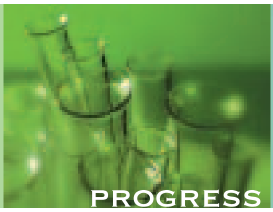
THE marker

Huntington's Disease Society of America

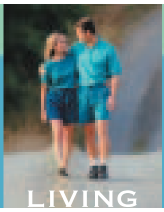
Commitment to Care

The HDSA Centers of Excellence for Family Services



PROGRESS
IN RESEARCH

HD
KIDS



LIVING
AT RISK

CARE

a message *from*

What a Difference a Friend Can Make

Friends are very important to me, as I am sure they are to you. Some friends last a few years; others, you have for a lifetime. They are there for every important, and not so important, moment of your life. My very good friend Nick Lopardo and I have known each other and worked together for years, before he joined State Street Corporation and rose to Chairman of State Street Global Advisors, the firm's investment management arm. Nick knows how deeply I care about finding a cure for Huntington's Disease and how much our HD families mean to me. So when I asked him if he would allow HDSA to honor him at our Boston Celebration of Hope Dinner on May 24, he was ready not only to accept the honor but to also show his support for our commitment to care and cure.

Nick Lopardo, Chariman and CEO, State Street Global Advisors

Nick is like so many of you who have chosen to join our HD family. You understand how important it is to move research ahead while providing support and services to those affected by the disease. Boston is a hub for HD research. It supports HDSA researchers at both Massachusetts General Hospital and Brigham and Women's Hospital. But more importantly, it is a primary nexus between the discovery of the HD gene in 1993 and current HD research.

Nick credits his parents with making him the concerned, dedicated citizen he is today. He expressed his hope that together with Diane, his wife of 33 years, the Lopardo legacy of honesty and hard work has been passed down to each of his three children. When he saw *Generation 2000 - Making This the Last Generation with HD*, Nick was so moved by the courage and determination of our HD families and by the optimism of our researchers, that he had to become involved. So Nick has joined with us in our commitment to the care and cure of Huntington's Disease. And now he wants to pass down something to our HD family - hope for a generation free of HD; hope for a cure. Nick Lopardo has demonstrated his commitment to HDSA, and has made it possible to guarantee community support in the future, with his generous $25,000 lead gift to the New England HDSA Center of Excellence at Massachusettes General Hospital and Tewksbury Hospital.

Help us find more leaders like Nick Lopardo. Help us move HD research forward by contributing generously to HDSA's new research campaign - *Generation 2000: Fulfilling the Promise*. Help us to raise $25 million for HD research in the next 5 years. Together, we can *fulfill that promise to make this the last generation with HD*.

Sincerely,

Donald A. King

Don King, Ph.D.

The Marker is an official publication of the Huntington's Disease Society of America, Inc. 158 West 29th Street, 7th Floor, New York, NY 10001-5300. (212) 242-1968

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The Marker, a periodical of the Huntington's Disease Society of America, Inc., is published twice annually. Its purpose is to provide information and opinion and to relay items of interest to individuals with Huntington's Disease and their families, health care professionals, and interested friends and supporters.

HDSA, Inc. is a national not-for-profit organization founded in 1986 to help individuals with Huntington's Disease and their families.

The Society is a member of the National Health Council, the National Foundation for Brain Research, the International Huntington Organization, the National Organization for Rare Disorders, the National Voluntary Health Agencies, the Alliance of Genetic Support Groups and the Independent Sector.

The Huntington's Disease Society of America meets all nine standards of the National Charities Information Bureau.

a message *from*

In 1967, Marjorie Guthrie created an organization dedicated to giving a voice to the thousands of people suffering in silence from Huntington's Disease. Today, the Huntington's Disease Society of America continues Marjorie Guthrie's commitment to finding a cure for this devastating disease while caring for our HD families. In the inaugural issue of *The Marker* magazine, HDSA featured its commitment to finding a cure through the many research initiatives funded by the Society.

This second issue of the new *Marker* magazine focuses on HDSA's commitment to providing care to those who suffer from the devastating effects of HD and those who offer support to them. We dedicate this issue to their courage and perseverance in the face of this deadly disease.

As we enter this new millennium, let us pause to look back in wonder at what we have accomplished. In 1967, before Woody Guthrie died, no one had heard of Huntington's Disease. In 1993, the HD gene was identified, and today, researchers and scientists view HD as a "model" for neurodegenerative diseases. They believe that the answers to the HD puzzle may also hold the key to solving the riddle of other diseases, such as Parkinson's and Alzheimer's.

For the HD community, the answers cannot come quickly enough. In 1997, HDSA launched the Center of Excellence program, an innovative concept in care that offers medical services for our HD families. From that first Center at Emory University, the program has grown to 11 Centers providing state-of-the-art services and support for our families in need including local resources and referrals, professional training and printed materials. This issue of *The Marker* profiles HDSA's current Centers of Excellence and the programs that make each unique.

Two Centers offer "telemedicine," the ability to consult with primary care physicians and our HD patients in outlying areas through videoconferencing. These two Centers are profiled in the *"In Focus"* section of this issue. You'll also read about HDSA's signature community-based events, the "Celebrations of Hope," that raise a portion of each Center's yearly grant.

In June 2000, the HDSA National Board of Trustees adopted an ambitious plan to expand the Center of Excellence program from 11 Centers to 25 by 2005. HDSA is pleased to announce that up to a maximum of 5 new Centers could be added later this month, bringing the current total to16 Centers of Excellence across the US. Total funding for the Center of Excellence program will increase from $705,000 in 2000 to more than $1,095,000 in 2001. HDSA, through its Center of Excellence program, volunteer-based chapters, affiliates and support groups, sustains a network of services and support that is unparalleled in its scope of coverage.

As we grow from 11 Centers of Excellence to 25 by 2005, we ask for your help and support. Every dollar raised for the Center of Excellence program is used in support of the yearly grants awarded to the Centers by HDSA. Whether you support the entire Center program or a specific Center, your gifts help to create and expand a national network of services for those affected by HD and their families. If you would like to help support the Center of Excellence program in your area, please call the National office.

Sincerely,

Barbara Boyle

Barbara T. Boyle
National Executive Director/CEO

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Our special thanks

to
AstraZeneca
for the
Community Service Grant
that made this
publication possible

HDSA CENTERS OF EXCELLENCE *for Family Services*

The cornerstone of HDSA's continued commitment to care is the HDSA Center of Excellence program. HDSA Centers of Excellence serve as the linchpin for HDSA's care initiatives for our HD families throughout the country. HDSA Centers of Excellence are multidisciplinary facilities that bring together the very best medical and social services under one roof. Centers of Excellence work in tandem with HDSA's volunteer-based chapters, affiliates and support groups to complete a nationwide network of resources and referrals.

History and Mission

In 1997, HDSA launched its very first HDSA Center of Excellence for Family Services at Emory University in Atlanta GA. This fledgling program has since grown, in just 3 short years, to 11 Centers of Excellence spread strategically across the United States to serve those affected by HD and their families and to be a resource for allied health care professionals. HDSA hopes to open a maximum of 5 additional Centers in 2001 and further expand the program to a total of 25 Centers of Excellence by 2005.

The HDSA Center of Excellence program is the gateway for our HD families to clinical trials and pilot programs that are not available at local health care facilities. All Centers of Excellence are also Huntington Study Group sites serving as recruitment centers for volunteers who wish to participate in clinical trials of possible therapeutics or in other projects that may lead ultimately to a cure for HD.

HDSA Centers of Excellence are catalysts for change. While each provides a core of services mandated by HDSA, each also participates in new research initiatives, pilot programs and special services that are unique to their scope of service. In this issue you'll read about the pilot "telemedicine" programs at the University of Iowa and the University of Virginia, as well as HD camps offered in several states.

The Making of a Center of Excellence

When HDSA developed the concept for a network of multidisciplinary health care facilities across the United States, a team of HD medical specialists also developed a set of standards that guide the Center program from an initial application request through the three-year probationary period and then annually to ensure that our HD families receive the very best care possible.

Before a facility is designated an HDSA Center of Excellence it goes through a rigorous review process that provides the Center Review Team with a comprehensive list of core services that will be provided as well as special services that a Center may offer outside the standard services. Each Center is ranked by the Center Review Team and the top Centers are identified for the next round of national funding.

Core Services

While each Center offers unique programs and services, all Centers have:

- Neurologists: Medical specialists in the treatment of the diseases and disorders that affect the nervous system, with expertise in Huntington's Disease.
- Speech, Physical and Occupational Therapists: Professionals trained to identify the challenges facing patients and to recommend exercise programs, therapies, strategies and aids to cope with the progression of HD.
- Genetic Counselors and Genetic Testing Services: Trained professionals who guide individuals and families "at risk" through the process of genetic testing and its outcome. HDSA maintains a national roster of genetic testing centers that conform with the guidelines developed in conjunction with HDSA for genetic testing.
- Trained Social Workers: Professionals who assist HD families in managing and coping with the disease, and also

act as advocates for the family with insurance companies, employers, etc.

- Regional Referrals: HDSA maintains a regional list of experts in various HD specialties through both the Center program and through its Chapters.
- Dietary Management: Information on maintaining weight in HD patients or referrals to nutritionists and dieticians.

Funding

HDSA funds each Center through a yearly grant that can grow from an initial $50,000 to a yearly grant of $100,000 based upon satisfactory completion of yearly goals. Each Center is evaluated annually and funding is determined by the level of services offered and by community support for that Center. Presently, HDSA funds in excess of $705,000 for its 11 Centers of Excellence for Family Services and this is projected to increase to $1,095,000 by June 2001.

Building Regional Support

A Center of Excellence does more than just provide medical services and support. Each Center serves as the regional representative of the Huntington's Disease Society of America. Through educational programs for allied health care professionals and health fairs for the general public, HDSA Centers of Excellence reach out to their respective communities to increase understanding and awareness about Huntington's Disease. By building alliances within the community, HDSA is able to raise a portion of each Center's funding through community-based "Celebration of Hope" events and contributions from local clubs, businesses and private donors. The balance of support for Center grants is derived from general donations made to HDSA throughout the year.

Clinical Trials

One criteria in the selection process for a potential HDSA Center of Excellence is that the proposed medical facility be a member of the Huntington Study Group (HSG). This then enables each HDSA Center of Excellence for Family Services to provide access to clinical trials. Members of our HD families can help researchers work toward therapies and ultimately, a cure for HD, by enrolling in these research trials. Some trials test treatments, such as medications that may slow the progression of the disease; other studies such as PHAROS and PREDICT-HD may be observational studies designed to aid in planning future therapeutic trials. A list of current HD clinical trials will be found on page 20.

HDSA Centers of Excellence fulfill an important part of the Society's mission to provide support and services for our HD families as we build toward a *future free of HD*.

HDSA *thanks*

OUR PARTNER

SunBridge Care

& Rehabilitation

for their support

NEW HD INFORMATION SLIDE KIT AVAILABLE SOON

The HDSA Slide Kit for Healthcare Professionals, appropriate for neurologists, genetic counselors and others involved in the care of Huntington's Disease, is in development. A "working model" of the slide kit was presented at the Centers of Excellence Conference for comments and review. This exceptional tool is made possible by an Educational Grant from AstraZeneca Pharmaceuticals.

HDSA CENTERS OF EXCELLENCE *for Family Services*

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**HDSA Satellite Program brings services to areas that are not yet supported by an HDSA Center of Excellence.*

spotlight
HDSA CENTER OF EXCELLENCE *for Family Services*

EMORY UNIVERSITY


Randi Jones, Ph.D., Director

Protective Fish Oil Derivative to be Tested in Research Trial

The HDSA Centers of Excellence for Family Services at Emory University and at Johns Hopkins University/Johns Hopkins Hospital are launching a clinical research study on ethyl-EPA (also known as ethyl-eicosapentanoic acid). Ethyl-EPA is a fatty acid that has potential for protecting brain cells damaged in Huntington's Disease. Ethyl-EPA is a pure derivative of fish oil, manufactured by Laxdale, Ltd., a drug company in Scotland. The substance has been administered to HD transgenic mice and to a small number of HD patients with promising results. Now the substance will be studied in a randomized double-blind clinical trial of approximately 120 patients at four sites worldwide.

For the first six months of the trial, half of the subjects will receive ethyl-EPA and half will be given a placebo without the substance. Neither the patient nor the physician will know which patients are receiving the placebo. Data will be analyzed to determine whether there is a large positive effect on HD symptoms within the treatment group. If there is, it may indicate that ethyl-EPA has a beneficial effect and all patients in the trial will then be offered the drug for another six months. At the end of the one-year study, all subjects will be offered the drug for another year.

