

# The Juvenile HD Handbook

*A Guide for Families and Caregivers – SECOND EDITION*

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## Disclaimer

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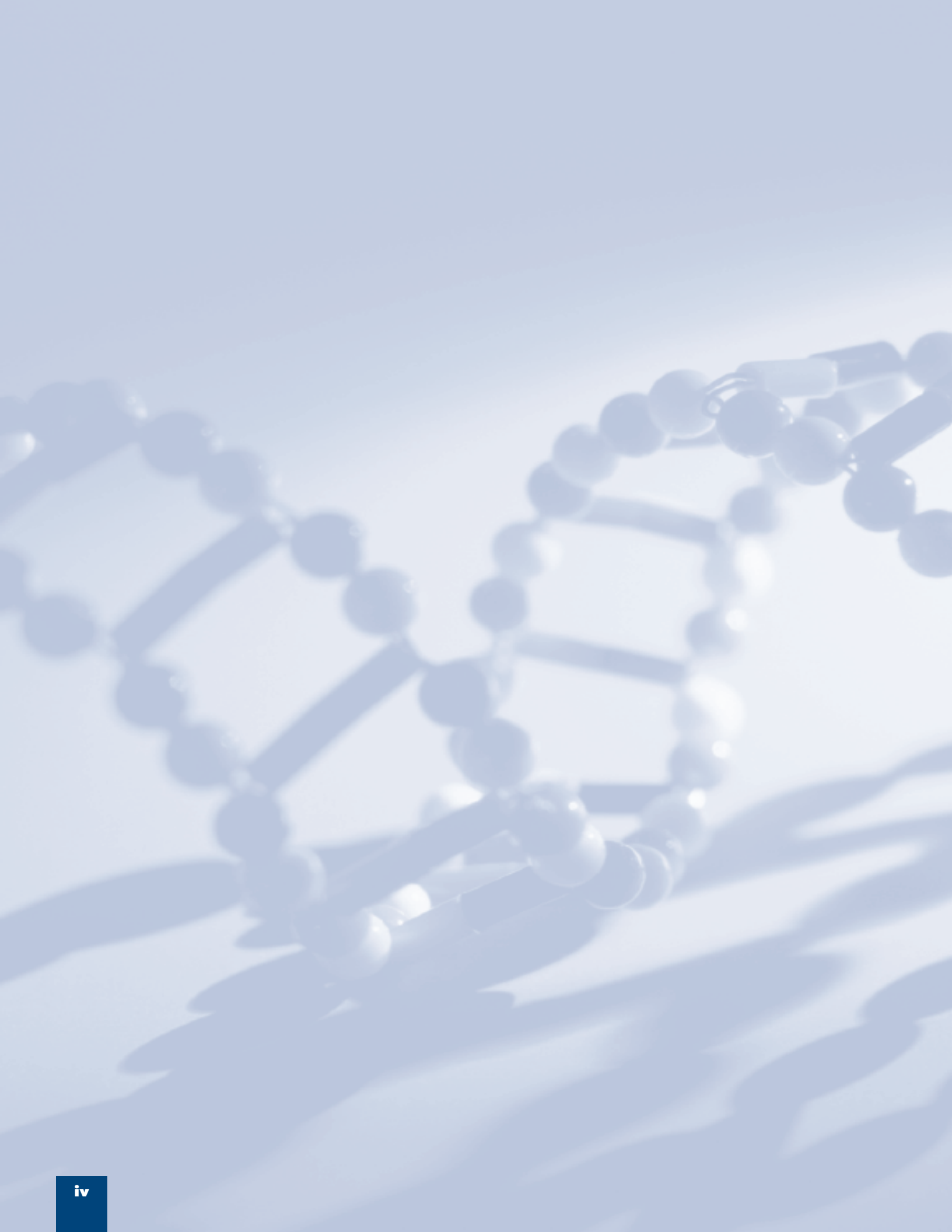
This book is dedicated to  
Danny, Davey, and Dawn

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## INTRODUCTION

*Huntington's Disease (HD) is a genetic neurologic disorder that leads to a movement disorder, dementia, and behavior disturbance. Although the gene that causes HD is present throughout a person's life, the symptoms do not usually begin until mid-adulthood. In the United States, the prevalence is about 1 per 10,000 individuals (30,000 Americans). 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 ten percent of individuals with HD develop symptoms before age 20<sup>1</sup>. Because it is uncommon and differs from typical adult-onset HD, in both the neurologic symptoms and the 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 uses to make the diagnosis of Huntington's Disease.*

*Many parents, families and health professionals are uncertain about the role of 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 of genetic testing for children is provided here; a 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, and 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.*

<sup>1</sup> Peter S. Harper, *Huntington's Disease*, NY: W.B. Saunders, 1996.





# The Diagnosis of HD in a Child

## When to Consider HD

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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 symptom or group of symptoms that are absolutely required for the diagnosis of Juvenile HD, most affected children have several of the features described (see box) 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.

### 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 more 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 diagnoses 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.

### **Typical initial symptoms of juvenile HD**

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- 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
- Chorea in an adolescent
- Behavioral disturbances

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 a 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 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, frequently leading to a diagnosis of attention deficit disorder (ADD) (of course, ADD is a common condition that can certainly be present in a child who does not have HD). In a younger child, increasing difficulty with previously attained cognitive or motor skills, such as speech, reading, math, throwing a ball, swimming or riding a bicycle, might be evident. In a young child or pre-teenager, the combination of declining school performance along with worsening motor skills would be 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 kinds and causes of dysfunction at school.

### **BEHAVIORAL DISTURBANCE**

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 and behavioral problems in the children. In younger children, when aggressive or disruptive behavior is related to HD, it is usually seen along with changes in cognitive function and motor disturbance. In adolescents, behavioral disturbance may be the first and only presenting symptom of HD. Behavior problems in adolescents with HD are often very severe, leading to psychiatric hospitalization, suspension from school, or involvement with law enforcement agencies. Examples have included arson, theft, 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 (chorea), 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, adolescents are more likely to have chorea as a presenting symptom.

## SEIZURES

Seizures are said to occur in about 25% of children with Juvenile HD<sup>2</sup>, 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 drug or toxin 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 neurologic examinations before mild or intermittent symptoms are recognized by the physician as being due to HD.

*A parent should understand that the diagnosis of Juvenile HD is unlikely to be made during the first visit and should be prepared to work with the physician through the process of diagnosis.*

## 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. 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 the diagnosis of Juvenile HD is unlikely to be made during 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 developmental 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

<sup>2</sup> Peter S. Harper, Huntington's Disease, NY: W.B. Saunders, 1996

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, developmental skills, and intelligence) can document areas of strength and weakness, suggest strategies for 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 to make appropriate referrals for individual or family counseling, county-based child protective services, school-based programs, or social services. Physical, occupational, and speech language pathologists can perform baseline assessments of motor skills with an emphasis on how the child is able to function in school and at home. Children in HD families can have 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 but may be helpful 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).

### Steps in the diagnosis of Juvenile HD

- History of a change in motor, behavioral, and cognitive function
- Family history of HD
- Abnormal neurological examination
- Abnormal neuropsychological test results
- Genetic test confirming the presence of an abnormally long "CAG repeat" sequence in the HD gene
- Family counseling

When the history, examination, and initial laboratory evaluation are strongly suggestive of HD, a genetic test may be the most efficient and accurate way to confirm the diagnostic impression. On the other hand, if the symptoms are not typical of HD, or the diagnosis of HD is not clear from the examination, then the doctor should not order the HD gene test immediately, as the gene test cannot determine the cause of a person's symptoms. 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. It is recommended that the neurologist evaluate the child twice, six to twelve months apart. If the initial

symptoms have remained or progressed despite optimal management, then it may be appropriate to do the HD gene test at the time of the second visit. This strategy makes it less likely that a child with temporary or non-progressive symptoms would be tested prematurely.

The diagnosis of Juvenile HD, whether based on a clinical examination alone, or secured with a gene test, gives an explanation for symptoms that may have confused and frightened the child and the family. This can relieve stress and can minimize "acting out" on the part of the child. It also gives

the family and doctors a more clear direction regarding the prognosis and care. Thus, it is equally important for the physician to make a diagnosis of Juvenile HD in a timely fashion, and not to postpone or refuse genetic confirmation of the diagnosis in a child whose symptoms and course suggest that the diagnosis of Juvenile HD is quite likely.

## Diagnostic Genetic Testing in Children

The ability to detect changes in the HD gene itself has made confirmation of the diagnosis of HD relatively simple. The genetic material 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, while any number higher than 35 can lead to the development of HD. There is a relationship between the repeat number and the age that HD symptoms begin, such that higher repeat numbers tend to result in a younger age of onset. Most adults with HD have between 40-50 CAG repeats in their abnormal HD gene. Usually, Juvenile HD is associated with CAG repeats of 50 or higher, although this is not always the case. CAG repeat lengths of over 200 have been reported in children with onset below age 3.

The gene test is close to 100% accurate. If the test shows two normal HD genes, the child will never develop HD and is not at-risk for passing HD on to his or her 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 HD gene test result, the physician should contact the laboratory or a genetic counselor to discuss the possibility of special testing to look for very large repeat lengths.

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 due to HD. For example, an adult whose HD gene test was felt to “explain” his blurry vision and headaches was incorrectly diagnosed as having HD and, as

### 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

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 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 detail, a genetic counselor may be asked to help with this part of the process. The table at left shows some of the risks of identifying a child as being gene positive prematurely.

## 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 abnormal HD gene, it is very important to consider the complex potential effects of the test result 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 include adverse psychological, social, financial and medical consequences. Most genetic testing experts believe that predictive genetic testing 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 the great majority of 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 she or he 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. Genetic testing of an infant prior to adoption also represents a predictive test of a non-consenting child, and is strongly discouraged by genetics professionals in the United States.

# 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 or 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 becomes impaired 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 age of onset, 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.

## 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 language pathologist 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 very few "experts" in Juvenile HD, but all pediatricians have seen children with chronic diseases, all speech language pathologists have seen patients with speech and swallowing problems, and physical

### **Effective team management is likely to include:**

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

### **Physical therapy in Juvenile HD**

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- 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

### **Occupational therapy in Juvenile HD**

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- Safety evaluation
- Assessment of activities of daily living
- Assistive devices

### **Speech Language Pathology in Juvenile HD**

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- Outpatient or home speech therapy/oral exercises
- Training with assistive devices
- Assessment of dysphagia
- Strategies to avoid choking
- Modification of food textures and types, eating/feeding strategies

### **Public health nurse home visit**

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- 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

### **Dietary consultation**

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- Assessment of ideal body weight, dietary habits, food preferences
- Identification of unhealthy food habits, obsessions, or food-related behavioral problems
- Instruction in preparation of modified diets
- Monitoring of weight and calorie intake
- Information about gastrostomy tube feedings

therapists work daily with children who have difficulty with walking and coordination.

The family and the 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 has established regional Centers of Excellence for Family Services where the services needed by an HD family 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, a local pediatrician who listens, is willing to learn, and is available when there is a crisis, may be more useful than traveling long distances to a specialist.

### **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 it 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 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 communities do have support groups for parents of children with chronic medical or 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.

It is doubtful that any parent can cope with his or her child's serious chronic illness without talking about it to someone sometime. Resources do exist for both short-term and long-term management of your own emotions. Getting the emotional support that you need is crucial for your health, the health of your HD affected child, and for the rest of your family. Resources are listed in the Appendix.

### *The Rest of the Family*

The child with HD does not live in isolation. More so 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 other 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.

#### THREE FAMILY SCENARIOS ARE COMMON

- 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.
- 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. Other children in the family may not be at-risk for HD.
- 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.

There is no single "right way" to tell the other children about a child's HD

*Explaining to other 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.*

*“Harry was accepting, the ramifications of HD unknown to him. Jane enjoyed life the best she could. Howard has been hostile and afraid. Through determination, faith, cauliflower-eared friends and humor, I have survived.”*

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 cancer or AIDS, 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 HDSA's web site at [www.hdsa.org](http://www.hdsa.org)).

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. We believe this approach is unproductive, and that families should try 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.

# Medical Care

## Overview

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The core features of HD in a child or an adult are 1) movement disorder (difficulty with movement), 2) cognitive disorder (dementia), 3) behavior and psychiatric issues (mood or behavior changes). Even these core features look different from one person to the next, and some symptoms that are common in adults are seen less often in children. Seizures, which are uncommon in adults, on the other hand, are quite common in Juvenile HD.

Death in Juvenile HD, unless it occurs unexpectedly due to accident or injury, is preceded by a time when the person is increasingly unable to communicate or eat safely. Because swallowing and communication problems are so universal in Juvenile HD, we devote a separate section to their management.

Until researchers find treatments that slow, stop, or reverse HD, families will likely find themselves frustrated as they watch the decline in function that accompanies the disease. A good relationship with the medical team can help to reduce the frustration, but even then, you may still find yourself wanting to try “alternative” therapies. Alternative therapies and medications are discussed at the end of this chapter.

Because Juvenile HD progresses gradually over years, it remains important to attend to the child’s general health needs, such as immunizations, dental care, and other age-appropriate evaluations. Every child with Juvenile HD should have a pediatrician or family doctor who takes care of these general health needs. As the disease progresses, the general physician can help to watch for and treat the expected medical complications of HD.

## The Movement Disorder

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### SYMPTOMS

HD is classified by neurologists as a “movement disorder,” although it also has profound effects on cognition and behavior. Children who show symptoms of HD before age 10 often develop stiffness of the limbs, usually most severe in the legs. The child may begin to walk on his/her toes, lose control of balance when running, hopping, or bicycling, or develop a scissoring or stiff-legged gait. Many young children have a loss of control of oral motor function, which can cause slurring of speech,

difficulty swallowing, or drooling. Later, the neck or trunk may also become stiff. These symptoms are caused by an increase in muscle tone, called “rigidity” or “spasticity”. Twisting movements or stiff, twisted postures may be called “dystonia”.

Older children (teenagers) with HD may develop chorea - the involuntary, irregular, fidgety or jerky movements of the arms, legs, trunk, neck, or face that are one of the most common symptoms in adults. Younger children with HD rarely have chorea.

In addition to an increase in muscle tone, children with HD have difficulty controlling voluntary movements. Clumsiness becomes noticeable, and the child may have difficulty with previously learned skills, such as throwing a ball, writing, or riding a bicycle. As the disease progresses, the child will have more difficulty performing a sequence of movements, such as those needed to bathe or dress. Eventually, even simple single movements can be difficult, and awkward stiff postures of the limbs or trunk, called dystonia, may become severe.

## TREATMENT

No medications improve control of voluntary movements, although there are treatments for rigidity, spasticity, dystonia, and chorea. Physical therapy and occupational therapy consultation may be very useful throughout the course of the disease. The role of speech-language pathology in addressing oral motor problems is discussed later in the chapter.

We wish to emphasize that early intervention is important with all therapies — even before it appears that the intervention or device is actually necessary. This is because cognitive decline is a major part of HD. A child may be better able to learn how to use a new device or therapy if it is introduced while he/she is able to understand and cooperate more easily. By the time the need for the device becomes more obvious, it is a familiar thing that can easily be incorporated into daily use.

The Juvenile HD 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 or other Juvenile HD families what adaptations have been helpful to them. It is good to bring your ideas or questions to an appointment with the therapist, so that you can work together to find ways to improve your child’s function and quality of life. It is also possible to request from the doctor an order for a public health nurse assessment. The nurse can come to your house to do a room-by-room assessment of safety and function, and make recommendations. It may be possible, with a doctor’s order, to have certain devices, equipment, or medically necessary home remodeling activities that are not covered by medical insurance be paid for from a pre-tax health savings account. Check with your health insurer or a social worker to learn more about your options.

### **Medications used to treat chorea**

- Dopamine-blocking agents
  - Haloperidol
  - Fluphenazine
  - Olanzapine
  - Risperidone
  - Others agents
- Dopamine-depleting agents
  - Tetrabenazine (not yet approved in the US)
- Benzodiazepines
  - Diazepam
  - Clonazepam
  - Lorazepam
  - Other agents
- Other agents and treatments
  - Amantadine

### **Treatments for spasticity and rigidity/dystonia**

- Spasticity
  - Baclofen
  - Tizanidine
  - Diazepam (and other benzodiazepines)
  - Dantrolene
  - Physical therapy
- Rigidity/dystonia
  - Anticholinergic agents (Trihexyphenidyl, Benztropine)
  - Carbidopa-levodopa
  - Dopamine agonists (Pramipexole, Ropinirole)
  - Botulinum toxin injection
  - Physical therapy

### **MEDICATIONS**

Doctors will often prescribe medications to relieve the stiffness that accompanies Juvenile HD, or to reduce chorea if it is present. Whenever a medication is prescribed, it should be clear what symptom it is being used for, what the starting dose is, whether there is a plan to increase the dose, when the effects of the medication will be checked, and what the possible side effects are, and what to do if undesirable side effects occur. Some children will have dramatic benefits from a particular medication, and, unfortunately, others may not.

Medications sometimes used to treat chorea are listed in the box at left. None has been proven to be more effective than any of the others, and none are FDA-approved for the treatment of chorea in HD. All can have significant side effects such as sedation, worsening of rigidity, or depression. A US research study recently showed significant benefits on chorea with the use of tetrabenazine, and this drug has been submitted to the FDA for consideration, but has not yet been approved for use in this country. Other drugs that have effects in the brain, such as anticonvulsant (seizure) medications, are rarely tried but occasionally have benefit.

