What is the ROSTER?

The National Research Roster for Huntington Disease Patients and Families is a unique nationwide information resource dedicated to assisting scientific research on Huntington disease. A goal of the Roster is to help researchers learn more about HD. To accomplish this goal, the Roster has involved nearly 3,000 families in HD research.

The Roster computerizes the names of families, including information about the history of HD in the family (family trees) and other related data. This information identifies HD patients and families who are interested in participating in research projects.

Scientists interested in studying HD may submit two types of requests for information contained in the Roster. The two types of information requested are anonymous information and identifiable information. Anonymous information is de-identified, meaning that names and personal identifiers are removed and can be given to researchers without having to contact Roster families. Identifiable information includes data such as names, dates of birth, and family structure. Identifiable information can be used to help researchers find volunteers who are willing to participate in HD research projects nationwide. In any instance where identifiable information may be given to a researcher, the Roster will contact participants to ask if they are willing to share their identifiable information for a research project.

Participation in the Roster is voluntary. You do not have to participate if you are not interested. All information received by the Roster remains completely confidential. No information about a member will be released without written permission from that individual. You are under no obligation to share your identifiable information with researchers or volunteer for any research project.

We would like to thank the many families who have participated in the Roster. We believe the Roster is a valuable scientific resource which may one day assist in the discovery of a cure for HD. We would like to extend the invitation to join the Roster to you and your family. We are always eager to accept new participants!

The National Research Roster for Huntington Disease Patients and Families

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Volume 1 • Fall / Winter 2004

REACH

Newsletter for the National Research Roster for Huntington Disease Patients and Families

The Roster and the NIH:
A Quarter-Century Partnership!

BY PATTI WOLF, RN BSN

October marks the 25 year anniversary of the National Research Roster for Huntington Disease Patients and Families, sponsored by the National Institutes of Health (NIH). The Roster was established at Indiana University in response to a recommendation from the United States Congress (Commission for the Control of Huntington Disease and its Consequences) in order to develop a program for wide-spread research on HD. The National Institute of Neurological Disorders and Stroke, a division of the NIH, agreed to fund the Roster on October 1, 1979. Over the last 25 years, the Roster has collected valuable information from over 2,700 families, consisting of nearly 135,000 individuals world-wide. With this information, researchers using the Roster have been able to learn more about the genetics, symptoms, and treatment of HD.

Throughout this newsletter, you will be learning about scientists who have used information from the Roster in their research projects. You will also be learning about the outcomes of that research, and what those outcomes have meant for the HD community. With continued enrollment of new families and steady support from scientists, the Roster will remain a source of vital information for HD research for many years to come.

NATIONAL INSTITUTES OF HEALTH

NATIONAL INSTITUTE OF NEUROLOGICAL DISORDERS AND STROKE

INDIANENSIS UNIVERSITATIS SIGILLUM ET VERITAS MDCCCXX

Coordinator's Corner

BY: PATTI WOLF, RN BSN

Exciting changes are occurring with the Roster!! These changes will offer many benefits to existing Roster families, as well as potential new families.

1) NEW INFORMATION MATERIALS! Our letters, booklets, hand-outs, and pamphlets have been completely rewritten! The new materials are user-friendly and provide easy to follow, step-by-step instructions to guide you through Roster participation. Also, we have new up-to-date resources available to answer your questions about Huntington disease symptoms and nationwide support groups.

2) NEW BROCHURE! A full-color brochure outlining the purpose, history, and recruitment process for the Roster is now available! Please contact us if you would like copies.

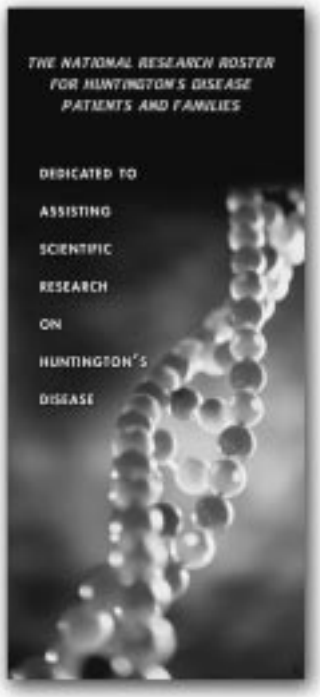
3) NEW NEWSLETTER! Members of the Roster can now look forward to this newsletter, sent out twice a year, in September and March. The newsletter will contain information on new research projects involving the Roster, as well as updates on research progress from previous studies involving Roster families.

