



Facing Huntington's Disease
A handbook for families and friends

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■ Introduction

THE PURPOSE OF THIS BOOK

This booklet is intended to give the reader information on Huntington's Disease (HD) and the issues that this condition raises, not only for the sufferer, but families, those with a 50% risk of inheriting the disease and professionals concerned with caring for or advising people.

It is not a replacement for more detailed medical texts or direct medical advice but presents the facts in what is hoped is a straightforward and helpful way.

Various Thank You's

The contents of this book owe much to other publications, especially material kindly made available by International Huntington's Disease Organisations

It has been edited by the Huntington's Disease Association of Ireland with assistance from Professor Andrew Green, National Centre for Medical Genetics. The 2003 edition is printed with financial assistance from the Northern Area Health Board, Department of Health, Dublin.

Feedback Invited

Huntington's Disease is a condition with many facets and in attempting to describe

these facets, the authors do not underestimate the range of problems that the condition brings; it is a feature of the disease that there are many examples of differences from the norm, and thus readers may have direct experience which differs from that detailed in this publication. Feedback and information would be most welcomed by the Huntington's Disease Association of Ireland (HDAI).

ABOUT HUNTINGTON'S DISEASE

Huntington's Disease (HD) is an inherited disorder of the central nervous system. It causes progressive deterioration with varying symptoms which may include involuntary movements, speech impairment and intellectual and emotional changes. In this booklet, a person with HD is referred to as a pHD.

Symptoms usually appear between the ages of 30 and 45, although they may appear earlier or later. The disease can also appear in children and this is known as Juvenile HD. HD is a genetic condition with each child of an affected parent having a one in two chance or 50% likelihood of inheriting the gene. Both men and women have equal chances of being affected. People with the abnormal gene will almost always develop the disease, unless they die of other causes



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prior to developing signs and symptoms. The illness usually lasts about 15-20 years and death is often from pneumonia or choking. Improvements in general health, such as adequate nutrition, will bring about improvements in the pHD's quality of life. It is estimated that pHDs need twice the normal calorie intake to sustain them. Many patients who gain weight also find their clinical state improves. See care of the pHD (P16) for more information.

People who do not inherit the HD gene will not develop the disease, neither will their children, or their children's children. The disease does not skip a generation.

Is there a cure for HD?

No. There is presently no cure for HD. A protein called "Huntingtin" is produced by the HD gene, and the structure of this protein in people with and without HD is being studied, to understand better how an altered HD gene causes the condition. However, although this area of research is highly active, no clear treatment has yet been identified.

It was estimated from a survey carried out by Dr. Morrison in Belfast (1992) that perhaps 400 people have the illness in the Republic of Ireland with a further 2,000 individuals at risk. Professor Andrew Green

of the National Centre for Medical Genetics believes the incidence in the Republic of Ireland is more common and that there are at least 400-500 people affected. While HD is relatively rare, over 9,000 family members in Ireland may require support and information.

ABOUT HDAI

The Huntington's Disease Association of Ireland (HDAI) provides consultation, information and individualised support to those diagnosed with HD, their families and their health care teams. HDAI's information is regularly updated through links with the International and European Associations. HDAI is a registered charity and was incorporated in 1998.

Because HDAI has been dealing specifically with HD in Ireland for almost 20 years, the Association has extensive expertise and knowledge in helping families. We give the family all the time, support and practical help we can to enable them to live with this disease.

The Association gives help and support to families and caring professionals through literature, newsletters and individualised advice and support. Information is available by contacting the HDAI office.



The first recorded meeting was held in May 1983 and HDAI was formally launched in 1985 to assist sufferers and their families. Its principal aims and objectives are:

- To provide specific advice to those who have the disease and their families.
- To provide practical help where possible
- To foster and promote research
- To provide up to date information to those interested in Huntington's Disease.
- To avail of every opportunity to highlight the needs of our members through the media.
- To update information regularly.
- To help establish the true incidence / prevalence of HD in the Republic of Ireland

HISTORY OF HUNTINGTON'S DISEASE

Huntington's Disease was first described in 1872 when a 22 year old American

doctor, George Huntington, had his paper published in the Medical and Surgical Reporter of Philadelphia and the hereditary disorder he described became known as Huntington's Chorea. The word "chorea" is derived from Latin and Greek words meaning chorus or group of dancers.

