

Huntington's New South Wales



Living with Someone who has Huntington Disease

Supported by NSW Health

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This fact sheet looks at the types of difficulties experienced by people affected by Huntington Disease. It looks at the symptoms and behaviours from the perspective of someone diagnosed with HD and someone caring for a person with HD. It then discusses what causes the behaviours and how to manage them.

A person diagnosed with Huntington Disease may describe the following types of difficulties:

Memory

People: I don't remember what people look like after I meet them. When I fell at the mall a while ago, a lady sat with me until I could stand up (at least 10 minutes). We talked and then I walked over to the washroom to see if I could walk. When I came back, I looked for her, but had no idea what she looked like.

My last job was a temporary one. It lasted five weeks. I never remembered the names of any of the six staff who worked there, or their jobs, or their faces.

I have seen my social worker five or six times over the last year or so. I don't remember what she looks like.

Tasks: I forget when I've done things. I worked over six hours one afternoon on a particular task. Next morning, when asked about it, I said I hadn't done it yet, but I'd get right to it.

I don't remember when I've watered the plants, got cleaned up, or filled the humidifier. I don't always remember to turn off the appliances in the kitchen.

Conversations: I don't always have a problem understanding - I just don't always remember a lot of what is said. I've made lists for years, but now I forget to look at the lists. I feel like a computer and someone is forgetting to press my 'save' button. I don't retain a lot of what I hear, or read.

Events: I forget things that are happening or have happened. My daughter got married last year and other than when someone

asked about it, I didn't think about it. I missed the excitement, because it wasn't with me all the time.

When I was asked for directions to the house where they were married, I couldn't remember without looking at the map, even though I knew where it was. I never remember the name of the house where the wedding took place, or the hotel where the reception was held.

My sister died last year. I forget unless someone else brings it up.

Sense of Time

I don't have a sense of time. When I look at my watch, I immediately forget what time it said. I used to be able to judge how long 15 minutes or half an hour was. Now it means nothing. I get up in the morning and before I know it, it's time to go to bed -time flies! I used to remember bus schedules but now I don't.

Speech

I don't always know, when I start a sentence, what the right thought is to finish it, even when I really know what I'm talking about. I have trouble enunciating sometimes. There are times when I really have to concentrate to get a word out and pronounce it properly. I often trip over words.

Confusion

Options throw me. I've always been a solitary sort who could take in a lot of information, sift through it and quickly make decisions that were sensible and comfortable to live with. Now, if I'm given a choice of two simple things, I'm slow to decide and not certain with my choice.

I was always a good navigator because I like maps and have a good sense of where things are. I could picture where to go when given directions. Now, I don't picture anything. I just get confused.

I've always been able to mentally keep track of what I'm spending in a grocery store. I could be within a dollar or two of the total when I checked out. Now, even if I only have three items, I not only get

confused when adding, I forget what my answer is, so I add things over and over.

Physically

I've fallen four or five times in the last couple of years. When I rush I stumble. I drop things. I've broken a dozen wine glasses. I break or chip dishes.

I have cut myself a couple of times, even though my 'sensible brain' told me my finger was in the way. I've stood in front of the cupboard door and opened it into my face. I've stuck my finger in to a pan full of hot stuff to see how hot it was.

I have no sense of rhythm anymore. I can't dance.

My printing is not great, but it is easier to read than my writing.

I fidget.

Social Skills

In social situations I'm slow to see when I should offer help. When friends visit with a problem, I don't always realise I could help until after they've gone. I never think to pick up the phone to talk to friends or family.

A person caring for someone with Huntington Disease may describe the following types of difficulties:

Mood Changes

Mood changes are common and often unpredictable. My partner has periods of feeling very "down" and becomes irritable and withdrawn.

Sometimes my partner becomes agitated and over-excited. He becomes overactive and restless.

Loss of Inhibitions

My partner seems unable to control his behaviour and does things in public that I find embarrassing.

My partner is extremely demanding and wants things done immediately. He cannot seem to wait, even when I am clearly in the middle of something else.

Recently, my partner has become angry and aggressive. This is unpredictable and frightening.

My partner has suddenly become extremely sexually demanding. I find this very difficult, as I am not always in the mood to be intimate.

Rigidity of Behaviour

My partner has become so stubborn.

My partner does not cope with change very well. It appears to distress and confuse him.

Lack of Insight and Denial

My partner used to be loving and gentle. He no longer seems to understand when I am upset or distressed.

My partner refuses to accept that he is no longer able to do things around the house anymore.

My partner refuses to seek help or see a doctor- he doesn't think anything is wrong

Self Monitoring and Awareness

My partner does not seem to realise how obvious his movements are. He denies that they are a problem.

My partner makes errors when completing tasks and doesn't seem to notice. He used to be so fastidious and yet now seems to make careless mistakes.

Guilt

My partner is consumed by guilt about the risk that he has passed on HD to his children.

Initiative and Drive

My partner used to take an active role in helping to run the household. Now he seems content doing nothing all day and leaving everything up to me. It's like he doesn't even notice that the house is a mess and he could help me by doing some tidying up or washing.

