



## HUNTINGTON'S DISEASE ASSOCIATION NORTHERN IRELAND

Families at the heart of all that we do

## Behavioural Problems in Huntington's Disease

### Introduction

Behavioural changes are a characteristic feature of Huntington's disease which, although less obvious to the casual observer than the involuntary movements, represent for many families the most distressing aspect of the condition. Some alterations in the person with Huntington's disease such as frustration, irritability and loss of confidence may occur to some extent as an understandable reaction to the presence of a disabling illness and to the loss of personal independence that it entails. However, behavioural changes also occur as an integral part of the disorder arising as a direct result of changes that take place in the brain. Some of these changes arise as a direct result of alterations in the way a person thinks (intellectual changes); others represent alterations in a person's mood.

Just as the nature and severity of the movement disorder varies from one person with Huntington's disease to another, so too do behavioural symptoms. In some people alterations in behaviour are mild without major social impact. However, in others they can be profound leading to disruption of family life and breakdown in social relationships, and placing a severe burden on carers. There are no easy solutions to difficult behaviour – precisely because the behaviour arises as part of the illness and is not under the patient's control. Nevertheless, understanding the behaviour can help: it can lessen the burden and provide clues to the most appropriate methods of management.

### Drive and Initiative

The person with Huntington's disease may show a loss of drive and initiative. If left to their own devices they may do nothing, stay in bed or spend the day watching television. This behaviour can be highly frustrating for family members, who may perceive the behaviour as “laziness” or the patient as “not pulling his or her weight”. It can be a great source of family conflict when the patient's partner is under stress from multiple responsibilities – for example, acting as breadwinner, caring for a young family, carrying out domestic chores. It is important to recognize that the patient's behaviour is not due to laziness. Huntington's disease affects the part of the brain (the deep part of the brain or subcortex and its connections to the regions at the front of the brain) which are crucial for drive and initiative. These are the parts of the brain that allow us to think ahead, to make plans, to generate the actions appropriate to those plans, and to persevere on tasks until goals are achieved. When those parts of the brain are damaged, the drive or foresight to self-initiate activity is severely compromised, even though the skill to carry out the activity is still retained. The patient may potentially be capable of carrying out a task, yet is unable to generate the motivation or initiative to embark on it. Arguing with the patient will not overcome his or her difficulty. Encouraging joint participation in activities, e.g. helping with the washing up, may be helpful, and is generally more successful than assigning the patient solitary pursuits. This is because the other participants in an activity act as an external stimulator or motivator. The patient does not have to rely on his or her own drive and initiative. Participation is important. Not only does it help to keep the patient active, it also helps to provide a sense of worth – that the patient is able to make a constructive contribution.

### Mental Flexibility

People with Huntington's disease may sometimes seem rather inflexible and mentally rigid. They may adhere to set behaviour patterns or routines and appear unwilling to adapt to new situations or altered circumstances. They are not being deliberately obstinate. The brain changes that occur in Huntington's disease can impair the ability to think flexibly and to adapt easily to novel situations. Patients generally feel most comfortable and confident in highly familiar situations, involving a fixed routine. If the patients appear to need routine, then it is worthwhile trying to accommodate this; it does not mean that variety or a stimulating environment is inevitably sacrificed, it is simply that a structure is imposed on the patients' day so that he or she knows what will happen and when.

## Doing two tasks at once

Many everyday situations involve people doing more than one activity at a time, e.g. answering the children's questions while cooking or watching the television while doing the ironing. Doing two things at once requires the mental flexibility to switch attention rapidly between tasks – however engrossed in the television programme one may be, it is essential to keep switching attention back to the ironing to avoid burning the clothes. Huntington's disease can impair rapid switching of attention, making it difficult for the person with HD to carry out two tasks at once effectively. In contrast many people with Huntington's disease are very good at sustaining attention on a single task, provided they are not distracted. "One thing at a time" is a good general rule. Overloading what the patient can cope with at one time may be one source of patients' irritability and disruptive behaviour.

It is worth remembering that a physical activity such as walking, requires much more conscious attention for the person with Huntington's disease than for other people. Someone with Huntington's disease may find it difficult to carry on a conversation at the same time as walking and may sometimes be noted to stop walking before answering a question. This is because both activities require conscious attention. To carry out both simultaneously would require rapid switching of attention from one to the other. Someone with Huntington's disease who has difficulty rapidly switching attention is obliged to stop one activity before embarking on the other.