Today the term Huntington's Disease rather than Huntington's Chorea is more common because we have learned that some patients display a more rigid form of the disease and some may show more intellectual and behavioural deterioration rather than the physical symptoms especially in the earlier stages of the illness.

In the past chronic adult hereditary chorea was poorly understood. While the gene which causes the disease was probably no less common than it is today, the disorder itself seemed rarer because fewer carriers lived long enough to manifest symptoms.

Early diagnosis and better management provides a better future for people with this disease. Ongoing research provides greater hope for future generations.



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■ The Facts

WHAT HAPPENS IN THE BRAIN

Huntington's Disease is caused by the destruction of brain cells, particularly in those parts of the brain known as the basal ganglia and the cerebral cortex. By some mechanism yet unknown, the causative dominant gene, which for years has remained inactive, begins to take its toll. Central nervous system neurons (brain cells) begin to die.

The gradual destruction of brain cells causes mood, memory and/or movement disorder. See diagram of HD BRAIN - on inside cover

SYMPTOMS

The illness begins to manifest itself by a change in usual behaviour, for example, depression, moodiness, unreasonable outbursts of anger out of character for the individual, or by unusual jerky, fidgety movements and perhaps unsteadiness of the hands or feet causing falls and a tendency to be clumsy. However, these

symptoms can arise for various reasons and may not be linked to the onset of HD.

These early signs are mild and increase so slowly that they may go unnoticed and it is only much later, when looking back that relatives realise all has not been well for some years.

It is worth noting that there are at least 50 different conditions where involuntary muscle movements may be present. If you fear that HD may be present the best way to resolve worrying questions is to see a specialist who is experienced in the diagnosis and treatment of neurological disorders. Ask your General Practitioner to refer you to a Neurologist.

WESTPHAL VARIANT

A minority of pHDs manifest a form of the disease characterised by muscular rigidity rather than chorea. This is called the Westphal Variant of the disorder after Dr. Westphal who described it in 1883. Before naming this variant he mistakenly believed that it was another condition. The emotional and intellectual problems common in the choreic form of the disease are also common in the rigid form.



DISEASE PROGRESSION

Over the years the illness gets more severe, though the rate at which it progresses varies from individual to individual. The ungainly jerky movements over which the sufferer has no control, increase, causing falls and making walking difficult. Some, though not all, will at times become confused and forgetful, at other times angry and unreasonable and possibly violent, although some people become quiet passive. This changing state is confusing and frustrating for both sufferers and their families.

A pHD can suffer from other unrelated

illnesses e.g. a chest infection etc therefore if in doubt it is best to see the GP.

At present there is no known cure, but some symptoms can be treated. Sympathetic and understanding care can help to keep the pHD more comfortable and less agitated.

Regular respite breaks and community support can help keep the patient at home. Long term care for those with neurological conditions has improved in recent years. The patient and the family are encouraged to make decisions regarding their care plan.



■ Genetic Aspects of Huntington's Disease

INHERITANCE OF HD

HD is passed from one generation to the next because of an alteration in one of the many genes each of us inherits from our parents. The gene that causes HD is called IT15. It is inherited in an autosomal dominant manner. This means that if either parent has the altered HD gene, each son and daughter has a 50% or 1 in 2 chance of inheriting or not inheriting HD.

A fifty per cent chance does not mean that exactly half the children will get the disease in a family where the altered HD gene is known to be present. Each individual child of a person with an altered HD gene stands a 50% or 1 in 2 chance at the moment of conception of inheriting the altered HD gene. This could mean, for example, that one child in a family of four children will develop HD, or two may get it, or three, or perhaps all four, or none. Each person faces his or her own fifty per cent chance irrespective of whether any of his or her brothers or sisters is affected or

not. An altered HD gene never skips a generation. It does not appear in one generation, skip the next, then reappear in a third or subsequent generation. If a person does not have an altered HD gene, they cannot pass HD on to their children.

However, not everybody with HD will have a clearly affected parent. Sometimes a parent who has an altered HD gene can die from something unrelated to HD, before ever showing signs of HD. In that case, their son or daughter could be affected, without an apparently affected parent.

