

The Juvenile Huntington's Disease Handbook

A Guide for Physicians, Neurologists and Other Professionals

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Table of Contents

Acknowledgments & Dedication

Introduction

Chapter 1-The Diagnosis of HD In A Child

Chapter 2-After The Diagnosis

Chapter 3: Specific Medical Issues

Chapter 4 - Daily Life

Chapter 5 - The Late Stages

Chapter 6 - Financial, Legal & Social Service Issues

Chapter 7 - Caring for the Caregivers

Chapter 8 - Hope for the Future

Appendices

This book is dedicated to Danny, Davey, and Dawn.

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Introduction

Huntington's disease (HD) is a genetic neurologic disorder that leads to a movement disorder, dementia, and behavioral disturbances. Although the genes that causes HD it is present throughout a persons life, though the symptoms do not usually begin until mid-adulthood. In the United States, the prevalence of HD is about one per 10,000 individuals. Although HD has been reported in people of almost every ethnic background, it is more common in Caucasians and less common in people of Asian, Native American, or African origin.

Fewer than 10% of individuals with HD develop symptoms before age 20. Because it is uncommon and differs from typical adult-onset HD, in both neurologic symptoms and ways that it changes the lives of the individuals and families that it affects, juvenile-onset HD presents unique challenges to affected individuals, their caregivers, and the various professionals who are called upon to assist them. Described in this handbook are the symptoms that are most commonly seen early in juvenile HD, as well as the approaches that the Physician can use to make the diagnosis of Huntington's disease.

Many parents, families, and health professionals are uncertain about the role of a genetic testing in the diagnosis of juvenile HD. Genetic testing must be used with particular caution in children, as the presence of the HD gene in a blood test does not mean that the child's symptoms are due to Huntington's disease. Discussion of the appropriate and inappropriate uses a genetic testing for children is provided here; the family or physician should contact a Genetic Counselor to discuss a specific child or situation in more detail.

One of the biggest problems that families face with a child who has HD is providing an educational program that meets the child's changing needs. Some suggestions for how to address these needs, or where to turn for help or more information, are provided in this handbook. Suggestions are also given about how to obtain financial, emotional, and spiritual support. Now, more than ever, advances in research will lead to better treatments for the future.

*Peter S. Harper, Huntington's Disease, NY: W.B. Saunders, 1996.

Chapter I - The Diagnosis of HD in A Child

When To Consider HD

The diagnosis of HD in an adult is usually made in a person who has memory or cognitive changes (dementia), and chorea (dance-like movements), often with behavioral or psychiatric problems such as depression, irritability, or mood swings, and usually with a family history of HD in a parent. The presenting symptoms may be a little different in a child, particularly a child under 10 years of age.

While there is no symptoms or group of symptoms that are absolutely required for the diagnosis of juvenile HD, most affected children have several of these features at the time that the diagnosis is made.

Chorea is uncommon in children developing HD within the first decade, but may be one of the first symptoms in a teenager. Severe behavioral disturbances may be the first symptom in an adolescent.

Typical Initial Symptoms of Juvenile HD:

- Positive family history of HD, usually in the father
- Stiffness of the legs
- Clumsiness of arms and legs
- Decline in cognitive function
- Changes in behavior
- Seizures
- Changes in oral motor function
- Family History

For reasons that only became clear after the gene responsible for HD was discovered in 1993, individuals with a very early onset of HD are far less likely to have an affected father than an affected mother. It is very unlikely for HD to appear in a child whose parent was not also affected with HD. If this situation appears to be present, the physician should consider diagnosis other than HD.

Occasionally an HD-affected child without an affected parent can be explained by the early death of a parent (before the parent's symptoms were evident), misdiagnosis or lack of diagnosis in a parent who was affected, non-paternity (a biological father who is not the same as the apparent father), onset of symptoms in the child before the parent's onset, or adoption.

Documenting the diagnosis of HD in other relatives can be helpful to the physician as the child is evaluated for HD. A parent who suspects that his or her child has juvenile HD can help the physician by assembling information about the family history.

If the family history is missing because the child was adopted, it may still be possible to obtain the missing information if it is important to the child's diagnosis.

Adoption agencies and county or regional social services departments, when given an understanding of the serious nature of HD and its hereditary pattern may be able to contact the birth parents to obtain more information.

Similarly, a mother or family who becomes aware of the family history of HD should be honest and open with the adoption agency, so the adopting parents are aware of the child's genetic risks and are able to plan appropriately.

Declining Cognitive Function

HD is a degenerative condition, which means that children who are symptomatic will begin to lose skills that they have had previously gained.

- In a school-aged child, this is often noticed first as an overall decline in grades or other measures of school performance. Attention and concentration may decline.
- In a younger child, increasing difficulty with previously attained cognitive or motor skills, such as speech, reading, throwing a ball, or riding a bicycle, might be evident.
- In a young child, declining school performance, accompanied by worsening motor skills, would be much more suggestive of HD than a change in grades alone.
- In an adolescent, many other common causes of poor school performance must be considered, including depression, drug or alcohol use, or disruptions of family or social life. Detailed information from teachers and school counselors may help the physician to pinpoint the different kids and causes of dysfunction at school.

Behavioral Disturbances

Behavioral disturbances are common in children with HD. Behavioral disturbances, depression, and attention deficit disorder-hyperactivity are also common in children without HD. The families of children at-risk for HD are often under significant financial and social stress, which increases the chance of social or behavioral problems in the children.

- In younger children, aggressive or disruptive behavior related to HD is usually seen along with changes in cognitive function and motor disturbances.
- In adolescents, behavior disturbance may be the first and only presenting symptom of HD. Behavior problems in these children are often very severe, leading to psychiatric hospitalization, suspension from school, or sexual promiscuity, physical or sexual abuse of younger siblings, severe drug or alcohol abuse, and depression with suicide attempts.

Rigidity

While adults with HD usually have involuntary movements, and only develop rigidity and dystonia (abnormal stiff postures) much later in the disease, children are likely to have stiffness of the legs, walking on the toes, or scissoring of the gait as initial or early symptoms.

Clumsiness of hand and arm movements, thickness of speech, drooling and poor oral motor control are also likely, particularly in very young children.

The earlier the symptoms begin, the less likely the child is to have chorea at any point in the course of the disease. On the other hand, the older the child, the more likely he or she is to have chorea as a presenting symptom.

Seizures

Seizures are said to occur in about 25% of children with juvenile HD, and may be a presenting symptom. They may be of any type, and they may or may not be severe.

The physician should never simply assume that seizures are caused by HD; any child with a seizure should have cerebral imaging studies and an electroencephalogram (EEG), as well as appropriate laboratory studies to rule out metabolic causes such as low blood sugar or poison ingestion.

This brief overview only describes the most typical presenting symptoms of HD; each child is unique and may have additional symptoms or symptoms that differ from the typical features described. It may take several visits to the physician or several neurological examinations before mild or intermittent symptoms are recognized by the physician as being due to HD.

Seeing The Physician

In many health care systems in the United States, a patient must first see a family physician or pediatrician to obtain a referral to a pediatric neurologist or HD specialist. The diagnosis of juvenile HD can be challenging, and it is appropriate to request such a referral.

In some localities, an adult neurologist specializing in movement disorders or HD may be more helpful than a pediatric neurologist who is unfamiliar with the condition; a telephone call from the referring pediatrician to the specialist can help the family to avoid unnecessary appointments.

A parent should understand that diagnosis of juvenile HD is unlikely to be made immediately at the first visit, and should be prepared to work with the physician through the process of diagnosis.

The physician will first take a medical and neurological history, a family history, and a development history, and perform a neurological examination. It is helpful for the parent to bring records to review, including any previous neurological examinations, psychological evaluations or school testing records.

It is important for the physician to review the neurological, behavioral, and functional problems before considering an HD gene test. If cognitive changes are present, a formal neuropsychological assessment (tests of memory, development skills, and intelligence) can document areas of strength and weakness, suggest strategies of management, and serve as a baseline for comparison later.

Either behavioral changes or changes in school performance in a child at-risk for HD should prompt the physician to evaluate the child's psychosocial situation and to make appropriate referrals for individual or family counseling, county child protection services, school-based programs, or social services.

Physical, occupational and speech therapist can perform baseline assessments of motor with an emphasis on how the child is able to function in school and at home.

Children in HD families are *not* immune from having developmental delay, attention deficit disorder, mental retardation, or other medical or neurological conditions entirely unrelated to HD, and both parents and physicians should consider these possibilities as they go through the diagnostic process.

Brain imaging, by computerized tomography (CT) or magnetic resonance imaging (MRI), is often normal early in the course of HD, and may help to rule out other conditions.

Similarly, routine blood tests, while not helpful in securing a diagnosis of HD, can help to rule out other diseases that can cause abnormal movements, such as hyper- or hypothyroidism, toxin or drug ingestion, systemic lupus erythematosus, or recent streptococcal infection (Sydenham's chorea).

When the history, examinations, and initial laboratory evaluations are strongly suggestive of HD, a genetic test may be the most efficient and accurate way to confirm the diagnostic impression.

If the diagnosis is not evident from the examination, then it is not appropriate to have genetic testing performed immediately, as the presence of the abnormal HD gene does not resolve the problem at hand, which is whether the person's symptoms are due to HD.

It is *extremely important* to limit diagnostic testing of the HD gene to children who clearly have clinical symptoms and a course that is consistent with HD.

Often it is recommended that a child be evaluated by the neurologist or other clinician twice, six to twelve months apart, to determine whether the initial symptoms have remained or progressed despite appropriate management, before the HD gene test is performed.

This strategy makes it less likely that a child with temporary or nonprogressive symptoms would be tested prematurely. The table shows some of the risks of identifying a child as an HD gene carrier prematurely.

Potential risks of premature gene testing in a child

- Incorrect attribution of symptoms to HD
- Failure to make the correct diagnosis
- Stigmatizing the child
- Insurability
- Employability
- Psychological effects on the child
- Social effects on the child

Diagnostic Genetic Testing In Children

The ability to detect changes in the HD gene itself has made confirmation of the diagnosis of HD much simpler. The HD gene can be isolated from a blood sample, and examined chemically; the abnormality in the gene that causes HD is called a "CAG repeat expansion." The HD gene normally has a variable number of "CAG repeats" - any number up to 35 repeats is normal.

There is a relationship between the repeat numbers and the age that HD symptoms begin, so that higher repeat numbers are associated with younger ages of onset. Most adults with HD have between 40-50 CAG repeats in their abnormal HD gene. Usually, juvenile HD is associated with CAG repeat numbers of 50 or higher, although it is not possible to define rigid boundaries. Very young age of onset are associated with very high CAG repeat numbers; children with HD onset at age 2-3 years and with over 100 CAG repeats have been reported.

The gene test is close to 100% accurate. If the tests show two normal HD genes, the child will never develop HD and is not at-risk for passing HD on to his children. If the test shows an abnormal HD gene, the child will someday develop HD.

The gene test, however, cannot predict when a particular person's symptoms will begin. Occasionally, individuals will have a CAG repeat length in the 36-39 range, which may or may not be associated with the development of HD symptoms during a normal life span. Results in this "intermediate range" are not usually a factor when testing children for possible symptomatic HD.

And rarely, very high CAG repeat numbers (over 100 CAG repeats) are not detected by the standard gene test. If HD is strongly suspected in a very young child who has a normal gene test results, the physician may want to contact the laboratory or a genetic counselor to discuss the possibility of a special analysis to look for very large CAG repeat numbers.

The potential risks of testing a child for the HD gene inappropriately or prematurely cannot be emphasized enough. Individuals who develop HD have the CAG repeat expansion in one of their HD genes from the moment of conception - years or decades before their symptoms begin.

There are two ways in which a premature gene test can be misleading or damaging to a child's care. First, an abnormal gene test result may incorrectly be assumed to "explain" a child's symptoms, when in fact the symptoms are not clearly related to HD. For example, an adult whose gene test was felt to "explain" his blurry vision and headaches was incorrectly diagnosed as having HD and, as a result, the diagnosis of his pituitary tumor was delayed.

Secondly, it is possible that a gene test will show a small CAG repeat expansion, one that is likely to be associated with adult-onset HD but not juvenile onset HD. This is equivalent to a predictive gene test and does not help to explain the child's current symptoms.

Due to the very sensitive nature of the gene test results, it is important for counseling to occur before the results are given, so there are no misunderstandings about their significance. If the physician is unable or does not have the time to explain the gene test in details, a genetic counselor may be asked to help with this part of the process.

Testing Children Who Do Not Have Symptoms

When one child has been diagnosed with HD, parents may want to have their other children tested as well. Testing of a person who does not have symptoms of HD is called predictive testing, in order to distinguish it from diagnostic testing for a person who has symptoms suggestive of HD.

Although, at first thought, it may seem reassuring for parents to find out that their other children do not carry the HD gene, it is very important to consider the complex potential effects of the tests results on the entire family, as well as on the individual or individuals being tested.

The risks of premature genetic diagnosis of HD have already been discussed and genetic testing experts believe that predictive tests should be reserved for individuals who are able to understand the potential risks and benefits of the test and who are able to give informed consent.

Experience in the United States has shown that most adults at-risk for HD do not choose to undergo predictive genetic testing, so a parent who requests predictive testing for a child is most likely doing something that the child would not want if he or she were able to make the choice.