Criteria for inclusion in this trial include:

- Clinical diagnosis of HD
- Early-stage symptoms of HD
- Age 30-70
- No use of fish-oil supplements for 60 days
- Absence of significant complicating medical problems
- A caregiver to accompany the subject to all visits and oversee the administration of the study drug

The HDSA Center of Excellence at Emory will be recruiting twenty subjects for the trial while the HDSA Center at Johns Hopkins University/Johns Hopkins Hospital will be recruiting thirty individuals. To be considered for this study, contact either Dr. Randi Jones, (404) 728-6849, at the HDSA Center of Excellence at Emory University or Debbie Pollard, (410) 955-2398, at Johns Hopkins University/Johns Hopkins Hospital.

JOHNS HOPKINS


Christopher Ross, M.D., Ph.D. and Adam Rosenblatt, M.D., Co-Directors

New Staff Added to Multidisciplinary Team

The Johns Hopkins HDSA Center of Excellence is expanding the scope of its services in 2001. As the number of individuals and families visiting the Center continues to grow, additional staff has been added to the multidisciplinary team. Kit McFarlane LGSW, utilizes her knowledge of the psychosocial aspects of the disease to complement the expert medical care provided by staff physician, Vani Rao, MD.

This cooperative approach is especially important to the many individuals who are referred to the HDSA Center for diagnosis or for a second opinion. At the Center, genetic testing for HD is offered within a framework of pre- and post-test counseling. A new support group for the local HD community has also been formed. Outpatient follow up is a feature of the Center, as are visits to care facilities located outside the Johns Hopkins facility.

HDSA CENTER OF EXCELLENCE *for Family Services*

HENNEPIN COUNTY MEDICAL CENTER

Martha Nance, M.D., Director

In-Service Program Informs Nursing Home Staff

In addition to its on-site services, the HDSA Center of Excellence for Family Services at the Hennepin County Medical Center (HCMC) provides educational outreach to nursing homes with HD patients. The Nursing Home In-Service Program is directed by Beryl Westphal, RN, CNP and HD Clinic Manager. This special pilot program is designed to keep the nursing home staff informed about the special needs and challenges of this progressively degenerative disease. When a patient is referred to the HD clinic from a nursing home, or when a call is received from an extended care facility, an in-service presentation is scheduled.

Since the program began in 1999, presentations have been made to about 300 health care professionals at 12 nursing homes throughout Minnesota. Those attending have included administrators, nurses, nurses aides, therapists (physical, occupational, speech and recreational), dietary technicians and housekeeping staff. Often, after a presentation is made, staff members choose to visit the HCMC for a day to learn more about the Centers' multidisciplinary approach to the care of HD patients. The Center at Hennepin County hopes to expand this program in 2001 by including additional members of the HCMC team from various disciplines at the presentations. The HDSA Center of Excellence at HCMC also plans to introduce a "Less Smoking is Better Campaign" at the University Good Samaritan Care Center's HD unit as a joint effort of the Center, Good Samaritan, the local HDSA chapter and local donors.

MASSACHUSETTS GENERAL HOSPITAL

Anne Young, M.D., Ph.D., Director

Collaboration for Multidisciplinary Care

The New England HDSA Center of Excellence is a collaborative effort of Massachusetts General Hospital with Tewksbury Hospital, SunBridge Care and Rehabilitation of Lowell and Boston University to provide the best clinical care possible to HD patients. In addition to clinical services and resources for families, educational lectures have been developed for patients and their families on topics such as advances in research and the medical and legal issues related to genetic testing. A web site will soon provide information about research and clinical trials.

Tewksbury Hospital and SunBridge Care and Rehabilitation of Lowell provide long-term care for HD patients in the more advanced stages of the disease. Tewksbury has hosted a caregiver support group for over 10 years and last year opened a newly renovated 32-bed unit dedicated to HD patients that includes short-term medical admissions to provide families with a break from their caregiving responsibilities. SunBridge Care and Rehabilitation of Lowell affords HD patients quality skilled nursing and long-term residential care.

Massachusetts General Hospital has long been a leader in HD research. The new Center of Aging, Genetics and Neurodegeneration (CAGn) is designed to foster a collaborative research effort to find treatments for HD and other neurodegenerative diseases. The Center is a site for both non-Huntington Study Group and Huntington Study Group (HSG) sponsored clinical trials, including CARE-HD, PHAROS, and PREDICT-HD.

For more information about the New England HDSA Center of Excellence and its affiliated programs, please call the Center or the National Office.

COLUMBIA HEALTH SCIENCES

Karen Marder, M.D., Director

Telling Children About HD

When HD is diagnosed in a family, children are often the last to be informed. Telling them about HD and what it may mean to their family is a sensitive and complex undertaking. Educating kids about HD and counseling their parents about how to discuss the subject are a special focus of the Center of Excellence at Columbia Health Sciences.

Children frequently ask the hard questions straight out. They may ask if a family member is going to die or if they are going to get the disease. Center Social Worker, Deborah Thorne noted, "The important thing is to present the information in as upbeat a manner as possible, while still telling the truth. Focus on the pace of research and the hope it represents."

Children should have an understanding of the disease before they reach adolescence. HD should be discussed in short sessions over time rather than as one full-blown discussion that may overwhelm them.

Last April, the Center hosted a professional training conference for 25 social workers and genetic counselors from other HD clinics and Centers. Presentations that shared professional expertise were given in order to add to the skills and abilities of those in attendance to work with HD families as they cope with the disease. For more information about talking to children about HD, contact the Center at Columbia Health Sciences or the HDSA National office.

UNIVERSITY OF VIRGINIA

Madaline Harrison, M.D. Director

Support, Outreach and Education

This Center offers an informal half-hour educational presentation, on a different aspect of HD, followed by separate support group meetings for patients and caregivers, as part of its regular 1/2 day monthly clinic.

Outreach programs at the Center include satellite Genetics clinics in Lynchburg, Roanoke, Abingdon, Danville and Winchester, Virginia and in Beckley, West Virginia. These clinics provide information about Genetic testing, as well as pre- and post-test counseling. As part of the outreach program, Dr. Madaline Harrison, a neurologist and program director, will offer services at some clinics. Clinical evaluation and follow-up, via the Center's innovative Telemedicine program, is an important part of the program at the University of Virginia. In addition to clinical services through existing Telemedicine sites, a pilot program for multidisciplinary follow-up care is currently being developed with the Telemedicine office at the Winchester Hospital. See the special report on "telemedicine" on page 11.

The HDSA Center of Excellence at the University of Virginia also offers symposiums like the one organized by the Center and the Tidewater area HD support group. This unique opportunity, held on November 4, 2000 in Newport News, Virginia, provided information about both the clinical and genetic features of HD, and was presented by Dr. Barbara Haskins and Dr. Madaline Harrison of the Center. Breakout sessions led by members of the UVA HD Team addressed caregiver concerns, daily and long-tem care, genetic testing and medication issues. A panel discussion by the UVA Program staff completed the program. Contact the HDSA Center at UVA for information about future events.

HDSA CENTER OF EXCELLENCE *for Family Services*

UNIVERSITY OF IOWA

Jane Paulsen, Ph.D., Robert Rodnitzky, M.D. and Henry Paulsen, M.D., Ph.D., Co-Directors

Computers Aiding Communication

When Jared, a long-time patient of the HDSA Center of Excellence at the University of Iowa's HD clinic, returned this fall, his doctors saw that he was losing his ability to communicate and express himself. He could no longer tell the jokes and stories the staff had come to expect. Dr. Jane Paulsen and Center coordinator Elizabeth Penziner decided to see if anything could be done to help. In consultation with Richard Hurtig, Ph.D. and Jodi Morgan, CCC-SLP, speech pathologists at the Center, the DynaMyte3100, a computerized device known as an "ambulatory communicator" was suggested. This durable, lightweight keyboard device "speaks" in an electronic voice when picture keys are touched. The device can also be programmed as the "remote control" for any consumer appliance that has infrared controls.

Soon after its introduction, Jared was able to assemble simple phrases and messages to communicate with his family and friends at the Center. The multidisciplinary approach taken at the University of Iowa Center of Excellence has made it possible for one young man with HD to reconnect with his loved ones and caregivers.

The loss of communication is a leading cause of depression in HD. Augmentative devices can dramatically improve the quality of life. For more information about augmentative devices, please contact your nearest HDSA Center of Excellence.

The Dynamite 3100 is manufactured by DynaVox Systems in Pittsburgh, PA. If you are a health care professional, and would like more information about DynaVox products, please call DynaVox Systems at (888) 697-7332.

The Huntington's Disease Society of America does not endorse or promote specific products mentioned in articles contained within this magazine nor does it recognize any claims made for the product by the manufacturer.

OHIO STATE UNIVERSITY

Sandra Kostyk, M.D., Ph.D., Director

HD Teens Meet for Support

The teen years are a time when peer interaction is especially important. For teens facing the challenges of HD in their family, this need is acute. To address these special needs, the HDSA Center of Excellence at Ohio State University College of Medicine & Public Health and the Central Ohio Chapter of HDSA have begun a support group for teens 10-18. The support group offers a confidential, non-judgmental outlet where teens can share their experiences in living and coping with HD in their family.

Support groups help teens cope with the changes that can confront their families. At-risk teens share a unique and complex set of problems. Joining a support group of people their own age can help to prevent depression, self-doubt and self-judgment. Here they can talk candidly about the realities of living in an HD family.

Peer support groups help relieve the burden of isolation as teens struggle to handle the difficult adjustments that are part of belonging to an HD family. Many HDSA chapters and Centers of Excellence organize support groups for teens, caregivers, HD affected individuals and adults living at risk. Contact HDSA at (800) 345-HDSA or www.hdsa.org to find a support group near you. For more information about the teen support group at Ohio State University, please contact Barbara Heiman, LISW at the HDSA Center of Excellence at OSU, at (614) 292-9960.

UNIVERSITY OF ROCHESTER

L to R bottom—**Charlyne Hickey, RN and Co-director Peter Como, Ph.D.** *Top*—**Co-directors: Ira Shoulson, M.D., and Karl Kieburtz, M.D.**

Expanding Comprehensive Care in Upstate New York

The HDSA Center of Excellence at the University of Rochester provides comprehensive care to HD families. The Center outreach program has been expanded to include several statewide teaching days, regular in-service trainings to direct care staff and more HD support groups to better serve the entire Upstate New York region.

In June 2000, a Saturday half-day workshop was held in conjunction with the HD Buffalo support group and Upstate HDSA chapter. In November, an all-day teaching conference in Syracuse focused on "Living Positively with Huntington's Disease." A joint HD conference will be held on April 28, with the Albany Medical College. This will be the Center's first event in the state capital district. These conferences have been facilitated by the Center's longstanding relationship with the HDSA Upstate NY chapter.

Through outreach, the Center offers monthly HD support groups in Buffalo, Rochester, Syracuse and Binghamton with plans to re-establish a group in the Albany area. Meetings are generally co-facilitated by an HD family member and a professional care provider.

Training meetings are conducted monthly to help de-mystify HD and provide resources for education and clinical support. "HD Chats," meet in various settings, from hospitals to home care agencies and nursing facilities.

Caregivers are also recognized by the Center. Amy M. Chesire, CSW-R, MSG, the Center's social worker, has designed a special psychosocial assessment tool, which helps to identify the most effective caregiver coping strategies throughout the phases of HD. It will be available to the HD community once it is established as a reliable and valid instrument.

NORTH SHORE UNIVERSITY HOSPITAL

Martin Bialer, M.D., Ph.D., Co-director and Andy Feigin, M.D., Director

Family Therapy Program Launched

HD is a disease with psychological effects as well as physical ones. These psychological stresses affect not only the person with HD, but the mental health of the entire HD family. Family and couple therapy can offer productive strategies for coping with the changes caused by Huntington's Disease. It can also provide a safe place to release strong feelings and emotions. As part of the Family Services program at the HDSA Center of Excellence at North Shore University Hospital, a Family Therapy program was launched in January 2001. Designed to augment and complement the multidisciplinary clinical services offered at the Center, this program will provide comprehensive family therapy to individuals and their families who are coping with the progression of Huntington's Disease. A monthly multiple-family group therapy program began in late February. Individual family and couple therapy is also offered at the Center.

