

Juvenile Onset HD



Huntington's Disease

Family Guide Series



Huntington's Disease
Society of America

Juvenile Onset HD

Family Guide Series

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Published with funding from a generous educational grant from The Bess Spiva Timmons Foundation, Inc.

Disclaimer:

Statements and opinions in this guide are not necessarily those of the Huntington's Disease Society of America, nor does HDSA promote, endorse, or recommend any treatment or therapy mentioned herein. The reader should consult a physician or other appropriate healthcare professional concerning any advice, treatment or therapy set forth in this book.

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I. What is Huntington's Disease (HD)?

Huntington's Disease (HD) is a hereditary brain disorder which leads to both physical and mental disabilities that increase over time. In people with the gene that causes HD, symptoms usually begin to appear between the ages of 30 and 50, but the disease may strike those as young as two or as old as 80. At present, there is no cure and no treatment known to stop, slow or reverse the progression of the disease.

Over time, HD causes the progressive loss of nerve cells in the brain affecting:

- Movement
- Cognition
- Emotions and Behavior

Fewer than ten percent of individuals with HD develop symptoms before age of 20. Juvenile onset HD (JHD) presents unique challenges to the affected individuals, their caregivers, and the various professionals who assist them. This publication describes how a physician makes a diagnosis of JHD, the most common symptoms, strategies for coping, and the current outlook for HD research.

II. Diagnosing JHD

Diagnosing juvenile onset HD is a complex task and a diagnosis may take time. This is because there is no single symptom or group of symptoms that absolutely indicate a diagnosis of juvenile onset HD. A child's symptoms may be caused by developmental delay, attention deficit disorder, mental retardation, depression, or other medical or neurological conditions entirely unrelated to JHD. No one should assume that JHD is causing a child's symptoms, even if a parent or sibling has the disease. For reasons outlined on page 3, a genetic test to confirm the presence of the HD gene mutation is typically not among the first steps in diagnosing JHD.

To begin the diagnostic process, the physician will take a medical and neurological history, a family history, a developmental history, and perform a neurological examination and neuropsychological assessment (tests of memory, developmental skills, and intelligence) to serve as a baseline for comparison later. The physician may order brain imaging or blood tests to rule out other conditions that may cause the child's symptoms. It is important for the doctor to see records of any previous neurological examinations, psychological evaluations or school testing.

Based on the child's behavior and school performance, the physician may ask physical, occupational, and speech therapists to assess the child. The pediatrician is quite likely to ask a neurologist, neuropsychologist or child psychologist to evaluate the child to assist with the diagnosis. The physician may also make referrals for individual or family counseling, school based programs, or social services.

What Might Suggest JHD

- Changes in motor, behavioral, and cognitive function
- Abnormal neurological examination
- Abnormal neuropsychological test results
- Progression of symptoms despite appropriate treatment and counseling
- Family history of HD

Changes in Motor, Behavioral, and Cognitive Function

Symptoms may include:

- Stiffness of the legs
- Clumsiness
- Loss of previously learned skills
- Difficulty learning new material
- Changes in behavior
- Seizures
- Swallowing or speech problems

The involuntary movements (chorea) typical of adult onset HD is uncommon in children who develop HD before age 10, but may be one of the first symptoms a teenager exhibits. Behavioral problems, such as the loss of impulse control or emotional outbursts may also be among the first symptoms in an adolescent.

Family History

HD is an inherited disease. For a child to have the disease, at least one parent must have the gene for the disorder. It is not contagious. You cannot "catch" HD from another person. You must be born with the abnormal gene that causes the disease in order to one day develop this disorder.

In some cases, a child may develop what appears to be JHD even though there is no known family history of the disorder. There are several possible reasons for this. Perhaps the parent who had the gene for HD died at an early age, before the symptoms appeared, or the parent's symptoms were misdiagnosed as Parkinson's disease or another disorder with symptoms similar to HD. It is also possible that the parent carried the genetic mutation for HD in what is called the "intermediate range of the CAG expansion for HD" in which case the parent did not develop the disease, but could still pass the abnormal gene on to the children. There may also be cases of nonpaternity. Finally, one of the birth parents of an adopted child might have carried the genetic abnormality that causes HD.

If the child was adopted, it may be possible to obtain the family medical history from the birth parents through the adoption agency or local social services department, once the serious nature of JHD and its hereditary pattern are communicated.