In addition, at this time, there is no medical advantage to knowing that someone carries the HD gene - treatments that prevent or delay the disease have not been developed yet. For all these reasons, most genetic professionals in North America decline parental requests for predictive tests on their asymptomatic children.

Occasional exceptions might be made for adolescents in adult situations, such as an "emancipated minor" or a married teenager.

In the United States, potential adoption is not usually felt to be an appropriate indication for HD predictive testing, because of the potential for social, financial, educational, insurance and employment discrimination based on the test results and the lack of medical treatment or care to balance the potential social harms. Other countries may have different practices.

Steps in the diagnosis of juvenile HD

- History of change in motor, behavioral and cognitive function
- Family history of HD

- Abnormal neurological examination
- Abnormal neuropsychological tests results
- Genetic test confirming the presence of an abnormally long "CAG repeat" sequence in the HD gene
- Family counseling

Chapter 2 After The Diagnosis

The Expected Course

HD is a chronic condition. At the present time, there is no cure and no medication that is known to slow down the progression of the disease. Treatment is directed at specific symptoms, which means that each person is likely to be treated a little differently.

HD progresses slowly over a number of years, so that the affected individual has more and more difficulty controlling his/her movements and increasing dementia. After a number of years, often a decade or more, affected individuals lose the ability to walk and speech becomes difficult or impossible to understand.

Swallowing problems become complicated and weight loss is common. The ability to perform personal care such as dressing, feeding, and bathing is gradually lost. After a number of years children (who by then may be adults) require 24-hour supervision and care.

Death usually comes 10-20 years after symptoms begin. Some children, particularly those with a very young onset age, follow a more rapid disease course over a shorter number of years. Physicians cannot predict, at the onset of the disease, which child is likely to have a longer or shorter disease duration; only by following a child over time can a more specific prognosis be given. (4)

Assembling A Team of Care Providers

One important way to plan ahead for the child with juvenile HD is to assemble a team of medical care providers. The child with HD is likely to need the services of a neurologist, psychologist or psychiatrist, a physical therapist, occupational therapist, and speech therapist as the years go by. But because HD has a slow course, it is also important not to neglect the child's general medical and dental health.

There are few "experts" in juvenile HD, but all pediatricians have seen children with chronic diseases, all speech therapists have seen patients with indistinct speech and swallowing problems, and physical therapists work daily with children who have difficulty with walking and coordination.

Effective Team Management Is Likely To Include:

- Diagnosis and general medical care
- Genetic test and counseling
- Neuropsychological test
- Drug therapy
- Diet counseling
- Physical and occupational therapy / speech-language pathologist
- Social services, including end of life planning

The family and medical team should work together to set reasonable goals and expectations, to plan ahead so that changes that are expected during the course of HD do not come as a surprise, and to manage or treat the symptoms that can be treated.

The Huntington's Disease Society of America [HDSA] has established regional Centers of Excellence for Family Services and Research, where the services needed by HD families can be obtained easily and conveniently in one location. However, a thoughtful, creative and aggressive physician can provide excellent care in any location if the physician and family work together. For those beyond the reach of an HDSA Center of Excellence, finding one local pediatrician who listens and is willing to learn may be more useful to some families than traveling long distances.

Care Plan For Juvenile Huntington's Disease

The goals of the team management for HD are:

- to provide an accurate and timely diagnosis
- to anticipate the changes that occur during the disease
- the aggressive management of the complications of the disease.

Managing Your Own Emotions

Many emotions run through the mind of a parent whose child has been diagnosed with HD. Anger, despair, depression, shock, disbelief, and hopelessness are among the common emotions parents have expressed. In order to provide properly for the affected child, the parent must find ways to manage his or her own feelings. It is helpful to recall that HD is slowly progressive – just because a diagnosis was made today or last week does not mean that the child's neurologic function will be different tomorrow, or even next month.

Parents must look beyond their child's physician for their own emotional support. Some fortunate parents have supportive family members who can help with either practical details, emotional support, or both. For some, friends and coworkers can provide the primary support; others find that their religious beliefs or church provide comfort and solace. In the medical system, psychologists, or family counselors can provide emotional support and practical approaches to managing difficult situations.

While it is unlikely that any community will have a juvenile HD support group, many larger communities do have neurologic diseases. In the global community in which people now live, email and computer chat rooms may provide a vital link to others around the country or world who are experiencing similar challenges.

The Rest of The Family

The child with HD does not live in isolation. More than many diseases, HD is truly a family disease. The affected child has parents whose lives are intimately affected by the diagnosis, and often there are other children at home. There is frequently an extended family. All of these individuals are likely to have opinions, Beliefs, misunderstandings, and helpful (or not so helpful) advice. Explaining to these family members the meaning of the child's diagnosis, and engaging them in improving the life of the affected child, can be a challenge in itself.

Typical Scenarios

Three family scenarios are common.

Single Provider

Since juvenile HD is most likely to have come from a father who himself is likely to have been affected at a relatively young age, the family of a child with juvenile HD is often headed by a single mother. The father may be deceased, divorced from the mother, or affected with HD to the extent that he is not working or no longer living at home. In any of these cases, the mother may be struggling financially,

psychologically, and socially, even prior to the child's diagnosis, and may not have a spouse to turn to for support.

Adopted Child

Another common scenario is that of an adopted child. In this situation, the family structure may be more intact, with two parents present to support each other, and other children in the family who may not be at-risk for HD.

A Relative

Finally, some families are headed by a grandmother who may already have seen two or three generations of HD in the family before the child comes to her care.

Telling Your JHD's Siblings

There is not a single "right way" to tell the other children about a child's HD diagnosis. It is important to provide children with correct information, however, or their imaginations or friends will supply them with misinformation. In particular, children of almost any age can be told that their sibling has HD, that it is not the same thing as AIDS or cancer, that it is not contagious, and that having HD doesn't mean that the sibling is dying soon.

Children are often more aware of problems than parents realize, but if parents do not talk to them about the problems, the children will quickly learn not to ask.

As the children grow they may have more detailed questions about HD, or realize that it is a genetic disorder that could affect them as well. If the parent is unable to answer a child's questions, ask the physician or genetic counselor for help or encourage the child to look for information from a reputable source on the Internet (such as the HDSA website at www.hdsa.org).

Telling Other Relatives

Telling other relatives about a child's diagnosis of HD is also a challenge. Each family is different; many still hide the knowledge about HD and may not welcome an open discussion. This approach is unproductive. Families should be encouraged to address this problem with openness, knowledge and understanding.

If requested by the family, the physician or genetic counselor can conduct a group counseling session with several family members to efficiently answer all of their questions about the child's illness at the same time.

With an understanding of what HD is and how it affects a child, family members will be better able to understand what the child and immediate family are experiencing, and will then be in a better position to help them.

Chapter 3 – Specific Medical Issues

The Movement Disorder

HD is classified by neurologists as a “movement disorder.” Certainly in adults, the most obvious symptom of HD is chorea, the involuntary, irregular, fidgety or jerky movements present in the arms, legs, trunk, neck, or face.

Some older children (teenagers) with HD develop chorea, but in children who develop HD symptoms before the age 10, chorea is often completely absent. Instead, young children develop stiffness of the limbs, which is usually most severe in the legs. The child may begin to walk on his toes, lose control of balance when running, hopping, or bicycling, or develop a scissoring or stiff-legged gait.

Other children have a noticeable loss of control or oral motor function, with slurring of speech, difficulty swallowing, or drooling. These symptoms are due to an increase in muscle tone, called rigidity or spasticity.

Treatment

No medication can improve control of voluntary movements. However, physical therapy and occupational therapy consultation may be useful at certain stages to address specific issues. Even a child with marked memory trouble may benefit from practicing a simple “touch-turn-sit” maneuver to keep from missing the chair. This simple instruction can help the child who has frequent falls.

Early intervention is important with all therapies, even before it appears that the intervention or device is actually necessary.

Assistive devices can be prescribed to help with dressing, walking, and feeding. A speech therapist can recommend exercises and strategies to make speech more intelligible, and to help the child and family learn how to use augmentative devices (such as communication boards or computerized devices) as speech becomes more difficult.

Early intervention is important with all therapies, even before it appears that the intervention or device is actually necessary. Because cognitive decline is a major part of HD, a child may be better able to learn to use a device or therapy if it is introduced or practiced while he is able to understand and cooperate more easily. Then, as the need for the device or therapy becomes more evident, it is a familiar or well-rehearsed thing which can be easily incorporated into the daily routine.

Home Assessment

The family is encouraged to make its own assessment of the home situation, to review catalogues of medical or therapeutic devices, and to find out from friends and acquaintances what adaptations have been helpful to them. Physical and occupational therapists are familiar with the most commonly used devices, but may not think of every item that might be of use to a particular family or individual.

Just as one might discuss medical treatments with the doctor, the family may bring a list of ideas to an appointment with the therapist for devices they have heard about or are considering purchasing. In this way, the family and the therapist can work together to find ways to improve the quality of the child’s life.

Public health nurse home visit

Dietary consultation

- Assessment of ideal body weight,

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| <ul style="list-style-type: none"> • Room-by-room assessment of the home • Attention to mobility • Safety • Assistive devices • Need for interior/ exterior remodeling (e.g. to accommodate a wheelchair) • Accessing emergency services | <p>dietary habits, food preferences</p> <ul style="list-style-type: none"> • Identification of unhealthy food habits, obsessions, or food-related behavioral problems • Instruction in preparation of modified diets • Monitoring of weight and calorie intake • Provide information about parenteral tube feedings |
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Medications

A number of medications have been developed to treat rigidity and spasticity, two forms of stiffness which are often present in a child with HD. Unfortunately, these medications have been found to be less effective in children with HD than in other conditions.

It is always important for the parents and the physician to be clear about what the goals of therapy are.

Medications can certainly be tried, and some children will have dramatic responses, but it is always important for the parents and the physician to be clear about:

1. what the goals of therapy are
2. how long the medication is to be used
3. when the effects of the medication will be checked, and
4. what to do if the medication causes undesirable side effects.

Rigidity

Medications which may help to reduce rigidity include anticholinergic drugs, such as trihexyphenidyl, and carbidopa-levodopa. Levodopa is likely to worsen chorea in those who have it and must be used very carefully for that reason.

Spasticity/Dystonia

Antispasticity drugs include lioresal, tizanidine, diazepam, and dantrolene, all of which must be used carefully because of the potential side effects. If rigidity or dystonia (a fixed posturing of a limb or other body part) affects a particular small muscle and interferes significantly with function (for instance, if jaw dystonia prevents chewing or opening of the mouth and therefore interferes with eating), local injection of botulinum toxin into the affected muscle might be attempted.

Chorea

For children who develop chorea, dopamine-blocking agents such as haloperidol, risperidone, olanzapine, or pimozide can be used, though these agents tend to make rigidity worse

Physical Therapy

Physical therapy is particularly important to children with rigidity, spasticity or dystonia. Daily exercises are recommended to maintain range of motion. Heat, massage, and stretching may help to loosen muscles with spasticity or rigidity. A therapist can also recommend exercises to maintain strength, as well as braces and other devices to keep an affected individual walking as long as possible. For children who develop chorea, dopamine-blocking agents such as haloperidol, risperidone, olanzapine or pimozide can be used, though these agents tend to make rigidity worse.

Maintaining aerobic capacity is also important for breath support for speech and to clear food and secretions from the throat. A therapist can also recommend exercises to maintain strength, as well as braces and other devices to keep an affected individual walking as long as possible.

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| <p><u>Physical Therapy in juvenile HD</u></p> <ul style="list-style-type: none"> • Home/school exercise program • Gait evaluation • Management of rigidity and spasticity • Safety evaluation • Training in the use of assistive devices • Seating for the classroom • Reassessment as the disease progresses | <p><u>Occupational therapy in juvenile HD</u></p> <ul style="list-style-type: none"> • Safety evaluation • Assessment of activities of daily living • Assistive devices <p><u>Speech therapy</u></p> <ul style="list-style-type: none"> • Outpatient or home speech therapy/oral exercise • Training with assistive devices • Assessment of dysphasia • Strategies to avoid choking • Modification of food textures and types, eating/feeding strategies |
|---|--|

The Cognitive Disorder

Unlike many adults, most children undergo frequent assessments of cognitive function in the form of school tests and standardized developmental and achievement batteries, so cognitive dysfunction is usually detected relatively quickly. Although there are many ways that cognitive problems may affect daily function, some of the most common first symptoms include difficulty starting or completing a task, or doing all the steps of a task in the right order. Anyone who has ever raised a teenager knows that these are also common symptoms of adolescence—parents, teachers, and physicians must be careful not to place too much significance on any single symptom, but rather to look for a pattern of change that suggests that HD has begun.

There are no medications that improve cognitive function. However, many of the medications used to treat mood, aggressive behavior, or other aspects of HD can have side effects that interfere with alertness; this potential risk must be evaluated whenever the use of a medication is considered. Just as exercising helps muscles to keep their range of motion and strength, continuing to "exercise" the mind helps the child to maintain a higher level of cognitive function for as long as possible.

Public schools are required to provide education for children with handicaps of all types. As soon as a diagnosis of HD is made, the family should meet with the school representatives to create an Individual Education Plan (IEP) for the child. This plan must take into account not only the child’s cognitive skills, but also the motor, behavioral, and social factors that can influence school performance. Because HD is a progressive disease, the educational plan must be reviewed each year with new goals appropriate to the

child's age and stage of the disease. The physician, or this handbook, may help teachers and other school officials to understand what problems to expect with a child who has HD, and the expected period of time over which these changes will take place.

