

Dealing with the Disadvantaged

Huntington's chorea—a family problem

ROSEMARY YALE



Huntington's chorea affects two main parts of the nervous system: some of the nerve centres controlling bodily movements, so that one of the main symptoms is a disturbance of movement; and some of the centres responsible for mental functions, so that patients may behave oddly or have emotional or other difficulties.

Each child of an affected parent has a 50-50 chance of inheriting and later developing the disease, which usually shows itself at the age of 35-40, though there are cases where it has developed earlier or later. Huntington's chorea is rare, affecting about 1 in 10 000 people in Britain. So many doctors see only one or two cases during their whole career, if that. The following points are therefore to alert doctors to the family stresses inherent in this dominantly inherited disease, and to offer some simple guidelines. A medical booklet describing the illness, by Dr David Stevens, may be obtained from the Association to Combat Huntington's Chorea, Lyndhurst, Lower Hampton Road, Sunbury-on-Thames, Middx (01-979 5055), price 20p. This booklet includes a list of genetic counselling clinics.

Simple guidelines

(1) Because this disease is incurable and combines mental with physical symptoms, families who know of its presence are often extremely anxious. They may react by denying its existence or by refusing to allow information to be given to children or siblings. Always find out straightaway what information the family has and their attitude to it, as this will affect all your efforts to help them.

(2) After diagnosis it may be helpful to see the spouse or caring relative of the patient, whether or not the diagnosis has been shared with the patient. Often a spouse has been too anxious—or has not had the opportunity—to discuss the illness with the consultant and may be suffering undue anxiety from misconceptions or uncertainties about the future.

(3) This interview will give you the opportunity to assess the need for referral to district nurse, social worker, or occupational therapist. If help is not needed immediately, you should reassure

the caring relative that he or she will not be left to cope alone with a chronic, degenerative illness. If appropriate, you could also tell the family about the Association to Combat Huntington's Chorea, through which they could meet other families and receive regular newsletters.

(4) Reactions in the patient who knows his or her condition may vary from unrealistic expectations of a cure to deep depression and temptation to commit suicide. While avoiding false reassurance it is important to explain carefully and positively the treatment that is available and never to give the impression that nothing can be done to help. Include discussion of different types of occupation, if appropriate—for example, day centres. It should be remembered that worldwide research is being undertaken into this disease.

(5) Choreic movements are embarrassing both to the patient and to the observer, who often mistakes them for drunkenness. When the family is willing for the diagnosis to be known, it is helpful to encourage them to share with friends and neighbours the fact that this is an illness. Equally, your receptionist should know. On the other hand, where the family does not wish the diagnosis known, it is important to be very careful how information is given and recorded.

(6) The patient may have slurred speech. Encourage him or her to take time and speak slowly. Patients are often frustrated in their attempts to communicate and may give up or react aggressively if they are not given time and attention. It is always safer to assume good awareness even when patients cease to speak.

(7) Young people at risk may sometimes need a medical reference for a job or life insurance. If you are obliged to disclose the diagnosis of Huntington's chorea in one of the parents, always discuss this fully with the person at risk because he or she may not realise that it might prevent getting some types of job or a mortgage. This also gives you a chance to find out exactly what information the young person has and to refer him or her for genetic counselling if necessary.

(8) Avoid giving advice about whether or not to have children. A doctor who has time and a good relationship with a family may have the best opportunity to open up this discussion with young people and to begin to help them clarify their feelings in the light of accurate knowledge of the illness and their own degree of risk. But people must ultimately make the best decision for themselves. If a young person is unduly anxious or undecided referral for specialised help may be more appropriate. Neither adoption nor long-term fostering is likely to be a solution available to families at risk to Huntington's chorea so be careful not to give false reassurance. Artificial insemination by a donor, if appropriate, needs very careful consideration.

(9) Examination of brain tissue after death can confirm a diagnosis of Huntington's chorea; this is particularly important where there has been some doubt as to the diagnosis. Additionally, brain tissue is used for research purposes to help establish the underlying biochemical disturbance. Relatives should be encouraged to agree to donate the brain tissue of patients with

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Huntington's chorea, or those thought to have the disease, by contacting the MRC Brain Tissue Bank, Department of Neurological Surgery and Neurology, Addenbrooke's Hospital, Hills Road, Cambridge CB2 2QQ (telephone: Cambridge 40011). Details should be sent to the brain tissue bank while the patient is still living, and arrangements will be made by them for tissue removal after death.

(10) Families caring for someone with Huntington's chorea can carry on in an amazing way if the strain on them is recognised and strenuous efforts made to give them a break from time to time. A doctor can help by using his influence to obtain a holiday or short-term admission to hospital. The Association to Combat Huntington's Chorea runs a holiday home near Epping, Essex, which is open all the year round.

MATERIA NON MEDICA

No smoke without fire?

Blowing my modest savings on an "investment property" has been, retrospectively, my only stroke of financial genius—for the capital appreciation, not the income. Perhaps it could work only in Cambridge, where there is such a need for rented accommodation and where, with reasonable luck and judgment, the tenants will be nice—much nicer than my generation was in our early 20s. The image being projected is not so much a rapacious landlord, more a social service. And the weekend tasks prevent one becoming too pompous, especially as they are so much more menial than those undertaken by the landlady's army of presumptive black economists (nothing ethnic, you understand) who do the skilled work—the carpentry, glazing, plumbing, and electrics—with the job satisfaction that goes with it.

