntington's Disease*. Marjorie had felt a lack of assistance when she had nursed Woody, a victim of Huntington's disease. The Society has an annual

“Walk on the (Capitol) Hill” in which “we schedule appointments” to see the capital's politicians. Fund-raising is also used as a way of advertising the existence of the disease. Support for increased medical facilities and research is sought by lobbyists in Washington, DC.

Sufferers are sent to VA hospitals, nursing homes, and mental institutions, depending on finances and local conditions. The disease is so often misdiagnosed and kept secret that accurate statistics are impossible.

The Society's position, like the other organisations mentioned above, is that Huntington's disease should not be seen primarily as a psychiatric disorder. Genetic counselling is useful for presymptomatic testing and for eliciting a family history of the disorder. It should be delivered by a sensitive expert. Support group meetings are attended by all interested parties, including patients.

Comment

Huntington's disease has been discussed in detail elsewhere (Falvey & O'Shea, 1983; O'Shea, 1984; O'Shea and Falvey, 1988). National Huntington support organisations owe a debt to the vision and energy of one American, Marjorie Guthrie. Her example was then followed elsewhere, chiefly by relatives distressed by the sufferings of kin. Some founders are themselves at risk of developing the disease.

Black's (1988) assertions that doctors in general provide little support for such organisations is supported by the material presented here. However, psychiatrists do generally know (when asked) what are the chief diagnostic features of the (well established) illness (O'Shea & Falvey, 1988). The teaching in medical schools that Huntington's disease is a rare disorder may inadvertently lower the diagnostic consciousness of practitioners. Also, lobbying by other groups (PKU, autism, multiple sclerosis, Alzheimer's disease) is bound to dilute the impact that Huntington societies may have.

Nevertheless, the disease strikes men and women at the peak of their productivity, often after they have formed a family. The social stigma of Huntington's disease has kept it largely underground. To some extent, recent recombinant DNA research (Gilliam *et al.* 1987) has highlighted the existence of the problem. However, the fact that this form of testing is not always availed of or heeded, and indeed does not provide a cure, has already created ethical dilemmas of no small magnitude (O'Shea, 1984; O'Shea & Falvey, 1988). There is a real danger that this debate may remain confined to a few interested practitioners.

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