Thinking Ahead

My partner is so focused on his own needs that it doesn't seem to matter about anyone else. If he decides he wants something, then he wants it immediately.

Organisation and Planning

My partner can't seem to organise himself to complete tasks. He seems to tackle tasks in a disorganised way and takes such a long time. Even a simple task like getting himself dressed seems to be difficult for him.

Attention

My partner seems to have difficulty doing two things at once. He can't have a discussion with me if he is doing something at the same time. At first I thought he was ignoring me, but he seems to have to focus all his attention on one thing at a time.

What is Causing these Behaviours?

The types of behaviours described previously are all possible presentations associated with HD.

It is important to recognise that the person with HD experiences emotional reactions to the disease as well as symptoms that are a direct result of irreversible changes to the brain that occur as part of the disease process.

The impact of changes to the brain (organic changes) and the emotional response to these changes (reactive features) may be affected by a number of factors:

- The extent of organic change

The extent of organic change in the brain will impact on the individual's ability to perform daily tasks, as described earlier in the fact sheet. If there is a high degree of intellectual impairment, the person with HD may lose insight into his or her own feelings and behaviour. They may also lack the understanding of the effect that behaviour has on other people.

- The pre-symptomatic personality

Personality changes are frequently a symptom of HD, but in some cases, the pre-symptomatic personality is emphasised and certainly in some way influences the reaction to HD diagnosis and onset of symptoms. Therefore, someone who was always active may find it very difficult to accept physical dependence, while

someone who has enjoyed intellectual pursuits may find their impaired memory and concentration very frustrating.

- Previous experience of Huntington Disease

People have differing perceptions of HD according to their past experience, and this is bound to affect their attitudes to their own diagnosis. Some family studies have shown that there are likely to be more difficulties where there has been a very disturbed family background. Also, a history of open discussion about HD and its consequences can lead to a better adjustment and willingness to continue to communicate.

- Support and counselling provided by family, friends and professionals.

The availability of support from family, friends and professionals can have significant impact on the cognitive and emotional changes associated with HD.

Managing the Difficulties Associated with Huntington Disease

Some of the practical difficulties associated with Huntington Disease can be managed by creative and flexible thinking. These strategies include ideas for assisting with memory (ie writing notes and placing them in places that are easily visible such as the fridge, keeping a diary/calendar), confusion (ie having a weekly schedule of tasks and activities to ensure routine), sense of time (ie using a timer when cooking).

These types of solutions are often most useful early on in the disease process and require insight and acceptance on behalf of the person with HD.

Although these problems can be very distressing for the person with HD, the family and carers, there are strategies that can assist with alleviating the problem.

In general, behavioural difficulties are much more of a problem, frequently causing family disruption or breakdown and posing dilemmas as to appropriate respite or long term care.

Managing Difficult Behaviours

There are no overall answers in dealing with behavioural difficulties and no one method that will deal with a particular problem in all people with HD, however, when developing management strategies, there are some key points to remember:

1. There are organic changes in the brain which cause a disturbance of the intellect and personality, resulting in difficulty managing daily tasks as well as changes in behaviour - *the person with HD has no control over this and it has to be accepted and accommodated.*
2. The person with HD has emotional reactions to the consequences of HD, real or feared, which increase the behavioural problems. These can be helped by understanding and foreseeing situations which will give rise to difficulties.
3. However much love, guilt and understanding the carer may feel, there are some aspects of behaviour that are unacceptable. The carer or family member must be clear with themselves and the person with HD as to what these are.

Given that many of the behaviours of people with HD are an interaction between organic changes to the brain and reactive responses, then both need to be considered when trying to manage the difficulties.

Support and Counselling

The time of diagnosis is a critical period for people affected by HD and their families, with many emotional adjustments having to be made. However, instead of sharing their feelings, many people with HD and their families, withdraw into themselves both because they find it painful to face the present and future, and because they do not want to add to the troubles of the others. Non-communication can become part of family functioning, and this feeds the fears of people

with HD and increases their feelings of isolation. Discussion within the family and/or with a professional counsellor at this stage is very important to deal with negative fantasies and immediate emotional and practical adjustment. If this openness can continue, the particular difficulties of each new stage can be discussed and coped with.

It is obviously very difficult to do this when there is strong denial or general lack of insight, and professional help may well be refused because the person will not admit to having a problem at all. Denial has the useful function of keeping depression at bay, but it becomes dysfunctional when it is so complete that it causes danger (eg. driving) or breakdown in support (eg. family can't cope). If the person will accept it, counselling geared to examining the person's feelings about their past experiences of HD may prove the way to slowly uncover their understanding of the current situation. On a daily management basis, denial is probably not best coped with by direct confrontation such as insisting that the person admits that they are ill, but rather by focusing on the consequences of their actions for others. Lack of insight may make this very difficult, but for instance, a person who is angry at having lost their job may be brought to see that the work was not being done to the employer's satisfaction, whatever the reason for that. There is a fine line to be drawn between this approach, and actual collusion in denying the diagnosis. Collusion is, in the long term, very unhelpful as it closes the door on the opportunity for the person to come to terms with their situation and inhibits close relationships.

