Ethics and Research on Huntington's Disease

Where Do Legal and Ethical Guidelines Come From?
All participants in studies connected with major medical centers are protected by federal laws that regulate the way research is conducted. The most important of these laws is entitled “Protection of Human Research Subjects” and is outlined in the Federal Register. These regulations require investigators to obtain the informed consent of the participants in the study, undergo review by an internal board known as an Institutional Review Board (an “IRB”), and comply with all applicable IRB policies and requirements. The most important consideration of investigators is directed to the perspective of the individuals most affected by the research - the subjects who volunteer to be participants in the research project. This article will briefly summarize the requirements of the law and the ethics that stand behind and sometimes beyond the law and form the trust that lies within the research-participant relationship.

The federal regulations grew out of a study commissioned in 1974 by the National Research Act which directed an interdisciplinary group of individuals to identify the basic ethical principles that should underlie the conduct of biomedical and behavioral research involving human subjects. The results of this study were published in The Belmont Report which outlined the principles of respect for persons, beneficence, and justice. As with most complex human endeavors, applying these principles requires thoughtful reflection and careful consideration. These three principles were developed at a time well before the gene for Huntington's Disease (HD) was identified, but they have been remarkably good ethical guides to the issues that have arisen in the era of genetic diagnosis and prediction.

How Are Huntington's Disease Participants Affected by Ethical Considerations?
The law and ethical guidelines require the informed consent of all participants. The idea that individuals have the right to consent, or not to consent, to participate in research acknowledges the respect we accord everyone to determine what happens to his or her own body. The law has long acknowledged the right to control whether a doctor can perform a medical procedure. Ethical principles demand the same respect be given participants in research, whether a medical procedure is performed or not. In the realm of genetics research we extend the idea of respect for persons to include the right to control not only what happens to one’s body, but also what can be done to blood or other tissues that are removed from the body. Thus, genetic testing on body fluids should not be conducted without the informed consent of the individual. In addition to physical dangers, such as bruising from a blood draw or other physical danger, all medical information that has a genetic component is treated as potentially damaging if certain individuals come into its possession. Medical information, including family history of disease or illness, is some of the most private and sensitive information anyone possesses. Researchers go to great lengths to make sure that participants consent to the disclosure of genetic and family information for use in research. In some cases identifying information can be removed from sensitive information prior to use in research, but in many instances links to other information prove invaluable to resolving difficult research questions. In either case respect for persons requires the participant be aware of the uses that may be made of their medical information and the efforts that are taken to safeguard this information from unauthorized disclosure. Neither the Belmont Report, written in 1979, nor the regulations enacted in 1991 specifically mention the protection of such sensitive information about human subjects, but common practices have developed in the past years that extend both the law and ethical principles to include such protections.

This is Part I of a three part series on ethics and HD research.

Written by Cheryl Erwin, JD, PhD, Assistant Professor, The University of Iowa College of Medicine

Part II- Respect, Beneficence and Justice
Part III- Concluding thoughts
Support Group Schedule

Iowa City
University of Iowa Hospitals and Clinics
Adult Psychiatric Outpatient Waiting Room
1st floor John Pappajohn, near elevator I

<table>
<thead>
<tr>
<th>Date</th>
<th>Topic</th>
</tr>
</thead>
<tbody>
<tr>
<td>January 25</td>
<td>Dr. Peg Nopoulos, What the Shrink thinks...about HD</td>
</tr>
<tr>
<td>February 22</td>
<td>Dr. John Harvey, Issues of Loss and Grief</td>
</tr>
<tr>
<td>March 28</td>
<td>Dr. Jane Paulsen, Understanding HD Behavior</td>
</tr>
</tbody>
</table>

What is a Neuropsychologist and What is Their Role in Understanding HD?

We know that individuals with HD can have difficulties in three different areas: cognitive (or thinking) changes, emotional or behavioral changes, and movement disturbance. Neuropsychologists are knowledgeable about brain functioning and how difficulties in these three areas affect people’s behaviors. Scientists know that these problems originate in specific areas of the brain.

Neuropsychologists are able to assess cognitive abilities by observation and testing. An individual with HD may have difficulty regulating emotions, organizing or planning day-to-day events, and being able to filter between important and random information. A neuropsychological evaluation is a comprehensive assessment of these cognitive and behavioral functions using a set of tests and procedures.

For example, neuropsychologists may assist in disability assessments for individuals who become too ill with HD to continue working. The assessment may involve tasks such as remembering a set of words to be repeated several minutes later or imitating a simple drawing. The neuropsychologist will use these results to write a report addressing areas of work that may have once been considered routine but now are increasingly more difficult.

The Huntington’s Center of Excellence recently hired two new neuropsychologists, Dr. Leigh Beglinger and Dr. Kevin Duff, to help us better address and treat cognitive changes in HD.