At home, the family can strive to make an environment that is stimulation and safe for the child. Routines are very reassuring to a child who has memory problems. Establishing safe routines early in the course of the disease can be helpful. For instance, making a clear habit of eating nutritional foods may help to prevent food obsessions or battles before they start. Moving the child's bedroom to the ground floor near the bathroom, incorporating safe swallowing techniques, and initiating an exercise program, may all be better accepted by the child earlier if they are introduced. If a child is likely to need appliances such as a wheelchair, electric toothbrush, helmet, or communication device or board, it is better to introduce the items before they are absolutely necessary. By the time they are needed, the child may not be able to learn how to use them correctly.

Behavioral and Psychiatric Issues.

Depression

Depression is the most common mood disturbance in children with HD. Depression can occur abruptly, in response to some upsetting life event, or more gradually. In addition to the obvious symptoms of sad mood, tearfulness, loss and a feeling of hopelessness or worthlessness, people who are depressed often show changes in sleep, appetite, energy level, or overall performance. Particularly in a child, who may not have verbal labels to describe his/her feelings, any significant change in sleep habits (either too much or too little), change in appetite or weight (in either direction), lack of interest in previously enjoyable activities, or poor performance at school or work should lead parents or a physician to consider the possibility of depression.

Parents should not be afraid to ask their child or the physician about the possibility of depression, as there are a number of ways to treat it and, if left untreated, it can be life threatening. Sometimes the primary physician will feel comfortable treating a mild depression with medications, while at other times counseling may be more appropriate, or the two can be used together. Severe depression, or depression in a young child, requires the specialized services of a child mental-health specialist. On rare occasions, even young children have required psychiatric hospitalization because they represent a threat to themselves or others. Severely depressed individuals should be asked whether they are having suicidal thoughts or plans. Asking about suicide does not plant the idea in the person's mind, but may provide a welcome opportunity for a young person to discuss troublesome thoughts.

The medications most commonly used to treat depression are the selective serotonin reuptake inhibitors, or "SSRI's." Fluoxetine (Prozac), sertraline (Zoloft), paroxetine (Paxil), and citalopram (Celexa) are most commonly used. Other newer antidepressants, which may be preferred by a particular physician, or in certain clinical situations, include bupropion, venlafaxine, nefazodone, and mirtazapine. If the depression is severe enough to have accompanying agitation, delusions, or hallucinations, then antipsychotic medications such as haloperidol, quetiapine or olanzapine to be added to the antidepressant. The initial dose of any of these medications depends on the age and size of the child, and can be adjusted according to response and side effects.

"After Davie found out he had HD (he was 14), he turned into a child from hell. Don't take me wrong, I love my son with all my heart and soul but his motto in life was 'if I'm not going to live I'm going to do it all,' and he did. From 14 to 18 Davie spent most of his time in and out of juvenile facilities for drugs, drinking, and trying to commit suicide. He was very sexually active and it scared me to death."

And occasional child will have what appears to be the opposite of a depressed mood. When an excessively elevated mood alternates with depressed mood, the term "bipolar disorder" may be used. In the time, elevated mood, a child may require very little sleep, may be buzzing with activity (although often getting little accomplished), and may be grandiose in plans, easily excitable, or irritable. This type of mood disturbance can be brought on or made worse by the use of stimulant drugs such as cocaine or amphetamines. Elevated mood can be treated with mood stabilizing drugs such as valproic acid, carbamazepine, or neuroleptic medications, all of which are preferred over the old drug, lithium.

Aggressiveness and Impulsiveness

Aggressive, violence and compulsive behaviors present the greatest management problem in children with HD. As children become teenagers, their rapidly growing bodies are confronted by the normal drives of sexuality, aggression, and independence, while at the same time HD is removing their ability to control these urges.

The control of aggressive, impulse, or violent behavior requires a multifaceted approach. The first concern should be the immediate safety of the individual or individuals who are at risk when the behavior occurs. Family members should know how to access emergency services when an explosive behavior occurs and should establish a plan to use these services in the time of crisis. Many families have devised plans that include, as needed: use of medications, a special room or rooms where the affected individual or other family members go for a specified period of time when a crisis is declared, a friend or neighbor who is called upon to remove one or another party from the scene of the crisis, or the threat of calling the police when a specified behavior occurs. Other families remove or lock up medications, guns, automotive and cleaning products because of suicide attempts, which often occur impulsively (for instance, after watching a television program showing a person committing suicide in a like manner).

The next step is to identify any factors that tend to trigger aggressive or explosive behaviors and address those factors or situations. If behaviors occur only at mealtime or bedtime, only in the presence of a certain care provider, or because of an obsession with soda, a certain food, sex, or cigarettes, then perhaps a compromise can be achieved. Reducing the frequency of baths, addressing food preferences or fears of choking, changing care providers or at least determining what it is about the care provider that triggers an inappropriate behavior, and administering treats or soda as rewards for good behavior are all the strategies that might be beneficial.

Many children with late staged HD have an impaired sense of time – telling the child that something will be done in 10 minutes might not mean much to the child. Setting a timer, and indicating that when the timer rings the activity will be done, may be helpful to both caregiver and child. Similarly, memory can be so poor that the individual does not recall that he just had a soda or cigarette 15 minutes earlier. Experienced parents advise that it is important to "pick your fights." If a situation escalates towards a crisis, it may be wise to back off and let it go - give the child and the caregiver both a break.

Families may be unable to identify specific situations that trigger aggressive or violent behavior and even if they do, the behaviors may not be controlled simply by changes in the environment as those described. For their safety, and that of others around them, aggressive, impulsive, or violent children may require medications to control their behaviors. Commonly used medications in this situation include valproic acid and carbamazepine, beta-adrenergic blocker drugs such as propranolol, sedatives such as lorazepam or clonazepam, and neuroleptic medications such as haloperidol, risperidone, olanzapine, or thiorazine. Each of these medications have potential side effects and none is effective in all children with behavior problems. Families should not rely on medications alone to control behavior; therapy with a skilled counselor or child's psychiatrists can help both the family and the child to understand and control the behavioral problems better.

In some children, symptoms of Attention Deficit Disorder may be present and may trigger problematic behaviors. In these children, treating Attention Deficit Disorder may lead to improvements in behavior. Similarly, treatments of an underlying depression may help to decrease violent, angry, or aggressive behavior. Many children are very sensitive to medications. Starting a new medication in a safe, quiet, control environment may be important to the success of the treatments.

When behavior is entirely out of control, psychiatric hospitalization may be necessary. This allows the child time away from the environment where he or she was having trouble, and allows time in a safe environment for medications to take effect. When behavior is chronically or unpredictably dangerous, particularly if other children are at risk for harm, it may be best to remove the child with HD from the home. Often, individuals with this type of severe behavior problems are older adolescents or young adults for whom other placement was already at consideration. For reasons that probably have no more to do with how children behave around their parents than with HD, some children or young adults show marked improvement in their behavior as soon as they're no longer at home with the parents!

Obsession

Obsessive thinking is very common in individuals with HD, and it may interfere significantly with the daily routines. This behavior is caused by changes in the brain that occur with progression of the disease. When the thoughts of the affected person and the requests or plans of the caregiver come into conflict, aggressive behavior often results, as noted in the previous section. Medications such as the SSRI's or clomipramine are used to suppress obsessive thoughts and behavioral strategies are often employed. By identifying what the obsession is about and why and when it occurs, it may be possible to reassure or distract the child or to use rewards or other strategies to satisfy the urge or obsession while also accomplishing whatever other tasks need to be done. As psychologist or psychiatrist may be needed if the problems are not easily managed by the caregiver.

Sexuality

Adolescent is a difficult time even for children who do not have HD. Managing a changing physical appearance, new and confusing sexual urges and desires, learning how to interact with peers who are undergoing similar changes, and moving away from relationships with parents into strong relationships with other adolescents or adults are all tasks for any teenager. Facing these challenges with a disability that diminishes the ability to communicate and to understand new information, and leads to impulsive or disruptive behavior, it is far more challenging.

For girls, the first challenge may be managing menstrual hygiene. Depending on how advanced HD is when menses begin, gentle and repeated counseling and assistance from a female nurse or relative may help the girl to understand the feelings that accompany maturation, treatments that are available for bloating and menstrual cramps, and when and how to use menstrual hygiene products. All girls should have a private discussion about what sexual activities include, how to prevent pregnancy, when sexual activity is inappropriate, and how to obtain help if problems arise. Group discussion in high school health class may proceed to quickly for a girl with HD to process all the important information or to ask questions. A school may be obliged to provide a personal care attendant if a girl is judged to be particularly vulnerable to the sexual or physical advances of others in the school.

Contraceptive devices or medications should be made available to girls with HD. Contraceptive patches or long acting injections (e.g. Depo-Provera) may be preferred to pills or devices that must be used daily or at that time of a sexual encounter. Depending on the social or clinical situation, some affected individuals may want to consider sterilization procedure. Early attention to these issues is important as many adolescents with HD are sexually promiscuous but not fully aware of the potential consequences of their sexual activity.

Boys with HD are also potentially vulnerable to sexual or physical abuse and their own impulses and aggressive behavior. Boys who are sexually or physically abused or threatened by their peers should be offered the same protection that vulnerable girls would be offered.

Boys who behave inappropriately may need both behavioral modification strategies and medication to manage their sexual urges and impulse of behaviors. For example, a boy who masturbates in public can be encouraged to use private areas such as his bedroom or bathroom, but the door closed, and be rewarded when the undesirable behavior stops. Impulse of or aggressive sexual behaviors can be severe in adolescent boys with HD. Antipsychotic medications such as risperidone, olanzapine, or haloperidol, seizure medications such as valproic acid or carbamazepine, sedatives such as diazepam or lorazepam, or beta-adrenergic blockers such as propranolol have all been used to curb aggressive behavior. When severe aggressive sexual behaviors are threatening, evaluation by a psychiatrist or counselor experienced in the treatment of sexual or conducts disorders, may be necessary, and in-patient treatment may be required. Anti-testosterone drugs can be considered for patients with severe and recurrent sexual behavioral dysfunction, but this is never a first choice.

Hallucinations

Hallucinations are uncommon in individuals with HD. When hallucinations occur for the first time, a thorough medical and psychiatric evaluation is appropriate. Euthanasia can be auditory (such as a hearing voices, which may simply make comments or may command the individual to do thing), visual, or sensory. They can occur in individuals with severe depression, as a result of prescription medications, or because of the use of mind altering drugs (such stimulates or hallucinogens). In the later stages, individuals with HD can easily develop delirium in response to systemic illnesses of any type, changes in medication, or other sources. Delirium, an acute and reversible change in mental function, may include severe confusion, combativeness, or hallucinations, and often requires treatment with high doses of medication.

The mainstays of treatment for hallucinogens are the antipsychotic or neuroleptic drugs. Older antidepressant drugs include haloperidol, thiorazine, and fluphenazine, among many others. A newer group of more "selective" antipsychotics, designed to have fewer side effects, includes risperidone, olanzapine, clozapine, and quetiapine. Which antipsychotic is used and in what doses depends on the previous experiences of the patient or physician and any other concerns, such as side effects or costs. Only a few of the older antipsychotic medications can be given intravenously or intramuscularly, so the choices are limited for an individual who is unwilling or unable to take oral medications.

Seizures

Seizures occur in about 25% of children with HD, but are uncommon in individuals with adult-onset HD. This is probably because the developing brain of a child is more likely to develop seizures in response to an insult or injury than the adult brain.

Seizures occur in about 25% of children with HD, but are uncommon in individuals with adult-onset HD.

The physician should never simply assume that a first seizure is due to HD. Children with HD are not immune to other reasons for seizures such as infections, brain tumors, diabetes, or traumatic brain injuries. Certain prescription medications can cause seizures, as can illicit drugs such as cocaine. The proper treatment of seizures first require that these possibilities be ruled out by blood and urine studies and brain imaging (MRI is strongly preferred over CT because it shows more detail). An electrocardiogram (EEG) to be performed, as the electrical characteristics of a seizure disorder can help to guide treatment.

The word "epilepsy" refers to an ongoing tendency to have seizures. Children with epilepsy usually need medication to reduce their chances of having seizures. Children with HD that have seizures typically have generalized or myoclonic epilepsy, although other seizure types can occur (focal or partial complex seizures).

Valproic acid is considered to be a first choice for the treatment of either general or myoclonic seizures, but clonazepam or lamotrigine can also be used. A number of other medications might be appropriate, depending on the circumstances. The selection of a seizure medication should be made carefully. After an evaluation has been completed, the family can help the physician to manage a child's seizure by:

- 1) keeping a log of seizure activity
- 2) becoming knowledgeable about the medicine that the child is taking (the pharmacist can often provide a computer printout with information about the medication)
- 3) in showing that the child is taking the medication exactly as directed, and
- 4) working with the physician to decide the goals of treatment, the medication dose, the length of time for trying to medication, and what to-do if there's a problem.

All the seizure medication can have serious side effects but proper use makes these side effects occur much less often. Epilepsy often decreases in severity of the child grows older, so seizures that were once difficult to manage might become less of a problem later. There is not a particular time or stage of juvenile HD when seizures are more or less like to begin. For the rare child's seizures cannot be managed despite careful use of the standard drugs, referral to a regional epilepsy specialty center may be helpful.

