



Huntington's Disease Association

Huntington's disease

Challenging Behaviour in Juvenile HD

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Fact Sheet

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CHALLENGING BEHAVIOUR IN JUVENILE HUNTINGTON'S DISEASE (JHD)

What is Juvenile Huntington's Disease?

Huntington's disease (HD) is an inherited, neurodegenerative condition that causes abnormal movements and other physical signs as well as emotional and cognitive problems. It usually manifests itself in middle life, although it can present at any age. HD gradually worsens with time and there is currently no cure. Juvenile Huntington's disease (JHD) is the name given to any affected person who has signs or symptoms before the age of 20 years.

About 5-10% of all people with HD have Juvenile-onset HD, and so it is a relatively rare condition in the general population. This can make dealing with JHD an isolating experience for the young people with HD, their families and also professionals involved with their care.

The clinical picture of JHD can sometimes be very different from that in adults. They are more likely to display rigidity, bradykinesia, dystonia and epilepsy, and tend to show choreic movements less often.

One problem in particular that can cause real problems for families and professionals is challenging behaviour. It seems to be more common in young people who first develop symptoms in their teens, and at first can be the only obvious symptom of HD. It is important to remember though, that many children with JHD do not have these problems. However, they can be very difficult to cope with when they do occur.

Challenging behaviour in young people with JHD may include irritability, frustration, aggression and inappropriate social or sexual behaviour. However, the behaviour itself is less important than looking at why that problem is occurring, which can be difficult to determine.

What is causing the problem?

This is the most important, but very difficult question.

In many cases, it may be unclear whether challenging behaviour in JHD is due to the HD or the situation. It is now generally recognised in some cases, particularly in those who develop Huntington's disease in their teens, that challenging behaviour can occur for some time without obvious signs of movement disorders, and many specialists would be reluctant to give a diagnosis of JHD (or to refer the young person for a genetic test) solely based on these symptoms. However, it is important to remember that simply because a young person has HD, it does not follow that every other problem is related to it and changes in behaviour can have many different causes. This is true whether the young person has a diagnosis of JHD or not.

For a child with HD, the family situation can be fairly disrupted. They may have been prevented from carrying out some normal childhood experiences. Children in families where their parent has or had HD, may also have been living with or may still be living with a parent who is very ill. The parent may have died and they may be grieving for them. This can all be very difficult for young people. Also, they may have been living with the knowledge that they are at risk from developing HD, and now they may be living with the fact that they have developed HD and all that this means

for them. Their understanding or ability to cope with this may not be as sophisticated as that of an adult. This is particularly true if they have not had good genetic counselling or support from friends and family.

At the same time as having JHD, they are also trying to be a normal young person and are trying to deal with the usual issues that young people have to deal with. For example, they may be finding it hard to develop some autonomy and independence. Sometimes, these problems are simply the normal reaction of a teenager to a difficult situation.

However, it is equally important to remember that the challenging behaviour could be due to the HD and should therefore be managed sensitively. We know that changes in behaviour are fairly common in adults with HD. It is not surprising that they may also occur in young people with HD too. In adults with HD, these changes often occur because they are more impulsive and find it harder to control their behaviour than normal, or out of frustration. There is more on this written in other books (see further information).

As you can see, there are many different factors that could possibly be causing the problem, including both HD and the situation. Looking at these problems as either caused by HD, or by the situation, is a very simplistic way to look at the issue. It is very important to bear in mind that a child who is having difficulties in a HD family needs special attention and support, regardless of whether they have the disease or not. Whether or not the changes in behaviour are directly caused by HD, the young person may have had some very difficult issues to deal with indirectly related to HD and anyone who has contact with them should be equally sympathetic to this.