Genetic Testing

A blood test is available that can determine, in almost all cases, whether a person has the abnormality in the HD gene that leads to the development of the disease. We distinguish between the use of a gene test in children who have symptoms suggestive of JHD, and children who are neurologically normal.

Testing Children Who Do Have Symptoms

The gene test can be very helpful in confirming a suspected diagnosis of JHD, but the medical community is very cautious about using a gene test too early in the evaluation of a child, so as to avoid revealing the genetic status of a child whose symptoms turn out to have a cause other than JHD. It would not be appropriate to

do a diagnostic gene test for JHD in a child because of headaches and a sore throat, for instance. Attention deficit disorder, poor grades in school, and depression, likewise, are so common in children without JHD that they may not justify the use of a diagnostic test for HD either. Because HD is a progressive disease, the worsening of neurological or behavioral symptoms over 6 months or year despite optimal management, might represent a situation in which genetic testing is appropriate. If a child is exhibiting symptoms of juvenile onset HD and a genetic test is being considered as a confirmatory measure, the family should consult with a neurologist who is familiar with HD and a genetic counselor. HDSA Centers of Excellence have neurologists and counselors who are extremely knowledgeable about the disease.

Testing Children Who Do Not Have Symptoms

Until there are treatments to slow, halt or reverse the disease, there is no medical advantage to testing a healthy child for the mutation that causes HD, even if a sibling has been diagnosed with the disease. The genetic test cannot predict when symptoms will begin, but the knowledge can have a severe emotional affect on the child. Testing of minors may also expose them to discrimination by insurance companies, future employers, and perhaps (consciously or unconsciously) by their parents. Most at-risk adults do not choose to have predictive testing for HD. For all these reasons, testing of an asymptomatic child is strongly discouraged. A possible exception could be an adolescent in an adult situation, such as an "emancipated minor" or a married teenager considering children.

In the United States, children who are eligible for adoption are not usually given a genetic test for HD because of the potential for discrimination based on the test results. Other countries may have different practices. Juvenile onset HD is a disease that affects the entire person and the way they move, think and behave. Eventually, affected individuals require 24-hour supervision and care with the goal to maintain the child's functioning and quality of life for as long as possible.

III. After the Diagnosis

Because there is no way to predict how rapidly symptoms will worsen, every effort should be made to help the child participate in school and continue to do the things he or she enjoys. The child's multidisciplinary medical team will have suggestions on maintaining the child's quality of life for as long as possible. The child's school will also have a major role to play in their educational experience. HDSA has created a comprehensive guide for school administrators and teachers entitled "Juvenile Huntington's Disease and the School Experience."

The juvenile onset HD multidisciplinary team typically includes:

- Family doctor or pediatrician
- Psychologist or psychiatrist
- Neurologist
- Physical therapist
- Occupational therapist
- Speech-language Pathologist
- Dentist
- Social worker, genetic counselor
- Dietician

The Huntington's Disease Society of America (HDSA) has established Centers of Excellence across the country to provide the full range of services needed by families facing HD.

Understanding JHD and Teaching Others

It is important to understand issues created by the many symptoms associated with JHD. Parents of a child with this disease will come into contact with many people who have no experience with the disease. They may have to teach doctors, teachers and other individuals about JHD. Although JHD is rare, many of the daily challenges are similar to those of children who have other disorders and disabilities.

The Movement Disorder

JHD affects movement, both through changes in the brain and through changes in muscle tone. Children who show HD symptoms before

age 10 often develop stiff or rigid muscles. Some adolescents with HD develop the involuntary, fidgety movements called chorea.

Symptoms of the movement disorder in JHD include:

- Walking on the toes
- Clumsiness
- Losing balance easily
- Developing a scissoring or stiff-legged gait
- Slurred speech, drooling or difficulty swallowing
- Reduced ability to write, throw a ball, or ride a bicycle

The Cognitive Disorder

The ability to think and process information is called cognition. JHD leads to the progressive loss of nerve cells in the brain, which affects the ability to concentrate, remember information and multi-task.

Changes in cognition may create difficulties with:

- Beginning/ending activities (initiation)
- Multi-tasking, attention and concentration
- Short term memory/new learning
- Repeating the same thought or ideas (perseveration)
- Controlling impulses
- Irritability/outbursts

There are currently no medications that improve mental function, but treating the movement or behavioral disorders associated with JHD may help reduce some cognitive problems. However, many medications can also have side effects that impact on cognition; changes for the worse after starting a new medication should be reported to the physician right away.