Ellen Landau, CSW, Director of Family Programs at the School Mental Health Alliance, directs these innovative and exciting new programs that focus on helping family members to identify interpersonal issues and discuss how HD is affecting their family. By providing an ongoing forum for dialog and education, the Center at North Shore helps HD families develop coping strategies.

In addition, the Center is planning a research project that will document the impact of genetic testing on individuals and families. By gathering this kind of information, more effective ways can be developed to counsel individuals and families before, during, and after genetic testing.

HDSA CENTER OF EXCELLENCE *for Family Services*

COLORADO NEUROLOGICAL INSTITUTE

Expanding Services to Meet Need

The HDSA Center of Excellence at the Colorado Neurological Institute (CNI) is located in the Denver metropolitan area, serving families from as far away as Utah, New Mexico, Oklahoma, Montana, North and South Dakota and Texas. The Center's multidisciplinary team of HD specialists includes a neurologist, neuropsychologist, clinical psychologist, genetic counselor, clinical social worker, occupational therapist and physical therapist.

As public awareness of HD grows, so too does the need to provide support and services for affected individuals, their family members and caregivers. To meet the growing demand, the Center at CNI has recently expanded its HD clinic to twice monthly, thereby increasing opportunities for initial appointments and follow-up visits.

Other services offered by the clinic include monthly support group meetings and a telephone hotline. Plans to expand the level of care available to local HD families in 2001 include incorporating a "telemedicine" program as part of an outreach initiative to hospitals in Colorado Springs, Grand Junction and Fort Collins, CO as well as increasing the knowledge and understanding of HD among health professionals in the area served by the Center at CNI.

Expanding the *Center of Excellence* Program

As part of HDSA's continuing commitment to care, the National Board of Trustees has directed the Society to undertake an ambitious expansion program that includes increasing the number of HDSA Centers of Excellence from its current 11 to 25 by 2005. To accomplish this task, the Huntington's Disease Society of America has solicited applications from prospective centers across the US and may select, during the March meeting of the National Board of Trustees, up to a maximum of 5 facilities that meet the rigorous criteria for an HDSA Center of Excellence.

The candidates from which the new Centers will be selected for funding in 2001 are:

Baylor College in Houston, TX

Indiana University School of Medicine in Indianapolis, IN

UCLA Medical Plaza in Los Angeles, CA

University of California in San Diego, CA

University of California, Davis Medical Center in Sacramento, CA

University of Washington School of Medicine in Seattle, WA

Washington University School of Medicine in St. Louis, MO.

Be sure to read about the new Centers of Excellence in the May issue of *Toward a Cure.*

May is HD Awareness Month

Each May, the Huntington's Disease Society of America reaffirms its commitment to the care and cure of HD. Please join us in remembering those who have lost their battle with HD and help to bring awareness about this devastating killer to your community. HDSA has Awareness kits available for you to take to your local newspapers, schools, clubs, organizations, television, cable and radio stations. If you would like to help raise awareness and increase funding for HD research, please call the National Office or email hdsainfo@hdsa.org.

Telemedicine Aids HD Care in Rural Virginia

For many HD patients and their families, traveling to major medical centers becomes more difficult as the disease progresses. Local health care facilities often lack the range of services available at a large hospital. Telemedicine programs, such as the one organized throughout the HDSA Center of Excellence for Family Services at the University of Virginia, are being expanded to connect these patients and their primary health care providers with a full multidisciplinary team of HD experts.

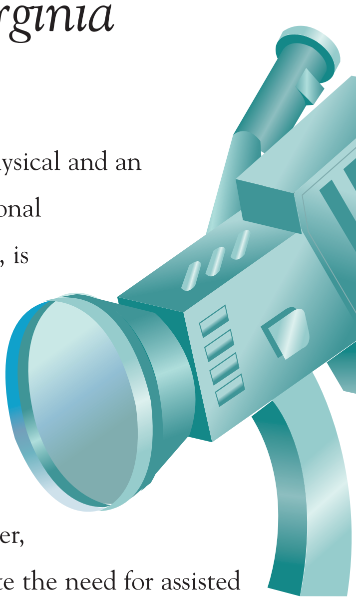
"This is potentially a very useful tool for hospitals serving large rural areas or those with distant satellite clinics," says Madaline Harrison, M.D., Director of the HDSA Center of Excellence at the University of Virginia in Charlottesville, VA.

Using a video camera and special interactive tools, Dr. Harrison can perform neurological examinations on patients at remote locations. Being able to interact with the patient and referring physician during the telemedicine appointment allows Dr. Harrison to discuss directly, with the patient and the physician, immediate strategies for managing the disease. "These exams are very effective," says Dr. Harrison. "It is possible to assess movement, balance and motor function. A lot can also be learned from simply seeing a patient's body language."

The UVA Telemedicine Program grew from a pilot program at the affiliated Virginia Neurological Institute's Center for Advanced Technology. "Prior to our designation as an HDSA Center of Excellence, I had used this strategy to evaluate several patients with movement disorders, one of whom had HD," noted Dr. Harrison. "The chance to expand our HD services under the Center of Excellence program offered a great opportunity to develop this further."

Where possible, initial evaluations are performed on-site, either in Charlottesville or at a satellite clinic, with the telemedicine function used primarily for follow-up visits. The focus of the program is to increase service in underserved, primarily rural, areas in Southwest Virginia, but the program covers the entire state, including Lynchburg to the south to Winchester in the north and connections are available with networks such as the Department of Mental Health that cover other areas. In the near future, clinical sessions, using an entire multidisciplinary HD team, will be available at satellite clinics throughout the telemedicine network.

Since receiving the designation as an HDSA Center of Excellence for Family Services in August of 2000, the Center has seen two HD patients who had been lost to follow up care since 1994 due to transportation problems. A multidisciplinary consultation, involving both a Physical and an Occupational Therapist, is being arranged for one patient in Winchester, to evaluate the need for assisted living. This live, interactive videoconferencing will be a first in Winchester, where telemedicine had been used only for conferences and transfer of diagnostic test results.



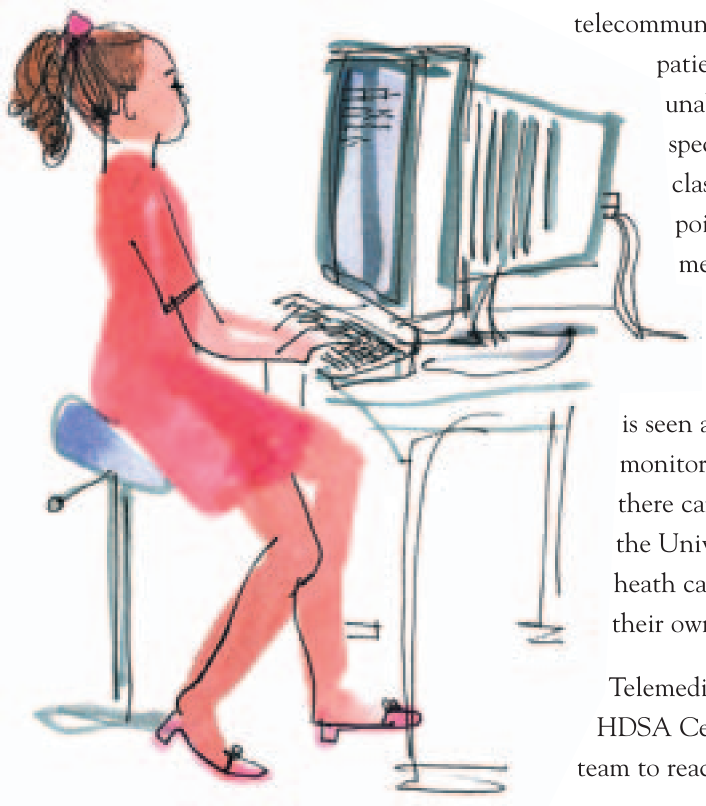
Recognizing the many possibilities this technology offers, the HDSA Center of Excellence at the University of Virginia hopes to include at least one regular telemedicine session per month per clinic when the program is fully established.

"I am very excited about the potential of telemedicine to expand our ability to reach patients and families with HD who can not travel to attend the clinics in Charlottesville," concludes Dr. Harrison. "It's a remarkably effective clinical vehicle and allows for a very interactive encounter both with the patient and family and their local provider. I anticipate that telemedicine will be a powerful tool in our efforts to reduce the isolation, including geographic isolation, experienced by so many of the patients and families with HD."

Telecommunications Aids Diagnosis and Patient Care

The HD helpline rings and Elizabeth Penziner answers in her usual calm and upbeat manner. Unfortunately, it won't help the situation at the care facility in Sioux Rapids, Iowa, where the Director of Nursing is, once again, surrounded by frustrated staff. "Joe is acting up again. He's soiled himself again and refuses to let the staff clean him up. He's being mean and simply not appreciating our efforts to help him. We can't get him to settle down. We don't want to put him in restraints, should we transfer him to your hospital?" The care facility is 5 1/2 hours away. It's a cold and snowy day and the unit at the University is full.

This scenario is not an unusual one for professionals caring for people with HD. What is unusual is the care facility's access to specialists in behavior management. "Telemedicine" is offering new ways for people to provide health care.



Telemedicine uses telecommunications technology for medical diagnosis and patient care. The Telecommunications Services at the University of Iowa, headed by Michael Kingsley, provide voice services for approximately 14,000 devices, data services for over 5,500 data devices, and support for over 2,500 radio pagers. In addition, the Iowa Communication Network (ICN) is a statewide fiber optic network supporting video, data and voice services. Iowa is fortunate to have the nation's first and only statewide fiber optic infrastructure available for educational and health-related applications. With connection in all 99 of Iowa's counties and over 400 endpoints currently networked, the ICN is a national leader in medical telecommunications. Even if a patient or facility is unable to access the specific ICN classroom, a point-to-point delivery method can reach almost any Iowa location. The remote site is seen and heard on a TV monitor and individuals there can, in turn, watch the University of Iowa heath care professionals on their own monitors.

Telemedicine allowed the HDSA Center of Excellence team to reach out to the frustrated staff in a remote northwest Iowa care facility on that snowy day in December. The Center staff was able to present an overview of Huntington's Disease and specifically address the behavioral concerns that were mentioned in conversations with the patient and facility staff. Joe's caretakers were able to voice their concerns and to engage in an interactive face-to-face discussion about Joe's individualized needs. In so doing, all those involved served as part of the solution and the intervention was collaborative rather than directive. Strategies were shared and all participants experienced a sense of teamwork. Several resulting benefits of this mutual effort included developing a "game plan" for Joe's on-going care that avoided the need for hospitalization or other costly disruptions to Joe's routine. More importantly, a support system was established for the facility staff that they could rely upon in their continued care of Joe.

Despite these exciting advances in telemedicine, the personal touch remains important. The Center of Excellence staff also provides on-site Huntington's Disease in-services when it can be planned more efficiently (i.e. in better weather or while visiting several facilities in one area of the state). Whenever possible, telemedicine is used as an ancillary rather than primary mechanism of service delivery to those living in rural areas. Nevertheless, providing much needed information to facilities state-wide enhances the overall quality of care for patients with HD.

IN focus

HD Camps and Retreats: Fun, Food and Friends

Picture a group singing around a campfire, laughing and smiling. The mood is relaxed and intimate, a group of friends enjoying the out-of-doors. The fact that everyone in the circle is either a person with HD or an HD health care specialist is the only exceptional part of the scene. This is the reality of HDSA-sponsored retreats and camp programs.

Through its Centers of Excellence for Family Services and Chapters, HDSA is committed to improving the quality of life for people with HD. As the disease progresses, it can limit opportunities for recreation and enjoyment. Finding a place that is fun and safe and welcoming to individuals with HD is a challenge. HD retreats are one way to provide a comfortable camping experience for people with early to mid-stage HD. They offer social and recreational opportunities and can break the isolation that often comes as the disease progresses. HD camps and retreats also give family members and caregivers a much-needed rest

Activities vary by camp, but most include: hiking, music or dance therapy, team sports, swimming, baseball, volleyball, horseback riding, campfires, singing, crafts and often a dance.

Eating is an important part of every day's schedule. Delicious, easy to eat, high calorie foods and drinks are served five times a day. People who attend the camp often gain five pounds!

The staff at an HD camp or retreat consists of a dedicated team of health care professionals that can include physicians, physical and occupational therapists, nurses, speech pathologists and swallowing experts.

Therapy is often combined with fun and relaxation.

