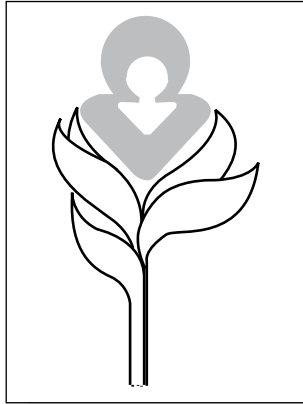


Huntington's Disease Association



A brief guide to Juvenile Huntington's Disease for Children's Hospices and palliative care services

Helen Brewer & Dr. Nicky King

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Fact Sheet

HUNTINGTON'S DISEASE (HD) IS AN INHERITABLE A BRIEF GUIDE TO JUVENILE HUNTINGTON'S DISEASE FOR CHILDREN'S HOSPICES AND PALLIATIVE CARE SERVICES

What is Juvenile Huntington's Disease?

Huntington's disease (HD) is a hereditary, 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. This means that Huntington's disease can be considered in group 3 of the Association of Children's Palliative Care (ACT) groups that can be described as having life-limiting conditions (progressive conditions without curative treatment options, where treatment is exclusively palliative and may commonly extend over many years) (ACT/ RCPCH, 1997). Huntington's Disease affects about 7-10 in every 100,000 people.

Juvenile Huntington's disease (JHD) is the name given to any affected person who began to exhibit signs or symptoms before the age of 20 years. This means that there may be some young adults in their twenties, or even older, who would be termed as having JHD. Throughout this leaflet the term 'child' will be used, although it should be recognized that in many cases the person with JHD may be older, and even a young adult.

About 5-10% of all people with HD have Juvenile HD. The Huntington's Disease Association (HDA) know of about 80 people in the country with 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 (or slowness of movement), dystonia (muscle contractions causing abnormal movement or postures) and epilepsy. They are less likely to have choreic movements (involuntary, dance-like movements commonly seen in adult-onset HD).

For more information about the clinical features of JHD, please see the book 'Huntington's Disease in Children and Teenagers' by Neil Glendinning, or the guides to JHD by the Huntington's Disease Society of America and the Huntington's Society of Canada (see 'Further Information' for details of these guides).

FAMILY ISSUES

Family issues are particularly important with both Huntington's Disease (HD) and Juvenile Huntington's Disease (JHD) because of the inherited nature of the condition. If a parent has the condition, then each biological child has a 50% risk of inheriting the condition. This means that there are often other family members who have the condition. In a family with a child with JHD there may often be one parent with HD who is still alive and they may also be being cared for at home by the same carer as the child with JHD. It also means that other members of the family may be at risk of developing HD, including siblings of the child with JHD. In some families, there may be more than one sibling with JHD, so there is also a possibility that the carer is caring for more than one child with JHD at home. Parents may also have concerns about whether other children in the family are developing, or will develop, HD.

The hereditary nature of HD means that there is often repeated loss in families affected by HD, and so bereavement / grief work is particularly important. It should also be remembered that previous experiences of loss and HD may impact on the family's responses to situations that they encounter with a child with JHD.

With Juvenile HD, the father is usually the affected parent (in about 70-80% of cases). This means that the person caring for the child with JHD is often the mother, who may be caring for the child without the support of another parent / partner.

GENETIC TESTING

A predictive test for Huntington's disease (HD) is available, which can tell someone at risk of HD whether or not they will go on to develop the condition. This test is not usually available to people under the age of 18 years old. In the case of JHD, the same genetic test may be used along with other tests to diagnose the condition. However, to avoid mistaking the child's symptoms for JHD when they are actually caused by some other condition, clinicians are usually very cautious about using the test. Further information about this can be found in Oliver Quarrell's book 'HD – The Facts', the Huntington's Disease Society of America guide to JHD, and in the book 'Juvenile Huntington's Disease and other Trinucleotide Repeat Disorders' (see 'Further Information').

TALKING TO CHILDREN

Parents may ask for advice or support in talking to their children, either about JHD or about being at risk of developing HD. Palliative care services may have an important role in supporting the family with this. Further information is available in the Huntington's Disease Association leaflets 'Talking to Children' and 'Talking to Teenagers', as well as Bonnie Hennig's book, 'Talking to Kids about Huntington's Disease'. Two books for children are also available from the Huntington's Disease Association: 'Huntington's and Me' (Alison Gray) (aimed at older children and teenagers) and 'Huntington's Disease in the Family' (aimed at younger children).

SYMPTOMS AND SYMPTOM CONTROL

There is currently little guidance on symptoms and symptom control, with each clinician using his or her knowledge of the other treatments and similar conditions to treat the different symptoms of Juvenile HD. This is one of the reasons why palliative care services can have such an important role in the care of a child with JHD.

