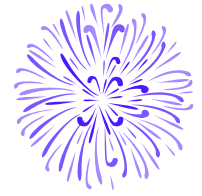


HIND-SIGHT



Huntington's Disease Information and New Discoveries

University of Iowa Hospitals & Clinics: Huntington's Disease Center of Excellence

A New Year brings a new update from Dr. Paulsen's lab at University of Iowa's Huntington's Disease Center of Excellence. The New Year has brought the start of a lot of exciting projects and developments. The HD Center is underway and we begin our clinic and research appointments in a new hospital location in an effort to centralize HD related services. Our new clinic location is in 3 South General Hospital. Visitors will take Elevator C up to the 3rd floor and turn right to meet us at the nurses station on the right hand side of the hallway. More detailed directions can also be obtained by calling the Center coordinator (Elizabeth) at 319- 353-4307. On clinic days (2nd Tuesday of every month), patients can obtain all of the Center's services at one place. This means no more hikes across the hospital to see a doctor in a different department. Now, if you need to see a neurologist and a psychiatrist, and maybe even visit with a nutritionist or social worker while you are here, appointments can be scheduled on the same day in the same location. Research appointments can also take place at the Center's clinic.

Please continue to explore our newsletter for stories about Center fundraisers, advancements in telemedicine and updates on HD conferences during this past year. We wish all of you happy reading and a wonderful New Year!



Laura Stierman and Elizabeth Penziner at the HD Holiday Benefit Sale booth.

Inside this issue:

- Conference Updates
- Alternative Therapies
- Nutritional Recommendations
- HD Center Events
- Telemedicine



HD CONFERENCE

UPDATES

Iowa State HD Conference

August, 2000 (reported by E. Penziner)

I am very excited to share news about the Iowa Huntington's disease conference that took place on August 12, 2000. Not only was this Iowa's first state conference on HD, but it also initiated a great partnership between the University of Iowa's HD Center and the Iowa HD Chapter. With the support of the Huntington's Disease Society of America, this partnership shall continue to develop and strengthen. I want to take this opportunity to share why I believe this is an important goal for the advancement of the HD community. I have a deep belief that only when the entirety of our community unites to compile and share our resources, will we be better able to serve all of the HD families. In fact, this was a message that seemed to resonate as a theme at this year's conference. We must all join forces and work together to see the greatest results. The conference began with introductions and a welcome from the Iowa Chapter's Mike and Diana Fisher. The morning keynote address was presented by Dr. Jane Paulsen and provided an educational overview of the disease and the current research efforts at finding a treatment and cure. We broke for lunch and enjoyed the inspirational words and lyrics of Carmen Leal Pollock. Although tears were abundant, this sorrow was tempered by a sense of community and compassion that unites us in this cause.

The remainder of the afternoon incorporated a forum discussion that allowed conference attendees to address a panel of HD 'experts'. The greatest experts were HD individuals and their families. We were fortunate to have had an at risk and an affected individual share with us the deeper meanings of their experiences. We also

gained valuable insight from the Huntington's Disease Center professionals who were able to attend including the Center's Psychiatrist, Physical Therapist and Genetic Counselor. It was truly remarkable to see the Center professionals, the Chapter, the families, and the community working together to discuss the work that needs to be done and what each of us can do to make a difference. I encourage all that were unable to attend to keep your calendars open for future gatherings and to consider ways that your involvement can have a great impact. Throughout this newsletter, you can find opportunities to get involved and to join forces with the families and professionals working to improve the lives of the HD community.

Huntington Study Group

October, 2000 (reported by L. Stierman)

The Huntington's Study Group's (HSG) 8th annual meeting took place in San Diego, California on October 26-28, 2000. This was my first conference and I am happy to say it was very encouraging. Professionals from all over the US, Canada, Europe, and Australia came together to discuss HD research in progress and possibilities for research in the future. I will begin with an update on research projects underway. The RID-HD study, looking at the short-term effects of riluzole on HD symptoms and the safety and tolerability of the drug, has been completed. The goal is to have results available by August. Another trial, testing the effectiveness of the nutritional supplement Co-enzyme Q₁₀ and the drug Remacemide in slowing HD's progression (CARE-HD), is also nearing completion. Results should be analyzed by June of this year. The PHAROS study is looking at initial signs of the disease in the at-risk population and possible factors that could influence the age of onset. Currently, over 400 of

the 1000 total participants have been enrolled. If you are interested in participating, between the ages 30-55, are at risk for HD, and have not been genetically tested, please contact the HSG coordination center at (800) 487-7671 for more information.