Medications sometimes used to treat rigidity and spasticity are shown in the second box. In general, any medication that reduces rigidity might worsen chorea. None are approved in HD, but all are used to treat similar symptoms in a wide range of medical conditions. Botulinum toxin (Botox) injections are given directly into a muscle to relax it; the effects of the toxin last for several weeks. Botox injections are not practical for large muscles, but if dystonic posturing of the jaw, hand, or a particular leg muscle, is interfering with a child's safety, care, or feeding, injection of one or a few muscles might be beneficial. The role of physical therapy is discussed below.

### **PHYSICAL THERAPY**

The physical therapist addresses large body movements, such as walking, and movement in and out of a chair, bed, or car as well as problems with posture, muscle tone, and positioning in a chair or bed. The physical therapist can help throughout the course of Juvenile HD. Initially, all children diagnosed with Juvenile HD should have an age-appropriate activity program that emphasizes full range of motion of the limbs, trunk, and neck, and aerobic exercise. The physical therapist can help to develop this program. Knowing that problems with balance are

*Knowing that problems with balance are in the future, the therapist can instruct the child and family on simple strategies to reduce the risk of falls when standing, turning, or sitting, and on how to help the child to get up after a fall.*

in the future, the therapist can instruct the child and family on simple strategies to reduce the risk of falls when standing, turning, or sitting, and on how to help the child to get up after a fall. As the disease progresses, balance and falling concerns will become evident. Assistive devices such as canes and walkers are not helpful if there is significant chorea, but if there is significant rigidity with little or no chorea, a walker with wheels (equipped for indoor and outdoor use) may be helpful. A child tends to fall the same way repeatedly; some always land on their knees, others hit their head or an elbow or the buttocks. Bicycle or hockey helmets and hockey or skateboard pads for the parts that repeatedly get injured can be helpful. It is useful to introduce a wheelchair before it is absolutely necessary, so that the child (and family) becomes familiar with it and learns how to use it.

Physical therapy consultation is particularly important for children with rigidity, spasticity, or dystonia. Heat, stretching, and massage can temporarily reduce increased muscle tone. Splinting or bracing of a limb may improve function. Positioning on the chair, wheelchair, or bed, and protection of stiff limbs or joints to prevent bruises or skin tears may be important. In the late stages, a child's mobility may be severely reduced, simple movements may be infrequent, and dystonia may be severe. By this time, special seating devices, cushions, mattresses, and padding may be needed. Heat, stretching, and massage may help to relieve discomfort caused by dystonia. Ensuring comfort is particularly important at this time, as the child may not be able to communicate his/her own needs well.

### OCCUPATIONAL THERAPY

The occupational therapist often focuses on small movements and the performance of daily cares such as bathing, dressing, and eating, with an emphasis on safety and improved and independent function. In the early stages of the disease, an assessment can provide a baseline for later comparison, and can also alert the family to things that should be watched. In the middle stages of HD, parents or teachers will notice changes in function and the occupational therapist should be brought in again to consider assistive devices, adaptive equipment, or environmental adaptations to improve safety and function while maximizing independence. For example, adaptive plates and utensils can help a child to continue eating independently, while the use of Velcro closures and elastic shoelaces rather than buttons, zippers, and tying laces may improve independence in dressing. The occupational therapist is a useful resource in assessing home safety and improving daily function. In the late stages of the disease, particularly if the family intends to keep the child at home, a reassessment may be necessary. The child may outgrow previous equipment or devices, and illness or disability in an aging parent may require a different strategy for care or safety.

*“Thinking becomes very literal. My child will take a teacher’s joke as the truth and obsess over it.”*

## The Cognitive Disorder

### SYMPTOMS

HD creates changes in the brain that cause difficulties in thinking and reasoning. These changes can be subtle, especially in the early stages of the disease. Sometimes symptoms appear as inattention or hyperactivity. The child may become anxious or aggressive because he or she is confused, distracted, or overwhelmed by stimulus. The child may have difficulty starting or completing a task, or trouble doing all the steps of a task in the right order. The cognitive symptoms of HD can be mistaken for “bad behavior,” when in actuality it is the disease that is interfering with the child’s ability to concentrate.

### MEDICATIONS

No medications have been proven to improve cognitive function in HD. However, if frequent seizures, attention deficit, or depression are interfering with a child’s ability to perform, treating these symptoms may reduce clouding of thought and improve the child’s ability to learn. At the same time, both prescription drugs and illicit drugs or alcohol can cloud a child’s thinking processes.

Several medications are FDA-approved and widely used for the treatment of dementia in people with Alzheimer’s disease (donepezil, rivastigmine, galantamine, and memantine), but none have been evaluated in more than a handful of adults with HD, and none can be recommended to treat Juvenile HD.

### Addressing Cognitive Symptoms

HD makes it difficult to organize incoming stimulus. A noisy room or complicated instructions can overwhelm the child and trigger outbursts of emotion or aggression. Many of the cognitive symptoms of HD can be reduced using simple techniques such as breaking tasks into a step-by-step format or using checklists to help the child keep on track. Some of these techniques are discussed in Chapter 4 – Daily Life.

### THE COGNITIVE DISORDER IN THE SCHOOL ENVIRONMENT

Changes in cognition will cause challenges at school. Over time, the child with HD will lose the ability to learn new material and academic skills will decline. Public schools are required to provide educational experiences for children with handicaps of all types. As soon as a diagnosis of HD is made,

*HDSA has created a book and CD-ROM set, Meeting the Challenges of JHD: Best Practices for the School Environment, to help parents to work effectively with teachers and schools in designing a good experience for the child with Juvenile HD.*

the family should meet with 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 at least once a year, although more frequent reviews may be needed as the diseases progresses.

HDSA has created a book and CD-ROM set, *Meeting the Challenges of JHD: Best Practices for the School Environment*, to help parents to work effectively with teachers and schools in designing a good experience for the child with Juvenile HD. The set contains information about the disease in text form and also as PowerPoint presentations which can be shown to a group. There are presentations appropriate for classmates as well as the teacher and school administrators. The CD-ROM also includes an extensive set of links to Internet resources for Juvenile HD. Please contact HDSA to request a free copy of this valuable resource.

Our hope is that the information presented will help teachers and other school officials to understand what to expect from a child who has HD, and the expected period of time over which changes will take place.

### **THE COGNITIVE DISORDER IN THE HOME**

At home, the family should try to "Keep it Simple." Routines are very reassuring to a child who has memory problems or dementia. Establishing routines early in the course of the disease can be helpful in maintaining a safe and stimulating environment. Routines can include everything from getting dressed in a certain order to establishing preset times for certain activities.

Routines that will help the child during the course of the disease should be put in place sooner rather than later, as the change is more likely to be accepted by the child. Helpful changes may include moving the child's room to the first floor near the bathroom or establishing good eating habits early on to reduce the instance of food obsession. The same is true with incorporating safe swallowing techniques, and starting an exercise program. If a child is likely to need appliances such as a wheelchair, electric toothbrush, or a communication device or word board, it is better to introduce the items before they are absolutely necessary. By the time they are needed, the child may not be as able to learn how to use them.



## The Behavioral and Psychiatric Disorders

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Damage to the brain caused by HD can cause changes in behavior. The child with HD may become more impulsive or aggressive. There may be sudden mood swings. Sometimes it is difficult to separate symptoms that are caused by the physical changes to the brain and symptoms that are caused by the psychological stress of the disease.

### SYMPTOMS

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

Parents should not be afraid to ask the child or the doctor about depression, as there are many treatments available to help — and left untreated, depression can be life-threatening. Sometimes the family doctor or pediatrician will feel comfortable treating a mild depression with medications, while at other times counseling may be helpful, or the two can be used together. Severe depression requires a psychiatric specialist, and on rare occasions, even a young child may need hospitalization because he/she represents a threat to himself or others. Severely depressed individuals should be asked whether they have suicidal thoughts or plans. Asking about suicide does not plant the idea in the person's mind, but may provide a welcomed opportunity for the young person to discuss troubling thoughts.

### MEDICATIONS

Medications commonly used to treat depression are shown in the box on page 18. The selective serotonin reuptake inhibitors (SSRIs) are the most commonly used in recent years, because of their favorable side effect profile. No antidepressant is specifically preferred over others in people with HD. Some are more sedating, while others are somewhat more energizing. If depression is severe and accompanied by agitation, delusions, or hallucinations, then antipsychotic medications such as haloperidol, quetiapine, or olanzapine are often added. The initial dose of all these medications depends on the age and size of the child.

Several antidepressants include a “black box” warning of the possibility of suicide early in the course of treatment. If a drug causes a depressed child to become more energized before the depression is relieved, he/she may be more likely to act on an impulse. All children with depression should be watched carefully by their parents and followed carefully by their doctors as they begin treatment.

### Commonly used antidepressants

- Tricyclic antidepressants
  - Amitriptyline
  - Nortriptyline
  - Imipramine
  - Clomipramine
- Selective serotonin reuptake inhibitors
  - Fluoxetine
  - Sertraline
  - Citalopram
  - Escitalopram
  - Paroxetine
- Serotonin-norepinephrine reuptake inhibitors
  - Venlafaxine
  - Bupropion
- Others
  - Trazodone
  - Mirtazapine

An occasional child will have what appears to be the opposite of a depressed mood. When an excessively elevated mood alternates with a depressed mood, the term “bipolar disorder” may be used. In times of an elevated mood, the child may require very little sleep, may be buzzing with activity (although often getting little accomplished), and may be grandiose in ideas or 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. Excessively elevated mood can be treated with mood-stabilizing drugs such as valproic acid or lamotrigine (carbamazepine, neurontin or topamax may also be considered), often in conjunction with an antipsychotic medication such as haloperidol or olanzapine. Lithium has also been used for years to treat mania and bipolar disorder, and remains an option.

## Behavior Management

Behavior management represents the greatest challenge for Juvenile HD families. As children become teenagers, their rapidly growing bodies are confronted by the normal drives for sexuality, aggression, and independence, while at the same time HD is removing their ability to control these urges.

### AGGRESSIVENESS, IMPULSIVENESS, EXPLOSIVE OR VIOLENT BEHAVIOR

Aggressive, explosive, or violent behaviors require a multifaceted treatment approach. The first concern should be the safety of the individual or individuals who are at risk for injury when the behavior occurs. Family members should know how to access emergency services when an explosive behavior occurs, and should use this plan in a time of crisis. Many families create a home management strategy for behaviors that are slightly less threatening, including the as-needed use of medication, a safe room where the child or others go for a specified period of time when a crisis is declared, or a friend or neighbor who is called upon to remove one or another party from the scene of the crisis. It is important to lock up or remove all medications, guns, knives, automotive or cleaning products, or other

dangerous substances if there is a potentially violent person in the house. Suicide attempts and other explosive behaviors often occur impulsively, so waiting until an event occurs to lock up or remove dangerous items may prove to be too late.

The next step is to identify and address any factors or situations that tend to trigger unwanted behaviors. If a behavior only occurs at mealtime or bedtime, only in the presence of a certain care provider, or because of an obsession with soda, cigarettes, or a certain food, then it may be possible to achieve a compromise. Reducing the frequency of baths, addressing food preferences or fears of choking, changing care providers, and using desired items as rewards for good behavior, are all strategies that have been successful for some families dealing with behavior problems.

Children in the middle or late stages of HD often lack a sense of time — telling the child that something will be done in ten minutes may not mean much to the child. Setting a timer, and indicating that when the alarm rings the activity will occur, may be helpful for both the caregiver and the child. Similarly, memory can be so poor that the child doesn't recall that he had a soda or piece of candy fifteen minutes earlier. Experienced parents advise caregivers to “pick your fights.” If a situation begins to escalate toward a crisis, it may be wise to back off and let it go — give both the child and the caregiver a break.

Some families may not be able to identify specific situations that trigger violent or aggressive behavior, or may not be able to control the behaviors using the techniques described above. For their own safety, and that of those around them, aggressive, impulsive, or violent children may need medications to help them control their behavior. Commonly used medications in this situation include mood stabilizers such as valproic acid and lamotrigine, antipsychotic agents such as haloperidol, olanzapine, or risperidone, sedatives such as lorazepam or clonazepam, or beta-adrenergic blockers such as propranolol. Each of these medications has potential side effects, and none of them is effective in all children with behavior problems. Families should not rely on medications alone to control behavior; a family counselor, child psychologist, or psychiatrist can help both the family and the child to understand and manage behavior problems better.

Some children may have attention deficit disorder or hyperactivity in addition to HD. For these children, treating these symptoms may lead to an improvement in behavior. Similarly, treating an underlying depression can lead to marked improvement in angry, aggressive, or dangerous behavior.

If a home situation is dangerous to the child or others in the family, a psychiatric hospitalization may be necessary. Removing the child temporarily allows both the child and the family to rest, reflect, and heal. The child can

begin treatment in a safe environment, and the family can learn different ways to manage the situation when the child returns home. When behavior is chronically or unpredictably dangerous, particularly if other children are at risk for harm, it may be best to remove the child from the home on a more permanent basis. Often, if behavior problems are this severe, the child is an adolescent or young adult, for whom placement outside the home was already a consideration. For reasons that probably have more to do with how children treat their parents than with HD, some children or young adults show dramatic improvement in their behavior as soon as they are no longer home with their parents!

*Obsessive thinking is very common in Juvenile HD, and it may interfere with daily routines. This behavior is caused by changes in the brain that occur with progression of the disease.*

### OBSESSION

Obsessive thinking is very common in Juvenile HD, and it may interfere with daily routines. This behavior is caused by changes in the brain that occur with progression of the disease. When the thoughts of the affected child 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 SSRIs or clomipramine can be used to suppress obsessive thoughts. Behavioral modification strategies can also be used to limit the bad behavior that can accompany obsession, or to restrict the activity to appropriate times or locations. By identifying what the obsession is about and why and when it occurs, it may be possible to reassure or redirect the child, or to find more appropriate ways to satisfy the urge. A psychologist or psychiatrist is best equipped to help the family manage obsessive behavior.

### SEXUALITY

Adolescence is a difficult time even for children who do not have HD. Managing a changing physical appearance, new and unfamiliar sexual urges, learning how to interact with peers who are undergoing similar changes, and moving away from relationships with parents into strong relationships with other adolescents and adults are tall tasks for any teenager. Facing these challenges with a disease that diminishes the ability to communicate and to understand new information, and reduces the ability to suppress impulsive or disruptive behavior, 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 trusted female relative or nurse may help the girl to understand the feelings that accompany menstruation, 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 activity includes, how to prevent pregnancy, when sexual activity is inappropriate, and how to obtain help if problems

arise. Group discussions in a school health class may proceed too quickly for a girl with HD to process all of the important information or to ask questions. A public school may be obligated to provide a personal care attendant if a girl is judged to be particularly vulnerable to the sexual or physical advances of others.

Contraceptive devices or medications should be made available to girls with HD. Contraceptive patches or long-acting injections (e.g. Depo-Provera) may be preferable to pills or devices that must be used daily or at the time of a sexual encounter. Depending on the social or clinical situation, some might consider a sterilization procedure. Early attention to these issues is important, as adolescents with HD may become sexually promiscuous without a real understanding of the potential consequences of their sexual activity.

Boys with HD are also potentially vulnerable to sexual and physical abuse and to the consequences of their own impulsive or aggressive behavior. Boys who are teased or physically abused or threatened should be offered the same protection that vulnerable girls would be offered.

Boys who behave inappropriately may need both behavioral modification strategies and medications to manage their sexual urges and impulsive behaviors. For example, a boy who masturbates in public can be encouraged to use private areas such as the bedroom or bathroom, with the door closed, but may need medication if the inappropriate behavior continues or interferes with other daily activities.

Impulsive or aggressive sexual behaviors can be severe in adolescent boys with HD. A number of medications, including risperidone, olanzapine, or haloperidol, valproic acid, carbamazepine, or lamotrigine, diazepam or lorazepam, or beta-adrenergic blockers such as propranolol, have all been used to curb aggressive behavior. Consultation with a psychiatrist or psychologist experienced in the management of sexual or conduct disorders may be helpful, and inpatient treatment may be appropriate in severe cases. Anti-testosterone drugs can be considered for those with severe and recurrent aggressive sexual behavior, but would never be considered a first choice.

## **HALLUCINATIONS**

Hallucinations are uncommon in Juvenile HD, but do occur in occasional individuals. They can be auditory (such as hearing voices, which may simply make comments, or may command the person to do something), visual, or sensory. They can occur in a person with severe depression, as a result of certain prescription medications, or because of the use of mind-altering drugs (such as stimulants or hallucinogens). In the later stages of HD, a

change in mental function called delirium can occur. Delirium is an acute and reversible change in mental function, which can include severe confusion, combativeness, and hallucinations. Delirium can be brought on by an acute medical illness, by changes in medications, or in response to other stressors.

The mainstay of treatment for hallucinations and for delirium are the antipsychotic (also called “neuroleptic”) drugs. Older antipsychotic drugs include haloperidol, thiorazine, and fluphenazine, among many others. A newer group of “selective” antipsychotics, designed to have fewer side effects, includes risperidone, olanzapine, clozapine, and quetiapine, among others. Which antipsychotic is used and in what dose depends on the previous experiences of the patient or physician, as well as other concerns such as potential side effects and cost. Only a few of the older antipsychotic agents 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 Juvenile HD, but are uncommon in those 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.