Given that children with JHD can present very differently to adult-onset HD, different treatments are often used (see Aubeeluck and Brewer, 2008).

Only one article has to date addressed the treatment of late stage JHD (King, 2005). Another useful chapter on the treatment of JHD can be found in the book 'Juvenile Huntington's Disease and Other Trinucleotide Repeat Disorders (see 'Further Information').

Prescribing needs to take a commonsense approach, directed at symptoms but bearing in mind sensitivity to medication and drug interactions, trying to keep medication to a minimum and avoiding poly-pharmacy. Close working with HD specialists and palliative care teams can enhance symptom control with each contributing their expertise and experience. It is particularly important to be aware that pain is a common symptom which is often under-treated due to the fear of strong medication and the difficulty in assessing pain due to communication difficulties. Morphine appears to be well tolerated and effective for the pain experienced by patients with JHD. Alternative analgesia may be required for muscle spasm related pain.

COMMUNICATION

Communication often becomes increasingly difficult during the course of the condition. It has been highlighted as one of the aspects of the condition that particularly impacts on children with the condition and their families (Smith et al., 2006). People with HD are able to understand more than they may appear to, as the condition will affect facial expressions and movements that show their understanding and some of their cognitive (or thinking) processes. In HD thinking is slower and it may take someone with HD longer to process information and reply. All this can make communication harder, and the emphasis is on the listener to help the person with HD communicate. Struggling to communicate can be a cause of agitation or frustration. For further information, please read the HDA factsheet 9, 'Communication Skills'. Some of these ideas are explained very well in the book, 'Hurry up and Wait' by Jim Pollard (see 'Further Information').

CHALLENGING BEHAVIOUR

Huntington's Disease can affect behaviour in some people, although this is not true for all children with JHD and can vary a lot between individuals. A guide to JHD and challenging behaviour can be downloaded through the Huntington's Disease Association website. Another useful book is the HDA guide, 'Understanding Behaviour in Huntington's Disease' and the HDA factsheet 10 'Behavioural Problems' (see 'Further Information').

SCHOOLING

A child with JHD may still be attending school, and the hospice or palliative care services may have an important role in supporting the school in their role. A guide to JHD for schools can be found at the Huntington's Disease Association website (see 'Further Information').

DISEASE PROGRESSION AND END OF LIFE ISSUES

Adult-onset Huntington's disease usually progresses over 15-20 years, with death often being caused by HD-related complications (e.g. pneumonia). There is some suggestion that the progression of Juvenile HD might be slightly quicker, but there is little evidence for this and it is likely that this is limited to a sub-set of those with JHD. The reality is that with both adult-onset HD and JHD it is very difficult to predict exactly how each individual's HD will progress.

Paediatric services should be aware that transition to adult services may be required, and should be planned for and supported.

The Huntington's Disease Association have a 'Standards of Care' document which can be downloaded from their website with a section on 'End of Life' as well as other useful sections on other aspects of care (e.g., diet, swallowing and communication). The Scottish Huntington's Association has a Care Pathway, which has information on many of the specific issues which may arise in the later stages of HD (see 'Further Information').

Both the Huntington's Disease Society of America and Huntington's Society of Canada have guides to JHD that contain information about later stages (see 'Further Information' for details of these guides) and the HDA has a 'Caregivers Handbook for Advanced-Stage Huntington's Disease'.

ADVANCED PLANNING

In view of the natural progression of HD and the anticipated loss of capacity it is good practice to make advance care plans encompassing decisions on End of Life issues such as place of care, resuscitation and feeding. The Huntington's Disease Association produces an information factsheet on Advanced Refusal of Treatment (see 'Further Information').

A high calorie intake is important for patients with JHD and can reduce symptoms. However swallowing often becomes a problem and tube feeding may be offered as an option. This can be beneficial in ensuring a good calorie intake and compliance with medication. Decisions should be taken early as inserting a feeding tube later in the disease can be risky. However it is important patients and families are fully informed of the risks and benefits and have the opportunity to consider end of life decisions, such as withdrawing feeds.

SOME FINAL COMMENTS

A recent audit of cases of Juvenile Huntington's Disease known to children's hospices in England and Wales showed that a relatively small proportion of children affected by JHD access children's hospices. However, it is also clear that children's hospices and palliative care services have a lot to offer these families, including respite breaks, hospice at home services, therapy services, key-worker / advocacy role, family support, symptom control advice, end of life care and bereavement support. Although those working with the family, or even the hospice, may not have had much, if any, experience of JHD, they

have significant experience and a valuable approach that can add a lot to the care of a child with JHD.

The Huntington's Disease Association has a team of Regional Care Advisers (and a Care Adviser for Juvenile HD) who offer information, advice and support to both families and professionals involved in the care of people with Huntington's Disease. They can work closely with palliative care services to provide support to families.