THE GENETIC TEST

Discovery of the HD gene

The gene which when altered causes HD was isolated in March 1993. The gene is called IT15, and is located on the tip of the short arm of chromosome 4. The abnormality which causes HD is an expansion of the DNA sequence of an otherwise normal gene. In people with HD, the HD gene contains between 40 and 100 copies of a repeated DNA sequence in the gene, whereas most people have less than 30 copies of the repeated DNA sequence. Rarely, individuals may have a repeat size of close to 38 copies, and interpretation of a result in this region can be difficult. The expanded gene alteration is found in



virtually all cases of HD, and can be used as a diagnostic test, as well as a predictive test.

What is a diagnostic and a predictive test?

A diagnostic test is used to confirm the diagnosis of HD in a person showing symptoms and signs of the disease. The test is carried out to confirm a clinical likelihood that a person already has HD. A predictive test for HD is carried out in a healthy person, who has no signs or symptoms of HD, but who has a family history of HD. The predictive test will determine whether that person has a HD gene alteration. If the person has a HD gene alteration, they will almost certainly develop the condition in their lifetime. Not everyone with a family history of HD wishes to pursue a predictive genetic HD test, and having the test remains an individual choice.

The results of the test would be one of the following three possibilities:

1. A person has inherited an altered HD gene
2. A person has inherited a normal HD gene.
3. Rarely, the test result is uninformative

Why have genetic counselling?

Genetic counselling gives people who wish to have a predictive HD gene test the opportunity to discuss the issues involved in testing, to have their questions

answered, and to consider the possible consequences of a predictive HD gene test result.

Once a person starts on the predictive test programme, a partner, close friend or family member should come to the appointments. At least three appointments are usually necessary before testing. A referral to a psychiatrist is made, to make sure that a person is able to deal with an unfavourable result, and also for follow-up if necessary when a result is unfavourable. A person receiving their test result needs to be accompanied to that appointment.

Why are partners or close friends asked to attend?

It is extremely important that a partner, close friend or relative accompanies the individual to their appointment. The result may have implications for members of the family, therefore they should have the opportunity to consider the implications of the result for themselves. Individuals accompanying a person should understand the anxieties the person may be experiencing.

Can children have a predictive test for HD?

No. Usually only adults are offered predictive testing. This is because the test is voluntary, and children usually cannot understand the complex issues involved in



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the decision to take the predictive test. In addition, children cannot legally themselves give consent for the test.

GENETIC CLINICS

If any person at risk of HD is interested in finding out more about the genetic aspects of HD, or about the predictive genetic test, they can talk to a clinical geneticist or genetic counsellor. The National Centre for Medical Genetics holds genetic clinics in Dublin, Cork, Galway and Limerick. More details are available from the National Centre for Medical Genetics (01 409 6739) or the HD association (01 872 1303).

GENETIC COUNSELLING

The definition of genetic counselling is "the process by which patients or relatives at risk of a disorder that may be hereditary are advised of the consequences of the disorder, the probability of developing or transmitting it, and of the ways in which this may be prevented, avoided or ameliorated" (P. Harper - 1983)

Genetic counselling is mainly an educational process that seeks to help those affected by HD or at risk of HD to understand the genetics of the disorder, the way in which it may have been inherited, and the options that are

available to them in management and further reoccurrence.

TELLING THE CHILDREN

Many parents and caring professionals find it hard to talk to children about HD. Nobody likes to be the giver of bad news and, although they may acknowledge that they should tell, the actual information-giving and possible repercussions seem overwhelmingly difficult.

If someone in the family has HD, it will be difficult to avoid giving a child some sort of explanation. Small children are more accepting than adults are, but become less so as they grow older and therefore need to know that someone is actually ill in order to understand and accept certain behaviour.

Children are also very sensitive to atmosphere and, if they realise that something is not being discussed, can imagine all kinds of things as the cause of the family 'secret'. They may feel they are in some way to blame and this anxiety and guilt can be more harmful than knowing the facts.

Ideally the telling should be a gradual sharing so that children can grow into a gradual understanding made possible by parents acting openly and answering questions appropriate to the child's age.



Parents should be as reassuring as possible without denying the risks. As the child gets older it is important that they can discuss their worries and feelings in an environment where their feelings are accepted and understood.