The physician should never simply assume that a first seizure is due to HD, but should do the same evaluation that would be appropriate for any other child, to rule out metabolic or structural brain problems that can cause seizures. Blood tests should be done to rule out an infection or problem with blood sugar, sodium, or other blood chemicals. Screening the urine for toxins such as cocaine may be appropriate. All children with a first seizure should have a brain imaging test (preferably magnetic resonance imaging [MRI] rather than computerized tomography [CT], as it shows more detail), and an electroencephalogram (EEG). The electrical characteristics of the seizures sometimes help to guide treatment.

The word “epilepsy” refers to an ongoing tendency to have seizures. Children with epilepsy usually take medication to reduce their chances of having seizures. Children with HD who have seizures usually have generalized or myoclonic epilepsy, although other seizure types are possible (focal or partial complex seizures). Valproic acid and lamotrigine are considered first choices for the treatment of myoclonic epilepsy, although other anticonvulsant drugs can be used. A wide range of anticonvulsant drugs are available to treat generalized seizures or partial complex seizures; none is specifically recommended or avoided because the child has HD. Phenytoin, carbamazepine, levetiracetam, topiramate, and zonisamide are among the favored drugs; the selection of a medication should be made carefully after the evaluation is completed.

*The physician should never simply assume that a first seizure is due to HD, but should do the same evaluation that would be appropriate for any other child, to rule out metabolic or structural brain problems that can cause seizures.*

### **The family can help the doctor to manage a child's seizures well by:**

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- 1) keeping a log of the seizure activity
- 2) becoming knowledgeable about the medicine that the child is taking (the pharmacist can provide a computer printout with information about the drug)
- 3) ensuring that the child is taking the medicine exactly as directed
- 4) working with the doctor to define the goals of treatment, the medication dose and schedule, and what to do if there are side effects.

All seizure medications can have serious side effects, but proper use makes these side effects less likely to occur. Epilepsy often decreases in severity as 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 likely to begin. For the rare child whose seizures cannot be managed despite careful use of the standard drugs, referral to a regional epilepsy specialty center may be helpful.

## **Other Medical Issues**

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### **CHOKING**

Choking, or dysphagia, is an expected complication of HD. While there are no medications that prevent dysphagia or improve swallowing, a speech-language pathologist can help a parent or child to understand what causes the problems and provide some practical tips to minimize the chance of choking. A chin-tuck maneuver, for instance, can help direct the food properly into the esophagus (in contrast, extending the head backwards, as actors are often seen doing in a soda commercial, opens up the airway and makes choking more likely!). Alternating solids and liquids helps to clear the mouth of food particles that were not completely swallowed the first time. Using a straw or a covered cup helps to avoid spills and to limit the amount of liquid taken with each swallow, thereby reducing the risk of choking. Minimizing distractions when the child is eating can also be helpful.

The speech-language 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 to thin liquids, making them thicker and easier for the tongue to push to the back of the throat. Family members should learn the Heimlich maneuver, so that they know what to

*The speech-language pathologist can also suggest ways to change the texture of the diet to adapt to a particular child's dysphagia.*

do if a food particle blocks the airway. Local hospitals, the Red Cross, or emergency service providers may sponsor training courses for this and other first aid procedures.

Some children with Juvenile HD develop such severe dysphagia that they are unable to maintain their weight or nutritional needs, or develop recurrent lung infections (because of food or saliva entering the lungs instead of the stomach). When this happens, the family should consider the use of a feeding gastrostomy tube. People tend to have strong feelings about the use of a feeding tube. While some say it only prolongs the later stages of a fatal, neurodegenerative disease, and that they cannot imagine using such artificial means of providing food, others say that the child should be able to live comfortably without the discomfort of hunger or the indignity of choking, and that a simple means of 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, how they are inserted, what the risks and potential complications are, and how nutritional supplements are administered through the tube. It is helpful to discuss the possibility of a feeding tube early, before a crisis situation arises. Parents or other caregivers do not need to wait for the doctor to bring up the topic at a clinic visit, but can include it on their list of questions or concerns to discuss at any time.

## **NUTRITION**

Most people who have HD, including children, experience a significant weight loss as the disease progresses. Early attention to nutrition is an important part of the management 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 such as Ensure® or Boost® are too expensive, simple options can include using cream instead of milk on cereal, milkshakes or ice cream for snacks, adding high-carbohydrate foods such as pastas, and protein/calorie additives such as Carnation Instant Breakfast™. As chewing and swallowing difficulties require changes in the texture or types of foods, consultation with a dietitian will help to ensure that the child is still getting enough calories, protein, and vitamins.

Many adults with HD develop strong preferences, or obsessions, for non-nutritious items such as soda, candy, coffee, or cigarettes. A parent can try to control such a problem in a child before it begins by making sure that only



nutritious foods are available for snacks. Giving a special or preferred food to a child in the later stages of the disease may offer a rare source of pleasure or a special reward for a child who can no longer express his/her own desires.

Oral hygiene is particularly important for people who have HD. Eating and speaking are two of the most basic pleasures, both of which are eventually limited because of the disease. Losing teeth or developing painful abscesses because of poor oral care makes both of these simple pleasures difficult or impossible. Early and aggressive attention to oral hygiene is necessary. Some caregivers have found that brushing with mouthwash is easier and better tolerated than toothpaste; others have found an electric toothbrush to be helpful. If the child cannot cooperate for routine dental visits, it is appropriate to seek out a dentist who specializes in, or who has been willing to treat, other children with disabilities.

*Because dementia will eventually impair the child's ability to learn how to use assistive devices, such as a communication board or electronic or computerized speech device, later in the disease, it is critical to introduce such devices early, before they are absolutely necessary.*

### COMMUNICATION

Children who have difficulty chewing and swallowing are also very likely to have speech impairment, as the same lip, tongue, and throat muscles are involved in both processes. The speech-language pathologist should evaluate communication as well as swallowing, even if there are no apparent problems. Because dementia will eventually impair the child's ability to learn how to use assistive devices, such as a communication board or electronic or computerized speech device, later in the disease, it is critical to introduce such devices early, before they are absolutely necessary. A variety of assistive devices and strategies are available. Each child must be assessed individually to find the device that works best for him/her, based on age, cognitive skills, motor skills, and the situation in which communication is needed. Because the disease is progressive, a device that worked well a few years ago may not be as useful today. The family and the speech-language pathologist should re-evaluate the child's needs annually.

## Alternative Therapies and Medications

Very little is known about the role of alternative therapies in HD, and even less is known about their use in Juvenile HD. In recent years, more research funding is being directed towards scientific studies of alternative therapies than ever before. We hope that this will lead to a better understanding of which therapies work and how, for which problems and in what doses, with what potential side effects. Physicians are often uncomfortable discussing such unstudied and unproven treatments, and have great difficulty making any specific recommendations to their patients about them. Typical concerns that physicians have about herbal preparations include 1) no scientific proof of benefits, 2) uncertainty as to

*It is very important that the parent inform the physician of any herbal supplements, vitamins, or other treatments that the child is taking, as there are potential effects, side effects, and interactions with other treatments that may be prescribed.*

what dose should be used, 3) variations in biological effects from one bottle or preparation to the next, 4) the possibility of side effects or interactions with other medications, and 5) the possibility that the patient will use unproven herbal treatments instead of prescribed treatments with known benefits and risks. It is very important that the parent inform the physician of any herbal supplements, vitamins, or other treatments that the child is taking, as there are potential effects, side effects, and interactions with other treatments that may be prescribed. Alternative therapies include but are not limited to vitamins, herbal preparations, homeopathic medicines, chiropractic manipulations, acupuncture, magnet therapy, and massage.

## VITAMINS

Vitamins are required in small amounts for normal body functions. If a person has a measured deficiency of a particular vitamin, then the use of supplements of that vitamin are of straightforward benefit. And many of us take a multivitamin tablet, “just in case” our diet is deficient in some way. However, the specific benefits of any particular vitamin in treating HD remain unproven.

Vitamin C, Vitamin E, and other compounds with antioxidant properties, are often discussed and would seem to be of potential benefit in people facing a neurodegenerative disease. However, recent research has shown no benefits from Vitamin E to people with Parkinson’s disease, and concerns have been raised that daily doses of Vitamin E over 400 IU can actually increase the risk of death. Excessive amounts of Vitamin C can interfere with the body’s ability to process iron, and megadoses of Vitamin A and certain B vitamins can also be harmful. However, if the daily diet is marginal or inadequate, a supplemental daily multivitamin might be reasonable.

Coenzyme Q<sub>10</sub> is a molecule, available over-the-counter, that is a part of the energy-production system in the nerve cell. It has been of particular interest to HD researchers, as it has shown to prolong life in a mouse genetically engineered to develop an HD-like condition. The CARE-HD study demonstrated no benefit to adults with HD from a daily Coenzyme Q<sub>10</sub> dose of 300mg, and a slight effect that was not statistically significant at a dose of 600mg/day. A later study in Parkinson’s disease showed evidence of benefit at a daily dose of 1200mg/day. Follow-up studies to look at even higher doses, different brands of Coenzyme Q<sub>10</sub>, and modifications to the Coenzyme Q<sub>10</sub> molecule that might make it more effective, are all underway in 2007.

An average adult weighs about 70 kg (154 pounds), so a dose of 1200mg of Coenzyme Q<sub>10</sub> represents about 17 mg for each kg of body weight, or about 8mg for each pound of body weight. Thus, a child weighing 50 pounds would take about 400mg of Coenzyme Q<sub>10</sub> each day if the goal is to take an amount equivalent to what was helpful to adults in the Parkinson’s disease study. Because the risks, the benefits, and the doses of such a

treatment are unclear, it is up to you to decide whether the possible benefits outweigh the rather high cost of the supplement. Discuss this and other treatments that you plan to try with your doctor!

### DIETARY SUPPLEMENTS

Three dietary supplements frequently discussed by HD families are fish oil, creatine and various glycoprotein preparations. Ethyl-eicosopentanoic acid (ethyl-EPA, a major component of fish oil or omega-fatty acid supplements) is currently being studied in a large scale clinical trial in adults with HD. Creatine, like Coenzyme Q<sub>10</sub>, supports the energy-producing parts of the nerve cell (the mitochondria), and has shown benefits in the mouse model of HD. Small studies in adults with HD have shown no severe toxicity or danger from the supplement. However, once again, the “right” dose remains entirely unknown. A large study in Parkinson’s disease patients is evaluating the long-term effects of 10gm/day of creatine in that disease.

Polysaccharide and glycoprotein compounds are often marketed by nutraceutical companies who promote their use for a wide variety of ailments. While it is possible that these compounds will someday show benefit to people with HD, the lack of convincing scientific evidence supporting their use makes doctors skeptical about their benefits.

### HERBAL TREATMENTS

Many herbal preparations are available in the grocery store or drugstore, often with a long history of traditional use. Herbs exert their effect on the body, just like prescription drugs, because of the properties of the chemicals in them. Just like prescription drugs, herbal treatments can have side effects as well as benefits, and chemicals in them can interact with each other and with the chemicals in prescription drugs. Because herbal treatments are “natural” and not strictly regulated in their preparation, there can be wide variation from one brand or bottle to the next as to the amount and effect of the chemicals in them. None of these preparations have been shown to slow or stop HD. The decision to use an herbal preparation should be based on an understanding of its potential risks and benefits, interactions with other treatments, and costs.

### HOMEOPATHIC TREATMENTS

Homeopathic treatments, by definition, have not been subjected to scientific study. Most physicians believe that the dilution process used to

*Because herbal treatments are “natural” and not strictly regulated in their preparation, there can be wide variation from one brand or bottle to the next as to the amount and effect of the chemicals in them.*

prepare a homeopathic remedy removes almost all of its active ingredients, and thus have difficulty understanding how such treatments could have a biological effect in the body.

### CHIROPRACTIC TREATMENTS

Chiropractic manipulations are generally directed at abnormalities of bones, joints, and muscles, and would not be expected to have a direct effect on HD itself. Patients with joint deformities or neck, back, or limb pain due to HD, however, might have relief from discomfort through chiropractic manipulation. Anyone choosing to see a chiropractor should verify the credentials and methods used by the particular practitioner, as well as the goals and planned duration of therapy, just as he or she would work with the physician to understand the specific purpose of each prescribed drug.

### ACUPUNCTURE

The role of acupuncture in 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 other treatments are ineffective. Magnet therapy has enjoyed a recent surge in popularity, but there is no known role for this treatment in HD.

### OTHER TREATMENTS

Other available “alternative” treatments include massage, T’ai Chi, yoga, and other forms of exercise. Any kind of exercise that is safe and emphasizes maintaining flexibility and range of motion is a good thing for a child with Juvenile HD. Finding an activity that the child enjoys and is willing to continue doing can be a challenge, but may also be fun for the whole family!

Each year, there are popular reports of dietary agents or supplements that might be helpful for one or another symptom. We recommend a thoughtful and moderate approach to these fads. The more unbelievable a claim sounds, the more unbelievable it probably is, and the more expensive a proposed treatment is, the more carefully one should consider it before starting it. Discuss your child’s specific situation with the doctor before starting a new or unusual treatment!

# Daily Life

This chapter discusses how HD affects the daily life of the child – school, home life, friends and daily activities. At the end of 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 many adults) love to have fun!

## The Parent as Advocate

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It is not easy being the parent of a child with Juvenile HD. Juvenile HD is uncommon and many healthcare professionals and school administrators have no real understanding of this devastating disease. Health professionals may have some knowledge of adult-onset HD, but have no concept of the interrelated motor, cognition and emotional disabilities that can affect a child with Juvenile HD. The parent may often find themselves in the position of advocate for their child and educator of the public. HDSA has developed a book and CD-ROM, *Meeting the Challenges of JHD: Best Practices for the School Environment*, which includes presentations and other information designed to help parents educate those around them about Juvenile HD. While the main focus of the presentations is on the school environment, they contain valuable information for everyone. Please contact HDSA for a free copy of this valuable resource.

While it can be frustrating to deal with people who do not understand the complexity of Juvenile HD, healthcare and education professionals may have valuable experience with other disorders and disabilities that pose similar challenges. In the interest of the child, it is worthwhile for parents to work patiently with the physicians, therapists, social workers and teachers. This will insure that important information is shared between professionals so that the child receives the best care possible.

It is important to recognize that Juvenile HD progresses differently in every child and that there is no set “formula” to follow when caring for a child with the disease. Even for professionals who have experience with Juvenile HD, each child presents unique concerns and challenges. The main goals of care should be to make the child’s life as safe, easy, happy and fulfilling as possible. All therapies and interventions should be reviewed with this in mind.

*“At school, you feel that you are continually explaining the disease. They have questions about behavior with every incident.”*

*The goal should be to provide and maintain a safe, supportive learning environment for the child.*

Talking with others who have had a child with Juvenile HD can be very helpful. Support groups and online discussion forums can assist a caregiver in becoming familiar with the medical, educational and legal issues that affect HD families. These forums can also help caregivers know that they are not alone in their struggle and offer an understanding place to share their experiences.

## School

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As soon as a diagnosis of Juvenile HD is made, it is important for the family to contact the school and begin to develop an Individual Education Plan (IEP) for the child. This plan, which is reviewed and updated at least once each year, outlines how the school will address disabilities caused by the disease that interfere with the child’s education. Public schools in the US are required by law to provide for the education of all children through the high school level up to age 21. Because Juvenile HD is a disease of many interrelated disabling symptoms, including movement, cognitive and behavioral disabilities, the plan may include adjustments or variations in 1) academic instruction, 2) physical education, 3) meals, 4) seating, 5) transportation and 6) behavior control. The goal should be to provide and maintain a safe, supportive learning environment for the child. Many HD families have found it helpful to have the IEP updated at the end of the school year, so that changes can be put in place before the start of the next academic year. Additional reviews of the IEP may be needed during the school year if the disease progresses.

Some children benefit by staying in a class with familiar classmates. Others may need the individualized attention available in a special education program. Some children remain in school throughout their illness; others may need a residential school environment to address their physical or behavioral disabilities.

### ACADEMICS

Young people with Juvenile HD often experience changes in memory, judgment and problem solving (cognition). There may be a marked decline in schoolwork and the ability to learn new material or remember what they learned previously. Sometimes these problems start before the physical symptoms of the disease appear. These changes can cause frustration in the child, the teacher and the parents. Cognition problems can often be managed if it is understood that the disease is causing the changes in performance and that the student cannot simply “try harder.” An understanding teacher can vastly improve the student’s school experience. The book and CD-ROM, *Meeting the Challenges of JHD: Best Practices for the School Environment*, which is available from HDSA, outline proven strategies to help schools work successfully with children with Juvenile HD. It is

recommended that all school personnel who will work with a child who has Juvenile Huntington's Disease participate in a training session which utilizes this tool.

## PHYSICAL EDUCATION

Juvenile HD affects balance and walking, as well as hand-eye coordination and depth perception. It can make it difficult to run or throw a ball. Children with Juvenile HD are prone to falls. Because of this, physical education classes can be very challenging and even dangerous for children with Juvenile HD. Competitive team sports may not be appropriate for them and the child may need protective gear, such as knee pads, in order to participate in other gym activities. A wide range of alternative activities have been developed by physical education instructors for students with disabilities.