Behavior and Psychiatric Issues

Damage to the brain caused by HD can cause changes in behavior. The child with JHD 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.

Changes in behavior may be among the earliest symptoms of the disease, especially among adolescents,

causing affected children to do things that seem completely “out of character.” Behavior changes often present the greatest management problems for children with JHD.

Some of the behavior disorders that can be caused by JHD include:

- Impulsive behavior and poor judgment
- Depression
- Anxiety and guilt
- Emotional and temper outbursts
- Obsessive thoughts, compulsive behavior
- Aggression

Managing behavior may become increasingly difficult as a child’s ability to reason and communicate declines. When things get frustrating, parents and other caregivers should remember that it is rarely the child’s desire to behave irritably or aggressively. It is the disease that causes the problems. In adolescents, aggressive behavior may also manifest with early or risky sexual behavior. This is discussed more thoroughly in the section addressing Special Issues of Adolescence and JHD starting on page 13.

Behavior Management

Behavior problems can have many causes and dealing with them may require a number of management strategies. Sometimes problems are caused by underlying medical conditions that can be addressed or by “trigger” situations that can be avoided. Therapy with a skilled counselor or child psychiatrist may help both the family and the child to understand and control the behavior problems.

The following approaches have been successfully used by many parents of children with JHD:

- Set simple goals
- Stick to a schedule
- Break tasks into small steps
- Avoid multi-part or open-ended questions
- Limit choices to those that are acceptable (i.e. “Do you want peas or carrots before your ice cream?”)
- Adopt “to-do” lists and calendars
- Be patient

Many apparent behavior problems come from frustration or confusion caused by the disease’s effect on the brain.

In some children, problematic behavior is related to attention deficit hyperactivity disorder (ADHD) or to an underlying depression that can be treated. Medications are available which may control some symptoms if used carefully; however, virtually all medications may rarely cause serious side effects. None is effective in all children with behavior problems. Information about medications used to treat behavioral and psychiatric problems in children with JHD is available through HDSA Centers of Excellence.

Swallowing, Speaking and Eating

Choking

Choking, or dysphagia, is an expected complication of HD. While there are no medications that improve swallowing, a speech-language pathologist can often provide practical tips to minimize choking. Choosing food that is not too chewy or watery and allowing the child enough time to eat can also reduce the risk of choking.

Communication

A speech-language pathologist can evaluate the child for communication problems and may suggest methods to improve the child’s ability to be understood. Communication boards or computerized devices may be prescribed as speech becomes more difficult. The child should be assessed periodically to determine appropriate strategies and devices.

Nutrition

Children with JHD may experience significant weight loss as the disease progresses. A child with JHD needs high-quality foods with extra calories and protein, along with plenty of calcium and vitamins. A parent may add weight-gain or protein/calorie supplements to the child’s diet, substitute cream for milk, offer ice cream for snacks, or serve high-carbohydrate foods such as pasta. As chewing and swallowing become more difficult, consultation with a dietitian may be helpful in order to arrange a nutritious easy-to-eat diet.

Many children and adolescents with JHD develop strong preferences, or even obsessions, for non-nutritious items such as soda or candy. Limiting snacks to nutritious foods can help manage this problem before it begins. Sometimes a prized food or snack item can be used as a reward for good behavior.

Maintaining the Activities of Daily Life

Physical therapists, occupational therapists and speech-language pathologists can offer a range of solutions to help the child continue to perform the activities of daily life. These might include:

- A program of exercises to maintain range of motion
- Special seating for the classroom
- Teaching strategies to reduce falls
- Introducing assistive devices to help with dressing, walking, and feeding
- Oral exercises and other individualized strategies for problems with speaking and swallowing
- A home safety assessment by a county public health nurse

The physician may also prescribe drugs to help reduce physical issues such as rigidity, spasticity (muscle spasms), dystonia (fixed posturing of muscles in a limb or jaw) or chorea. Prescription drugs may also be recommended for behavioral or psychiatric problems.

Assistive Devices

Assistive devices such as wheelchairs, helmets, and communication boards can improve the child's safety and quality of life as symptoms progress. However, because the disease affects the ability to learn new skills, it is necessary to gently introduce assistive devices well before they are absolutely needed, so that they are slowly incorporated into the child's routine.