Carol Moskowitz, RN-C, NP, co-directs the HD camp program affiliated with the HDSA Center of Excellence at Columbia Health Services/New York Psychiatric Institute. She also provides valuable assistance to other HDSA Centers and Chapters to establish new camps and retreats. Carol speaks of last summer's campers with pride. “They took the risk, came to camp and created a magic space where everyone belonged and no one was bored or alone. They declared ‘We are normal’ and ‘I left my HD at the gate’. The true heroes of the camp are the people who taught us by example and touched our lives forever.”

Camps are planned for 2001 in Massachusetts, Michigan, New York (Rochester), Georgia, and Pennsylvania. To learn how to you can enroll in an HD camp for 2001 or organize a camp through your local HDSA Chapter, contact the National Office at 800-345-HDSA.

HDSA *thanks*

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HDSA CENTERS OF EXCELLENCE
September Conference 2000

The Huntington's Disease Society of America (HDSA) hosted its inaugural Centers of Excellence Conference in Elk Grove Village, Illinois, on September 16-17, 2000. Approximately 40 individuals, including Center of Excellence directors, neurologists, social workers, genetic counselors, nurses, prospective center directors and HDSA staff members, attended.

The conference provided an opportunity for the various Center staffs to meet and learn about the different programs at each clinic, as well as to provide a forum to discuss and exchange ideas on problems relating to "clinic day". Directors from each center presented information about their clinics' activities, staff availability and types of services offered, coordination of nursing home care, number of patients seen on an ongoing basis and relationships with HDSA and HDSA chapters.

During the conference, the master plan for the future development of the Center of Excellence program was discussed. One of the strategic goals adopted last year by the HDSA Board of Trustees includes designating and funding a total of 25 centers by the year 2005.

Other conference sessions included a discussion of HDSA's new grant and renewal process for Centers and a presentation on the development of educational and fundraising materials that will be available to all HDSA Centers of Excellence. The continuing development of the Center of Excellence at Emory University School of Medicine was also featured. This presentation illustrated how the Center has increased its activities to support our HD families and which programs worked the best. As the first site to receive an HDSA Center of Excellence designation, this Center serves as the "model" and vanguard of the program.

Throughout the conference, those in attendance reaffirmed their continuing commitment to caring for individuals and families affected by HD. The 2001 conference is already in the planning stages and will include additional forums and training opportunities for the many health care professionals and others who provide medical and social support to our families.

> *Throughout the conference, those in attendance reaffirmed their continuing commitment to caring for individuals and families affected by HD.*

The meeting was made possible by a generous Educational Grant from AstraZeneca Pharmaceuticals.

Coming in June 2001

HDSA is pleased to announce the addition of two new publications to the very popular *Family Guide Series*. Two new titles will premier in June and will focus on *Long–Term Care* and *Juvenile HD*. *The Family Guide Series to Long–Term Care* has been authored by Suzanne Imbriglio, PT, Virginia Goolkasian, LSW, MS and Ann Murfitt, MSW, LICSW. *The Family Guide Series to Juvenile HD* has been made possible by a generous grant from the American Contract Bridge League Charity Foundation Corporation.

Also arriving in June is the fourth title in HDSA's *Professional Handbook Series* entitled *The Juvenile HD Handbook*. This valuable resource, written by Martha Nance, M.D., provides information about diagnosis, treatment, issues relating to school and peer interaction, to allied health care professionals and HD families alike. A resource directory also offers helpful information. *The Juvenile HD Handbook* was made possible through a generous grant from the American Legion Child Welfare Foundation.

Please contact the National Office in June to find out how you can order your copies of *The Family Guide Series to Long-Term Care* and *Juvenile HD* as well as HDSA's *Juvenile HD Handbook*.

Chapters

HDSA's 31 volunteer-based chapters and 3 affiliates serve as the vital link between the Society and our HD families. HDSA chapters and affiliates bring the mission of the Society to those in need at the local level by providing services, support and education in the field. Chapters and affiliates also work to increase awareness about HD in their community and to raise the necessary funds in the battle against HD. For the chapter or affiliate closest to you, please consult the listing below.

*Alabama Affiliate**

3384 Cedar Brook Circle
Trussville, AL 35173
(205) 655-7090
Ktolleson@epenergy.com

Arizona Chapter

P.O. Box 7666
Phoenix, AZ 85011-7666
(602) 222-9312
(888) 267-3411 (helpline)

*Arkansas Affiliate **

P.O. Box 556
Morrilton, AR 72110
(800) 558-3370
(501) 791-0909 (helpline)

Central Ohio Chapter

Suite 317, McCampbell Hall
Columbus, OH 43210-1296
(614) 292-9660

Connecticut Chapter

P.O. Box 719
Southington, CT 06489
(1-800) 840-9907

Delaware Valley Chapter

529 Swede Street, Suite 100
Norristown, PA 19401
(610) 277-1006
Mae O'Brien, Executive Director

Georgia Chapter

P.O. Box 15298
Atlanta, GA 30333
(770) 279-9207
Ron Jackson, Executive Director

Greater Los Angeles Chapter

9903 Santa Monica Blvd., Suite 106
Beverly Hills, CA 90212
(800) 686-9868
(888) 4-HDSA LA (helpline)

Illinois Chapter

P.O. Box 597045
Chicago, IL 60659
(630) 443-9876

Indiana Chapter

P.O. Box 2101
Indianapolis, IN 46206
(317) 271-0624

Iowa Chapter

P.O. Box 3
Bedford, IA 50833
(712) 523-2291
(800) 535-4797 (helpline)

Kentucky Chapter

c/o Kosair Charities Center
982 Eastern Parkway
Louisville, KY 40217
(502) 637-4372
(800) 784-3721 (helpline, KY only)

Maryland Chapter

711 West 40th Street, Suite 354
Baltimore, MD 21211
(410) 467-5388
Kathy Alexander, Executive Director

Massachusetts Chapter

1253 Worcester Rd. Suite 202
Framingham, MA 01701
(508) 872-8102
Kimberly Gleason, Executive Director

Michigan Chapter

601 North Waverly Road
Lansing, MI 48917-2851
(517) 321-6511 (voice mail)
(800) 909-0073 (helpline)

*Mid-South Affiliate **

8950 Evening Grove Cove
Cordova, TN 38018
(901) 755-0205
midsouthhd@usa.com
Kathy Bergenson

Minnesota Chapter

22-27th Ave. SE Suite 211
Minneapolis, MN 55414
(612) 371-0904
(612) 673-6268

New Jersey Chapter

114 South Main Street,
Box 67A
Cranbury, NJ 08512
(609) 448-3500
(800) 424-2494 (helpline)
Pat Velasco, Executive Director

North Carolina Chapter

P.O. Box 681
Cary, NC 27512
(919) 469-0222

Northeast Ohio Chapter

c/o Alan M. Tartakoff, Ph.D.
Chapter President
Institute of Technology,
School of Medicine
Case Western Reserve University
Cleveland, OH 44106
(216) 368-5544 *or*
Eve Weisman **(216) 226-6633**

Northern California Chapter

9110-B Alcosta Blvd., #234
San Ramon, CA 94583
(510) 287-5969
(888) 828-7343

Northwest Chapter

P.O. Box 33345
Seattle, WA 98133
(206) 464-9598 (SW)
(888) 264-4372 (helpline)

Ohio Valley Chapter

3537 Epley Lane
Cincinnati, OH 45247
(513) 741-4372

Oklahoma Chapter

525 NW 13th Street
Oklahoma City, OK 73103
(405) 236-4372

Rocky Mountain Chapter

1045 Lincoln, Suite 100
Denver, CO 80203
(303) 321-5503
Dave Anderson, Executive Director

San Diego Chapter

P.O. Box 19524
San Diego, CA 92159-0524
(760) 752-1844

Sioux Valley Chapter

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

South Florida Chapter

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

St. Louis Chapter

8039 Watson Rd., Suite 132
Webster Groves, MO 63119-5325
(314) 961-4372 (also helpline)

Texas Chapter

P.O. Box 204086
Austin, TX 78720
(800) 910-6111

Upstate New York Chapter

115 Hardwood Lane
Rochester, NY 14616
(716) 341-7400

Washington Metro Area Chapter

8303 Arlington Blvd., Suite 210
Fairfax, VA 22031-2900
(703) 204-4634
(703) 323-1403 (helpline)

Western Pennsylvania Chapter

P.O. Box 4102
Pittsburgh, PA 15202
(412) 833-8180
(888) 779-HDSA

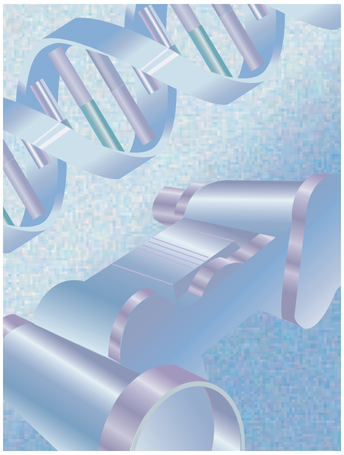
Wisconsin Chapter

2041 N 107th
Wauwatosa, WI 53226
(414) 257-9499
(879) 330-2699 (helpline)

research UPDATES

Why is Huntington's Disease So Important to Researchers?

Huntington's Disease (HD) is a hereditary degenerative brain disorder that slowly robs the affected individual of the ability to walk, talk, think and reason. HD is caused by the mutation of a single gene. It was identified in 1993 and since that discovery, much has been added to our understanding about HD.



Though Huntington's Disease affects an estimated 30,000 Americans and places another 200,000 "at risk" for inheriting the disease, the research now being performed has far wider implications. In 1997, the Huntington's Disease Society of America launched the HDSA Coalition for the Cure. What began as a handful of researchers in labs around the world, who shared the vision of collaboration with HDSA, has today grown to 17 labs worldwide, all working to solve the puzzle of HD. And as they apply their theories to unlocking the mysteries of the single gene that causes HD, they are beginning to see that the answers can be applied to many other diseases.

PROGRESS IN RESEARCH

In the past four years, amazing strides have been made in developing mouse models that mimic the symptoms of HD. These "transgenic" mice are used to find ways to delay the onset of symptoms, repair brain cells, and test promising new drugs. Other research targets large-scale screening of compounds, called a library, in a process known as high throughput screening, which holds the potential for identifying promising new therapeutics in the treatment of HD. Still other research studies the role that caspases play in cell death, specifically in the brain, and the cellular pathways that lead to disease progression. Once these pathways are identified, then therapies can be developed to block them and arrest progression of the disease.

SHARING COMMON GROUND

Today, scientists view HD as a "model" disease and believe that the answers they find can be applied to other "neurodegenerative" disorders that affect millions of individuals and include Amyotrophic lateral sclerosis (ALS), Parkinson's, Alzheimer's, Multiple Sclerosis and Muscular Dystrophy. Huntington's may also provide insight into other dominantly inherited genetic diseases, and other movement disorders such as dystonia and ataxia.

Huntington's Disease is also a trinucleotide (polyglutamine) repeat disease. HD creates an abnormal clumping of proteins in brain cells. Other repeat diseases that could be treated by advances in HD research include Spinocerebellar Ataxia (SCA) and Machado-Joseph Disease (MJD or SCA 3).

While the primary goal of HD research remains focused on discovering what "triggers" HD, and finding the means to delay onset, treat symptoms and ultimately, cure HD, the research we fund today will also help millions of people with related diseases. Our scientists tell us that each day we grow closer to finding the answers; to solving the mystery of HD.

FULFILLING THE PROMISE

HDSA is committed to fulfilling the promise made in 2000 to *make this the last generation with HD*. With your help, we can do that. HDSA researchers are sponsored by families and friends who join together each year to pledge a gift in support of a specific researcher. In return, they receive progress reports from "their" scientist and thus actively engage in the battle to unlock the secrets of HD. If you would like to establish a Family Fund for your friends and family or if you would like to underwrite an HDSA scientist or research project or if you would like to make a gift to HD research, please call the National Office.

HDSA Coalition for the Cure

In 2000, HDSA committed more than $2.5 million to the HDSA Coalition for the Cure and other research programs. It is projected that the Society's overall commitment to research will top $3 million in the coming year.

The HDSA Coalition for the Cure is a group of HDSA-funded scientists, representing 17 labs worldwide, who are dedicated to discovering effective treatments for HD while working toward a cure for this devastating disease. The Coalition targets four specific areas for research: bio-chemistry, cell biology, cell models and animal models.

The HDSA Coalition for the Cure is made possible through the following Founding Leadership Gifts: The Anonymous Fund for a Cure, The Helen Becker Memorial Research Fund, The Woody and Marjorie Guthrie Research Fund and The Kent Westbrook Endowed Research Fund.