## MEALS

Meals and eating become more difficult as Juvenile HD progresses. While children with Juvenile HD often need extra calories to maintain their weight, they may have a hard time getting enough nutrition in the lunchroom. The time allotted for lunch in schools is usually not long enough for a child with Juvenile HD to eat a meal.

Children with the disease are often easily distracted and may forget to eat. They may also be embarrassed by their clumsiness with utensils. Some children with the disease have their lunch in the resource room or other quiet area. Others require the help of a personal aide at lunchtime.

Certain foods can cause choking in children with Juvenile HD. The risk of choking can be reduced by allowing plenty of time for eating, avoiding very chewy food and using strategies such as the chin-tuck maneuver (see Choking in Chapter 3). The speech language pathologist and school dietitian can work together to find a diet that is safe, nutritious and enjoyable to the child. As the disease progresses and choking becomes more of a concern, an attendant with proper training may be needed for the safety of the child.

## SEATING AND TRANSPORTATION

The main issue of seating and transportation is safety. Bus travel, whether to and from school or on a school trip, may present a challenge. As a disabled person, the child may be open to teasing or harassment from other students. Noise and activity on the bus may be overwhelming and the child may react with impulsive or aggressive behavior. As the disease progresses, it may be best for the student to be seated directly behind the bus driver and provided with a seatbelt.

*While children with Juvenile HD often need extra calories to maintain their weight, they may have a hard time getting enough nutrition in the lunchroom. The time allotted for lunch in schools is usually not long enough for a child with Juvenile HD to eat a meal.*

*"Open continuous communication with schools is essential. Don't always count on the IEP team leader to coordinate all of it"*

*"Changing schedules can be devastating to a kid with JHD."*

Seating in the classroom is less of an issue. Until Juvenile HD is fairly advanced, the child should be able to use a regular chair in school. If balance, rigidity or spasticity becomes an issue, the physical or occupational therapist can make specific recommendations to make school life easier. For example, an adaptation such as a pommel can keep the child from sliding out of the chair if trunk control is poor. Special padding or a recliner chair may also make the child more comfortable.

## SAFETY AND HYGIENE

Hygiene may become an issue for children with Juvenile HD. The disease may make them unsteady in the bathroom and buttons and zippers may be difficult to manipulate. They may get distracted or forget what to do. Adolescent girls with Juvenile HD may require assistance in managing menstrual hygiene. The child may need help using the bathroom.

Personal safety is another concern. With disabilities that are cognitive and emotional as well as physical, young people with Juvenile HD are highly vulnerable to physical or psychological abuse. The disease also makes a young person impulsive and willing to take risks, especially as they become adolescents. It can be easy for another student to bully a child with Juvenile HD into dangerous or inappropriate behavior. Some school districts, with a physician's order, are able to provide a personal care attendant to accompany the child throughout the school day.

Young people often taunt anyone who is different – out of fear or ignorance. Teaching classmates about Juvenile HD can help prevent dangerous bullying. Children need to be reassured that HD is not contagious and that it progresses over a number of years. The "best practices" book and CD-ROM mentioned at the start of this chapter contains suggestions to reduce bullying and increase understanding. For example, a team of student "mentors" can be organized to accompany the Juvenile HD student during the school day and alert adults to bullying situations. In some schools, students and their families have worked to increase public understanding by organizing "Hoop-a-thon" fundraisers for HD research.

## Residential Schools and Tutoring Options

Occasionally, a school district is unable to address the motor, cognitive and emotional disabilities caused by Juvenile HD in the public school setting. The student may be unable to keep up with academic coursework, or may be exhibiting aggressive or inappropriate behavior in the school. Under the provisions of the Americans with Disabilities Act (ADA), public schools must meet the needs of all individuals. If the local public school is unable to meet a child's needs, it must provide an alternative. One alternative that schools frequently suggest is home-based tutoring. In some cases, this can be



an acceptable solution. However, home-based tutoring may create greater social isolation for the child and prevent the family from getting needed respite from caregiving.

Residential schools represent another option, especially those which focus on meeting the educational, social and physical needs of multiply-handicapped children. For some families who have struggled with other options, residential placement has greatly enhanced the child's educational and social experience. Residential schools can be a positive option, not a "last resort." Frequently, familial relations improve when the parents are no longer the primary caregivers.

Schools and parents must work together in a positive way to identify the best alternative for the child. That is why it is important for parents and schools to have a positive working relationship. Local schools fund residential placement when it is deemed "educationally necessary." It is important that the family have advocates who support the child's residential placement, such as psychologists, physicians and case managers.

## Daily Routines

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For children, HD may strike at a time when certain skills, such as dressing, tying shoes, cutting meat, and grooming are just being learned. It is important to remember that the child is not simply being "lazy" if they forget or refuse to perform a task. With support and repeated instruction, children with Juvenile HD can retain past skills and even learn some new ones, particularly if they become part of a comfortable routine or if others around them are doing the same activities.

When confronted with an apparent inability to perform a task, such as eating or getting dressed, the caregiver must assess the situation before reacting. Is the child being given enough time to complete the task? Is he/she embarrassed by choking, drooling, slowness, or motor impairment? Do they simply dislike a certain piece of clothing? Is there an individual involved who triggers a negative response in the child? Has a change in daily routine caused the outburst? Individuals with HD often do best with set daily routines and may retain skills for a longer time if they are performed in a particular order at a certain time.

Once the trigger has been identified, anger and frustration are reduced and management options become available. Often the environment, time frame/schedule or goals can be adapted to avoid a crisis. Simple questions and limiting choices often work. For example, rather than asking, "What vegetable do you want with dinner?", a parent might say, "We are having ice cream after you finish your vegetables. Do you want peas or beans before your ice cream today?"

*With support and repeated instruction, children with Juvenile HD can retain past skills and even learn some new ones, particularly if they become part of a comfortable routine or if others around them are doing the same activities.*

*“For my teen, socialization is the most important part of school. She loves to meet her other ‘special needs’ friends for bowling or at the therapy pool.”*

Managing behavior can become increasingly difficult as the child’s speech becomes more indistinct. Inability to communicate can lead to irritability, aggressiveness and combative behavior. Working with a speech-language pathologist to devise simple communication devices and schemes can help maintain communication and reduce frustration. These devices must be introduced before they are needed, while the child can still learn new skills.

It is rarely the child’s desire to behave irritably or aggressively. It is the disease that is triggering the behavior. One family crocheted the words “Huntington’s Disease” on a pillow and whenever the child (or the parent!) became angry or frustrated, they were allowed to punch, throw or kick the pillow, thereby acknowledging that the disease was the source of the frustration, while venting their anger.

Parents often are aware of certain routines, favorite treats and other desires that please their child, bring them comfort and calm them down. As communication declines, it is important for the parent to share this knowledge with others who may be caring for the child, as it may avoid conflict and crisis.

### ACTIVITIES

As HD progresses, the child may not be able to handle a complicated schedule of multiple activities. Behavior problems may be the result of an overtired or confused child who needs a simpler schedule. There are many activities that children with HD and their families can enjoy, especially those based on familiarity, routine, comfort and enjoyment. Sunday morning church, Friday night pizza, or swimming at the YMCA may all fit into this category. A respite caregiver may also be a great help, taking the child to an activity while the rest of the family has a much needed break.

### FRIENDS

Children with HD ultimately lose their ability to maintain friendships. As HD advances, the movement disorder will limit physical activities and the cognitive differences between the child affected by Juvenile HD and other children become more striking. Reduction in impulse control and other behavioral changes also make friendships challenging.

It is worthwhile to maintain the child’s social interactions as long as possible. Some children continue to respond well to small groups, surrounded by familiar and friendly faces. Other children may thrive in a large group if it is a friendly one and is supervised appropriately.

## PETS

As the ability to make and sustain friendships with peers diminishes, some families have found a remarkable new outlet for their child 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, an emergency alarm, and a physical therapy helper, among other roles.

The type of pet depends on the child's specific needs and temperament, who else is available to care for the pet, how large the house is, and other issues. The safety of the animal must also be considered and children with a history of aggression may not be good candidates for pet therapy.

Some animals are specially trained to assist individuals with disabilities. Interested families can contact a local veterinarian, the ASPCA or a service such as Canine Companions (See Appendix III) 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 – 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. Summer camps, Special Olympics and weekend retreats for HD-affected individuals are examples of pleasurable activities to consider.

Fun means different things to different people. For some, it is swinging on a tire swing; for others it is a trip with the family to the zoo, the beach or a favorite relative's house. The parent may wish to create a scrapbook of fun activities, as a reminder of the good times that can be shared in future years when the child has limited mobility. For some children, spiritual beliefs and activities provide remarkable comfort and reassurance when daily life becomes difficult or lonely.

In the United States, the Make-A-Wish Foundation and the Starlight Foundation (see Appendix II) provide funding for children with serious, life-threatening conditions to make a special trip with their families, participate in a special event, or make a purchase. Juvenile HD qualifies as such a condition and many children with HD have enjoyed trips to Disneyland or other places with Foundation support. Families, as well as children with HD, benefit from these once-in-a-lifetime events!

*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.*

*“My teen makes some poor choices. She thinks anyone who is nice to her is her friend.”*

*“Not being able to drive was huge for my son. He felt such a sense of loss.”*

## Adolescence and JHD

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Adolescents with Juvenile HD face different challenges than younger children who develop the disease. Just as the young person is becoming physically more mature, their cognitive, emotional and motor control abilities will plateau and then decline. This will affect their ability to safely participate in many of the activities of adolescence.

### DRIVING

Driving is one of these activities. While getting a driver's license is a major milestone of adolescence and represents a big step toward independence, children who develop HD symptoms before age 15 cannot view learning to drive as a reasonable goal. Families can help themselves by preparing years in advance, reminding the affected child that he/she should not expect to drive. Older adolescents who have learned to drive before the onset of HD symptoms pose even a greater challenge. Impairments in judgment or denial of the illness may lead the young person to drive erratically or unsafely without even recognizing it. Families and physicians must reinforce the fact that the ability to drive will be lost at some point during the progression of the disease. Many larger communities have driver education programs for individuals with disabilities; written and behind the wheel examinations can be administered through these programs. If the young person cannot pass the examination, then he/she will not be licensed to drive.

Adolescents with Juvenile HD also face social challenges. Many young people with HD become lonely during the teenage years as HD robs them of their ability to initiate or maintain social relationships. Dating may be difficult for the adolescent with HD, as it requires learning a new set of behaviors and responses to other's behavior. Social contacts can be maintained through school, church or club activities. Activities involving small groups are often the most successful.

Parents and other caregivers should be alert to indications of sexual behavior. Adolescents with Juvenile HD may have reduced impulse control and are unlikely to understand fully the implications of sexual behavior. Girls with Juvenile HD may not understand the attention being paid to them by boys and some boys (and girls) with Juvenile HD become sexually aggressive. All young people with Juvenile HD should be regarded as vulnerable to promiscuity or sexual abuse, and heightened supervision may be required. Inappropriate or aggressive sexual behavior should be recognized as a behavioral health problem, which can be addressed by a psychologist or psychiatrist skilled in sexual disorders.

Behavior problems, including aggression, can escalate in adolescents with Juvenile HD. While young children may not entirely understand the severity of their illness, an adolescent may become clinically depressed over their diagnosis. The young person may also begin to exhibit dangerous risk-taking behavior. He or she may become defiant or delinquent. The cognitive and behavioral changes caused by Juvenile HD can be as devastating as any physical symptom of the disease. Parents should work closely with physicians and mental health professionals to monitor the behavior of the child and to be prepared to intervene if behavior becomes erratic or a danger to the child or others.

## Staying Safe – Mental Illness and Juvenile HD

*Caregivers may wish to put together a CARE kit (Critical Advocacy Resources for Emergencies) of materials and information in a three-ring binder or file box that can be quickly shared with treatment professionals in a crisis.*

Juvenile HD can cause serious mental health conditions and even psychosis. As the disease progresses, the young person may become confused or act aggressively, even toward family members and siblings. Verbal abuse, threats, temper tantrums and even physical violence are a possibility. Juvenile HD can cause a young person to become involved in dangerous behavior, or become unwilling to take needed medications. Early intervention and sustained treatment when symptoms of mental illness are present is essential to maintaining quality of life for the individual with Juvenile HD, their caregivers and the rest of the family.

Because the emotional outbursts of a person with Juvenile HD have the potential to escalate into a full-blown crisis, it is prudent for caregivers to prepare a plan to deal with explosive situations that may develop. Caregivers may wish to put together a CARE kit (Critical Advocacy Resources for Emergencies) of materials and information in a three-ring binder or file box that can be quickly shared with treatment professionals in a crisis. This kit might include a one-page summary of psychiatric history, a recent picture and description, a list of emergency numbers, a copy of the criteria for emergency evaluations and civil commitments in the jurisdiction where the individual with HD lives, copies of involuntary commitment forms (if that state permits it), and if possible, a signed medical release that allows a designated caregiver access to the individual's medical information. Guidelines for preparing a CARE kit are included in Appendix VI of this Handbook.

Whether or not a caregiver has prepared a CARE kit, caregivers and family members should have an "escape" plan ready to implement if a situation arises where they are not safe in the house with the individual with Juvenile HD. If law enforcement is involved, the police should be informed that the individual with Juvenile HD has a severe mental illness and a medical condition that may cause them to ignore the verbal instructions of the police.



# The Late Stages

## The Stages of HD

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Because there is currently 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, behavior problems are mild and working with the school or other local resources to find appropriate activities or, as the child becomes a young adult, an appropriate place to live may be the biggest obstacle.

Physicians often use functional scales 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 Drs. Ira Shoulson and Stanley Fahn, is commonly used to describe functional capacity in adults with HD. This 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 scale is in assessing individuals with adult-onset HD, it is difficult to apply to children. Provided here is a suggested way to modify the Shoulson Scale to apply to children. Although this scale has not been "validated" (proven to be scientifically or statistically accurate), it may be clinically useful in judging how a child's HD is progressing.

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 at home, but who is still attending some classes and performing appropriate self-cares would be classified as being in Stage 2. Stage 3 is a transitional stage. Entering this stage, the child would typically still be attending school, but performing little, if any, academic work. In this stage, children need increasing assistance with self-cares, and extra help may gradually be needed in the home. Often children in Stage 4 and 5 have reached young adulthood 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 behavior 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 behavior problems respond well to treatment. However, despite its limitations, this scale can be applied to children with HD, to help families understand where the affected person is in the course of the disease, and to help guide treatment plans in the school and at home.

**The stage of HD is determined by adding the points, as shown:**

11-13 points	Stage 1
7-10 points	Stage 2
3-6 points	Stage 3
1-2 points	Stage 4
0 points	Stage 5

**A Functional Scale for Assessing Juvenile-Onset HD**

A. School attendance

- 3—attends school, no special assistance needed
- 2—attends school, some regular classes, some special or modified classes
- 1—attends school, few or no regular classes
- 0—unable to attend school or work program

B. Academic/developmental performance

- 3—reading/writing/math skills appropriate to age
- 2—mild decrease in academic performance but still able to take a test or to write
- 1—unable to write legibly but able to communicate orally
- 0—unable to read/write/communicate orally

C. Chores

- 2—able to assist in age-appropriate manner with household chores
- 1—occasionally assists with chores
- 0—unable to participate in household chores

D. Activities of daily living

- 3—performs self-cares in an age-appropriate manner
- 2—requires some assistance for bathing, dressing, grooming, or feeding
- 1—assists others who bathe, dress, or feed him/her
- 0—unable to assist in self-cares

E. Lives

- 2—at home with only family assistance
- 1—at home/group home/foster care with assistance from non-family members
- 0—living in a long-term care facility



## Medical Problems in the Late Stages

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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 or passive movement of the limbs. Scoliosis due to HD is likely to recur or worsen despite bracing or surgical procedures because, regardless of treatment, the underlying disease (HD) will continue to progress.

In the late states 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 a 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 ideas or solutions than to have a crisis arise and never have considered the options. An older family member, a chaplain, a minister, a counselor, a social worker, or a physician or nurse can help parents or caregivers to consider and discuss these issues.

### **Issues to discuss in advance with professional caregivers might include:**

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- What adaptations can the school provide for a child with slowly declining motor and mental function?
- When is the school environment no longer appropriate for the child?
- What adaptations or devices are needed at home to ensure the child's safety?
- Under what circumstances would a feeding/gastrostomy tube be considered?
- How are falls managed, and when is a wheelchair introduced?
- What does "do not resuscitate/do not intubate" mean, and when is this an appropriate medical order?
- Are there other limitations to medical care that are appropriate for a particular child, and how can these limitations be noted?
- What sort of home services or respite care are available, and at what cost?
- When is nursing home placement appropriate?
- Should there be an autopsy or donation of tissue for medical research or care?
- Funeral service plans

In the late stages of HD, the affected individual is not usually able to communicate his/her own desires or choices regarding medical care. It is best for the family to discuss in advance any limitations in care that they would like in the end stages. Issues that are commonly discussed include feeding, hydration (fluids), use of antibiotics, hospitalization and resuscitation. The affected person's wishes can be written down in the form of "Advance Directives." It is wise for another family member to serve as a 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

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Many different kinds of help can be provided within the child's home, either by professionally-trained care providers or by family members or volunteers. The needs of each family depend upon the specific problems that the child is having, what the home environment is like, and the skills of available family members or friends. The solutions to the 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 enough 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 "custodial" tasks, such as bathing, dressing, and feeding are impossible to manage because of the child's size or behavior, or because of their own emotional, health, or work issues (or the needs 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 and the child

### **Issues that might be discussed with other family members or friends might include:**

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1. Who can provide help or supervision for the child with HD or his family?
2. Who can be called upon in case of a medical or behavioral emergency?
3. What to do when behavior problems become overwhelming?
4. Concerns or disagreements among family members about end of life care limitations, gastrostomy tube feeding, autopsy or brain/tissue donations.
5. What would happen to the child (either in the short-term or the long-term) if the parent or primary caregiver suddenly died or became seriously ill?

physically and emotionally for death, and to maintain dignity and privacy in the process. The physician can order a visiting nurse assessment. The visiting nurse can then work with the family to determine what specific needs they have and 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

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*There are several facilities that have specialized care units for adults or young adults with HD, so that placement of a young adult with Juvenile onset HD in a specialized care setting may be possible.*

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, because there are enough family members able to help, 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 give the primary caregiver a daily or frequent respite. However, most families do not have the physical, financial, or emotional resources to provide all the care that a child with HD needs while also providing for the needs of the rest of the family, particularly if the child's behavioral problems are severe.