## Quality of Performance

It is not uncommon for people with Huntington's disease to carry out everyday tasks less efficiently than before. For example, in writing a letter, a patient may miss out words; in washing dishes, plates may not be properly cleaned. Indeed, for many people with the illness reduced efficiency is the precipitant of medical retirement from work. The poorer quality of performance on tasks may be a source of irritation to the person's family who may perceive the person as being 'slapdash' or 'not bothered'. It is not the case that the person is simply not trying. In fact many people with HD put an enormous amount of effort into their activities. The errors arise as a result of the changes that take place in the brain. The person with Huntington's disease does not forget how to do a task. What becomes impaired is the ability to self-motivate and check the results of one's own performance. The patient is often not aware of errors that are apparent to others. Encouraging the person to carry out tasks is a good thing. However, it is worth being aware of – and trying to accommodate – the person's possible limitations. In the case of the person with Huntington's disease who lacks initiative, it falls on other family members to act as a 'stimulator to action'. So too, in the case of a person with HD who carries out tasks inefficiently, 'checking' procedures are dependent upon others.

## Hygiene and Self-Care

It is quite common for people with Huntington's disease to show less interest in their personal appearance and their standards of hygiene and self-care decline. Loss of drive and initiative undoubtedly contributes to this change. Also of relevance is that Huntington's disease can impair personal and social awareness and blunt emotions. The person is likely to be unaware of the change in him/herself and insensitive to the effect that an unkempt appearance has on others. Moreover, he or she may not experience the feelings of shame or embarrassment that under normal circumstances act as a strong motivator to self-care. The person may need to be prompted to bathe or change clothes. A prompt often suffices. However, some people with HD still adamantly refuse to wash or change their clothes. It is worthwhile to try to establish bathing and clothing changes as part of a fixed routine – for example occurring at a specific time on specific days. It is also worth considering whether certain conditions influence the persons' level of cooperation. The manner in which the prompt is given may be relevant. A person may react badly to being told what to do, yet respond positively when he or she is encouraged to participate in making decisions. People with HD who refuse, for example, to put on the clean clothes given to them, may be willing to put on clothes that they themselves help to select. The person with HD who participate in making decisions are less likely to be behaviourally disruptive than those from whom all choices have been taken.

## Disinhibition

Some people with Huntington's disease may act in a disinhibited way that is embarrassing to others. Disinhibited behaviour may take a variety of forms. People may act impulsively or rashly without thought, such as making a sudden purchase of a car that they cannot afford. They may make socially inappropriate remarks, for example making personal comments about a person who is within earshot. They may behave in a sexually disinhibited way, such as making sexual advances to a partner in front of the children. Such behaviour results from a breakdown in patients' social awareness and ability to think through and appreciate the social consequences of actions. They do not see the repercussions of actions. The person with Huntington's disease may also no longer experience so acutely the feelings of embarrassment, guilt and shame that under normal circumstances place constraints on social behaviour. People with HD cannot simply be "made to see" the consequences of their actions if the capacity to do so has been damaged by the disease process; nor can they be made to feel guilt, shame or embarrassment if those emotions have been taken from them by the disease. Disinhibited behaviour may have the inevitable and unfortunate consequence that it leads to a restriction in a person's freedom: for example, a partner being obliged to take control of family finances. Some disinhibited behaviour, such as socially inappropriate sexual advances, are best managed by imposing limits, by letting the person know what is acceptable and what is not, and as far as possible adhering to those "rules".