Further Information

INFORMATION AVAILABLE FROM THE HDA

Glendinning, N. 'Huntington's Disease in Children and Teenagers', 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

Huntington's Disease Association 'Standards of Care' www.hda.org.uk/download/hda/Standards-Of-Care.pdf

'Challenging Behaviour in Juvenile Huntington's Disease' Leaflet, available to download from www.hda.org.uk/download/acrobat/JHD-001.pdf

'Information for Schools on Juvenile Huntington's Disease', available from www.hda.org.uk/juvenile/schools.html

Huntington's Disease Association Factsheet 'Talking to Children about Huntington's Disease', available to download from www.hda.org.uk

Huntington's Disease Association Factsheet 'Information for Teenagers', available to download from www.hda.org.uk

Huntington's Disease Association Factsheet 'Communication Skills', available to download from www.hda.org.uk

Huntington's Disease Association Factsheet 'Behavioural Problems', available to download from www.hda.org.uk

Huntington's Disease Association 'Huntington's Disease in the Family', available from the HDA, www.hda.org.uk

Gray, A. (2000) 'Huntington's and Me: A Guide for Young People', available from the HDA, www.hda.org.uk

Paulsen, J. 'Understanding Behaviour in Huntington's Disease', available from the HDA, www.hda.org.uk

Pollard, J. 'Caregivers Handbook for Advanced-Stage Huntington's Disease', available from the HDA, www.hda.org.uk

Pollard, J. 'Hurry up and Wait!', available from the HDA, www.hda.org.uk or <http://www.lulu.com/content/2517713>

INFORMATION AVAILABLE FROM OTHER HD ASSOCIATIONS

Nance, MA (2007) 'The Juvenile HD Handbook: A Guide for Families and Caregivers', available from the Huntington's Disease Society of America, www.hdsa.org

Huntington Society of Canada (2000) 'Juvenile Huntington Disease: A Resource for Families, Health Professionals and Caregivers', available from http://www.huntingtonsociety.ca/english/uploads/Juvenile_HD_2008.pdf
Scottish Huntington's Association 'Integrated Care Pathway' www.hdscotland.org

JOURNAL ARTICLES / BOOKS

King, N. (2005) 'Palliative care management of a child with juvenile onset Huntington's Disease' *International Journal of Palliative Nursing*, 11: 278.

Aubeeluck, A. & Brewer, H. (2008). Huntington's disease. Part 2: treatment and management issues in juvenile HD. *British Journal of Nursing*, 17(4):260-3.

Brewer, HM, Eatough, V, Smith JA. Stanley, CA, Glendinning, N. & Quarrell, OWJ (2008). The impact of Juvenile Huntington's Disease on the family: The case of a rare childhood condition. *Journal of Health Psychology*, 13 (1): 5-16.

Brewer, H.M., Smith, J.A., Eatough, V., Stanley, C.A., Glendinning, N.W. & Quarrell, O.W.J. (2007). Caring for a child with Juvenile Huntington's Disease: helpful and unhelpful support. *Journal of Child Health Care*, 11(1): 40-52.

Smith, J.A., Brewer, H.M., Eatough, V., Stanley, C.A., Glendinning, N.W. & Quarrell, O.W.J. (2006). The personal experience of Juvenile Huntington's Disease: an interpretative phenomenological analysis of parents' accounts of the primary features of a rare genetic condition. *Clinical Genetics*, 69: 486-496.

Quarrell, O. & Nance, M. (2009) 'The Diagnostic Challenge' in Quarrell et al (eds) 'Juvenile Huntington's Disease and Other Repeat Disorders, Oxford: Oxford University Press.

Nance, M. (2009) 'The Treatment of Juvenile HD' in Quarrell et al (eds) 'Juvenile Huntington's Disease and Other Repeat Disorders, Oxford: Oxford University Press.

OTHER INFORMATION

Hennig, B (2003) 'Talking to Kids about Huntington's Disease', available through www.talkingtokidsabouthd.com

Association for Children with Life-threatening or Terminal Conditions and their Families / Royal College of Paediatrics and Child Health (1997): Guide to the Development of Children's Palliative Care Services. ACT, Bristol and RCPCH, London

"Tools and support for assessing palliative Care needs" CD ROM / Tool kit produced by Forum Business Media <http://www.forumbusinessmedia.co.uk/>

Fact sheets available from the HDA:

- General Information about Huntington's Disease and the HDA
- 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 Carer's 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 for free from our website:
www.hda.org.uk or ordered direct from Head Office

For a publication price list/order form, membership form, details of our Regional Care Advisers and local Branches and Groups, please telephone, email or write to:

Huntington's Disease Association

Neurosupport Centre, Norton Street, Liverpool L3 8LR

Tel: +44 (0)151 298 3298 Fax: +44 (0)151 298 9440

Email: info@hda.org.uk Web: www.hda.org.uk