When living at home is no longer appropriate for the child or family, placement into 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 care units for adults or young adults with HD, so that placement of a young adult with Juvenile 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 amounts 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 types of living situations vary greatly, and the parent is encouraged to speak at length with a social worker or case manager, and to visit each facility, before making any decision.

The selection of a long-term care facility 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” the particular person. For a young person with HD, the local nursing home that has never had a resident under the age of 50 may seem like a very poor match. However, there have been instances when 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. Some young adults with Juvenile HD have moved to community group homes, and have been able to remain in the group home throughout the course of their disease until death.

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 behavioral modification programs), dietary changes, and restraints that are used in daily management. Families should maintain an open discussion with care facilities, and should not be afraid to ask questions, offer recommendations, and work with the facility to provide the best quality of life possible for the child or young adult with HD.

## Hospice Care

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Hospice care is a special kind of nursing care which focuses on easing the transition between life and death for both the ill individual and the 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 the personal touches and support that seem more useful and important at that time. Hospice can be provided in the hospital, in a care facility, 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 need someone 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 the 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.

# Financial, Legal and Social Services Issues

Taking care of a child with Juvenile HD involves taking the long view and preparing for the entire continuum of the illness. There are legal and financial issues that must be addressed when the child is young and other issues that may arise for children with HD who reach the age of majority at 18.

For example, while parents may consent to medical treatment for their minor children, when a young person turns 18, he or she is considered an adult in most states and acquires the right to consent to medical treatment in their own behalf. In the case of Juvenile HD, the young person may already be severely disabled and it will be important for an adult caregiver to assume guardianship. This is a legal procedure, usually handled in the probate court, wherein the caregiver files a petition for guardianship, which is then evaluated by an independent evaluator and an attorney for the proposed ward before a hearing is held. If guardianship is granted, the caregiver will have the legal authority to make personal and financial decisions for the young person with Juvenile HD.

## Supplemental Security Income

Caring for a child with Juvenile HD can put a heavy financial strain on any family. In the US, there are various State and Federal government programs which may help pay some expenses, including Supplemental Security Income (SSI) and Medicaid. Qualifying for these programs can be a complicated task, but many families have found it worth the effort because of the benefits that become available to children who qualify.

Supplemental Security Income is a federal program of cash assistance which offers benefits to disabled individuals, who have little income and few assets. Benefits to disabled children are made under this program rather than under Social Security Disability Insurance (SSDI). A child under 18 can qualify if he or she meets Social Security's definition of disability for children, and if his or her income and resources fall within the eligibility limits. The amount of SSI payment depends on the level of income and varies from state to state because some states add to the SSI payment. The maximum Federal payment is \$623 a month.

As of January 2007, in order to qualify for SSI benefits, the child must: 1) have a physical or mental condition, or a combination of conditions, that results in “marked and severe functional limitations”; 2) have disabilities that must have lasted, or are expected to last, at least 12 months or are expected to result in death; and 3) not be working and earning more than \$900 a month in 2007 (This earnings amount changes every year. Please go to [www.ssa.gov](http://www.ssa.gov) for updated information).

When applying for benefits for the child, parents will be asked for detailed information about the child’s medical condition and how it affects his or her ability to function on a daily basis. They will also be required to give permission for the doctors, teachers, therapists and other professionals who have information about your child’s condition to release information to the state agency that will review the application. Financial information will also be required. Not only are the child’s income and resources looked at, but those of family members living in the child’s household as well. Other rules apply for children with parents currently drawing Social Security benefits. However, resources of the parents are no longer counted when the child turns 18 years old, even if he or she still lives at home.

**Assets that are not counted when determining financial eligibility for SSI benefits include:**

- The family home and the land it is on
- Life insurance policies with a face value of \$1,500 or less
- The family car (usually)
- Burial plots or burial funds up to \$1,500

## Medicaid

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The Medicaid Program provides medical benefits to certain low-income people who may have no medical insurance or inadequate medical insurance. Children who are considered disabled under SSI are automatically qualified for Medicaid. In some states, disabled children may qualify for Medicaid, even if their family income is too high to receive SSI benefits, under the category of “medically needy.”

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. Medicaid will also pay for Medicare premiums, deductibles and coinsurance for Qualified Medicare Beneficiaries (QMB) — individuals whose income is at or below 100% of the Federal poverty level

### States that have medically needy programs for the disabled include:

Arkansas  
California  
Connecticut  
Dist. of Columbia  
Florida  
Georgia  
Hawaii  
Illinois  
Iowa  
Kansas  
Kentucky  
Louisiana  
Maine  
Maryland  
Massachusetts  
Michigan  
Minnesota  
Montana  
Nebraska  
New Hampshire  
New Jersey  
New York  
North Carolina  
North Dakota  
Pennsylvania  
Rhode Island  
Tennessee  
Utah  
Vermont  
Virginia  
Washington  
West Virginia  
Wisconsin  
Puerto Rico

and whose resources are at or below twice the standard allowed under SSI. In addition, all states provide community Long Term Care services for individuals who are Medicaid eligible and qualify for institutional care. More information about Medicaid in each state is available on the website of the Centers for Medicaid and Medicare Services, [www.cms.hhs.gov/medicaid/consumer.asp](http://www.cms.hhs.gov/medicaid/consumer.asp).

It is important for parents not to assume that their child is not eligible for Medicaid because of family resources. The Children's Special Health Care Needs program, also called Crippled Children's Services or Children's Medical Services in some states, is a program that starts with medical eligibility and then considers income eligibility. Most pediatric health providers and medical social workers are familiar with the access points in each county and state.

## Other Financial and Placement Considerations

Programs exist to help families care for the child with Juvenile HD at home. Because many states do not have nursing home beds for people less than 16 years of age, with some very specific exceptions for short-term care, home-based services have been developed for children with progressive, severe and ultimately life-threatening diseases such as Juvenile HD. These can include aides who assist with the daily activities of life, occupational and physical therapists and other services. Insurance or Medicaid may cover some of these services if they are deemed medically necessary. Respite care services have also begun to emerge as a means to provide short-term care for the child with Juvenile HD in order to give a break to the regular caregivers, such as the child's mother. While home health services are widespread, there are wide variances in availability and sophistication of respite care programs that are available.

Another source of services for families, including respite, can be a hospice provider. Today, many hospices have transitional care programs that deliver comprehensive, compassionate services to patients with advanced or life-threatening illnesses and their families. Hospice services may include assistance to help the individual remain at home with quality of life and physical comfort or they may involve in-patient placement that allows family members unlimited visitation. Hospice is typically paid for through the Medicare Hospice Benefit, Medicaid Hospice Benefit, and by private insurance.

TEFRA 134(a), a provision of the Tax Equity and Fiscal Responsibility Act of 1982, allows states to extend Medicaid coverage to certain disabled children under the age of 19. TEFRA is a category of Medicaid that provides care to disabled children in their homes rather than in institutions.

Appropriate medical services must be available to provide care for the child in the home and the estimated cost of care in the home cannot exceed the estimated cost of care for the child in an institution. Children who live in institutions or who receive extended care in institutions are not eligible in the TEFRA category. To qualify for TEFRA benefits, the child must be disabled according to the Supplemental Security Income (SSI) definition of disability and meet the “medical-necessity” requirement for institutional care. With TEFRA, parental income and resources are not considered. Only the income and resources of the child are counted. Children who receive SSI but lose coverage intermittently due to fluctuating parental income may be eligible for TEFRA benefits in the months they do not receive SSI. The child's parent or guardian can apply for TEFRA at the same office where they apply for Medicaid benefits.

Some resources may be available to a child through their school. Provisions of the Individuals with Disabilities Educational Act (IDEA) require that public schools accommodate students with severe disabilities with services that will allow them to continue their education. In Chapter 4 of this book, the section on School includes information on the responsibilities of public schools in the education of a child with Juvenile HD.

Even professionals experienced in disability benefits may struggle to find entry points for obtaining services and often have to follow up with phone calls and letters to clarify eligibility and other issues. It is recommended that families embarking on obtaining services try to collaborate with professionals working within the particular system or agency. The rules can be complicated, but most professionals want to help a family with a disabled child obtain all appropriate benefits. Caregivers should expect to keep track of phone calls, correspondence and visits to agency offices. They can also expect to become frustrated with the amount of paperwork and follow up that is needed to get and retain services for their child.

Social workers at HDSA Chapters and Centers of Excellence are very knowledgeable about the resources in their individual state and region. They can assist caregivers in getting started with services and help with advocacy as needed. Please refer to the Appendix in the back of this book to find the closest HDSA Chapter or Center of Excellence.



# Caring For the Caregivers

## Parents

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The parents of a child with Juvenile 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 family is 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 is little wonder that the parents of children with HD report 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 toward their spouse and his/her family and estrange themselves from the family. Some parents, unable to manage an affected spouse and an affected 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/her, arrange for foster placement. A few lucky families have the support of 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 too only leads to trouble. A tired, 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, than to fight with the affected teenager until his aggressive or confrontational behavior leads to the 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 are or how much help is needed unless someone tells them! The Huntington's Disease Society of America provides literature, support

groups, newsletters, and annual convention, and a website ([www.hdsa.org](http://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 you are the best resource for 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

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In addition to addressing the needs of the 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 of families 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 his/her course. In other families, the rage or impulsive behavior of the affected person seems to be directed at the sibling, even to the extent of physically or sexually abusive behavior. In still others, the family becomes fractured, 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 manage the affected individual leaves insufficient 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 notice an increase in “escaping” behaviors such as cigarette, alcohol, or drug abuse, or if dangerous behaviors such as driving while intoxicated or physical or verbal aggression towards others 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 and should make a special effort to provide a safe haven where the child can retreat and talk. In situations where a school counselor, minister or clergyman, or special “big brother” or relative are not enough, a psychologist, family counselor, or social worker can be consulted. A parent who is engaging in harmful or destructive 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 a family counselor.

*Basic 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 the hospital, have surgery, feel sick, or die immediately*

In general, open communication about HD is important. That does not mean that all the 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 their sibling. Parents should do their best to answer questions honestly when they arise. Basic 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 the hospital, have surgery, feel sick, or die immediately.

Since the advent of gene testing for HD, many junior high and high schools discuss HD in health, biology, genetics, religion, and ethics classes. Thus, many children will 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 websites, such as the HDSA website at [www.hdsa.org](http://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/her 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. It is important to make sure siblings have an accurate understanding of their risk of developing HD, but it is not necessary to over-emphasize it. When a child moves into adolescence and has the potential to be sexually active, it is good to review how the HD gene is passed on from parent to child. Once an at-risk child has become an adult and is able to make his/her 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

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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 who 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 symptoms occurs. Just as a

parent cannot provide good care when he or she is exhausted, angry, or seriously depressed, an uninterested or unhappy professional caregiver may be harmful to the affected individual.

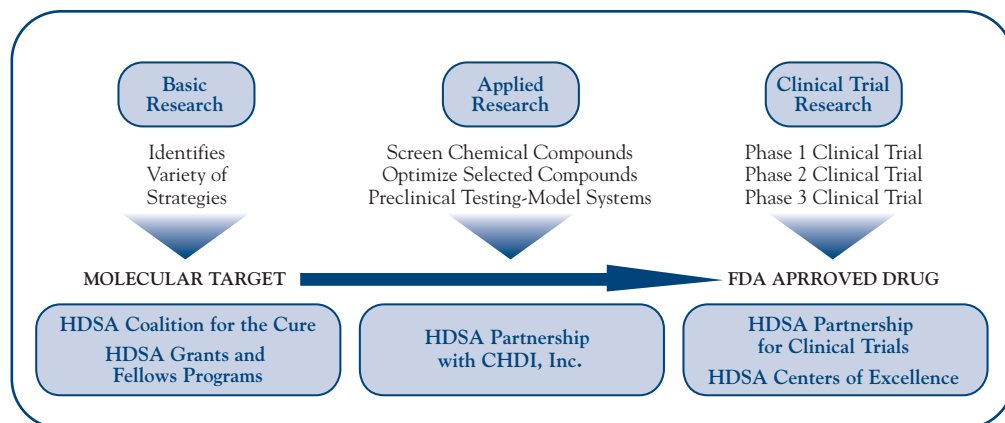
Sometimes two individuals simply do not get along. This can happen with physician-patient and physician-parent relationships as well as caregiver-child pairs. Both parties should be candid about the problems, and if the problems cannot be worked out, then other provisions for care should be arranged. 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 a disabling and destructive disease. If for any reason they cannot at least try to achieve this goal, then the child is better off with a different caregiver.

## Hospice

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Another source of services for families, including respite, can be a hospice provider. Today, many hospices have (transitional) care programs that deliver comprehensive, compassionate services to patients with advanced or life-threatening illnesses and their families. Hospice services may include assistance to help the individual remain at home until death, with a focus on comfort and relief of pain, or they may involve in-patient placement that allows family members unlimited visitation. Even after years of experience with HD, a caregiver may be unfamiliar with and need help getting through the very end stages of the disease. Chapter 5 describes hospice in detail.

# Hope for the Future



Now, more than ever before, there is reason to hope for improved treatments for Juvenile Huntington's Disease. While the discovery of the HD gene in 1993 was a momentous event, it was just the first step toward the development of treatments for the disease.

Since 1997, HDSA has supported an aggressive research program which has led to numerous advances in our understanding of HD. In mid-2004, HDSA created a Pipeline for Drug Discovery that swiftly moves innovative ideas from basic research through applied or translational research to clinical trials that may yield promising therapies.

The Pipeline begins with the HDSA Coalition for the Cure and HDSA Grants and Fellows researchers, who perform basic research that identifies targets for drug development while also continuing to work toward a cure. The most promising avenues are then sent on to HDSA's strategic partner in drug discovery and applied research, CHDI, Inc., where potential compounds and targets are screened and tested in animal models. The most promising candidates derived from this translational research are then sent onto clinical trials that are frequently conducted at HDSA Centers of Excellence.

## WHAT DO WE KNOW TODAY AS A RESULT OF THIS RESEARCH EFFORT?

- In the laboratory, researchers use fruit flies, roundworms and mouse models that have had an abnormal human HD gene (one with too many CAG repeats) or even just the fragment of the gene containing the CAG repeat segment inserted. These animals are called transgenic and are very useful in investigating different aspects of the disease and its progression. In a sense, almost all of the transgenic animal models of HD could be considered models of Juvenile HD, as researchers have used human HD genes with very large CAG repeat numbers as those seen in Juvenile HD for these experiments.
- The longer the CAG repeat number is the earlier symptoms tend to begin. However, the length of the CAG repeat number does not determine which symptoms will be worse or how long a person will live after the symptoms begin.
- CAG repeat numbers in the HD gene are the same in all of the body cells throughout a person's life. Other factors besides the CAG repeat number influence the onset age, but we do not yet know exactly what those factors are.
- Everyone has two copies of the HD gene. However only those with an expanded CAG repeat number will produce the abnormally long huntingtin protein, develop HD, and have the possibility of passing the abnormal gene onto each of their children.
- A child receiving a normal HD gene from a parent usually has the same CAG repeat number as the parent, but a child receiving an abnormal HD gene may not have the same CAG repeat number (for example, if the parent's HD gene has 17 repeats, the child's gene will also have 17 repeats, but if the parent's HD gene has 44 repeats, the child's HD gene may have 46 or 43 or some other repeat number). This is called "meiotic instability", and partly explains a clinical phenomenon called "anticipation", in which a child's HD symptoms start at a younger age than the parent's.

## WHERE ARE RESEARCHERS LOOKING FOR TREATMENTS?

- Several lines of evidence have shown that mitochondria (the cell's "energy furnace") inside the nerve cells that are dying become stressed and function poorly. Coenzyme Q<sub>10</sub> and creatine are two compounds that support the function of mitochondria; both are currently being studied by clinical researchers as possible treatments for HD.