Some parents may be able to face this task on their own, but many parents will find it difficult and may need to find someone with whom to talk over their own feelings and the needs of the child. This could be a relative or close friend, doctor, social worker or other professional. The important thing is to find someone with an understanding both of HD and of children, who will give good advice and help you to work out the best approach. It may help too, to get together with other parents and share your ideas, provided you don't feel that everyone must agree on how to act, because each family will have different circumstances.

Moodiness and irritability can make life very difficult for the children and spouse. The first and probably most difficult step to face is the truth, that a family member is affected. The second step is to speak openly about the situation. This won't come easily but it is important to work toward it. The whole family must understand what is happening.

Truth and honesty within the family helps friends and relatives feel more comfortable in their relationships with an HD person and makes it easier to lend much needed help and support.

COPING WITH BEING AT RISK

Studies of people at risk have shown that each individual reacts to this challenge in a variety of ways. Even though there is just as great a mathematical chance that the person will escape the disease as there is that they will get it, for some people being at risk means a constant struggle to master the odds and they may live lives of dread, denial, fear, emotional disarray and gloom.

Understandably at times this struggle makes their situation seem much worse but at other times they can live with it.

Some people respond to the risk situation by trying to ignore the disease. Pretending that it either does not exist or couldn't possibly touch them, they push the reality of the disease out of their minds. They do not talk about it nor do they seriously consider it when they contemplate marriage and a family. Unfortunately, the disease itself may not co-operate with this attitude of denial. It cannot be wished away.



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Families may be helped by recognising a five step coping process we may all experience:

1. **DENIAL:** The person at risk to HD may refuse to accept the information and says "No, its couldn't happen to me." Sometimes the person accepts the information well, but later cannot recall the details of what was said.
2. **ANXIETY:** The person can suffer headaches, fatigue, insomnia and irritability as a response to fears. They may benefit from emotional support at this time.
3. **ANGER:** The person "sees his/her misfortune as the act of a cruel and uncaring world" and may be openly hostile to friends, relatives and health professionals. There may be a great deal of angry behaviour. Resentment builds between family members. Counselling helps to channel the guilt and redirect the anger in constructive ways.
4. **DEPRESSION:** This is a critical phase, necessary for eventual readjustment. The person will need support to accept changes, try new behaviours and plan for a new way of life.

5. **STABILITY:** The person at risk "cycles" back and forth between these five phases but with family and professional support and reinforcement, can be helped to live with the threat of HD.

MARRYING SOMEONE AT RISK

Thinking of marrying someone at risk can pose problems. If you have never seen or even heard of HD, you may try to ignore the risks and possible consequences. Some of you will feel so frightened that you break off the relationship almost immediately without going further into the subject. Learning the real facts and weighing up the risks and what they mean to you, against the quality of the relationship, is the only way a proper decision can be taken. If you understand the implications and can work out plans for the possibility that your partner either may or may not get HD, then your marriage can be happy and a success whatever happens.

This is an extremely complex area but the right of all individuals to make decisions, provided they are well informed, should be respected.



If people have sufficient information it will help them to make an informed decision.

If you are married to someone at risk they may not have told you because they may never have "told" themselves. See section on "Coping with being at risk" (page 9).

WHETHER OR NOT TO HAVE CHILDREN?

The hereditary nature of HD, makes the prospect of starting a family particularly difficult. However, many individuals at risk to HD have already established families before they learn about HD or fully understand the hereditary nature of it. Children of an at risk parent have a 25%

risk themselves. Some who fully understand HD and its hereditary implications may choose to have their own children. Others at risk may decide not to have children of their own in order to avoid passing the disease on to another generation.

Through genetic counselling the full implications of the genetic characteristics of HD should be discussed and all the alternatives available should be considered.

In other words people at risk to HD should know all the facts before making decisions appropriate to their individual situations.



■ Practical Problems

ROLE CHANGES

Roles within the family are likely to change as the pHD is unable to complete all their previous tasks. Carers/spouses may require support to adapt to increased responsibilities. They may also need financial assistance due to:

- Loss of income
- Cost of care
- Childcare
- Additional food.

Over time the marriage relationship will become altered, and the HD person will be less of a friend, companion and lover. This adds personal grief to a complex situation and both spouse and patient may need support to deal with these challenges.

Although working outside the home brings it's own problems and worries about what is happening in your absence, it can also be a relief from the demands of home and in fact can help you cope with the physical and emotional problems because you are not subjected to them all the time.