“THE LABEL”

When trying to deal with challenging behaviour both in a child with JHD (and in at-risk children and siblings) it is important to bear in mind the effect that labelling the child may have in the particular context of the family. This could be true both if you label the child as ‘difficult;’ rather than having a medical condition, or if you label them as having JHD. For example, in some cases, being labelled as ‘difficult’ can be more stigmatizing than having a diagnosis of HD. People may be more sympathetic to a child when they believe there is a medical reason for the child’s behaviour, but when there is not, there may be a temptation for people to place the blame for the young person’s behaviour on the family. Even if this isn’t intended, it could be how it is perceived by the family.

What can be done?

The reason for discussing the number of different causes for challenging behaviour was not to suggest that there is nothing that can be done to deal with the situation, but rather to help investigate the factors that could be causing the changes to behaviour in the young person. Although the precise cause of the problem can be difficult to identify, there are various options that can be tried.

The first important thing to do is to try and establish what might be causing the challenging behaviour. It is useful for family, schools and anyone else who has frequent contact with the young person to try and keep a record of exactly what is happening, and when. It is useful to try and build up a picture of the context surrounding the behaviour, the nature of the problem (exactly what happens), and when it usually happens. What you are trying to look for are any patterns in the behaviour that might give a clue as to why it is happening. People often tend to make their own interpretation of why the behaviour is happening, and so more objective information (by keeping a careful note of the behaviour) is useful to help investigate possible causes. Talking to the young person can be useful if trying to build up this picture, although it must be remembered that people

with HD can sometimes have very little awareness into the situation and the events surrounding the problem.

Sometimes the solution may be very simple and involve making small changes. For someone who cannot communicate well, some physical problems (e.g., pain, constipation, feeling sick or infection) can cause changes in behaviour and it is worth considering some of these first.

We also know that in Huntington's disease, certain changes in the brain make it very difficult for the person to do certain functions, or to cope with certain issues. In particular, people with HD usually have specific problems with multi-tasking and planning ahead. For example, it may be that the environment is too overwhelming for the young person with HD, who may be finding it difficult to absorb what is going on. Also, he or she may be frustrated for various reasons. For example, they may be finding it difficult to communicate, be having problems adjusting to changes in their routine, or be feeling frustrated by their lack of independence. Equally, they may just simply be very tired. There is a lot more information about this in the book 'Understanding Behaviour in Huntington's Disease,' by Jane Paulsen, or 'Hurry up and Wait' by Jim Pollard; both of which are available from the HDA (see 'Further Information').

In many cases a better understanding can help people cope with the behaviour more easily. It might be worth thinking about whether you could think differently about the behaviour. For example, if the young person is not doing what you ask them to do, it might be that they have difficulty getting started, and that they are not trying to be 'difficult.' When it is thought of in this way, then it becomes easier to accept their behaviour, as it is out of their control. Often, the best way to deal with challenging behaviour, as in this example, is to make some simple changes. For example, it may be possible to make certain changes to the environment that removes as much as possible the cause of challenging behaviour. At the same time, better understanding of why the behaviour is occurring can help family and friends to cope better with it and to be more tolerant.

SEEKING PROFESSIONAL HELP

The young person's problems may pass after a period of time, or disappear after making small changes to the environment. However, if the young person's behaviour is causing real difficulties, or doesn't seem to be going away, then it may be worth seeking the advice of a paediatric psychiatrist or an educational psychologist. It is worth remembering that specific psychiatric diagnoses, such as depression or psychosis can present with challenging behaviour, and a psychiatrist may be able to look at some of these other possibilities.

Also, they may be able to help by developing a behavioural modification plan (i.e., where good behaviour is rewarded). Although people with HD often have a lack of insight, behavioural modification plans can work well. In fact, programmes based on rewards and reinforcement of good behaviours can work even better in some people with HD than people without HD. Keeping good records of the young person's behaviour might help the psychologist or psychiatrist plan a programme suited to the specific circumstances.

Sometimes we can inadvertently reinforce bad behaviour without realising it, which can encourage challenging behaviour. For example, when the young person is given attention because of specific behaviours, this may inadvertently reinforce them. A psychologist or psychiatrist might be able to help you identify the ways in which this might be happening.