- Adding a molecular “on-off” switch to the HD gene in a transgenic mouse model has led to reversible accumulation of huntingtin protein in the nerve cells. This is a particularly encouraging finding, since it suggests that, even after the disease has begun, it might be possible for nerve cells to “clean up” some of the damage.
- A group of enzymes called caspases are involved in the processing of huntingtin protein within the cell. Chemicals that block caspases, such as the antibiotic minocycline, are being investigated as possible treatments for HD. Strategies to remove toxic proteins from the cell, and strategies to either enhance or reduce the formation of protein aggregates, are being studied. Recent research with a mouse model has shown that inhibiting caspase 6 can prevent aggregates from forming in the mouse cells.
- As huntingtin accumulates in the degenerating nerve cell, it affects the regulation of many other genes and the production of many proteins. Histone deacetylase (HDAC) inhibitors such as phenylbutyrate may help to restore the correct balance of gene regulation and are being studied as possible treatments for HD.
- Stem cells are being explored as a possible treatment for several different neurodegenerative disorders. The simple idea of stem cell therapy is to replace the damaged or lost cells with new, healthy cells. Many scientists are working on the techniques to develop nerve cells from stem cells and to successfully and safely use them to replace lost or damaged cells.
- Trophic factors support the growth and health of existing cells. Researchers are studying several different ways to administer trophic factors to cells in the brain to support and nourish them.
- Researchers are working with RNA, the “Xerox copy” of the gene used by the cell when it wants to make a protein, with the hope of creating a molecule that selectively “sticks to” the RNA corresponding to the abnormal HD gene. This “interfering” or “silencing” RNA, by binding to the RNA copy of the abnormal HD gene, prevents the gene from being used to synthesize the huntingtin protein. In a sense, it would turn off the abnormal HD gene. Abnormal huntingtin protein would not be made and the disease caused by the abnormal protein would not occur. While this appears to work in the test tube, scientists are working hard to see whether it works safely in animals as well.
- It is possible that some drug or chemical that currently exists will unexpectedly have a favorable effect in HD. Now that researchers have created simple test tube models of the nerve cell damage that occurs in HD, they are screening thousands of existing drugs and chemicals to identify any that reduce damage to the nerve cells.

## Support for Today

While HDSA works to create a future free of HD, it also provides resources for families facing the daily challenges of living with the disease. One such resource is *The Marker* magazine, which is published twice a year by HDSA. It is designed to provide information and opinion and relay items of interest to individuals with Huntington's Disease and their families, health care professionals, interested friends and supporters. Back issues of *The Marker* can be downloaded from the HDSA website, [www.hdsa.org](http://www.hdsa.org).

The HDSA website offers a vast amount of information and news about HD, including reports from our researchers, publications and Family Guides for download or purchase, current news and events. The website can also be used to locate HDSA Chapters, genetic testing centers, medical resources, support groups, social services, and the HDSA Centers of Excellence.

HDSA Centers of Excellence serve as the cornerstone of HDSA's commitment to care. The Centers of Excellence are multidisciplinary facilities that provide a vast array of medical and social services to HD families across the US. The Centers work in collaboration with HDSA chapters, affiliates, regions and support groups to form a seamless national network of resources and referrals for those affected by HD, their families and the allied healthcare professionals who care for them. A list of HDSA Centers of Excellence as well as HDSA Chapters and Affiliates can be found in Appendix IX and X of this book. Please consult the HDSA national web site for updated information.

## Summary

While facing the daunting challenge of Juvenile HD, it is important to know you are not alone. There is a global community of HD researchers, clinical specialists, and other HD families that are ready to help offset the feeling of isolation that many HD families feel. The HDSA Chapters and Centers of Excellence offer support groups in many larger communities, and there are Internet HD support groups that are available to all. With their help and support, you can face the difficult journey that is Juvenile HD.



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# General Resources

## **American Speech-Language-Hearing Association**

10801 Rockville Pike  
Rockville, MD 20852  
(301) 897-5700  
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, speech and hearing science, and to advocate for people with communication disabilities.*

## **Hereditary Disease Foundation (HDF)**

3960 Broadway, 6th Floor  
New York, NY 10032  
(212) 928-2121  
www.hdfoundation.org

## **Social Security Administration (SSA)**

Office of Public Inquiries  
Windsor Park Building  
6401 Security Boulevard  
Baltimore, MD 21235  
www.ssa.gov/SSA\_Home.html

*This government agency oversees the Social Security, Supplemental Security Income (SSI) and Medicare programs. Social Security provides a monthly income for eligible elderly and disabled individuals. SSI supplements 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 kidney failure, 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 to any of these programs.*

## **National Organization for Rare Disorders (NORD)**

55 Kenosia Blvd.  
P. O. Box 1968  
Danbury, CT 06813  
(800) 999-NORD (voicemail)  
(203) 744-0100

www.rarediseases.org • orphan@rarediseases.org  
*NORD provides information on rare disorders and assists patients in obtaining medications they might otherwise not be able to afford. It also coordinates a patient networking service and enhances awareness of clinical trials.*

## **National Assistive Technology Technical Assistance Partnership (NATTAP)**

(703) 524-6686  
www.resna.org/taproject • resnaTA@resna.org  
*The Technical Assistance Project helps increase access to assistive technology devices and services for consumers with disabilities of all ages. Follow the links to a state-by-state listing of state programs providing access for persons in need of assistive technologies.*

## **National Mental Health Information Center**

(800) 789-2647  
http://mentalhealth.samhsa.gov/  
*The National Mental Health Information Center provides information about mental health services and direct users to Federal, State, and local organizations dedicated to treating mental illness.*

## **NICHCY – National Dissemination Center for Children with Disabilities**

(800) 695-0285  
www.nichcy.org • nichy@aed.org  
*The NICHCY serves the nation as a central source of information on disabilities in children, US education laws, and effective educational practices. The NICHCY website also features “zigawhat!,” a link for resources just for kids.*

# General Resources by State

The following resources were compiled from states with the largest HD populations.  
Please visit your state web site for similar resources.

## CALIFORNIA

### **Children's System of Care Initiative (CSOC)**

[www.dmh.cahwnet.gov/CFPP/csoc\\_initiative.asp](http://www.dmh.cahwnet.gov/CFPP/csoc_initiative.asp)

*The CSOC serves seriously emotionally disturbed children, adolescents and families with special needs in their homes, schools, and their community.*

### **Special Education – CA Dept. of Education**

[www.cde.ca.gov/sp/se/](http://www.cde.ca.gov/sp/se/)

*Information and resources to serve the unique needs of persons with disabilities so that each person will meet or exceed high standards of achievement in academic and nonacademic skills.*

### **Caregiver Resource Centers (CRCs)**

[www.dmh.cahwnet.gov/CRC/regional\\_crc.asp](http://www.dmh.cahwnet.gov/CRC/regional_crc.asp)

*11 regional Caregiver Resource Centers providing information, education and support for caregivers.*

### **United Advocates for Children of California (UACC)**

1401 El Camino Ave, Suite 340

Sacramento, CA 95815

(866) 643-1530

[www.uacc4families.org](http://www.uacc4families.org)

[information@uacc4families.org](mailto:information@uacc4families.org)

*UACC's mission is to improve the quality of life for all children and youth with mental, emotional and behavioral challenges; to eliminate discrimination and social stigma; and to promote the empowerment of families to meet the mental health needs of children.*

### **Family Caregiver Alliance Statewide Helpline**

(415) 434-3388

(800) 445-8106

*Will make statewide referrals.*

### **Genetically Handicapped Persons Program**

(800) 639-0597

[www.dhs.ca.gov/pcfh/cms/ghpp](http://www.dhs.ca.gov/pcfh/cms/ghpp)

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

## FLORIDA

### **The Advocacy Center for Persons with Disabilities, Inc.**

(850) 488-9071

[www.advocacycenter.org](http://www.advocacycenter.org)

*The Advocacy Center for Persons with Disabilities, Inc. is a nonprofit organization providing protection and advocacy services in the State of Florida.*

### **Children's Medical Services (CMS)**

(850) 245-4200

[www.cms-kids.com](http://www.cms-kids.com)

*CMS provides a continuum of medical and support services to medically and financially eligible children. Services are provided through an integrated statewide system that includes local, regional, and tertiary care facilities and providers. CMS is a program of the Florida Department of Health and is directed by the Deputy Secretary of CMS.*

### **Florida Department of Children & Families**

(850) 487-1111

[www.myflorida.com/cf\\_web/](http://www.myflorida.com/cf_web/)

*The Florida Department of Children and Families provides a variety of programs to ensure the protection of the vulnerable, to promote strong and economically self-sufficient families, and to advance personal and family recovery and resiliency.*

### **My Florida Web 211**

Dial 2-1-1

<http://flweb211.myflorida.com/>

*My Florida Web 211 is a centralized point of access to find a variety of human services offered by the State of Florida and their business partners. Calling 2-1-1 connects people with trained and supportive professionals who help identify needs, problem-solve with callers to identify solutions, and guide families through the confusing maze of public and private health and human services.*

## ILLINOIS

### **Illinois Department of Human Services – Mental Health Services**

(800) 843-6154

[www.dhs.state.il.us/mhdd/mh/](http://www.dhs.state.il.us/mhdd/mh/)

*The Division is responsible for ensuring that children, adolescents, and adults have access to publicly funded mental health services, provided by well-trained, highly professional, and compassionate people.*

## NEW YORK

### **Children with Special Health Care Needs Program (CSHCN)**

(800) 522-5006 (Growing up Healthy Hotline)

(212) 676-2950 (NYC)

*The CSHCN Program is a public health program that provides information and referral services for health and related areas for families of CSHCN.*

### **Genetic Services Program**

(518) 474-7148

*The Genetic Services Program's goal is to ensure that individuals with genetic disorders, whether affected, at-risk for transmitting, or simply concerned, have access to comprehensive genetics services which include diagnostic, counseling and preventative services.*

## TEXAS

### **Children with Special Health Care Needs (CSHCN) Services Program**

(800) 252-8023

[www.dshs.state.tx.us/cshcn/](http://www.dshs.state.tx.us/cshcn/) •

[cshcn@dhs.state.tx.us](mailto:cshcn@dhs.state.tx.us)

*The CSHCN Services Program provides services to children with extraordinary medical needs, disabilities, and chronic health conditions. The program's health care benefits include payments for medical care, family support services, and related services not covered by Medicaid, CHIP, private insurance, or other "third party payors."*

### **Mental Health Services – TX Dept. of State Health Services**

[www.dshs.state.tx.us/mhservices/](http://www.dshs.state.tx.us/mhservices/)

*Search for your local Mental Health Authority to learn more about DSHS community mental health services that are available in your area for adults and children.*

### **The Texas Information & Referral Network**

P.O. Box 13247, MC – 2077

Austin, TX 78711

Dial 2-1-1

[www.hhsc.state.tx.us/tirn/tirnhome.htm](http://www.hhsc.state.tx.us/tirn/tirnhome.htm)

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



# Huntington's Disease Organizations

## **Huntington's Disease Society of America, Inc.**

505 Eighth Avenue Suite 902

New York, NY 10018

(800) 345-HDSA

(212) 242-1968

(212) 239-3430 fax

www.hdsa.org • 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

(800) 998-7398 (Canada only)

www.hsc-ca.org • info@huntingtonsociety.ca

*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 Disease, and to improve the quality of life for people with HD and their families.*

## **International Huntington's Association**

Callunahof 8

71217 ST Harfsen, The Netherlands

+31-573-431595

+31-573-431719 fax

www.huntington-assoc.com

iha@huntington-assoc.com

*The International Huntington 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 HDSA.*

*For information about European, South American, Australian, New Zealand and Asian Huntington's Associations, please contact the International Huntington Association web site listed above.*

## Additional Organizations

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### **The Childhood Apraxia of Speech Association of North America (CASANA)**

1151 Freeport Road, # 243

Pittsburgh, PA 15238

(412) 767-6589

[www.apraxia-kids.org](http://www.apraxia-kids.org)

*CASANA is a national nonprofit organization representing the needs and interests of children and families affected by apraxia. Their website is the internet's largest, most comprehensive, and trusted site for information on childhood apraxia of speech and children's speech and language topics – including evaluation, speech therapy, research, and other childhood communication topics.*

### **National Hospice and Palliative Care Organization (NHPCO)**

1700 Diagonal Road, Suite 625

Alexandria, VA 22314

(800) 658-8898 (helpline)

(703) 837-1500 (mainline)

[www.nhpco.org](http://www.nhpco.org) • [nhpco\\_info@nhpco.org](mailto:nhpco_info@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.*

### **Make-A-Wish Foundation of America**

3550 North Central Avenue, Suite 300

Phoenix, AZ 85012

(800) 722-WISH

[www.wish.org](http://www.wish.org)

*The Make-A-Wish Foundation grants the wishes of children with life-threatening medical conditions to enrich the human experience with hope, strength and joy.*

### **Starlight Starbright**

5757 Wilshire Blvd., Suite M100

Los Angeles, CA 90036

(310) 479-1212

[www.starlight.org](http://www.starlight.org) • [info@starlight.org](mailto:info@starlight.org)

*Starlight Starbright brings together experts from pediatric health care, technology, and entertainment to create programs that educate, entertain, and inspire seriously ill children. There are both inpatient and outpatient programs including online social networks, fun centers, great escapes, and more.*



# Medical Equipment & Supplies

## Medical Equipment

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### **Aid DL products**

[www.alsa.org/resources/product.cfm](http://www.alsa.org/resources/product.cfm)

### **Broda Seating**

(800) 668-0637

### **Canine Companions for Independence**

(800) 572-BARK

[www.caninecompanions.org](http://www.caninecompanions.org) [info@caninecompanions.org](mailto:info@caninecompanions.org)

*Canine Companions for Independence is a nonprofit organization that enhances the lives of people with disabilities by providing highly trained assistance dogs.*

### **Clothing for children with special needs**

[www.special-clothes.com](http://www.special-clothes.com)

### **The Family Center on Technology & Disability**

1825 Connecticut Avenue, N.W.

Washington, DC 20009

(202) 884-8068

[www.fctd.info](http://www.fctd.info) • [fctd@aed.org](mailto:fctd@aed.org)

*The Family Center is a resource designed to support organizations and programs that work with families of children and youth with disabilities.*

### **Identi-Find**

P.O. Box 567

Canton, NC 28716

(828) 648-6768

[www.identifind.com](http://www.identifind.com) • [labels@identifind.com](mailto:labels@identifind.com)

*Iron-on clothing labels, wallet cards*

### **iMed.com**

(800) 940-iMed

[www.imed.com](http://www.imed.com)

*An online store for medical supplies at discounted prices.*

### **National Assistive Technology Technical Assistance Partnership (NATTAP)**

(703) 524-6686

[www.resna.org/taproject](http://www.resna.org/taproject) • [resnaTA@resna.org](mailto:resnaTA@resna.org)

*The Technical Assistance Project helps increase access to assistive technology devices and services for consumers with disabilities of all ages. Follow the links to a state-by-state listing of state programs providing access for persons in need of assistive technologies.*

### **New Care Therapies**

(800) 432-6249

*Vail Beds and other supplies*

### **SOS America, Inc.**

114 Hampton Blvd.

Massapequa, NY 11758

(800)999-1264

[www.sosamerica.com](http://www.sosamerica.com) • [info@sosamerica.com](mailto:info@sosamerica.com)

*Bracelets, pendants, watch and sneaker attachments.*

# Medic Alert Systems

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*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:*

## **American Medical Alarms, Inc.**

4414 SE 16th Place, Suite 4  
Cape Coral, FL 33904  
(800) 542-0438  
[www.americanmedicalalarms.com](http://www.americanmedicalalarms.com)  
[office@americanmedicalalarms.com](mailto:office@americanmedicalalarms.com)

## **LifeFone**

(888) 691-8876  
[www.lifefone.com](http://www.lifefone.com) • [info@lifefone.com](mailto:info@lifefone.com)

## **Lifeline**

(800) 797-4235  
[www.lifelinesys.com](http://www.lifelinesys.com)

## **Medic Alert Foundation**

2323 Colorado Avenue  
Turlock, CA 95382  
(888) 633-4298  
[www.medicalert.org](http://www.medicalert.org)

## **ResponseLINK**

(800) 894-1428  
[www.responselink.com](http://www.responselink.com)

# Health Care Resources

## Home Health Care

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### **National Association for Home Care (NAHC)**

228 Seventh Street, SE

Washington, DC e

(202) 547-7424

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.*

### **Visiting Nurse Association of America**

99 Summer Street, Suite 1700

Boston, MA 02110

(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, medial 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.*

### **Girling Health Care, Inc.**

(800) 447-5464

www.girling.com

*Girling is a dynamic health care company offering a full range of services in Texas, Oklahoma, New York, Florida, Illinois, and Tennessee.*

## MASSACHUSETTS

### **Boston Center for Independent Living**

60 Temple Place, Floor 5

Boston, MA 02111

(617) 338-6665

*Contact information and referrals for personal care attendants.*

### **Massachusetts Rehabilitation Commission Home Care Assistance Program**

(800) 245-6543

*A statewide organization that provides homemaker service for people under sixty who have a disability and a limited income.*

### **Visiting Nurse Associations of New England**

200 Friberg Parkway, Suite 3005

Westborough, MA 01581

(508) 475-0210

www.vnane.org

*Covers Connecticut, Massachusetts, and Rhode Island*

## NEW YORK

### **Visiting Nurse Service of New York**

(888) 867-1225

www.vnsny.org

*Providing a full range of home health care services to the New York metropolitan area.*

## TEXAS

### **The Visiting Nurse Association of Texas**

1440 West Mockingbird Lane

Dallas, TX 75247-4975

(214) 689-0000

www.vnatexas.org

*Coverage throughout Texas, call for specific area office.*

# Hospice

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## **National Hospice and Palliative Care Organization (NHPCO)**

1700 Diagonal Road, Suite 625

Alexandria, VA 22314

(800) 658-8898 (helpline)