SEXUAL AND MARITAL ISSUES

Problems related to sexual adjustment for people with a lengthy illness are of great concern both to the patient and the spouse. These anxieties are often hidden. Even in the midst of the current abundance of information about sexuality, one can find little about sexual adjustment in conditions such as HD.

Hopefully each partner will feel free to discuss their needs with the other. In the words of someone in this situation- "thoughtfulness and consideration can work wonders" Professional guidance may also be sought through the family doctor.

STOPPING WORK

The sufferer's ability to continue working is frequently a critical issue. The length of time somebody can work will depend on the progression of the disease and the kind of job. It may be difficult for the person concerned to admit that he or she can no longer do the job. They may need some help to accept that there is a problem.

Occupations which are potentially dangerous such as welding, or intellectually demanding such as accountancy, may have to be given up earlier than others.



Some employers can be sympathetic and may be able to offer alternatives, though this may not be acceptable to the sufferer in terms of income or status. Your doctor could intervene if, for instance, you were worried about the person driving or where their actions could put themselves or others in danger.

INCOME AND FINANCIAL MATTERS

The employment of the carer is also an important factor. You may already have been the main earner in which case you will have to decide whether to carry on as before while bearing the extra stresses of organising the home and looking after the sufferer.

If you were the secondary or part-time earner before, you will have to think about whether your income will be sufficient to maintain the family, or how you can supplement it. If you were not going out to work at all, you may consider whether a job is an option or whether family

demands are too great. It is advisable to consult your solicitor at an early stage in the illness to discuss your legal affairs, e.g. the making of wills etc.

Normal outgoings can be set against expected income. New expenses may have to be taken into account, for example, extra food, extra heating if the house has not been used in the daytime before. Some people move to a house or ground floor flat which is physically more practical and cheaper to run, though this can be disruptive to neighbourhood social life. Probably some financial sacrifices will have to be made, though different people have different priorities as to which they should be.

OTHER WORRIES

Other important worries about HD exist in relation to insurance, mortgages and so on. If you need information in relation to these or other issues, contact the HDAI office.



■ Care of the Huntington's Patient

DIET

" I am writing this note for the benefit of patients with Huntington's Disease who require higher than normal calorific intake, often up to 4000 kilo calories a day. This is a well recognised requirement in this condition, which is characterised by weight loss. Many patients who manage to gain weight also find that their clinical state improves".

Dr. Niall Quinn, Professor in Clinical Neurology, University of London.

Weight loss in Huntington's Disease has long been observed and frequently attributed to the involuntary movements. The cause of weight loss is still unclear and only theories have been put forward, one being that the weight loss may be caused by an underlying biochemical defect. Whatever the cause the fact remains clear that a greater calorie intake is required.

A normal diet usually takes in 2,000 kcals per day for energy needs. A pHD may require more than twice that. A diet in the region of 5,000 kcals per daily intake is recommended to help promote weight stability.

As the disease progresses, food will need to be cut in small pieces or liquidised to facilitate chewing and swallowing. Hunger and lack of hand control can lead to "cramming" at a time when the patient can least cope with such large amounts. Meals should be smaller but more regular (5-6 per day) with nourishing drinks in between.

Many food supplements, rich in protein and calories are available. They can be taken on their own or added to the patients favourite food. Talk to your G.P. as these supplements may be available on prescription.

Feeding HD patients is a costly business. They need a lot and often. The Community Welfare Officer may be able to help if you need financial assistance towards extra costs involved.

HEALTH PROFESSIONALS

In addition to The General Practitioner (GP), services available from Health Centres include:

Public Health Nurse - The Public Health nurse works from the local Health Centre and can give advice and support on many aspects of daily living. His/her work is preventative and rehabilitative and she/he will provide nursing care (e.g. help with bathing, dressing, skin and basic care). She/he will advise the family on nutrition



and be happy to advise on any problem area. (e.g. incontinence).

Occupational Therapist (O.T) - Will provide advice on aids to daily living and can assess what aids or even structural alterations (e.g. home extensions) may be needed to help the pHD. It is advisable to consult an occupational therapist before paying out money for aids.

Physiotherapist - Physiotherapy can help reduce balance and co-ordination difficulties. It may also increase muscle power and endurance which allows further independence for the HD sufferer.