Other Medical Issues

Seizures

Epileptic seizures occur in about 25% of children with JHD, but it should never be assumed that JHD is the cause of the seizures until other possible causes have been ruled out by imaging studies, blood tests, and an electroencephalogram (EEG). Seizures may begin at any time during the disease, and range from mild or infrequent episodes to frequent and severe seizures. There are a number of anti-epileptic (seizure) medications, which can reduce the chances of having seizures; all medications must be used carefully to minimize their side effects. Referral to a pediatric epilepsy specialist may be appropriate for some children.

Hallucinations

Hallucinations are uncommon in JHD, but can occur.

Hallucinations can be triggered by severe depression, by certain drugs, or in the late stages of the disease, by medical illness. They may be controlled with medication.

General Medical and Dental Care

A child's general health should not be neglected because of the need for special care related to JHD. Immunizations should be given on time, and growth and development should be monitored as they would be for any other child.

Oral hygiene may be difficult for a child with JHD. Tooth brushing requires small muscle coordination. Rigidity in the jaw muscles may make brushing difficult. An electric toothbrush may be helpful. The parent may need to seek out a dentist who treats children with disabilities.

Alternative Therapies and Supplements

Very little is known about the benefits of alternative therapies in treating HD. Alternative therapies might include vitamins, herbal preparations, homeopathic preparations, chiropractic manipulations, acupuncture and magnet therapy. The decision to use alternative therapies must be made by the family after considering what is known about the benefits, the potential risks, and the financial cost. The medical team can help the family to decide about these treatments.

The Rest of the Family

Telling the Siblings

Children are often more aware of problems than the parents realize and deserve honest answers. Children of almost any age can be told that their sibling has JHD, that JHD is not contagious, and that having the disease does not mean that the sibling is dying soon. An important part of your explanation to a child is that the affected parent did not pass on the gene deliberately.

Finding Emotional Support

JHD is truly a family disease, affecting parents, siblings and the extended family. No one, especially parents, should try to cope with the child's disease alone. Friends, family members, mental health professionals or a religious institution can be good sources of emotional support. Joining an HD support group may be helpful. There are HD support groups in virtually every state. Your local chapter of the Huntington's

Disease Society of America (HDSA) or the HDSA national office can help you locate a support group in your area. You can contact HDSA at 800-345-HDSA or visit www.hdsa.org, click on "About Huntington's Disease Society of America," then click on "Find Your Local HDSA Chapter, Affiliate or Regional Office".

Professional counseling should be encouraged for any family member who feels overwhelmed, depressed or out of control. Many families find it helpful to schedule regular visits with a family counselor. School counselors should be made aware that siblings of a child with JHD may need help with depression or stress.

IV. JHD in the School Environment

Public schools are required to provide education for children with handicaps of all types. As soon as a diagnosis of JHD is made, 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. Parents of children with JHD may request assistance or adjustments in:

- Academic classes
- Physical education
- Meals
- Seating and transportation
- Safety and hygiene
- Behavior control

Teachers, administrators and other students should be helped to understand JHD and how it affects the child's performance at school. "Juvenile Huntington's Disease and the School Experience," a PowerPoint, CD-ROM and book combination, can help explain the disease to teachers, administrators and other students. It is available from the HDSA national office at 800-345-HDSA.

Changing Abilities

As JHD progresses, the child will become less able to learn new information and may lose skills previously mastered. Behavioral problems may occur when a child is frustrated by material that is too difficult or presented too quickly. While remaining in a class with familiar classmates may be important to some children, others may prefer or need the individualized attention available in a special education program.

Children with JHD should have individual education plans (IEPs). Because JHD is a progressive disease, the educational plan must be reviewed frequently (at least once a year), to set new goals appropriate to the child's age and stage of disease.

Other Schooling Options

The Americans with Disabilities Act (ADA) requires that public schools meet the needs of all individuals. If a local school is unable to meet a child's needs, it must provide an alternative for the family. One alternative is home-based tutoring, which may or may not be appropriate. Residential schools for multiply handicapped children are another option. Local schools must fund residential placement when it is deemed "educationally necessary." For some families, residential placement greatly enhances the child's educational and social experience, while reducing the family's day to day burden.