## Sympathy and Empathy

People with Huntington's disease may sometimes seem self-centred, uncaring and thoughtless. The person's apparent disregard for the emotional needs of a partner can be particularly hurtful, and is especially poignant when it contrasts with a former loving and caring relationship. The natural tendency is for a partner to feel personally slighted. It is important to emphasize that the person with HD is not being deliberately awkward, wilful or unkind. Apparent self-centredness is in part a consequence of the loss of mental flexibility associated with Huntington's disease: the person may no longer be able to put themselves in another person's shoes, to see another's point of view, to weigh up all sides of an argument. They may genuinely fail to see how their remarks or actions affect others. Moreover, Huntington's disease can impair the ability to experience the complex range of subtle emotions that contribute to interpersonal relationships, so that persons' emotions are more shallow or 'blunted'. The adverse effect of Huntington's disease on the person's capacity for sympathy and empathy with others is a major reason that Huntington's disease can have such a devastating effect on families. Relationships that ought to be mutual may seem one-sided. There are no magical remedies: it is not possible to put back emotions and feelings that have been lost by disease. But remember it is the disease that is at fault. The person is not being deliberately uncaring. The emotional changes are not under his or her control.

## Depression

Depression is a relatively common problem in Huntington's disease, although by no means all people are affected. Its recognition is important because its effects can be profound, yet it can be effectively treated resulting in dramatic improvements in persons' well-being and ability to function efficiently. The loss of drive and initiative, which is an integral part of the mental changes that occur in Huntington's disease, does not necessarily indicate a depressed mood. Nevertheless, the possibility that it represents a symptom of depression should always be considered, particularly if the change in patients' level of motivation and interest has occurred relatively rapidly. Do not hesitate to seek medical advice if depression is suspected.

## Irritability and Aggression

Although some people with Huntington's disease may be even-tempered throughout the disease course, it is not uncommon for people with HD to become emotionally volatile. They may 'flare up' for no apparent reason, or over trivial issues. People with HD may experience a feeling of internal agitation and be aware that they are easily 'worked up', yet the sudden surge of anger often comes without warning and is outside the persons' control. In these circumstances it is best to avoid confrontation, which will tend only to add fuel to the fire. If necessary leave the room, particularly if there is a threat of physical aggression. There is no foolproof solution to prevent emotional outbursts. However, it is worthwhile considering whether there are specific precipitating factors that can be avoided. Experience may suggest a range of everyday situations which 'work up' the person. These might appear quite trivial – for example, someone switching over television channels while the person is watching a programme. Some people may become irritable whenever they do not get their own way or their own views are opposed.

Remember that people with HD, by virtue of the changes that take place in their thinking, may have difficulty seeing another's point of view. Continually arguing a point is unlikely to convince the person – it is more likely to increase his or her emotional agitation. Another point is worth reiterating: The person with Huntington's disease find it more difficult than other people to do two things at once. Overloading what the person can cope with at any one time may provoke in the patient feelings of agitation and potential loss of temper. Avoid whenever possible placing multiple simultaneous demands on the person. One thing at a time is best. Symptoms of irritability and aggression, like depression, can be treated medically. You should be conscious of the safety of yourself, the person with HD and anyone else involved. Do not take simply 'put up' with a risky situation or allow anyone to be in danger as a result of HD symptoms. If you are concerned do consult your doctor for advice.

## Denial of illness

The onset of Huntington's disease inevitably leads to life changes, which require accommodation and adjustment from both the person with HD and their families. The process of adjustment is made difficult if the person with HD refuses to accept that there is anything wrong. To an external observer, the involuntary movements may be obvious, and it may be evident that the person no longer carries out occupational and domestic tasks as efficiently as before. It is easy to assume therefore that these changes must be equally apparent to the person with HD. The conventional interpretation of lack of acceptance of illness is that the person is 'in denial', that the patient at a subconscious level is aware of the reality that he or she refuses consciously to acknowledge. Such an interpretation may be an oversimplification. Research suggests that people with Huntington's disease may not have normal experience of their involuntary movements: if persons' own direct physical experience does not 'match' with their perception of the illness in others, it is perhaps not surprising that they feel that they do not have that same illness.

There are additional factors that may contribute to none acceptance. The disease itself can impair the ability to self-monitor and to reflect on one's own performance: people with HD may genuinely be unaware of mistakes that are evident to others. It can impair too, the ability to draw inferences: a person may be aware of clumsiness or forgetfulness, yet fail to see the implications of these symptoms for Huntington's disease. Refusal to accept illness is not simply a result of obstinacy on the part of the person – it is a feature that occurs in some (not all) people with HD as a consequence of the disease process itself. Carers and professionals may need to accept that confronting the person with a diagnosis of Huntington's disease will not always induce immediate acceptance. An approach that focuses on specific symptoms rather than diagnosis can sometimes be helpful (for example, a suggestion that the person is 'a bit clumsy and coordination is not so good' may be accepted more readily as a reason for curtailing driving than 'having Huntington's disease'). Most people do come to accept the diagnosis given time.