Speech Therapist - The speech therapist can advise on methods of maintaining communication skills.

Community Psychiatric Nurse - The Community Psychiatric Nurse will be happy to advise the family if the patient has behavioural or psychological problems. The Community Psychiatric Service can be contacted via the local Health Centre. A referral letter from your G.P. is necessary before contacting the Psychiatric Service.

Social Worker - The Social Worker can provide information and advise on various issues.

Home Helps - The Home Help Service offers the family assistance with housework and other duties. A small charge may be made if the family can afford it. A family can be referred to the home help service via the General Practitioner, Social Worker, or Public Health Nurse.

Community Welfare Officer - The C.W.O give the family advice on financial benefits and entitlements.

See Entitlements and Benefits (Page 18)

■ OTHER SERVICES

Meals-on-Wheels - This is a voluntary service in most areas. A hot mid-day meal is delivered to the patient's home. A small charge is usually made. The service is for people who cannot cook for themselves. If a special diet is required it should be requested (e.g. - a minced diet or a high calorie diet).

Day Centres - Many voluntary organisations have day care centres, clubs and various facilities throughout the country which can be accessed by patients or carers. Ask your Citizens Information Centre or Health Centre for relevant information.



CARE OF THE pHD IN THE HOME

The following points should be considered when caring for the HD sufferer at home.

When walking becomes difficult one of the best ways to hold someone with HD when walking around the house is:

- stand in front of him/her (face to face)
- take hold of both the patients' wrists
- then walk backwards with him/her following you.

Walking frames may be helpful in the early stages when the individual is unsteady. Banisters are essential for going up and down stairs. The OT will assess the person at home and arrange for rails and banisters to be put in the hall and stairs. Handrails must be sturdy and firmly fixed.

Coming downstairs one needs to stand behind the person and give support by holding their belt/waist. Information regarding wheelchairs can be obtained from the Irish Wheelchair Association.

TIPS TOWARDS AVOIDING PROBLEMS

Dental care is important as the patient will have difficulty using dentures.

Incontinence occurs in the later stages of HD because control is lost over the muscles of the bladder and bowels. A regular toilet routine (a two hourly pattern) should be observed. Pads, protective mattresses and absorbent bed sheets provide comfort for the patient and eases the workload of the carer.

Clothes should be comfortable and easy to wash and dry. Items of clothing which are easily worn (e.g. few buttons or hooks) allow the patient greater independence.

Safe Environment - Any prospective hazards in the house etc. should be removed, guards placed in front of fires etc. Ensure all electric appliances are safe.

The Community care Team can help in the management of choking, infections, chiropody and stress. They can provide advice on communication & recreation, respite care, day centres and day hospitals.

General Practitioner - If the person is unwell do not always assume it is HD. It may be another problem, e.g. chest infection. It is best to contact the General Practitioner.



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■ Tips to Cope

CARERS

Caring for a sick or elderly person can be very rewarding but it is also very demanding at times. Benefits include: knowing you are doing something meaningful, learning to live in the present and appreciate the simple things in life, re-thinking priorities and values, and an increased insight into your own strengths and limitations. Caring however can be physically, psychologically, emotionally and even financially draining.

You will experience a certain amount of stress but it is essential to manage stress successfully and avoid burnout. Chronic exhaustion, significant weight gain or loss and frequent illnesses are physical signs of high stress levels. Frequent crying, frequent irritation, feelings of hopelessness and inadequacy and difficulty controlling ones temper are emotional signs.

In order to prevent being overwhelmed by stress you should:

- Look after your own physical health: eat nutritiously and get adequate exercise and rest.
- Stay connected to friends and outside activities

- Take time daily to relax by doing something you enjoy for example : read something uplifting, listen to music you enjoy, take a walk or phone a friend etc
- Avoid wasting time and energy on unimportant things
- Acknowledge when you need some help and ask for it!
- Find at least one person close to you who will listen and understand
- Find out about respite facilities in your area
- Take things one day at a time

THE IMPORTANCE OF RATIONAL THINKING

FEAR: can be an unreasonable overestimation of some threat, coupled with the underestimation of your ability to cope.

PANIC: You panic when you are anxious, depressed self critical or upset.

When you feel Fear or Panic - STOP..ask yourself ...