Some standard behavioural therapies may not work well or be logical, give the cognitive aspects of the disorder.

MEDICATIONS

In some adults with HD that suffer from impulsivity and aggression, certain medicines can be useful in controlling these behaviours (e.g., anticonvulsants, and a group of drugs called Selective Serotonin Re-uptake Inhibitors, or SSRIs). Also, adults with HD have an increased risk of depressive illness, and to a less extent psychosis, which can lead to challenging behaviours. Some people with JHD in their mid to late teens may also develop similar problems, but these can be responsive to appropriate medication.

However, there are a number of issues about drugs to control challenging behaviour that can be even more complicated when these drugs are used in young people (e.g., finding the correct dose for the person, side-effects, taking too many different medications and the safety of certain medications). Medications that are used for these behaviours are often unlicensed for use in people under the age of 18 years, but may be prescribed on the grounds that they are used in children with similar behaviours from other causes, as well as adults with HD and this type of problem. A paediatric psychiatrist will be able to give you advice about this.

It is also important to remember, that medications will only help where the cause of the problems is organic (i.e., caused by brain changes). It is therefore important to consider any other possible causes and solutions first, before considering medications. Again, keeping a good record of the behaviour will help a psychiatrist to decide whether medications might be helpful in a particular situation.

YOUNG PEOPLE AT-RISK IN A FAMILY WITH HD

One group that hasn't been considered so far here is young people living at risk of HD. Again, they could have problems and there could even be some question that they may have JHD as a result of their behaviour (although JHD is very rare). Some of the issues that they are dealing with may be similar to those with JHD. They may also have a disrupted family background, and may have been helping to care for a parent with HD. They may also be living at-risk from developing HD. In some ways these young people need to be supported in a similar way to other young people with disrupted family backgrounds. However, as for the young person with JHD, it is important to bear in mind the implications of the way their behaviour is interpreted in the context of the family history of HD (see the paragraph on labelling).

The thought that the young person has HD is likely to be quite scary and worrying for him or her for many reasons, such as having seen affected relatives suffer from the condition. However, it could equally be quite upsetting for them, and for the family, for them to be labelled as 'difficult' when, in fact, their behaviour is at least partly caused by HD. This can make the family or the child feel blamed for the difficult behaviour; if the issue is not dealt with sensitively. There is no answer as to the best way to deal with this situation and it depends a lot on the individual situation. It is important, however, to be aware that challenging behaviour could still be caused by a number of factors and to discuss any issues surrounding any difficulties sensitively with the young person and their family, and to support the young person regardless of the cause.

SOME FINAL COMMENTS...

Challenging behaviour can be difficult to cope with in any child, but when the people having to cope with it are coping with many other issues to do with Huntington's disease as well, it becomes even harder. There is no simple answer to dealing with challenging behaviour. However, carefully noting down the situations in which the behaviour happens and trying to look for patterns, may lead

to a better understanding of why the behaviour happens. When you have done that, you have taken a big step towards finding some way of improving the situation.

It is worth stressing again that challenging behaviour does not occur in all children with JHD. Given that this leaflet has looked in detail at these problems, it is tempting to think that this happens in most young people with JHD. This is not the case and many have no serious difficulties in this area. As with all young people, challenging behaviour can also occur for a while and then go away again. There may be a simple reason for the young person's changes in behaviour and some ideas for what this could be have been discussed. However, this leaflet is primarily aimed at people who are having real difficulties dealing with the problems challenging behaviour can cause in JHD.