4) NEW WEBSITE! Please take a moment during your web browsing to stop by the new home for the Roster at http://hdroster.iu.edu

The new website contains a wealth of information about family participation, researcher utilization, and general HD facts. There is also a special section highlighting the most recent findings from five very important HD research projects that have recruited hundreds of Roster families for study participation. Even more information is just a click away on our links page, which connects users to several reliable and knowledgeable websites devoted to HD research and support. Gain greater knowledge about HD using the Roster on the web!! Visit us today at **http://hdroster.iu.edu**

We are pleased to offer these improvements to the National Research Roster for Huntington Disease Patients and Families. We are very excited to be more user-friendly and accessible to our existing families and those individuals interested in becoming new participants. We welcome any questions, concerns, and requests that you may have. Feel free to contact the Roster at (317) 274-5744 or email the coordinator: pswolf@iupui.edu

Patti Wolf joined the Roster team as the new research coordinator on December 1, 2003. She is a Registered Nurse with over 3 years of clinical research experience. As the coordinator for the Roster, Patti is responsible for recruiting families and scientists interested in HD research opportunities. She is also an active participant in HD Clinic at the Indiana University Medical Center, as well as Indiana HDSA support group meetings and events.



A full-color brochure outlining the purpose, history, and recruitment process for the Roster is now available.

SCIENTISTS USING THE ROSTER: CURRENT STUDIES - WHAT IS THE LATEST HD RESEARCH?

Longitudinal Studies Among At-Risk HD Gene Carriers

BY: TATIANA FOROUD, PH. D.

Over a decade ago, researchers at Indiana University undertook one of the largest studies of its kind to better understand the early clinical findings of Huntington Disease (HD). The study was entitled Huntington Disease: A Neurological Marker of Aging. Beginning in 1989 and during the next 6 years, 657 individuals at-risk for HD came to Indiana University for the one day study. Study participants completed many different tests including a detailed neurological examination, computerized tests to evaluate their speed of response to lights and sounds, questionnaires asking about their mood and feelings as well as measurements of their ability to provide and recall information.

A number of important findings were reported from the analysis of this significant study. We found that early, subtle changes in thinking (cognition) may occur even before the onset of definite clinical symptoms of HD. For example, individuals who have inherited an expanded number of CAG repeats and therefore are classified as HD gene carriers, but were not yet diagnosed as having HD, on average, had worse performance on several tests of cognition (thinking/information recall). In addition, these same individuals who were asymptomatic HD gene carriers also had, on average, slower reaction times and slower speed of movement of their hands and fingers.

Another important result of the study was found after careful examination of the eye movements of all study participants during the neurological examination. One particular area of focus was saccadic eye movements. Saccadic eye movements are observed when we use our eyes to view a moving object. Our eyes typically don't move smoothly when tracking an object; rather, they make little jumps from place to place. These jerky motions are called saccades. In our study, we found that even among those individuals with minimal evidence of chorea, whose symptoms would be too mild to make a diagnosis of HD, there often were some abnormal saccades. These abnormalities included the accuracy of the little jumps, the speed with which the saccades moved, the length of time it takes the eye to start a saccade, etc.

It is important to stress that all the findings from our study report trends in the data. The abnormalities which have been observed are all very mild and are significant because we have included so many individuals in the study. As we know, there is great variation in the onset of symptoms of HD. Therefore, results from our studies are meant to point us toward areas of the brain affected by the disease; however, the onset of symptoms can be different in each individual. Results of our study should not be interpreted as the pattern of disease onset for each individual with HD.