- What am I telling myself to make me feel this way?
- Do I really want to do this to myself?
- Do I really want to stay upset?



Relax or Distract

Do something physical, walk, talk, read or listen to music.

Question the negative belief

- What is the evidence for this?
- Is this always true?
- Has this been true in the past?
- What are the odds of this happening?
- What is the worst that could happen?
- What is so bad about that?
- What would I do if that happened?
- Am I looking at the whole picture?
- What would I say to a friend in this situation?

WORRYING HAS NO EFFECT ON SOLVING PROBLEMS, TAKING ACTION DOES.

FINANCIAL SUPPORT

Entitlements and Benefits - You may need to apply for supplementary benefit if your income is insufficient. Your local Health Clinic, Social & Family Affairs Welfare Office and Citizens Information Centre can provide you with relevant information.

Medical Card - Application for the General Medical Services Card is made to your local Health Board office. This is a means tested benefit but special consideration is given in the case of HD.

Disability Benefit - Disability Benefit is a payment made to insured people who are unable to work due to illness.

Disability Allowance - Disability Allowance is a weekly allowance paid to people with a disability who are aged 16 or over and under age 66. Your disability must be expected to last for at least one year and the allowance is subject to both a medical suitability and a means test.

Invalidity Pension - Invalidity Pension is a payment made to people who are permanently incapable of work because of an illness or incapacity.

Carer's Benefit - Carer's Benefit is a payment made to insured persons who leave the workforce to care for a person(s) in need of full-time care and attention.

Carer's Allowance - Carer's Allowance is a payment made to carers on low incomes who care for a person(s) in need of full-time care and attention.

Respite care Grant - If you qualify for Carers Benefit or Carers Allowance you may be entitled to an Annual Respite Care Grant. This is a once off payment and can be spent as you wish.

Bereavement Grant - A Bereavement Grant is a payment based on PRSI contributions which is payable on the death of: an insured person; the wife or husband of an insured person; the widow or widower of an insured person; a child



under age 18, or under age 22 if in full-time education (where either parent or the person that the child normally lives with satisfies the PRSI contribution conditions); a contributory pensioner; the wife or husband of a contributory pensioner; the qualified adult of a contributory pensioner, including those who would be a qualified adult but are getting another social welfare payment, e.g. Carer's Allowance ; a qualified child or an orphan in receipt of Orphan's (Contributory) Allowance.

ADDITIONAL SOURCES OF INFORMATION

Department of Social Community & Family Affairs - The Department of Social Community & Family Affairs provides information on the relevant supports and services provided by the Department. For general enquiries: Telephone 01-8748444,
Email: info@welfare.ie
Website: www.welfare.ie

The Information Officer in your Social Welfare Local Office gives advice on all social welfare matters. Local offices are listed in the Eircom telephone directory under the Department of Social Community & Family Affairs section.

Department of Health & Children - The Department of Health & Children has overall responsibility for the development of health policy and for the planning of health services. Their website provides information and links to their services: - <http://www.doh.ie>

Information is also available by phone:-01 635 4000 or by writing to Department of Health & Children, Hawkins House, Hawkins Street, Dublin 2.

Healthboard details are listed in the Eircom telephone directory under the Department of Health & Children. Local Health Centres are listed in the Golden pages.

Comhairle - Comhairle is the national support agency responsible for the provision of information, advice and advocacy to members of the public on social services.

Comhairle can be contacted by telephone: 01 605 9000, Fax: 01 605 9099 or email at comhairlecid@comhairle.ie, Their website address is:- <http://www.comhairle.ie/>

Comhairle have developed a user-friendly database called OASIS that provides information about public services. You can access information on healthcare, education, employment, housing, taxation, retirement and much more by logging on to:- www.oasis.gov.ie



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Citizens Information Database - The Citizens Information Database (CID) is an electronic database produced by Comhairle which covers all aspects of civil and social rights and entitlements in Ireland. It was developed as a common resource for all information providers in the independent and statutory sectors.

The Citizens Information Database is directly accessible at www.cidb.ie or by a link from the Comhairle homepage <http://www.comhairle.ie>

Citizens Information Centres - Citizens Information Centres (CICs) provide free confidential and impartial information, advice and advocacy services to members of the public, on their rights and entitlements. CICs are countrywide voluntary organisations, they are stocked with a full range of information on civil and social rights and entitlements and on state and other services. All Key CICs provide specialist services in addition to their generalist information service. Many provide a legal advice service and a specialist financial/ tax advice service, a number provide consumer advice service (in association with the Office of the Director of Consumer Affairs) and others

offer the services of the Ombudsman and Threshold for special housing advice.