(703) 837-1500 (mainline)

[www.nhpco.org](http://www.nhpco.org) • [nhpco\\_info@nhpco.org](mailto:nhpco_info@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.*

# Legal Resources

## **American Bar Association**

321 North Clark Street  
Chicago, IL 60610  
(800)285-2221  
www.abanet.org

*A not for profit agency that provides listings for lawyer referral agencies as well as information on the law.*

## **Judge David L. Bazelon Center for Mental Health Law**

1101 15th Street NW, Suite 1212  
Washington, DC 20005  
(202) 467-5730  
www.bazelon.org • info@bazelon.org

*The Judge David L. Bazelon Center for Mental Health Law is the nation's leading advocate for people with mental disabilities. Their mission is to protect and advance the rights of adults and children who have mental disabilities.*

## **NICHCY – National Dissemination Center for Children with Disabilities**

(800) 695-0285  
www.nichcy.org • nichy@aed.org

*The NICHCY serves the nation as a central source of information on disabilities in children, US education laws, and effective educational practices. The NICHCY website also features “zigawhat!,” a link for resources just for kids*

## **Patient Advocate Foundation**

700 Thimble Shoals Blvd., Suite 200  
Newport News, VA 23606  
(800) 532-5274  
www.patientadvocate.org  
help@patientadvocate.org

*The Patient Advocate Foundation helps patients with insurance denial, job discrimination and debt crisis. It is a nonprofit organization that does not provide financial aid but has numerous resources for patient referral. It also has a legal network to refer patients to representation.*

## **U.S. Department of Justice – American Disabilities Act Homepage**

(800) 514-0301  
www.ada.gov

*Information and technical assistance for the Americans with Disabilities Act.*

## **Wrightslaw – Special Education Law & Advocacy**

www.wrightslaw.com

*The mission of Wrightslaw is to provide parents, advocates, educators, and attorneys with accurate, up-to-date information about special education law and advocacy so they can be effective catalysts. Their website provides articles, publications, information on laws and regulations, tips and more on dozens of topics.*

## FLORIDA

### **Florida Justice Institute-ACLF Residents**

First Union Financial Center, # 2870  
200 South Biscayne  
Miami, FL 33131  
(305) 358-2081

## IOWA

### **Legal Hotline**

(800) 532-1108  
(515) 282-8161 (Outside Des Moines, IA)  
*Will help find a lawyer specializing in a certain area of law.*

### **Legal Services Corporation of Iowa**

(800) 532-1503  
*Provides pamphlets and information*

### **Lawyer Referral Service**

(513) 381-8359



# Preparing for a Crisis: Creating a CARE Kit

## CARE Kit: List of Emergency Numbers

Keep a list of emergency numbers in your CARE kit, including the following. If you have made direct contact with any key people (like the Crisis intervention team/CIT team coordinator), include their contact names and any cell phone or pager numbers.

<b>HELPLINES</b>			
Suicide prevention	(1-800-SUICIDE)	Local NAMI	
<b>RESOURCE</b>	<b>PHONE/CELL</b>	<b>RESOURCE</b>	<b>PHONE</b>
Psychiatrist		Police department	
Case manager		Local hospital	
Primary Physician		Local emergency room	
ACT/PACT team		Civil commitment Court	
Community mental health center		Mental health court	
Mobile crisis team		Homeless shelter(s)	
Crisis intervention team			
<b>Other Key Contacts:</b>		<b>Family member friends:</b>	

### INSERT THIS IN THE FRONT OF YOUR CARE-KIT

<b>Critical Advocacy Resources for Emergencies [CARE] Kit</b>															
ChecklistFor:	Last Update:														
<p><b><i>This CARE-kit contains:</i></b></p> <table border="0"> <tr> <td><input type="checkbox"/> One-page psychiatric history summary</td> <td><input type="checkbox"/> Signed Medical release</td> </tr> <tr> <td><input type="checkbox"/> Recent picture and description</td> <td><input type="checkbox"/> Medical Power of Attorney</td> </tr> <tr> <td><input type="checkbox"/> List of emergency numbers</td> <td><input type="checkbox"/> Patient's Advance directive</td> </tr> <tr> <td><input type="checkbox"/> Copy of criteria for emergency evaluation</td> <td><input type="checkbox"/> _____</td> </tr> <tr> <td><input type="checkbox"/> Copy of criteria for civil commitment</td> <td><input type="checkbox"/> _____</td> </tr> <tr> <td><input type="checkbox"/> Petition form for emergency evaluation</td> <td><input type="checkbox"/> _____</td> </tr> <tr> <td><input type="checkbox"/> Petition form for civil commitment</td> <td><input type="checkbox"/> _____</td> </tr> </table>		<input type="checkbox"/> One-page psychiatric history summary	<input type="checkbox"/> Signed Medical release	<input type="checkbox"/> Recent picture and description	<input type="checkbox"/> Medical Power of Attorney	<input type="checkbox"/> List of emergency numbers	<input type="checkbox"/> Patient's Advance directive	<input type="checkbox"/> Copy of criteria for emergency evaluation	<input type="checkbox"/> _____	<input type="checkbox"/> Copy of criteria for civil commitment	<input type="checkbox"/> _____	<input type="checkbox"/> Petition form for emergency evaluation	<input type="checkbox"/> _____	<input type="checkbox"/> Petition form for civil commitment	<input type="checkbox"/> _____
<input type="checkbox"/> One-page psychiatric history summary	<input type="checkbox"/> Signed Medical release														
<input type="checkbox"/> Recent picture and description	<input type="checkbox"/> Medical Power of Attorney														
<input type="checkbox"/> List of emergency numbers	<input type="checkbox"/> Patient's Advance directive														
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<input type="checkbox"/> Copy of criteria for civil commitment	<input type="checkbox"/> _____														
<input type="checkbox"/> Petition form for emergency evaluation	<input type="checkbox"/> _____														
<input type="checkbox"/> Petition form for civil commitment	<input type="checkbox"/> _____														

<b>PSYCHIATRIC HISTORY SUMMARY FOR:</b>		Inset current photo	
<p>_____</p> <p>[print or type patient's full name]</p> <p>Date of birth: _____ / _____ / _____</p>			
Last known street address:			
City:	County:	State:	Zip:
Home Phone:	Cell Phone:	Guardian's Phone:	
Current Age:	Height:	Weight:	Hair: Eyes:
Other [scars, birthmarks, etc.]:			
<b>MEDICAL CONDITIONS DIAGNOSED</b>			
Psychiatric Diagnosis:		Age when diagnosed:	
Other:		Age when diagnosed:	
<b>EMERGENCY CONTACT INFORMATION</b>			
Name:			
Address:			
City/State/Zip:			
Phone/Cell Phone:			
Relationship to patient:			
<b>PRIMARY INSURANCE INFORMATION</b>			
Company:	Policy:	Emergency Phone:	
<b>PHYSICIAN CONTACT INFORMATION</b>			
Psychiatrist:			Phone:
Primary Care:			Phone:
<b>PATIENT'S CURRENT SYMPTOM/SITUATION</b> <i>[check all that apply]:</i>			
Suicidal	Homeless	Violent	Vulnerable Substance abuse
Other:			
<b>MEDICATION HISTORY</b>			
Current medications:			
Medication:	Dosage:	Taken Last:	
Medication:	Dosage:	Taken Last:	
Medication:	Dosage:	Taken Last:	
Past medications that have helped:			
Past medication(s) that have not helped:			
<b>MOST RECENT HOSPITALIZATION AND/OR ARREST</b>			
Date:	Hospital:	Reason:	
<b>MOST RECENT ARREST OR INCARCERATION:</b> <i>[Violence, abuse, out-of-control, stealing, etc.]</i>			
Date:	Police Station/Jail:	Charge:	
<b>HISTORY OF SYMPTOMATIC BEHAVIOR:</b>			
<i>[failure to care for basic needs, threatening family member or others, repeated car accidents, running up huge debts, etc]</i>			



## *MATERIALS: Maintain Detailed Records*

For long term use, there are materials in addition to those in your CARE (Critical Advocacy Resources for Emergencies) kit that will be useful in dealing with caregivers and providers, whether in an outreach team, outpatient clinic, inpatient facility, or a jail or prison.

### FULLY DOCUMENTED MEDICAL HISTORY

Remember that patients have the right to request copies of their own medical records, which can be compiled and saved for future use. Any information/records that can be gathered along the way should be saved in a central file, including the names, addresses, and phone numbers of all previous or current treatment providers. Having this basic one-page summary is still important to maintain and share – the full history supports more in depth advocacy.

### INFORMAL INCIDENTS JOURNAL

Keep a journal that regularly documents the person's illness, medications and reactions to or side effects of medications, and any significant/related symptoms or problems.

- Journal entries should concentrate on observable facts, and use action words.
- They should be descriptive (not “we had a fight” but “he picked up a heavy frying pan and waved it at my head”).

Being able to provide specific dates and detailed descriptions of events is a substantial advantage for someone testifying in a treatment hearing or trying to convince a treating professional of the severity of a person's illness.

## *RESEARCH: Investigate Options*

### WHAT TREATMENT OPTIONS ARE AVAILABLE?

A person seeking the best possible care for a person suffering from a severe mental illness must first research what treatment options are available. The best and most obvious place to turn is to the professionals presently managing the person's care, but that is only the beginning of a thorough investigation.

The leaders and staff of local NAMI (the National Alliance for the Mentally Ill) affiliates will not only be familiar with service options, but may offer a better “real world” assessment of what is available than employees of mental health departments, hospitals, or private community providers. You can call the NAMI national hotline at 1-800-950-NAMI (6264) or visit their website: <http://www.nami.org>. Another basic resource is state or local mental health administrators.

### WHAT ARE THE ELIGIBILITY CRITERIA?

Most specialized services are reserved for specific populations. Those advocating for a particular service must also learn the eligibility criteria for these programs.

## DOES YOUR LOVED ONE MEET THE CRITERIA?

Don't assume the answer is "no." Ask for written policies governing eligibility. Use the person's treatment history (for example, repeat hospitalizations) to establish whether they are eligible for those services.

Never accept a first answer, and know what questions to ask. For example, does the community have intensive case managers, intensive family support services, and residential support services? Visit the local mental health center or mental health service providers and request a tour.

## WHAT ARE THE FINANCIAL INCENTIVES TO TREATMENT?

There is often a financial disincentive for providers to serve those who are most acutely ill – they may require intensive and expensive treatment. One way to counteract this is to document what it costs not to provide those services. For a person who is repeatedly hospitalized, multiply the cost per day by the total number of days. A state or county mental health administrator may appreciate that providing the needed community services or additional inpatient days to more fully stabilize the person's condition will be less expensive in the long run. Your local law enforcement may be able to provide you with the costs of repeated jailing for someone in your city.

## *ADVOCACY: Fight for the Best Cure*

Whether the person is in an inpatient facility or needs psychiatric care in the community, it can be frustratingly difficult to obtain appropriate mental health services from often unresponsive mental health care systems. However, a persistent family member or friend can secure action from these bureaucracies.

## BE AN ADVOCATE FIRST.

Building relationships with a service provider and letting them see how much you care about your loved one is vital and demonstrates your value as a member of the treatment team. But don't get too cozy. It is more important to be an advocate than to be friends with service providers and mental health officials. Feel free to disagree with politeness and persistence when cooperation is not possible. It sometimes may even be necessary to go over someone's head.

## DON'T BE STYMIED BY MEDICAL PRIVACY LAWS.

Sometimes treatment providers raise concerns about breaching confidentiality as a reason not to talk to someone trying to help another person who is receiving or in need of treatment. Sometimes confidentiality is used inappropriately.

Remember that listening is not against the law. Even if treatment providers cannot tell you about someone's condition or treatment, no privacy law prohibits them from taking information concerning a patient's condition and psychiatric history. In fact, some would argue that a mental health professional is negligent if they fail to gain as

much information about the person's condition as possible. If nothing else, you may establish a relationship that could at some point be invaluable. You can read a detailed overview of medical privacy laws and some tips on navigating them here: <http://www.psychlaws.org/JoinUs/CatalystArchive/CatalystSpringSummer2005.htm#hippa>

### **CALL THE DEPARTMENT OF MENTAL HEALTH.**

Call the county department of mental health and explain that you are having a “service delivery problem.”

- Keep a record of who you talk to at the county level.
- Try to get in writing whatever information they give you.
- Make sure you write down exactly what they say.
- Note the dates of conversations and any actions promised.

If you are still not getting a satisfactory answer, follow the same steps with the state department of mental health.

### **WORK YOUR WAY TO THE TOP.**

Jails, hospitals, and treatment facilities have established chains of command. Work your way up the chain until you get results. For example, the chain of command in an inpatient setting may start with a social worker. From there, contact a nurse, psychologist, or psychiatrist. Next, ask for a treatment team leader or section chief.

If those attempts fail, contact the hospital administrator. Every county and state has its own methods for handling grievances, so you need to find out what these procedures are.

### **MOST ORGANIZATIONS HAVE OVERSIGHT FROM A BOARD OF DIRECTORS OR TRUSTEES.**

The identity of board members is a matter of public record. Write them a short letter documenting the problems and lack of responsiveness. The board has a fiduciary responsibility to ensure that the organization's mission is met and will likely investigate and address your complaints.

For publicly funded services, you can work your way up from the Director of Mental Health Services to the very top. We have seen well-documented cases get a Governor's attention – and results.

### **KEEP TRACK OF EVERYTHING.**

- Get the name, address, phone and fax number for everyone you talk to, and their supervisor.
- Document each attempted contact or conversation that you make by email or fax (which will also politely make it known that you are creating a paper trail that can later be reviewed by supervisors or even in court).
- If you have to, send a certified letter or even a telegram to the Commissioner of Mental Health in your state highlighting your concerns, detailing your conversations with other county and state staff, and requesting an immediate response.

Once a common way to deliver urgent messages, telegrams are rarely used today because there are faster means of delivering messages. Because they are less common, they definitely get people's attention.

### HIRE AN EXPERT.

If you can afford it, an expert can be a huge help. Different situations call for different experts. For instance, care providers may react more readily to second opinions from fellow treatment professionals while hospital and community service administrators are often more responsive to contacts from attorneys. Getting help in a jail or correctional facility can be easier with the assistance of a correctional expert.

### GET HELP FROM LAW ENFORCEMENT.

If the person you are trying to help has repeated contacts with law enforcement, you may be able to get a sheriff or police chief to intervene. Help them understand that getting treatment for your loved one is in everyone's best interest - not only would appropriate treatment benefit the person, it could help avert a tragedy and remove the burden on local law enforcement created by the person's symptomatic actions. A call from a sheriff or police chief can be very influential in prioritizing services for someone you care about.

### GET THE MEDIA INTERESTED.

Television and newspaper reporters cannot cover every story they hear about. But if your situation is particularly egregious, heart wrenching, or representative of a systemic problem, your local media outlet might be interested. Find contact information on their website and focus on a reporter who is in your community, especially if they have covered mental illness or crime issues before.

When you call, summarize your story in one sentence and keep your comments focused on one main issue (such as treatment, insight, or criminalization). Picture the headline that you want to see and make that your theme. ("Man jailed for tenth time in five years because the law can't help him: Mother demands answers.")

If an article is printed that is useful to you, email, fax, and mail copies to those you are trying to influence. Don't forget to send copies - with a personal note - to your legislator and governor.

For more information on creating a CARE kit, please visit the Treatment Advocacy Center web site ([www.treatmentadvocacycenter.org](http://www.treatmentadvocacycenter.org)).