Based on the results of our first study, we decided that it would be important to carefully document when these changes in thinking, reaction time and saccades occurred as individuals entered the early stages of disease. Understanding when these abnormalities begin to occur is important, since this may identify factors that allow physicians to establish an earlier diagnosis of HD. In addition, these early changes may prove to be very important disease predictors that could be significant in future clinical trials of new HD medications.

Continued on page 4.

Dr. Tatiana Foroud

is an Associate Professor for the Department of Medical and Molecular Genetics at Indiana University School of Medicine. Her research interests include the study of neurodegenerative diseases such as Huntington disease, Parkinson's disease and Alzheimer's disease. She is the principal investigator of an ongoing NIH study that seeks to better understand the early clinical symptoms of Huntington disease. She is also the Chair of the Patient and Family Services Committee for the Indiana Chapter of the Huntington Disease Society of America.

Highlighting HD Help: Water Exercise Therapy

BY: PATTI WOLF, RN BSN

Exercise is recommended for Huntington patients to help retain muscle mass and body weight, as well as improve quality of life. However, balance problems and incoordination make exercising difficult. Depending on the style chosen, exercise may even place an individual at greater risk for falls. A new exercise alternative for HD patients experiencing balance and in-coordination symptoms is water exercise therapy.

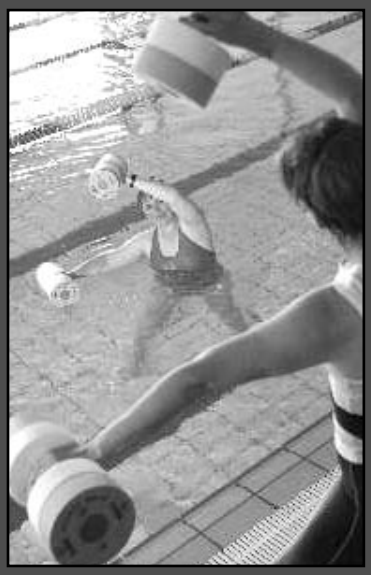
Why exercise in the water? Water exercise therapy offers a chief benefit not available in any land-based exercise style: security against falling and injury. If an individual does lose their balance in water, the water buoys the body and prevents a hard-surface fall. By being able to exercise in a safe environment, participants feel less hindered and will challenge themselves more because they do not fear falling and hurting themselves.

Water exercise is just as effective as many land-based exercises in improving muscle strength and coordination. Whether you are old or young, fit or not so fit, exercising in the water may help you to manage and maintain a healthy fitness regimen.

Contact your local YMCA or community health center for more information on available water exercise classes. Remember to contact your healthcare provider before beginning any new exercise regimen to determine if water exercise therapy is right for you.

To find the YMCA nearest you, visit www.ymca.net

Enter your zip code and click on the "Find Locations" button. The website will provide information about your nearest YMCA branch, including their address, phone numbers, programs offered, and hours of operation.



DID YOU KNOW?

Water exercise allows you to target specific muscle groups to tone your body, train for good posture and body movement, move in all directions for an aerobic-style workout, and stretch through greater ranges of motion.

Continued from page 3.

Longitudinal Studies Among At-Risk HD Gene Carriers

About two years ago, we began a new study at Indiana University which seeks to address these crucial questions regarding early symptom progression in HD. This new study is performing many of the same tests that were used a decade ago which will allow us to examine the change in an individual's performance on the test over time. There are also some new tests and questionnaires which focus particularly on a more careful examination of saccadic eye movements as well as more detailed information about possible personality and mood changes early in the disease.