Ways to manage seizures

- Keep a log of seizure activity
- Become knowledgeable of child's medications
- Ensure medication is taken as directed
- Work closely with the physician on goals of treatment

Oral Motor Problems

Choking

Choking, or dysphagia, is an expected complication of HD. While there are no medications that can prevent dysphagia or improve swallowing, a speech pathologists can help a parent or child to understand what causes this swallowing problem and can often provide simple practical tips to help minimize choking. A chin-tuck maneuver, for instance, can help to direct food into the esophagus (extending the head backwards, as actors are often seen doing in soda or beer commercials opens up the airway and makes choking more likely!). Alternating solids and liquids can help to clear the mouth and throat of food particles that were not completely slow look first time. Using a straw or a cover cop can help to avoid spills and limit the amount of liquid taking with each swallow, thereby reducing the risk of choking. Minimizing distractions when the child is eating can also be helpful.

"She can still feed herself certain foods, but prefers that a family member feed her. She is totally dependent on family for any and all activities. All of her friends have deserted her. Her parents have no social life outside the nursing home."

The speech pathologist can also suggest ways to change the texture of the diet to adapt to a particular child's dysphagia. For instance, commercially available powders can be added in liquids that often cause choking, to make them thicker and easier for the tongue to push to the back of the throat. Family

members can feel more comfortable around a person who has choking problems if they will learn the Heimlich maneuver, a method for expelling particles that are blocking the airway. Local hospitals, the Red Cross, or emergency service providers may sponsor training courses for this and other first aid procedures.

Tips to minimize choking

- Chin-tuck maneuver
- Alternate solids and liquids
- Use a straw or covered cup
- Minimize distractions

Feeding Tubes

Some children with HD develop such severe dysphagia that they are unable to maintain their weight or nutritional needs, or developed real-time pneumonia from food or saliva that end up in the lungs. When this happens, the family should consider the use of a feeding gastrostomy tube. Many people have very strong feelings about the use of a feeding tube. While some say it only prolongs the later stages of a degenerative disease and that they cannot imagine using such "artificial" means of providing food, others say that their child should be able to live comfortably without the discomfort or indignity of hunger and choking and that a simple, though artificial means, by providing nutrition is desirable.

The decision about a feeding tube should be a thoughtful and careful one. What works best for one person or family may not be the right decision for another person or family. The physician or dietitian can provide more information about the types of feeding tubes that can be used, how they are inserted, what the risks and potential complications are, and how nutritional supplements are given through such a tube. It is helpful to consider and discuss issues early, before crisis situation arises. Parents or other caregivers do not need to wait for the physician to bring up the topic at a clinical visits, but can include it on their list of questions or concerns to discuss at any time.

Nutrition

Most people who have HD experience significant weight loss as the disease progresses. Early attention to nutrition is an important part of the management all of HD, as it can help to slow down or minimize weight loss. Access to high quality foods with extra calories and protein and adequate supplies of calcium and vitamins are important.

For a child who has difficulty maintaining weight, supplementation with high calorie snacks is often appropriate. If prepared supplements are too expensive, it is recommended that affected individuals substitute cream for milk (for instance, on cereal), use proteins/calorie additives such as Carnation Instant Breakfast, use milkshakes or ice cream for snacks and eat high carbohydrate foods such as pastas. As chewing or swallowing difficulties lead to changes in food types or textures, consultation with a dietitian may be helpful to ensure that the diet still includes enough calories, protein, and vitamins.

Many adults with HD develop strong preferences, or even obsessions, for non-nutritious items such as soda, candy, coffee or cigarettes. The parents can try to control this problem in a child with HD before it begins by making only nutritious food available for snacks. Giving a preferred or special food to a child in later stages of HD may offer a real source of pleasure or social reward for a child who may no longer be able to express his desire or needs.

Oral Hygiene

Oral hygiene is particularly important for people who have HD. Eating and speaking are two of the few pleasures left when one loses the ability to walk, go to school or work, interact with peers. Loosing teeth or developing painful abscesses may make a person less able to speak and more likely to choke and may even lead to our refusal to eat. Early and aggressive attention to oral hygiene is necessary. Some caregivers have found that brushing with mouthwash is easier and better tolerated than using toothpaste; others have found that electric toothbrush, introduced early, is helpful. If the child cannot cooperate for routine dental visits, it is appropriate to seek out a dentist who specializes in or has been willing to treat children with disabilities.

Communication

Children who have difficulty swallowing properly are very likely to have speech impairments as well, as the same lip, tongue and throat muscles are involved in both processes. The speech pathologist should evaluate communication as well as swallowing, even if there are no apparent problems. Because dementia will impair the child's ability to learn how to use assistive devices, such as a computer or a communication board later in the disease, introducing devices or communication strategies, before they are absolutely necessary, is key to maintaining good communication for as long as possible. A variety of assistive devices and strategies are available. Each child must be assessed individually to find a device that works best for him, based on age, cognitive skills, motor skills, and the situation in which communication is needed. Because the disease is progressive, a device that works well for them years ago may not be as useful today. The family and the speech therapist must be willing to reevaluate the child needs annually.

General Medical Care

In the early stages of HD, a pediatrician or family physician should ensure that the child general health is not neglected because of the special care related to HD. Immunizations should be given on time and growth and maturation should be monitored as it would be for any other child. As the disease progresses, the general physician can also monitor for and treat any medical complications of the disease.

Alternative Therapies and Medications

Very little is known about the role of alternative therapies a HD, and even less is known about their use in juvenile HD. In recent years, more research funding has been directed towards scientific studies of alternate therapies than ever before. In the next two years, this will hopefully lead to a better understanding of the appropriate indications and doses of different treatments.

Physicians often feel uncomfortable discussing alternative therapies because of the lack of scientific studies/data and their own lack of knowledge about them. A parent should always mention to the physician any herbal preparations, vitamins, or other therapeutic treatments that a child is taking, and be prepared to describe why the preparation is being used and any apparent benefits or side effects. Bringing the bottle along to the physician's can help to avoid misunderstandings. Some physicians are more accepting of alternative treatments than others and there may be medical reasons to avoid some treatments. Alternative therapies might include vitamins, herbal preparations, homeopathic preparations, chiropractic manipulations, acupuncture, and magnet therapy.

"I went to an HD convention and talked to a man there who claims that his HD was arrested by drinking DM cough medicine three times a day. That's quite a story here with our family, how I fed her cough medicine three times a day for, I think, pretty near eight months. She sees me coming with the bottle and she would run. Of course, there didn't turn out to be any truth to it, but we were desperate to try anything."

Vitamins are required in small amounts for normal body functions. Except for the treatment of individuals who have measured deficiencies of the particular vitamin or vitamins, it has been difficult to prove that large doses of any vitamins delay or improve chronic neurodegenerative diseases. Vitamin C and vitamin E are sometimes promoted in particular literature because they have an antioxidant function, meaning that they protect damage cells against further damage or death. Neither vitamin has been shown to slow down or stop HD or other similar neuro-degenerative diseases, although another antioxidant (Coenzyme Q10) is currently being studied in the CURE-HD trial, scheduled by the [Huntington Study Group \[HSG\]](#) to be completed in 2001. Excess amounts of vitamin C can interfere with the progression of iron in the body, so it is important not to take excessive amounts of this vitamin. Similarly, mega-doses of vitamin A and some of the B vitamins can be harmful. However, if the daily diet is marginal or inadequate, these vitamins might be appropriate.

Other dietary supplements frequently asked about include creatine (a muscle protein) and various polysaccharide and glycoprotine compounds. Several clinical trials are underway to assess the safety, dose, and effects of creatine in HD, and by 2001 these studies should have provided additional information. Polysaccharides or glycoprotines are unlikely to change the underlying cause of nerve cell deterioration in the brain.

The affected person or family must carefully weigh the potential risks of each compound, their costs, and the reason for using them. Most physicians are reluctant to support the use of expensive nutraceuticals which have not been studied in any detail in either laboratory or clinical trials of neurodegenerative disorders.

Typical concerns that physicians have about herbal preparations include:

- Appropriate doses are unknown
- The risk and benefits have not been analyzed scientifically
- Her ballpark rations are not subject to the same standards of purity and accuracy as our FDA – approved or prescription medications
- Patient to spend large amounts of money on herbal preparations may be reluctant to use prescription medications of known benefit
- If patients believe that their doctor does not want them to use herbal preparations, they may be reluctant to notify the doctor of side effects or to mention the use of herbal compounds to Dr.

Many herbal preparations are available in the grocery store or drugstore, most with a long history of traditional use. None of these preparations have been shown to slow or stop HD. The decision to use a herbal preparation, like any other medical decision, must be made by the individual or the parent after considering the potential benefits, risk, and interactions with other medications or treatments, and the financial costs.

Homeopathic preparations almost by definition have not been subject to scientific study. Most physicians believe that the dilution process, used to prepare a homeopathic remedy, removes almost all the active ingredients from the preparation and they have difficulty explaining how such a compound could have a biological effects.

Chiropractic manipulations are generally directed at abnormalities of the bones, joints, and muscles and would not be expected to have a direct effect on HD itself. Patients with joint deformities or neck or back pain as a consequence of HD might respond to chiropractic manipulation, although studies documenting any benefits have not been reported. Anyone who chooses to see a chiropractor should assess the credentials and methods of a chiropractor, as well as the goals and planned duration of the treatment, just as he or she would work with a physician to understand a specific purpose of each prescribed drug.

The role of acupuncture and HD is also likely to be a minor one, but older children who have chronic pain or stiffness because of their disease could certainly try acupuncture if their therapies are ineffective. Magnet therapy has enjoyed a recent surge in popularity and is said to improve or cure a number of ailments. There is no rule of magnet therapy in HD.

Other treatments might include massage, Tai Chi, and other forms of exercise. Some benefits have been noted in the balance in elderly individuals with Parkinson's disease who have participated in Tai Chi, as well as improvements in pain and stiffness from massage therapy. Exercise in individuals of any age who have a movement disorder, and for children, is particularly important to maintain strength and flexibility, to aid in normal growth and to avoid the development of contractures

Chapter 4 - Daily Life

This chapter discusses how HD affects the deal a life of a child -- --school, home life, friends, and daily activities. At the end on the chapter, special emphasis is given to the importance of fun. This is perhaps *the most* important thing to remember in caring for children with HD, as all children (and maybe even some adults) love to have fun!

"Linda liked to have fun, and she still does. She still gets out and goes places and has quite a good sense of humor and a good disposition. So, like I said, even after all she's been through she still has the biggest smile around and one of the biggest hearts."

The Parent As Teacher

Many parents are frustrated or angry when they find out that "no one knows anything about juvenile HD." Parents must understand that juvenile HD is uncommon and they will eventually have more practical knowledge of the disease, because all of their experiences, than almost anyone they meet throughout their child's life. However, many of the daily concerns that parents and children with juvenile HD face are similar to the problems that professionals in the school or clinic face with children who have other disorders or disabilities. Health professionals can draw on their experience with other similar chronic diseases to help a family with juvenile HD.

Rather than being angry or turning away in frustration, parents should learn to work with the physicians, therapists, social workers, and teachers assigned to help their child so that information is passed in both directions. Even for professionals who are familiar with juvenile HD, each child presents unique concerns and problems -- -- there's no "formula" or "solution" for taking care of a child with HD. For those who care for someone with juvenile HD, their main goals are to make the individual's life, however long or short it may be, easier, happier, and more filling.

When a parent or family member gets balled down with paperwork or meets a professional who seems to have forgotten these goals, it maybe helpful to pause and review the overall goals as well as the smaller goals for the particular intervention or treatment. Talking to others who have had a child with juvenile HD may help a parent or caregiver to become more familiar with the medical, educational, and legal terms that go along with the disease, which will make it easier to speak with the various professionals about the child's needs.

Some parents come to enjoy the chance to teach other people what they have learned about juvenile HD, and realize that through teaching others they can make life a little more better for the next child that the develops the disease. Most parents who have taken care of someone with juvenile it's deep are happy to talk to parents were new to disease and struggling with their own new roles as teacher, caregiver, and advocate.

School

As soon as the diagnosis of juvenile HD is made, it is important for the family to begin what will be an ongoing relationship with the school about how to meet the special needs of the child. All the schools in the United States are required to provide for the education of all children through the high school level.

As the child's disease progresses, the school may need to make adjustments for, or allow for, variations in 1) academic classes, 2) physical education, 3) meals, 4) seating and transportation, 5) safety and hygiene, and 6) behavioral control. An Individual Education Plan (IEP) should be written each year for the child and periodic assessments of the goals should be made during the school year as needed. Many HD families have found it helpful to visit their HD physician at the end of the summer so that any "doctor's orders" can be written, new medication started, or forms completed, just before the start of the school year.

"Had I known about Huntington's disease, like I know now, we would never have allowed her to go through so much with the Ritalin and trying to learn so much she just couldn't learn. It probably started altogether when she was in the third or fourth grade, maybe even younger."

Academic Classes

As HD progresses, children become less able to learn new material due to disturbances in memory, concentration, and loss of ability to initiate and continue complex or even simple actions. Increases in behavioral disturbances may occur when a child is frustrated by material that is too difficult or presented too quickly. Often the child is unable to describe or understand why he's angry or frustrated.