*Excerpted with permission from Catalyst Summer 2005, the newsletter of the Treatment Advocacy Center ([www.treatmentadvocacycenter.org](http://www.treatmentadvocacycenter.org)), a national non profit organization dedicated to eliminating barriers to the timely and effective treatment of severe mental illnesses.*

# Support for Parents & Children

## **Band-Aids & Blackboards**

<http://www.lehman.cuny.edu/faculty/jfleitas/bandaides>

*Helpful site to educate children about growing up with medical problems. Information for teens and parents available as well.*

## **Brave Kids**

1223 Wilshire Blvd., # 1411  
Santa Monica, CA 90403  
(800) 568-1008 (West Coast)  
(904) 280-1895 (East Coast)

[www.bravekids.org](http://www.bravekids.org) • [info@bravekids.org](mailto:info@bravekids.org)

*Brave Kids is a nonprofit organization that helps children with chronic, life-threatening illnesses or disabilities. Brave Kids provides an online community for children and their caretakers through message boards, a healthcare directory of resources, and recent and accurate medical information.*

## **Creating Solutions, Disability Solutions**

(503)443-2258

[www.disabilitysolutions.com](http://www.disabilitysolutions.com)

*Creating Solutions publishes Disability Solutions, a publication for family and others interested in Down Syndrome and Developmental Disabilities with extremely relevant information for HD families.*

## **Food Recipes**

<http://www.hdac.org/recipe/index.php>

*An online resource with recipe ideas for those who suffer from swallowing difficulties.*

## **Huntington's Outreach Project for Education, at Stanford (HOPES)**

<http://www.stanford.edu/group/hopes/>

*HOPES is a student-run project at Stanford University that offers a public web resource on Huntington's Disease. Focusing on the scientific side of HD, the site also features an online forum, short stories, and a section just for kids.*

## **Journey of the Heart: A Healing Place in Cyberspace**

[www.journeyofhearts.org](http://www.journeyofhearts.org)

*A place for resources and support for people in the grief process following a loss or significant life change.*

## **Parents' Common Sense Encyclopedia**

[www.drhull.com/EncyMaster/index.html](http://www.drhull.com/EncyMaster/index.html)

KidsHealth

[www.kidshealth.org](http://www.kidshealth.org)

*KidsHealth is the largest and most visited site on the Web providing doctor-approved information about children, with separate areas for kids, teens, and parents. There are thousands of in-depth features, articles, games, and more resources available.*

## **SchoolBehavior.com**

[www.schoolbehavior.com](http://www.schoolbehavior.com)

*The SchoolBehavior.com website provides practical knowledge and tools for educators to better learn about neurobehavioral disorders, as well as information for parents.*

## **Sibling Support Project**

6512 23rd Avenue NW, # 213

Seattle, Washington 98117

(206) 297-6368

[www.siblingsupport.org](http://www.siblingsupport.org) [donmeyer@siblingsupport.org](mailto:donmeyer@siblingsupport.org)

*The Sibling Support Project is the only national effort dedicated to the interests of over six million brothers and sisters of people with special health, mental health, and developmental needs.*

## **VerySpecialCamps.com**

[www.verypecialcamps.com](http://www.verypecialcamps.com)

*An online resource for individuals with special needs to obtain information about special needs camps*

## Online Support Groups/Bulletin Boards

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### **Brave Kids**

[www.bravekids.org](http://www.bravekids.org)

*Brave Kids provides an online community for children and their caretakers through message boards, a healthcare directory of resource, and recent and accurate medical information.*

### **Huntington's Disease Advocacy Center**

[www.hdac.org](http://www.hdac.org)

*The HDAC website provides a wealth of resources for HD families including message boards for support.*

### **Huntington's Outreach Project for Education, at Stanford (HOPES)**

<http://www.stanford.edu/group/hopes/>

*The HOPES site features an online forum, short stories, and a section just for kids.*

### **Online Discussion Groups**

[health.groups.yahoo.com/group/HDSAYouth](http://health.groups.yahoo.com/group/HDSAYouth)

[health.groups.yahoo.com/group/HDCaregivers](http://health.groups.yahoo.com/group/HDCaregivers)

[health.groups.yahoo.com/group/JHDCargivers](http://health.groups.yahoo.com/group/JHDCargivers)

### **Our-Kids**

[www.our-kids.org](http://www.our-kids.org)

*An online resource for special needs kids and their caregivers.*

### **Special needs children bulletin board**

<http://messageboards.ivillage.com/iv-psdisability>

### **Starbright World**

[www.startbrightworld.org](http://www.startbrightworld.org)

*A site for chronically ill teens to come together, from the Starlight Starbright foundation.*

# Research

## **DNA Bank**

Indiana University Medical Center  
Indianapolis, IN 46202  
(317) 274-1056

*The DNA Bank was established for the purpose of storing genetic material for possible future use. Cost to store a DNA sample is \$70.00*

## **Harvard Brain Tissue Resource Center (HBTRC)**

McLean Hospital  
115 Mill Street  
Belmont, MA 02478

(800) BRAIN-BANK  
[www.brainbank.mclean.org](http://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 diseases, including Huntington's Disease. It is essential that the donation procedure occur within 24 hours from the time of death of the donor.*

## **HD Roster**

Indiana University Medical Center  
HS 4000, 410 West 10th Street  
Indianapolis, IN 46202  
(866) 818-0213  
[hdroster.iu.edu](http://hdroster.iu.edu)

*The HD Roster is a 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.*

## **Huntington Study Group (HSG)**

1351 Mt. Hope Avenue, Suite 223  
Rochester, NY 14620  
(716) 275-9138  
(800) 487-7671

[www.huntington-study-group.org](http://www.huntington-study-group.org)

*The Huntington Project brings together under one expansive umbrella the clinical research efforts for Huntington's Disease that are underway throughout the world. The entire HD community, regardless of geographical location, is able to take part in the Huntington Project.*

## **National Neurological Research Bank**

Los Angeles, CA  
(310) 268-3536

## **NIH Clinical Trials Database**

[www.clinicaltrials.gov](http://www.clinicaltrials.gov)

*The U.S. National Institutes of Health ([www.nich.gov](http://www.nich.gov)), through its National Library of Medicine ([www.nlm.nih.gov](http://www.nlm.nih.gov)), has developed [www.ClinicalTrials.gov](http://www.ClinicalTrials.gov) to provide patients, family members and members of the public current information about clinical research studies. Check often for regular updates.*





# HDSA Chapters

HDSA's 29 volunteer-based chapters and 10 affiliates represent HDSA at the local level. They bring a voice and a face to the Society's mission, delivering services and support to the HD community in their area while working to increase awareness about HD and raise vital funds to support HDSA research initiatives. HDSA chapters and affiliates are the local link between the Society and HD families. HDSA chapters are constantly growing, so please contact HDSA by calling (800) 345-HDSA or visit [www.hdsa.org](http://www.hdsa.org) for updates.

## CALIFORNIA

### **Greater Los Angeles Chapter**

9903 Santa Monica Blvd., Suite 106  
Beverly Hills, CA 90212  
(888) 686-9868 • (888) 4-HDSA-LA (helpline)

### **Northern California Chapter**

2860 Gateway Oaks Drive, #300  
Sacramento, CA 95833  
(916) 927-4400 • (888) 828-7344 (helpline)

### **San Diego Chapter**

P.O. Box 19524  
San Diego, CA 92159  
(800) 473-4014

## COLORADO

### **Rocky Mountain Chapter**

6545 West 44th Avenue, Unit 1  
Wheat Ridge, CO 80033  
(303) 321-5503 • (877) 740-HDSA (toll free)

## DELAWARE

### **Delaware Chapter**

P.O. Box 7235  
Wilmington, DE 19803  
(410) 467-5388

## FLORIDA

### **South Florida Chapter**

12555 Biscayne Blvd.  
North Miami, FL 33181  
(305) 274-7411

## GEORGIA

### **Georgia Chapter**

P.O. Box 15007  
Atlanta, GA 30333  
(770) 729-9207

## ILLINOIS

### **Illinois Chapter**

P.O. Box 8383  
Rolling Meadows, IL 60008  
(630) 443-9876

## INDIANA

### **Indiana Chapter**

P.O. Box 2101  
Indianapolis, IN 46206  
(317) 271-0624 • (866) 488-0008 (toll free)

## IOWA

### **Iowa Chapter**

304 Market Street, Suite 2  
Audubon, IA 50025  
(712) 542-4976 • (866) 248-IAHD (toll free)

## KENTUCKY

### **Kentucky Chapter**

c/o Kosair Charities  
982 Eastern Parkway  
Louisville, KY 40217  
(502) 637-HDSA • (800) 784-3721 (helpline)

## HDSA Chapters (cont'd)

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### MARYLAND

#### **Maryland Chapter**

c/o HDSA Mid-Atlantic Region  
525 Plymouth Road, Suite 314  
Plymouth Meeting, PA 19462  
(410) 467-5388 • (877) 384-3721 (toll free)

### MASSACHUSETTS

#### **Massachusetts Chapter**

1253 Worchester Road, Suite 202  
Framingham, MA 01701  
(508) 872-8102 • (888) 872-8102 (toll free)

### MICHIGAN

#### **Michigan Chapter**

1174 James Savage Road  
Midland, MI 48640  
(989) 832-4170 • (800) 909-0073 (toll free)

### MINNESOTA

#### **Minnesota Chapter**

22 27th Avenue, Suite 212  
Minneapolis, MN 55414  
(612) 371-0904

### MISSOURI

#### **St. Louis Chapter**

8039 Watson Road, Suite 132  
St. Louis, MO 63119  
(314) 961-HDSA • (866) 707-HSDA (helpline)

### NEW JERSEY

#### **New Jersey Chapter**

114B South Main Street, Box 67A  
Cranbury, NJ 08512  
(609) 448-3500

### NEW YORK

#### **Upstate New York Chapter**

P.O. Box 25237  
Rochester, NY 14625  
(585) 341-7400

### NORTH CAROLINA

#### **North Carolina Chapter**

P.O. Box 240353  
Charlotte, NC 28224  
(919) 669-2560

### OHIO

#### **Central Ohio Chapter**

490 City Park, Suite C  
Columbus OH 43215  
(614) 460-8800 • (866) 877-HDSA

#### **Northeast Ohio Chapter**

1567 18th Street  
Cuyahoga Falls, OH 44233  
(440) 423-HDSA

#### **Ohio Valley Chapter**

3537 Epley Lane  
Cincinnati, OH 45247  
(513) 741-HDSA

### OKLAHOMA

#### **Oklahoma Chapter**

1313 Val Genes Road  
Edmond, OK 73003  
(405) 236-HDSA

### PENNSYLVANIA

#### **Delaware Valley Chapter**

525 Plymouth Road, Suite 314  
Plymouth Meeting, PA 19462  
(610) 260-0420 • (877) 384-3721 (toll free)

#### **Western Pennsylvania Chapter**

P.O. Box 110223  
Pittsburgh, PA 15232  
(412) 833-8180 • (888) 779-HDSA (toll free)

### NORTH & SOUTH DAKOTA

#### **Sioux Valley Chapter**

P.O. Box 1311  
Sioux Falls, SD 57101  
(605) 334-9917

## HDSA Chapters (cont'd)

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### WASHINGTON (ALASKA, IDAHO, OREGON, WASHINGTON)

#### **Northwest Chapter**

P.O. Box 33345  
Seattle, WA 98133  
(206) 464-9598 • (888) 264-HDSA (helpline)

### WASHINGTON, D.C.

#### **Washington Metro Chapter**

8303 Arlington Blvd., Suite 210  
Fairfax, VA 22031  
(703) 204-4634

### WISCONSIN

#### **Wisconsin Chapter**

2041 N. 107th Street  
Wauwatosa, WI 53226  
(414) 257-9499 • (877) 330-2699 (toll free)

## Affiliate Chapters

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#### **Alabama Affiliate**

(205) 325-3877

#### **Arizona Affiliate**

(888) 267-3411 (toll free)

#### **Central Florida Affiliate**

(407) 719-2291

#### **Connecticut Affiliate**

(860) 267-5980 (toll free)

#### **Long Island Affiliate**

(631) 786-8529

#### **Maine Affiliate**

(207) 286-3315

#### **Orange County Affiliate**

(949) 824-3061

#### **South/Southwest Region**

(Arkansas, Kansas, Texas Affiliates)  
(866) 712-0523 (toll free)



# 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 U.S. These multidisciplinary medical facilities bring together allied healthcare professionals, experienced in Huntington's Disease or movement disorders, to provide services and support to HD families. Centers of Excellence work in tandem with HSDA chapters, affiliates and support groups to form a national network of referrals and support. Center information can change periodically, so please contact HDSA for updates.

## ALABAMA

### **HDSA Center of Excellence at the University of Alabama at Birmingham**

Birmingham, AL 35233  
(205) 996-7865

## CALIFORNIA

### **HDSA Center of Excellence at the University of California Davis Medical Center**

Sacramento, CA 95817  
(916) 734-6278

### **HDSA Center of Excellence at the University of California, Los Angeles**

Los Angeles, CA 90095  
(310) 794-1225

### **HDSA Center of Excellence at the University of California, San Diego**

San Diego, CA 92103  
(858) 622-5854

## COLORADO

### **HDSA Center of Excellence at the Colorado Neurological Institute Movement Disorders Center**

Englewood, CO 80113  
(303) 768-4600

## FLORIDA

### **HDSA Center of Excellence at the University of South Florida Health Sciences Center**

Tampa, FL 33612  
(813) 974-6022

## GEORGIA

### **HDSA Center of Excellence at the Emory School of Medicine Wesley Woods Health Center**

Atlanta, GA 30329  
(404) 728-6364

## ILLINOIS

### **HDSA Center of Excellence at the Rush University Medical Center**

Chicago, IL 60612  
(312) 563-2900

## INDIANA

### **HDSA Center of Excellence at the Indiana University School of Medicine**

Indianapolis, IN 36202  
(866) 488-0008

## IOWA

### **HDSA Center of Excellence at the University of Iowa Hospitals and Clinics**

Iowa City, IA 52242  
(319) 353-4307

## MARYLAND

### **HDSA Center of Excellence at Johns Hopkins University Johns Hopkins Hospital**

Baltimore, MD 21205  
(410) 955-2398

## HDSA Centers of Excellence (cont'd)

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### MASSACHUSETTS

#### **New England HDSA Center of Excellence**

Charleston, MA 02129  
(617) 726-5532

### MINNESOTA

#### **HDSA Center of Excellence at the Hennepin County Medical Center**

Minneapolis, MN 55415  
(612) 873-2595

### MISSOURI

#### **HDSA Center of Excellence at the Washington University School of Medicine**

St. Louis, MO 63110  
(314) 362-3471

### NEW YORK

#### **HDSA Center of Excellence at Columbia Health Sciences/ NYS Psychiatric Institute**

New York, NY 10033  
(212) 305-9172

#### **HDSA Center of Excellence at the University of Rochester**

1351 Mount Hope Avenue, Suite 220  
Rochester, NY 14620  
(585) 273-4147

#### **George C. Powell HDSA Center of Excellence at North Shore University Hospital**

Great Neck, NY 11021  
(516) 570-4477

### OHIO

#### **HDSA Center of Excellence at Ohio State University**

Columbus, OH 43210  
(614) 688-8672

### TEXAS

#### **HDSA Center of Excellence at Baylor College of Medicine**

6550 Fannin, SM 18011  
Houston, TX 77030  
(713) 798-3951

### VIRGINIA

#### **HDSA Center of Excellence at the University of Virginia**

Charlottesville, VA 22903  
(434) 924-2665

### WASHINGTON

#### **HDSA Center of Excellence at the University of Washington**

Seattle, WA 98195  
(206) 598-4030

# HDSA Publications

The Huntington's Disease Society of America has an extensive library of informative books, magazines, and pamphlets available. To order copies of these and other publications, please contact HDSA at (800) 345-HDSA or visit [www.hdsa.org](http://www.hdsa.org).

## **The Marker**

*The Marker Magazine, an outstanding source of information published twice a year, is designed to provide information and opinion and relay items of interest to individuals with Huntington's Disease and their families, health care professionals, interested friends and supporters.*

## **Family Guide Series**

*Our informative Family Guide Series offers an overview of key areas.*

- *Coping with Speech and Swallowing Difficulties in Huntington's Disease*
- *Genetic Testing for Huntington's Disease*
- *Huntington's Disease*
- *Juvenile Huntington's Disease*
- *Long Term Care*
- *Nutrition and Huntington's Disease*
- *Physical and Occupational Therapy for Huntington's Disease*

## **Additional Publications**

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Additional publications are available for download from the HDSA website including:

- **A Physician's Guide to the Management of Huntington's Disease**
- **A Caregiver's Handbook for Advanced-Stage Huntington's Disease**
- **Understanding Behavior in Huntington's Disease**
- **Fast Facts About HD**
- **Toward a Cure**

*Disclaimer: Kindly note that the information provided within these publications is not meant as an indicator for a diagnosis or medical treatment.*

# JHD Handbook CD-Rom

Websites listed in the preceding appendices can be found on the JHD Handbook CD-Rom, including HDSA Chapter websites and HDSA Center of Excellence websites. Additional resources are also provided on the CD-Rom in Adobe Acrobat PDF format.

## HD Care

- Caring for People with HD
- Certified Nursing Assistants First Shift
- HD Palliative Care Study by The Robert Wood Johnson Foundation
- Coping With Speech and Swallowing
- Huntington's Disease and Diet
- Nutrition and Huntington's Disease

## HD Information

- Facing HD
- Fast Facts
- Genetic Testing for HD
- Huntington's Disease
- Juvenile HD

## JHD and the School Experience

The complete Juvenile Huntington's Disease and the School Experience publication is included on the JHD Handbook CD-Rom. Contents from the original School Experience CD-Rom are included in addition to newly added material.

- Questions and Answers for School Staff and Administrators
- Questions and Answers for Teachers, Students and Parents
- Meeting the Challenges of JHD: Best Practices for the School Environment
- Bullying and Teasing of Youth With Disabilities
- Effective Interventions in Dropout Prevention
- Moving Beyond Punishment to Promote Effective Interventions for Children with Mental or Emotional Disorders
- Pointing Fingers: Things That Teachers and Parents May Say That Foster Distrust
- Sexuality Education: Building a Foundation for Healthy Attitudes
- Sexuality Education: Building on the Foundation of Healthy Attitudes

*Documents included were produced by: the National Information Center for Children and Youth with Disabilities (NICHCY); Consortium for Appropriate Dispute Resolution in Special Education (CADRE); the Parent Advocacy Coalition for Education Rights (PACER); the Families and Advocates Partnership for Education (FAPE) at the PACER Center; the National Center on Secondary Education and Transition (NCSET); the National Dropout Prevention Center for Students with Disabilities (NDPC-SD); Creating Solutions, Terri Couwenhoven, MS – [www.disabilitysolutions.com](http://www.disabilitysolutions.com); the Judge David L. Bazelon Center for Mental Health Law; and Leslie E. Packer, PhD – [SchoolBehavior.com](http://SchoolBehavior.com). PACER Center materials used with permission from PACER Center Inc., Minneapolis, MN, (952) 838-9000. [www.pacer.org](http://www.pacer.org). All materials used with permission. All rights reserved.*