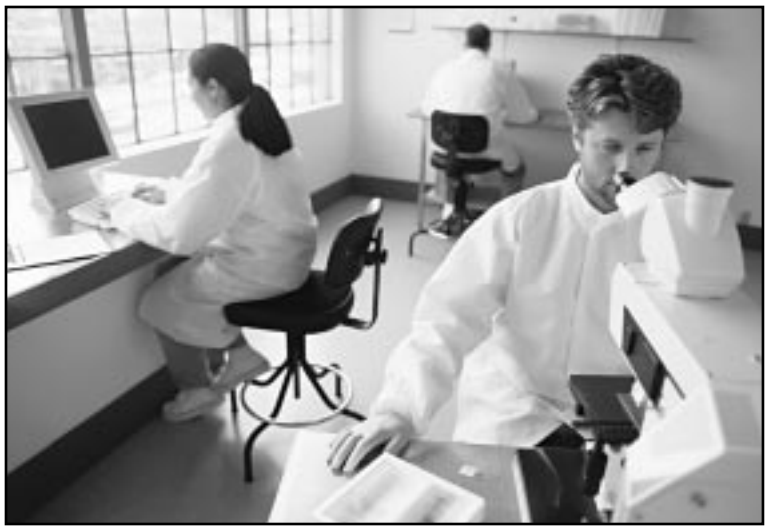
This current research entitled, Longitudinal Studies Among At-Risk HD Gene Carriers, is recruiting individuals to come to Indiana University to complete a single study visit. Qualifying individuals must:

- Be under the age of 65 years, since older individuals may have changes on some of these tests due to aging.
- Either not been diagnosed with HD, or have been clinically diagnosed with HD only within the past two years.

If you are interested in learning about this study, please feel free to contact the study coordinators: Kerry White (317) 278-8411 or jldatzma@iupui.edu.

SCIENTISTS USING THE ROSTER: PAST STUDIES - WHAT DID THEY DISCOVER ABOUT HD?

Basal Ganglia Neurobehavioral Evaluation



BY: JULIE STOUT, PH. D.

Two studies have been completed by Dr. Stout that focused on thinking and memory abilities of people with Huntington disease. This research aims to improve measurement methods so that behavioral changes in Huntington disease can be detected with high sensitivity. This work is important because it helps identify the brain mechanisms that account for the difficulties in behavior that people with HD experience in daily life. Sensitive measurement techniques are also essential for successful clinical trials that could help to treat or prevent HD.

In the first study, a number of key findings were made about HD:

- People with HD can have difficulties suppressing distracting information, and this can interfere with their later processing of new information
- People with HD may have difficulties with decision making related to problems remembering the outcomes of their past choices.
- People with HD initiate conversations less frequently and tend to use fewer words to describe their thoughts than healthy controls.
- People with HD have elevated levels of apathy.

In the second study, which was funded by the National Institutes of Health, difficulties with decision making were explored further, and the following findings were made:

- People with HD tend to react less strongly to negative consequences on a decision making task than do healthy controls.
- This reduced reaction to loss may lead to more short-sighted choices or a difficulty considering possible negative outcomes in the choice-making process.

Dr. Julie Stout has been involved with research on Huntington disease (HD) since her post-doctoral fellowship in San Diego from 1992-1995. She is a clinical neuropsychologist with training in neuroscience and cognitive science. She is also an Associate Professor at Indiana University in Bloomington where she offers courses and runs a laboratory which provides research training to undergraduates, graduate students, and post-doctoral fellows.

Dr. Stout currently has a lead role in the ongoing international study called PREDICT-HD. Dr. Stout's laboratory is in charge of the development and implementation of the specialized computer testing of cognition for PREDICT-HD which is being carried out at 24 sites in the United States, Canada, and Australia. She is also a collaborator with Dr. Tatiana Foroud and others on the Longitudinal Studies Among At-Risk HD Gene Carriers study at Indiana University, Indianapolis. Dr. Stout maintains involvement in the Huntington Study Group, an organization of scientists and clinicians who coordinate their efforts to work toward understanding, preventing, and curing Huntington disease.

For more information about Dr. Julie Stout, you can view her website at:

http://www.indiana.edu/~psych/faculty/stout.html

My Experiences with Huntington's Disease: A Personal Story

BY: JOHN CHARLTON

(feature article published 8/6/2004 in the Huntington's Disease Advocacy Center website)

Growing up with a father and an aunt with Huntington's disease, I have always known that there was a chance that I too would get this illness. Knowing this, I had always tried to prepare myself just in case I ever had to hear those words, "You have Huntington's, I'm sorry."