Diet

It has been noted in a few cases that the general mood of a person with HD has improved when there has been an increase in food intake. People with HD can need up to 5,000 calories a day, and if they are not eating sufficiently then they will be hungry, not feel well, and therefore be difficult. An increase in the number of meals rather than the amount at anyone time may be helpful (*Fact Sheet "Guidelines for Meeting the Nutritional Needs of People with Huntington Disease"* is available from the Association).

Adapting to the changes

Organic changes lead many people with HD to become inflexible and obsessive so that routines such as meal times, going to the lavatory etc., become entrenched, and they become agitated if there are many changes. Attempts to alter this behaviour are unlikely to be successful, and where possible it is probably better to adapt to the sufferer, since rational argument won't have much effect. Obsessions and rigid thought processes or behaviour may be irritating for the carer to deal with, but do provide the person with HD with a sense of security and also, perhaps, an alternative to expressing their feelings in other ways.

Avoiding frustration

Some frustration is inevitable, as the person with HD gradually becomes unable to function in the same manner as they did before. However, many situations which seem to spark off aggressive or generally difficult behaviour can be avoided. An awareness of the person's capabilities is very important, so that they are encouraged to be as independent as possible and allowed to take risks without causing them constant exposure to failure. Close attention should be paid to the signals, verbal or non-verbal, that the person with HD is upset or wanting something, so that they do not have to get to the stage of making a fuss before they receive attention. The frequent inability to tolerate delay or refusal, plus any communication difficulties, can make efforts to avoid frustration quite exhausting if not sometimes impossible. However, knowledge of the person and sensitivity to their needs means that some situations can be anticipated and potential frustration defused. It may be possible to identify 'trigger' circumstances, and avoid these, or have a stock of diversional activities.

Imposing Limits

The previous comments have stressed the need to fit in with the person with HD where possible, but there are circumstances where this is not possible or appropriate.

Violence and sexual harassment cannot be tolerated, and other family members also have a right to a reasonable quality of life and a need to have time to themselves. It is very important to take the needs of children into account, and sometimes difficult decisions have to be taken when the behaviour of the person with HD is in opposition to the physical or emotional health of the children. Personal experience of some carers has been that they were surprised that when they felt really strongly about something and were very firm about their needs, then the person with HD was able to accept the limits set, and understand the reasons behind them. It is not easy to achieve this as a person with HD may well perceive any ambivalence in the carer who may by this stage be feeling exhausted and unsure how justified they are in asserting their own needs. Nevertheless, it is advisable for a carer to be clear about what is permissible behaviour, and what is not, for their own sakes, but also for the person with HD whose behaviour may otherwise precipitate crises and possibly even an admission to residential care which might have been avoided.

Medication

There are medications that can be effective for managing mood disturbance as well as excessive agitation and restlessness. There is also medication to assist with management of the chorea (involuntary movements).

However, these medications may have side effects. It is important to discuss the options with you GP or Specialist.

Severe Psychiatric Disturbance

In a minority of cases the behavioural problems will be of an order to require psychiatric treatment. Symptoms may be violence, extreme paranoia or delusions. It is unlikely that such behaviour will be sufficiently modified by any of the previously described methods, and involvement from psychiatric services may be required.

Assistance with the Management of Difficult Behaviour

Not only will the behavioural symptoms of people with HD vary enormously from one individual to another, but so will the reactions of carers, family or professionals. What is experienced as intolerable for one person may not be a problem for another. The carer may well benefit from some professional help to sort out what behaviours of their family member may be amenable to change and what may not. It is also useful to determine how flexible they are able to be and what is the base line beyond which they can be firm that the behaviour is unacceptable. Both sides need to have realistic expectations of each other and of themselves, but unresolved feelings of guilt, anger or grief may impede clear communication.

The NSW Huntington Disease Service which is funded by the NSW Department of Health has a team of health professionals who are experienced in all aspects of the effects of Huntington Disease on the individual and the family. They have close links with other health and welfare workers who may be involved with the family and they are able to put people affected by HD in contact with various support systems within their own community.

Fact Sheets

- Huntington's Disease in the Family – A Booklet for Young Children
- Huntington's Disease – A Fact Sheet for Teenagers
- Talking to Children about Huntington's Disease
- Presymptomatic Testing – The Consumer Experience
- Understanding Challenging Behaviour in Huntington's Disease
- Living with Someone who has Huntington Disease
- Making a Decision About Residential Care
- Guidelines for Meeting the Nutritional Needs of People with Huntington's Disease
- Eating and Swallowing Difficulties in Huntington Disease
- The Importance of Dental Care in Huntington Disease
- Communication and Huntington Disease

*These Fact Sheets are available from the
Huntington NSW
Please see the back page for contact details.*

Helpful Contacts

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NSW Huntington Disease Service

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For further information on Huntington Disease, a membership form, or details of HD services in NSW, please contact Huntington's NSW

Acknowledgements: Huntington's Disease Association (England & Wales)
Huntington's Victoria