Remaining in a class with familiar classmates may be important to some children; others may prefer or need individual attention available in special education programs. For example, a patient was given time each day in the high school office, where she helped to Xerox and staple, while other students pursued academic class work. Another child is placed in different school because his behavior problems were too frequent, severe, and disruptive to allow him to remain in his old school. Another child remain with his class until his last days even though he was unable to speak for several years before his death; his classmates helped him in different classes, included him in activities, and in general found his presence to be a valuable part of their school experience.

Physical Education

Juvenile HD produces noticeable motor disturbances vary early in the disease. Because physical skills are such an important part of self-esteem and social experience in school, it's maybe best to remove the child from the embarrassment being the clumsiest kid in the gym class. On the other hand, if a particular group or class is excepting of a child's physical disabilities, including the child in regular gym class, in whatever capacity he can participate, may be very important in helping him to maintain self-esteem and social interactions. Safety must be considered as well.

Children who fall frequently, or have the potential to fall, should avoid activities that present a high risk of injury. For children with rigidity or spasticity, exercises that include stretching and relaxation are particularly beneficial.

Meals

Meals become difficult as HD progresses. Any child whose speech is indistinct is likely to have a degree of swallowing difficulty of well. Because nutrition is so important, and the risk of choking is real, it is probably best to have a child with significant dysphagia eat in a quiet and closely supervised setting. It may also be more comfortable for the child to eat apart from his or her peers and receive help with the food tray.

The risk of choking can be decreased by using a few simple maneuvers, such as chin-tuck maneuver (see Choking in Chapter 3). Some children with advanced HD need special diets that avoid certain types of

foods or texture. The speech pathologist and dietitian can work together to find a diet that is safe and provides adequate nutrition but is also enjoyable for the child to eat.

Seating and Transportation

The main concerns of seating and transportation relate to positioning and safety. Children with handicaps of any kind are vulnerable to the comments or taunts of other children on the school bus. Children with HD often have impulsive, rapid, or aggressive behavior which may be dangerous to other children or to the safety of the whole bus.

Until HD is fairly advanced, children should be able to use regular chairs in school, but as rigidity and spasticity worsen, or if chorea develops, a wheelchair may be needed for transportation and special recliner chairs with appropriate padding may be appropriate for prolonged sitting. A pommel or other adaptations may be needed to keep the individual from sliding out of the chair trunk control is poor. A physical or occupational therapist can make specific recommendations about seating and other devices that may make their school life easier.

Safety and Hygiene

The older a child is, and the further along he or she is in the course of HD, the greater the concerns about safety and hygiene. For adolescent girls, managing menstrual hygiene may be difficult and require assistance. And, for any child with physical, mental, or behavioral handicaps (and children with HD are likely to have all three), vulnerability to physical or psychological abuse by other children or adults is a concern.

Some school districts, with a physician's order, are able to provide a personal care attendant to accompany the child throughout the school day. Educating, and then reminding, the child about what to do in an emergency or uncomfortable situation is important.

Brothers and sisters can use the opportunity to teach other students about HD, turning the jokes and jibes, which often are initiated because of the misunderstandings or fear, into encouragement and offers to help. Children can take a leading role in educating their classmates (or the whole school) about HD and by organizing their own "Hoop-A-Thon," an HDSA program that has raised thousands of dollars to support HD care and research. Children need to be reassured that HD is not contagious and that it progresses slowly over a number of years. By understanding this, they may better be able to regard the affected individual as a member of the group.

Residential Schools and Tutoring Options

Occasionally, an aggressive or depressed child may pose a threat to himself or others. In addition, some school districts may be unable to meet the needs of a child with HD. In either case, an alternative to public-school may be appropriate. There are residential schools for optically handicapped children that may be better able to meet the child's educational, social, and physical needs than the local school.

The American with Disabilities Act (ADA) requires that public schools meet the needs of all individuals. If a local school is unable to meet our child's needs, it must provide an alternative for the family. One alternative that the school will frequently suggest is home-based tutoring. Although this may be an acceptable alternative for some, it creates even greater problems for others in meeting the child's social needs and the rest of the family's needs for respite from care duties.

Residential schools represent another option. Local schools fund residential placement when it is deemed "educationally necessary." It is helpful if the parents have developed a working relationship with the school before such a step is taken. Then the school and parents can work together in a positive way to

identify the best alternatives for the child, rather than having the appearance that the parent is taking the child away because the school is “bad” or deficient in some way. It is important that the family have advocates to support the child’s residential placement, such as a psychologist, physicians, or case managers. For some families who struggled with other options, residential placement has greatly enhanced the child’s educational and social experience. Residential placement should be considered as a good option rather than a “last resort.”

Daily Routines

For children, HD may strike at a time when certain skills, such as dressing, tying shoes, cutting meat, combing hair, and shaving are being learned. The onset of HD does not mean, however, that a child should never be asked to do more than he or she is currently doing. With support and repeated instruction, children can learn new skills, particularly if they become part of a comfortable group routine or if others around them are doing the same activities.

When there is a problem, parents of children with HD often asked whether their child really can’t do a particular task or whether this refusal or apparent inability represents an “HD behavior.” It is often difficult for the physician or psychologist to answer this question, and the solution to the problem, no matter its cause, is likely to be behavioral (changing the situation or environment) rather than medical.

"He could feed himself then, that it was a major project. He became severely emaciated (110 pounds on a 6 foot frame). At times, he was restrained into padded bed."

When confronted with and new apparent inability to perform some tasks, such as eating or getting dressed, the parents should:

- 1) **examine the situation carefully.** Is the job not given enough time to complete the task? Is he embarrassed by choking/drooling/slowness/motor impairment/the choice of clothing? Is there a specific person or situation associated with this problem? Some individuals are so obsessive about their schedule; does a slight change in schedule on the weekend precipitate a crisis?
- 2) **consider the management options available**, which often include changing the environment or schedule to adapt to the individual with HD, or changing the goals.
- 3) **set simple goal**, discuss them with the individual, and allow only choices that are acceptable. Using open-ended questions such as, “*How do you think we should schedule meals so that you will you to better?*” are likely to lead to no answer or any answer that the caregiver doesn’t want to hear (such as, “*Give me potato chips at midnight when I am hungry!*”). Instead, the caregiver should provide acceptable choices such as “*we are having your favorite ice cream after you eat your vegetables. Do you want peas or beans before your ice cream today?*”

Managing behavior becomes increasingly difficult as the child’s ability to communicate declines. Increases in irritability, aggressive and combative behavior are often seen as speech becomes more difficult and less intelligible. Once again, the parent is more likely to know a particular child’s comforts, special treats, and usual needs and desires better than anyone else. If the child is living or staying elsewhere, it is important for the parent to help other caregivers by noting some of the special things about the child. Working with a speech pathologists to devise a simple communication device or schemes may postpone this time and reduce everyone’s frustration.

When things get frustrating, parents and other caregivers should remember that it is rarely the child’s desire to behavior irritable or aggressively. It is HD that causes the problems. For example, one family

crocheted the words "Huntington's Disease" onto a pillow and whenever frustration levels were high (for the child or the parents), they were allowed to hit, throw, or kick the pillow. This approach both acknowledged that it was HD that was causing the frustration and provided a safe way to relieve the feelings!

Activities

In the middle stages of HD, children with HD become less able to handle complicated schedules filled with multiple activities. Apparent behavior problems may be the result of a tired or confused child who needs a less complicated régime. On the other hand, there may be certain familiar activities that are particularly enjoyable. Sunday morning church, Friday evenings at the pizza place, or swimming at the YMCA may all fit into this category.

As HD progresses, the emphasis should be on familiarity, comfort, in an enjoyment rather than working too hard to finish all the tasks on a busy family schedule. A respite caregiver may also be a great help, taking the child to activities while the family has a much-needed break.

"Her church was supportive and included her in all the activities from Bible studies to bus washing to eating to camping in the Boundary Waters. She finished high school in regular classes, staying to the last of the all-night parties after marching proudly around the football field on graduation night."

Driving

Getting a driver's licenses is a major milestone for adolescents and represents a big step towards independence. For children who developed HD symptoms before the age of 15, learning to drive is not a reasonable goal. The combination of cognitive, behavioral, and motor impairments prevent the adolescent with established HD from learning to use a car properly and safely. This is a very important issue, as it can impact the safety of other family members and strangers, as well as that of the affected child. Families can help themselves by preparing for the conflict about driving, years before it comes, by gently but firmly reminding the affected child that he should not expect to drive.

For the child whose HD symptoms develop later in adolescence, driving privileges may already have been established and impairment in judgment or denial of his illness may lead the child to drive very erratically or unsafely without recognizing that he is doing so. Many larger communities have driver education programs for individuals with handicaps: written and behind the wheel exams can be administered through these programs. If the child or young adult cannot pass the examination, then he cannot get a license and should not drive.

The family and the physician should work together to help the child to understand, in advance, that the ability to drive will be lost at some point in the disease and that yearly assessments may be appropriate. To avoid injury to the affected person or others, the family must be firm with a young adult who does not have a license or whose license has been revoked, even to the point of keeping car keys locked up and notifying authorities that the person is driving without a license.

Friends

Children with HD ultimately lose the ability to maintain friendships. There are number of factors that contribute to this loss. The movement disorder is likely to limit the child's ability to keep up physically with his classmates. As HD progresses the cognitive differences between the affected person and his peers become more and more striking; the friends progress, follow new trends and learn new things while the affected individual's ability to process new information plateaus and then begins to decline. Finally, severe behavior problems may bring the child into more contact with other children who have

behavioral problems, as well as cutting off friendships with those who find the abnormal behavior frightening or strange.

Maintaining social interactions is as important to the child who has HD as it is to the child who does not. Depending on the child's personality and the behavioral patterns, it may be easier for him to succeed in small groups, surrounded by familiar and friendly faces, rather than a large group. An occasional child, however, may thrive in a large group if it is a friendly one and is supervised appropriately.

Dating may be difficult for child with HD, as he requires learning a new set of behaviors as well as responses to the behaviors of others. Many children with HD become lonely in their teenage years as HD robs them of their ability to initiate or maintain social relationships. Children should be encouraged to maintain social contacts for school, churches, or other resources. Activities involving small groups are often the most successful.

Any child in their teenage years who does have a close relationship should be encouraged to talk about the relationship, with special attention given to indications of sexual behavior. Children with HD are unlikely to understand fully the implications of sexually transmitted diseases or pregnancy, so they should be considered vulnerable in this regard. An occasional child with HD is sexually aggressive. Parents and other caregivers should recognize this as a behavioral health problem and seek treatment from a psychologist or psychiatrist skilled in sexual disorders.

Pets

As the ability to make lasting friendship with peers diminishes, some families have found a remarkable new outlet for their children in the form of non-human friends. A pet can be a tireless companion, a housekeeping and personal hygiene assistant, a friend in times of sadness or anger, and emergency alarm, and a physical therapy helper, among many other roles.

The type of pet depends on the child's specific needs, who and how much time is available to care for the pet, how large the house is, and other issues. Some animals are trained specifically to assist individuals with disabilities. Families can contact a local veterinarian, the ASPCA or a service such as Canine Companions for more information.

Fun

In the midst of all the sorrow and losses that accompany the diagnosis of juvenile HD, it is easy to overlook one of the main things that life is about, which is having fun. The child with HD has a limited life-span. It is important to think of ways to have fun this year, since next year may bring a reduced capacity to participate in or enjoy activities that require cognitive or motor function.

Fun means different things to different people. For some, it is swinging on a tire swing, for others a trip with the family to the zoo, the beach, or at favorite relative's house. The parent can keep a scrapbook of fun activities, partly to remind himself or herself when activities were fun, but also to remind a child in later years about these enjoyable memories. For some children, spiritual benefits and activities provide remarkable comfort and reassurance even when daily life becomes difficult or lonely.

In the United States, the Make-A-Wish Foundation and the Starlight Foundation both provide funding for children with serious or life-threatening conditions to make a special trip with their families, to participate in a special event or make a purchase. HD qualifies as such an illness and many of the children have enjoyed trips to Disneyland or other places with Foundation support. Summer camps, Special Olympics and weekend retreats for HD affected individuals are examples of pleasurable activities.

Chapter Five - The late stages

The Stages of HD

Because there is no treatment that stops or reverses HD, the symptoms will always progress over a period of years. Just as each child is different, each child's HD will be a little different too. For some families, aggressive or impulsive behavior provides the greatest challenge. For others, behavioral problems are mild and working with school or local resources, to find appropriate activities or, as a child becomes a young adult, an appropriate place to live, maybe the biggest obstacle.

Physicians often use functional skills to classify or group patients, expecting that certain medical or social issues are likely to occur at certain stages of the disease. A 13– point functional scale, devised a number of years ago by Dr.'s Ira Shoulson and Stanley Fahn, is commonly used to describe function in adults with HD. The scale measures a person's ability to work, manage money, do household chores, perform self-care activities, and live independently. Affected individuals are grouped into five stages by this scale according to the number of points scored, with Stage 1 representing the earliest stages (11 – 13 points) and Stages 4 (1-2 points) and 5 (0 points) representing the late stages.