In 2000, HDSA committed more than $2.5 million to the HDSA Coalition for the Cure and other research programs. It is projected that the Society's overall commitment to research will top $3 million in the coming year.

The Coalition for Cure meets twice a year. Below is an update of the fall meeting.

Research Report

HDSA Coalition for the Cure Investigators, Steering Committee members, and Medical and Scientific Advisory Committee members gathered in La Jolla, California to conduct the 8th semi-annual HDSA Coalition for the Cure Research Conference. Coalition members met from October 25-26, 2000 and shared in exciting, oftentimes bold and frank, discussions regarding the latest HD research emerging from Coalition laboratories.

Several ancillary events were also held in conjunction with the meeting.

- Madeline Kuhn, founding member of HDSA's Council for Research, hosted a Research Advancement Council Dinner.
- A gala reception brought together members of the HDSA San Diego chapter with Coalition investigators.
- A video-conference, featuring renowned HDSA Coalition investigators, Gillian Bates, Ph.D. and Jang-Ho Cha, M.D., Ph.D., provided interested viewers in San Diego and Los Angeles with an exciting overview of HD research, as a part of a California educational program held in conjunction with the California Caregivers Alliance.
- The annual joint session of the Huntington's Disease Society of America and the Huntington Study Group (HSG) followed the Coalition meeting.

Research Progress

The HDSA Coalition for the Cure targets the most promising new research for further study. Investigators meet to discuss the latest unpublished work developing within their individual laboratories and to share ideas that may avoid costly duplication of effort. Each presentation provides valuable pieces to the HD puzzle that could result in promising new therapeutics and a cure.

Though each Coalition conference brings renewed enthusiasm and excitement to Coalition investigators, there are always one or two topics that generate passion and avid attention. During the Fall Coalition for the Cure meeting, much of the discussion centered on basic research with several investigators focusing upon the role that proteins play in Huntington's Disease.

Hope Through Research

Presentations included research targeted toward explaining

- how the huntingtin protein disrupts the arrangement and activity of specific genes
- how the use of chaperones and heat shock proteins* might help the huntingtin protein to fold properly
- how these molecular chaperones can be used to suppress protein aggregation (the accumulation of mutant huntingtin in cells and nucleus) using an animal model (c.elegans)

- why and how proteins misfold and how this process can lead to abnormal cell formations in an animal model (*Drosophila*)

This exciting expansion on basic research illustrates the different avenues taken by our Coalition for the Cure investigators. And while basic research adds to our understanding of the disease and its pathways, the HDSA Coalition Sub-Committee is also moving forward with a Therapeutics Initiative that will focus upon experiments that are therapy driven. Much of the work being done in Coalition labs today represents the next step in the road to a cure for HD.

HSG Research

The joint session of the Huntington's Disease Society of America and Huntington Study Group followed the Coalition for the Cure Research Conference. Each year, HDSA meets with the Huntington Study Group to discuss the best ways to move potential therapeutics from the lab into clinical trials.

Dr. Robert Ferrante's work on clinical trials in mouse models of HD was used to illustrate the kind of research that bridges the gap between the bench and the bedside. Dr. Ferrante, in his work entitled, *"Experimental Therapeutics of HD: From the Bench to the Clinic (and Back)"* shared data on clinical trials in mouse models that utilized five compounds, each with a potential for use by humans: creatine, Coenzyme-Q10 (CoQ10), remacemide, dichloroacetate (DCA) and cystamine. The use of cystamine in Dr. Ferrante's clinical trials in mouse models underscores the collaborative effort that is fostered by the HDSA Coalition for the Cure.

Creatine

The National Institutes of Health recently approved a clinical trial for creatine in humans though recruitment has not yet begun. Preliminary data has demonstrated that HD mice given creatine showed a 19% increase in survival. Degeneration in the striatum, the brain region most affected by HD, also decreased.

CoQ10 and Remacemide

CoQ10 and remacemide are currently being tested in a human clinical trial, conducted by the Huntington Study Group, called CARE-HD. While the results of the human trial will not be available until next year, the mouse studies have indicated that survival rates are enhanced 33%, with improved motor ability, when CoQ10 and remacemide are administered individually. The effect is greater when the two treatments are combined. The effect of these two drugs on the HD mouse models was the most promising therapeutic combination suggested to date.

Dichloroacetate (DCA)

DCA is a compound that may show therapeutic effects in mitochondrial diseases. In patients with HD, it is believed that the mitochondria (organelles which make the energy for the cell) do not function properly. Thus, a mouse trial testing DCA was developed. The results indicate a more modest increase in HD mouse survival (6-9%) than other compounds used in clinical trials listed above.

Other Research

Riluzole, another drug in human clinical trials (also being conducted by HSG), is currently being studied in this battery of mouse clinical trials. The results are not yet in. The Ferrante mouse clinical trials have also explored two other drugs, nicotinamide, and nicolic acid, neither of which demonstrated any therapeutic effects.

Funding the Future

When the HD gene was identified in 1993, after an exhausting 10-year search, we began the first step in the road to a cure for Huntington's Disease. Since 1993, we have moved further down the road of discovery and much has been added to our understanding about how the HD gene works, including what may trigger the onset of the disease and some pathways that might be blocked to halt disease progression. As we continue along the road to a cure and move from exclusively funding basic research into our new therapeutic directive and clinical trials arena, we ask for your help. As each year brings increased opportunities, it must also bring increased funding. Every dollar donated to HDSA research programs goes to fund the HDSA Coalition for the Cure, the Grant and Fellowship program as well as other research initiatives including the Huntington Study Group. Our HD families and HD researchers are counting on you to help us to *make this the last generation with HD.*

* See June 2000 *Toward a Cure* for Coalition article spotlighting chaperones and heat shock proteins.

Tracking Energy Requirements in People with HD

By Ann Gaba and Carol Boozer

How do you persuade 13 individuals with HD to spend a night sealed in an air-tight hospital room, where they are tracked by radar and then, climb into a giant plastic egg the next day? More importantly, why?

These 13 individuals were participants in a clinical research study about energy requirements of people with HD and these activities were actually precise measurements designed to reveal exactly how much each participant would need to eat to maintain their body weight. The results will be used to help clarify which factors, such as chorea, dystonia, or increased metabolic rate, contribute to the total calorie needs of individuals with HD.

The research project originated at the Columbia Health Sciences HDSA Center of Excellence at the New York State Psychiatric Institute. Ann Gaba, a nutritionist there since 1992, noted that many people with HD have trouble maintaining their weight. High calorie recipes and advice about eating "a lot," did not bring consistent results. Some individuals did well; others continued to lose weight. The usual formulas that dietitians use to calculate calorie needs didn't seem to work for those affected by HD.

Ann decided to pursue this question in the dissertation research for her doctorate in nutrition. With the help of her academic advisor, Dr. Carol Boozer, Ann arranged to use the high-tech metabolic measurement equipment at the Obesity Research Center at St. Luke's Hospital. An HDSA Initiative Grant provided the funding needed to support the project.

Recruitment for the study was done through the HD clinic at Columbia Presbyterian. Though some patients were initially concerned about the tasks required, they bravely agreed to get involved in the study.

Each participant spent a day in the Metabolic Chamber (also called the Human Respiratory Chamber) at St. Luke's. This small air-tight room contained a bed, table, TV and VCR, a toilet and a sink. All meals were passed in through a hatch and food intake was carefully monitored. The diet was not restricted and personal food preferences were accommodated as much as possible. Each participant had plenty of food. Their movements were tracked by radar, and also by a special motion-sensing floor. Measurements of oxygen and carbon dioxide, recorded by a computer, told the researchers how many calories each subject was using every two minutes. The following day, the participant was released from the special room and escorted to the Body Composition Lab where their percent of body fat was measured very precisely using air displacement by a piece of equipment called "the Bod-Pod."

Continued on page 29

CLINICAL RESEARCH IN PRESYMPTOMATIC INDIVIDUALS

PHAROS (**Prospective Huntington At-Risk Observational Study**) is seeking men and women between the ages of 30-55 years of age who are at-risk for HD and who have never undergone genetic testing. If you or a member of your family meets the eligibility requirements for **PHAROS**, please consider joining the investigators of the Huntington Study Group in this effort.

Most studies that focus on identifying a treatment for HD are conducted in individuals who already have symptoms, in hopes of halting, slowing or reversing the disease. Research is just beginning on at-risk individuals who are presymptomatic. These trials may reveal whether treatments for prevention are the same as or very different than treatments designed to delay the progression of illness. However, preventive therapies can only be tested on individuals at-risk for HD but not yet experiencing symptoms.

Crucial Questions

There are many issues to consider before testing possible treatments in at-risk individuals. What criteria should neurologists use to estimate onset of illness? Can a study be performed in this vulnerable population while protecting the confidentiality of their clinical and genetic information? And how many individuals have to be followed and for how long before it is known if a treatment works? These are some of the crucial questions that the **P**rospective **H**untington **A**t-**R**isk **O**bservational **S**tudy (**PHAROS**) is designed to answer.

Men in Clinical Trials

Since July of 1999, PHAROS has enrolled more than 450 subjects at 42 research centers. Curiously, enrolling men into the study is proving particularly challenging. During the first year and a half of study enrollment, more than twice as many women as men have signed up to participate in PHAROS. Since HD affects men and women equally, there should be just as many men as women eligible for participation. PHAROS investigators are eager to discover what factors affect male participation. Is it the time commitment, a heightened awareness of being at-risk for HD, a concern about confidentiality or some other unknown factor? Whatever the reasons, they must be overcome in order to achieve optimum study results.

Phase Two

The PHAROS study is now ready to move into a second phase of recruitment with the challenging goal of enrolling the full complement of 1000 subjects by November 2001. PHAROS is seeking men and women between the ages of 30-55 years of age who are at-risk for HD and who have never undergone genetic testing. If you or a member of your family meets the eligibility requirements for PHAROS, please consider joining the investigators of the Huntington Study Group in this effort. To learn more about PHAROS and view frequently asked questions, visit the Huntington Study Group (HSG) web site: www.huntington-study-group.org or phone the HSG toll-free number: 800-942-0424.

HDSA *thanks*

The American Legion Child
Welfare Foundation

for their support of
HDSA's Juvenile HD programs
and sponsorship of the Juvenile HD
section of ***The Marker***

Medicare Coverage of Clinical Trials

In September 2000, the Health Care Financing Administration released a decision that directed Medicare to cover routine patient care costs associated with clinical trials.

Many federally funded trials will be automatically covered under this decision. Other clinical trials must be certified for Medicare coverage.

Currently, a multi-agency panel of the Agency for Healthcare Research and Quality is determining the criteria for this certification.

HDSA *thanks*

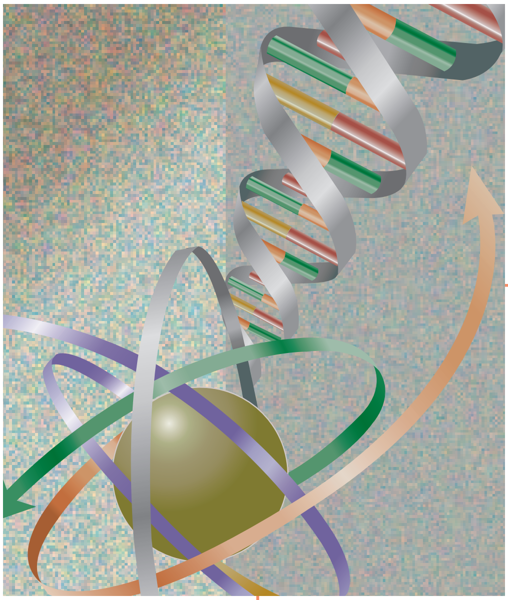
Kelly E. Miller
Juvenile HD
Memorial Fund

for its support

Living At-Risk

A LOOK AT GENETIC TESTING, PRIVACY AND DISCRIMINATION

For those living "at-risk," the decision of whether they should be tested for the HD gene often leads them to wonder how knowing will change their lives. Added to that is the concern that others, such as insurance companies and employers, may also obtain the results of the testing, perhaps without the "at-risk" individual's knowledge and approval. There is a fear that genetic test results will be misused to deny health benefits and employment. Fear of genetic discrimination is very real.



The Health Insurance Association of America (HIAA) maintains that there is no evidence indicating the existence of genetic discrimination. "None of our members requests applicants take genetic tests in order to get coverage (or set premiums), and none have plans to do so," said Richard Coorsh, a spokesperson for the HIAA. "It is against the law to discriminate against anybody with health coverage on the basis of any test, including a genetic test." But many genetic counselors and patient advocate groups believe that some level of discrimination by health insurers is already widespread.