Shortly before my sister passed away, also a victim of HD, I began to notice the tell-tale signs of HD, like how I kept dropping things more frequently. I also began moving without wanting to, and I had become very moody as well.

A few months after my sister passed away, I decided it was time to get tested not only to put my mind at ease but also my wife and mother. I had a theory that the symptoms were only in my mind, that I had convinced myself that I had it or was showing movements because I had learned the behavior of moving after watching my father and sister with the sickness, not because I actually had it.

The test usually is a rather long process involving the following: 1 - a visit with the geneticist; 2 - a visit with the neurologist; 3 - the visit with the psychologist; 4 - the actual blood test; 5 - the follow-up with the neurologist as well as 2 more follow ups with the psychologist if the results were positive. Fortunately, because of my past history with my neurologist (was diagnosed with Meniere's disease which left me deaf in my left ear) as well as the fact that he also saw my sister as well, he decided to just give me the test. I had explained to him about my thoughts regarding my movements being all in my mind, and he told me that there was a woman at risk who had also began to show movements as well and actually tested negative.

The big day finally came to go get tested in June of 97. They took a sample of my blood, and two weeks later I went in for my follow up appointment, and unfortunately, the results were not good news. I was only 26 years old and HD+.

Shortly after, I left my job, and applied for disability, which was hard for me because I had been working since I was 15 and suddenly I had to depend on others which really does wonders to one's self esteem. I was still able to do the basics, like cook for my then wife, as well as clean the house, which helped a little bit to regain it back. I was embarrassed to go out in public for fear that people would assume the worse (he is drunk! He is on drugs! etc) This really was a change of pace for me considering my entire life I never cared what others thought of me (I'm best known as being a free spirit!) It was also getting frustrating how almost anytime that I would go for a drive I was always being pulled over.

I have had this disease for 7 years now and during this time have really grown to accept my limitations. After my divorce, my parents were kind enough to take me in and keep an eye on me. I no longer am afraid to go out anymore, in fact I even enjoyed going out and go dancing! I also no longer drive; I must rely on friends and family to take me out to activities. I have also gotten used to the fact that every night before I go to bed I will have a new set of bumps, bruises and cuts from running into various

John (left) and his Kumdo instructor

Continued on the back cover.

We invite you to share your story on living with HD! Selected stories may be reprinted in future newsletter issues. Please send stories to:

The National Research Roster for Huntington Disease Patients and Families

Attention: Patti Wolf

Department of Medical and Molecular Genetics,
975 West Walnut Street,
Indianapolis, IN 46202

Or email: pswolf@iupui.edu

Statistics – What do they mean?

Statistics are a toolkit for problem solving. Data is collected from a population and examined for certain characteristics or traits. Scientists use statistics to describe, understand, and draw conclusions about data being researched. The following article was written by Tracy Costello, Ph.D. from MD Anderson Cancer Center in Houston, Texas. The Roster was able to provide Dr. Costello with family history data collected from 2754 families. She explains how data from the Roster helped her develop a research method to help detect age of onset within a family unit over several generations.

Ask the Researcher: How do Statistics Help HD Research?

BY: TRACY J. COSTELLO, PH.D.

1) Why did you choose to use Roster information in your research?

My dissertation research involved developing a statistical method to identify a phenomenon called "genetic anticipation." Genetic anticipation is defined as a decrease in age of onset of a disorder as it is passes from one generation to the next. This has been shown to be the case in Huntington disease, with the identification of the gene on chromosome 4. Once I had heard about the rich resource that was available through the Roster, I requested de-identified data from the Roster so that I would be able to show that my method could identify the age of onset pattern in real data.

2) What kinds of things were you looking for from the information you received from the Roster?

The type of information that I needed for my research was a data set that included family-based data. Since my statistical method was based on identifying a relationship between age of onset and generation, I also needed a data set in which this phenomenon was apparent. The HD roster fit these two criteria.