As with all life styles, there is an element of stress, encapsulated by the law of natural cussedness or sod's law. This entails that each minor procedure shall escalate to the proportions of heroic surgery. Example: electric socket by the bed becomes wobbly. Examination reveals anterior plate of box has become subluxed. Diagnosis—avulsion of screw due to degenerative changes in plastic. No, the face plate cannot be purchased separately from entire socket—price almost £5. No, the pattern I have brought along is no longer in production. Purchase new type box, return to premises. Remove face plate of new box to attach to existing. Screw holes in wrong place, necessitating replacement of entire box. Remove existing box to replace by new box. Screw holes in wall in wrong place for new box. *Almost* in right place but not quite—not far enough away, though, to make totally new holes. Fill existing and re-drill. Switch on electric supply to drill. Forgot to ensure protruding wires not touching, inevitable consequence. . . . Tenant charming academic, but no fuse wire. . . . Cycle back to DIY shop: Flanders and Swan said it all.

The other major source of stress is the neighbour who observes and complains about each door closure, each coming and going, each disc played louder than 1 decibel by the long-suffering tenants, who accept him as a part of life's rich pattern. But the neighbour's researches show that where the second floor of a property is occupied by a tenant certain fire regulations must be complied with. A series of visits by the local authority Gestapo at 7.30 am (our rates are high) reveal that the notice on the attic door "For storage only" is being ignored. Smoke detectors are eventually installed, costing many hundreds of pounds. They go off in the middle of the night, causing aforementioned neighbour and family to vacate their home hastily in their nightclothes. They go off whenever anyone cooks dinner. Tenant telephones. What shall she do? Switch off the smoke detectors. But would that not be defeating the object? Not at all, I assure her. The object is to comply with the fire regulations. It has nothing to do with fire at all. It has a great deal to do with nosy neighbours, excess bureaucracy, an overpaternalistic state, politics, jobs for the boys, but nothing to do with safety. Her house, which is occupied by young healthy adults, is at no greater risk than mine, where I happily accommodated my much-loved family when they were tiny children, unmolested by the council means-of-escape officer. Protection against fire cannot be enforced through oppressive

legislation but is more effectively ensured through education, as anyone who has experienced the shattering impact of the excellent Australian film *Hospitals Don't Burn, Do They?* can testify.—NICHOLAS CONT (consultant geriatrician, Cambridge).

Stede Bonnet, Pirate of Barbados

Within every schoolboy must lurk a sense of awe and admiration for those legendary sea rovers. No one loves today's hijackers with guns and bombs at 30 000 feet, but the romantic pirates of the past seem to have escaped all moral considerations. The evil and terrifying Blackbeard was pure legend. Henry Morgan the buccaneer was white-washed, made a baronet and Vice-Governor of Jamaica. And who, brought up on English history, does not regard the privateers like Raleigh and Drake as heroes?

It came as a surprise a year ago to learn that my home once belonged to a pirate. It has made me an avid piratologist, with special reference to a lesser pirate of Blackbeard's day, Stede Bonnet of Barbados.

Major Stede Bonnet was apparently an army officer who became a planter in Barbados at the end of the seventeenth century. The rest of his life story makes an interesting case study in psychopathology. He was reputed to be so disenchanted with his wife that he bought a ship in Bridgetown, hired a crew of 70, and set sail as a pirate—perhaps the only pirate to buy rather than seize his ship. Contemporary accounts suggest that he had a mental disorder and had been acting strangely for sometime. A London cartoon lampooned him as leaving home hastily under a shower of plates. Had his domestic turbulence made him mad? He sailed off the Carolinas and pillaged merchantmen until he met up with Edward Teach, alias Blackbeard. Teach was clearly his hero (both men's ships were named *Vengeance*). Bonnet teamed up with Teach, either to bask in his glory or to learn the trade from its undoubted master. But Teach despised him and he was so humiliated he went his own way again. Yet such was his admiration for Teach that he promptly copied his ruse of seeking pardon from the Governor of North Carolina, to give the pretence of legality to his piracy. While he was on shore negotiating, Teach abducted his ship's crew and pillaged his ship. A faithful few escaped and enabled him to rescue the others. He then had a second and much more successful spell of piracy before being captured by the authorities.

His last days were a series of crazy paradoxes. He engaged the sloops sent to capture him, then surrendered unexpectedly, and went to prison. He escaped and when attacked again fought fiercely but on losing his second-in-command he was demoralised and surrendered again. In court he defended himself and claimed that necessity drove him to piracy. He and his men were condemned to be hanged.

He appealed pitifully to the king: "I employ you to spare my life, even if I have to cut off my arms and legs to obtain my pardon, leaving me only the use of my tongue to pray and make penance and live clad in sack cloth and ashes." Despite his appeal he was hanged, on 18 November 1718. His last request was granted: to die with a bunch of flowers in his hands. He died as he had lived—a strange and almost pitiful figure, yet still described and sung of as a daring pirate.

An old house is full of unexplained noises, and as I sit in my study alone on a windy night it takes little effort to imagine Bonnet and Blackbeard stalking the decks above.—HENRY S FRASER (consultant physician, Bridgetown, Barbados).