As useful as this article is in assessing individuals with adult-onset HD, it is difficult to apply to children. Provided here is suggested way to modify the Shoulson Scale to apply to children. Although this still has not been "validated" (proving to be scientifically or statistically accurate), it may be clinically useful in judging how a child's HD is progressing.

| A fundamental scale for assessing Juvenile-Onset HD | The stages of HD is determined by adding the point, as shown: |
|--|--|
| A. School attendance 3 - attend school, with those special assistance needed 2 – attend school, some regular classes, some special or modified classes 1 – attend school, few or no regular classes 0 – unable to attend school or work program | 11-13 points Stage 1 7-10 points Stage 2 3-6 points Stage 3 1-2 points Stage 4 0 points Stage 5 |

| | |
|--|--|
| <p>B. Academic/developmental performance 3- reading/ writing/ math skills appropriate to age 2 -violence decrease in academic performance but still able to take a test or to write 1 -unable to write legibly but able to communicate orally</p> <p>C. Chores 2- able to assist an age -appropriate manner with household chores 1 – vocationally assists with chores 0 – unable to participate in household chores</p> | <p>D. Activities for daily living 3 -performs self – care unit at age-appropriate manner 2 -require some assistance for bathing, dressing, grooming, or feeding 1 -assist others who bathed, dressed, or feed him/her 0-unable to assist in self-cares</p> <p>E. Lives 2 – at home with only family assistants 1 – at home/group home/foster care with assistance from non-family members 0 – living in a long – term care facility</p> |
|--|--|

- According to this scale, a child who is attending regular classes, but showing a slight decline in academic performance or ability to help with chores would be classified as having Stage 1 HD.
- A child requiring some help at school and home, but was still attending some classes and performing appropriate self-care would be classified as being in Stage 2.
- Stage 3 is transitional stage-entering that stage, children would typically still be attending school, but performing little if any academic work. In this stage, children need increasing assistance with self-care, and extra help may gradually be needed in the home.
- Often children in Stages 4 and 5 have reached young adult and are placed outside the home where they can obtain advanced nursing care.

Neither the Shoulson Scale nor the scale proposed here account well for severe behavioral or psychiatric problems. Thus, patients with early HD who have very severe behavioral problems may score more poorly than their mental or motor skills would suggest that they should, and apparent improvement in disease stage could be seen if severe behavioral problems respond well to treatment. However, despite its limitations, it is believed that this skill can be applied to children with HD, to help families understand where the affected person is in the course of their disease, and to help guide treatment plans in the school and at home.

Medical Problems In The Late Stages

Stiffness of the limbs may become very severe in some children. Scoliosis (curvature of the spine) and limb contractures can occur because of changes in muscle tone and strength. Sometimes early referral to a physical therapist can help the family or other caregivers to learn exercises to stretch tight muscles and to maintain the range of joint motion through active and passive movements of the limbs. Scoliosis due to HD is likely to occur or worsen despite bracing or surgical procedures because, regardless of treatment, underlying disease will continue to progress.

In the late stages of HD, children are bed bound and prone to malnutrition, urinary tract infections, pneumonia, and skin sores. With excellent nursing care, some of these complications can be prevented and children can live in the late stages for number of years.

Although it is difficult to talk or even think about the progression of symptoms or the late stages of any disease, it is important to do so. By having a realistic idea of the challenges ahead, parents or other caregivers can ask questions, make plans, clear up any misunderstandings, and avoid surprises or crisis situations.

As difficult as it is to discuss some of these issues, it is better to have discussed them and have some idea or solutions than to have a crisis arise and never have considered the options. An older family member, a chaplain or minister, a counselor, a social worker, or physician or nurse can help parents or caregivers to consider and discuss these issues.

In the late stages of HD, the affected individual is not usually able to communicate his own desires or choices regarding his medical care. It is best for the family to discuss any limitations in care they would like in the end stages (issues commonly discussed include feeding, hydration (fluids), the use of antibiotics, hospitalization and resuscitation). The affected person's wishes can be written down in the form of "Advanced Directives." It is wise for another family member to serve as legal guardian, so that he or she is permitted to make medical care decisions on behalf of the affected person.

Professional Help In The Home

Many different kinds of help can be provided within the child's home, either by professionally -trained care providers or by family or volunteers. The need of each fairly depends on specific problems the child is having, what the home environment is like, and the skill of available family members or friends.

The solutions to problems also depend on the family's creativity, financial resources, and the community in which they live. Some families, with numerous relatives or a very active church or school group, can find volunteers to assist with the affected child so that professional assistance is never necessary.

Other parents simply desire help with household chores such as cleaning and cooking, so that they have more time to spend with their HD affected child or the other children. Still other parents find that there are "custodial" tasks such as bathing, dressing, and feeding that are impossible to manage because of the child's size or behavior, or because of their own emotional, health, or work issues (or the need of other family members).

Professional home health aides or certified nursing assistants can provide assistance with some of these tasks. A visiting nurse can help to set up medications and supervise the activities of the aides or nursing assistants. For those in the terminal stages of HD, in-home hospice care can be invaluable in helping family members to prepare themselves physically and emotionally for the child's death, and to maintain dignity and privacy in the process. The physician can order a visiting nurse assessment. The visiting nurse and then work with the family to determine what specific needs they have in which services are appropriate.

A social worker will be able to discuss what programs are available in a particular county or community. Networking with other families with disabled children can be very valuable as well.

Placement Outside The Home

Individuals with juvenile HD require 24-hour nursing care by the time they are in the late stages of the disease. Some families are able to accomplish this at home, either because family members who were trained in nursing are able to help, or because they have been able to access in-home services, or because they are able to qualify for or afford school or community-based day programs that provide all the care that a child with HD needs as well as also providing for the needs of the rest of the family, particularly if the child's behavior problems are severe.

When living at home is no longer appropriate for the child or family, placement at a long term care facility becomes necessary. There are no facilities in the United States that specialize in the care of children with juvenile HD. However there are several facilities that have specialized units for adult or young adults with HD, so that placement of the young adult with the onset HD in a specialized care setting may be possible.

For those who are not in the late stages of the disease, but who, as young adults, need or desire placement outside the home, a number of different living situations, with varying amount of care or supervision, are possible. These include fully independent living without assistance; subsidized but independent housing; adult foster homes; group homes; board and care facilities; assisted-living facilities; and nursing homes.

The financial and care aspects of these different parts of living situations vary greatly, and the parent is encouraged to speak at length with the social worker or case manager, and to visit each facility, before making any decisions.

This selection of long-term care facilities is an individual and difficult one. The choice may be determined on the basis of the programs available at the facility, geographic location, financial considerations, religious affiliations, or often - like choosing a new home - by how it “feels” or “matches” a particular person. For a young person with HD, the local nursing homes have never had a resident under the age of 50, so this may seem like a very poor match. However, there have been circumstances where young men with HD flourish in a facility filled with grandmotherly women!

If behavior is a major problem, a facility with a special “behavioral unit” or a “head-injury unit” may be a good choice. Over time, as the motor and cognitive symptoms progress and outweigh the behavioral symptoms, the specialized services of the behavioral unit may no longer be necessary and a nearby community care facility may be more convenient or appropriate.

The family should always tour a long-term care facility before making a selection. Once the affected individual is living in a facility, there are ongoing opportunities (in fact, requirements) for the facility and the family to discuss and modify the Care Plan. The Care Plan includes not only medications, but also any treatments, restrictions (for instance, rationing of cigarettes or other behavior modification programs), dietary changes, and restraints that are used in the daily management. Families should maintain an open discussion with care facilities, and should not be afraid to ask questions, offer recommendations, and to work with facilities to provide the best quality of life possible for the child or young adult with HD.

Hospice Care

Hospice care is a special kind of nursing care which focuses on using the transition between life and death for both the ill individual and family. As medicine and nursing become more and more technological and less personal, many families have turned to Hospice care in the last few days or weeks, to find personal touches and support that seem more useful and important at the time.

Hospice can be provided in the hospital, in the nursing home, or at home, by nurses who have special training or experience in the kinds of problems and concerns that occur in the last few days of life.

Families often have questions or worries about medical issues such as pain, nutrition, infections, and whom to call when there is a change in mental state or a fever or difficulty breathing. Hospice nurses can make sure that these questions are answered and support the family in whatever care decisions they make.

The hospice team can also gently help the family prepare for the tasks that must be done at time of death, such as planning a funeral or memorial service, contacting relatives, employers, and insurers and finding resources to help cope with grief. Families of children with juvenile HD, who have made use of hospice services, have found them to be very helpful.

Chapter 6 - Financial, Legal and Social Service Issues

Medical Consent

Every state has a Medical Consent law that allows certain people to consent to medical treatment for another person. For example, a parent may consent for his or her minor child, one spouse may consent for another, and a grandparent may consent for his or her minor grandchildren if the parent is absent.

The Age of Majority is a term used by lawyers to describe the time in life after which a person is legally no longer considered a child; it is an arbitrary time when a child becomes an adult in the eyes of the law. Historically, the age of majority was 21 in most states. However, after the Twenty-sixth Amendment to the United States Constitution was ratified, giving 18-year-olds the right to vote in federal elections, most states lowered their age of majority to 18. At the age of majority individuals acquire the right to consent to medical treatment on their own behalf.

When a child with HD reaches the age of 18, he is considered an adult in most states. Since the young adult who has had HD for a period of years is usually fairly advanced in the illness, an adult is needed to assume guardianship. This is a legal procedure handled within the probate court of most counties. The caregiver must file a petition, then an independent evaluator or an attorney for the proposed ward is assigned, and a hearing is held. If guardianship is granted, the caregiver will have the legal authority to make personal and financial decisions for the person with HD.

Financial options

Supplemental Security Income

Supplemental Security Income (SSI) is a federal program for cash assistance for aged, blind and disabled individuals who have little income and few assets. Disability benefits to children are made under SSI (rather than SSDI). An important component of this program is the fact that, after an individual is determined eligible for SSI benefits, he is automatically qualified to receive Medicaid benefits as well. The program provides monthly checks from the federal government of up to \$530 (in 2001) for an individual. Some states provide an additional cash supplement. The exact amount of SSI for which the claimant is entitled depends on what the individual owns and how much income he or she has.

The basic purpose of the SSI program is to assure a minimum level of income to individuals who are disabled and who do not have significant income and resources to maintain a standard of living at the established federal minimum income level.

SSI pays benefits to disabled needy individuals of any age, including children. Under SSI children are considered disabled if they have a physical or mental condition which is so severe that it results in marked and severe functional limitations. The individual's condition must last, or be expected to last at least 12 months, or be expected to result in the child's death.

When the applicant is an individual under 18 years of age the resources of the parents are deemed to the applicant, under a formula that takes into account the number of individuals living in the household. However, resources of the parent are no longer counted the month after an individual turns 18 years of age, even if he still lives with his parents.

“Countable” resources include cash, stock, bonds, mutual funds, promissory notes, mortgages, life insurance policies, and others similar property that can be easily converted to cash.

Special rules apply for children with parents currently drawing Social Security. Upon diagnosis of juvenile HD, it is important to obtain pamphlets and information from your local Social Security office. You may check the web site for Social Security at <http://www.ssa.gov> or call (800) 772 -1213.

The disability Law Attorney can also provided state—specific information and guidance in planning for eligibility for SSI benefits.

| Assets that are not counted when determining financial eligibility for SSI benefits include: | |
|---|--|
| <ul style="list-style-type: none"> • The value of the family home • Household goods having a total value of no more than \$2,000 • One automobile if it is used for transportation to implement or to obtain • medical services • Life-insurance policies having no cash value | <ul style="list-style-type: none"> • Life-insurance policies with cash value having a face value of no more than \$1,500 • Burial services for the individual and his/her immediate family |

Medicaid

Medicaid is a jointly funded and administered state and federal welfare program that pays the qualifying medical expenses for those individuals whose financial resources fall below the programs established minimums.

Medicaid may pay for such services as hospital and doctor bills not covered by insurance, home health care services, medical transportation, and nursing home care. Many states also have waiver programs under which an individual who is disabled, but does not meet the financial criteria, may still become eligible for benefits.

Because the eligibility criteria and available benefits very greatly by state, one should contact the state or county department that administers the Medicaid program in his/her state (called Medi-cal in California) or a disability Law Attorney for information specific to the state.

Children Special Health Care Needs

The Children Special Health Care Needs arose from title V. of the Social Security Act. This program exists in every state and territory in the USA, but it may be called Crippled Children’s Services or Children’s Medical Services or Handicapped Children’s Program. Most pediatric health providers and medical social workers are familiar with the access point in each county and state. The program starts with medical eligibility and then considers income eligibility.

Do not assume you’re not eligible as “cut-off” points are different then in programs for adults and may also vary from state to state. Different states request “waivers” from the Federal government regarding specific eligibility rules from the core program of Medicaid. Many states add their own money to special programs to supplement the federal money for services.

Other Financial and Placement Considerations

Many states do not have nursing home beds for people under 16 years of age, with some very specific exceptions for short-term care. People entering nursing homes are subject to an OBRA screening which

refers to the Omnibus Budget Reconciliation Act. The Act impacts all nursing homes excepting Medicaid, and overall has been viewed as standardizing and improving long-term care.

One outcome has been the increase in home-based services for people of all ages and medical conditions of a severe and chronic nature. Because more children are cared for at home through the entire course of a progressive, severe, and ultimately life-threatening disease, efforts have been made for respite care services to be developed. Respite care offer short-term care for the medically involved person in order to give a break to the regular caregivers, frequently the parents. Throughout the country there are wide variances and availability and sophistication of respite care programs.