V. Special Issues of Adolescence and JHD

Adolescents with JHD 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.

Behavioral Problems

Behavior problems, including aggression, can escalate in adolescents with JHD. 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 JHD 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.

Sexuality

Parents and other caregivers should be alert to indications of sexual behavior. Adolescents with JHD may have reduced impulse control and are unlikely to understand fully the implications of sexual behavior. Girls with JHD may not understand the attention being paid to them by boys and some boys and girls with JHD become sexually aggressive. Parents should talk with their physician about contraceptives; girls who are sexually active should have an appropriate annual evaluation by a gynecologist.

The school may be obligated to provide a personal care attendant if a boy or girl is judged to be particularly susceptible to teasing, physical threatening or sexual abuse by others in the school.

All young people with JHD 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.

VI. The Progression of Juvenile Onset HD

The Stages of JHD

A functional scale to help parents and physicians judge where a child is in the progression of JHD (based on the scale for adults devised by

Drs. Ira Shoulson and Stanley Fahn) can be found below. This scale rates motor and cognitive functions, but does not rate severe behavioral or psychiatric problems. Treatment of behavior problems may result in an increase in a child's score. Despite its limitations, this scale can help families to understand generally where the child is in the course of the disease, and may assist in the development of treatment plans for school or home.

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-care

E. Residence

- 2 - at home with only family assistance
- 1 - at home/group home/foster care with assistance from nonfamily members
- 0 - living in a skilled nursing care facility

Add the points to determine the stage of HD

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

Preparing for Late Stage JHD

Although it is difficult to think about the progression of symptoms and the late stages of the disease, it must be done. Before a crisis arises, families should discuss such issues as long term care placement, end of life care limitations, tube feeding, tissue donation and autopsy. In the late stages of JHD, the child or young adult may not be able to participate in discussions regarding medical care or other wishes. Talking about these issues early on allows the child to make his/her wishes known and to have them recorded in the form of "Advance Directives." When an affected teen reaches the legal age of maturity, it is wise to designate a family member as a legal guardian who can make financial and medical care decisions on his or her behalf. These arrangements are usually made through the probate court of the county of residence.

Home Care and Nursing Homes

Parents of a child with JHD should plan to ask for outside assistance in the late stages of the disease. Some families find enough volunteers to help care for the affected child and never require professional help. Other parents may hire someone to help with household chores. Still other parents need professional home health aides or certified nursing assistants for "custodial" tasks such as bathing, dressing, and feeding. A visiting nurse can work with the family to determine which services the aides or assistants will supply. Home health services are often administered by a hospital or county government agency.

Individuals with late-stage JHD require 24-hour nursing care. Placement in a skilled nursing care facility may become necessary. While there are no facilities in the United States that specialize in the care of children with JHD, there are several facilities that have specialized care units for adults or young adults with HD.

For young adults, who are not yet in the late stages of the disease, a number of different living situations are possible with varying amounts of care or supervision. These include adult foster homes, group homes, board and care facilities, assisted living facilities, and nursing homes.

Hospice Care

Hospice care is a special kind of clinical care, which focuses on easing the transition between life and death, for both the ill individual and the family. Hospice care is provided by nurses, social workers and pastoral counselors who have special training in the unique concerns that arise at the end of life. Hospice can be provided in the hospital, in the nursing home, or at home. Hospice nurses can answer questions about medical issues such as pain, nutrition, or infections and help the family prepare for the tasks that must be taken care of at the time of death.

VII. Legal, Financial, and Social Service Issues

Medical Consent

Parents are generally the ones who make medical decisions for their children until they reach the age where they are legally responsible for themselves (generally eighteen). In the case of a young adult with JHD, a parent or grandparent may need to assume guardianship, which will allow them to consent to medical treatment for the young person. This is a legal procedure handled within the probate court of most counties. The caregiver must file a petition, an independent evaluator and attorney for the proposed ward is assigned, and a hearing is held. If guardianship is granted, the caregiver will have the legal authority to make personal and financial decisions for the person with JHD.

Supplemental Security Income

Supplemental Security Income (SSI) is a federal program of cash assistance to disabled needy individuals of any age, including children. Under SSI, children are considered disabled if they have a physical or mental condition which is so severe that it results in marked and severe functional limitation. The individual's condition must last or be expected to last at least twelve (12) months, or be expected to result in the child's death. An important component of this program is the fact that after an individual is determined eligible for SSI benefits

he/she is automatically qualified to receive Medicaid benefits as well. Some states provide an additional cash benefit to individuals receiving SSI.