> *"…many genetic counselors and patient advocate groups believe that some level of discrimination by health insurers is already widespread."*

The Department of Public Health Sciences, Wake Forest University School of Medicine, Winston-Salem, North Carolina sponsored a study on the issue, "Laws Restricting Health Insurers' Use of Genetic Information: Impact on Genetic Discrimination" by Mark A. Hall and Stephen S. Rich, which was published in the American Journal of Human Genetics in December 1999. Using data from both in-person interviews with insurers and a direct market test, they found that a person with a serious genetic condition who is presymptomatic faces little or no difficulty in obtaining health insurance. Whether those individuals would be charged prohibitively high premiums was not included in the study.

In the United Kingdom, the government has announced that insurers may use the results of genetic testing to refuse coverage or to set premiums for life insurance. They may also require individuals to reveal any family history of genetic disorders. Professor Durant, chairman of the Genetics and Insurance Committee, an advisory board to the Department for Health, stated that while no one could be asked to take a genetic test prior to getting life insurance coverage, they would be expected to reveal the results of any test already performed. Although the majority of people rely on the National Health Service for their medical insurance, the government's approval of the use of genetic information to set life insurance premiums may lead to other uses in the future.

The National Consumers' Council expressed their concern that this would lead to people to choose not to be tested. "If you don't know the information, you can't put it on the form," a spokeswoman said. Sue Watkin, chair of the Huntington's Disease Association, called on the government of the United Kingdom to establish a fund to provide insurance for people at risk of HD.

In December, after four years of work, the United States Congress directed outgoing president Bill Clinton to issue the first comprehensive federal standards to protect the privacy of medical records, by requiring doctors and hospitals to obtain consent from patients before disclosing health information. Patients now have a federal right to review and correct their medical records. If a request for access to medical records is denied, the patient may file a complaint with their health care provider or the federal government. These rules cover electronic records, paper records and even oral communications between health care providers and health plans. The rules, however, do not allow patients to sue for damages. The new standards will help to provide at-risk individuals with more control over access to the results of their genetic tests. The health insurance industry, though, has stated its concern regarding the cost and burden of implementing the standards and may lobby to have them modified.

While the question of genetic discrimination by insurers moves through the halls of Congress and the courts, there are things that those considering genetic testing can do.

- **If possible, get health insurance coverage before you are tested.** The Health Insurance Portability and Accountability Act (remember that HIPAA does not cover everyone) states that individuals can buy insurance no matter what their health status, but the premiums for persons with existing conditions may be higher. As the Wake Forest study found, presymptomatic individuals experience few problems finding insurance.

- **Don't lie.** Falsifying or withholding information can be grounds for an insurance company to deny coverage for a given condition or to cancel the insurance. Get your coverage, then get your tests.

- **Get counseling.** HDSA Centers of Excellence provide pre- and post-test counseling. HDSA also maintains a list of genetic testing centers, for those areas with no Centers of Excellence, that provide counseling before, during and after genetic testing. This is not the time to "go it alone." Benefit from their experience and resources.

- **Participate in research.** The only way that effective treatments will be found for HD is through clinical research trials, though there remain legitimate concerns as to the fair use and privacy of the genetic information gathered. Francis Collins, M.D., Ph.D., and Director of the National Human Genome Research Institute, feels that until appropriate policy protections are established, potential research participants should be informed of the state and federal protections currently in place regarding the collection, use and disclosure of genetic information in order to weigh the benefits and risks for themselves. HDSA encourages you to read the article on clinical trials found on page 20 or view the trials currently underway at http://www.clinicaltrials.gov. People who search this web site or call the HSG will not be contacted by sponsors of the trials or anyone else.

- **Become an HDSA advocate.** With the assistance of AstraZeneca Pharmaceuticals, HDSA has established a nationwide network of advocates to help educate public policy makers on issues of importance to the HD community, such as health insurance coverage issues and genetic privacy. See the form on page 33 for more information on becoming an HDSA advocate.

HDSA *thanks*

The **Bess Spiva Timmons Foundation,** a Founding Benefactor, for its sponsorship of *Caregivers' Link*, which includes the following articles this month: *At-Risk for Abuse in HD*, *Strategies for Speaking and Eating with HD*, and *Living with Grief and Loss*.

At-Risk for Abuse in HD

By Richard M. Dubinsky, M.D.

The physical and mental changes that occur during the progression of HD can make formerly independent people targets for abuse. Abuse of the person with HD can take many forms, ranging from verbal abuse to financial or sexual exploitation. Abuse can come from caregivers and family, as well as unscrupulous outsiders. Friends, family and caregivers all have a role to play in reducing the incidence of abuse and in reporting abuse when it occurs.

Verbal Abuse

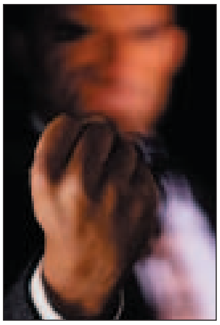
Verbal abuse occurs when the person with HD is constantly "put-down" for their actions or inactions. Comments may be as mild as "You're going to wear that again?" to statements as harsh as, "You're worthless. If I didn't take care of you, no one would." Social isolation is one of the most common methods used by an abusive person to keep their victim dependent upon them. The abuser may try to prevent the individual from having contact with other family members or with friends.

Verbal abuse can also occur in nursing homes. It may be due to a lack of knowledge about HD in the nursing home personnel. Family members must monitor the tone of the health care professionals who look after their loved ones. Report your concerns and any instances of verbal abuse to the nursing supervisor or administration.

Not all abuse is intentional. When a family member or caregiver is extended beyond their ability, they may react with anger, rather than love and assistance. Caregivers can become as socially isolated as the person with HD. Family therapy can address the underlying depression and frustration, of both the person with HD and the caregiver, that can lead to this kind of abuse.

Physical Abuse

Physical abuse occurs when a person is struck with the intention to cause harm. Black eyes, cut lips, cigarette burns or bruises can be evidence of physical abuse. Some abusers only strike where the bruises will not show, such as the stomach or back. Victims, feeling ashamed or without options, may try to cover up for their abusers, saying that they have fallen or stumbled. Vigilant friends and health care providers must trust their "gut feelings" and investigate possible cases of abuse. It may be best to take the person aside to a safe, quiet, and private place to ask them if they have been abused. Fear often keeps the abused person isolated.



Help by providing them with a line of communication to the outside world and with the phone numbers of local social service agencies and shelters for victims of abuse. Report suspected abuse to adult or child protective authorities. If in doubt, it is better to report suspected abuse than to possibly leave that person in peril.

Financial Exploitation

Financial exploitation of persons with HD is another form of abuse. Mental changes and dementia occur during the progression of the disease. As judgment and cognition become impaired, dishonest people may try to manipulate the person with HD into unreasonable purchases, investment schemes or donations. Steps can be taken to protect a person with HD from this kind of exploitation. These steps should be taken early in the progression of the disease, when the individual is not under duress.

- Appoint a financial guardian
- Cancel credit cards and ATM cards
- Arrange for direct deposit of disability payments, social security checks, etc.
- Establish a durable power of attorney for health care decisions (US)

Sexual Exploitation

Sexual changes in HD can present difficult questions. As HD progresses, there is a loss of social inhibition. The person with HD may develop new or non-traditional sexual interests that are startling or disturbing to their families. They may display inappropriate sexual behavior. Reduced social inhibition, combined with diminished judgment and dementia, puts people with HD at-risk for sexual abuse and exploitation. AIDS and other sexually transmitted diseases are also a concern. Unfortunately, there are no easy solutions. There is a fine line between allowing a person's natural sexual expression and keeping them safe from illness and exploitation.

Juveniles

Adolescents with Juvenile HD may be particularly susceptible to sexual promiscuity and aggressive behavior. Normal adolescent feelings, augmented by the reduced inhibitions that accompany HD, can make an explosive combination. A psychologist may suggest behavior modification strategies to reward appropriate behavior. Medications may also be prescribed to reduce sexual urges and aggression. A personal care attendant must be provided by the public school if the teen is judged to be especially vulnerable to sexual or physical advances. (See page 29 to learn more about your child's rights in school.)

Resources

Whatever the form of abuse, isolation helps the abusers. The more we stay connected and involved in the lives of people with HD, the fewer opportunities will exist for abuse. HD Clinics and support groups, HDSA chapters and helplines, and HDSA Centers of Excellence for Family Services are there to offer resources to assist HD families in protecting their loved ones from abuse. Local Departments of Health and Human Services can provide information about abuse and it is their duty to investigate the possibility of abuse. Therapy and support groups are available for caregivers, as are a full range of medical and mental health services to address any underlying problems that are opening the door to abuse.

About the author – Richard M. Dubinsky, M.D. is an Associate Professor of Neurology at the University of Kansas Medical Center. His article about Abuse originally appeared on the Huntington's Disease site he maintains at KUMC. To visit this valuable HD site, go to http://www.kumc.edu/hospital/huntingtons/discuss

Strategies for Speaking and Eating with HD

By Jeanne E. Thomson, M.S., CCC-SLP

People with HD face a progressive degeneration in their ability to speak, communicate, eat and swallow. Management strategies have been developed to help individuals with HD and their caregivers to maintain a higher quality of life during the progression of the disease.

Managing communication in HD

Communication difficulties are common in persons affected by Huntington's Disease and are a leading cause of depression. People with HD often experience changes in memory, judgment and problem-solving (cognition) as well as a breakdown or impairment in their ability to speak (dysarthia). While HD has a progressive, debilitating effect on a person's ability to communicate, there are techniques that assist and augment understanding. Your doctor may have additional suggestions or may recommend a skilled speech language pathologist who can perform a comprehensive language assessment.

Cognition Management Strategies

- Maintain a structured environment with established daily routines
- Use calendars and to-do lists
- Ask that information be kept concise and simple
- Use a journal to record feelings and perceptions
- Have important information written down
- Keep written information on new people and events

Difficulties with speech

People with Huntington's Disease often find it hard to make themselves understood. Slurring of the speech is a common problem. Coordinating breathing and speaking may become difficult due to sudden involuntary movements (tics). Unnatural hesitations while speaking as well as sudden changes in volume may occur. The rhythm and rate of speech are sometimes affected. These are a few management strategies to help patients and caregivers improve the quality of their communication.

Speaking Tips for the Person with HD

- Watch your listener for signs of misunderstanding
- Restate your message using different words (rephrasing)
- Consider using a picture, alphabet or phrase board
- Ask a speech therapist for speaking drills to help maintain your rate and rhythm of speech

Communicating Strategies for Caregivers

- Stop the affected individual and ask them to repeat themselves
- Suggest to the individual with HD that they rephrase their message
- Set up a reliable yes/no system or code

It is always a good idea to introduce and gradually begin to use a new communications tool or strategy before the person with HD actually needs it. In that way, the person with HD will be comfortable using the tool or strategy and will be able to continue to communicate.

Improving eating and swallowing in HD

Difficulty with eating and swallowing (dysphagia) is a common complication of HD. This creates not only an uncomfortable situation, but one that can be life-threatening. Food that is swallowed without adequate chewing; coughing caused by food entering the air passages; respiratory tics (hiccups) and air being swallowed all undermine the ability of a person with HD to eat and enjoy meals.

Paradoxically, a person with HD needs more calories to maintain a healthy weight than a person without HD. The challenge of maintaining adequate intake and safety during meals can be daunting. It is recommended that the person with HD or a family member/caregiver should consult with a licensed speech language pathologist for a full eating assessment. These are a few techniques to improve the mealtime experience.

Because of the danger of choking, all caregivers should know the Heimlich maneuver: a simple technique that dislodges food caught in the throat. Your local hospital, Red Cross or ambulance corps can tell you where to learn this life-saving technique.

If weight loss, coughing or choking become a constant problem for the person with HD, a feeding tube may be the best way to ensure adequate and safe food intake. It is important to discuss this before it becomes absolutely necessary, so that everyone involved understands the reasons for the change. Tube feeding does not prevent eating for pleasure, it merely ensures that the person with HD receives adequate nutrition. Nor is a feeding tube always a permanent solution. It can be used to help manage an eating, hydration or weight loss crisis until the person with HD is stable and able to again eat by mouth. The goal of all HD management strategies is to increase the quality of life for the person facing HD.