A future project includes a study of Minocycline, which has been found to delay the progression of HD in mice. A study to test the safety and tolerability of the drug in the HD population is being reviewed and may begin as early as

April of 2001. Finally, the PREDICT-HD Study is also under review. If approved, we will be recruiting individuals ages 30-55 that have tested positive for the HD gene expansion, but are not exhibiting motor symptoms. As always, we will continue to do evaluations of everyone at risk for or having HD. If interested, please contact us. We are anticipating a great year in HD research, bringing us closer to ending this devastating disease.

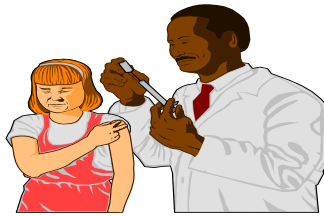
ALTERNATIVE THERAPIES AND TREATMENTS FOR HUNTINGTON'S DISEASE



Recent attention has been given to several new therapies and treatments for longitudinal Huntington's disease. We are continually exploring the latest news to discover the myths and facts at the heart of these discoveries.

It is our feeling that health consumers need to be especially cautious about purported new therapies. We make these recommendations, not because we want to keep anyone from benefiting from anything that might help, but rather because we fear the potential harm that mis-use of information can cause. Here's an example: Let's think about vitamins and the benefit that comes from meeting all of our nutritional needs. We've all heard that Vitamin A is an essential part of our diet. This anti-oxidant is especially important in the prevention of eye and skin problems and is needed for the healthy formation of bones and teeth. It also has shown some benefit in protecting our cells from oxidation (breakdown) and can even promote cell growth. However, you might not have heard that taking large amounts of vitamin A over long periods of time can be toxic to the body, mainly the liver. Toxic levels of vitamin A are associated with abdominal pain, loss of menstrual periods, enlargement of the liver and/or spleen, stomach disturbances, hair loss, itching, joint pain, nausea, vomiting and water on the brain. These not so pleasant effects of taking too much of a needed nutrient, exist with almost all of the fat-soluble vitamins (A, D, E, and K). The most common problem with a lack of iron is a condition called anemia, but too much iron has been shown to increase a person's risk of heart attack. This is why the FDA has upper-level recommendations: to remind us that more is not always better! In the same token, combinations of certain vitamins can interfere with the absorption of others. For example, large amounts of calcium and zinc can interfere with iron absorption, however taking enough vitamin C can enhance our absorption of iron. All of this is confusing, so if you can take one main message from these facts, it is to be cautious with what we put in our bodies. The best way to remain safe is to make sure your physician knows everything you are taking and why.

RESEARCH VERSUS RUMORS



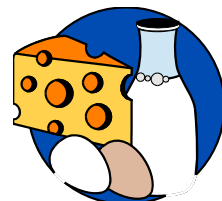
To better understand the subtleties of experimental design, keep in mind that the experiences of one individual are a far cry from scientific validity. For example, your neighbor Joan (a sample size of one and no control group) says she takes vitamin C when she feels a cold coming on and “it works every time.” She thinks she knows what she’s taking, she has faith in its efficacy, but she tends not to notice when it doesn’t work. Before concluding that an experiment has shown that a nutrient cures a disease or alleviates a symptom, ask these questions:

- Was there similarity between a comparison group and the experimental (or treatment) group?
- Was the sample size large enough to rule out chance variation?
- Was a placebo effectively administered? (Blind study)
- Was the experiment double-blind (even the investigator doesn’t know which group had the treatment)

Well-designed research has enabled scientists to study the actions of nutrients in the body. Such research has laid the foundation for quantifying how much of each nutrient is needed by the body. In summary, scientists learn about nutrition by conducting experiments following the protocol of scientific research. Researchers make sure to establish similar control and experimental groups, use large sample sizes, placebo, and blind treatments. Their findings must be reviewed and replicated by other scientists before being accepted as valid. [Understanding Nutrition: by Whitney, Rolfes. 1999.]

NUTRITIONAL RECOMMENDATIONS FOR PERSONS WITH HUNTINGTON’S DISEASE

Weight loss is a common problem in Huntington’s disease. The reasons for this are not fully understood, and weight loss may occur from a variety of reasons such as swallowing problems, fatigue, and/or depression. Two strategies can be employed to increase the caloric intake of someone with HD: increase the number of meals, or increase the calorie content of the food. Consultation with a nutritionist can help in selecting the most appropriate foods and supplements to meet the patient’s needs. Physicians have reported that regaining lost weight sometimes results in improved alertness and responsiveness, and often appears to reduce chorea as well. Maintaining hydration is crucial, so it is very important to consume a lot of fluids throughout the day.



The following has been adapted from:
Understanding Nutrition, by Eleanor Noss Whitney and Sharon Rady Rolfes. Wadsworth Publishing Company, 1999.