There are special programs for a child without family health insurance, or for a child who has “topped out” the lifetime limits of the family insurance. The Tax Equity Fiscal Reform Act (TEFRA) is a waiver to help in such a situation. It will help cover necessary treatment and support, often in a home setting.

In another direction families must consider the services available through the public education programs of their particular state. Specialized services are often coordinated by the intermediate school district servicing several local school districts. Under the Individuals with Disabilities Education Act (IDEA), children with severe disabilities must be accommodated with services such as transportation, variable school hours [as needed], and personal assistance to participate. Accessibility provisions through the American With Disabilities Act (ADA) apply to children. An Individual Education Plan [IEP] committee must be held to develop a learning plan for each child in special education.

Services available to private schools and to charter schools may vary from jurisdiction to jurisdiction. States may add services to federally mandated programs, but they cannot detract.

Community mental-health programs, which in many states cover virtually every corner of the state, are frequently involved with children deemed to be “developmentally disabled”. These programs typically administer aspects of the (OBRA) program described earlier.

Experienced professionals working with people with complex health problems, and with several programs within the State and Federal network services, often have to search for entry points for people and often have to follow up with phone calls and letters to clarify eligibility and to gather information. It is recommended that families embarking on obtaining services try to become politically astute, to be assertive and to be collaborative with professionals working in the system. The rules can be complicated, but most professionals want to help a family with a disabled child obtain the appropriate benefits. Expect to make numerous phone calls, to keep logs of calls, correspondence, and visits. Expect to be a bit discouraged by the necessary paperwork.

The HDSA Chapter social workers and the Center of Excellence social workers are very knowledgeable about the resources in their state and region. Social workers will help you get started with services and help with advocacy as needed. Please refer to the appendix to find a local HDSA Chapter or Center of Excellence.

“I have just found in the last few years, after beating myself up with guilt, I did the best I could do. I loved him.”

Chapter Seven - Caring for the caregivers

Parents

The parents of a child with HD bear the brunt of years of care, adapting to the child’s increasing disability, always with limited resources, helped by people who are almost always unfamiliar with the disease, and usually without any thanks from the child. Not uncommonly, the families headed by a

single, divorced, or widowed woman who is working, caring for an ill child, and raising other children, often with the fear that they too could develop HD.

With all of this going on, it's of little wonder that the parents of children with HD record high levels of stress, anxiety, depression, anger, guilt and frustration. People who marry into an HD family and are not told about the disease may direct their anger towards the spouse and his/her family and estrange themselves from the family. Some parents, unable to manage an affected spouse and an infected child at the same time, may divorce or place the spouse outside of the home so that they can focus their attention on the child or children. Others, fearful that they will harm the affected child or be unable to provide adequately for him, arrange for foster placement. A few lucky families have the support of the aunts, grandparents, or friends and are able to rely on this extended family for day-to-day assistance as well as emotional support.

Most parents, however, struggle with their emotions without feeling that they have a moment to spare to sort through them. Caring for someone with HD without caring for oneself can only lead to trouble. A tighter, angry, or depressed caregiver cannot provide loving care, which is what children with HD need the most! In the end, it may be less expensive for a parent to spend time and money on counseling for him or herself, or respite care for the child, then to fight with an affected teenager until his aggression or confrontational behavior leads to a behavioral crisis, hospitalization, or incarceration.

Professional counseling can be provided by a member of the clergy, a psychologist or psychiatrist, a marriage counselor, or the family doctor or nurse. Emotional support can be provided by other family members and by friends - remember, friends and family will never know how bad things or how much help is needed unless someone tells them! The Huntington's Disease Society of America provides literature, support groups, newsletters, an annual convention, and a web site (www.hdsa.org), all of which are there to help families to connect with other families and to find accurate and up-to-date information. Any parent who is confronting juvenile HD should be reminded that your best resource is others in the same situation. Although others may live far away, the telephone and Internet can easily link people who are miles or even countries apart.

The other children

In addition to addressing the needs of a child with HD, the parent is faced with providing as normal a life as possible for the other children in the family. Although the details of the disease are slightly different, the overall picture is not much different from that a family struggling with any handicapped or chronically ill child.

Families adapt in many ways to HD. In some families, the other children seem to understand, love, and support their brothers or sisters who have HD, and the affected person remains part of a cohesive family throughout this course. In other families, the rage or impulse of behavior of the affected person seems to be directed at the siblings, even to the extent of physical or sexually abusive behavior. In still others, the family becomes fractionate, because of HD in one of the parents, because of the strain of caring for affected people in two generations at the same time, or because the energy required to maintain the affected individual leaves insignificant time or energy for the other children.

A family in which there is a child with juvenile HD is a family under stress. Everyone in the family should consider seeking professional counseling any time things begin to feel overwhelming; if performance (at work, home, or school, sleep, or mood begin to suffer, if they noticed an increase in "escaping" behaviors such as the driving while intoxicated or physical or verbal aggression towards other begins to emerge. School counselors should be aware that children in HD families, whether they are affected or not, are at-risk for depression or stress-related problems, should make a special effort to provide a safe haven where the child can retreat and talk. In situations where the school counselor, a Minister or clergymen, or a special "big brother" or relative are not enough, there are psychologists,

family Counselors, or social workers which can be consulted. A parent who is engaging in harmful or descriptive behaviors is unlikely to be able to redirect his or her child when the child acts out. In this situation, the parent must first regain control of his own mood or behavior, and then work to improve the child's situation. Many find it helpful if the entire family has regular visits with the family counselor.

In general, open communication about HD is important. That does not mean that all children in the family need to be told everything in detail about the late stages of the disease on the day that the diagnosis is made in his sibling. Parents should do their best to answer questions honestly when they come up. The sick misunderstandings that young children may have about HD should be put to rest right away. HD is not contagious and having HD doesn't mean that the affected child will have to be in a hospital, have surgery, feel sick, or die immediately.

"I have had so many years to prepare myself for the day of finding out for sure that Linda had the disease, so I know what helped me handle things better, emotionally. It was much better than having it all dumped on your lap at once. The rest of my family sympathizes very much and there are a few members who still visit her and come to see her on a regular basis, but the rest just kind of ask about her from time to time and that's about it."

Since the advent of gene testing for HD, many junior high and high school's discuss HD in health, biology, genetics, religion, and ethnic classes. Thus, many children would hear about HD at school even if it is not discussed at home. Children also have ways, particularly now in the computer era, of getting answers to their questions on their own. Rather than discouraging this activity, parents should encourage their children to access reputable web sites, such as the HDSA web site at www.hdsa.org, so that they get accurate and up-to-date information. An occasional physician may be willing to help a parent tell his teenage children "the facts" about HD and to answer questions that the parent is unable to answer. Writing a term paper for school may be a very useful way for an at-risk child to learn more about HD and to express some of his or her feelings and experiences with the disease.

Siblings of a child with HD have a growing awareness of their own risk of developing HD as they grow up. Although understanding how HD is passed on genetically is important, and that understanding includes knowing that individuals repeatedly emphasize an at-risk child's status unless the child asks, or if the child is sexually active and at-risk for passing on the gene. Once an at-risk child has become an adult and is able to make his own medical decisions, he or she can consider undergoing a gene test to determine whether the HD gene is present or not. Chapter 1 discussed why physicians do not ordinarily agree to test asymptomatic at-risk children.

Other Caregivers

Late in the course of HD, care is typically spread among a number of providers. Depending on the particular child, these caregivers may become fatigued or despairing. Nurses or aides that find themselves irritable, verbally or physically abusive to a child or young adult with HD, for whom they are caring, should ask to be removed from the case, at least temporarily. Ongoing education of caregivers is important as well. There are many caregivers who seem not to understand that HD is a progressive disease and appear surprised when the expected evolution of the symptoms occur. Just as a parent cannot provide good care when he or she is exhausted, angry, or seriously depressed, and uninterested or unhappy professional caregiver may be harmful to the affected individual.

Sometimes two individuals simply do not get along - this can happen with physician-patient and physician-parent relationships as well as caregiver-child pairs. Both parties should be candid about the problems, and if the problems can not be worked out, then other provisions for care should be arranged. All caregivers - parents, nurses, therapists, and doctors - should remind themselves that their role is to relieve the suffering and protect the dignity of a young person with at disabling and destructive disease.

If, for any reason, they cannot at least try to achieve this goal, then at the child is better off with a different caregiver.

Chapter 8 - Hope for the future

"I never give up hope that she will be 'healed' or that they will find a cure through research."

Now, more than ever before, there is reason to hope for improved treatments for HD. Although clinical research in HD began with the identification of the disease over 100 years ago, the discovery of the HD gene in 1993 has resulted in an explosion of experimental research, with new findings and insights about the disease announced almost every month.

To physicians, the discovery of the HD gene was of great benefit in the accurate diagnosis of HD and in allowing interested individuals to know in advance of any symptoms whether they were gene carriers. To researchers, the discovery of the HD gene allowed basic experimental studies of HD to begin. With the tools of modern molecular biology, it is possible to "cut and paste" genes and parts of genes and to identify genes in different animal species that are similar to the human HD gene. Using these tools, scientists have inserted the abnormally elongated HD gene in a culture flask, into the eye of a fruit fly, into a tiny worm called *Caenorhabditis elegans*, and into a mouse. These "HD models" can be used in experiments to understand different aspects of the gene, the HD protein, or the disease. For example, when the abnormal HD gene is inserted into the eye of a fruit fly, the nerve cells in the eye degenerate. Because scientists have already identified many of the fruit fly's other genes, this model can be used to determine what other genes in the fruit fly might hasten, slow or prevent the loss of nerve cells caused by the HD gene. But to study changes in the location or function of the abnormal HD protein within the cell, it may be easier to look at isolated cells grown in a culture flask, or to study the changes that take place in the brain during the course of the disease, or to study new chemicals that might delay or prevent the disease. However, it is better to study the human and develop a neurological disease comparable to HD.

The first exciting lead that came from this new air of research was discovery that, unlike the normal HD protein which is exclusively located in the outer part of the cell, three unusual things happen to the abnormal HD protein. It is cut by enzymes called "caspases." Right at that abnormally long part out of the protein where the amino acid glutamate appears too many times in a row. Then the cut piece of the HD protein gets into the nucleus, or center of the cell (where it doesn't belong). Finally, the abnormal protein, along with a few other proteins, forms a "glob" (called an "intranuclear inclusion" or "aggregate") in nucleus of the cell. Since the discovery, scientists have focused their attention on how these three steps lead to HD and what compounds can be used to prevent each step. Several reports in 1999 and 2000 suggest that the information of intranuclear inclusions may not cause HD, but that the cutting of the HD protein by caspase-1 probably is an important step in the disease. Several compounds that inhibit caspase-1 are being studied in the HD mouse and the early results have been promising.

HD researches are benefiting from related studies of other diseases. Encouraged by reports of improvement in individuals with Parkinson's disease who have undergone brain cell transplant procedures, several groups of investigators are looking at the feasibility and safety of cell transplants in HD. Other researchers are looking at the delivery of nerve growth factors into the brain, either through a catheter or by "gene therapy," with the thought that these factors may promote repair or regrowth of brain cells damaged by HD. Still others are looking at the possibility of using biological tools similar to those used by cancer researchers that "turn off" genes that are too active in cancer cells, to selectively "turn off" the abnormal HD gene.

At the same time, clinical researches around the world are learning how to conduct clinical trials of therapies in HD. Experience with other degenerative disorders such as Multiple Sclerosis, Alzheimer's disease and Parkinson's disease have shown that clinical research studies must be designed very carefully

for their results to be meaningful. The Huntington's Study Group, a consortium of over 50 centers in North America and around the world, as well as research trial groups in Europe, have already begun trials of new therapies in HD.

Thus, the dawn of a new century begins with it a great hope for new treatments for this difficult disease. With a reason for optimism, there is hope that it will be easier for the worldwide community of those with juvenile HD, their families and friends, their physicians and the researchers to walk together into the future, improve the lives of those who already have the disease, and find the final pieces to the puzzle of HD.