EATING TIPS FOR THE PERSON WITH HD

- Slow down the pace of eating. Put the utensil down between mouthfuls.
- Use a "commuter" cup with a lid or a straw to limit the amount of liquid to reduce the chance of choking.
- Try weighted dishes and plates with higher edges to make eating easier.
- Select utensils with bigger, softer handles, like those designed for people with arthritis.

MEALTIME STRATEGIES FOR CAREGIVERS

- Include more fat and calories to maintain weight during the early and middle stages of the illness.
- Chop foods to compensate for lack of chewing.
- Reduce the use of liquids to "wash down" foods. Try purees, such as applesauce.

About the author – Jeann
Thomson, M.S., CCC-SLP
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Hospital in NYC.

Living with Grief and Loss

—Barb Heiman, LISW

> *"To live in this world you must be able to do three things: to love what is mortal; to hold it against your own bones knowing your own life depends on it; and when the time comes to let it go, to let it go."   - Unknown*

What is this thing called grief?

Grief is a normal, healthy human response to any important change or loss in our life. It is "love" not wanting to let go. Throughout life we grieve; an infant grieves the loss of his bottle or his mother's breast; a child grieves the security of his home when he goes off to school. Every major change in our life involves "letting go" of the familiar to step into the unfamiliar. All along life's way we are confronted with losses. Each loss brings with it a time to grieve.

How can I stop feeling this way?

We have all heard the phrase "time heals", but in reality, time does nothing more than "pass." It is what we do with that time that matters in our recovery. We may call upon our friends, family, or co-workers; put our faith in God or a higher power; join a grief support group; get on-line with a grief-related Internet "chat room;" or draw on our own inner strength and courage. What is important is that grief be expressed. Repressing these feelings will only prolong the process. If you let yourself feel bad now, you will feel better sooner.

My friends tell me to "get over it"

As harsh as it seems, the world does not stop while we grieve. Even our friends and families may assume that we will be "over it" in three to six months. Their expectations may have more to do with their discomfort regarding loss, than with the level of our grief. The truth is that grief is a process. We have the right to be as sad as we are for as long as we need to be. When faced with a significant loss, that can mean two to five years.

Why do some people seem to cope with loss better than others?

No two losses are the same, so there is no way to "compare" grief. Our personal grief experience will depend on many things: What is my loss history (Is this my first or tenth loss?) When, where and how did I suffer this loss? Are there other stresses in my life, such as conflicted relationships or the loss of a job? Was this a sudden loss or the result of a prolonged situation? All of these factors make a difference in how a person comes to terms with that loss.

Where can I find help and hope?

People facing loss related to HD are confronted with complex grief issues over a long period of time. As a member of an HD family, one may face the same challenges over and over again. But you are not alone. HDSA is here to ensure that you do not have to face your grief alone. The Huntington's Disease Society of America consists of volunteer-based chapters, affiliates and support groups across the United States. They are there to provide you with support and resources to help you through the challenge of HD. Their trained social workers can work with you and your family to cope with your grief. In addition, HDSA Centers of Excellence also provide trained social workers and support groups and, because each Center of Excellence is a major medical facility, many will have grief counseling available.

By continually increasing our financial commitment to HD research, the Huntington's Disease Society of America remembers those who have lost their battle with HD and recognizes the families that carry the hope that together, we can make this the last generation with HD.

About the author –

Barb Heiman is a Licensed Independent Social Worker with more than 18 years experience working with the terminally ill and their families. She received her Master's degree from Ohio State University and certification as a bereavement counselor from the American Academy of Bereavement. She is currently the Social Worker with the HDSA Central Ohio Chapter.

NEW RESEARCH—continued from page 19

The research may determine if patients with HD use more calories because the HD gene alters cellular metabolism and makes it less efficient, so that the body needs extra calories to maintain weight. This result would point to a need for new ways to improve cellular metabolic processes. Alternately, the research may reveal that HD patients need extra calories simply because they get more muscle exercise due to chorea and dystonia. If this is the case, then nutritional advice may be useful to help patients maintain their weight.

Once completed, the research will be submitted for scientific publication. The results will appear in a future issue of *The Marker* magazine, so that our HD families and caregivers may have this important information.

About the Authors: Ann Gaba MS, RD, is a Clinical Nutritionist at Columbia Presbyterian Medical Center and the HDSA Center of Excellence at the New York State Psychiatric Institute. She is also a doctoral student at Teachers College, Columbia University.

Carol Boozer, D.Sc., is on the faculty of the Institute of Human Nutrition at Columbia University and directs the Energy Metabolism Core Laboratory at the Obesity Research Center at St. Luke's - Roosevelt Hospital Center. Her research includes basic and clinical studies on aspects of nutrition and physical activity that affect the regulation of energy balance.

HDSA *thanks*

The American
Contract Bridge League
Charity Foundation
for their support of
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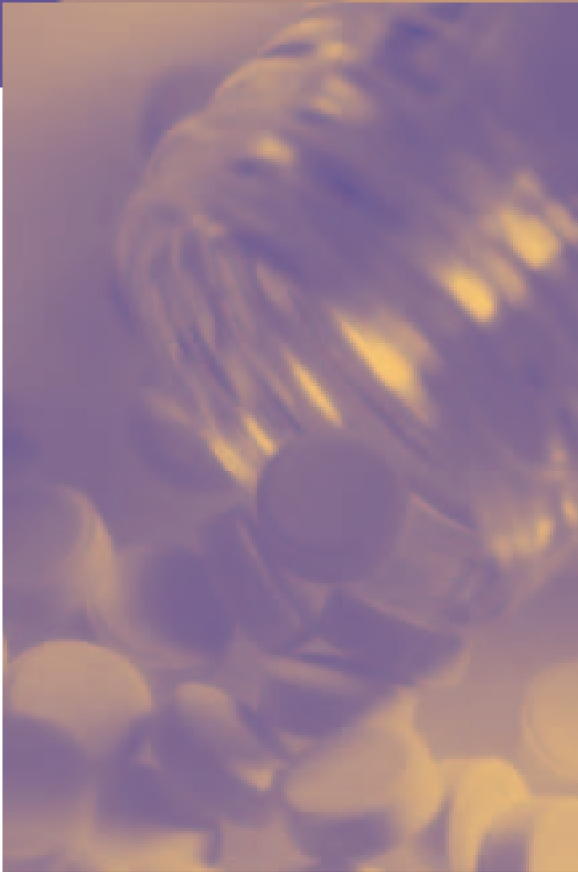
DIAGNOSIS AND TREATMENT OF SEIZURES IN JUVENILE HD

New Drugs for the New Millennium



While seizures are uncommon among adults with HD, approximately 25-30% of children with juvenile onset HD will have seizures at some time during the course of their illness. Some will develop an ongoing risk of having seizures, a condition also called "epilepsy." Tests have been developed to help diagnose seizures in juvenile onset HD and several new medications seem to help some children. Learning more about seizures may help the patient and family manage these unexpected events.

Causes

Seizures can be caused by metabolic abnormalities, such as a low blood sugar or an abnormal sodium or calcium level; by ingestion of toxins such as cocaine; or as a side effect of medications, including antidepressants, stimulant drugs such as Ritalin, and neuroleptic drugs. Seizures can also be caused by structural abnormalities in the brain, such as brain tumors, scars from prior brain injuries, or areas where the brain was formed abnormally. All children who experience a first seizure should be brought to a hospital Emergency Room immediately.

Diagnosis

The tendency to have seizures, sometimes runs in families and can be unrelated to HD. Experts estimate that between 5-10% of all people experience a seizure at some time in their life! Physicians attending a patient who has had a seizure must take a family history, analyze the blood for toxins or chemical abnormalities, perform a brain imaging procedure (CT or MRI), and an electroencephalogram (EEG), before deciding what has caused the seizure and what the appropriate course of treatment should be. Tests such as an EEG can help a doctor determine if there is an ongoing risk of additional seizures.

Often, even after all the appropriate tests have been performed, there may be no definitive proof as to what caused the seizure or group of seizures, and the physician will have to make a judgment about the cause. While no test can prove that HD caused the seizure, it can rule out other possible causes.

Treatment Options

Although a single seizure is not usually dangerous to the child, a series or flurry of seizures may not stop without medication, and left untreated can lead to tongue lacerations, breathing difficulties, irregular heartbeats, and other more serious complications. A single seizure may not require treatment with antiepileptic medication, but if a person with HD has had more than one unprovoked seizure, treatment to prevent further seizures will probably be necessary.

The "first-line" drugs used to treat seizures include phenytoin (Dilantin), carbamazepine (Tegretol), and valproic acid (Depakote/Depakene). These drugs are familiar to most physicians and to all neurologists, and are effective for most people with seizures, though an occasional child may continue to have seizures. Researchers have been hard at work developing new seizure medications, and within the last five years, at least seven new seizure drugs have been released in the United States. These include: gabapentin (Neurontin), lamotrigine (Lamictal), levetiracetam (Keppra), oxcarbazepine (Trileptal), tiagabine (Gabitril), topiramate (Topamax), and zonisamide (Zonegran).

Because these drugs are new, your physician and neurologist may not be familiar with them. If a child is not responding well to medication, you may wish to visit a neurologist who sub-specializes in pediatric epilepsy in order to determine if one of the newer medications will be helpful in managing the child's seizures. Contact the HDSA National Office for a referral to a specialist in your region.

About the author –

Martha Nance, M.D., is the director of the HDSA Center of Excellence at Hennepin County Medical Center. She is an HDSA Scientific Editorial Board Member.

Education & Juvenile HD

BEING YOUR CHILD'S ADVOCATE

While most parents are involved in their child's education through parent-teacher conferences and perhaps the PTA or after school activities, the parents of a child diagnosed with juvenile-onset HD face a different level of participation. They must become their child's front-line advocate in the school system.*

Kids with HD can continue to learn and interact with their peers during the early stages of the disease. As the parents of a child with HD, you can help make that happen. A determined parent, working closely with teachers and school administrators, can make a tremendous difference in the amount of help the school system will provide to a child with HD.

To ensure that your child receives all the benefits and services available, you should speak with the Director of Special Education in your school district or with your school principal, even before your child with HD needs special assistance in school. They will have copies of the federal, state, and local guidelines that govern the education of students with disabilities. These guidelines describe what the school must do to help your child and exactly what you have to do to request those services.

School is an important part of a child's life. Your determination and advocacy will help your child with HD to stay in school and continue to learn for as long as is possible.

You may find that you need to educate teachers and administrators about the physical and mental changes that occur as HD progresses. For example, if they are advised that emotional outbursts and lowered inhibition may be displayed by a child with HD, they are less likely to think that the child is simply "acting up."

As the disease progresses, placing your child in a special education class may prove beneficial. Special Education teachers are skilled at developing Individualized Education Plans (IEP) for students with challenges. They can organize information so that the child with HD can understand. They are also better prepared than most classroom teachers to deal with emotional or erratic behavior. If home schooling becomes necessary, they may provide appropriate learning activities.

Once your school has been alerted to the special needs of your child with HD, be sure to keep them up to date on physical and mental changes that you observe in your child. The Special Education department may only assess the needs of special students on a semester or yearly basis, rather than on a monthly basis as may be required for a child with HD.

While a good relationship with the school administration will help to ensure that your child's needs are met, it is possible that you and the school will not always agree upon what is best for your child. When the school and family cannot agree on the needs, placement, or program of a student, both parties have the right to request a due process hearing with their State Education Department to resolve the problem. You and your child have rights under the law.

Social workers are available, through HDSA and the HDSA Centers of Excellence for Family Services, who can guide you as you work with your school system. HDSA can also provide information and resources for parents of children with HD, including a comprehensive Handbook about Juvenile HD. For referrals or more information, contact HDSA at 1-800-345-HDSA.

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* The term "parent" is used in this article to include foster parents, legal guardians, or any primary caregiver who is functioning as a parent.

WAYS YOU CAN MAKE A DIFFERENCE

Your gifts to HDSA enable us to fulfill our mission to fund vital research while providing support and services to those affected by HD and their families. Through the HDSA Center of Excellence program, our 31 volunteer based chapters, 3 affiliates and numerous support groups, HDSA offers our HD families the very best in medical services and referrals to existing local resources. HDSA research initiatives fund the most promising projects that will lead to effective treatments and, ultimately, a cure. Your contributions also help the Society to increase public awareness and educate health care professionals about this devastating disease. Your continued support is most urgently needed as we build toward a future free of Huntington's Disease.