3) How has the information provided by the Roster helped your research?

The HD Roster contains information from a large number of patients and their families. These individuals have been recruited from many sites within the United States. The ability to use such a comprehensive set of data in genetics research is rare and valuable.

Dr. Tracy J. Costello

received her Bachelor of Science degree, magna cum laude, with the majors of mathematics and physics in May 1994 from The University of Southern Mississippi in Hattiesburg, Mississippi. In the fall of 1994, she entered The University of Texas Health Science Center at Houston, Graduate School of Biomedical Sciences. She was awarded the Master's Candidate Award from the Association for Women in Science, Gulf Coast - Houston Chapter in 1997 and earned her Masters of Science degree in Biomathematics in August 1998. She was awarded an NCI R25 Predoctoral Fellowship and an American Legion Auxiliary Cancer Research Fellowship in 1999. She completed her Doctor of Philosophy degree in Biomathematics and Genetics in May 2004. She is currently an NCI R25 Postdoctoral Fellow in the Department of Epidemiology at The University of Texas M.D. Anderson Cancer Center. Her current research interests include the development of statistical methods and their application to the analysis of complex disease and cancer.

Continued from page 6.

My Experiences with Huntington's Disease: A Personal Story

Whenever I begin to explain to some one the basics about HD, probably the most frequently asked question would have to be "Does it hurt or can you feel it when you move?" "Well, only if I hit something hard," I always reply! Actually, it's amazing how much pain you can endure after a while of constantly moving into things. Another limitation is that my speech seems to be suffering quite a bit, which is a shame considering how much I loved to talk.

I even started Kumdo (Korean swordsmanship) for the past 5 years using Japanese swords and wooden swords for teaching my class. I recently got my second degree black belt in Kumdo as well and now I'm teaching for my instructor. My instructor knew about my sickness from the get-go and did not charge me a dime during the five years at his school. As I practice my various forms with my swords in class, or even sparring with one of my students with a bamboo sword, it always amazes me how I almost leave my body and am somewhere else. For that brief time, all of a sudden my coordination returns, and my movements disappear. I always look forward to my next class because it gets me out of the house and also for just for a while I get to feel normal again.

It seems strange, because in some ways I'm worse than before, like my movements as well as having to be completely dependent for alot of things, but I'm also better in my overall attitude towards my life. I am very fortunate to live with my parents who are very, very understanding. I am also very lucky to have great friends that care as much as they do. Every now and then, I will miss my old life, like being able to live on my own as well as my old job as a social worker. But then I remember that I did not choose my illness, I was born with it. God has decided to put my faith to the test, and it is a test that I hope I pass. There are alot more painful sicknesses in the world. I am grateful for the time I have on the earth before passing on to see my family and friends in heaven.

Useful Sources for Information and Support

Stanford HOPES - Huntington's Outreach Project for Education, at Stanford University
Website developed and maintained by a team of students and faculty at Stanford University. Provides information on the scientific aspects of Huntington's disease.
http://www.stanford.edu/group/hopes

HDAC - Huntington's Disease Advocacy Center
Website developed and maintained by families living with HD. Provides helpful information for caregivers, families, and patients with HD.
http://www.hdac.org/

HDSA - Huntington Disease Society of America
Tel: (800) 345-HDSA
http://www.hdsa.org/

NINDS - National Institute of Neurological Disorders and Stroke
Tel: (800) 352-9424
http://www.ninds.nih.gov/health_and_medical/disorders/huntington.htm

HDF - Hereditary Disease Foundation
Tel: (212) 928-2121
http://www.hdfoundation.org

We Move - Worldwide Education and Awareness for Movement Disorders
Website resource for movement disorder information. Provides comprehensive educational materials for patients and health care professionals
http://www.wemove.org

YGYH - Your genes your health
Multimedia website guide for genetic disorders.
http://www.ygyh.org

Don't Forget!

Visit the Roster's NEW website at
http://hdroster.iu.edu
Browse through a wealth of information for individuals and families interested in HD research.