## Behaviour and Disease Progression

Certain aspects of behavioural change become more pronounced with disease progression. For example, a person typically show less and less initiative over the course of the disease; they show progressively less concern over their own appearance; they become systematically less aware of the feelings of others. However, there is not an inevitable association between the length of time that the person has been ill and the severity of behavioural disturbance. Indeed, some behaviours may become more manageable as the disease becomes more advanced: for example, irritability and aggression may gradually give way to apathy and unconcern. Similarly, disinhibited behaviour may be most pronounced early in the disease when the person is most active, and diminishes and becomes less of a problem later as the person loses drive and initiative. Mood disturbances such as depression tend to occur sporadically and are unrelated to the duration, severity or progression of the disease. Behavioural Problems: action and reaction There is no doubt that the behavioural changes described above are an integral part of the condition, arising as a direct consequence of physical changes that take place in the brain. However, the person with HD has to contend with a whole host of obstacles as a consequence of the disease: loss of a job, reduced independence, impoverished social life, impaired mobility, lack of understanding and tolerance from others. It is not surprising that person should feel frustrated and even angry with the apparent injustice. The persons' behaviour often represents an interaction between the direct effects of the disease and the reaction to its consequences: a person, for example, who assaults someone who falsely accuses him of being drunk, is showing an understandable feeling of anger at being victimised and falsely accused (reaction to consequence of illness), combined with the lack of emotional control resulting from Huntington's disease itself (effect of disease).

## Understanding Huntington's Disease

Huntington's disease is a disabling condition that creates significant difficulties for the person and their family. Nevertheless, the traditional notion of the 'mental' changes associated with Huntington's disease as 'global' are both misleading and inaccurate. Huntington's disease damages selective parts of the brain, leading to specific difficulties in thinking, which in turn give rise to specific and predictable changes in behaviour. Some aspects of persons' mental or 'intellectual' functioning remain well preserved even when the condition is advanced: people can, for example, see and hear, and understand the meaning of what they see and hear. The task for carers and professionals working with Huntington's disease is both to recognize persons' abilities so these can be harnessed most effectively, and to understand persons' limitations, so that these can be compensated for. Behavioural changes in Huntington's disease represent the greatest challenge. The profound impact of disordered behaviour on families is at last being recognized by professionals. Researchers into new treatments for Huntington's disease recognize that improving mobility and reducing involuntary movements are not enough. Treatments must also be directed at improving behavioural aspects of the condition.

## Contacts

You can speak to our support workers about any questions you have in relation to behaviour and mood.

Any changes in symptoms should be discussed with your GP who may advise a referral to a neuropsychiatrist specialising in medications for HD or to the community mental health team. Medication review is extremely important with HD and you should note any side effects or issues with medication and see your GP regularly.

## Support for anyone affected by HD

HDANI run monthly **support groups** with counsellors which provide an opportunity to share experiences and vent in safe space.

You can also speak to HDANI about arranging one to one counselling and we do this either through our own counsellors if there is availability in your area or via GP or community counselling organisations where we will provide information and training about HD to help improve the counsellors understanding of your situation.

There are also courses provided free of charge throughout the year in Northern Ireland in the areas of Mental Health First Aid, Managing Aggression and other relevant areas. Contact HDANI for further information.

## Support for Carers

### CAUSE

Founded in 1995, CAUSE is a unique peer-led regional charity offering services to families, partners and friends across Northern Ireland caring for a loved one who has experienced serious mental illness. CAUSE is run by carers for carers which gives us an organisation a true peer-led ethos and belief in the power of connecting with other people who have shared similar experiences of caring.

Helpline: 0845 603 0291

### Women's Aid

Offer specialist services for women, children & young people who have experienced domestic violence.

Helpline: 0808 802 1414

### Men's Advice Line

MAL are a team of skilled professionals offering practical advice, information and emotional support to male victims of domestic violence, as well as to concerned friends and family and frontline workers.

Helpline: 0808 801 0327