The exact amount of SSI for which a claimant is entitled depends on what the individual (or the parents in the case of a child) owns and how much income he or she has. Eligibility requirements and benefit payments for federal SSI benefits are identical throughout the 50 states and the District of Columbia. For more information, check the website for Social Security at <http://www.ssa.gov> or call (800) 772-1213. A disability law attorney can also provide state-specific information and guidance in planning for eligibility for SSI benefits.

HDSA Chapter Social Workers and Center of Excellence Social Workers are very knowledgeable about the resources in their state and region. Social workers will help you get started with services, and then help with advocacy as needed.

Medicaid

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

Medicaid may pay for such services as hospital and doctor bills not covered by insurance, home healthcare services, medical transportation, and nursing home care. Many states also have waiver programs under which an individual who is disabled but does not meet the financial criteria may still become eligible for benefits. Because the eligibility criteria and available benefits vary greatly by state, one should contact the State or County department that administers the Medicaid program in his/her state (called Medi-Cal in California) or a disability law attorney for information specific to the individual's state.

It may be helpful to ask your pediatric healthcare provider or medical social worker about the Children's Special Healthcare Needs programs. This program exists in every U.S. state and territory, but it may be called "Crippled Children's Services" or "Children's Medical Services and Handicapped Children's Program." The

program starts with medical eligibility and then considers income eligibility. These programs may provide "waivers" for families who do not meet the standard eligibility requirements for Medicaid.

Other Financial and Placement Considerations

Many states do not have nursing home beds for people under sixteen years of age, with some very specific exceptions for short-term care. Because of this, there has been an increase in home-based services for people with severe and chronic illnesses and also in respite services for their caretakers. Respite care offers short-term care in order to give a break to the regular caregivers. Community Mental Health programs, which in many states cover virtually every corner of the state, are frequently involved with children deemed to be "developmentally disabled" and may be a source of information on respite programs.

The Tax Equity Fiscal Reform Act (TEFRA) is one special program for children without family health insurance, or for a child who has "topped out" the lifetime limits of the family insurance. TEFRA will help cover necessary treatment and support, often in a home setting.

Assistance may also come from the public education programs of the state. Under the Individuals with Disabilities Educational Act (IDEA), children with severe disabilities must be accommodated with services such as transportation, variable school hours as needed, and personal assistance to participate. Services available to private schools and to charter schools may vary from jurisdiction to jurisdiction. Some states may offer additional benefits and services. No state can reduce the benefits of federally mandated programs.

Finding entry points for care is difficult, even for experienced professionals. The rules can be complicated, but most professionals want to help a family with a disabled child obtain all appropriate benefits. Expect to make numerous phone calls and follow up letters to clarify eligibility and to gather information. Keep a log of calls, correspondence and visits. Expect to be a bit discouraged by the necessary paperwork, but know that persistence pays off in the end.

VIII. Hope for the Future

Now, more than ever before, there is reason to hope for improved treatments for HD. The discovery of the HD gene in 1993 has resulted in an explosion of experimental research with new findings and insights about the disease announced almost every month. With the tools of modern molecular biology, it is possible to "cut and paste" genes and parts of genes and to create "HD models" in fruit flies, mice and other animals that can be used in experiments to understand different aspects of the gene, the HD protein, or the disease.

HD researchers are beginning to collaborate with researchers of other brain disorders, such as Parkinson's and Alzheimer's diseases. With good reasons for optimism, we foresee the worldwide HD community of families, friends, clinicians and researchers working together to find the final pieces of the HD puzzle, and to develop effective therapies for those facing this devastating disease.

For More Information: visit the HDSA website at www.hdsa.org or phone the HDSA National Office at 1-800-345-HDSA. HDSA can direct you to local chapters, HDSA Centers of Excellence for Family Services, and other sources of information.

The Huntington's Disease Society of America (HDSA) is dedicated to:

- promoting and supporting research to find a cure for HD
- helping people and families affected by the disease, and
- educating the public and healthcare professionals about HD

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**Huntington's Disease
Society of America**

Published with funding from
a generous educational grant from
The Bess Spiva Timmons Foundation, Inc.