Citizens Information Centres are listed in the Golden pages and on the Comhairle and OASIS webpages. You can also find information on your nearest centre by calling 01 605 9000.

Money Advice and Budgeting Service - The Money Advice and Budgeting Service (MABS) is a free and confidential service for people with debt problems and money management problems. Their services include: Help in dealing with debts and making out a budget, examining your income to make sure you are not missing out on any of your entitlements and contacting creditors on your behalf with offers of payment if you are not able to do it yourself.

There are 53 MABS offices in Ireland with trained Money Advisers. You can contact MABS directly yourself by phone, email, letter or by calling in person. You can find out about your local MABS office through Directory Enquiries, Citizens Information Centres or their Web Site address at <http://www.mabs.ie>.



■ Facts at a Glance

Increase Calorie Intake - It is estimated that some pHDs need up to 5000 kilo calories a day to maintain normal weight. See care of the pHd (page 17).

Westphal Variant - A minority of pHDs manifest a form of the disease characterised by muscular rigidity rather than chorea.

Genetic Inheritance - If either parent has the altered HD gene, each son and daughter has a 50% or 1 in 2 chance of inheriting or not inheriting HD (see page 6).

A diagnostic test - Used to confirm the diagnosis of HD in a person showing symptoms and signs of the disease. The test is carried out to confirm a clinical likelihood that a person already has HD.

A predictive test - Is carried out in a healthy person, who has no signs or symptoms of HD, but who has a family history of HD.

Genetic counselling - People who wish to have a predictive HD gene test will have an opportunity to discuss the issues involved in testing, to have their questions answered, and to consider the possible consequences of a predictive HD gene test result.

Adenine, A: One of the 4 bases that makes up the rungs in the DNA ladder.

Base, bases: Two bases join to form the rungs of the DNA ladder. In DNA the four bases are called A, G, C and T.

Cell: The building blocks which make up all living things.

Chromosome: Chromosomes are threadlike structures found in the middle of your cells that contain your genes. Humans have 46 chromosomes in each of their cells.

Code: A code is a system of symbols or letters that represents a message. In DNA the four bases A, G, C and T form a code, telling a cell how to make the proteins it needs.

Cytosine, C: One of the 4 bases that makes up the rungs in the DNA ladder.

DNA, deoxyribonucleic acid: DNA (deoxyribonucleic acid) is the chemical that genes and chromosomes are made of. DNA is a very long molecule that is simply an acid that is found in every cell of your body which has lots of sugar groups (ribo) attached to it and each sugar is missing an oxygen (deoxy).

Gene: a gene is made of DNA. A gene is a recipe telling your cells to make a certain protein.

Gene therapy: adding DNA to a person with a genetic disease to try to make them healthy.

Genetics: Genetics is the science investigating similarities and differences between people and how your genes help determine these.

Guanine, G: One of the 4 bases that makes up the rungs in the DNA ladder.

Human Genome Project: scientists all over the world are working together to write down all the bases that make up the genetic code of humans.

Mutation: Any change in the DNA of someone.

Protein: Proteins are important substances in your cells. They make your cells look and work the way they should to keep you healthy.

Thymine, T: One of the 4 bases that makes up the rungs in the DNA ladder.



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■ How You Can Help!

You can make a donation by forwarding a cheque made payable to The Huntington's Disease Association of Ireland and sending it to:

The Huntington's Disease Association of Ireland,
Carmichael House,
North Brunswick St.,
Dublin 7

Or by Direct Debit to:

Huntington's Disease Association of Ireland Account
Bank of Ireland, Smithfield, Dublin 7
Account No 54757711
Branch Code 90-00-92

Please accept a donation of € _____
for your work

Name: _____

Address: _____



■ Information Request

If you need further information on any issues mentioned in this booklet please advise

The Huntington's Disease Association of Ireland,
Carmichael House,
North Brunswick St.,
Dublin 7



Please forward information on:

Name: _____

Address: _____

Please forward information on:

Name: _____

Address: _____