FURTHER INFORMATION

Jane Paulsen, 'Understanding Behaviour in Huntington's Disease', available from the Huntington's Disease Association (www.hda.org.uk)

Jim Pollard, 'Hurry up and Wait!' Available from www.lulu.com/content/2517713

Huntington's Disease Association Factsheet: 'Behavioural Problems in Huntington's Disease', available from the Huntington's Disease Association (www.hda.org.uk)

Quarrell, O. (2008) 'Huntington's Disease: The Facts (2nd Edition)', available from the Huntington's Disease Association(www.hda.org.uk)

Quarrell, O. et al (2009) 'Juvenile Huntington's Disease and other Trinucleotide Repeat Disorders' published by Oxford University Press ISBN 978-0-19923612-1

Glendinning, N. 'Huntington's Disease in Children and Teenagers: A Guide for Professionals', available from the Huntington's Disease Association(www.hda.org.uk)

Huntington's Disease Association 'Standards of Care' available from:
www.hda.org.uk/download/fact-sheets/HD-Standards-Of-Care.pdf

Nance, MA (2007) 'The Juvenile HD Handbook: A Guide for Families and Caregivers', available from the Huntington's Disease Society of America(<http://www.hdsa.org/images/content/1/1/11702.pdf>)

Huntington Society of Canada (2000) 'Juvenile Huntington Disease: A Resource for Families, Health Professionals and Caregivers', available from:
www.huntingtonsociety.ca/english/uploads/Juvenile_HD_2008.pdf

The Challenging Behaviour Foundation (www.thecbf.org.uk)

TOP TIPS FROM FAMILIES FOR FAMILIES

TIPS FROM FAMILIES AT THE JHD FAMILY WEEKEND 2008

We talked about the challenges facing the family when one or more members has JHD. These are some of the tips we shared for surviving together as a family.

When you disagree with a family member..... what might help is to:

Walk away for 10 minutes (and come back calmer.....)

Change the focus of the discussion from a negative to a positive (what they can do, not what they cannot do)

Bring someone else into the room/home to deflect attention

Present a choice

Take an indirect approach to a request

Use other people in the family to negotiate, e.g. one says no, the other gives in, with conditions

What doesn't help is:

Confrontation

Trying to rationalise with the person

Expecting them to see your point of view

Sometimes family members focus on a particular happening for hours, days or weeks. It can cause distress if the planned event doesn't take place.

It might help to:

Avoid mentioning treats until very close to the time they will take place. Also ask others to avoid subjects that cause excitement and could influence behaviour e.g. birthdays.

ALWAYS have a plan B, to avoid disappointment if plan A fails. Introduce both plan A and plan B as possibilities, ahead of time. For example, we might go to X, but if it rains or the car is not running well, we might stay home and get a video.

Everyone likes to have some control over their life.....

Give the person options and allow them to make some choices, e.g. what they will wear. When asking the person to make a choice, put the possible options in front of them e.g. this shirt or that shirt, this food or that food. This is far easier than asking them to think abstractly.

To reduce unsettled behaviour

Have a routine and stick to it

Look after yourself by:

Setting boundaries. If you give in completely, you lose yourself, so set boundaries beyond which you do not go, and be consistent.

Not being too hard on yourself.

Celebrating the small successes!

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Fact sheets available from the Huntington's Disease Association:

- General information about Huntington's Disease and the Huntington's Disease Association
- Predictive Testing for Huntington's Disease
- Talking to Children about Huntington's Disease
- Information for Teenagers
- Eating and Swallowing Difficulties
- Huntington's Disease and Diet
- The Importance of Dental Care
- Communication Skills
- Behavioural Problems
- Sexual Problems
- Huntington's Disease and the Law
- Huntington's Disease and Driving
- Advice on Life Assurance, Pensions, Mortgages etc.
- Seating Equipment and Adaptations
- Checklist for Choosing a Care Home
- Advance Decision to Refuse Treatment (ADRT)
- A Carers Guide
- A Young Adults Guide
- Challenging Behaviour in Juvenile Huntington's Disease
- A Brief Guide to Juvenile Huntington's Disease for Children's Hospices and Palliative Care Service
- A Young Person with Juvenile Huntington's Disease at School

All fact sheets can be downloaded free of charge from our website www.hda.org.uk or ordered from Head Office.

For a publication price list/order form, membership form, details of our Specialist HD Advisers and local Branches and Groups, please call Head Office on:

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