Weight -Gain Strategies

People attempting to gain weight sometimes have a hard time because they choose low-calorie, high bulk foods that make it hard to consume enough energy. Weight gain strategies center on eating foods that provide many calories in a small volume as well as exercising to build muscle. Conventional advice is to provide about 700 to 1000 calories a day, above what normal energy needs and to exercise to build lean tissue. Table 1 provides diet patterns for energy intakes at 3000 calories a day (recommended for individuals with HD). These patterns follow the Daily Food Guide plan and incorporate many of the principles for planning a healthy diet.

- **Energy-Dense Foods**
 Pick the highest calorie items from each group—that is, milk shakes instead of nonfat milk, avocados instead of cucumbers, a cup of grape juice instead of a small apple, and whole wheat muffins instead of whole-wheat bread.
- **Regular Meals Daily**
 Make meals a priority and take time to plan, prepare, and eat each meal.
- **Large Portions**
 Learn to eat more food at each meal. Put extra slices of ham and cheese on the sandwich for lunch, drink milk from a larger glass, and eat cereal from a larger bowl.
- **Extra Snacks**
 Eat more frequently. Between-meal snacking offers a solution. Snacking on dried fruit, nuts, and seeds will also add calories easily.
- **Juice and Milk**
 Beverages provide an easy way to increase energy intake. Consider that 6 cups of cranberry juice adds almost 1000calories to the day's intake. Calories can be added to milk by mixing in powdered milk or packets of instant breakfast.
- **Exercising to Build Muscles**
 Use strength training primarily and increase energy intake to support that exercise. About 700 to 1000 calories a day above normal energy needs is enough to support both the exercise and the building of muscle.

TABLE 1

| FOOD GROUP | RECOMMENDED NUMBER OF SERVINGS IN 3000 KCALORIE DIET |
|--|---|
| Bread, Cereal, Rice and Pasta | 15 |
| Meat (lean) and meat alternates | 8 |
| Vegetable | 6 |
| Fruit | 6 |
| Milk and Milk Products | 3 |
| Fat (tsp.) | 12 |

UPCOMING PLANS AND PROJECTS IN THE HD

CENTER:

HDSA's recent designation of the University of Iowa as an HD Center of Excellence brings with it certain responsibilities and expectations. It is the mission of our center to accomplish these three specific goals:

- To provide comprehensive services for patients and their families.
- To educate patients and families on all aspects of Huntington's Disease.
- To provide opportunities for patients to enroll in research trials for treatments that may slow the progression of the disease while working toward a cure through clinical and basic research.

Obviously, there is much to do and we want to be honest about our need for your help. Part of the ongoing effort to unite the community and get everyone involved includes figuring out what each of us can do and how we can contribute. For example, Dr. Paulsen's research lab recently raised funds to support HD through a Holiday Benefit Sale sponsored by Younkers. Michelle, Laura, and Elizabeth even woke up one Saturday at 5 AM (an event in itself) to sell tickets for the big day! Fundraising is going to be a big part of sustaining an Iowa HD Center. We fully understand that not everyone is able to make a financial contribution, however, everyone is able to be involved in some way, whether through research participation, educating others, or even organizing a bake sale.

HD COMMUNITY WISH LIST

The HD Community needs every one of us, so if you are able to help in ANY way, please contact Elizabeth at the Center to get involved.

The following are a few requests we've had for patients and families seeking support:

- Long distance calling cards (for individuals living in a facility far from their family and friends).
- Sponsor a child to attend summer camp for one week (providing valuable family respite).
- Organizing a local fundraiser in your own community.
- Participate in research to make this the last generation of HD.
- Help out with transportation for individuals no longer able to drive.
- Educate your local schools and physicians about HD.
- Be kind; take some time to discover how you might bring joy to a person with HD.

If you or anyone you know have 'wishes' for the community and would like your requests to be included in our next newsletter, please let us know.

ADVANCES IN TECHNOLOGY ASSISTING HD PATIENTS AND FAMILIES



“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 remains a major leader in worldwide 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 they, in turn, can watch the University of Iowa health care professionals on their own monitors.

Telemedicine has already allowed the HDSA Center of Excellence team at the University of Iowa to reach out to staff in a remote northwest Iowa care facility. Continued use of these new methods of delivery will continue to bring information to folks in rural locations and others who may not have access to HD Center services. 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 statewide enhances the overall quality of care for patients with HD.



HD RESOURCES AND CONTACT INFORMATION



To schedule an HD Center appointment: Contact Elizabeth Penziner at (319) 353-4307
For information about genetic testing: Contact Cathy Evers at (800) 260-2065
To learn about research opportunities: Contact Laura Stierman at (319) 353-3716
To receive 24-hour emergency support: Call the helpline at (800) 777-8442 ext. 34307
To reach us by email: hdinfo@uiowa.edu
To connect to our web site: www.uihealthcare.com/huntingtonsdisease/
To reach the national office: Contact HDSA at (800) 345-HDSA
To contact the Iowa Chapter: Call the chapter office at (800) 535-4797

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