Appendices

Table of contents

| | |
|---------------|------------------------------------|
| Appendix I | General Resources |
| Appendix II | Huntington's Disease Organizations |
| Appendix III | Medical Equipment & Supplies |
| Appendix IV | Health Care Resources |
| Appendix V | Additional Resources |
| Appendix VI | Research |
| Appendix VII | HDSA Chapters |
| Appendix VIII | HDSA Centers of Excellence |

Appendix I

General Resources

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| <p>American Speech-Language-Hearing Association 1081 Rockville Pike Rockville, MD 20852 (301) 897-5700 Website: www.asha.org The Mission of the American Speech-Language-Hearing Association is "To promote the interests of and provide the highest quality services for professionals in audiology, speech language pathology, and speech and hearing science, and to advocate for people with communication disabilities"</p> <p>Choice in Dying, Inc. 1035 30th Street, NW Washington, DC 20007 (800) 989 WILL (202) 338-9790 _ (202) 870-2003 Fax Website: www.choices.org</p> <p>Choice in Dying is a national, non-profit organization dedicated to fostering communication about complex end-of-life decisions among individuals, their loved ones and health care professionals. It provides the national hotline to respond to patients and their loved ones in end-of-life-crises. UPDATE: Choice in Dying is now called Last Acts. Their website is www.lastactspartnership.org</p> <p>LifeWise Family Financial Security, Inc. 136 South Main Street, Suite 700 Salt Lake City, UT 84101 (800) 219-7385 Website: www.lifewisefinancial.com This company provides loans using an existing life insurance policy as collateral.</p> | <p>Hereditary Disease Foundation (HDF) 11400 W. Olympic Boulevard, Ste. 855 Los Angeles, CA 90064 (310) 575-9656 Website: www.hdfoundation.org</p> <p>LifeWise Foundation 136 South Main Street, Suite 701 Salt Lake City, UT 84101 (800) 942-2338 Website: www.lifewisefoundation.org The foundation is a public charity created to help alleviate the financial burdens of terminally ill patients and their families. Services include giving grants to terminally ill patients who are facing significant financial challenges and are unable to meet eligibility requirements for state and private aid.</p> <p>Social Security Administration (SSA) Office of Public Inquiries 6401 Security Boulevard Room 4-C-5 Annex Baltimore, MD 21235 Website: www.ssa.gov/SSA_Home.html The government agency oversees the Social Security, Supplemental Income (SSI) and Medicare programs. Social Security provides a monthly income for eligible elderly and disabled individuals. SSI provides Social Security payments for individuals who have certain income and resource levels. Medicare is a federal health insurance program for those who are 65 or older, people of any age with permanent disability, and disabled people under age 65 who have received Social Security payments for at least 24 months. Contact the SSA to get information on eligibility and coverage and to find out how to apply for these programs.</p> |
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National Organization for Rare Disorders (NORD)

P.O. Box 8923
New Fairfield, CT 06812
(800) 999-NORD (voice mail)
(203) 746-6518
Website: www.rarediseases.org
Email: orphan@rarediseases.org

It provides information on rare diseases and helps patients in obtaining medications they otherwise not be able to afford. It also facilitates patient networking services and increases awareness of clinical trials.

California

Family Caregiver Alliance Statewide Helpline
Will make statewide referrals
(415) 434-3388
(800) 445-8106

Genetically Handicapped Persons Program

This is a state funded program which coordinates care and helps pay for medical costs of persons with various genetic transmitted conditions

(619) 543-3500

Southern Caregivers Resources Center

3675 Ruffin Road, Suite 230
San Diego, CA 92123

268-4432

Texas

The Texas Information and Referral Network

4900 North Lamar, 4th Floor
Austin, TX 78751

(512) 424-6520

25 area information centers across the state providing information about general health and human services.

Appendix II Huntington's Disease Organizations

Huntington's Disease Society of America

158 West 29th Street, 7th Floor
New York, N.Y. 10001-5300
(800) 345-HDSA
(212) 242-1968
(212) 239-3430 fax
Website: www.hdsa.org
Email: hdsainfo@hdsa.org

The Huntington's Disease Society of America (HDSA) is dedicated to finding a cure for Huntington's Disease while providing support and services for those living with HD and their families. HDSA promotes and supports both clinical and basic HD research, aids families in coping with the multifaceted problems presented by HD and educates the families, the public and health care professionals about Huntington's disease. Our HD families give a face to Huntington's disease. HDSA is its voice.

Huntington Society of Canada

151 Frederick Street, Suite 400
Kitchener, Ontario N2H 2M2
(519) 749-7063
(519) 749-8965 fax
1-800-998-7398 Toll Free in Canada
Website: www.hsc-ca.org
Email: info@hsc-ca.org

The Huntington Society of Canada (HSC) is a national network of volunteers and professionals united in the fight against HD since 1973. Our goal is to find new treatments and ultimately a cure for Huntington's disease, and to improve the quality of life for people with HD and their families.

International Huntington's Association

Callunahof 8
7217 ST Harfsen
The Netherlands
+31-573-431595
+31-573-431719 fax
Website: www.huntington-assoc.com
Email: iha@huntington-assoc.com

The International Huntington's Association (IHA) is a federation of national voluntary health agencies that share common concern for individuals with HD and their families. Each agency promotes lay and professional education; individual and family support; psychosocial, clinical and biomedical research; and ethical and legal considerations related to Huntington's Disease in its respective country. The IHA website lists the contact information for each of its member agencies, including the HDSA.

Appendix III Medical Equipment and Supplies

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| <p>Aid In Daily Living products http://www.alsa.org/resources/product.cfm Email: alsinfo@alsa-national.org</p> <p>Broda Seating (800) 668-0637 http://www.brodaseating.com/</p> <p>Canine Companions for Independence (407) 834-2555 or Toll Free 1-800-572-2275 http://www.caninecompanions.org/</p> <p>Clothing for children with special needs www.special-clothes.com www.stitchesfromtheheart.com www.conforthouse.com</p> <p>Food thickeners Diamond Crystal (206) 623-7140 www.diamondcrystal.com</p> <p>THICK IT® 1-800-333-0003 http://www.precisionfoods.com/consumer/thick.asp</p> <p>Power wheelchair purchase http://www.lougehrigsdisease.net/images/images%20equip.htm/wcpurchase.htm</p> <p>Identi-Fund Iron-on clothing labels, wallet cards P.O. Box 567 Canton, NC 28716 (828) 648-6768 www.identifund.com Email: labels@primeline.com</p> <p>Florida Durable Medical Equipment Services Advocacy Center- Technology Assistance Program (TAP) – Main Office 2671 Executive Center Circle West Webster Building #100 Tallahassee, FL 32301-5024 (850) 488-9071 TAP is authorized to provide investigational, negotiation and litigation to promote the provision of assistive technology services.</p> <p>Medic Alert Systems Medic Alert Systems are also known as Emergency Response Systems. Typically it is a home monitoring device for individuals who are alone during the day or evening. There are several different types. The following is a list of Medic Alert Systems.</p> <p>Medic Alert Foundation Body worn emblems, wallet cards 2323 Colorado Avenue Turlock, CA 95382-2018 (800) 432-5378 (209) 669-2495 fax www.medicalert.org Email: postmaster@medicalert.org</p> | <p>Information Technology (800) 331-3027 A free information and referral service on assistive technology. They provide free, up-to-date product information on commercially adaptive equipment for people with disabilities or people who are elderly.</p> <p>Resna Technical Assistance http://www.resna.org/taproject/at/connections.html Offers a state-by-state contact list. The programs are designed to help children and adults with disabilities across all aspects of their lives - education, employment, recreation and independent living.</p> <p>Mcard Corporation Plastic Wallet card, identification seal 6320 W. 159th Street, Suite A Oaks Forest, IL 60452 (708) 535-7215</p> <p>New Care Therapies Vail Beds and other supplies Shelbyville, KY (800) 432-6249 http://www.newcaretherapies.com</p> <p>SOS America, Inc. Bracelets, pendants, watch and sneaker attachments P.O. Box 260 Massapequa, NY 11758 (516) 795-3960 _ (800) 999-1264 www.sosamerica.com Email: info@sosamerica.com</p> <p>Texas</p> <p>Baylor Home System Dallas (204) 820-2229</p> <p>LifeLine Metro/Johnson County (817) 551-2790</p> <p>LifeLine/SPAN Denton and surrounding areas (940) 382-2224</p> <p>Lutheran LifeLine Covers the entire state of TX (800) 598-0520</p> <p>Memorial LifeLine Huston Area (800) 358-4999</p> |
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Appendix IV - Health Care Resources

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| <p>Health Care Resources</p> <p>Home Health Care National Association for Home Care (NAHC) 228 Seventh Street, SE, Washington, DC 20003 (202) 547-7424 (202) 547-3540 - Fax www.nahc.org - info@nahc.org NAHC's mission is to promote quality care to home care and hospice patients, preserve the rights of caregivers, effectively represent all home care and hospice providers and place home care at the center of health care delivery.</p> <p>Nursefinders Central Office (817) 460-1181</p> <p>Massachusetts Boston Center for Independent Living 95 Berkeley St., Suite 206 Boston, MA 02116 Contact information and referrals for personal care attendants. (617) 338-6665</p> | <p>Visiting Nurse Association of America 99 Summer Street, Suite 1700 Boston, MA 02110 (617) 737-3200 (617) 737-1144 Fax (888) 866-8773 www.vnaa.org - vnaa@vnaa.org A national toll-free referral line provides callers with information about the nearest Visiting Nurse Association in their area. The Association's services include general nursing, physical, occupational and speech therapy, medical and social services, case management, personal care, high-tech therapies, adult day care, parent aid, care for the dying, nutritional counseling, friendly visit services and Meals on Wheels.</p> <p>Massachusetts Rehabilitation Commission Home Care Assistance Program A statewide organization that provides for decor service for people on the 60 who have a disability and a limited income. (800) the 245-6543</p> |
| <p>Visiting Nurses Association Covers Massachusetts, southern New Hampshire, and Rhode Island (800) 562-4700</p> <p>New York Visiting Nurses Service Covers all NYC Boroughs (212) 290-3800</p> <p>Texas Girling Health Care Coverage throughout Texas, call for specific area office. (800) 580-3683</p> <p>NurseFinders Coverage throughout Texas, call for specific area office. (800) 264-5445</p> | <p>Olsten Health Services Coverage throughout Texas, call for specific area office. (800) 341-9597</p> <p>Hospice National Hospice and Palliative Care Organization (NHPCO) 1700 Diagonal Road, Suite 300 Alexandria, VA 22314 (800) 658-8898 (helpline) (703) 243-5900 (main line) (800) 338-8619 www.nhpco.org NHPCO is dedicated to improving quality of life and end-of-life care and increasing access to hospice care. NHPCO's National Hospice Helpline provides information and referrals to local hospice programs.</p> |

Appendix V-Additional Resources

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| <p>Web Sites Children of books to assist healthy children to understand children with disabilities. www.joniandfriends.org/helps/child-bk.htm no longer valid</p> <p>Journey of the Heart: A Healing Place in Cyberspace www.journeyofhearts.org A place for resources and support for people in the grief process following a loss or significant life change.</p> <p>Listing of schools for special-needs children www.funrsc.fairfield.edu/~jfleitas/contents.html no longer valid</p> <p>Parents' Common Sense Encyclopedia www.drhill.???/EncyMaster/index.html no longer valid</p> | <p>Special-needs children's bulletin board http://boards.parentsplace.com/messages/get/ppspeci/needs5.html New link: http://messageboards.ivillage.com/iv-psdisability</p> <p>Special-needs tips www.geocities.com/Hotsprings/Spa/6889/SNTips.htm no longer valid</p> <p>Support for adult caregivers of special-needs children http://www.our-kids.org/OKAdults/</p> <p>What is a gene? http://kidshealth.org/kid/talk/qa/what_is_gene.</p> |
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Appendix VI Research

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| <p>DNA bank Indiana University medical Center Indianapolis, IN 46202 (317) 274-5744 The DNA bank was established for the purpose of storing genetic material for possible future use. Costs to store a DNA sample is \$70.00.</p> <p>Harvard Brain Tissue Resource Center (HBTRC) McLean hospital 115 Mill St. Belmont, MA 02478-9106 (800) BRAIN-BANK www.brainbank.mclean.org the HBTRC has been established as a centralized resource for the collection and distribution of human brain specimens for research in a broad range of neurological disorders, including Huntington's disease. It is essential that the donation procedure or within 24 hours of the time of death of the donor.</p> <p>HD Roster Indiana University medical Center 975 W. Walnut St. Indianapolis, IN 46202 the HD Roster is our Vital link between scientists and HD families to facilitate clinical trials/research. Volunteers are invited to complete a questionnaire about their HD history. This confidential information is then stored until a research project requires volunteers that match specific criteria. Those volunteers are then contacted by the Roster and asked to participate in a trial or study. There is no charge to register with the Roster.</p> | <p>Huntingtons Study Group (HSG) 1325 Mt. Hope Avenue, Suite 160 Rochester, NY 14620 (716) 275-9138 (800) 487-7671 www.huntington-study-group.org The Huntington's Study Group conducts clinical trials for patients with Huntington's disease.</p> <p>National Neurological Research Bank Los Angeles, CA (310) 268-3536</p> <p>NIH Clinical Trials Database http://clinicaltrials.gov The U.S. National Institutes of Health (www.nih.gov), through its National Library of Medicine (www.nlm.nih.gov), has developed www.ClinicalTrials.gov to provide patients, family members, and members of the public current information about clinical research studies. Check often for regular updates.</p> |
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| <p>Appendix VII – HDSA Chapters</p> <p>This information is not listed in this eBook. The HDSA chapter information is subject to changes. For the most current information or to locate the nearest HDSA Chapter to you, go to this HDSA webpage and then to your state: http://www.hdsa.org/chapters/Locations.pl</p> <p>Appendix VIII - HDSA Centers of Excellence - HDSA Centers of Excellence for Family Services are the cornerstone of HDSA's commitment to caring for HD families across the US. These multidisciplinary medical facilities bring together allied healthcare professionals, experienced in HD or</p> | <p>Appendix VIII continued</p> <p>movement disorders, to provide services and support to HD families. Centers of Excellence work in tandem with HDSA chapters, affiliates and support groups to form a national network of referrals and support. New Centers are added each year, so please contact HDSA at (800) 345-HDSA [4372] or visit the HDSA Webpage http://www.hdsa.org/getting/centers.pl to locate the HD Center of Excellence nearest to you.</p> <p>Added: You may also want to check out this State-By-State HD support information: http://huntingtondisease.tripod.com/statebystatesupport/</p> |
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