Leigh Beglinger completed her doctorate at Washington State University and her postdoctoral fellowship at Indiana University’s Department of Neurology. Dr. Beglinger is a member of the Huntington Study Group (HSG) and her research plan includes using brain imaging to explore cognitive changes in Huntington’s disease. Her other research interests include learning how best to measure cognitive changes in people taking medications for research and assessing how much individuals improve on cognitive tests with practice.

Kevin Duff completed his doctoral training at State University of New York at Albany, and he completed his postdoctoral fellowship at the University of Oklahoma Health Sciences Center. Dr. Duff is a member of the HSG and will focus his research on cognitive and psychiatric changes present throughout the course of Huntington’s disease. Outside of work, Kevin enjoys spending time with his family and is quickly becoming a big Hawkeye fan.

Welcome, Dr. Leigh Beglinger and Dr. Kevin Duff! We look forward to a long and productive relationship with both of you.
How Can I Help?

Brain Bank Donations - A Valuable Gift

Brain donations can be arranged through brain banks at various medical centers throughout the country. Brain banks accept brains from disease-free individuals as well as from people who have died in the early, middle or late stages of a particular disease.

The brain tissue is accepted and stored by various brain banks free of charge to the donor’s family. However, there is a charge to remove the brain and send it to the nearest designated bank. Some brain banks will cover these fees.

Anyone interested should contact the brain bank that they want to use for specific information about necessary arrangements and costs.

The following link contains common questions about brain donation as well as a list of steps to take when death occurs: http://www.lib.uchicago.edu/~rd13/hd/brainban.html, or specific questions to www.hdny.org click contact us. Furthermore, a recent edition of the Huntington’s Disease Society of America Marker magazine included a list of brain banks that accept all stages of HD-affected brains as well as disease-free brains.

If possible, families should openly discuss the idea of donation in order to avoid misunderstandings and to facilitate the donation process. At the time of death of the donor, the surviving family members will need to verify the donor’s intent-to-donate, and to offer authorization to the brain bank to acquire all medical records. At the time of death, an individual’s body becomes the property of the spouse, or if there is no spouse, then the adult children or parent. Although an individual can make a personal request to donate his/her brain, ultimately it is the surviving family members who have the privilege and responsibility of deciding whether this unique and valuable gift will be made.

Thank you to Jennifer Moran and AVON for a successful fundraiser to benefit the University of Iowa HD Center of Excellence.

What does it mean when my doctor uses the word _____?

definitions continued C-G

Chronic: A long duration or frequent recurrence; always present. A chronic condition is one that lasts longer than three months or more.

Degenerative: Deterioration of a tissue or an organ.

Delusional: Something that is falsely believed; a persistent false belief. The belief that one’s house is contaminated is an example of delusional thinking when others in the household do not hold the same belief.

Disorientation: To lose sense of time, place or identity. Your doctor may check for disorientation by asking your name, the date, what time it is and where you are.

Dysarthria: Disordered or impaired articulation or expression of speech due to disturbances of muscular control often associated with HD. This may make speech more difficult to understand in patients with HD.

Extremity: Limb of the body; hand or foot. The doctor may state that he notices movements especially in lower extremities (legs).

Gait: Manner or style of walking. Many HD patients develop a distinctive manner of walking (gait) that may be unsteady, disjointed or lurching.

Dateline 1/2/04

NBC’s Dateline aired a 60 minute story about Huntington’s disease. Previously slotted for August, 2003, the program focused on the plight of Carol Carr, a Georgia woman, accused of murdering her 2 sons who were in a nursing home with late stage HD. Carol was convicted of assisting her sons’ suicides and is currently serving a 5 year sentence. The Huntington’s disease community has expressed mixed feelings about this show. It is important to increase awareness of HD in the public sector. However, the case of Carol Carr is not a typical HD case and many people wish for more opportunities to educate the public about HD and show other families’ struggles of caring for their loved ones with HD.
10 Tips for Family Caregivers

- Choose to take charge of your life, and don’t let your loved one’s illness or disability always take center stage.

- Remember to be good to yourself. Love, honor and value yourself. You’re doing a very hard job and you deserve some quality time, just for you.

- Watch out for signs of depression, and don’t delay in getting professional help when you need it.

- When people offer to help, accept the offer and suggest specific things that they can do.

- Educate yourself about your loved one’s condition. Information is empowering.

- There’s a difference between caring and doing. Be open to technologies and ideas that promote your loved one’s independence.

- Trust your instincts. Most of the time they’ll lead you in the right direction.

- Grieve for your losses, and then allow yourself to dream new dreams.

- Stand up for your rights as a caregiver and a citizen.

- Seek support from other caregivers. There is great strength in knowing you are not alone.

printed by permission of NFCA (National Family Caregivers Association)