Generation 2000: Fulfilling the Promise

Generation 2000 - Making This the Last Generation with HD came to a close on December 31, 2000. Through your generous support, over $2.7 million was raised for HD research. We sincerely thank every contributor, large and small, who helped to make our dream a reality. A special thank you to the Vaughan Family, the Smith Family, the Dobis Family and an anonymous donor who provided *Generation 2000* with $1 million in matching funds.

We now stand on the threshold of a new campaign: *Generation 2000 : Fulfilling the Promise*. Building upon the incredible momentum created by our original 18-month campaign, the Society has now set a goal to raise **$25 million** for HD research in the next five years. Our scientists tell us that each day we grow closer to solving the HD puzzle. And while our scientists cannot yet predict which avenue of HD research will fulfill the promise, each of us can do our part by dramatically increasing the funding available for HD research.

Matching Gift Fund for Research

Once again, two HD families have come forward to pledge their commitment to make this the last generation with HD by agreeing to match new contributions to HD research. By the time this article is printed, we hope that two more families will have stepped forward to join with our two other benefactors to establish a $2 million Matching Gift Fund that will propel this campaign forward toward its goal of raising $25 million for HD research by 2006. HDSA will announce our generous Matching Gift Fund benefactors in the May issue of *Toward a Cure*.

Whether your donation is large or small, whether you give through a Family Fund, a Tribute/Memorial, a bequest or with your corporation's Matching Gift program, every dollar raised in *Generation 2000: Fulfilling the Promise* WILL be matched by this new matching gift fund. If you would like to help HDSA fulfill the promise to our future generations, please send your contribution today or contact the National Office for other ways that you can help.

HDSA Family Funds

A Family Fund gives family members and friends an opportunity to contribute a larger gift than one might be able to individually. Family Funds can recognize

All Family Funds, cash gifts, pledges, tributes, memorials or bequests established or renewed in 2001 will be matched through the

Generation 2000: Fulfilling the Promise

Matching Gift Fund.

TUNITIES

the commitment of an entire family to HDSA or they can create a proactive remembrance of a loved one. With an annual gift of $10,000 or more, each Family Fund is invited to co-sponsor a research project or any of HDSA's educational or family service programs. To create your Family Fund, contact the National Office today.

Cash and Pledges

Charitable cash gifts or pledges provide our benefactors with significant tax benefits. Pledges made to HDSA may be paid over a period of three years or longer.

Tribute Gifts

Tribute and memorial gifts honor or remember a loved one while supporting the goals of HDSA. A personal acknowledgement is sent to the individual or family in whose honor a gift is made and a separate receipt is sent to the donor for tax purposes. Tributes and memorials may also be made through the national web site at www.hdsa.org (click on Making a Difference.)

Stock, Securities or Real Estate

Gifts of stock, securities or real estate provide tax benefits by avoiding all capital gain taxes and allowing the donor to claim the current market value of the gift (not the purchase price.)

Heritage Club -

• Individual Planned Giving

Remember HDSA in your estate plans by joining the Heritage Club. Use your will, trust, or estate assets to make a contribution to HDSA while receiving valuable tax benefits. Below are types of planned giving that may be considered.

• Bequests

Remember HDSA in your will.

• Gifts of Personal Residence or Farms with a Retained Life Estate

• Life Income Gifts - Charitable Gift Annuity, Pooled Income Fund or Charitable Remainder Trust.

• Gifts of Insurance

HDSA's planned giving experts can work with your tax or legal advisor to create your planned gift.

Corporate Matching Gift Fund

To double the value of your gift, simply enclose your company's matching gift form with your gift to HDSA. To quadruple your gift, be sure to specify that both your gift and your employer's matching gift are included in HDSA's Matching Gift Fund for *Generation 2000: Fulfilling the Promise.*

United Way/Combined Federal Campaign

If you give through United Way or Combined Federal Campaign at your work place, be sure to identify Huntington's Disease Society of America (HDSA) as your charity of choice.

WAYS YOU CAN SUPPORT HDSA ALL YEAR LONG

Sending **flowers**? Use HDSA's new affiliation with FTD that donates 15% of your purchase to support HDSA's programs. It's easy to use. Simply call 800 SEND-FTD and be sure to mention **code 3015** (so HDSA receives the donation) or order through the national web site at www.hdsa.org

If you like to **shop on-line** then the HDSA Marketplace is for you! It offers great discounts and donates a percentage of your purchases from vendors like L.L. Bean, Land's End, The Sharper Image and more to HDSA! And, HDSA receives a $2 donation the first time you register on the site. To access the HDSA Marketplace, just click the button at the bottom of the HDSA homepage at www.hdsa.org.

Save money on your monthly **Internet** connection and support HDSA through AOL/CompuServe's new service for non-profits. If you already have AOL, then switching over to the HDSA program is easy. You can keep your screen name(s) and save $2 each month on your connection. If you're new to AOL/CompuServe, then loading the program and registering your screen name(s) is easy using the CD Rom provided in the info packet. For more information, contact HDSA today!

Donate your **used vehicle** to benefit HDSA. It's easy to do, Just make one phone call to (800) 562-7919 and your car will be removed free of charge. All paperwork will be done for you and best of all, you'll receive a tax deduction. Call today to find out how your used car can help the cure.

HDSA 2001 Annual Convention

The HDSA 2001 Annual Convention will be held at the Town and Country Resort in San Diego, CA (Mission Valley) from Friday, June 29 through Sunday, July 1. With a theme that underscores HDSA's commitment to make this the last generation with HD, "Fulfilling the Promise" defines the goal of the Society as we enter this new millennium.

The HDSA Annual Convention renews your connection to the larger HD community while keeping you up to date with the most recent research. The convention offers opportunities to expand your knowledge with interesting and informative forums on current Research and on Family Services. You can take part in workshops and panels including Learning about HD, Family Services and Care, Family Issues and more, and participate in sharing and networking sessions for all members of the HD family and their care givers. The social highlight of the convention is the gala Awards Dinner where we honor those who make a difference in our lives. All these events come together to create an unforgettable convention experience. For more information about the 2001 Annual Convention programs, please contact the National Office at (800) 345-HDSA, extension 35. A convention registration form is also conveniently provided for your use in this magazine. Please note the special Early Bird registration discount.

HDSA 2001 Annual Convention pricing

Convention Registration (per person)

* Early bird special (postmarked by April 30)	$130
* Full registration (postmarked May 1 - June 22)	$150
* Late or on-site registration (after June 22)	$180
* One day only (Friday or Saturday)	$ 80
* Saturday night dinner	$ 65

Town & Country Hotel Reservations

Hotel reservations are to be made directly with the hotel by calling (800) 772-8527. Please be sure to tell them that you are attending the HDSA Annual Convention.

Hotel pricing

Garden accommodations -
$105 (single or double occupancy) $125 (triple) $145 (quadruple occupancy)
Regency Tower/Courtyard -
$125 (single or double occupancy) $145 (triple) $165 (quad occupancy)
All accommodations are subject to occupancy tax.

Airline Reservations

Certified Travel can assist you with airline reservations
(800) 237-1517, ext. 330.

Scholarship Fund

Through the generosity of registrants and donors, HDSA is able to offer a limited number of scholarships to individuals and families who might not otherwise be able to attend the Convention. Help us to expand our scholarship opportunities by including a scholarship donation with your registration.

Cancellation policy: Cancellations of convention registration that are received before May 18 will be refunded in full. Those received on or after May 19 but before June 8 will receive a 50% refund. Cancellations received on or after June 9 are non-refundable.
All cancellations must be received in writing by mail or fax.

Celebration of Hope **Events**

Spotlight on Celebration of Hope

Celebration of Hope events are held annually across the country in support of the HDSA Center of Excellence program. These gala events bring together the community served by a Center or potential Center and demonstrate the positive effects an innovative program can have on a region.

Celebration of Hope Dinners recognize outstanding community leaders in the fields of business, medicine and philanthropy. On May 24, 2001 HDSA will host a "Celebration of Hope" Dinner to benefit the New England HDSA Center of Excellence at Massachusetts General Hospital & Tewksbury Hospital that will be held at the Westin Copley in downtown Boston, MA

The dinner will pay tribute to Nicholas A. Lopardo, Chairman and CEO of State Street Global Advisors (SSgA), Dr. Edward Bird, pioneer in HD research and Director Emeritus Harvard Brain Tissue Resource Center and Ron Burton, Executive Consultant for John Hancock Mutual Life Insurance and Executive Director of the Ron Burton Training Village. A special HD Family Award will be presented to the Boothbie Family. HDSA is privileged to have Congressman Barney Frank as the Honorary Chair of the event.

For more information about the Celebration of Hope Dinner in Boston, please contact the National Office at 800-345-HDSA, extension 17.

The following cities will also host Celebration of Hope events. To volunteer, attend or support any of these events, please contact the National office. Full coverage of all Celebration of Hope events will appear in the August *Marker* magazine.

April 16, 2001: Iowa City, Iowa
Iowa Memorial Union Center, University of Iowa

April 24, 2001: Baltimore, MD
Hyatt Regency Baltimore

May 9, 2001: Atlanta Georgia

May 24, 2001: Boston MA
Westin Copley Downtown Boston

June 21, 2001: Denver, Co

June 2001: Seattle, WA

September 12, 2001: San Diego, CA

September 13, 2001: Orange County, CA
Hyatt Newporter

September 21, 2001: Columbus, OH

September 2001: St. Louis, MO
Hyatt Regency Convention Center

September 2001: Houston, Texas

October 3, 2001: St. Paul, MN

Become an HDSA Advocate!

Your voice can make a difference in Congress! With the assistance of AstraZeneca Pharmaceuticals, HDSA has established a nationwide network of advocates who will help educate public policy makers about issues of importance to the HD community, such as NIH funding for HD research, health insurance coverage issues, and genetic privacy and discrimination. This year's goal: an HDSA Advocate in every congressional district in the country!

If you would like to know more about becoming an HDSA Advocate, please complete and return the form below or contact Mike Lamma, Director of Field Development at (212) 242-1968, ext. 12 or at mlamma@hdsa.org.

Your Name ____________________

Mailing Address ____________________

City ____________ State ____________ Zip ____________

Evening Phone ____________ Email ____________

Be sure to include your e-mail so that we can alert you to important bills in Congress.

Mike Lamma, HDSA, 158 W. 29th Street, 7th Floor, New York, New York 10001-5300

WEB-BASED ADVOCACY FOR THE HD COMMUNITY

HDSA advocacy efforts have focused upon educating and informing elected officials, community leaders and the general public about Huntington's Disease, with the goal of influencing legislation that may have a direct impact upon HD families. To broaden the base of our advocacy efforts, HDSA is introducing a new online web based advocacy tool, known as CapWeb.

CapWeb is an easy way to contact your elected officials in Congress. It provides an electronic directory of all House and Senate members (contact information, district office locations, key staff members, etc) with direct e-mail links. HDSA also offers Legislative Alert Posting, which provides additional background information about important legislative issues, such as patient's rights, genetic discrimination, Medicare funding for clinical trials, HD research funding and caregiver issues.

CapWeb

Using CapWeb

You can link to CapWeb through HDSA's national web site at www.hdsa.org. Once in the CapWeb system, you can speak out on legislation that may impact our HD community by writing your own letter or by using the letters created by HDSA. You may either print out your letter and mail or fax it to your representative or you may email it directly through CapWeb. Your letters will go directly to your elected representatives or even the President of the United States. And, when a letter is e-mailed, CapWeb tallies it, thus allowing HDSA to track the volume of e-mailed letters sent on its behalf. HDSA can then provide updated information to the advocates who contacted Congress, via email or the national web site, in the form of Advocacy Bulletins. CapWeb is user-friendly and allows supporters of HDSA and HD research to speak out in ever-greater numbers. Be sure to visit CapWeb today!

Huntington's Disease won't kill you right away.

It takes its time destroying your mind and body.

~~Fir~~ First to go is motor control. Simple things like writing become difficult.

Little by little you start to twitch uncontrollably. It continues destroying brain cells, as well as your memory, ability to think, [illegible] speak, and even swallow. Unfortunately, ~~to~~the worst is yet to come.

There is currently no cure for the hereditary brain disorder known as Huntington's Disease. But we're closer than ever to finding one. Your contribution could help stop the suffering of thousands. Please make your gift today and help HD research to continue.

You can double your gift today by indicating that you want it to be matched by HDSA's Matching Gift Fund for *Generation 2000: Fulfilling the Promise*. Together, we CAN *make this the last generation with HD!* Visit our website at www.hdsa.org or call **1-800-345-HDSA.**

Huntington's Disease Society of America
158 West 29th Street, 7th Floor, New York, New York 10001-5300

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